A STUDY OF AUTISM SPECTRUM DISORDER AND SENSORY IMPAIRMENT IN ADULTS WITH INTELLECTUAL DISABILITY

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ABSTRACT

Autism spectrum disorder (ASD) and sensory impairment are common in people with intellectual disability (ID), but little is known about the relationship between them.

The primary aim of this thesis was to explore the relationship between deafness, blindness and ASD. The secondary aim was to determine the prevalence of sensory impairment, ASD and other co-morbidities in adults with ID.

This thesis comprised a comprehensive literature review followed by a 2-stage study. Stage 1 involved cross-sectional analysis of data on adults with ID on a population-based case register. Stage 2 involved investigating adults with congenital deafness and their controls (deaf subgroup), and congenital blindness and their controls (blind subgroup) using medical case file review and face-to-face interviews, including the Pervasive Developmental Disorder in Mental Retardation Scale to identify ASD. Data were analysed using chi-squared tests, estimated probabilities (to explore interactions) and general linear, conditional and non-conditional logistic regression modelling.

Stage 1 identified 3183 adults with ID, 634 (20%) of whom had sensory impairment (congenital and acquired), comprising partial (n=447), total (n=165), or dual/deaf-blindness (n=22). Both visual and hearing impairment were associated with degree of ID, age and having Down syndrome but only visual impairment was associated with epilepsy. Neither visual impairment nor hearing impairment was associated with ASD at this stage of the study.

In stage 2, those with an acquired sensory impairment were excluded and only 60 congenitally blind cases, 21 congenitally deaf cases and their controls (matched on degree of ID and gender) were included. Congenital blindness, but not deafness, was associated with ASD (OR=3.03; 95% CI: 1.34–6.89; $p<0.008$) after adjustment for potential confounders.

This thesis supports previous findings of high prevalence of sensory impairment among adults with ID. For the first time, an independent relationship was observed between congenital blindness and ASD in a cohort of adults with ID. The implications of these findings are discussed.

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I started my PhD project when I was a senior registrar. What seemed to be an idealistic project at the start, by an enthusiastic trainee, soon became a mammoth task to incorporate into an already busy clinical schedule. To make matters more complicated, after becoming a full time NHS consultant clinician, the task of meeting the deadlines and completing the assessments of patients at the slow rate of 1 or 2 cases per week to meet the requirement of the power calculation, turned into a daunting prospect which I thought I might never be able to overcome in spite of all the enthusiasm that I had initially felt about a subject that was very close to my heart.

The project was not easy; I had to sacrifice a lot of personal time, evenings, weekends and annual leave over the years. However, I have met so many inspirational people who enthused me with hope and optimism along the way to be able to carry on despite all the challenges.

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ABBREVIATIONS/GLOSSARY OF TERMS

ABA	APPLIED BEHAVIOURAL ANALYSIS
ABC	ABERRANT BEHAVIOUR CHECKLIST
ADHD	ATTENTION DEFICIT HYPERACTIVITY DISORDER
ADI	AUTISM DIAGNOSTIC INTERVIEW
ADOS	AUTISM DIAGNOSTIC OBSERVATION SCHEDULE
ASD	AUTISTIC SPECTRUM DISORDER
BSL	BRITISH SIGN LANGUAGE
CAH	CONGENITAL ADRENAL HYPERPLASIA
CACDP	COUNCIL FOR THE ADVANCEMENT OF COMMUNICATION WITH DEAF PEOPLE
CB	CHALLENGING BEHAVIOUR
CHARGE	CLOBOMA, HEART DEFECTS, ATRESIA CHOANAE, RETARDATION OF GROWTH, GENITAL AND EAR ABNORMALITIES
CHL	CONDUCTIVE HEARING LOSS
CI	CONFIDENCE INTERVAL
CP	CEREBRAL PALSY
CRS	CONGENITAL RUBELLA SYNDROME
CT	COMPUTED TOMOGRAPHY
CVI	CERTIFICATE OF VISUAL IMPAIRMENT
DAS	DISABILITY ASSESSMENT SCHEDULE
DB	DECIBEL
DH	DEPARTMENT OF HEALTH
DISCO	DIAGNOSTIC INTERVIEW IN SOCIAL AND COMMUNICATION DISORDERS
DSM-IV	DIAGNOSTIC AND STATISTICAL MANUAL OF MENTAL DISORDERS-4TH EDITION
ECG	ELECTRO CARDIOGRAPHY
EEG	ELECTRO ENCEPHALOGRAPHY
GER	GASTRO-OESOPHAGEAL REFLUX
GP	GENERAL PRACTITIONER
HI	HEARING IMPAIRMENT
ICD-10	INTERNATIONAL CLASSIFICATION OF DISEASES-10TH REVISION
ID	INTELLECTUAL DISABILITY
IHD	ISCHAEMIC HEART DISEASE
IQ	INTELLIGENCE QUOTIENT
LD	LEARNING DISABILITY
LID	LEICESTERSHIRE INTELLECTUAL DISABILITY
LLDR	LEICESTERSHIRE LEARNING DISABILITY REGISTER

MDT	MULTI-DISCIPLINARY TEAM
MELAS	MITOCHONDRIAL ENCEPHALO-MYOPATHY, LACTIC ACIDOSIS, AND STROKE-LIKE EPISODES
MENCAP	A CHARITY ORGANISATION FOR PEOPLE WITH INTELLECTUAL DISABILITY
MERRF	MYOCLONIC EPILEPSY WITH RAGGED RED FIBERS
MMR	MUMP, MEASLES, RUBELLA
MRI	MAGNETIC RESONANCE IMAGING
NAS	NATIONAL AUTISTIC SOCIETY
NICE	NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE
NIMEH	NATIONAL INSTITUTE FOR MENTAL HEALTH
OR	ODDS RATIO
PDD-MRS	PERVASIVE DEVELOPMENTAL DISORDER IN MENTAL RETARDATION SCALE
PECS	PICTURE EXCHANGE COMMUNICATION SERVICE
PEHO	PROGRESSIVE ENCEPHALOPATHY WITH OEDEMA, HYPSARRYTHMIA AND OPTIC ATROPHY
PUD	PEPTIC ULCER DISEASE
PTSD	POST-TRAUMATIC STRESS DISORDER
RH	RHESUS DISEASE
RNIB	ROYAL NATIONAL INSTITUTE OF BLIND PEOPLE
RNID	ROYAL NATIONAL INSTITUTE FOR DEAF PEOPLE
SE	STANDARD E RROR
SPELL	STRUCTURED, POSITIVE, EMPATHIC, LOW AROUSAL AND LINKS
SI	SENSORY IMPAIRMENT
SNHL	SENSORY NEURAL HEARING LOSS
SS	SCOTOPIC SENSITIVITY
TEA	TOWARDS EQUITY AND ACCESS
TEACCH	TEACHING AND EDUCATION FOR AUTISTIC AND OTHER COMMUNICATION HANDICAPPED CHILDREN
TOM	THEORY OF MIND
TORCHES	TOXOPLASMOSIS, OTHER INFECTIONS, RUBELLA, CYTOMEGALOVIRUS, HERPES SIMPLEX AND SYPHILIS
VI	VISUAL IMPAIRMENT

1. INTRODUCTION

1.1. Background

Intellectual disability (ID), or learning disability, is a life-long condition with onset before adulthood, characterised by deficits in learning new skills, coping independently and understanding new or complex information (Department of Health, 2001). The World Health Organisation (1992) defines ID as ‘‘a condition of arrested or incomplete development of the mind, which is especially characterised by impairment of skills manifested during the developmental period, which contribute to the overall level of intelligence, i.e., cognitive, language, motor, and social abilities’’.

Globally, the reported prevalence of ID varies between 1% and 3% (Harris, 2006). Although there is variation in reporting, a meta-analysis by Maulik *et al*. (2011) of 52 studies, reported an overall ID prevalence of 10.37/1000. The highest rates were seen in low and middle income countries, in studies conducted in children and in those studies that used psychological assessments or scales for case identification (Maulik *et al*. 2011).

The Government’s 2001 White Paper ‘*Valuing People*’, specifically for people with ID, initiated a commitment to inclusion and enabling people with ID to “make use of mainstream services” (Department of Health, 2001). Since then, a number of policy initiatives have focused on the need to improve access to generic services for people with ID, so that they remain independent and integrated in their communities without suffering from social isolation (Department of Health, 2005). These include ‘*Making change happen*’ (Department of Health, 2003), the Foundation for People with Learning Disabilities (2004), Learning Disability Task Force (2004) and Disability Rights Commission (2006).

The provision of high-quality healthcare for people with ID is a national priority, as highlighted by ‘*Valuing People Now’* (Department of Health, 2009).

Reports such as '*Healthcare for All*' (Michael, 2008) and '*Death by Indifference*' (Mencap, 2007) have given increasing recognition to the substandard levels of healthcare and institutional discrimination experienced by people with ID. The follow-up report to '*Death by Indifference*', '*Six Lives: The Provision of Public Services to People with Learning Disabilities*' (Local Government Ombudsman and Health Service Ombudsman, 2009), raised serious questions and concerns about how well-equipped the NHS and Local Authorities were to plan for and provide services tailored to the needs of people with ID.

The Department of Health (Michael, 2008) has made it explicit that all healthcare providers should make reasonable adjustments to service delivery to meet the complex needs of vulnerable groups of service users in accordance with the Equality Act. Following one of the recommendations of the '*healthcare for All*' (Michael, 2008), the Learning Disabilities Observatory was established to provide better and more accessible information on the health of people with ID. The observatory aims to help hospitals and other providers to better understand the complex needs of people with ID and their carers which, in turn, should improve outcomes for this vulnerable client group (Public Health England, 2015). A better understanding of the challenges and barriers (Box 1.1) (Marston & Perry, 2013) to the delivery of better healthcare to people with ID will, therefore, enable clinicians to assess and treat various health difficulties in people with ID more effectively.

In July 2002, the Department of Health launched a consultation document ('*A Sign of the Times*') on modernising mental health services for deaf people. In 2005 also, the Department of Health and the National Institute for Mental Health in England (NIMHE) published best practice guidance on mental health and deafness: '*Towards Equity and Access*' (TEA) to show how mental health services for deaf people can be improved. There are, however, great concerns at the imbalance of emphasis placed on deafness in ID, as the disability in deafness is often down-played and thus, the importance of deafness in individuals with ID is frequently under-reported (Miller & Courtney, 2006).

Box 1.1: Barriers to accessing specialist care by people with intellectual disability (ID)*

Barriers related to service users

- Co-morbid physical disability and ill health.
- Accompanying Autistic Spectrum Disorder (ASD) and mental health problems.
- Severity of ID and communication difficulties.
- Atypical presentation of physical ailments through change in behaviour.
- Fear of hospital and investigation.
- Some people with ID might not understand the importance of their symptoms or hide them from healthcare professionals for variety of reasons.

Barriers related to service provision

- Lack of accessible/pictorial information.
- Under funding/lack of resources.
- Poor access to the clinics and general hospitals.
- Environmental barriers (e.g. lack of user friendly sign postings).
- Disconnection between teams (social and health services).
- Not taking into consideration needs of individuals with special needs for appointments (e.g. no flexible appointments, no waiting time for someone with ASD, not offering a quiet waiting area).

Barriers related to healthcare professionals and carers

- Attitudes and assumptions.
- Considered as low priority.
- Lack of awareness and training.
- Institutional discrimination.
- Marginalised status of sensory work.

*Taken from Marston & Perry (2013).

'Valuing People' (Department of Health, 2001) suggests that health action plans for people with ID should include details of the need for health interventions, such as interventions for individuals with sensory impairment (deafness and blindness).

People with ID and sensory impairments have complex needs which are often overlooked by professionals. These may include epilepsy, challenging behaviour, mental health problems and neurodevelopmental disorders, such as autism spectrum disorder (ASD). Such co-morbidities are often missed in clinical practice owing to diagnostic overshadowing and lack of awareness and training on the part of the healthcare professionals (Box 1.1).

Furthermore, there is no specialist inpatient or outpatient health care provision for people with ID and accompanying deafness, blindness and ASD. Community-based ID teams, at best, have few professionals with very basic awareness and knowledge of communicating with these individuals (Miller & Courtney, 2006) Teams usually do not include professionals with sensory impairment, or those who have a working knowledge of blindness and/or deafness and know how to facilitate access to appropriate services. Based on clinical experience, such service users frequently are placed in expensive homes that are often out of the county, but can offer greater expertise in dealing with sensory needs. Clearly, this has a significant emotional impact upon the service users and their family members as well as having financial implications on the NHS and local authorities.

Research into sensory impairments and ASD has the potential to improve service provision for people with ID. There are, however, potential challenges and barriers to the design of an appropriate research project investigating presence of sensory impairment and ASD in this population. These include heterogeneity of people with ID, sensory impairment and ASD, difficulties in finding a relatively large sample size and appropriate control group, presence of additional comorbidities (e.g. epilepsy) and a lack of valid/standardised assessment tools for diagnosing ASD in people with sensory impairment (Pring, 2005; Tager-Flusberg, 2005; Salt, 2010).

The majority of research available on the relationship between sensory impairment and ASD has either been conducted on children and those without significant ID or lacked an appropriate control group or statistical power to detect differences (i.e. small sample sizes) and not taken into account the main confounders (degree of brain damage [ID] and gender). The current thesis aimed to address this gap in knowledge.

Appendix 1 provides a tabulated critical review of the main published literature on the association between ASD and sensory impairments.

1.2. Aims of the thesis

The primary aim of this thesis was to explore the relationship between ASD and blindness/deafness in adults with ID, controlling for the nature and severity of brain damage and gender. The secondary aim was to determine the prevalence of sensory impairment, ASD and other co-morbid medical and mental health problems in adults with ID.

1.3. Ethical approval

The project was registered with the University of Leicester in Sep–Oct 2007. Ethical approval was granted by the Nottinghamshire Research Ethics Committee in April 2008 (Appendix 2).

Following a preliminary study conducted on the data available on an ID population register, the Leicestershire Learning Disability Register (LLDR) database, and after the first annual meeting between the researcher and the members of the thesis committee (Professor Terry Brugha, Professor Sabyasachi Bhaumik and Dr Michelle O'Reilly) on 9th February 2009, the study was approved as a PhD project, so that it could be completed by the researcher, a full time clinician working in the NHS.

1.4. Overview of the thesis

Chapter 2 describes the search strategy for the literature review and gives an overview of the literature in terms of prevalence of sensory impairment, its association with ASD, challenging behaviours and mental ill-health and its assessment and management in the general population and in individuals with ID. Chapter 3 lists the aims and objectives of the thesis, research question and hypotheses. Chapter 4 contains information about the methodology of the cross-sectional studies carried out for this thesis, describing the two-stage process and the statistical analyses. Chapter 5 describes the challenges and barriers to completing this thesis.

Chapter 6 presents the results from stage 1 of the research project, based on data from the LLDR. This chapter describes the study population, the relationship between sensory impairment, ASD and other potential confounders, the relationship between sensory impairment by individual autistic traits and the relationship between sensory impairment and challenging behaviours.

Chapter 7 presents the results from stage 2 of the research project, based on face-to face interview and direct clinical examination using objective assessment tools, involving two separate subgroups of blind and deaf service users and their controls. This chapter describes the aetiology of ID, demographic characteristics, co-morbid conditions and explores the relationship between sensory impairment, ASD, epilepsy, age, gender, ethnicity and degree of ID. Different assessment methods for diagnosing ASD are compared and the relationship between ASD, sensory impairment, gender, degree of ID and epilepsy is explored in more detail. Carers' views are also briefly described in this chapter.

Chapter 8 contains the discussion and conclusions of the thesis, giving a summary of the findings, discussing them in the context of the literature, describing strengths and limitations and making recommendations for future work.

2. LITERATURE REVIEW

2.1. Chapter overview

This chapter describes the literature review on the prevalence of sensory impairment and its association with ASD, challenging behaviours and mental ill-health in the general population and in individuals with ID. Although attempts were made to include all the published studies available to provide a narrative review on this topic, it must be emphasised that it was beyond the scope of the current research project to carry out a systematic review.

The review was conducted using the electronic literature databases *Ovid Medline*, *PsycLIT* and *PsycINFO* and internet searches (google, NHS evidence) for grey literature. The reference lists of key book chapters and articles related to the project were also researched. Searches included terms: intellectual disability, learning disability, mental retardation, developmental disability, Down syndrome, cerebral palsy; sensory impairment, visual impairment, hearing impairment, blindness, deaf-blindness, deafness, autism, autism spectrum conditions, autism spectrum disorders. The Boolean operator ‘’and’’ was used to combine searches with different terms such as sensory impairments, deafness, blindness, intellectual disability and autism spectrum disorder. Studies written in non-English language were excluded. Studies between 1950 and 2008 were initially included, and subsequently the review was updated to include research up until 2015. The researcher read the titles and abstracts of the studies to determine eligibility. A clinical narrative of the published studies on this subject is provided below. In addition, Appendix 1 provides a tabulated review summary of the main studies published on the association between ASD and deafness, blindness and deaf-blindness.

2.2. Prevalence Studies

2.2.1. Hearing impairment

According to the literature, hearing impairment is the most common sensory disorder in humans, with approximately 1–2 per 1000 children born deaf or

developing deafness during early childhood. Both incidence and prevalence of hearing impairment increase progressively with age (Kitson & Fry, 1990; Morton, 1991; Fortnum & Davies 1997; Fortnum *et al.* 2001; Agrawal *et al.* 2008).

In the UK, 1 in 7–8 people (over 10 million) have a hearing loss, and there are around 45,000 children who are deaf. Effective communication in the healthcare setting is, therefore, a priority for people with hearing impairment (Royal National Institute for Deaf People, 2003; Middleton *et al.* 2010). Degree of hearing impairment can be classified by audiometry (Table 2.1), which measures the intensity of a sound (decibel; dB) required for someone to hear at a particular frequency (Hertz; Hz). A person with normal hearing ability can hear sounds as low as 0–20 dB (a whisper is about 10–20 dB). A hearing impairment is defined as an average hearing loss in the best ear of 20 dB or more at 1, 2 and 4 kHz (Evenhuis *et al.* 2001; Acker & Crocker, 2004; Graham, 2004; Baines, 2007).

Table 2.1: Degree of hearing impairment based on audiogram

Degree of hearing impairment	Hearing loss in decibel (dB)
Mild	20–39
Moderate	40–69
Severe	70–94 (difficulty even with a hearing aid)
Profound	>95 (no perception)

Prevalence of hearing impairment is considerably higher in individuals with ID compared with the general population (Ellis, 1986). One study found a prevalence of 3.4% for severe to profound deafness in 18,657 people with ID in England and Wales (Kropka, 1984, cited in Carvill, 2001). This is approximately 35 times higher than that observed in the general population. Another study at the *German Special Olympics Summer Games 2006* showed a high proportion of undetected hearing impairment, even among those with a mild or moderate ID who were otherwise physically fit (Hild *et al.* 2008); nearly a quarter of 524

athletes with ID were positively screened for hearing impairment and 74 had an undetected hearing loss.

2.2.2. Visual impairment

According to the World Health Organisation (WHO), a visual acuity of less than 0.3 (a normal visual acuity is 1.0) or a visual field of below 30 degrees (a normal visual field is 180 degrees) is an indicator of visual impairment (van Den Broek *et al.* 2006). Blindness is an inability to see with a visual acuity of less than 3/60 and a visual field of fewer than 5 degrees (World Health Organisation, 2004). This has also been defined as a visual acuity of less than 0.05 or a visual field of fewer than 10 degrees by van den Broek *et al.* (2006).

While the prevalence of visual impairment has been reported to be 0.5%–2.0% in the general population (van den Broek *et al.* 2006), a number of studies have shown that this is several times higher in people with ID (Warburg, 1994 & 2001). Evenhuis (1995 & 2000) reported that the prevalence of visual impairment was at least 10 times higher in people with ID compared with the general population. Similarly, van Splunder (2003) and van Splunder *et al.* (2004 & 2006) found that 5% of people with ID in Holland were blind and 14% were partially sighted, which is between 10 and 30 times higher than the prevalence in the general population.

In the UK, an estimated 96,500 adults with ID are either blind or partially sighted. The estimated prevalence of blindness and partial sightedness in the adult ID population has been reported to be 9.3% (Robertson & Emerson, 2010).

2.2.3. Co-morbidity of visual impairment and hearing impairment

Approximately 40%–50% of congenitally blind children have additional disability, including hearing impairment, epilepsy, motor difficulties and ID (Hirst *et al.* 1993). Those with deafness also have a high co-morbidity of visual impairment. One visual screening programme of deaf students found that almost half (48%) also had significant eye problems (Brinks *et al.* 2001). A study in the USA reported that 27% of their deaf and hard-of-hearing students (6–19 years old)

had additional disabilities, e.g. ID 9%, developmental delay 5%, specific learning difficulties 8%, visual impairment 4%, and ASD 2% (Gallaudet Research Institute, 2008). In the UK, a literature review by the National Deaf Children's Society published in 2012 provides detailed information on the additional disabilities in children with hearing impairment (www.ndcs.org.uk).

It is estimated that there are about 365,000 people with some level of combined hearing and sight loss (deaf-blindness or dual sensory loss) in the UK (Robertson & Emerson, 2010). The prevalence of deaf-blindness is about 1 in 10,000 in school age children in the UK (Bond, 2000). The aetiologies of deaf-blindness are usually extreme prematurity, congenital rubella syndrome, meningo-encephalitis, Usher and other rare genetic syndromes, e.g. CHARGE syndrome. A US survey (Jensema, 1980) reported that over 60% of people who were deaf-blind had an Intelligence Quotient (IQ) below 50.

Most cases of deaf-blindness in people with severe to profound ID could easily go undetected if no objective assessment tools were used (Fellinger *et al.* 2009). Fellinger and colleagues (2009) found that most of their study subjects with deaf-blindness had profound ID (87.5%). They also reported a rise in the prevalence of deaf-blindness from 3.6% to 21.4% in their study population of adults with ID (n=224) following completion of objective assessments for both hearing and visual impairment, and concluded that deaf-blindness is usually missed in people with ID, especially in those with severe and profound ID.

Deaf-blindness has a huge impact on cognitive, psychosocial and language development to the extent that the behavioural and social manifestations can be very similar to ASD (Hoevenaars-van den Boom *et al*. 2009). Deaf-blind people are highly reliant on carers to actively participate in activities; therefore, interventions to engage this group of service users in social interaction and to promote their independence are extremely important to avoid isolation and social exclusion (Prain *et al*. 2010).

2.3. Factors that influence the prevalence of sensory impairment

2.3.1. Age

Using data from a New York register of 45,000 adults (aged 35 years and older) with ID, Janicki and Dalton (1998) found that prevalence of hearing impairment increased significantly with age (16% in 35–59 years olds vs 35% in 60–79 year olds). Thus studies on older sample populations yield higher prevalence estimates of sensory impairment (Cooke, 1989; Evenhuis *et al.* 1995 & 2000); for example, studies in Holland reported a prevalence of 16.7% for blindness and 67% for partial visual impairment in adults with Down syndrome who were aged 50 years or over (van Splunder, 2003; van Splunder *et al.* 2004 & 2006).

2.3.2. Degree of ID

The prevalence of sensory impairment is also affected by the degree of ID; the more severe the degree of ID, the higher the prevalence of sensory impairment (Evenhuis *et al.* 2001). A study by van den Broek *et al.* (2006) found that almost all their sample of people with severe and profound ID had visual impairment.

2.3.3. Study population

Most individuals with mild ID live in the community and are not registered with local ID services. Therefore, studies using ID case registers or on those in receipt of specialist social and healthcare services are unable to accurately measure the prevalence of mild ID in the general population. However, such studies tend to comprise a representative sample of people with moderate to profound ID because they have additional support and healthcare needs (Smiley, 2005). The prevalence of sensory impairment increases in people with severe forms of ID; therefore, studies in this population tend to render relatively high prevalence rates of sensory impairment. This is also the case for studies of adults with ID living in inpatient units or long-stay hospitals, as these populations are weighted heavily towards the severe end of the ID spectrum.

2.3.4. Methods of case ascertainment

Many people with ID lack effective communication skills; therefore, they find it difficult to complain or express their symptoms effectively, which leads to

unrecognised sensory impairment (Evenhuis *et al.* 2001). Warburg (1994) found that the concordance rate in diagnosing visual impairment between carers' report (questionnaire) and objective clinical assessment was less than one-third (32%). Similarly, it has been reported that the prevalence of hearing impairment is lower if cases are identified by subjective reports from carers (9.4%), as opposed to objective clinical assessments (38.9%) (Lavis *et al.*1997).

2.3.5. Ethnicity

Deafness may be more prevalent among the immigrant population. This can be a consequence of marriage within close family networks, greater chance of poverty and inadequate access to healthcare and immunisation in this population (Admiraal & Huygen, 2000; Royal National Institute for Deaf People, 2003). A study in Pakistan indicates that there is a strong association between poverty and an increased rate of blindness (Gilbert *et al.* 2008).

2.3.6. Aetiology of ID

There are number of conditions that cause both ID and sensory impairment (Table 2.2), including Waardenburg syndrome, Usher syndrome, Down syndrome and congenital rubella syndrome (Chess, 1971; Jones, 1997; Toriello *et al.* 2004; Firth *et al.* 2005). The aetiology of hearing impairment, based on whether it is congenital or acquired, ranges from genetic conditions to infectious causes (Admiraal & Huygen, 2000). Approximately half of cases of congenital deafness are due to genetic causes, mainly recessive genes, e.g. gene GJB2 for Connexin 26 protein (Marazita *et al.* 1993; Steel & Bussoli, 1999). Approximately 80% of non-syndromic hearing impairment is inherited through autosomal recessive genes, with the remaining 20% through autosomal dominant mode. Hearing loss is a common clinical feature in mitochondrial syndromes such as MELAS syndrome (mitochondrial encephalopathy, lactic acidosis and stroke-like episodes), MERRF syndrome (myoclonus epilepsy, ragged red fibres) and Kearns-Sayre syndrome. However, these account for only about 2% of inherited cases and X-linked disorders are the causes in about 1% of cases of hearing impairment (Hutchin *et al.* 2001; Cryns & Van Camp, 2004; Hsu *et al.* 2005). An interesting mitochondrial DNA mutation (m.1555A>G), which is exclusively inherited maternally, is known to predispose

a person to vestibular and ototoxic effects of even therapeutic doses of aminoglycosides. The association is so strong that it has been suggested that everyone, who is going to have multiple courses of aminoglycosides, should be screened for this mutation to detect the vulnerability to hearing loss (Bitner-Glindzicz & Rahman, 2007).

Cerebral palsy (CP) is another cause of sensory impairment and ID. CP occurs in approximately 2 per 1000 live births in developed countries, and is an umbrella term used for a group of conditions that cause movement problems. It is associated with additional disabilities such as ID, epilepsy and sensory impairment. Common risk factors for CP are prematurity, maternal iodine deficiency, and Rhesus incompatibility. With advancement of peri-natal screening, it is now argued that a considerable number of cases of CP occur during pregnancy, rather than as a consequence of complicated delivery, and indeed can precede postnatal adverse events (Rosenbaum, 2014). A recent population-based epidemiological study in Norway (Tollanes *et al.* 2014) using the Medical Birth Registry reported a 15-fold increase in the risk of CP in the second twin, 9-fold in the non-twin sibling and 3-fold in a half sibling. This suggests that a genetic cause, a shared early environment, or both, are implicated as risk factors for CP.

Structural anomalies of sensory organs, such as narrowed ear canals and Keratoconus, are common in people with Down syndrome. As a result, age-related hearing loss (pres-bycusis) occurs several decades earlier in people with Down syndrome compared with the general population (Prasher & Janicki, 2002; Meuwese-Jongejeugd *et al.* 2005 & 2006). Nearly all adults with Down syndrome have anomalies of the ear with a predisposition to infection which needs aggressive treatment to avoid irreversible hearing loss (Shott *et al.* 2001). Hearing impairment due to impacted earwax is relatively common. A study on people with Down syndrome also found that 57% of those aged 35–62 had a bilateral loss greater than 40dB and only 25% had been diagnosed before the study (Evenhuis *et al*, 2001). Down syndrome can cause both conductive and sensorineural hearing impairment.

Usher syndrome is one of the most common causes of deaf-blindness in adults, causing 5–10% of cases of deafness, 18% of cases of retinitis pigmentosa and visual impairment and over 50% of cases of deaf-blindness. The prevalence in the general population is 3–5 per 100,000 (Rosenburg *et al.* 1997). Usher syndrome causes gradual loss of vision due to progressive retinitis pigmentosa. Retinitis pigmentosa, which is essential for diagnosis, can be confirmed by electro-retinography. The visual impairment usually starts with night blindness during adolescence and progresses to tunnel vision and blindness. Usher syndrome may also cause problems with balance.

Table 2.2: Examples of genetic syndromes associated with sensory impairment

Genetic syndrome	Signs and symptoms
Alport	Kidney abnormalities, deafness, ocular abnormalities
CHARGE	Coloboma, heart defects, atresia choanae, retardation of growth, ear anomalies, deaf-blindness
Coffin Lowry	Hypotonia and short stature, hearing impairment
Jervell and Lange-Nielson	Fainting and long QT interval in ECG, deafness
Klippel Feil	Short webbed neck, visual and hearing impairment
MELAS	Myopathy, encephalitis, lactic acidosis and stroke, associated with hearing impairment
Neurofibromatosis	Café au lait spots, visual and hearing impairment
Pendred	Hypothyroidism, deafness
Stickler	Joint hyperflexibility, cataract, glaucoma and retinal detachment, visual and hearing impairment
Treacher Collins	Facial dysmorphology and coloboma, visual and hearing impairment
Usher	Retinitis pigmentosa, deaf-blindness
Waardenburg	Blue iris and white forelock, deafness
Down	Heart and digestive system defects, visual and hearing impairment

QT interval in cardiology: time between Q wave and T wave; ECG: electrocardiography

Rubella in pregnancy can cause sensorineural deafness, central auditory imperceptions, visual impairment and developmental delay, all of which may be under-diagnosed in people with ID. Chess (1971 & 1977) & Chess *et al.* (1978) studied 243 preschool children with congenital rubella syndrome and found that 37% had ID, 15% had reactive behaviour disorder and 7% had autism. When followed up at the age of 8–9 years, the prevalence of ID had decreased; however, challenging behaviour (i.e. behavioural problems) had increased owing to neurological damage. The authors also reported new cases of ASD and few remissions at follow up and hypothesised that the course of ASD was that of a chronic infection, with remission and delayed emergence of symptoms. In an update of the literature on congenital rubella, Berger and colleagues (2011) concluded that the measles, mumps and rubella (MMR) vaccine had, between 2001 and 2010, prevented several thousand cases of ASD, ID and sensory impairment in the USA. In addition to the above, it is recognised in the literature that other adverse events during pregnancy can also cause sensory impairment and ASD including infection with Cytomegalovirus (Sweeten *et al.* 2004).

2.4. Relationship between sensory impairment, mental illness and ASD

Pre-lingual and profound sensory deficits are reported to adversely affect acquisition of theory of mind and abstract thinking, either independently or through accompanying brain damage, which might result in ASD/autistic-like symptoms (Pring, 2005; Hoevenaars-van den Boom *et al.* 2009).

ASD, a polygenic neuro-developmental (lifelong) condition, is characterised by qualitative impairment in a triad of social interaction, reciprocal communication and imagination. It is one of the most invisible causes of lifelong disability, with an estimated annual cost in support and lost productivity of more than 28 billion pounds in the UK (Knapp, 2009). Since Kanner first described the condition in 1943, an increased awareness of the condition and changes in the diagnostic criteria (Wing, 1996; Wing & Potter, 2002) have led to a substantial increase in the estimated prevalence of the condition in the general population from about 4–6/10000 to 0.1% and 0.3% (Lotter, 1966; Bouras *et al*. 1999; Fombonne,

1999; Kielinen *et al.* 2000) and most recently to 1.0% to 1.1% (about 1 in 100) and 1.5% (1 in 63 children) (Baird *et al*. 2006; Fombonne *et al*. 2006; Brugha *et al*. 2011 & 2012; Centre for Disease Control and Prevention, 2014). ASD is more prevalent in boys and those with severe and profound ID (Baird *et al*. 2006; Mandy *et al*. 2012).

People with ASD are more likely than the general population to have ID, language impairment, sensory impairment, academic under-achievements, movement disorders and mental and behavioural disorders (Simonoff *et al.* 2008; Charman *et al*. 2011). This can have the undesired consequence of delayed diagnosis in adulthood because priority is given to the other co-morbidities, which create challenges for health and social care professionals and add considerably to the burden of care (Baron-Cohen *et al.* 2009a). Inadequate identification of adults with ASD leads to inappropriate provision of care and can, therefore, result in inadequate treatment of co-existing mental and physical health conditions such as challenging behaviours, epilepsy and insomnia. The prevalence of co-morbid challenging behaviours is high in people with ID and ASD which, in turn, has a negative impact on independence and community integration, and can lead to institutionalisation (Jordan, 2001).

ASD is highly prevalent in individuals with ID compared with the general population (Bhaumik *et al.* 1997). Prevalence rates vary between 8% to 27% (Wing & Gould, 1979; Deb & Prasad, 1994; Beadle-Brown *et al.* 2002; de Bildt *et al.* 2005; Bhaumik *et al.* 2008), depending on diagnostic criteria used and ID severity in the sample population. ASD is more prevalent as the severity of ID increases (Deb & Prasad, 1994; Brugha *et al.* 2012) and in men, although the sex differences do not appear to be as pronounced as in the general population (Brugha *et al.* 2012).

The rate of ID is also higher in people with ASD compared with the general population but the figures vary substantially, ranging from as low as 6.8%–8.0% (Corbett *et al.* 1979; Bhaumik *et al.* 1997), to 15%–17% (Gillberg & Soderstrom, 2003; de Bildt *et al.* 2005), to as high as 42%–70% (Bouras *et al.* 1999; Kielinen *et al.* 2000; LaMalfa *et al.* 2004; ADDMNS Year 2000 Principal Investigators,

2007) and 65%–88% (Gillberg, 1995; Berney, 2000) based on the assessment tools used and differences in diagnostic threshold and the study population. A study of a cohort of young people (under 17 years old) living in Sweden reported a positive association between maternal depression and pre-natal use of antidepressant agents (selective serotonin reuptake inhibitors and monoamine oxidase inhibitors) and ASD. The association was particularly strong in those without ID, after adjusting for confounders (age and gender). The authors controlled for other confounders, such as recorded psychiatric problems in the parents, birth parity, parental ages, migration status, income, education and occupation (Raj *et al*. 2013).

2.4.1. Hearing impairment and ASD

If not exposed to sign language early on in life, a deaf child will miss out on incidental learning, such as family gatherings and conversations, TV and radio programmes, traffic outside, birds singing, thunder, waves and wind rustling. This has a huge impact on cognitive, psychosocial and language developments to the extent that the manifestation can be very similar to ASD.

Early deafness in children born into hearing families may delay the development of theory of mind due to a lack of exposure to sign language. Children with an existing ID are at a higher risk of such developmental delays. These children are generally found to perform no better than autistic individuals of a similar mental age based on theory of mind tests (Russell, *et a*l. 1998; Peterson & Siegal, 1995 & 2000).

Hearing impairment and ASD are both disorders of communication. Children eventually diagnosed with ASD are often initially thought to be deaf by the parents (Grewe *et al.* 1994). However, both conditions may be present in a child simultaneously. Therefore it is not surprising to see an increased prevalence of hearing impairment reported in people with ASD in the literature.

Studies conducted by the Gallaudet Research Institute report a gradual increase in dual diagnosis of hearing impairment and ASD among deaf and hard-of-hearing children (Gallaudet Research Institute, 2008). In 1999,

Rosenhall and colleagues reported on the presence of hearing impairment in those with a diagnosis of ASD and found that 9.5% (19 out of 199 autistic children) had a hearing impairment. The prevalence of profound hearing impairment in their study was approximately 3.5%. In 1991, Jure and colleagues reported a 4% prevalence of ASD (n=46) in a sample of 1150 children with hearing impairment, with a male to female ratio of 2:1. They reported that this co-morbidity had led to a delay of approximately several years in diagnosis of either condition. Their research also showed that one-third of their sample (35%) had accompanying visual impairment, 17% epilepsy and 24% neurological signs. Over 80% of their sample had severe to profound hearing impairment with 17% of them having a genetic syndrome as the aetiology of their deafness. The authors also found a relationship between the degree of ID (not degree of deafness) and prevalence of ASD, therefore it was argued that the high prevalence of ASD reported was mediated through accompanying brain damage (a literature review on the following website provides more information on this topic: http://aucd.org/).

2.4.2. Visual impairment and ASD

Infants with congenital blindness lack visual and social experiences as they begin their life with a major developmental disadvantage i.e. they cannot see the world, the items around them and other people's body language or facial expressions (Pring, 2005). It is believed that this is one of the reasons why children with congenital visual impairment are delayed in theory of mind development, though an accompanying brain damage has been generally regarded as the main reason for deficit in the development of theory of mind (Begeer *et al.* 2014). Fraiberg (1977) observed that the concept of distinguishing between oneself ("I"), others ("You" or "He"') and objects is delayed in congenitally blind children, in comparison to sighted children. The development of imaginative play in children with visual impairment also occurs later than sighted children. Repetition, or echoing, of words spoken by another person (echolalia) has also been reported in blind children, irrespective of ASD diagnosis (Conti-Ramsden, 2005). Echoing appears to be associated with difficulty in using pronouns (pronominal reversal or using ''He/She'' for ''I'' or "You"') and is also seen in sighted people with ASD (Fraiberg, 1977; Andersen

et al. 1984 & 1993; Conti-Ramsden, 1999 & 2005). Some blind children present with several symptoms suggestive of ASD. Although common in congenitally blind children, these do not amount to a formal ASD diagnosis. The symptoms are known in the literature as 'blindism' or autistic-like features (Fraiberg, 1977; Hobson *et al.* 1999). Warren (1986) described blindism as a result of somatosensory deprivation and social isolation in congenitally blind children (e.g. eye poking, light gazing, rocking, tapping, twirling, flicking fingers in front of lights and spinning). Jan and Groenveld have reported (1990) that eye pressing or eye poking can be seen in blind children; this might be self-stimulatory in nature but there might also be other reasons for this which need to be explored individually in each case.

A diagnosis of ASD has however been reported more commonly in people with congenital blindness (Hobson, 2002) and this association is not limited to only Leber's congenital amaurosis or retinopathy of prematurity (ROP) as once was thought to be the case (Chase, 1972; Brown *et al.* 1997; Fazzi *et al.* 2007). Although it has been suggested that ASD and autistic-like symptoms in children with conditions such as Leber's congenital amaurosis may be related to a neurological disorder rather than absence of visual and social experiences (Rogers & Newhart-Larson, 1989), this association has also been noted in congenital blindness due to other aetiological conditions. For example, previous research also revealed that, children with congenital rubella syndrome and visual impairment had a high rate of autism/autistic-like symptoms (Chess, 1971 & 1977; Chess *et al.* 1978). In 1997, Brown and colleagues described a study on 24 blind children, aged 3–9 years, at several special schools for blind children and found out that the majority had autistic-like features. The authors subsequently compared 9 of these children with 9 sighted autistic children who had been matched on age and IQ (all had IQ<70) and determined that blind children had autistic-like symptoms that were qualitatively different in comparison to the sighted autistic children. They reported that a formal diagnosis of ASD could only be made in two congenitally blind children and that blindness had added to the ASD symptomatology which could be amenable to interventional strategies early on in life.

Cass and colleagues (1994) studied 102 infants with congenital blindness and found that 11% showed set-back in their development. Nearly one-third (31%) of those with profound visual impairment (10 out of 32) developed a developmental set-back at 15–27 months, compared with 1 in 72 of children with severe visual impairment (those who had form vision) who developed the developmental set-back. The symptoms were similar to ASD. The set-back phenomenon occurred regardless of the aetiology of congenital blindness but was related to its severity. Set-back was defined in those whose early development had a normal direction but then stopped progressing or regressed (Dale, 2005). This was reported to be more common in more severe forms of visual impairment (owing to the variety of causes), in males, and in those with accompanying brain damage with absence of form vision (Dale, 2005). The authors concluded that even slight form vision, particularly where present from the first few months of life, seemed to be a protective factor for developmental set-back. Other risk factors reported for this developmental set-back were the number of lesions in the brain magnetic resonance imaging (MRI), failure in or lack of the brain myelination process (Waugh *et al.* 1998), degree of abnormal connectivity (Sonksen & Dale, 2002), ID, deprivation of visual input negatively affecting the functional systems of the brain (Dale, 2005) and qualitative factors within the family environment (Dale, 2005; Dale & Salt, 2008). Given the latter risk factor, it has been suggested that early intervention starting within the first two years of life might be beneficial to improve development. It has been shown that, with early intervention, some cases of set-back will reverse (Cass *et al.* 1994; Dale & Sonksen, 2002; Sonksen & Dale, 2002; Dale, 2005). However, it has been argued that this reversibility cannot take place with a neurodevelopmental condition such as ASD, as symptoms by definition have a life-long course (Perez-Pereira & Conti-Ramsden, 1999 & 2005). Perez-Pereira and Conti-Ramsden (1999 & 2005) argued that the majority of blind children studied by Cass *et al.* (1994) were not autistic and those who had ASD had additional disabilities such as brain damage and ID. They concluded therefore that visual impairment per se did not cause the autistic symptoms.

Similarly, it has been argued that blind children might experience delay in their development of theory of mind, but that it does eventually develop; children

make up for their lack of sight with other sensory modalities to overcome developmental challenges that they are facing because of congenital blindness (Perez-Pereira & Conti-Ramsden, 1999 & 2005). Some researchers do not believe in a top-down or bottom-up theory of ASD pathogenesis but more in the interplay of these in a bi-directional interaction; for example, they argue that sensory difficulties can affect neurological development during early infancy and vice versa (Frith, 2003; Leekam & Wyver, 2005). Therefore, there is still debate as to whether ASD or autistic-like symptoms in blind children directly originate from a pure sensory deprivation early in life (e.g. blindness) or an accompanying brain damage.

To understand the effect of visual impairment on development, a pure research sample of people with congenital disorders of peripheral visual pathway is therefore needed (Dale, 2005; Pring, 2005), i.e. without accompanying neurological or posterior visual pathway damage which comprises possibly minority of all those with congenital blindness. In reality, however, it would be extremely difficult to have a 100% pure sample in the ID population where the presence of co-morbidities is a norm. Box 2.3 highlights the challenges that researchers face when studying the relationship between ASD and sensory impairment (Pring, 2005).

Box 2.3: Challenges in studying the relationship between Autism Spectrum Disorder (ASD) and sensory impairment

- Within group heterogeneity of people with sensory impairment (e.g. different degree of blindness or deafness).
- Within group heterogeneity of people with ASD & Intellectual Disabilities.
- Achieving sufficient sample size.
- Finding an appropriate control group.
- Presence of additional disabilities (e.g. brain damage).
- Lack of valid assessment tools.
- Low incidence rate.
- Difficulty in determining exclusion and inclusion criteria.
- Geographical dispersion.

2.4.3. Deaf-blindness, mental illness and ASD

Studies exploring the relationship between deaf-blindness and ASD are often based on small sample sizes because deaf-blindness is relatively rare. Diagnosing ASD is also extremely difficult in this population. In one study, 5 deaf-blind service users with a clinical consensus diagnosis of ASD were compared with 5 service users without a clinical consensus diagnosis of ASD (Hoevenaars-van den Boom *et al*. 2009). The authors developed a semi-structured instrument, "Observation of characteristics of Autism in persons with Deaf-Blindness: O-ADB'' based on item checklists of the Autism Diagnostic Observation Schedule (ADOS; Lord *et al*. 2006), Autism Diagnostic Interview (ADI-R Le Couteur *et al*. 2003), a "Hands on" assessment and Autism Screening Instrument for Educational Planning (Krug *et al*. 1980). The authors standardised the tool by using a set of materials suited for persons with deaf-blindness and prompts by the assessor to increase the chance of deaf-blind people engaging in the study and showing their competencies in the relevant domains. Even with the additional measures, diagnosis of ASD in deaf-blind people with ID was found to be a challenge, primarily because their communication skills were impaired as a result of their dual sensory loss. The situation was worse for those who had an additionally significant ID. However, despite a large overlap in symptom presentation of those who were deaf-blind with those who were both deaf-blind and autistic, it was possible to diagnose ASD by thorough consensus assessment. The authors found that autistic deaf-blind people were different in terms of their openness for contact, reciprocity/joint attention and communicative signals/functions from their non-autistic counterparts. However, the two groups did not differ in relation to stereotypies, play, exploratory activities, coping with changes and problem solving techniques. Both groups had similar levels of ID.

In children with deaf-blindness, development of language first and then other skills such as cognitive ability, socio-emotional development and motor development are affected most (van Dijk & Janssen, 1993; Gense & Gense, 2005). People with deaf-blindness with or without ASD can present with stereotyped behaviours, which are also commonly reported in people with ID with or without ASD, as a compensation for social and sensory deprivation.

However, over time and as children with deaf-blindness grow older, these behaviours tend to reduce in frequency and intensity if children are offered more opportunities to communicate with others and explore the environment around them (van Dijk *&* Janssen, 1993; Murdoch, 2000; Frith, 2003; Hartshorne *et al*. 2005). Many symptoms in deaf-blind children such as lack of initiation and social withdrawal are secondary to their dual sensory loss and their dependence on carers (Knoors & Vervloed, 2003). These symptoms do not differentiate between autistic and non-autistic deaf-blind children. Although communication and social interaction are impaired in all people with deaf-blindness, these impairments are more prominent and have a greater adverse effect on the quality of life of people when there is a co-morbid ASD (Hoevenaars-van den Boom *et al*. 2009).

Those with CHARGE (coloboma, heart defect, atresia choanae, retarded growth and development, genital hypoplasia, ear anomalies/deafness) syndrome, one of the most common causes of deaf-blindness, have been reported to have more delayed language development and autistic symptoms when compared with those who have other developmental conditions, such as Down syndrome and William Syndrome (Graham *et al*. 2005; Peltokorpi & Huttunen, 2008).

In 2002, Carvill and Mraston reported a high rate of ASD (n=15; 83%) in their sample of 18 adults with sensory impairment referred to their service for self-injurious or aggressive behaviours. Of these, 11 had deaf-blindness, 4 were blind (but not deaf) and 3 were deaf (but not blind). The aetiology of sensory impairment was congenital rubella syndrome in 12 of their sample (67%). Those remaining had Joubert syndrome (n=1), infection during infancy (n=1), Leber's congenital amaurosis (n=2), self-injury (causing blindness; n=1) and rhesus haemolytic disease (n=1). The majority were males, with an average age of 31 year old. The authors reported that it was challenging to reach a final psychiatric diagnosis owing to their subjects' unusual presentations, with only 7 cases being diagnosed with postulated depression and 4 with a definite diagnosis of depression. Service users responded to a variety of treatment strategies including medication, staffing support and environmental adaptation. The authors reported that the majority of individuals (n=15; of which 10 were deaf-

blind, 3 were blind and 2 deaf) had atypical ASD or autistic-like features based on ICD-10 criteria (World Health Organisation, 1992); they concluded that this high rate was less of a clinical concern and more of academic interest, since the management of all these presentations were the same in clinical practice. Psychotic symptoms have also been reported in the context of deaf-blindness in people with Usher syndrome (Hess-Rover *et al.* 1999).

Appendix 1 provides a tabulated summary of some of the published studies on the association between ASD and deafness, blindness and deaf-blindness.

2.4.4. Other sensory impairment in ASD

In addition to visual and hearing impairments, there are a number of other sensory difficulties reported in people with ASD that can affect their cognitive and psycho-social development (Bogdashina, 2003). Although investigating these is beyond the scope of the current research project, a summary of relevant literature on sensory processing and motor issues in ASD, based on a publication by Hilton (2011), is provided below.

People with ASD have atypical sensory processing behaviours and therefore struggle to respond to personal and environmental demands (Dunn, *et al*. 2002), which has a negative impact on self-esteem, symbolic play, cognitive development and social skills (Bundy *et al*. 2002). There are several types of sensory processing disorders (Hilton, 2011): for example, in sensory discrimination disorder, the individual is unable to recognise the similarities and differences between stimuli because they have difficulty in interpreting the quality of these stimuli (Miller *et al*. 2007).

In sensory modulation disorders, the individual might react with an abnormal behaviour, such as sensory seeking, sensory avoiding, over responsiveness or under responsiveness and an association has been found between these behaviours and the degree of their social impairment (Hilton *et al*. 2007). Studies with quantitative MRI and electroencephalography (EEG) have shown disturbances of sensori-motor gating (an ability to modulate responses to the external sensory stimuli) in ASD (McAlonan *et al*. 2002; Perry *et al*. 2007;

Orekhova *et al*. 2008). Sensory processing abnormalities in ASD might restrict an individual's participation in group games and social activities that involve spontaneity during their development (Ben-Sasson *et al*. 2007)

Two processes are disrupted in people with ASD; that of habituation (recognising familiar stimuli) and sensitisation (heightened response to an important stimulus) (Hilton, 2011). The neuro-anatomical abnormalities found in post-mortem studies confirm neurological foundations for the above (Casanova, 2007). Research has also shown differences in cortical organisation and the neuronal networks of people with ASD in comparison to people without ASD (Coskun *et al*. 2009) and that these sensory processing disorders involve various senses that are interconnected to each other (Kern *et al*. 2007b).

Abnormalities, such as deficits in vestibular modulation, can result in depression or anxiety disorders and might be a cause of stereotypical and repetitive behaviours, such as swinging and rocking, commonly seen in service users with ASD in clinical practice (Pfeiffer *et al*. 2005; Kern *et al*. 2007a; Baker *et al*. 2008).

Another clinical presentation of ASD is hyposensitivity and hypersensitivity to sound (Khalfa *et al*. 2004). In 2003, Bonnel and colleagues reported that autistic children had an enhanced sensitivity compared with non-ASD controls when given tasks on pitch discrimination and categorisation, and musical pitch (Heaton *et al*. 1998; Lepisto *et al*. 2008). Various electrophysiological measures such as cortical auditory evoked potentials have been used to explore the underlying auditory processes in people with ASD (Bomba & Pang, 2004; Tharpe *et al*. 2006; Nieto Del Rincon, 2008).

Auditory hyper-responsiveness, under responsiveness and hyperacusis are also well known in clinical practice in people with ASD (Osterling and Dawson, 1994; Rosenhall *et al*. 1999). Similarly, visual hypo-responsiveness might lead to staring intensely at others, while hyper-responsiveness can lead to avoidance of bright lights (Dunn, 1999). Other visual perceptual abnormalities in people with ASD reported in the literature include problems with pursuit eye

movements, motion perception (Takarae *et al*. 2008), Nystagmus (Tantum, 2012) and increased visual acuity (Ashwin *et al*. 2009). Multisensory processing disorder, problems in managing multiple sensory inputs at the same time, can also occur; this appears to be significantly associated with the degree of ASD (Baier *et al*. 2006; Kern *et al*. 2007b). Peripheral auditory asymmetry has also been reported in autism (Khalfa *et al*. 2001).

Similarly, cortical deafness and blindness have been reported in people with a normal structure of sensory organs. These are mainly as a result of the damages (e.g. hypoxia, haemorrhage, trauma etc.) to those parts of the brain that process and integrate the sensory stimuli from the eye or ears (Groenveld *et al*. 1990 & Cavinato *et al.* 2012).

In addition to the above, research has also been conducted on talent, musicality, blindness and ASD, which are believed to be related to weak central coherence in these individuals (Pring &Tadic, 2005; Ockelford *et al*. 2006; Ockelford & Matawa, 2009). In autistic savants, there are certain areas of the brain which have the ability to detect patterns and complete missing information; this might be related to their hypersensitivity toward pattern recognition (Baron-Cohen *et al.* 2009b).

2.4.5. Association of sensory impairment with mental illness in the general population

Being deaf in a hearing world can results in discrimination; people who are deaf are less likely to have appropriate jobs and more likely to have restricted access to education and social and health services. Deaf people tend to use more body contact and appear to be more direct and abrupt with their comments, with no intention of offending others, mainly trying to gain the attention of others. Miscommunication is therefore very common between deaf people and hearing people. Sign language is not commonly used among services, such as psychiatric, police, probation, education and health and social services. Therefore, deaf people experience more barriers in accessing services, which can be extremely frustrating and challenging for them (Hindley & Kitson, 2000; du Feu & Fergusson, 2003; Royal National Institute for Deaf People, 2003;

Austen & Crocker, 2004; Miller & Courtney, 2006; Gentili & Holwell, 2011; Fellinger *et al.* 2012; Sessa & Sutherland, 2013). Deaf people's language, whether in sign, speech or writing, may be limited or idiosyncratic. Written English can be poorly presented in British Sign Language (BSL) word order, which gives the appearance of formal thought disorders. For professionals who are not 'deaf aware', features of deaf culture and sign language might be misinterpreted as challenging behaviour or mental illness. Therefore, misdiagnosis (both over and under diagnosis) of mental illness is common (Hindley & Kitson, 2000; du Feu & Fergusson, 2003; Austen & Crocker, 2004; Miller & Courtney, 2006; Austen & Jeffery, 2007; Gentili & Holwell, 2011; Fellinger *et al.* 2012; Sessa & Sutherland, 2013). As a result, various personality traits such as immaturity, egocentricity, lack of empathy, lability and explosive personalities have all been unfairly attributed to deaf people. Some researchers have gone on to categorise such personality traits as the deaf mind (surdophrenia) (Basilier, 1964; Carvill, 2001).

In addition, deaf children commonly experience marginalisation, scapegoating, physical, emotional and sexual abuse, which are risk factors for mental illness (Hindley & Kitson, 2000; du Feu & Fergusson, 2003; Austen & Crocker, 2004; Hindley, 2005; Miller & Courtney, 2006; Gentili & Holwell, 2011; Wright, 2011; Fellinger *et al.* 2012; Sessa & Sutherland, 2013). Studies of the effects of deafness on mental health in adult populations have rendered different outcomes (Hindley & Kitson, 2000). Earlier studies report lower rates of depression and anxiety in the deaf population; however, later studies argued that an absence of appropriate service provision for this population may have caused this reduction. The rate of schizophrenia in the deaf population is similar to that of hearing people and it is believed that schizophrenia in this population is less responsive to antipsychotic medication and may require higher dosages and augmentation with mood stabiliser (Kitson & Fry, 1990; Hindley & Kitson, 2000; du Feu & Fergusson, 2003; Austen & Crocker, 2004; Hindley, 2005).

If no remedial measures are taken early in life, congenital deafness can adversely affect language, identity and psychosocial/cognitive development (Hauser *et al*. 2006; Woll, 2008). Research shows that between 40% to 45–

50% of deaf children suffer from mental illness, emotional or behavioural problems, in comparison to 25% of their hearing peers (Hindley *et al.* 1994; Hindley, 1997; Vostanis *et al.* 1997; Hindley, 2000; Austen & Crocker, 2004; Department of Health, 2005; Gentili & Holwell, 2011; Wright, 2011; Fellinger *et al.* 2012; Sessa & Sutherland, 2013). The prevalence of attention deficit hyperactivity disorder (ADHD) in children who are deaf from non-inherited causes is reported to be higher than that of hearing children (Hindley & Kroll, 1998; Hindley, 2005). A study on deaf adolescents and young children has shown an excess of behavioural and emotional problems (van Gent *et al.* 2007) mediated through difficulties in communication and experience of stigma and oppression in a predominantly hearing world. Some of the causes of challenging behaviour and mental illness in deaf children, however, might be due to associated ID, co-existing physical health problems, including the genetic syndrome causing the deafness, and environmental issues in relation to communication breakdown (Roberts & Hindley, 1999; Bond, 2000; Austen & Crocker, 2004; van Gent *et al*. 2007; Gentili & Holwell, 2011; Wright, 2011; Fellinger *et al.* 2012; Sessa & Sutherland, 2013).

Being born in a hearing family, which is the case for 90 to 95% of congenitally deaf children, appears to be a risk factor for the development of mental illness and emotional problems, because hearing parents with deaf children are usually communicating through speech which further jeopardises language acquisition in children with deafness (Moores, 1987; Lederberg & Everhart, 1998 & 2000; Meadow-Orlans & Erting, 2000; Austen & Crocker, 2004). It has been found that delayed language development (either speech or sign language) increases the likelihood of mental illness (Hindley *et al*. 1994; Austen & Crocker, 2004; Gentili & Holwell, 2011; Wright, 2011; Fellinger *et al.* 2012; Sessa & Sutherland, 2013). In contrast, deaf children, born to deaf families or to hearing families with signing parents, develop language (sign language) in time, and research shows that in contrast to those deaf children with delayed language development, these children have a similar risk of developing mental illness as their hearing counterparts (Bishop, 1983; Sinkkonen, 1994; Sutton-Spence & Woll, 1998; Austen & Crocker, 2004; Strong, 2007). This confirms the positive impact of mutual communication between parents and deaf children. Using sign language

early on in the family environment helps to facilitate development of speech as well as psychosocial and emotional development in deaf children; these are all protective factors against mental health problems (Mahshie, 1995; Pickersgill & Gregory, 1998; Austen & Crocker, 2004; Sutherland, 2004; Gentili & Holwell, 2011; Wright, 2011; Fellinger *et al.* 2012; Sessa & Sutherland, 2013).

Previous reports also suggest an association between acquired deafness with psychotic illness (late paraphrenia) (Eastwood, *et al*. 1985) and acquired blindness with visual hallucinations (Charles Bonnet syndrome) (Jacob *et al*. 2004) in elderly people. Similarly, people with acquired deafness or blindness may undergo a bereavement reaction. Problems acquired as a result of a sensory impairment in old age may be missed or attributed to depression or dementia, which can lead to isolation (du Feu & Fergusson, 2003).

2.4.6. Relationship between sensory impairment and mental illness in people with ID

A population-based study in Scotland (Cooper *et al*. 2007) found no independent association between mental illness and sensory impairment in people with ID. Other studies (Hindley & Kitson, 2000) reported an association between different disabilities such as ID, visual problems and central nervous system damage and development of psychiatric illness. Deaf and blind adults with ID are vulnerable to high prevalence of mental illness for a number of reasons including biological, psychological, social and developmental factors (Cooper, 2007). They may also be marginalised or become a victim of emotional, physical and sexual abuse (Hindley & Kitson, 2000). Challenging behaviour may also be common in deaf adults with ID. A community-based study found that 62% of deaf adults with ID living in the community displayed challenging behaviour (Timehin & Timehin, 2004). One population study of people with ID (both deaf and hearing) found that people who displayed challenging behaviour tended to have more restricted expressive and receptive communication (Emerson *et al.* 2001). Another study showed an increased rate of depression and self-injurious behaviours in deaf-blind adults with ID (Carvill & Marston, 2002). In 1959, Hallgren reported an association between deaf-blindness in Usher syndrome and psychosis in up to 23% of people. In this

study, most of the individuals with psychosis had ID. A follow-up study of young adults in a birth cohort who experienced in utero exposure to rubella in the 1964 rubella epidemic, found a 5-fold increase in non-affective psychosis (Brown *et al.*, 2000).

2.5. Assessment of sensory impairment

In spite of the detrimental impact of undiagnosed sensory impairment on the ability of people with ID to carry out their activities of daily living (Evenhuis *et al*. 2009), one study showed that 39% of people with ID received less eye care than those in the general population (Starling *et al*. 2006). Another study reported that 30% of people with ID and hearing impairment had never had their hearing tested (Timehin & Timehin, 2004). Deficits in communication in people with ID pose a significant challenge to the assessment of sensory impairment. In practice, diagnostic overshadowing may occur, whereby changes in behaviour and loss of skill may be attributed to ID, dementia or mental illness (e.g. depression) rather than to a sensory impairment. In such cases, the underlying health needs of an individual may not be addressed and treated appropriately and accurately (Lindsey, 2002). Carers may perceive a person to be non-co-operative when, in reality, they cannot hear or see properly; alternatively, some people will try to cover up their sensory loss which can be misinterpreted by carers, leading to statements such as “he can hear/see when he wants to” (Box 2.4) (Kiani & Miller, 2010).

McGlade *et al.* (2010) reported that on a quarter of their cases with ID, they needed three or more sessions to complete their optician assessment reflecting the complexity of the client group. Over 50% of their study population needed glasses for refractive error. It is therefore imperative that adults with ID have access to specialist sensory assessment in order to help with the identification of sensory impairment, which may go unrecognised by carers. Furthermore, it is vitally important to ensure that carers have training in blind and deaf awareness and are able to access appropriate aids (hearing aids or eye glasses) and environmental adaptations if needed. Training carers and staff can help them to identify sensory impairment, which can then be followed up by specialist

assessment. Specialist assessment can provide service users and carers with information on what a service user can and cannot see or hear; it also ensures that they have access to appropriate aids and services (Domokos, 2000; Pring, 2005; Miller & Kiani, 2008).

Box 2.4: Examples of carers' comments about unrecognised sensory impairment

- "He has taken to ignoring us."
- "He can talk so he is not deaf."
- "He can hear/see when he wants to."
- "He understands what we say."
- "She keeps breaking things."
- "She hits me with her white stick and won't use it anymore."
- "She won't go out."
- "We put the fluorescent light on in the kitchen then she starts breaking things."
- "She sees more than you think."

Yeates (1991, 1995 & 2000) found that, given access to specialist audiology services, 56% of adults with ID were able to complete a pure tone audiometery. She concluded that people with a deficit or lack of linguistic abilities should not be considered as unable to benefit from diagnosis of their hearing loss. She also emphasised that it would be ideal for adults with ID to have access to specialist audiology services, as generic audiology services are too busy to meet these service users' needs. Meuwese-Jongejeugd and colleagues (2005, 2007) set up a prospective study of a new audiological rehabilitation programme designed to meet the needs of people with ID with a recently diagnosed hearing loss. They implemented a detailed, well-thought and thorough multidisciplinary protocol for audiological rehabilitation in ID and audiological services. Even with all this in place, they were able to rehabilitate successfully only three of thirty-one adults with ID, because they found that screening adults with ID revealed a huge amount of unmet need for which services were not ready, and that rehabilitation required co-ordinated work between different agencies and professionals. They concluded that, for successful rehabilitation, there should

be changes in both. If it is decided that an individual needs to undergo specialist assessment, the process of desensitisation and preparation should be started as soon as possible. By law, hospitals are required to make reasonable adjustments for people with ID to access health services. This includes access to hearing and visual services. More information on this topic is provided on the Public Health England website (Public Health England, 2015). Boxes 2.5–2.7 provide further details on assessing individuals with sensory impairment. The following provision of information is extremely helpful if used well in advance to prepare service users for their appointment:

- Use of illustrated information leaflets, audio materials or information in Braille and other alternative communication strategies to take into account sensory impairment, before attending audiology or ophthalmology clinic;
- Use of text and audio messages;
- Information in DVD or CD format;
- Role play

History taking in acquired hearing loss, or when a hearing impairment deteriorates, includes determining: (i) whether the hearing loss is unilateral or bilateral, sudden onset or gradual, fluctuating, waxing and waning or progressive; (ii) whether it is associated with symptoms such as vertigo, tinnitus, otorrhoea, otalgia, head and neck lumps, swelling, pain, nasal block, epistaxis, itching, discharge; and (iii) its effect on quality of life, communication/work and family life.

Risk factors for acquired hearing loss include family history of otosclerosis, diabetes, autoimmune or vascular diseases, infections, noise exposure, trauma to ears, surgery and iatrogenic causes (ototoxic medications). Examination follows the history taking and includes examining external ears, wax impaction, tympanic membrane, head and neck and then cranial nerve examination (V, VII, VIII for facial weakness, abnormal sensation and taste, etc.). The whispered voice test is a very simple screening test and can be used at arm's length behind the service user, occluding the contra-lateral ear. Tuning fork (512 Hz)

test can also be used (Rinne and Webber tests) to distinguish between conductive hearing loss (CHL) and sensorineural hearing loss (SNHL). None of these tests are 100% reliable and, therefore, necessary referrals, either routinely or urgently, should be sent to the ear, nose and throat (ENT) or audiology services (Edmiston & Mitchell, 2013). Same principles could be adopted when assessing a visual impairment (Box 2.5). The invisible nature of some of these problems often complicates investigation in people with ID. For example, in this population there is also a high prevalence of central (cortical) auditory processing disorders, which are difficult to diagnose in a person with an otherwise normal ear anatomy and structure (Neumann *et al*, 2007).

People with central auditory processing problems may not be recognised as having hearing difficulties because they do not have trouble detecting sound or recognising speech in ideal listening situations. Since they appear to 'hear normally', the difficulties these individuals experience are often assumed to be the result of an attention deficit, a behaviour problem, lack of motivation or depression. In the aforementioned study on Special Olympic athletes, 20 of those who received tests had previously undetected central processing disorders; there were disturbances at the cortical level in all and in a considerable proportion of the subjects at the brainstem level (Neumann *et al*, 2007). For more information please refer to a review article by Hitoglou *et al*. 2010.

Other examples are tinnitus or visual agnosia, which have been acquired later in life due to a variety of reasons. Adding to this complexity is the knowledge that the organ affected might be perfect in observation or examination, such as in someone with prosopagnosia whose main problem is in recognising faces. This can make it difficult for others to understand and may affect their attitude towards the individual as such issues are difficult to explain, even by people without ID. Raising the awareness of the condition and accessing specialist input including neuropsychological approaches are therefore paramount (Farah, 2004; Basu, 2012).

Box 2.5: Points to consider when taking a service user's history to assess sensory impairment*

- Family history of sensory deficits and genetic disorders.
- History of kernicterus, peri-natal asphyxia and in utero exposure to TORCHES (Toxoplasmosis, Other infections, Rubella, CMV, Herpes Simplex and Syphilis).
- Childhood history of meningo-encephalitis.
- Developmental milestones.
- Any recent change in behavior.
- Previous injury to ears/eyes.
- Discharge, itchiness and pain in sensory organs.
- Problems with earwax.
- Dizziness, vertigo, loss of balance and tinnitus.
- Double vision or blurred vision.
- Previous assessments and provision of hearing aids or eyeglasses.
- Past operations on sensory organs.
- Past or current use of medication affecting sensory organs (e.g. aminoglycosides).
- Motor abnormalities.

*Taken from Firth *et al.* 2005; CMV: Cytomegalovirus

Box 2.6: Assessment of visual impairment*

Examples of functional visual assessment

- External appearance of the eyes (coloboma, squint).
- Abnormal eye movements.
- Watching from angle of eyes.
- Head tilting.
- Finger flicking in front of the eyes.
- Eye poking.
- Bringing objects very close to eyes.
- Not recognising familiar faces.
- Groping to find things.
- Preference for bright objects.
- Bumping into things.
- Difficulty using steps.
- Not looking confident when walking.

Examples of specialist assessment

- Visual acuity tests*.
- Visual field tests.
- Ophthalmoscopy.
- Contrast sensitivity tests.
- Binocular vision tests.

*During examination of visual acuity, Kay pictures (http://www.kaypictures.co.uk/) and the Cardiff Acuity Test (preferential looking pictures) can be used instead of the Snellen chart: http://www.cardiff.ac.uk/optom/eyeclinic/downssyndromegroup/thecardiffacuitytest.html

Box 2.7: Assessment of hearing impairment*

Examples of functional hearing assessment

- Size and shape of the ears (absent or very small ears).
- Talking unusually loudly or in a whisper.
- Not taking notice of prolonged or loud noises such as fire alarms.
- Startled by people approaching who are not in sight.
- Liking TV/radio on louder than normal.
- Responding only to certain voices (inconsistent in response).
- Misunderstanding instructions.
- Covering, poking, slapping ears.
- Experimenting with noises.
- Getting close to sounds.

Examples of specialist assessment

- Otoscopy.
- Pure tone audiometry.
- Warble tone audiometer.
- McCormick Toy Discrimination Test.
- South London Object Test.
- Tympanometry.
- Otoacoustic emission.
- Brainstem evoked response.

*Taken from Hindley & Kitson (2000); Austen & Crocker (2004).

It is important to note that stereotypical movements (e.g. flicking fingers) and fascination with or avoidance of certain stimuli (sensory seeking and avoiding behaviours) have been commonly described in people with ASD. Care should

therefore be taken not to confuse these with symptoms of visual or hearing impairment in people with ID.

In addition, a number of sensory assessment tools can be used for the assessment of sensory perceptual difficulties in autistic individuals, such as the Adult Sensory Questionnaire (ASQ) (Kinnealey & Oliver, 2002) and Sensory Profile Checklist (Bogdashina, 2003). Ideally a referral should be sent to a colleague from the occupational therapy department for a sensory integration assessment (please see below for more detail on sensory integration).

Health guidelines, published by the International Association for the Scientific Study of ID (IASSID, 2002) recommend that specialist screening for age-related visual and hearing loss in people with ID should be started at the age of 45 years and repeated every 5 years thereafter. People with Down syndrome should also have a one-off visual screening assessment at the age of 30 years and their hearing should be monitored more frequently at 3 year intervals. For more details on the assessment of sensory impairment in people with ID, refer to Northfield (2008) and the Health Guidelines published by the IASSID (2002).

2.6. Management strategies for people with sensory impairment and ID

When sensory impairment and ID are present together from early childhood, their psychosocial and cognitive effects are much greater than the sum of each, as they not only interact with each other, but can also be accompanied by other disabilities (e.g. mental illness, CP, challenging behaviour and epilepsy) (Carvill, 2001; Butler, 2002).

It has been argued that better management of service users with multi-morbidities requires putting greater emphasis on: (i) clinical judgement when assessing service users and their carers' needs to manage more than one problem at a time; and (ii) coordination of care that can promote good therapeutic relationship between the professionals and the service user. In this model, it is important that a generalist's skills are used by the specialist team for the better management of problems in the community (Rolan & Paddison,

2013). Effective management in people with ID is difficult as medical treatments form only one component of the management strategy (Table 2.8) (Kiani & Miller, 2010). Management requires an approach that is not simply multi-modal and multi-disciplinary, but also multi-agency, combining education, social services and other agencies such as charity organisations to ensure a holistic response to service users' difficulties. Intensive long-term intervention with different components is the best strategy to address service users' complex needs and training the carers and staff in sensory impairment (Domokos, 2000) is a key component to ensure successful long-term management.

People with ID are able to express their wishes and ideas regarding hearing aids, provided that they are given sufficient information tailored to their cognitive abilities. Several elements (e.g. benefit, cosmetics, sound quality/acoustics, and comfort/ease of use and service delivery) may play a role in satisfaction with hearing aids (Austen & Crocker, 2004; Meuwese-Jongejeugd *et al*. 2005 & 2007), but research shows that while 70% of the service users had been seen by audiology services at some time in their life, only 24% had ongoing assessments and hearing-aid maintenance (Timehin & Timehin, 2004). The study also showed that only 20% of them wore their hearing aids regularly and that only 2% of care home staff had received training on maintaining hearing aids despite the positive effect of hearing aids on communication and behavior.

Boxes 2.9 and 2.10 show the reasons for and ways to improve non-compliance with wearing hearing aids and glasses (Hindley & Kitson, 2000; Austen & Crocker, 2004).

Table 2.8: Management strategies for people with sensory impairment and ID

Medical	Environmental	Psychosocial	Communication	Other
Medication	Appropriate lighting and contrast colouring	Social skill training (e.g. social stories)	PECS†	TEACCH†
Eye glasses	Consistency of rooms	Psycho-education	Braille and Moon	Sensory integration
Hearing aids	Uncluttered rooms Railing for stair or driveways (outside the house) Adapted bathroom	One-to-one support from a support worker conversant in sensory impairment	Sign language and Makaton	Irlen tinted Glasses (for SS*)
Surgery	Light or vibrating alarms and clocks Pagers for doorbells and calendar boxes Use of special carpet and audio, touch and smell orientation Good signage, big calendar and clock and use of magnifying lenses, loop system	Commissioning structured day time and leisure activities Access to sensory room	Deaf-blind manual and block alphabet Objects of reference	ABA Intensive interaction

ABA: Applied Behavioural Analysis (Lovaas, 1987); PECS: Picture Exchange Communication System; SS: Scotopic Sensitivity or Irlen syndrome; TEACCH: Treatment and Education of Autistic and related Communication handicapped Children.

*A recent article by Williams (2014) questioned the validity of Irlen syndrome and prescription of filtered lenses.

†PECS and TEACCH can be adapted for people with visual impairment by replacing pictures with tactile symbols and objects of reference (Lund & Troha, 2008; Taylor & Preece, 2010).

Box 2.9: Reasons* for non-compliance with hearing aids or eyeglasses

- Badly fitting or painful device.
- Broken or lost device.
- Poorly maintained device.
- Ineffectual device owing to wrong assessment/diagnosis.
- Feeling stigmatised.
- Teasing by others.

*Taken from Hindley & Kitson (2000); Austen & Crocker (2004).

Box 2.10: Ways* to improve compliance with hearing aids and eyeglasses

- Allow clients to choose their preferred model and colour.
- Gradually extend usage from one setting to another.
- Give the client the responsibility for using and cleaning them.
- Positive reinforcement and lots of praise.
- Integrate devices into everyday life.
- Establish a routine around them.
- Practice role-play and modelling.

*Taken from Hindley & Kitson (2000); Austen & Crocker (2004).

Communication with people with ID who have sensory impairment is challenging and complex. Staff working in such settings must receive regular training and be given the opportunity to practice, in order to be able to communicate effectively with service users (Table 2.11).

Table 2.11: 'Dos' and 'Do nots' when communicating with people with sensory impairment

Do:	Do not:
Always tell them where you are, and where you are going to go.	Assume the person is totally blind and deaf. They may have some residual visual or hearing ability.
Consider supplementing verbal communication with simultaneous signs and symbols.	Shout or speak very loudly unless you are asked to do so.
Facilitate lip reading by allowing them to see your mouth clearly.	Misinterpret head tilt for extra-pyramidal symptoms; they may be using their better ear to listen to you!
Encourage those with macular degeneration to look at objects through the angle of their eyes and teach them to use magnifying lenses.	Assume the service user lacks eye contact (e.g. as in autism and Fragile X syndrome); they may be looking slightly off-axis due to loss of central vision.
Respect confidentiality by not talking too loudly.	Mumble or exaggerate your lip movements

Psychological approaches (e.g. psycho-education), tailored to the individual service user's level of language and cognitive ability, are important to help clients make sense of the social world around them. These can be complemented with social skills training (e.g. social stories) to improve the service user's understanding of other people's emotions and minds as well as their own.

There are also several management strategies which can be implemented by the occupational therapists. One of the most famous of such strategies is the sensory integration approach, which helps service users to be able to use their body effectively in the environment (Ayres, 1972). A study on sensory processing in ASD has shown that there are global abnormalities in the five main sensory modalities (tactile, gustatory, olfactory, visual and auditory) and that these seem to be inter-related (Kern, 2007b). Some biographical accounts of people with ASD show that sensory integration is helping service users

integrate their senses so that they can use more than one sense at a time. For some people with severe ASD and ID, intensive interaction has also been helpful in establishing attention and emotional engagement (Caldwell & Horwood, 2008).

Sensory integration interventions provide an opportunity for more than one sensory experience along with challenging activities, which is enjoyable and motivating, through building a trusting relationship with the service user and arranging an appropriate and safe environment that is conducive to optimal level of arousal (Parham *et al*. 2007).

Treatment and education of autistic and related communication handicapped children (TEACCH) can reduce anxiety by providing a structured and predictable daily timetable for different activities (Schopler & Mesibov, 1995). TEACCH and the Picture Exchange Communication System (PECS) can be adapted for people with visual impairment by replacing pictures with objects of reference (Lund & Troha, 2008). Objects of reference are taught by pairing an event with an object to facilitate the learning process, for example handing a cup to a deaf-blind service user when it is time for a drink.

It has been shown that it is extremely important to keep carers aware and provide advice to them to improve the quality of low vision rehabilitation for people with visual impairment (Sjoukes *et al*. 2010). Those who are experts in the field of visual impairment advise mothers of blind children to use other senses and activities such as touch, feel, smell, sound, swinging and tickling to make up for the lack of eye contact between themselves and their children so that blind babies could be engaged in joint attention activities with their mothers (Pring, 2005; McLinden, 2012). It is also important that the blind person's learning partner takes on a greater role in ensuring that haptic (tactile) information is mediated to meet the person's needs though exploratory strategies, given that limited information is available through visual modality. These exploratory haptic procedures could be classified in different ranges, e.g. lateral motion, pressure, static contact, enclosure, unsupported holding and contour following (McLinden, 2012). The learning partner has the responsibility

of employing a range of prompts to facilitate engagement and participation of the person in structured activities. The partner can use hand over hand support along with verbal commentary to usher the learning session which, over time, can result in the blind person becoming more independent in carrying out the activities (McLinden, 2012).

Most people with ID who are deaf use a very simple version of sign language. Some might know Makaton or Signalong, which have basic sign language vocabulary and structure. PECS can also be used to facilitate service users' autonomy by showing pictures of the items they need. For people who are blind, communication can be via Braille, Moon, objects of reference or audio materials. For those who are deaf-blind, it is essential to communicate through the deaf-blind manual, hands-on signing, visual frame signing or block alphabet. A person with dual sensory loss experiences a greater degree of impairment than the sum of the visual impairment and hearing impairment alone. Dual sensory loss is a major risk factor for falls and injuries; therefore, the environment should be adapted in a way that those with a dual sensory loss can orientate themselves through touch and smell (Butler, 2004).

For those service users with a diagnosis of ASD, the environment must be autism friendly, so that it appears less distracting and at the same time safe, uncluttered and easy to use. Each room should serve a clear function and unnecessary noises should be eliminated. Use of remaining eyesight should be encouraged by reducing the glare and providing appropriate lighting and contrasting colours (Butler, 2004). An occupational therapist or charity organisation (Box 2.12) could be consulted for the environmental adaptation of a daycare centre or the home. The Royal College of Ophthalmologists has created a Best Practice Guide for GPs on visual impairment in people with ID, which was launched in June 2012, highlighting key areas to ensure this group of service users has access to appropriate eye health care. VISION 2020 UK also facilitates collaboration and co-operation between organisations that focus on visual impairment (Vision 2020 UK, 2012). There are also organisations that provide further information on hearing impairment and visual agnosia (Box 2.13).

Box 2.12: Charity organisations for people with sensory impairment

Look Up	www.lookupinfo.org
SeeAbility*	www.seeability.org.uk
Deafblind UK	www.deafblind.org.uk
VISTA	www.vistablind.org.uk
Sense	www.sense.org.uk
Royal National Institute of Blind People†	www.rnib.org.uk
Royal National Institute for Deaf People‡	www.rnid.org.uk

*Seeability is a web based charity organisation providing training and consultancy services for people with ID and visual impairment. It provides accessible information for patients and those who are involved with their care (www.seabaility.org).

†RNIB Technology support squad can help clients to regain their independence through provision of support and use of new technology (www.rnib.org.uk/techsupport). Action for blind people (part of the RNIB group) (www.actionforblindpeople.org.uk) provides advice and support in dealing with employment issues, financial entitlement, social activities, events and sport opportunities. There is also an Eye Clinic Liaison Officer (ECLO) service, which help people access their appointments in hospitals.

‡RNID has now changed to Action on Hearing Loss.

The National Autistic Society also provides information on the association of hearing and visual impairment with ASD:

http://www.autism.org.uk/about-autism/related-conditions/visual-impairment-and-autism-spectrum-disorders.aspx

http://www.autism.org.uk/about-autism/related-conditions/asds-and-hearing-impairments.aspx

Box 2.13: Useful websites on sensory impairment

Useful websites with information about hearing impairment	
Rotherham Primary Ear Care Centre and audiology services	www.earcarecentre.com
Deafness Research UK*	www.deafnessresearch.org.uk
Patient.co.uk	www.patient.co.uk/doctor/Deafness-in-Adults.htm www.patient.co.uk/doctor/Hearing-Tests.htm
BMJ Learning	http://learning.bmj.com/learning/module-intro/hearing-loss-and-tinnitus-in-adults--a-guide-for-gps-.html?moduleId=10029379
Websites with information about visual agnosia	
National Portage Association (a home-visiting education service for preschool children with additional support for service users and families)	www.portage.org.uk
Websites with information about face blindness	
National Institute of Neurological Disorders and Stroke	www.ninds.nih.gov/disorders/prosopagnosia/prosopagnosia.htm
Faceblind.org	www.faceblind.org

*Now merged with Action on Hearing Loss

3. AIMS AND OBJECTIVES OF THE THESIS

3.1. Aims and Objectives

The primary aim of this thesis was to explore the relationship between ASD and blindness and deafness in adults with ID, controlling for the nature and severity of brain damage and gender. The secondary aim was to determine the prevalence of sensory impairment, ASD and other co-morbid medical and mental health problems in adults with ID.

The thesis topic was chosen because of the higher prevalence of ASD found in blind and deaf children that many researchers attribute to the confounding effects of underlying brain damage. Although a number of studies and case series have attempted to disentangle this association by researching those with brain damage, the debate still continues. In addition, the majority of studies highlighted in the previous chapter (literature review) were carried out in children. Therefore, there was a need to fill the gap in literature by carrying out a study that was able to explore the relationship between blindness/deafness and ASD in adult service users, but that also controls for the nature and severity of brain damage.

The objectives were:

1) To conduct a literature review on the prevalence of sensory impairment and its association with ASD, challenging behaviours and mental ill-health in the general population and in individuals with ID.

2) To conduct a cross-sectional study on a representative population of adults with ID living in Leicester city, Leicestershire and Rutland, UK, and to use this to:
   a) estimate the prevalence of sensory impairment, ASD and co-morbid physical and mental health problems;
   b) explore the relationship between ASD and sensory impairments, adjusting for potential confounders;

c) explore the relationship between individual autistic traits and sensory impairment, adjusting for potential confounders;
d) explore the relationship between challenging behaviour and sensory impairment, adjusting for potential confounders.

3) To identify a subgroup of adults with congenital deafness and a randomly selected group of adults without sensory impairment matched by degree of ID and gender using objective assessment tools and to use this cohort to:
   a) describe the characteristics of people with congenital deafness;
   b) compare differences in diagnostic methods used to identify ASD;
   c) explore the relationship between ASD and congenital deafness, adjusting for potential confounders.

4) To identify a subgroup of adults with congenital blindness and a randomly selected group of adults without sensory impairment matched by degree of ID and gender using objective assessment tools and to use this cohort to:
   a) describe the characteristics of people with congenital blindness;
   b) compare differences in diagnostic methods used to identify ASD;
   c) explore the relationship between ASD and congenital blindness, adjusting for potential confounders.

3.2. Research Questions

This thesis aimed to address the following research questions:

1) What is the prevalence of sensory impairment, ASD and comorbid physical and mental health problems among adults with ID?
2) What is the relationship between ASD and sensory impairment after adjusting for potential confounders?
3) What is the relationship between individual autistic traits and sensory impairment after adjusting for potential confounders?

4) What is the relationship between challenging behaviour and sensory impairment after adjusting for potential confounders?
5) What is the aetiology of ID in blind and deaf cases?
6) What are the differences in diagnostic methods used to identify ASD in the blind and deaf subgroups?
7) What is the relationship between ASD and congenital deafness after adjusting for potential confounders?
8) What is the relationship between ASD and congenital blindness after adjusting for potential confounders?

4. METHODOLOGY

In order to achieve the aims of this thesis, a two-stage cross-sectional comparative study was carried out to explore the association between sensory impairment and ASD after adjusting for confounders (degree of ID and age).

4.1. Study population

Both stages of the project involved utilising routine data from a case register of adults with ID called the Leicestershire Learning Disability Register (LLDR) (McGrother *et al*. 1993). Additional information was collected from the Leicestershire adult ID service if the data were missing or incomplete in the LLDR database. The register contains details of adults with ID living in the unitary authorities of Leicester city, Leicestershire and Rutland who receive support from a network of specialist health and social care providers. Adults on the register make up about 4.9 per 1000 population of this geographical location (approximate total adult population size: 0.7 million [National Statistics, 2001]). The register contains a representative population of adults with moderate to profound ID who are seen and notified by specialist service providers and a proportion of adults with mild ID who have more complex needs and require support in managing on a day-to-day basis, such as adults with challenging behaviour, sensory impairment and severe co-morbidities. Detailed home interviews with carers of adults with ID on the register are carried out every 5–7 years, for which the acceptance rate is 90%.

Appendix 3 shows the detailed questionnaire used by the LLDR information officers when interviewing service users and their carers. At the outset of this work, the register held data on 3,138 adults (aged 18 years and over) with ID who (or whose carers/family members) had provided written consent to be approached and for their data to be used for research purposes (Watson, 2002).

Information collected by the LLDR includes demographics, skill level, adaptive behaviour, social functioning, behaviour problems, psychological symptoms and carers' health. In 2008, Tyrer and colleagues developed a proxy measure for severity of ID, the Leicestershire ID (LID) tool, which displays a sensitivity of 95% and a specificity of 65% in identifying adults with moderate to profound ID compared with the recognised standard of Vineland developmental age (Sparrow *et al.* 1984). However, the authors advised exercising caution when using the tool to assess people with substantial physical disability, acquired cognitive impairment such as dementia, and people with ASD who tend to perform worse in adaptive behaviour assessments (Carpentieri & Morgan, 1996).

Case registers, such as the LLDR, are believed to provide reasonably accurate estimates of the underlying prevalence of moderate to profound ID within countries with well-developed education and welfare systems (ten Horn *et al*. 1986).

Information on autistic traits on the LLDR database is derived from the Disability Assessment Schedule (DAS) (Holmes *et al.* 1982) and there are additional questions on support and care need developed for use in this client group (McConkey & Walsh, 1982). Historically, in Leicestershire, information on autistic traits have been collected based on five items derived from the DAS (Box 4.1) that act as a proxy measure for diagnosing ASD in the absence of other indicators (Bhaumik *et al*. 1997). The five traits comprise impairments or deficit in speech, social skills and empathy and presence of elaborate routines and stereotypies. Previous research suggests that these traits occur in approximately half of adults who use specialist ID services (Bhaumik *et al*. 2010).

For stage 1 of the research project, a diagnosis of ASD was considered if an individual had 4 or more of the autistic traits outlined in Box 4.1. For this thesis, a person was considered to have an autistic trait if it was present in any form, no matter how minor. For further details please refer to question items Q3.28 to Q3.32 in Appendix 3 (The LLDR interview schedule). Having no trait or just one

trait ruled out a diagnosis of ASD. Having 2–3 traits was regarded as having autistic traits, but not having a diagnosis of ASD. This process was agreed in advance between the researcher and his two supervisors. Previous research (Bhaumik *et al*. 2010) had shown that some of these traits were commonly observed in the ID population without a diagnosis of ASD, and therefore having some of the traits did not necessarily mean that an individual had ASD. In addition, people with autistic traits but no ASD were more likely to have Down syndrome, CP and mobility problems and it was debated that clinicians might have attributed some of the autistic traits to their underlying condition rather than to a diagnosis of ASD.

Making an accurate diagnosis of ASD in individuals with severe to profound ID can be challenging as autistic-like symptoms can be present without a diagnosis of ASD. For example, impairments in communication and socialisation might be indicative of a low IQ, congenital sensory impairment and problems with developmental adaptive skills, rather than ASD. Individuals may also have limited behavioural and language repertoires which makes diagnosis of ASD very difficult. Therefore, a proxy diagnosis of ASD was set at the higher threshold of 4 or more autistic traits in stage 1 of this project to avoid false positives and type I error.

Box 4.1: Autistic traits on the Leicestershire Learning Disability Register*

Minimal speech

- Unable to ask for things s/he wants or talk about things s/he has done.†
- Uses a few words or signs (e.g. hello, bye-bye, drink).
- Uses words or signs for practical needs – variety of needs.
- Uses words or signs to comment on his/her own personal experience (e.g. tells people s/he has new clothes, that s/he has been on an outing, that someone has done something wrong etc.).

Poor quality of social interaction

- Does not interact – mainly aloof, indifferent or bizarre.†
- Interacts to obtain needs only – otherwise indifferent.†
- Responds to and may initiate physical contact.
- Does not initiate social contact, but responds passively if other people make approaches.
- 'Unwarm' – does make social approaches, but these are peculiar, naïve or even bizarre. The person does not modify behaviour in light of these responses, needs or interests of those whom s/he approaches. The interaction is one-sided and dominated by the person being rated. †
- Some warm qualities in addition to the above.

Lack of empathy

- No or limited empathy†

Simple stereotypies

- Marked repetitive activities (e.g. rocking, flicking fingers etc.), especially when unoccupied, although may be controlled by close supervision or being kept fully occupied – often a constant feature, present each day. †
- Present, but minor aspect of behaviour pattern.

Elaborate routines/Obsessional behaviour

- Has elaborate routines of the kind and intensity found in early childhood autism. †
- Has minor routines, or obsessional behaviour, such as hand washing.

*Taken from the Disability Assessment Schedule (Holmes *et al.* 1982) – see question items Q3.28 to Q3.32 in Appendix 3 (The LLDR interview schedule). Those marked with † were those traits included in the articles on autistic traits by Bhaumik et al. 1997 and 2010. A wider threshold has been chosen for this thesis.

4.1.1. Case ascertainment of ID service users with a sensory impairment

A preliminary investigation of the data available on the LLDR and the local adult ID service showed that there were 51 adults with severe to profound deafness and 175 adults with partial deafness. Similarly, 114 individuals had profound blindness and 272 individuals had partial blindness.

Twenty-two people with deaf-blindness were identified, of whom 10 had partial deaf-blindness and those remaining had total deaf-blindness (n=12). Half of the latter group had congenitally born deaf-blindness (n=6) and half had acquired deaf-blindness (n=6).

In stage 1 of the project all adults with a sensory impairment (n=612) who had given consent that their LLDR data could be looked at for research purposes, regardless of whether it was congenital or acquired, unilateral or bilateral, partial or total, were included. Additional information was collected from the Leicestershire adult ID service if the data were missing or incomplete in the LLDR database.

Stage 2 of the project involved investigating 2 subgroups of adults from stage 1; all adults with congenital and total visual impairment and a random sample of 'controls' and all adults with congenital and total hearing impairment and a random sample of 'controls'. All controls were matched with 'cases' on degree of ID and gender. Therefore, each subgroup comprised cases with congenital and bilateral blindness or deafness and their controls. For both subgroups the relationship between sensory impairment and ASD was investigated.

Despite the LLDR and local adult ID service databases providing useful data, it was important to exclude those with borderline IQ, unilateral sensory impairment, deaf-blindness and partial (with residual sight or hearing) and acquired sensory impairment for stage 2 to fully explore the association between a pre-lingual sensory impairment and a diagnosis of ASD. Stage 2 of the project, therefore, involved assessing all service users diagnosed with a co-morbid sensory impairment on the LLDR or the local adult ID service databases using face-to-face interviews and objective assessment tools to determine the

exact nature and degree of their sensory impairment and ID. Medical case files and electronic data records of the service users open to the local adult ID service were also investigated to obtain further information for the project. When a service user had input from other members of the multidisciplinary team such as a speech and language therapist or an occupational therapist, these information were also looked into to ensure detailed information is collected for each service user.

In stage 2, adults with a profound and irreversible absence of sight bilaterally (without form vision) that had occurred congenitally were included. These visual impairments could not be corrected with spectacles, contact lenses, medication or other procedures such as surgical interventions. Using this definition of blindness, light vision may or may not be present. Similarly, only those with deafness were included if they had a severe to profound bilateral hearing impairment (a loss of equal to or above 75 dB) occurring congenitally which could not be corrected using hearing aids, surgical procedures or other interventions such as ear drops or ear wax removal.

Data on two subgroups in the stage 2 of the research project were analysed, each containing data on adults with ID with and without sensory impairment. The first subgroup comprised adults with congenitally and bilaterally total visual impairment (without accompanying hearing impairment), the 'cases', and adults with normal vision and hearing, the 'controls'. The second subgroup comprised adults with congenitally and bilaterally total hearing impairment (without accompanying visual impairment), the 'cases', and again adults with normal vision and hearing, the 'controls'. The cases were mutually exclusive because adults with deaf-blindness (n=22) were excluded (see later); however, controls could occur in both subgroups.

For the cases, those with unilateral sensory impairment (one-sided deafness: n=1 or one-sided blindness: n=6 – with no deafness or blindness on the other side), those with no ID (n=9 blind adults and n=9 deaf adults with borderline IQ= IQ>70) and those with acquired sensory impairment (n=24 adults with acquired

blindness and n=11 adults with acquired deafness; occurred later in life) were excluded from the final analysis.

The control groups were randomly selected from those adults (aged 18 years or over) with an ID and both normal hearing and vision who were living in the geographical locations of Leicester City, Leicestershire and Rutland. Controls were matched with the cases by gender and degree of ID (mild, moderate, severe and profound ID groups) using SAS (Kawabata *et al.* 2004) in a way that the first person in the list with blindness or deafness was matched with the first person on the control list based on the degree of ID and gender and so on (Appendix 14). Matching was done to account for confounders. Equal numbers of control group from various age categories (18–29, 30–39, 40–49 and ≥50 years old) were randomised for each age category of cases.

In addition, deaf-blind people (n=22) were excluded since these were a very heterogeneous group of service users, primarily with partial deaf-blindness (n=10) or acquired deaf-blindness (n=6). Those with congenital severe to profound deaf-blindness were in the minority (n=6) and therefore were not included in the final analyses.

Appendix 4 shows the characteristics of service users who were excluded from the study.

4.2. Power calculation for stage 2

In order to find a difference of 15% in the prevalence of ASD between individuals with and without sensory impairment with 80% power and 5% level of significance (2-tailed), a sample size of 60 people in each group was needed. This calculation was based on having 20% discordant pairs. To achieve the samples determined by the power calculation, all service users with sensory impairment who were eligible for stage 2 as per inclusion criteria described above were included to compensate for the attrition rate, considered to be as high as 30% (i.e. individuals who later refused to participate, moved out of the county or to an unidentified address or died).

All cases with visual impairment were therefore included if they had a certificate of visual Impairment (CVI) from the ophthalmology department and were registered as blind (with or without light perception but without form vision) with VISTA, the local charity organisation supporting people with visual impairment (n=75). Equal numbers for the control group from various age categories were randomly selected from the LLDR database, matched with the cases by degree of ID (mild, moderate, severe and profound ID categories) and gender (n=75).

Similarly for the subgroup with deafness, all individuals with a report from Ear, Nose and Throat or hearing services showing congenitally severe to profound deafness (a hearing loss of ≥75 dB) were included (n=30). Equal numbers for the control group from various age categories were randomly selected from the LLDR database, matched by degree of ID and gender (n=30).

4.3. Taking informed consent

Information about the project was presented to the multidisciplinary ID teams working in different catchment areas of Leicester City, Leicestershire and Rutland. At the same time, information leaflets were given to the teams to distribute among service users and their carers in a number of settings, including private residences, residential homes, supported living placements, day centres, short break/respite facilities and inpatient settings. The project was also discussed with colleagues at social services and charity organisations, including Vista, RNIB, and action on hearing loss (formerly RNID).

Details about service users with sensory impairment, including names and addresses, were gathered from the LLDR and the local adult ID service databases. The researcher then wrote to the service users and their carers/families to take informed written consent to proceed with interview and completion of assessment tools. All service users and their carers/families received an invitation/information letter and a consent form (Appendices 5–8). To facilitate the process of taking consent, specifically designed accessible information leaflets and consent forms (illustrated), which provided pictorial

information in simple language tailored for use by people with ID were also used (Appendix 6.2).

The information letter was later translated to Braille for blind service users who had mild ID (Appendix 5), with the help from the RNIB staff based at Stan Bell College at Loughborough. For services users who were deaf, accessible pictorial information was used and, if they were able to sign or use Makaton, it was explained to them by the researcher, their carers or an independent interpreter in simple sign language.

Individuals who did not send the consent form (Appendix 8) back within one month were sent another letter of invitation and a reminder. A telephone contact was then made for those who did not respond after sending 2 invitation letters. When an address was deemed incorrect after following the above steps, attempts were made to find the service user's new address by contacting the GP surgeries, with which they were originally registered, and social services. Only those who provided written informed consent could be assessed in the 2nd stage of the project. In the majority of cases, the participant lacked capacity to consent and the carers/family members were consulted and signed a form to say that the participant could take part in the study, in accordance with mental capacity guidance (DH Scientific Development and Bioethics Division, 2008). A proportion of service users were able to provide informed consent and sign the form with support from their carers. For those with severe to profound ID who were not able to give consent, signatures were obtained from carers and next of kin. In such cases, a common sense approach was adopted, whereby the assessment was only carried out if the service user appeared to be calm and relaxed during the assessment and did not withdraw or show any sign of discomfort and unhappiness during the process.

In addition, an information letter was sent to the service user's general practitioner and written informed consent was requested from the responsible ID consultants for the care of the patients in the community (if open to one) so that they were aware of the study and the consequences of taking part in the assessment (Appendices 7.1 & 7.2).

4.4. Interviews and objective assessment tools

For stage 2 of the thesis, face-to-face interviews were carried out alongside collection of data from clinical case files, which allowed time for more in-depth objective assessments to be conducted. To test the main hypothesis of the research project, only data gathered from face to face interview using an objective assessment tool for identifying ASD (PDD-MRS, please look below for more detail) were used. The main interview was conducted with the service users and their families, carers or relevant key workers, but all of the service users were also observed in their familiar setting and examined. This included a basic examination such as looking for abnormal appearance of sensory organs and carrying out the whispered voice test, eye movement exam, etc., provided they agreed to this and did not withdraw.

All interviews were carried out by the researcher. The following tools were used:

- A sensory impairment proforma (Appendix 9), designed by the researcher, to collect as much as information as possible on demographic characteristics and other variables, such as aetiology of ID (if known), presence of genetic syndrome, and co-morbid physical problems (e.g. mobility problems, CP, incontinence and epilepsy) and mental illness. Additional information on whether a patient had mental or physical illness was collected from the psychiatric case file and the summary information sheet provided by their primary care physician which contained information on various co-morbidities and prescribed medication.

- Screening tools for sensory impairment: a visual and a hearing impairment screening checklist, devised by the speech and language therapy department at the local adult ID service, were sent to all cases and controls prior to the interview session to ensure that no accompanying sensory impairment was missed (Appendices 10 & 11). All service users were also briefly examined by the researcher, a consultant ID psychiatrist. Those who were found to have additional

health or social concerns were referred to relevant health and social care professionals for further assessment and management (please see the section 5.2, addressing the unmet needs, for more details).

- The Pervasive Developmental Disorder - Mental Retardation Scale (PDD-MRS) (Kraijer & Bildt, 2005; Kraijer, 2007) was used to determine the presence of ASD for the hypothesis testing. PDD-MRS (Appendix 12) has been successfully standardised and validated for use in adults with all degrees of ID, most notably for those with severe/profound ID and those with additional conditions and disabilities, where the administration of detailed and longer diagnostic tools, such as the Diagnostic Interview for Social and Communication Disorders; DISCO (Wing *et al*. 2002) and Autism Diagnostic Interview - Revised; ADI-R (Lord *et al*. 1994), is clinically impractical owing to time constraints. It can be completed through a combination of observation of the client's current interaction, behaviour and communication and collection of collateral information from a carer/family member. The PPD-MRS consists of 4 categories; social interaction with adults (one item with three stems; a maximum score of six); social interaction with peers (one item with two stems; a maximum score of four); language and speech (three items, with a maximum score of four); and other behaviours (seven items, with a maximum score of ten). It can be administered in around 45 minutes and is supported by a concise manual informing the professionals about various aspects of the scale, including directions for use and scoring. Based on the final score, the PDD-MRS ascertains whether a service user has ASD (score=10–19), doubtful ASD (with some autistic traits but not qualifying for a diagnosis of ASD) (score=7–9) or no ASD (score=0–6). For the purpose of the current study, people with a score of doubtful ASD were grouped with those who did not have ASD and only those who scored 10 or above were considered as having ASD. This was mainly done to avoid false positive outcome and type I error as autistic traits have been commonly reported in people with ID and sensory impairment (please look at chapter 2; literature review for more detail).

PDD-MRS has a sensitivity of 94.4%, specificity of 92.7%, α coefficient of 0.8 and inter-rater reliability of 0.83 (Krajer, 2007) for detecting ASD. The questionnaire was devised from the Diagnostic Manual of Mental Disorders (DSM) (American Psychiatric Association, 1994) item checklist for diagnosis of ASD. PDD-MRS has been shown to be successful in identifying previously undiagnosed ASD in the ID population (LaMalfa *et al*. 2004). LaMalfa and colleagues (2004) reported a diagnosis of ASD in 39.2% of their studied population after using PDD-MRS, whereas, prior to the screening only 7.8% had a confirmed diagnosis of ASD. A study in the West Midlands (Morgan *et al.* 2002) using PDD-MRS found a 30% prevalence of ASD in a community sample of 571 adults with ID.

- The Aberrant Behaviour Checklist (ABC) was used to identify challenging behaviours (Aman *et al*. 1985). The ABC is a symptom checklist, which has high reliability and validity for assessing challenging behaviours in adults with ID. The ABC was developed using factor analysis on data from 1,000 persons with ID. It has five subscales: (i) irritability and agitation; (ii) lethargy and social withdrawal; (iii) stereotypical behaviour; (iv) hyperactivity and non-compliance; and (v) inappropriate speech. Each subscale consists of several items and overall there are 58 items. Each item checklist can be scored according to the severity of the behaviour for the last 4 weeks (0= no problem, 1= slight problem, 2= moderately serious problem, 3= severe problem). For the purpose of the statistical analysis, only the final data on the presence or absence of challenging behaviours were used i.e. challenging behaviour variable was dichotomised according to whether it was or was not present. Only those behaviours that carers/families considered to be difficult to manage without input from multidisciplinary members in the primary or secondary services were rated (Appendix 13).

- To carry out further statistical analyses and compare the results of objective assessments by the researcher with the assessments carried out by other clinicians, information on the list of co-morbid medical or psychiatric diagnoses (including a diagnosis of ASD) based on ICD-10

clinical criteria were collected from the service users' case files and electronic data records. The ICD-10 coding based on clinical diagnoses of various medical and psychiatric co-morbidities had already been recorded by the consultant psychiatrists/responsible clinicians in the medical case files and/or the electronic data records of the service users registered with the local adult ID service. For example the ICD-10 coding for a service user with a diagnosis of childhood autism had been recorded as F84.0.

4.5. Statistical analyses

All data for stage 1 and stage 2 were analysed using Stata statistical package version 12.0 (StataCorp, 2011). The following analyses were conducted:

4.5.1. Statistical analyses for stage 1

The demographic characteristics and prevalence of sensory impairment, ASD and comorbid physical and mental health problems in the study population were described (research question 1). The relationship between sensory impairment and potential confounders were further investigated using Pearson's chi-squared tests. To explore the relationship between number of autistic traits, sensory impairment and potential confounders, general linear modelling was used. Autistic traits were then dichotomised into ASD (4 or more traits) and no ASD (<4 traits) and the relationship between ASD, sensory impairment and potential confounders was further explored using logistic regression modelling (research question 2). Logistic regression modelling was also used to explore the relationship between individual traits, sensory impairment and potential confounders (research question 3) and challenging behaviour, sensory impairment and potential confounders (research question 4). All main effects and 2-way interactions were tested. The relationship between significant interactions was visualised using graphs of predicted probabilities for ease of interpretation.

4.5.2. Statistical analyses for stage 2

The demographic characteristics, aetiology of ID and co-morbid physical and mental health problems of both the deaf and blind subgroups were described (research question 5). Rates of ASD, defined as a PDD-MRS score of 10 and above, in both deaf and blind subgroups were compared using Pearson's chi-squared test. Rates of epilepsy in the blind and deaf subgroup were also compared. To compare differences in diagnostic methods used to identify ASD (research question 6), the PDD-MRS, ICD-10 clinical diagnoses and traits on the LLDR database were compared and agreement was evaluated using Cohen's kappa statistics. Conditional logistic regression and logistic regression (see below) were used to explore the relationship between ASD (outcome as measured by using PDD-MRS) and congenital deafness and ASD and congenital blindness, adjusting for potential confounders (research questions 7 & 8).

There was a high attrition rate for this stage of the study, which had the unintended consequence of 'orphaning' some of the blind/deaf service users and their corresponding controls. Thus, when applying the conditional logistic regression model during the data analysis, blind/deaf service users were excluded if they had no matching control and, similarly, controls were excluded if they had no matching case. As a result, the conditional logistic regression included only those people who were alive, could be contacted and consented to take part in the study. Therefore, for more detailed analyses, logistic regression (i.e. no longer 'conditional') was used to explore the relationship between ASD (outcome as measured by PDD-MRS) and congenital blindness or deafness. For these logistic regression analyses, as well as adjusting for potential confounders, the models also adjusted for the original matching variables, degree of ID and gender. For all analyses, a p-value of ≤0.05 was used to denote statistical significance.

5. Challenges faced when implementing the research project methodology plan

5.1. Introduction

To carry out the assessments in a highly complex group of service users, the researcher attended a number of training courses (Appendix 16) and accreditations:

- Makaton modules 1, 2, 3 and 4, Grantham, 2005
- Describing and presenting data, Leicester, 2006
- Analysing data and further data analysis using SPSS, Leicester, 2006
- Having confidence in data, Leicester, 2006
- Designing a questionnaire, Leicester, 2006
- The senses sight and sound, Bristol, 2006
- British Sign Language (BSL) Level 1, 2, 3 and pre-level 4 from the Council for the Advancement of Communication with Deaf People (CACDP), Lincoln, Leicester & Birmingham, 2006–2009
- DISCO training course (Lorna Wing centre), Bromley, 2007–2008
- Deaf-blind manual and alphabet level 2, Deaf-blind UK, Peterborough, 2008
- An introduction to visual impairments and autism spectrum conditions (RNIB/NAS), Birmingham, 2011
- ADOS-2 training course (Spectrum Specialist Consultancy), Leicester, 2012
- Better eye care for people with LD, Public Health England, Peterborough, 2013 (https://www.improvinghealthandlives.org.uk/)
- Mary Kitzinger Workshop on visual impairment, Institute of Child Health, UCL, London, 2012, 2013 & 2014

Box 5.1 summarises some of the challenges faced when completing the project.

Box 5.1: Challenges faced when implementing the research project methodology plan

Ethical

- Contacting service users who were receiving clinical input from the ID service.
- Consent taking from those who did not have capacity e.g. profound ID.

Logistical

- Locating the service users.
- Time.
- Distance and travelling across the county.
- Administration.

Assessments

- Assessing ASD/mental illness in client groups with complex needs:
  - Training on DISCO and ADOS;
  - Training on PDD-MRS/ABC;
  - Training on British Sign Language and deaf-blind sign language/manual.
- Assessing and following up a sensory impairment:
  - Involving other services e.g. opticians, hearing services, ophthalmology and ENT departments;
  - Basic physical exam and issues related to non-compliance;
  - Encouraging carers to complete the hearing and visual checklists.

Addressing unidentified/unmet needs of the patients

- Determining the aetiology of ID.
- Determining the aetiology of sensory impairment.
- Unmet clinical (e.g. monitoring response and side effects to medications) and social (e.g. level of support) needs.
- Referrals needed to other specialists.
- Encouraging compliance with hearing aids/spectacles.
- Encouraging service users/carers to keep up the appointments.
- Completing the investigations necessary.
- Flexibility in offering home visits for assessment.

ADOS: Autism Diagnostic Observation Schedule; DISCO: Diagnostic Interview for Social and Communication Disorders; ENT: ear, nose, throat; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale

The main challenge of this stage of the project was its slow pace. Only one or two service users could be seen each week as, being a full time NHS clinician, the researcher needed to prioritise care and any clinical emergencies that occurred, even if they clashed with the time allocated to the research project.

The nature of the research project and its robust methodological design demanded a detailed assessment to ensure only those with severe to profound and congenital sensory impairment could be included in stage 2 of the project. Time and commitment over a period of 4–5 years was therefore required to see all service users registered with sensory impairment, over and above the numbers needed for the completion of the research project that had been ascertained by the power calculation.

On a similar note, the relatively long period of data collection affected attrition rate because some service users sadly died during the course of the project and some moved out of the county before having the opportunity to be assessed.

The ethical conflict of conducting research on the same service users who were receiving clinical input from the researcher or his colleagues were discussed in detail during supervision hours and annual thesis committee meetings so that the vulnerable service users and their families/carers felt safeguarded and not pressured to participate in the project if they wished not to (Bravo *et al*. 2003; Lader *et al*. 2004; O'Neill, 2004; Cameron & Murphy, 2006; DH Scientific Development and Bioethics Division, 2008). Therefore, attempts were made to engage service users and carers group from the early stages of the project through the local Trust board. In addition, service users representatives who were active in the local charity organisations were invited to make their views known about the project. This was facilitated by producing accessible information leaflets, both pictorial and in Braille, and engaging sign language interpreters while presenting the projects in different settings at the initial stages. The other practical challenge was an inability to take consent from those with severe and profound ID. English consent procedures are complex

with regard to adults who lack capacity for this type of research project. Proxy consent from a carer or another adult cannot be taken, but they need to be consulted and asked whether they think the individual might be willing to take part in the study and to advise on any potential distress that participation might incur (DH Scientific Development and Bioethics Division, 2008). Importantly, carers' own views of taking part in research more generally should not influence their role as a 'consultee'. Unfortunately some of the carers/families chose to deprive the service users from participating in the research project despite having all the information and knowing about the benefits of participation. Carers/families who refused participation included both cases and controls in the deaf and blind subgroups and, as most of the service users could not be contacted directly, there were times when it was frustrating not to be able to proceed because the relevant carers/families would not allow service users to be approached by the researcher or, indeed, any other members of the multidisciplinary team. Fortunately, this situation occurred in a relatively small number of cases.

It has previously been observed that there are a number of challenges with regard to recruiting participants with ID into research studies. In 2010, Oliver-Africano reported a number of refusals from managers in establishments because priority was given to the day-to-day running of the establishment rather than to research participation. Our own experiences in Leicestershire suggest that managers often cite unacceptable resident disruption as a reason for refusal (Leaver *et al.* 2012).

Similar to the above findings, some of the families and paid carers refused to allow access to potential participants because they simply could not see any benefit to the service user's involvement and believed that any encounter with health professionals might make their mental health worse. Some families felt stigmatised at the notion of assessment for ASD and were concerned that the assessment might adversely affect the service users' financial benefits, and some mentioned that the service users were already receiving input from their general practitioners and other health professionals and did not want to put them through any additional stress.

Travelling to see service users at their place of residence was a real challenge, as the majority of service users were living across a vast geographical area within Leicester city, Leicestershire and Rutland. For some, this required freeing up one or more hours travelling time in addition to the time for the assessment.

Identifying the aetiology of ID or a sensory impairment in a considerable number of the service users proved quite challenging as some of the service users' parents had passed away and no other family members were involved in their care; therefore the researcher had to make arrangement to collect as much information as possible to be able to liaise with other medical specialists such as clinical geneticists, neurologists, ENT specialists and ophthalmologists to carry out detailed examination and investigations to determine aetiological factors.

Although the researcher's medical secretary dealt with administrative support for the project, the unmet clinical and social needs unravelled by detailed assessment for each service user created a lot of additional clinical work that the researcher could not ignore or pass on to other colleagues and, therefore, added considerably to the burden of work and meant that sacrifices had to be made at the expense of non-working hours, weekends and annual leave. For some service users, the researcher was the sole professional and, therefore, carers/families used him as a first-point of contact to request help regarding practical support, such as an environmental adaptation or level of support, even after the assessment was completed. Although this was not a condition of the ethics approval, the researcher, as the only physician in a position to recognise unmet needs, had to intervene in some cases, on ethical ground, to ensure that they could have access to necessary medical or social/practical support (please see section 5.2, addressing unmet needs, for more details). Although clinically challenging and time consuming, this aspect of the research was extremely rewarding because the researcher managed to deal with the issues that mattered most to the service users and their families, which had not hitherto been picked up, either in the primary care or hospital setting. As a result, and with permission from the service users and their carers/families, the researcher needed to make referrals to other colleagues in primary/secondary care or

social care services, request necessary investigations or directly recommend treatment options to ensure that the service users were able to be followed up in the community. The next section provides a summary of these interventions.

5.2. Addressing the unmet needs of service users with blindness

5.2.1. Epilepsy management

For 8 patients, epilepsy management needed to be addressed to ensure safety. This included discussion on the risk of Sudden Unexpected Death in Epilepsy (SUDEP) and the ways in which it could be prevented, e.g. adjusting the dose of antiepileptic medications, referral to the epilepsy clinic at Leicester General Hospital, preparing a rescue medication protocol and prescribing a rescue medication (such as Midazolam), arranging for basic epilepsy training and requesting an epilepsy bed sensor.

5.2.2. Metabolic monitoring

For 22 service users, metabolic monitoring needed to be completed because the service users were on several psychotropic medications and no annual blood tests or electrocardiography (ECG) monitoring had been requested. These were carried out to help to prevent serious side effects of psychotropic medications such as diabetes and arrhythmias.

5.2.3. Certificate of Visual Impairment (CVI)

There were 10 service users who, despite being clinically blind, did not have a CVI which is needed to register them with the local charity, VISTA, to receive rehabilitation, day services and environmental adaptations. They were therefore referred to the opticians/ophthalmologists at the Leicester Royal Infirmary for further assessment and confirmation of their blindness to be issued with a CVI. Most of these service users had moved to Leicestershire from other parts of the country and, therefore, had been lost to follow up. In some cases, blindness and its degree were simply taken for granted and no attempt had been made by the professionals to ascertain the nature or the aetiology of the visual impairment, partly because, in some, other co-morbid health issues were given priority (e.g. epilepsy, challenging behaviour, mental ill-health).

5.2.4. Training staff and families on visual impairment

In the course of conducting the research project the researcher, with the help of the nursing manager and the deputy clinical director of the service for the short break/respite staff/inpatient unit, managed to secure limited funding to receive training from VISTA to be able to clinically support service users with blindness. This included training for different nursing staff and nursing assistants carried out over 3 separate days in various settings to improve the quality of service delivery for this highly vulnerable group of service users.

In addition to the above, 6 families/carers were given a CD-ROM and booklet (published by RNIB) purchased by the researcher on how to manage challenging behaviour in people with blindness and ASD (Bell & Bell, 2010; Bell *et al.* 2011a & b; Bell, 2013). For a number of service users, the researcher also liaised with the training officer at VISTA to arrange and facilitate training for the carers and families at their place of residence.

5.2.5. Social care referrals

Five cases needed referral to colleagues at social services for a carer's needs assessment, environmental adaptation, and allocation of a respite facility/short break or support at home.

5.2.6. Multidisciplinary input

The inputs required from the multidisciplinary team were as follows:

- Occupational therapy input for environmental, life skills or sensory assessments, assessment of falls and dementia care mapping (n=9).
- Speech and language therapy input for advice on communication skills or formulating an eating and drinking plan to avoid choking (n=6).
- Support from health facilitators (primary care liaison nurses) (n=2) to develop a Health Action Plan for the service user.
- Referral to community nurses for desensitisation prior to investigations, monitoring mental health and side effects of medication (n=4).

- Input from physiotherapists (n=3) for assessment of mobility and allocation of a wheelchair.
- Psychology input (n=2).
- Support from the outreach nursing staff (n=2) for the management of challenging behaviours.

5.2.7. Primary care input/involvement

Numerous letters were sent to the general practitioners (GPs) to request the management of insomnia, menstrual difficulties, vitamin D deficiency or other physical health problems, which had not been discussed with the GPs prior to the research project. GPs were directly informed when a diagnosis of ASD or other condition, such as a genetic syndrome, was made, and they received copies of requests made for blood tests, ECG, EEG or other investigations. They also received copies of all letters sent to other agencies, so that they could keep their records up to date.

For 6 individuals, the researcher had to write to the GP to recommend either adjusting the dose of a psychotropic medication, request a change from tablets to syrup or vice versa (to reduce the likelihood of non-compliance and choking), or initiate a service user on a regular or “as needed” (PRN) medication. One service user needed a referral to the district nurses for the management of incontinence and prevention of pressure sores.

5.2.8. Secondary care input

Six service users were referred for dental work. One service user needed neurology input for torticollis. Five service users had to be referred to the acute liaison nurses based at the University Hospitals of Leicester Trust for support regarding reasonable adjustments in accessing their investigations or treatment in a hospital setting. A genetic referral was made in 5 of the cases to rule out or confirm a diagnosis of a genetic syndrome. Three service users needed cardiology input and for one, further investigations were requested to assess a previous cardiac arrest in the community. One service user needed brain MRI scanning to investigate a neurodegenerative disorder. In addition, referrals were

made to the specialist hearing services with expertise in dealing with complex needs of people with ID to ensure that 2 of the service users with blindness did not also suffer from a hearing impairment.

5.3. Addressing the unmet needs of the controls for the blind subgroup

Without going into details of exactly what was done for every service user, Box 5.2 describes the referrals and contacts made with regard to addressing unmet needs of the controls, and number of service users affected.

Tables 5.3 and 5.4 summarise the number of referrals and investigations requested in the blind subgroup.

Box 5.2: Referrals and contacts made to address the unmet needs of the controls for the blind subgroup

Epilepsy management strategy

- 6 service users affected.

Metabolic monitoring

- Blood tests and ECG monitoring for 33 service users (approximately half of the control group).

Social care referral

- For 2 service users.

Multidisciplinary input

- Physiotherapy input for 7 service users.
- Occupational therapy input for 4 service users.
- Speech and language therapy input for 4 service users.
- Community nursing input for 2 service users.
- Health Action plan provided for 2 service users.

Primary care input/involvement

- GPs received copies of all relevant clinical letters.
- GP received a request to adjust dosage of antidepressant medication for 1 service user.
- GP received a request to initiate medication for hypothyroidism (n=1) and genital thrush (n=1).
- Referral to incontinence nurses for 1 service user.

Secondary care input

- Referral to the genetic clinic for 3 service users.
- Referral to the neurology clinic for 2 service users.
- Referral to cardiology clinic for 2 service users.
- Referral to orthopaedics clinic for 1 service user.
- Brain scanning (MRI/CT) for 6 service users and EEG for 1 service user.
- Referral to opticians for 3 service users.
- Referral to the acute liaison nurses for 1 service user.
- Referral to the advocacy services for 1 service user

CT: computed tomography; ECG: electrocardiogram; EEG: electroencephalography; GP: general practitioner; MRI: magnetic resonance imaging

Table 5.3: Number of referrals needed to be sent to different agencies for blind service users and their controls

Number of letters	Blind service users	Sighted service users
1	26	26
2	17	20
3	8	4
4	2	2
6	1	0
Total	**54**	**52**

Table 5.4: Number of investigations requested for blind service users and their controls

Investigation	Blind service users	Sighted service users
Bloods	30	34
ECG	31	32
EEG	1	1
Neuroimaging (Brain MRI or CT scan)	2	4
Total	**64**	**71**

CT: computed tomography; ECG: electrocardiogram; EEG: electroencephalogram; MRI: magnetic resonance imaging

5.4. Addressing the unmet needs of the service users with deafness

5.4.1. Metabolic monitoring

Nine service users needed metabolic monitoring (blood tests and ECG).

5.4.2. Social care referrals

Seven service users required input from social services and advocacy groups for help with environmental adaptation and also for best interest meetings and multidisciplinary team meetings for an appropriate accommodation and for adjustment to be made regarding their hearing impairment.

5.4.3. Multidisciplinary input

Six service users needed referral to the acute liaison nurses for support during their admission or appointments in general hospital settings. One service user needed referral for support from the psychology and community nurses.

5.4.4. Secondary care input

Seven service users were referred to the clinical genetics department to screen for a genetic syndrome. Three service users were referred to opticians and 3 to hearing services for monitoring of their hearing aids and for information and support on how to maintain and clean their hearing aids. One service user was referred to the local community dentist with expertise in addressing the needs of people with ID. One service user needed urgent clinical input due to severe anaemia, which was picked up by carrying out the blood tests following the research interview session

5.4.5. Additional input

One service user needed referral for legal input regarding forensic problems and one carer required a supporting letter to her employer to handle the burden of care more effectively by working flexibly.

5.5. Addressing the unmet health needs of the controls for the deaf subgroup

Box 5.5 describes the referrals and contacts made with regard to addressing unmet needs of the controls, and number of service users affected.

In brief, the information collected during the research project was used to not only update the data on the LLDR database and the local adult ID service as well as primary care/GP practice data bases but also address the unmet social and health needs of service users. This was even the case for those whose data was not included in the final analysis i.e. those who were excluded (Appendix 4); while taking part in the research project, service users and their families/carers were supported to access services by using the principle of reasonable adjustments.

Box 5.5: Referrals and contacts made to address the unmet needs of the controls for the deaf subgroup

Metabolic monitoring

- Blood tests and ECG monitoring for 10 service users.

Social care referral

- Practical and environmental support for 4 service users.

Multidisciplinary input

- Physiotherapy input for 2 service users.
- Occupational therapy input for 3 service users.
- Speech and language therapy input for 1 service user.
- Community nursing input for 3 service users.

Primary care input/involvement

- GPs received copies of all relevant clinical letters.
- Request for adjustment to dosage of anti-epileptic medication for 1 service user.

Secondary care input

- Referral to genetic clinic for 1 service user.
- Referral to hearing services for 2 service users.
- Referral to ENT services for 2 service users.
- Brain scanning for 2 service users suspected of neurodegenerative disorder.
- Referral to dermatology clinic for 1 service user.

ECG: electroencephalogram; ENT: ear, nose, throat; GP: general practitioner

6. RESULTS FROM STAGE 1: ANALYSIS OF DATA FROM THE LEICESTERSHIRE LEARNING DISABILITY REGISTER (LLDR)

6.1. Introduction

At the time of the study, there were 3,138 service users registered on the LLDR, of whom about 66 (2.1%) had an IQ above 70 and 60 (1.9%) had no data on their degree of ID (Table 6.1). The percentage of service users with mild ID (21.7%), moderate ID (22.5%) and profound ID (22.5%) were quite similar, but those with severe ID had the largest distribution at 29.3%.

Fifty-seven per cent of the participants were men. For the purpose of the statistical analysis, the participants were grouped into 4 distinct age categories; 18–29 (24.1%), 30–39 (23.5%), 40–49 (22%) and ≥50 years old (30.4%).

The majority of service users were white (83.2%) or South Asian (13.7%) and were either living in a care home setting (48.5%; various categories of residential facilities or supported living accommodation) or with family members (41.6%). Only 9.2% were living with a spouse, a partner or independently and 3.3% (n=104) were registered as married.

Table 6.1 provides more detail on the demographic characteristics of the study population.

Table 6.1: Demographic characteristics of the study population based on the data available on the LLDR (n=3138)

Demographic data	N	(%)
Gender:		
Male	1789	(57.0)
Female	1349	(43.0)
Age group (years):		
18–29	757	(24.1)
30–39	737	(23.5)
40–49	691	(22.0)
≥50	953	(30.4)
Degree of ID:		
Borderline (IQ>70)	66	(2.1)
Mild (IQ≤70)	679	(21.7)
Moderate (IQ<55)	707	(22.5)
Severe (IQ<35)	919	(29.3)
Profound (IQ<20)	707	(22.5)
Missing data	60	(1.9)
Married:	104	(3.3)
Ethnicity:		
White	2612	(83.2)
Asian	429	(13.7)
Black	45	(1.4)
Mixed	34	(1.1)
Other/unknown	18	(0.6)
Accommodation:		
Living independently/with partner/spouse	289	(9.2)
With parents/family/foster carers/guardians	1304	(41.6)
Residential/nursing home/Supported living/NHS facilities	1522	(48.5)
Missing data	23	(0.7)
Total	**3138**	**(100.0)**

ID: intellectual disability; IQ: intelligence quotient; LLDR: Leicestershire Learning Disability Register

6.2. Prevalence of sensory impairment among adults with intellectual disability (Research Question 1)

About half of the adults registered with the LLDR were prescribed glasses because of a refractive error (47.7%). The most common serious co-morbid condition was epilepsy, with a rate of 25%, showing that 1 in 4 service users were suffering from this chronic neurological condition. This was followed by mobility problems in 34.9% (including those who could not walk independently and those who needed mobility aids e.g. wheelchairs or help from carers to mobilise), urinary incontinence in 23.6% and faecal incontinence in 16.2% of the service users. Down syndrome was reported in 13.5% and a smoking status in 12.5% of the study population.

Approximately 3.6% of the sample (n=114) had total blindness and 1.6% had total deafness (n=51). The rate of partial visual impairment and hearing impairment were respectively 8.7% (n=272) and 5.6% (n=175).

Twenty-two service users had various degrees of deaf-blindness (0.7%). Sign language or Makaton users constituted 6.7% of the service users and 4.8% of the whole target population were using a hearing aid. This included those whose hearing impairment could be corrected by using a hearing aid.

An unpublished report by VISTA (directly obtained by the researcher) in Feb 2012 showed that there were 2,534 people with total blindness in the general population (which gives a prevalence of 0.3% given the total adult population of Leicester City, Leicestershire and Rutland of about 700,000). Blindness, therefore, was around 12 times more prevalent in the ID population than those without ID. The rate of partial blindness in the general population of the Leicester City, Leicestershire and Rutland was reported to be 0.4% (n=3,221), which is 22 times lower than the rate seen in people with ID within the same geographical area. In addition, given a deaf-blindness prevalence of around 1 in 10,000 of the general population, this figure shows that deaf-blindness is occurring about 70 times higher in the population with ID. There was no report by VISTA regarding the prevalence of deafness in the general population of

Leicestershire for a comparison with the prevalence figure found in this study for people with ID who had deafness. Table 6.2 provides more details of co-morbid conditions in the registered population of adults with ID in the LLDR (n=3,138).

Table 6.2: Co-existing conditions in the study population, based on the data available on the LLDR and local adult ID service databases (n=3138)

Co-morbid condition	N	(%)
Epilepsy	786	(25.0)
Down syndrome	425	(13.5)
Mobility problems	1095	(34.9)
Urinary incontinence	742	(23.6)
Faecal incontinence	508	(16.2)
Visual impairment:		
Blindness	114	(3.6)
Partial visual impairment	272	(8.7)
Using spectacles for refractive error	1498	(47.7)
Hearing impairment:		
Deafness	51	(1.6)
Partial Hearing Impairment	175	(5.6)
Signing	209	(6.7)
Using hearing aids	151	(4.8)
Deaf-blindness		
Total:	12	(0.4)
Partial	10	(0.3)

LLDR: Leicestershire Learning Disability Register

The relationship between sensory impairment, degree of ID, age and other co-morbidities could not be investigated in all service users with sensory impairment on the LLDR database because some had either a borderline IQ or a unilateral sensory impairment (Table 6.3).

Table 6.3: Relationship between visual impairment, age, degree of ID, gender, Down syndrome and epilepsy in the study population (n=3138)

	Blindness (n=99) N (%)	Partial visual impairment (n=272) N (%)	No visual impairment (n=2737) N (%)	Missing data (n=30) N (%)	Pearson Chi^2	Chi^2 p-value
Age group (years):						
18–29	16 (15.2)	52 (19.2)	681 (24.8)	9	17.38	0.04
30–39	27 (27.6)	61 (22.4)	645 (23.6)	4		
40–49	18(18.4)	74 (27.2)	590 (21.6)	9		
≥50	38 (38.8)	85 (31.2)	821 (30.0)	8		
Degree of ID:						
Borderline	1 (1.0)	2 (0.7)	63 (2.3)	0	161.16	<0.001
Mild	7 (7.1)	32 (11.8)	638 (23.3)	2		
Moderate	3 (3.0)	42 (15.4)	658 (24.0)	4		
Severe	29 (29.3)	91 (33.5)	789 (28.9)	10		
Profound	58 (58.6)	99 (36.4)	536 (19.6)	14		
Missing data*	1 (1.0)	6 (2.2)	53 (1.9)	0		
Gender:						
Male	58 (58.6)	148 (54.4)	1565 (57.2)	18	0.99	0.80
Female	41 (41.4)	124 (45.6)	1172 (42.8)	12		
Down syndrome	8 (8.1)	71 (26.1)	341 (12.5)	5	42.16	<0.001
Epilepsy	41 (41.4)	91 (33.5)	641 (23.4)	13	33.57	<0.001
-Prescribed spectacles	-	171 (62.9)	1300 (47.5)	6	606.14	<0.001
-Missing data*	-	1 (0.4)	3 (0.1)	8		

ID: intellectual disability

*Not included in the analysis

More details on the number of patients, who were excluded from the data analysis, e.g. those who had borderline IQ or unilateral sensory impairment are provided in Chapter 7 (Flowcharts 7.1 & 7.2) and Appendix 4.

Pearson Chi-Square analysis of those with complete information revealed that both total and partial visual impairment (congenital and acquired) were significantly associated with degree of ID ($p<0.001$) and age ($p=0.04$). People with visual impairment were also more likely to have co-morbid epilepsy or Down syndrome ($p<0.001$ for both; Table 6.3): 63.5% of those with Down syndrome (n=270) were using spectacles. Gender was not associated with visual impairment.

Similarly, deafness (congenital and acquired) was significantly associated with degree of ID (Table 6.4). However, for those with partial deafness (congenital and acquired) the association pattern was more complex as the majority of these individuals had moderate ID, followed by severe, and then profound ID, suggesting an underestimation in carers' reports of hearing impairment in service users with severe and profound ID. Overall, however, hearing impairment had a statistically significant association with the degree of ID ($p=0.03$).

Similarly, a relationship was found between age group and hearing ($p<0.001$); older individuals were more likely to have hearing impairment, with the exception of 18–29 years old, where the rate of partial hearing impairment was higher than that in 30–39 and 40–49 years old.

Neither gender nor co-morbid epilepsy was associated with hearing impairment, but hearing impairment was more commonly reported in service users with Down syndrome ($p<0.001$). Approximately 1 in 10 people (n=41/425) with Down syndrome used hearing aids.

Table 6.4: Relationship between hearing impairment, age, degree of ID, gender, Down syndrome and epilepsy in the study population (n=3138)

	Deafness (n=41) N (%)	Partial hearing impairment (n=175) N (%)	No hearing impairment (n=2892) N (%)	Missing data (n=30) N (%)	Pearson Chi^2	Chi^2 p-value
Age group (years):						
18–29	6 (14.7)	45 (26.0)	696 (24.2)	10	35.84	<0.001
30–39	3 (7.3)	24 (13.9)	705 (24.5)	5		
40–49	11 (26.8)	30 (17.3)	642 (22.3)	8		
≥50	21 (51.2)	76 (42.8)	849 (29.4)	7		
Degree of ID:						
Borderline	0 (0.0)	1 (0.6)	65 (2.2)	1	27.48	0.03
Mild	5 (12.2)	25 (14.3)	646 (22.4)	3		
Moderate	11 (26.8)	56 (32.0)	636 (22.0)	4		
Severe	12 (29.3)	52 (29.7)	844 (29.2)	11		
Profound	13 (31.7)	38 (21.7)	645 (22.3)	11		
Missing data*	-	3 (1.7)	56 (1.9)	-		
Gender:						
Male	30 (73.2)	98 (56.0)	1642 (56.8)	19	5.00	0.17
Female	11 (26.8)	77 (44.0)	1250 (43.2)	11		
Down syndrome	6 (14.6)	57 (32.6)	359 (12.4)	3	57.62	<0.001
Epilepsy	7 (17.1)	32 (18.3)	737 (25.5)	10	7.041	0.07
-Prescribed hearing aids	20 (48.8)	69 (39.4)	†62 (2.1)	0	683.01	<0.001
-Missing data*	1 (2.4)	1 (0.6)	3 (0.1)	-		
-Signing	22 (53.6)	28 (16.0)	157 (5.5)	2	167.57	<0.001
-Missing data*	0 (0.0)	6 (3.4)	62 (2.1)	-		

ID: intellectual disability; *not included in the group analysis; †those whose mild to moderate hearing impairment could be corrected by hearing aids (reversible).

In addition to the above, a strong relationship was observed between both partial and total visual impairment/hearing impairment and lack of speech, incontinence and mobility problems ($p<0.001$). However, these skills are known to be a function of the severity of the brain damage rather than being directly associated with either hearing impairment or visual impairment.

6.3. Relationship between ASD and sensory impairment (Research Question 2)

As discussed in the methodology section in chapter 4, the register holds data on 5 key autistic traits for each individual (presence of stereotypies, presence of elaborate routines, lack of empathy, use of speech and quality of social interaction; Box 4.1). The population was restricted to 2,940 service users who had complete data on vision, hearing, severity of ID (mild, moderate, severe and profound only) and autistic traits (Table 6.5). This population included data on both total and partial sensory impairment regardless of whether these were congenital in nature or acquired, because the data in the LLDR did not differentiate between a congenital sensory impairment and an acquired sensory impairment.

Table 6.5: Data exclusions

Description	Number
Total adults on LLDR	3138
Missing data on visual impairment	30
Missing data on hearing impairment	20
Borderline or unknown severity of ID	125
Incomplete information on autistic traits	23
Total remaining	2940

ID: intellectual disability; LLDR: Leicestershire Learning Disability Register

Using the agreed threshold of 4 or more traits, 506 individuals (17.2%) met the criteria for ASD; 476 adults (16.2%) had 3 traits and 1299 (44.2%) had 1–2 traits. Only about 1 in 5 of the population (n=659; 22.4%) had no traits at all (Table 6.6).

Table 6.6: Prevalence of autistic traits based on 5 key indicators on the LLDR database (n=2940)

Number of autistic traits	Frequency	(%)
0	659	(22.4)
1	661	(22.5)
2	638	(21.7)
3	476	(16.2)
4	344	(11.7)
5	162	(5.5)

LLDR: Leicestershire Learning Disability Register

6.3.1. Relationship between sensory impairment and number of autistic traits

General linear modelling (multi-variable analysis) did not reveal a significant relationship between number of autistic traits (entered into the model as a continuous variable) and visual or hearing impairment (Table 6.7). However, older age and being South Asian were associated with having fewer traits ($p<0.001$ and $p=0.001$ respectively) and being male and having more severe ID were associated with having more traits ($p=0.01$ and $p<0.001$ respectively). No interactions were observed between variables in this model.

Table 6.7: General linear model showing the relationship between number of autistic traits as a continuous variable, with visual impairment/hearing impairment (n=2940)

Variable	Coefficient	95% CI	p-value
Visual impairment	-0.07	-0.18 to 0.03	0.17
Hearing impairment	-0.10	-0.14 to 0.13	0.89
Age group	-0.14	-0.18 to -0.10	<0.001
Ethnicity			
White	0.00	(reference)	-
South Asian	-0.22	-0.35 to -0.09	0.001
Black	-0.01	-0.39 to 0.37	0.96
Other/Unknown	0.31	-0.06 to 0.68	0.10
Gender Male	0.11	0.03 to 0.20	0.01
Degree of ID	0.81	0.77 to 0.85	<0.001

CI: confidence interval; ID: intellectual disability

6.3.2. Relationship between sensory impairment and ASD

The relationship between sensory impairment and ASD (4 or more traits), as a dichotomous variable, is shown in Table 6.8. People of South Asian origin were less likely to have ASD, as measured by the carer-reported traits (p<0.001). Younger age groups and people with severe and profound ID were more likely to have ASD, but there was an interaction between both, so they could no longer be interpreted independently. The interaction between age group and degree of ID is shown graphically in Figure 6.9. As expected, the graph shows that the predicted probability of having ASD was highest in people with profound ID, but that this probability peaked at age 30–39 years old. In contrast, the predicted probability of having ASD in those with mild, moderate and severe ID was highest in the youngest age group (<30 years), and peaked again in those aged 40–49 years old with moderate and severe ID.

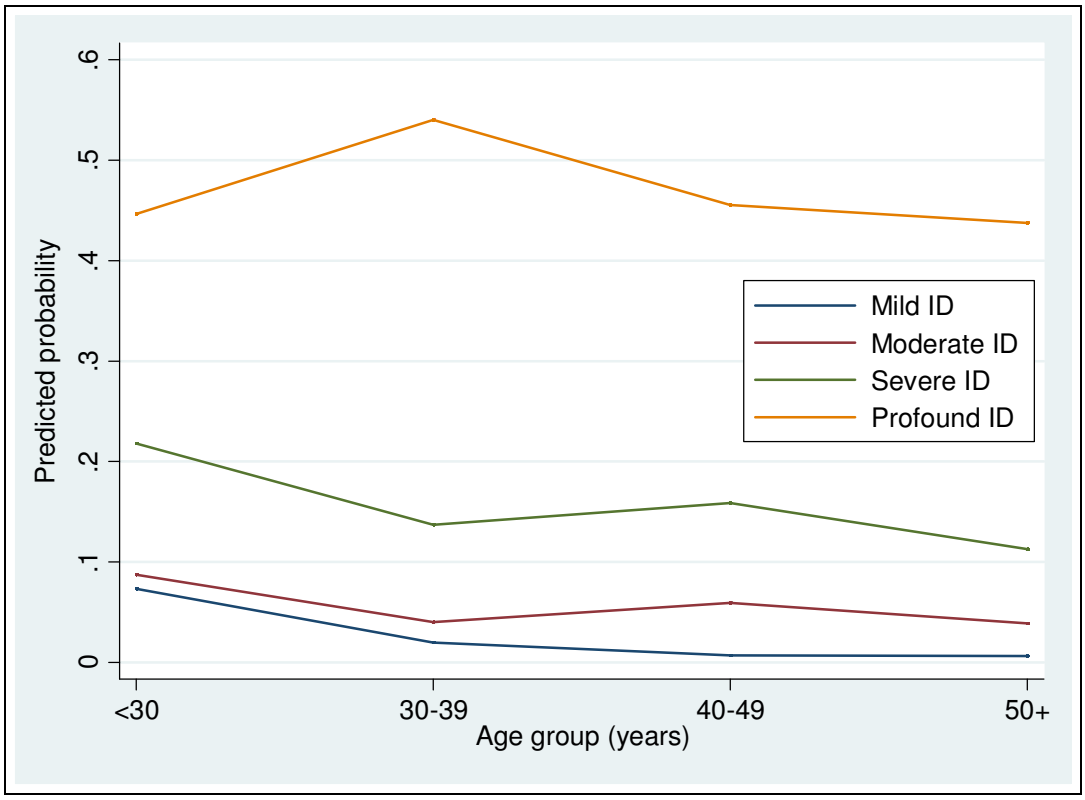
A relationship was not observed between ASD and hearing impairment, visual impairment or gender.

Table 6.8: Logistic regression model showing the relationship between ASD (outcome) and visual and hearing impairment after adjustment for potential confounders (n=2940)

Variable		OR	95% CI	p-value
Visual impairment:	None	1.00	(reference)	
	Partial	0.96	0.67 – 1.36	0.81
	Blind	0.95	0.57 – 1.58	0.85
Hearing impairment:	None	1.00	(reference)	
	Partial	1.10	0.69 – 1.75	0.67
	Deaf	1.08	0.44 – 2.66	0.86
Ethnicity:	White	1.00	(reference)	
	South Asian	0.57	0.42 – 0.78	<0.001
	Black	0.96	0.42 – 2.18	0.92
	Other/Unknown	1.79	0.85 – 3.78	0.13
Age group (years):	<30	1.00	(reference)	
	30–39	0.26	0.08 – 0.81	0.02
	40–49	0.09	0.01 – 0.69	0.02
	50+	0.08	0.01 – 0.60	0.02
Gender:	Male	1.16	0.93 – 1.45	0.18
Degree of ID:	Mild	1.00	(reference)	
	Moderate	1.25	0.58 – 2.67	0.57
	Severe	3.70	1.85 – 7.41	<0.001
	Profound	11.82	5.96 – 23.45	<0.001
Interaction: Age group * Degree of ID				
30–39 years	* Moderate	1.42	0.75 – 2.67	0.28
	* Severe	2.25	1.11 – 4.56	0.02
	*Profound	1.95	1.01 – 3.76	0.05
40–49 years	* Moderate	0.67	0.36 – 1.25	0.21
	* Severe	1.91	0.97 – 3.73	0.06
	*Profound	1.69	0.90 – 3.16	0.10
50+ years	* Moderate	2.47	1.14 – 5.36	0.02
	* Severe	3.37	1.52 – 7.49	0.003
	*Profound	2.30	1.10 – 4.80	0.03

CI: confidence interval; ID: intellectual disability; OR: odds ratio

Figure 6.9: Graph of predicted probabilities for ASD, showing the relationship between age group and degree of ID (n=2940)



ID: intellectual disability

6.4. Relationship between individual autistic traits and sensory impairment (Research Question 3)

6.4.1. Relationship between stereotypies and sensory impairment

There were 1008 service users with stereotypical behaviours (34%).

Using logistic regression with stereotypical behaviour as the outcome variable and, after evaluating the main effects and testing all 2-way interactions, people in the 40–49 and 50+ age groups and those from South Asian origin were found to be less likely to display stereotypical behaviours ($p<0.01$ for all). Similarly, people with severe and profound ID were more likely to display stereotypical behaviours ($p<0.001$ for both) (Table 6.10).

A relationship was not observed between stereotypical behaviours and visual impairment, hearing impairment or gender.

Table 6.10: Logistic regression model showing the relationship between stereotypical behaviours (outcome) and visual and hearing impairment after adjustment for potential confounders (n=2940)

Variable		OR	95% CI	p-value
Visual impairment:	None	1.00	(reference)	
	Partial	1.27	0.96 – 1.68	0.09
	Blind	1.18	0.75 – 1.84	0.49
Hearing impairment:	None	1.00	(reference)	
	Partial	0.85	0.59 – 1.23	0.39
	Deaf	0.91	0.44 – 1.88	0.79
Ethnicity:	White	1.00	(reference)	
	South Asian	0.69	0.54 – 0.88	0.002
	Black	1.37	0.71 – 2.66	0.35
	Other/Unknown	0.70	0.36 – 1.38	0.30
Age group (years):	<30	1.00	(reference)	
	30–39	0.92	0.73 – 1.16	0.48
	40–49	0.68	0.54 – 0.87	0.002
	50+	0.52	0.41 – 0.65	<0.001
Gender:	Male	1.14	0.97 – 1.34	0.12
Degree of ID:	Mild	1.00	(reference)	
	Moderate	1.20	0.92 – 1.56	0.18
	Severe	2.65	2.08 – 3.36	<0.001
	Profound	6.47	5.01 – 8.37	<0.001

CI: confidence interval; ID: intellectual disability; OR: odds ratio

6.4.2. Relationship between sensory impairment and empathy

Less than half of the service users either lacked or had limited empathy (n=1336; 45%). Men were more likely than women to show deficits in empathy (p=0.003) (Table 6.11). A relationship was not observed between deficits in empathy and visual impairment, hearing impairment or ethnicity.

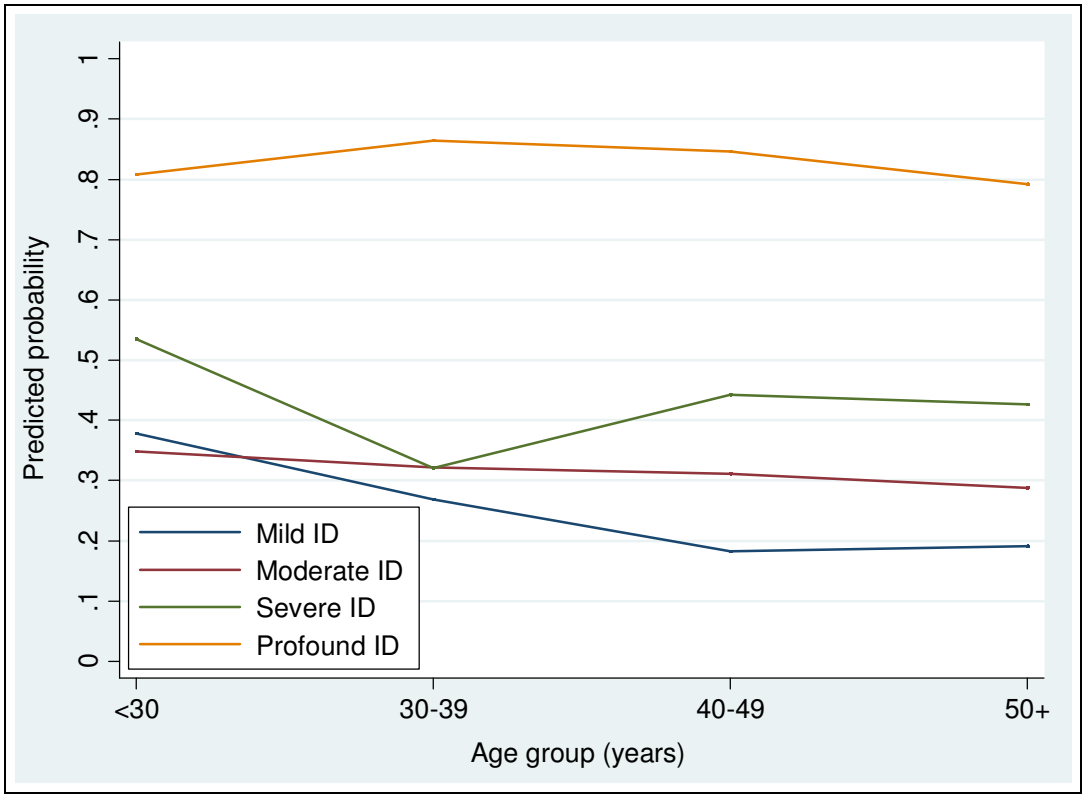
Table 6.11: Logistic regression model showing the relationship between deficits in empathy (outcome) and visual and hearing impairment after adjustment for potential confounders (n=2940)

Variable		OR	95% CI	p-value
Visual impairment:	None	1.00	(reference)	
	Partial	0.77	0.57 – 1.03	0.08
	Blind	0.62	0.38 – 1.02	0.06
Hearing impairment:	None	1.00	(reference)	
	Partial	0.82	0.58 – 1.17	0.28
	Deaf	0.89	0.43 – 1.84	0.75
Ethnicity:	White	1.00	(reference)	
	South Asian	0.81	0.63 – 1.04	0.10
	Black	0.67	0.33 – 1.39	0.28
	Other/Unknown	1.19	0.60 – 2.38	0.62
Age group (years):	<30	1.00	(reference)	
	30–39	0.61	0.39 – 0.95	0.03
	40–49	0.37	0.22 – 0.63	<0.001
	50+	0.39	0.23 – 0.64	<0.001
Gender:	Male	1.28	1.09 – 1.51	0.003
Degree of ID:	Mild	1.00	(reference)	
	Moderate	0.90	0.59 – 1.38	0.63
	Severe	1.95	1.26 – 3.03	0.003
	Profound	7.89	4.62 – 13.47	<0.001
Interaction: Age group * Degree of ID				
30–39 years	* Moderate	1.42	0.75 – 2.67	0.28
	* Severe	2.25	1.11 – 4.56	0.02
	*Profound	1.95	1.01 – 3.76	0.05
40–49 years	* Moderate	0.67	0.36 – 1.25	0.21
	* Severe	1.91	0.97 – 3.73	0.06
	*Profound	1.69	0.90 – 3.16	0.10
50+ years	* Moderate	2.47	1.14 – 5.36	0.02
	* Severe	3.37	1.52 – 7.49	0.003
	*Profound	2.30	1.10 – 4.80	0.03

CI: confidence interval; ID: intellectual disability; OR: odds ratio

An association was found between deficits in empathy and with being younger ($p<0.05$ for all age groups when compared with <30 years) and having severe ($p=0.003$) or profound ID ($p<0.001$) when compared with mild ID. However, an interaction was also observed between age group and degree of ID, which meant that these effects could no longer be interpreted independently. Graphing the predicted probabilities revealed that the effect of degree of ID on deficits in empathy was more pronounced in the 40–49 and 50+ year age groups, while deficits in empathy were very similar in those aged <30 years who had mild or moderate ID (Figure 6.12).

Figure 6.12: Graph of predicted probabilities for lack of empathy, showing the relationship between age group and degree of ID.



ID: intellectual disability

6.4.3. Relationship between sensory impairment and elaborate routines

In total, 1,143 service users had elaborate routines (38.9%). Presence of elaborate routines was more common in men ($p=0.003$), those with other/unknown ethnicity ($p=0.001$) and in those with severe and profound ID

when compared with those with mild ID ($p<0.001$ for both). Conversely they were less common in those of South Asian origin ($p=0.01$). They were also less common in people aged 30–39 years when compared with people <30 years ($p=0.01$), although this may be a spurious finding as no other trends were observed in the age group categories (Table 6.13).

A relationship was not observed between presence of elaborate routines and visual impairment or hearing impairment.

Table 6.13: Logistic regression model showing the relationship between presence of elaborate routines (outcome) and visual and hearing impairment after adjustment for potential confounders (n=2940)

Variable		OR	95% CI	p-value
Visual impairment:	None	1.00	(reference)	
	Partial	0.76	0.58 – 1.01	0.06
	Blind	0.82	0.53 – 1.27	0.37
Hearing impairment:	None	1.00	(reference)	
	Partial	0.95	0.68 – 1.33	0.77
	Deaf	1.60	0.83 – 3.09	0.16
Ethnicity:	White	1.00	(reference)	
	South Asian	0.75	0.60 – 0.94	0.01
	Black	0.95	0.50 – 1.81	0.88
	Other/Unknown	3.02	1.57 – 5.84	0.001
Age group (years):	<30	1.00	(reference)	
	30–39	0.75	0.60 – 0.93	0.01
	40–49	0.95	0.76 – 1.19	0.68
	50+	0.81	0.66 – 1.01	0.06
Gender:	Male	1.26	1.08 – 1.47	0.003
Degree of ID:	Mild	1.00	(reference)	
	Moderate	1.21	0.96 – 1.53	0.11
	Severe	2.32	1.87 – 2.89	<0.001
	Profound	2.16	1.70 – 2.73	<0.001

CI: confidence interval; ID: intellectual disability; OR: odds ratio

6.4.4. Relationship between sensory impairment and use of speech

Approximately half of service users (50%) had not developed an efficient and qualitatively normal speech (n=1468). Unsurprisingly, people who were deaf were more likely to have speech deficits (p=0.02); this effect was of borderline significance in those with a partial hearing impairment (p=0.06). People under the age of 30 were also more likely to have deficits in speech when compared with the older age groups (p<0.005 for all) (Table 6.14). It is worth noting the marked effect of degree of ID on deficits in speech, in that, people were

significantly more likely to have deficits in speech as their severity of ID increased; people with profound ID had 358 times the odds of having speech deficits compared with people with mild ID ($p<0.001$). However, an interaction was observed between ethnicity and degree of ID which means that this effect must be interpreted with some caution.

The interaction between degree of ID and ethnic group is shown graphically in Figure 6.15. The figure illustrates a gradient effect in both South Asian and white groups, in that the predicted probabilities of having deficits in speech increased as the degree of ID became more severe. This effect was more pronounced in people of South Asian origin with moderate and severe levels of ID. However, a gradient effect was not observed in people who were black: the predicted probability of having deficits in speech was similar in those with mild and moderate ID and was again similar, but much higher, in those with severe and profound ID.

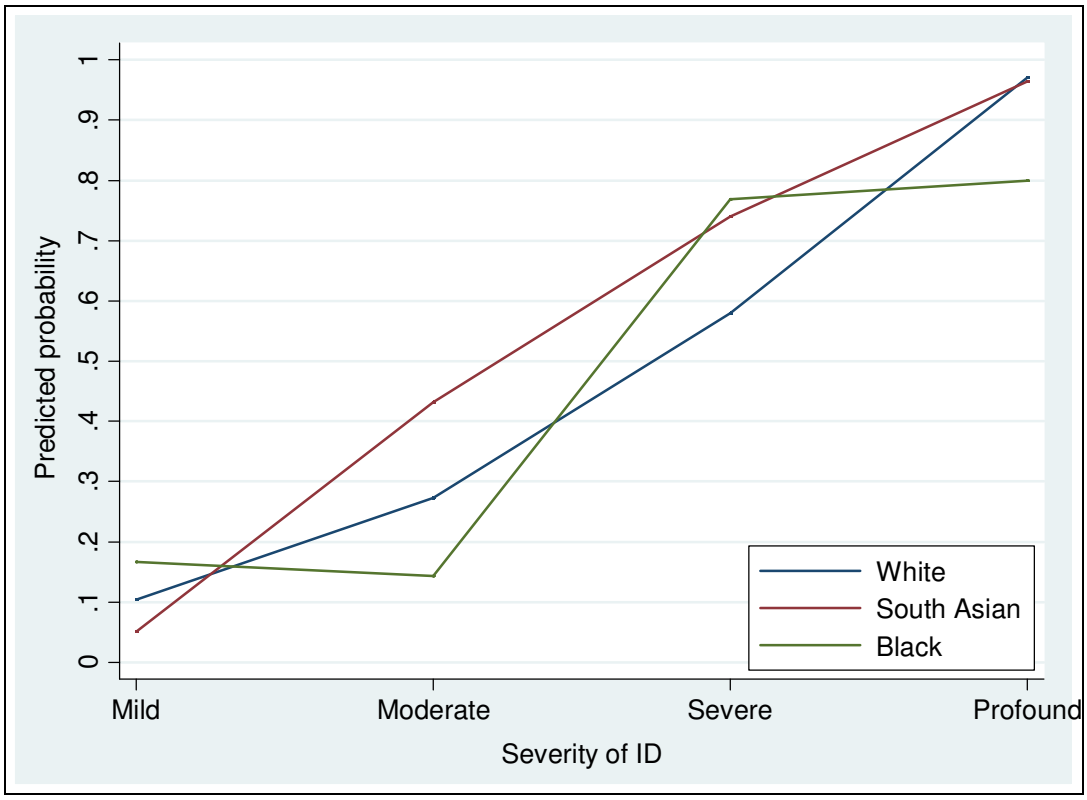
Table 6.14: Logistic regression model showing the relationship between use of speech (outcome) and visual and hearing impairment after adjustment for potential confounders (n=2897*)

Variable		OR	95% CI	p-value
Visual impairment:	None	1.00	(reference)	
	Partial	0.98	0.68 – 1.40	0.91
	Blind	0.60	0.32 – 1.13	0.11
Hearing impairment:	None	1.00	(reference)	
	Partial	1.48	0.99 – 2.22	0.06
	Deaf	2.93	1.23 – 6.96	0.02
Ethnicity:	White	1.00	(reference)	
	South Asian	0.39	0.09 – 1.69	0.21
	Black	1.51	0.17 – 13.28	0.71
Age group (years):	<30	1.00	(reference)	
	30–39	0.57	0.43 – 0.76	<0.001
	40–49	0.66	0.49 – 0.87	0.004
	50+	0.36	0.27 – 0.47	<0.001
Gender:	Male	0.84	0.69 – 1.02	0.08
Degree of ID:	Mild	1.00	(reference)	
	Moderate	3.25	2.36 – 4.48	<0.001
	Severe	14.08	10.36 – 19.14	<0.001
	Profound	358.40	198.94–645.66	<0.001
Interaction: Ethnicity * Degree of ID				
South Asian	* Moderate	4.41	0.96 – 20.26	0.06
	* Severe	4.70	1.04 – 21.36	0.05
	*Profound	1.84	0.31 – 10.96	0.50
Black .	* Moderate	0.23	0.01 – 4.95	0.35
	* Severe	1.58	0.12 – 20.02	0.72
	*Profound	0.07	0.01 – 0.93	0.04

*Other/unknown ethnic group was dropped from the model because it predicted the outcome perfectly

CI: confidence interval; ID: intellectual disability; OR: odds ratio

Figure 6.15: Graph of predicted probabilities for deficits in speech, showing the relationship between degree of ID and ethnic group (n=2897*)



*Other/unknown ethnic group was excluded to correspond with the logistic regression model (see Table 6.15)

ID: intellectual disability

6.4.5. Relationship between sensory impairment and social interaction

Approximately 20% of the service users had qualitative deficits in social interaction (n=596). People of South Asian origin appeared to be less likely to have deficits in social interaction compared with white groups ($p<0.001$) and, again, older individuals appeared to be less likely to have deficits in social interaction (Table 6.16). Men were also more likely to have deficits in social interaction, but this relationship was found to be complex, as an interaction was observed between both gender and visual impairment and gender and degree of ID.

Table 6.16: Logistic regression model showing the relationship between poor quality of social interaction (outcome) and visual and hearing impairment after adjustment for potential confounders (n=2940)

Variable		OR	95% CI	p-value
Visual impairment:	None	1.00	(reference)	
	Partial	1.06	0.41 – 1.05	0.82
	Blind	0.94	0.53 – 2.56	0.70
Hearing impairment:	None	1.00	(reference)	
	Partial	0.66	0.41 – 1.05	0.08
	Deaf	1.17	0.53 – 2.56	0.70
Ethnicity:	White	1.00	(reference)	
	South Asian	0.56	0.42 – 0.76	<0.001
	Black	0.97	0.45 – 2.09	0.94
	Other/Unknown	1.54	0.75 – 3.13	0.24
Age group (years):	<30	1.00	(reference)	
	30–39	0.70	0.52 – 0.93	0.01
	40–49	0.78	0.59 – 1.04	0.10
	50+	0.70	0.54 – 0.92	0.01
Gender:	Male	3.89	1.96 – 7.70	<0.001
Degree of ID:	Mild	1.00	(reference)	
	Moderate	3.30	1.62 – 6.73	0.001
	Severe	5.90	3.03 – 11.47	<0.001
	Profound	26.47	13.77 – 50.91	<0.001
Interaction: Gender * Visual impairment				
Male	* Partial	0.46	0.23 – 0.92	0.03
	* Blind	0.74	0.27 – 2.05	0.56
Interaction: Gender * Degree of ID				
Male	* Moderate	0.31	0.13 – 0.71	0.006
	* Severe	0.33	0.15 – 0.71	0.004
	*Profound	0.28	0.13 – 0.60	0.001

CI: confidence interval; ID: intellectual disability; OR: odds ratio

Further analysis of the interactions revealed that men who did not have any visual impairment were more likely than their female counterparts to have deficits in social interaction. However, women with visual impairment were more

likely than men with visual impairment to have deficits in social interaction (Figure 6.17).

Similarly, men with mild ID were more likely than women with mild ID to have deficits in social interaction, but these gender differences were less pronounced as the person's degree of ID became more severe, and women with profound ID were more likely than men with profound ID to have deficits in social interaction (Figure 6.18).

A relationship was not observed between poor quality of social interaction and hearing impairment or visual impairment.

Figure 6.17: Graph of predicted probabilities for deficits in social interaction, showing the relationship between visual impairment and gender (n=2940)

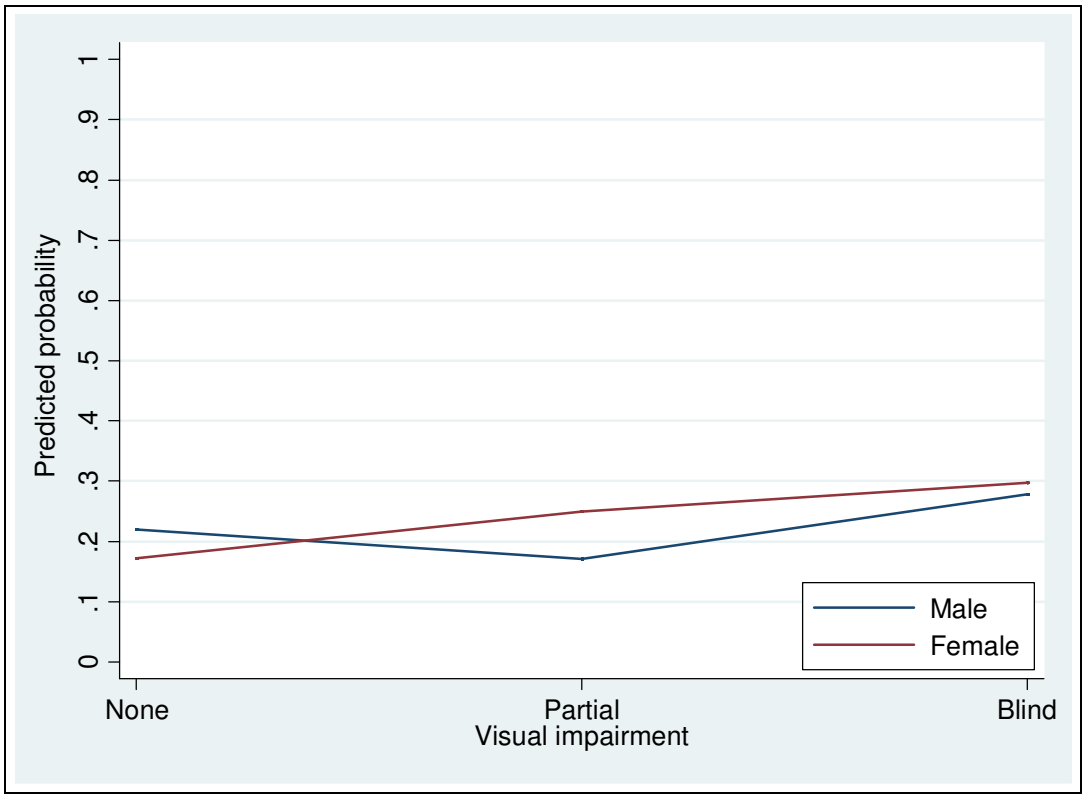
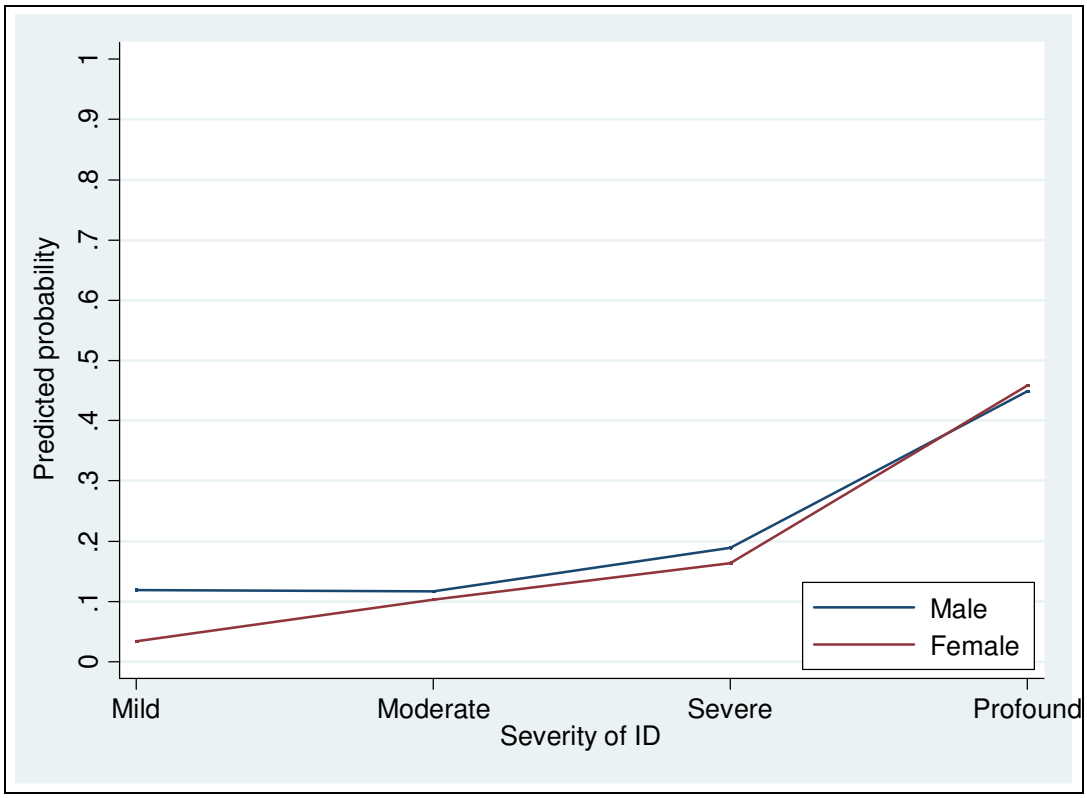


Figure 6.18: Graph of predicted probabilities for deficits in social interaction, showing the relationship between degree of ID and gender (n=2940)



ID: intellectual disability

6.5. Relationship between challenging behaviour and sensory impairment (Research Question 4)

In total, there were 2,056 (70%) service users with challenging behaviour of any type, as reported by their carers. People with ASD ($p<0.001$) and of unknown/other ethnicity ($p=0.03$) were more likely to display challenging behaviours. Neither hearing impairment nor gender were found to be associated with challenging behaviour (Table 6.19). An interaction was observed between both age group and visual impairment and age group and degree of ID.

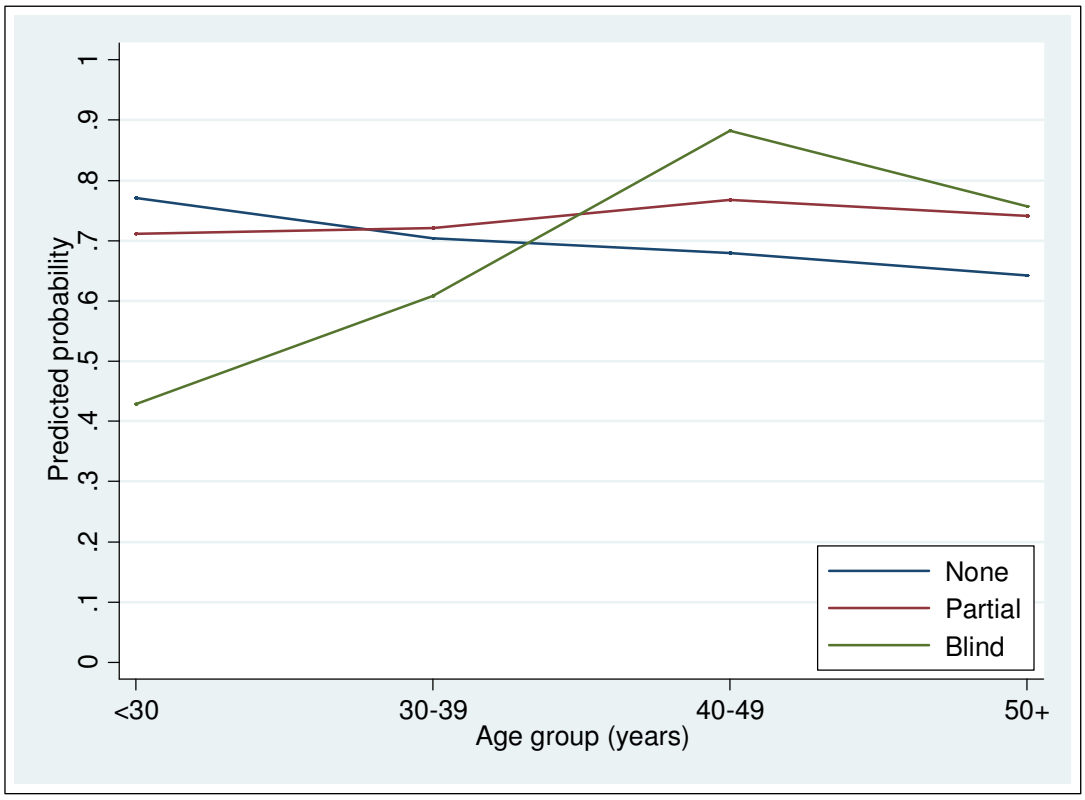
Table 6.19: Logistic regression model showing the relationship between challenging behaviour (outcome) and visual and hearing impairment after adjustment for potential confounders (n=2940)

Variable		OR	95% CI	p-value
Visual impairment:	None	1.00	(reference)	
	Partial	0.56	0.27 – 1.14	0.11
	Blind	0.10	0.03 – 0.35	<0.001
Hearing impairment:	None	1.00	(reference)	
	Partial	1.15	0.79 – 1.66	0.46
	Deaf	1.46	0.65 – 3.24	0.36
Ethnicity:	White	1.00	(reference)	
	South Asian	0.86	0.67 – 1.11	0.26
	Black	0.83	0.40 – 1.73	0.63
	Other/Unknown	3.31	1.12 – 9.78	0.03
Age group (years):	<30	1.00	(reference)	
	30–39	0.80	0.52 – 1.23	0.01
	40–49	0.65	0.41 – 1.04	0.10
	50+	0.51	0.33 – 0.80	0.01
Gender:	Male	1.13	0.96 – 1.34	0.15
Degree of ID:	Mild	1.00	(reference)	
	Moderate	2.00	1.26 – 3.18	0.003
	Severe	2.29	1.37 – 3.83	0.002
	Profound	2.40	1.31 – 4.39	0.005
ASD (4 or more traits):	Present	5.57	3.77 – 8.24	<0.001
Interaction: Age group * Visual impairment				
30–39 years	* Partial	1.76	0.68 – 4.55	0.24
	* Blind	3.61	0.75 – 17.31	0.11
40–49 years	* Partial	1.72	0.65 – 4.51	0.27
	* Blind	25.10	3.45 – 182.51	0.001
50+ years	* Partial	2.21	0.89 – 5.48	0.09
	* Blind	8.40	1.88 – 37.57	0.005
Interaction: Age group * Degree of ID				
30–39 years	* Moderate	0.85	0.44 – 1.62	0.62
	* Severe	0.99	0.50 – 1.95	0.98
	*Profound	0.85	0.38 – 1.90	0.69
40–49 years	* Moderate	0.50	0.26 – 0.98	0.04
	* Severe	1.13	0.56 – 2.27	0.73
	*Profound	1.69	0.73 – 3.92	0.22
50+ years	* Moderate	0.73	0.39 – 1.36	0.32
	* Severe	0.98	0.52 – 1.87	0.96
	*Profound	1.70	0.78 – 3.71	0.19

ASD: Autism Spectrum Disorder; CI: confidence interval; ID: intellectual disability; OR: odds ratio

The graphs of predicted probabilities for the interaction variables revealed that, in those without visual impairment, the likelihood of displaying challenging behaviour decreased with age (Figure 6.20). In contrast, in those with visual impairment, the probability of displaying challenging behaviours increased with age. This effect was particularly marked in people who were blind; they were the most likely to display challenging behaviours past the age of 40 years.

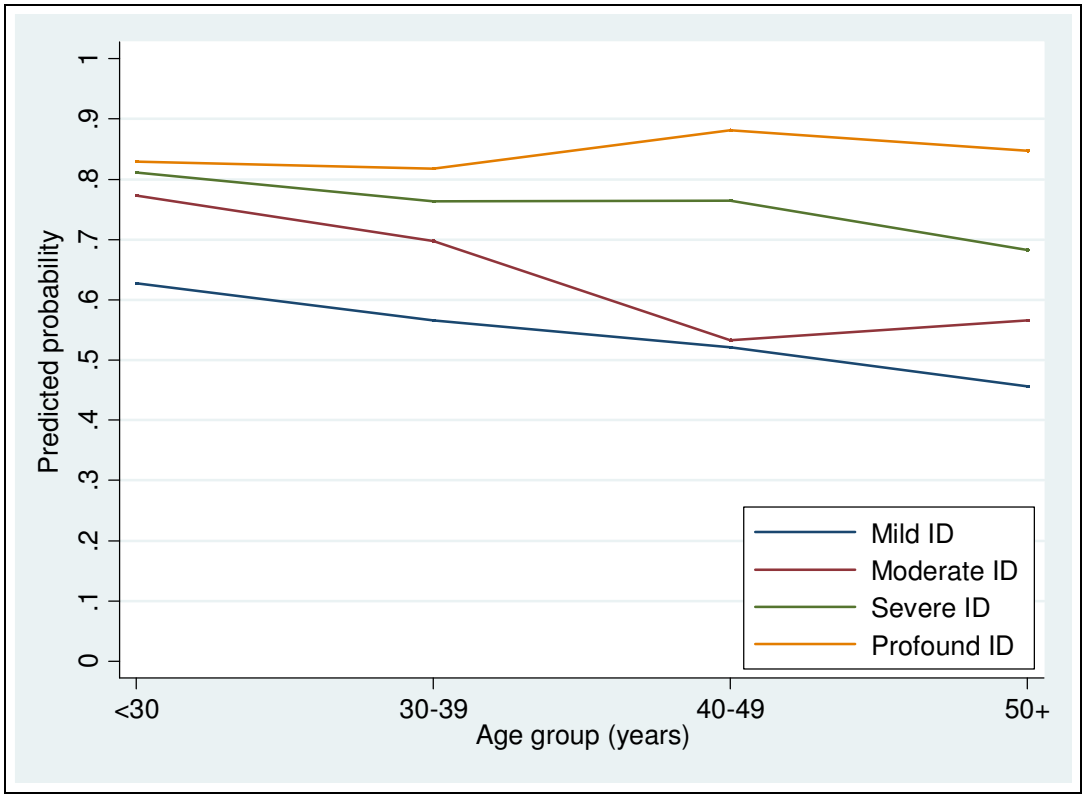
Figure 6.20: Graph of predicted probabilities for challenging behaviour, showing the relationship between age group and visual impairment (n=2940)



Similarly, challenging behaviour appeared to decrease with age in those with mild ID, whereas there was a marginal increase in challenging behaviour in those with profound ID. Differences between the predicted probabilities of

displaying challenging behaviours were most marked in the 50+ age group for all levels of ID (Figure 6.21).

Figure 6.21: Graph of predicted probabilities for challenging behaviour, showing the relationship between age group and degree of ID (n=2940)



ID: intellectual disability

7. RESULTS FROM STAGE 2: FACE-TO-FACE INTERVIEWS USING OBJECTIVE ASSESSMENT TOOLS

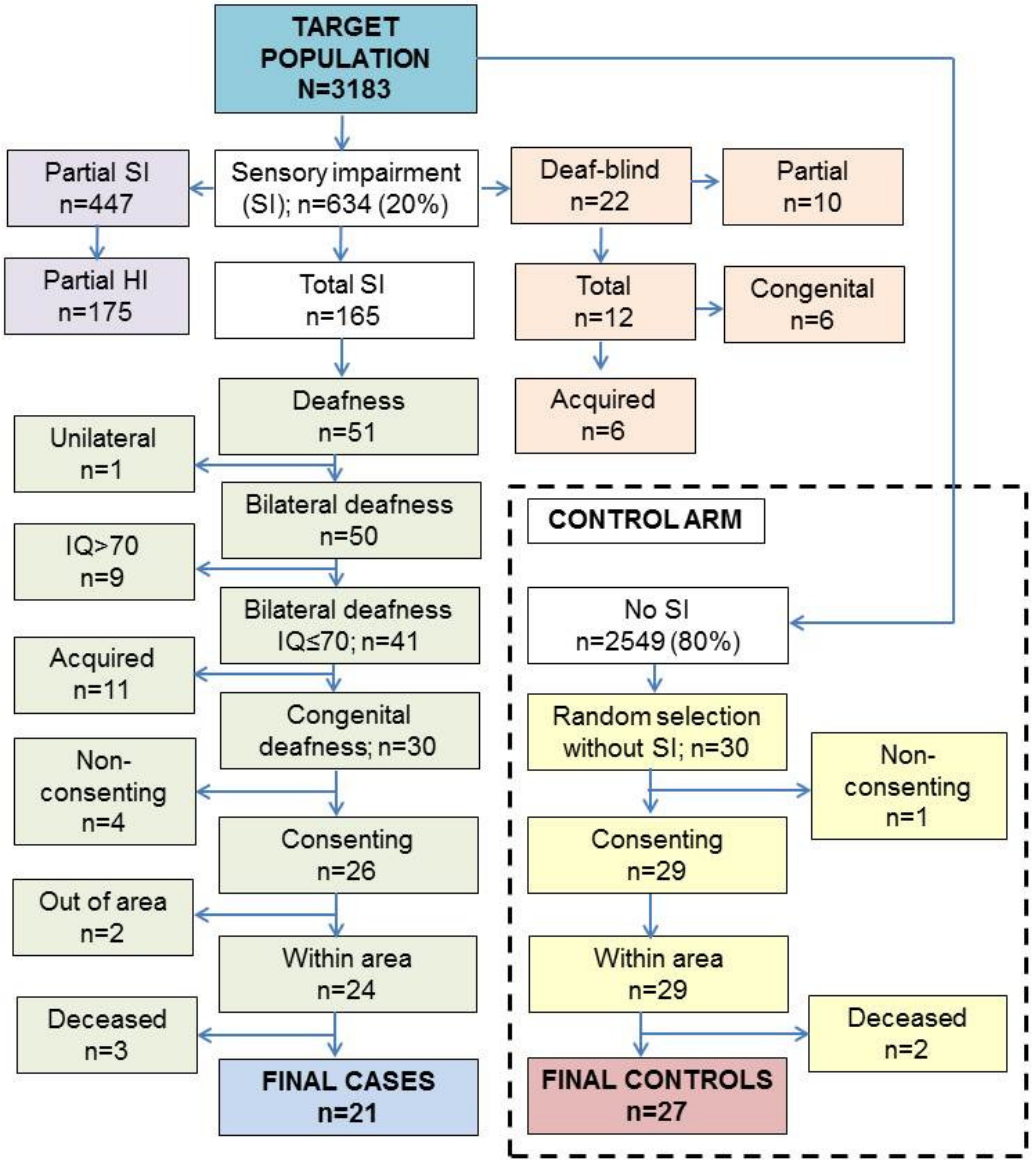
7.1. Introduction

Overall, 30 service users with a total congenital and bilateral deafness were included in the deaf subgroup and equal numbers of randomly selected controls, normally sighted and hearing, were matched to deaf cases by degree of ID and gender.

For the blind subgroup, 75 service users with a total congenital and bilateral blindness were included and these were complemented by equal numbers of those with normal sight and hearing as their controls who were electronically randomised and matched to blind cases by degree of ID and gender.

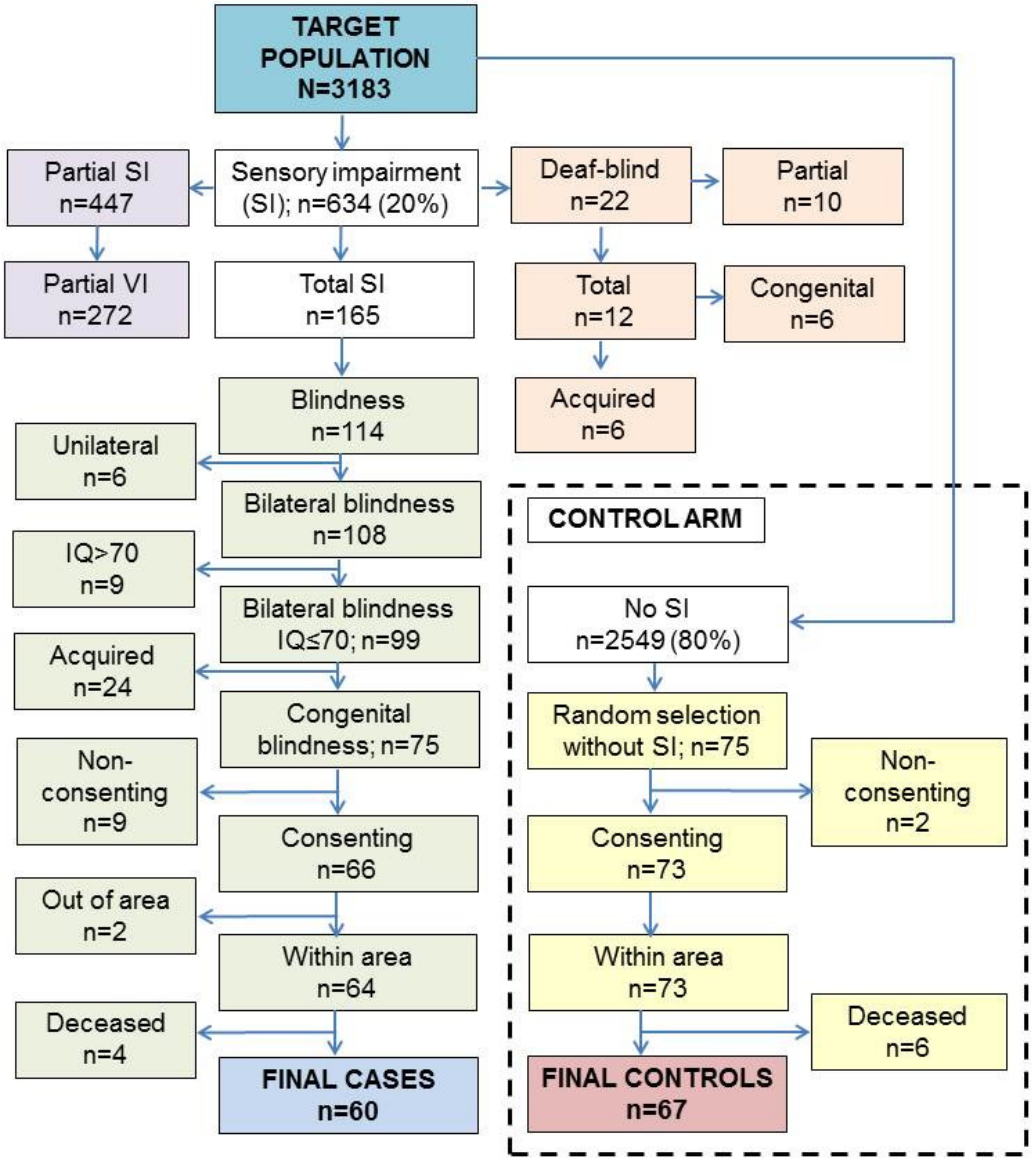
Figures 7.1 and 7.2 summarise the selection of cases and controls, exclusions and attrition rates in the deaf and blind subgroups. Owing to a high attrition rate, the final statistical analysis was carried out on 60 blind service users and 67 of their controls and on 21 deaf service users and 27 of their controls.

Figure 7.1: Flowchart showing the selection of cases and controls for the deaf subgroup



HI: hearing impairment; IQ: intelligence quotient; SI: sensory impairment

Figure 7.2: Flowchart showing the selection of cases and controls for the blind subgroup



IQ: intelligence quotient; SI: sensory impairment; VI: visual impairment

Attrition (n=35) occurred for the following reasons:

- Carers/families did not want to participate in the research.
  - Blind service users; n=9.
  - Controls in blind subgroup; n=2.
  - Deaf service users; n=4.
  - Controls in deaf subgroup; n=1.

- Service users were non-contactable because they had moved out of the county or to an unknown address.
  - Blind service users; n=2.
  - Deaf service users; n=2.

- Service users sadly died before being interviewed.
  - Blind service users; n=4.
  - Controls in blind subgroup; n=6.
  - Deaf service users; n=3.
  - Controls in deaf subgroup; n=2.

In total, 257 service users (163 with sensory impairment and 94 controls) were assessed to ensure that they all had confirmation of their degree of ID, degree of sensory impairment and information on whether these were unilateral, bilateral, partial, total, congenital or acquired. Only 175 (81 with sensory impairment and 94 in control groups) of these individuals were included in the final analyses, because they fulfilled the inclusion criteria for stage 2 of the research project.

Out of 82 service users who were assessed but, whose data were not included in the final analyses, 22 were deaf-blind, 6 had unilateral blindness and 1 had unilateral deafness. Nine deaf service users and 9 blind service users were found out to have borderline IQ. Eleven service users had acquired deafness and 24 had acquired blindness.

The results of the face-to-face interviews for those with a congenital total bilateral sensory impairment, using objective assessment tools discussed in the methodology, are discussed in detail in the next section.

7.2. Results from stage 2: deaf service users and their controls

7.2.1. Aetiology of ID and sensory impairment in cases with deafness (Research Question 5)

All cases with deafness were either born deaf or developed deafness in infancy (sensory-neural deafness). There was no aetiology of ID identified in 5 cases (24%). For the remaining cases (76%, n=16) the following conditions were determined as the cause of their ID or deafness:

- Down syndrome (n=4);
- Meningo-encephalitis during infancy (n=3);
- Extreme prematurity (n=2);
- Coffin Lowry syndrome (n=1);
- Congenital Rubella Syndrome (n=2);
- Halt Oram syndrome (n=1);
- Waardenburg syndrome (n=1);
- 18q deletion syndrome (n=1);
- **15q24 micro-deletion (n=1)**.

The genetic condition of one service user, in **bold** print (with 15q24 micro-deletion syndrome), was diagnosed after examination by the researcher and referral to the clinical genetics department for confirmation through genetic testing.

7.2.2. Aetiology of ID in the controls for the deaf subgroup

There was no information on the aetiology of ID for 16 service users (59%) in the controls for the deaf subgroup. Of those remaining (n=11), the aetiology of ID was as follows:

- Down syndrome (n=5);
- Brain damage as a result of complication of pregnancy/delivery: CP (n=3);
- Fragile X syndrome (n=1);
- 18q12.3 deletion (n=1);
- Tuberous sclerosis (n=1).

These aetiologies were all known before the start of the project (Appendix 15).

7.2.3. Demographic characteristics of deaf service users and their controls

Table 7.3 shows the demographic characteristics of the 21 deaf service users and their controls (n=27). The majority of the service users in the deaf subgroup had moderate to severe ID (followed by profound and mild ID respectively), were white and male. The average (mean) age of deaf subgroup as a whole was 53.9 years.

Of the 21 deaf service users, only 53% (n=12) had received input from the medical team at specialist adult ID services. This compared with slightly large numbers of controls, 63% (n=17).

None of the service users in the deaf subgroup were married. Two deaf service users and 2 controls had supported employment. Four deaf service users and 4 controls were not in touch with any family members because they were either abandoned by their families during infancy or lost touch after first-degree relatives, mainly parents, passed away.

Table 7.3: Demographic characteristics of service users with deafness and their controls (n=48)

	Deaf (n=21)		Hearing (n=27)	
Demographic data	**N**	**(%)**	**N**	**(%)**
Gender:				
Male	17	(81.0)	21	(77.8)
Female	4	(19.0)	6	(22.2)
Age; mean (±SE)	51.3	(±2.27)	55.9	(±2.48)
Degree of ID:				
Mild (IQ≤70)	2	(9.5)	3	(11.1)
Moderate (IQ<55)	7	(33.3)	10	(37.0)
Severe (IQ<35)	7	(33.3)	9	(33.3)
Profound (IQ<20)	5	(23.8)	5	(18.5)
Aetiology of ID known	16	(76.2)	11	(40.7)
Ethnicity:				
White	17	(81.0)	26	(96.3)
Asian	2	(9.5)	1	(3.7)
Black	1	(4.8)	0	(-)
Mixed	1	(4.8)	0	(-)
Accommodation:				
Living independently	1	(4.8)	0	(-)
Family home	5	(23.8)	4	(14.8)
Supported living	1	(4.8)	5	(18.5)
Residential home	14	(66.7)	18	(66.7)
Total	**21**	**(100.0)**	**27**	**(100.0)**

ID: intellectual disability; IQ: intelligence quotient; SE: standard error

Information on positive family history of ID was available for 3 of the deaf cases, but in none in the controls, suggesting the possibility of a genetic syndrome in the former as an aetiological factor.

Over two-thirds of the deaf service users and controls (67%) were living in a residential home. One deaf service user was living independently. The remaining deaf service users and controls were living in either family homes or supported living accommodation.

More than half of the deaf service users (57%, n=12) had never received any input from a charity organisation, such as RNID, despite having a major sensory deficit.

All but one of the deaf service users had been prescribed hearing aids, but only 8 were compliant in using them. More than half of the deaf service users (n=11) were able to use a basic form of sign language e.g. Makaton, Signalong, Sign English or idiosyncratic signing. Only one of the controls could use a basic sign language.

Seventy-one per cent of the deaf service users (n=15) had been prescribed spectacles for refractive errors, of whom 7 were compliant in using them. This compares with 44.5% of the controls (n=12), all of whom were compliant in wearing spectacles. All of the controls in the deaf subgroup were regularly monitored for their eyesight and hearing. However, three deaf service users did not have any follow-up arrangements for their sensory impairment.

7.2.4. Co-morbid conditions among deaf service users and their controls

Information on ICD-10 clinical diagnoses of various conditions was extracted from service users' medical case files, electronic data records and their GP referral letters or primary care summary sheets, in addition to directly probing carers for information on any co-morbidities. The co-occurrence of various medical and psychiatric problems, which needed medical and psychiatric attention, was common in deaf service users and their controls.

None of the service users had substance misuse or alcohol difficulties, but one deaf service user and one control (both with a mild degree of ID) had challenging behaviour of sexually inappropriate type, but only the service user with deafness had been through the criminal justice system for this behaviour. Apart from challenging behaviour, mood disorders were the most common co-morbid psychiatric problems followed by psychosis and anxiety disorders in both deaf service users and controls. Aggression and self-injurious behaviours

were the most common types of challenging behaviour requiring referral to specialist services for further input.

Tables 7.4 and 7.5 show the breakdown of co-morbid psychiatric and medical disorders in deaf service users and their controls based on ICD-10 diagnostic criteria collected from the medical case files and electronic data records.

Table 7.4: Prevalence of co-morbid* psychiatric conditions and challenging behaviour requiring treatment among deaf service users and their controls based on ICD-10 clinical diagnosis recorded in the medical case files and electronic data records (n=48)

	Deaf (n= 21)		Hearing (n=27)	
Psychiatric disorders	**N**	**(%)**	**N**	**(%)**
Aggression	13	(61.9)	19	(70.4)
Self-injury	3	(14.3)	12	(44.5)
Antisocial behaviour	3	(14.3)	0	(-)
Depressive disorder	3	(14.3)	1	(3.7)
Psychotic disorder	2	(9.5)	2	(7.4)
Bipolar affective disorder	1	(4.8)	2	(7.4)
Anxiety disorder	1	(4.8)	3	(11.1)
Personality disorder	1	(4.8)	1	(3.7)
PTSD	1	(4.8)	0	(-)
Forensic history	1	(4.8)	0	(-)
Sexually inappropriate behaviour	1	(4.8)	1	(3.7)

ICD-10: International Classification of Diseases-10th Revision; PTSD: post-traumatic stress disorder as a result of abuse during childhood

*Some service users had more than one condition.

With regard to physical health co-morbidities, speech difficulties, constipation, gastro-oesophageal reflux (GER), motor problems and diabetes were more common in the deaf service users. Conversely, skin/hair diseases, incontinence, vitamin deficiencies, hay fever, swallowing difficulties, ischaemic heart disease and hypothyroidism were more common in the controls.

Chronic pain, high lipid profiles, smoking and hyper-uricaemia were seen in similar rates in both cases and controls.

Table 7.5: Prevalence of co-morbid physical conditions requiring generic or specialist input among deaf service users and their controls (n=48)

	Deaf (n= 21)		Hearing (n=27)	
Co-morbid physical health problem	**N**	**(%)**	**N**	**(%)**
Speech difficulties	21	(100.0)	8	(29.6)
Constipation	7	(33.3)	6	(22.2)
PUD/GER	5	(23.8)	5	(18.5)
Motor problem	5	(23.8)	1	(3.7)
Vitamin D or B12 deficiency	3	(14.3)	7	(25.9)
Chronic pain*	3	(14.3)	4	(14.8)
High lipid profile	3	(14.3)	4	(14.8)
Smoking	3	(14.3)	4	(14.8)
Skin or hair diseases	2	(9.5)	9	(33.3)
Incontinence	2	(9.5)	5	(18.5)
Hypothyroidism	2	(9.5)	3	(11.1)
Diabetes Mellitus	2	(9.5)	0	(-)
Swallowing difficulties	1	(4.8)	4	(14.8)
Hay fever	1	(4.8)	4	(14.8)
IHD	1	(4.8)	2	(7.4)
Taking vitamin and food supplements	1	(4.8)	2	(7.4)
Hyperuricemia	1	(4.8)	1	(3.7)

GER: gastro-oesophageal reflux; IHD: ischaemic heart disease; PUD: peptic ulcer disease

*Requiring prescription of regular analgesics

7.2.5. Prevalence of mental illness, ASD, challenging behaviour, epilepsy and prescribed medication in deaf service users and their controls

The co-morbid diagnoses of mental illness, ASD (as measured using PDD-MRS), challenging behaviour (as measured using ABC) and epilepsy and prescribed medication (psychotropic, antipsychotic and medication for physical health problems) among deaf service users and their controls were compared using the chi-squared test (Table 7.6).

Approximately similar numbers of deaf service users and controls had a diagnosis of ASD or mental illness. Although controls were diagnosed more with epilepsy, challenging behaviour and physical health problems, these differences were not statistically significant.

Table 7.6: Comparison of deaf service users and controls by rates of ASD, mental illness, challenging behaviour, epilepsy and prescribed medication (n=48)

Variables	Deaf (n=21)	Hearing (n=27)	Pearson Chi^2	Chi^2 p-value
ASD (PDD-MRS)	8 (38.1%)	11 (40.7%)	0.03	0.85
Mental illness (ICD-10)	9 (42.9%)	10 (37.0%)	0.17	0.68
Challenging behaviour (ABC)	11 (52.4%)	18 (66.7%)	1.01	0.32
Epilepsy	3 (14.3%)	7 (25.9%)	0.97	0.33
Antipsychotic medication	9 (42.9%)	14 (51.9%)	.0.38	0.54
Other psychotropic medication	11 (52.4%)	13 (48.2%)	0.08	0.77
Medication for physical health	5 (23.8%)	11 (40.7%)	1.52	0.22

ABC: Aberrant Behaviour Checklist; ASD: Autism Spectrum Disorder; ICD-10: International Classification of Diseases-10th Revision; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale

7.2.6. Differences in diagnostic methods used to identify ASD (Research Question 6)

The presence of ASD based on three different assessment methods was compared in deaf service users and their controls (Table 7.7). Information on autistic traits (as described in the previous chapter) from the LLDR database was available for all 48 service users in the deaf and hearing groups. Information on clinical diagnosis of autism was available on 41 service users in the deaf and hearing groups.

Table 7.7: Rate of identifying ASD based on different assessment methods in deaf service users and their controls

	4 or more traits from the LLDR	Clinical diagnosis	PDD-MRS
Deaf	5/21 (23.8%)	0/18 (-)	8/21 (38.1%)
Hearing	4/27 (14.8%)	6/23 (26.1%)	11/27 (40.7%)
Total	**9/48 (18.8%)**	**6/41 (14.6%)**	**19/48 (39.6%)**

ASD: Autism Spectrum Disorder; ICD-10: International Classification of Diseases-10th Revision; LLDR: Leicestershire Learning Disability Register; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale

Using PDD-MRS, considered to be the most accurate method, rendered higher rates of identifying ASD in both deaf service users and controls (40%), followed by using 4 or more traits (as described in the previous chapter) and ICD-10 clinical assessment. Clinicians had missed a co-morbid ASD in all who were deaf. Of the 19 people with ASD (as measured by PDD-MRS), only 4 (21%) also had 4 or more carer-reported traits on the register. Using Cicchetti's criteria for interpreting Cohen's kappa statistics (1994) as 0.75–1.00=excellent, 0.60–0.74=good, 0.40–0.59=fair and <0.40=poor, there was poor agreement between identifying ASD using 4 or more traits from the LLDR (kappa=0.042) and identifying ASD using an objective assessment tool (PDD-MRS) administered by a trained professional (Table 7.8). Similarly, agreement between identifying ASD using an objective assessment tool (PDD-MRS) and identifying ASD using

ICD-10 clinical criteria was also poor (kappa=0.39) (Table 7.9), although agreement was significantly higher than chance alone.

Table 7.8: Kappa agreement between identifying ASD using PDD-MRS and 4 or more autistic traits on the LLDR in deaf service users and their controls (n=48)

Agreement	Expected agreement	Kappa	Standard error	Z	p-value
58.3%	56.5%	0.042	0.127	0.33	0.37

ASD: Autism Spectrum Disorder; LLDR: Leicestershire Learning Disability Register; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale

Table 7.9: Kappa agreement between identifying ASD using PDD-MRS and clinical criteria in deaf service users and their controls (n=41)

Agreement	Expected agreement	Kappa	Standard error	Z	p-value
73.1%	56.0%	0.390	0.124	3.15	<0.001

ASD: Autism Spectrum Disorder; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale

7.2.7. Prevalence of autistic traits in deaf service users and their controls using the Leicestershire Learning Disability Register (LLDR) database

Service users who were deaf did not differ from their hearing counterparts in terms of the rate of occurrence of carer-reported autistic traits on the LLDR (Table 7.10). In the hearing group, lack of empathy was the most common trait, followed by presence of elaborate routines and stereotypies. In the deaf group, presence of elaborate routines was the most common trait, followed by poor use of speech and deficits in empathy. The results were similar (statistically non-significant) when rates of autistic traits were compared between autistic deaf and autistic hearing service users (Table 7.11).

Table 7.10: Comparison of autistic traits in deaf service users and controls by presence of traits on the LLDR (n=48)

Autistic traits	Deaf (n=21)	Hearing (n=27)	Pearson Chi^2	Chi^2 p-value
Stereotypies	8 (38.1%)	13 (48.2%)	0.49	0.49
Deficits in empathy	9 (42.9%)	15 (55.6%)	0.76	0.38
Elaborate routines	14 (66.7%)	14 (51.9%)	1.07	0.30
Poor use of speech	13 (61.9%)	12 (44.4%)	1.44	0.23
Poor quality of social interaction	3 (14.3%)	7 (25.9%)	0.97	0.33

LLDR: Leicestershire Learning Disability Register

Table 7.11: Comparison of autistic traits in autistic* deaf service users and autistic* controls by presence of traits on the LLDR (n=19)

Autistic traits	Autistic deaf (n=8)	Autistic hearing (n=11)	Pearson Chi^2	Chi^2 p-value
Stereotypies	4 (50.0%)	8 (72.7%)	1.03	0.31
Deficits in empathy	5 (62.5%)	7 (63.6%)	0.003	0.96
Elaborate routines	6 (75.0%)	7 (63.6%)	0.28	0.60
Poor use of speech	5 (62.5%)	6 (54.6%)	0.12	0.73
Poor quality of social interaction	1 (12.5%)	4 (36.4%)	1.36	0.24

LLDR: Leicestershire Learning Disability Register; *Defined using PPD-MRS

7.2.8. Relationship between ASD and congenital deafness (Research Question 7)

When deaf service users and their controls were compared using conditional logistic regression modelling (to take account of matching), the controls had 1.33 times higher odds of having ASD (based on the PDD-MRS) compared with the deaf service users, but this was not statistically significant (p=0.71), and the confidence interval contained the null value of 1 (Table 7.12).

Table 7.12: Conditional logistic regression showing the relationship between ASD (outcome as measured by PDD-MRS) and deafness

Variable	OR	SE	95% CI	p-value
Hearing (vs deafness)	1.33	0.764	0.30 – 5.96	0.71

ASD: Autism Spectrum Disorder; CI: confidence interval; OR = odds ratio; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale; SE: standard error

Table 7.13 shows the results from the single-variable logistic regression conducted. The table shows that being male and having epilepsy raised the odds of having co-morbid ASD (ORs of 3.2 and 1.7 respectively), but the 95% confidence intervals and the p-values were not statistically significant. The odds of having ASD in the background of deafness was less than 1 but again the p-value and 95% confidence intervals were not statistically significant. In the multi-variable logistic regression model (Table 7.14), the lower odds of deafness and higher odds of having ASD in men were more pronounced, but again, this did not reach statistical significance.

Table 7.13: Single-variable logistic regression showing the crude relationship between ASD (outcome as measured by PDD-MRS), deafness, gender, degree of ID and epilepsy (n=48)

Variable	OR	SE	95% CI	p-value
Deafness	0.90	0.53	0.28 – 2.88	0.85
Gender (Male)	3.24	2.77	0.61 – 17.31	0.17
Age (years)	0.98	0.02	0.94 – 1.03	0.50
Degree of ID:				
Mild and moderate	0.38	0.30	0.08 – 1.78	0.22
Severe	1.00	0.81	0.21 – 4.86	1.00
Profound	1.00	-	(reference)	-
Epilepsy	1.71	1.23	0.42 – 6.98	0.45

ASD: Autism Spectrum Disorder; CI: confidence interval; ID: intellectual disability; OR: odds ratio; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale; SE: standard error

Table 7.14: Multi-variable logistic regression showing the relationship between ASD (outcome as measured by PDD-MRS), deafness, gender, degree of ID and epilepsy (n=48)

Variable	OR	SE	95% CI	p-value
Deafness	0.73	0.49	0.20 – 2.70	0.64
Gender (Male)	3.70	3.52	0.57 – 23.86	0.17
Age	0.98	0.03	0.92 – 1.04	0.43
Degree of ID:				
Mild and moderate	0.29	0.26	0.05 – 1.68	0.17
Severe	1.25	1.11	0.22 – 7.16	0.80
Profound	1.00	-	(reference)	-
Epilepsy	1.13	0.91	0.24 – 5.44	0.87

ASD: Autism Spectrum Disorder; CI: confidence interval; ID: intellectual disability; OR: odds ratio; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale; SE: standard error

7.3. Results from stage 2: blind service users and their controls

The attrition rate in people with blindness group was 20% (n=15). This included those who did not consent or whose carers did not agree to participation, could not be located or who died before being assessed by the researcher. In the control group, 12% (n=8) could not be seen because they did not consent or their carers did not agree to participation, they moved to an unknown address or they died before assessment.

7.3.1. Aetiology of ID in cases with blindness (Research Question 5)

Twelve blind service users and 16 of their controls were not open to the medical team of the local adult ID services. The reasons for blindness were quite varied e.g. absence or underdeveloped eyes (anophthalmia or microphthalmia), cortical blindness, abnormal eye structures such as aniridia, optic atrophy, congenital bilateral cataract, retinal dystrophy e.g. Leber's congenital amaurosis, coloboma, retinopathy, sclera pupil, infantile glaucoma, either alone or in combination.

In 27% of the service users with blindness (n=16), no background aetiology of ID could be found, despite suspecting a condition such as TORCHES (Toxoplasmosis, Other infections, Rubella virus, Cytomegalovirus, Herpes Simplex virus, Syphilis) or a genetic syndrome, and carrying out detailed investigations. In the remaining cases (n=44; 73%) with blindness, the underlying aetiologies of ID in order of the frequency were as follows:

- Extreme Prematurity (n=7);
- Brain damage or CP due to peri-natal complication e.g. hypoxia, brain haemorrhage (n=6);
- Meningo-encephalitis with or without hydrocephalus (n=5);
- TORCHES (mainly congenital rubella syndrome; n=4 and one case of congenital Toxoplasmosis);
- Laurence-Moon-Bardet-Biedl Syndrome (n=2);
- Osteogenesis Imperfecta (n=2);
- Down syndrome (n=2).

The rest of the cases had one of the below conditions, of which some are extremely rare. The conditions in **bold** print were diagnosed after the researcher suspected a genetic syndrome and made a referral to the clinical genetics department of the Leicester Royal Infirmary for confirmation (Appendix 15):

- **Progressive Encephalopathy with Oedema, Hypsarrythmia and Optic atrophy** (PEHO, n=1);
- **Mitochondrial genetic condition** (n=1);
- Shaken baby syndrome (n=1);
- ID due to complication of Renal Tubular Acidosis during infancy (RTA) (n=1);
- Infantile spasm (n=1);
- Norrie disease (n=1);
- Cohen syndrome (n=1);
- Sturge Weber syndrome (n=1);
- Joubert syndrome (n=1);

- Hypoglycaemic brain damage during infancy (n=1);
- Congenital Adrenal Hyperplasia (CAH) (n=1);
- Septo-optic dysplasia with pan hypopituitarism (n=1);
- Batten disease (neuronal ceroid lipofuscinosis) (n=1);
- Mucopolysaccharidosis (n=1);
- Beta thalassaemia complicated with hydrocephalus during infancy (n=1).
- One service user was suspected of an X-linked recessive disorder but this could not be confirmed despite extensive genetic testing.

For the remaining cases of blindness, the aetiology of ID was not known; this is in contrast to the literature available which stipulates that, in most cases of severe to profound ID, the underlying aetiology could be identified.

7.3.2. Aetiology of ID in the controls for the blind subgroup

For 57% of the control group (n=38) the aetiology of the ID was unknown. The rest (n=29, 43%) had a confirmed diagnosis of the following conditions:

- Down syndrome (n=9);
- Brain damage due to birth complications i.e. CP (n=5);
- Meningoencephalitis (n=3);
- Angelman syndrome (n=2);
- Sturge Weber syndrome (n=2);
- Rett syndrome (n=2).

Other conditions were:

- Klinefelter syndrome (n=1);
- Infantile spasm (n=1);
- Tuberous sclerosis (n=1);
- Cri du chat syndrome (n=1);
- Cornelia de lange syndrome (n=1); and
- Lennox-Gastaut syndrome (n=1).

Infantile spasm and Lennox-Gastaut syndromes are mainly epilepsy syndromes secondary to a variety of underlying genetic or metabolic causes. Therefore, in these cases there is a high suspicion that the underlying causes could not be ascertained in spite of various investigations and, as such, these epilepsy syndromes have been reported as the aetiology of ID.

7.3.3. Demographic characteristics

For 5 blind cases and 15 controls, families were not in touch with the service users at all. There was a positive family history of blindness/ID in 6 of the blind service users compared with only one of the controls who had a family history of ID. Table 7.15 provides more details on the demographic characteristics of the 60 congenitally blind service users and their controls (n=67).

Table 7.15: Demographic characteristics of service users with blindness and their controls

	Blind (n=60)		Sighted (n=67)	
Demographic data	**N**	**(%)**	**N**	**(%)**
Gender:				
Male	42	(70.0)	42	(62.7)
Female	18	(30.0)	25	(37.3)
Age; mean (±SE)	45.3	(±1.73)	46.0	(±1.64)
Degree of ID:				
Mild (IQ≤70)	4	(6.7)	4	(6.0)
Moderate (IQ<55)	3	(5.0)	3	(4.5)
Severe (IQ<35)	18	(30.0)	23	(34.3)
Profound (IQ<20)	35	(58.3)	37	(55.2)
Aetiology of ID known	44	(73.3)	29	(43.3)
Marital status	1	(1.7)	0	(-)
Ethnicity:				
White	46	(76.7)	64	(95.5)
Asian	12	(20.0)	3	(4.5)
Black	1	(1.7)	0	(-)
Mixed	1	(1.7)	0	(-)
Accommodation:				
Living independently	1	(1.7)	2	(3.0)
Family home	21	(35.0)	12	(17.9)
Supported living	10	(16.7)	23	(34.3)
Residential home	28	(46.7)	30	(44.8)
Total	**60**	**(100.0)**	**67**	**(100.0)**

ID: intellectual disability; IQ: intelligence quotient; SE: standard error

The study sample was representative of people with more severe ID, with around 90% of blind service users and their controls having severe and profound ID.

The majority of blind service users and their controls were male and the average age of the study population was 46 years old. Interestingly, more of the

blind service users were from ethnic minority backgrounds compared with the controls (23.4% vs 4.5%), primarily South Asian backgrounds. None of the service users could work and only one with mild ID in the blind group was married. The majority of blind service users and controls lived in residential or supported living accommodation. The proportion of people living within the family home, however, was nearly twice as high in the blind service users compared with the controls.

7.3.4. Co-morbid conditions among blind service users and their controls

The most common types of mental health issues based on ICD-10 clinical criteria, experienced by both blind service users and their controls, were challenging behaviours of aggressive and self-injurious types, followed by mood disorders, and then psychotic and anxiety disorders; these were generally seen more frequently in the sighted group than the blind service users.

There were no reports of forensic history or drug and alcohol misuse in any of the service users, but there were 2 cases of personality disorders and one case of ADHD, Tourette syndrome and post-traumatic stress disorder (PTSD) reported only in the controls (Table 7.16).

Table 7.16: Prevalence of co-morbid* psychiatric conditions and challenging behaviour requiring treatment among blind service users and their controls based on ICD-10 clinical diagnosis recorded in the medical case files and electronic data records (n=127)

	Blind (n=60)		Sighted (n=67)	
Psychiatric disorders	**N**	**(%)**	**N**	**(%)**
Self-injury	25	(41.7)	29	(43.3)
Aggression	22	(36.7)	37	(55.2)
Depressive disorder	4	(6.7)	8	(11.9)
Bipolar affective disorder	2	(3.3)	6	(9.0)
Anxiety disorder	2	(3.3)	2	(3.0)
Psychotic disorder	1	(1.7)	3	(4.5)
Personality disorder	0	(-)	2	(3.0)
PTSD	0	(-)	1	(1.5)
ADHD	0	(-)	1	(1.5)
Tourette syndrome	0	(-)	1	(1.5)
Forensic history	0	(-)	0	(-)
Sexually inappropriate behaviour	0	(-)	0	(-)
Antisocial behaviour	0	(-)	0	(-)

ADHD: attention deficit hyperactive disorder; ICD-10: International Classification of Diseases-10th Revision; PTSD: post-traumatic stress disorder as a result of childhood abuse

*Some service users had more than one condition.

Co-morbid medical conditions were common in both blind service users and controls, with very similar rates except for some differences in the prevalence of constipation, poorly controlled epilepsy, anaemia, gynaecological problems, hypertension, chronic pains, skin and hair disorders and spasticity, which were more commonly seen in the cases than controls. In contrast, speech and swallowing difficulties, asthma and allergic conditions (asthma and hay fever), vitamin deficiencies and issues related to weight loss were more common in the controls. None of the blind service users smoked or had been diagnosed with

Parkinson disease while some of the controls were smokers and had a diagnosis of Parkinson disease (Table 7.17).

While all of the controls had regular follow up for their sight, 25 (41.7%) of blind service users were lost to ophthalmology or optician follow up. Seventeen of the blind service users (28.3%) were not open to RNIB or VISTA in spite of having a major sensory deficit.

Table 7.17: Prevalence of co-morbid physical conditions requiring generic or specialist input among blind service users and their controls (n=127)

	Blind (n=60)		Sighted (n=67)	
Co-morbid physical health problem	**N**	**(%)**	**N**	**(%)**
Constipation	33	(55.0)	26	(38.8)
Speech difficulties	32	(53.3)	43	(64.2)
Chronic pain*	21	(35.0)	20	(29.9)
Skin or hair diseases	20	(33.3)	20	(29.9)
PUD/GER	18	(30.0)	19	(28.4)
Swallowing difficulties†	16	(26.7)	22	(32.8)
Epilepsy, poorly controlled	16	(26.7)	13	(19.4)
Vitamin D or B12 deficiency	15	(25.0)	20	(29.9)
Anaemia	10	(16.7)	6	(9.0)
Hypertension	10	(16.7)	7	(10.4)
Hay fever	7	(11.7)	9	(13.4)
Hypothyroidism	7	(11.7)	7	(10.4)
Diabetes Mellitus	7	(11.7)	5	(7.5)
Hormone therapy for gynaecological problems	7	(11.7)	3	(4.5)
On food supplements for weight loss	6	(10.0)	8	(11.9)
High lipid profile	6	(10.0)	6	(9.0)
Spasticity	5	(8.3)	1	(1.5)
Asthma	4	(6.7)	8	(11.9)
IHD	2	(3.3)	2	(3.0)
Osteoporosis	2	(3.3)	1	(1.5)
Hyper-uricaemia	1	(1.7)	0	(-)
Smoking	0	(-)	4	(6.0)
Parkinson disease	0	(-)	3	(4.5)

GER: gastro-oesophageal reflux; IHD: ischaemic heart disease; PUD: peptic ulcer disease

*Requiring prescription of regular analgesics

†Needed input from speech and language therapist

7.3.5. Prevalence of mental illness, ASD, challenging behaviour, epilepsy and prescribed medication in blind service users and their controls

The co-morbid mental illness (based on ICD-10), ASD (based on PDD-MRS), challenging behaviour (based on ABC), epilepsy and prescribed medication (psychotropic, antipsychotic and medication for physical health problems) in the blind service users and their controls were compared using the chi-squared test (Table 7.18). Prevalence of ASD and epilepsy were higher among blind service users and these differences were statistically significant (p=0.007 and p=0.03 respectively). Mental illness was higher in the controls and challenging behaviour was higher in blind service users, but these findings were not significant at the 5% level.

Table 7.18: Comparison of blind service users and controls by rates of ASD, mental illness, challenging behaviour, epilepsy and prescribed medication (n=127)

Variables	Blind (n=60)	Sighted (n=67)	Pearson Chi^2	Chi^2 p-value
ASD (based on PDD-MRS)	46 (76.7%)	36 (53.7%)	7.27	0.007
Mental illness (ICD-10)	8 (13.3%)	16 (23.9%)	2.30	0.13
Challenging behaviour (ABC)	41 (68.3%)	40 (59.7%)	1.02	0.31
Epilepsy	36 (60.0%)	27 (40.3%)	4.91	0.03
Motor problems	28 (46.7%)	22 (32.8%)	2.54	0.11
Asthma/Allergy	22 (36.7%)	28 (41.8%)	0.35	0.56
Metabolic problems	25 (41.7%)	21 (31.3%)	1.46	0.23
Incontinence	26 (43.3%)	29 (43.3%)	0.00	1.00
Any other medical problem	35 (58.3%)	38 (56.7%)	0.03	0.85
Antipsychotic medication	25 (41.7%)	31 (46.3%)	0.27	0.60
Other psychotropic medication	24 (40.0%)	35 (52.2%)	1.91	0.17

ABC: Aberrant Behaviour Checklist; ASD: Autism Spectrum Disorder; ICD-10: International Classification of Diseases-10th Revision; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale

7.3.6. Differences in diagnostic methods used to identify ASD (Research Question 6)

The identification of ASD based on three different assessment methods was compared between blind service users and their controls (Table 7.19). As with the deaf subgroup, differences in numbers can be attributed to availability of service users for assessment by the researcher and availability of medical records of service users open to the medical team of the Leicestershire adult ID service.

The most accurate diagnostic method was considered to be the objective assessment tool (PDD-MRS): 64.5% of people in the blind subgroup were diagnosed with ASD using this method. Autistic traits from the register (4 or more traits, as defined in the previous chapter) identified 39.2% of people as having autism. Clinical diagnosis (ICD-10 criteria) identified 25% of people as having autism (Table 7.19).

Table 7.19: Rates of identifying ASD based on different assessment methods in blind service users and their controls

	4 or more traits from the LLDR	Clinical diagnosis	PDD-MRS
Blind	18/59 (30.5%)	10/49 (20.4%)	46/60 (76.7%)
Sighted	31/66 (47.0%)	18/63 (28.6%)	36/67 (53.7%)
Total	**49/125 (39.2%)**	**28/112 (25.0%)**	**82/127 (64.5%)**

ASD: Autism Spectrum Disorder; LLDR: Leicestershire Learning Disability Register; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale

Although there was agreement between identifying ASD using the objective tool (PDD-MRS) and using 4 or more traits on the LLDR and clinical autism diagnoses (62% and 61% respectively), the level of agreement was relatively poor (kappa = 0.28 and 0.29 respectively; Cicchetti, 1994) (Tables 7.20 and 7.21). Of the 82 people with ASD (as measured by PDD-MRS), 81 had

complete data set on the LLDR, of which 41 (51%) had 4 or more carer-reported traits. Of interest, only 38% of those with ASD who were blind also had 4 or more traits, compared with 67% of those with ASD who were not blind, perhaps suggesting that traits may be under-reported, not observed, attributed to blindness by the carers or present differently in the blind population.

Table 7.20: Kappa agreement between identifying ASD using PDD-MRS and 4 or more traits on the LLDR in blind service users and their controls (n=125)

Agreement	Expected agreement	Kappa	Standard error	Z	p-value
61.6%	44.8%	0.278	0.08	3.55	<0.001

ASD: Autism Spectrum Disorder; LLDR: Leicestershire Learning Disability Register; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale

Table 7.21: Kappa agreement between identifying ASD using PDD-MRS and clinical criteria in blind service users and their controls (n=112)

Agreement	Expected agreement	Kappa	Standard error	Z	p-value
60.7%	44.6%	0.290	0.07	4.02	<0.001

ASD: Autism Spectrum Disorder; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale

7.3.7. Prevalence of autistic traits in blind service users and their controls using the Leicestershire Learning Disability Register (LLDR)

The prevalence of autistic traits (defined in the previous chapter) in the blind subgroup is shown in Table 7.22. Following a similar pattern to that found in the analysis of the stage 1 data, autistic traits were generally more commonly observed among sighted service users. The presence of stereotypies (p=0.03), deficits in empathy (p=0.02) and poor quality of social interaction (p=0.02) were all significantly more prevalent among sighted individuals. In both blind and sighted service users, poor use of speech was the most commonly reported

autistic trait, followed by deficits in empathy and presence of stereotypies. Among autistic blind and sighted service users, the presence of stereotypies (p=0.03), deficits in empathy (p=0.002), poor use of speech (p=0.02) and poor quality of social interaction (p=0.001) were all significantly more prevalent among autistic sighted individuals (Table 7.23).

Table 7.22: Comparison of each autistic trait in blind service users and their controls by presence of traits on the LLDR (n=125)

Autistic traits	Blind (n=59)	Sighted (n=66)	Pearson Chi^2	Chi^2 p-value
Stereotypies	28 (47.5%)	44 (66.7%)	4.71	0.03
Deficits in empathy	35 (59.3%)	52 (78.8%)	5.58	0.02
Elaborate routines	25 (42.4%)	28 (42.4%)	0.00	1.00
Poor use of speech	42 (71.2%)	53 (80.3%)	1.42	0.23
Poor quality of social interaction	17 (28.8%)	33 (50.0%)	5.83	0.02

LLDR: Leicestershire Learning Disability Register

Table 7.23: Comparison of each autistic trait in autistic* blind service users and autistic* controls by presence of traits on the LLDR (n=81)

Autistic traits	Autistic Blind (n=45)	Autistic sighted (n=36)	Pearson Chi^2	Chi^2 p-value
Stereotypies	23 (51.1%)	27 (75.0%)	4.71	0.03
Deficits in empathy	28 (62.2%)	33 (91.7%)	9.33	0.002
Elaborate routines	17 (37.8%)	19 (52.8%)	1.83	0.18
Poor use of speech	34 (75.6%)	34 (94.4%)	5.30	0.02
Poor quality of social interaction	15 (33.3%)	25 (69.4%)	10.43	0.001

LLDR: Leicestershire Learning Disability Register

*Defined using the PPD-MRS

7.3.8. Relationship between ASD and congenital blindness (Research Question 8)

As before, conditional logistic regression was carried out, but the data had to be restricted to a smaller sample from that which was originally randomised, due to attrition, and did not allow adjustment for other potential confounders (Table 7.24). Thus, for the subsequent analyses, logistic regression only was carried out to study the relationship between ASD (outcome based on PDD-MRS) and each potential confounding variable (visual impairment, degree of ID, gender and epilepsy) either individually (using single-variable logistic regression models) or in combination (using multi-variable logistic regression models).

Table 7.24: Conditional logistic regression showing the relationship between ASD (outcome as measured by PDD-MRS) and blindness

Variable	OR	SE	95% CI	p-value
Blindness	4.29	2.220	1.55 – 11.85	0.005

ASD: Autism Spectrum Disorder; CI: confidence interval; OR: odds ratio; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale; SE: standard error

Table 7.25 shows the findings from the single-variable models. The odds of ASD were around 3 times higher among service users with congenital blindness compared with their sighted counterparts and this association was independent and highly significant (p=0.008).

Although being male and having a diagnosis of epilepsy increased the risk of ASD, these associations were not statistically significant. Both groups had been matched during randomisation on their degree of ID and gender; therefore, there was no statistically significant association detected between having ASD and degree of ID or gender.

Similarly, although people with epilepsy had higher odds of having a co-morbid ASD, this was not statistically significant after adjusting for other confounders.

Table 7.26 shows the findings from the multi-variable models.

Table 7.25: Single-variable logistic regression showing the crude relationship between ASD (outcome as measured by PDD-MRS), blindness, gender, degree of ID and epilepsy (n=127)

Variable	OR	SE	95% CI	p-value
Blindness	2.82	1.11	1.31 – 6.09	0.008
Gender (Male)	1.44	0.55	0.67 – 3.06	0.35
Age (years)	1.00	0.01	0.97 – 1.02	0.82
Degree of ID:				
Mild	0.38	0.29	0.09 – 1.69	0.21
Moderate	0.19	0.17	0.03 – 1.13	0.07
Severe	0.54	0.22	0.24 – 1.22	0.14
Profound	1.00	-	(reference)	-
Epilepsy	1.38	0.51	0.66 – 2.86	0.39

ASD: Autism Spectrum Disorder; CI: confidence interval; ID: intellectual disability; OR: odds ratio; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale; SE: standard error

Table 7.26: Multi-variable logistic regression showing the relationship between ASD (outcome as measured by PDD-MRS), blindness, gender, degree of ID and epilepsy (n=127)

Variable	OR	SE	95% CI	p-value
Blindness	3.03	1.27	1.34 – 6.89	0.008
Gender (Male)	1.41	0.59	0.62 – 3.19	0.41
Age (years)	0.99	0.02	0.96 – 1.02	0.61
Degree of ID:				
Mild	0.31	0.24	0.06 – 1.47	0.14
Moderate	0.14	0.14	0.02 – 1.00	0.05
Severe	0.52	0.22	0.22 – 1.21	0.13
Profound	1.00	-	(reference)	-
Epilepsy	0.78	0.34	0.34 – 1.83	0.57

ASD: Autism Spectrum Disorder; CI: confidence interval; ID: intellectual disability; OR: odds ratio; PDD-MRS: Pervasive Developmental Disorder in Mental Retardation Scale; SE: standard error

7.4. Carers' views

Although this project did not aim to explore carers' views qualitatively, the carers did give their views on how the services could be improved for those with ID, sensory impairment and ASD. When probed directly what else could have helped to improve the individual's quality of life in the community, the carers thought the following would be important:

- Access to sensory integration assessment and therapies through Occupational therapists (OT) or sensory rooms (Snoezelen).
- More level of one-to-one support and provision of support at home.
- An allocated social worker to help with day care, social/leisure opportunities and structured activities in the community or on site.
- Voluntary jobs and supported employment for those with mild to moderate ID.
- Living in a residential home with smaller groups of service users where the age mix and the level of skills and abilities of the service users were matched.
- More input from the multidisciplinary team e.g. speech and language therapists for communication strategies, nursing team for support for challenging behaviour and occupational therapists for environmental adaptation and skills assessment.
- More training & support from charity organisations on sensory impairment (e.g. RNIB, RNID).
- Dignified accommodation which could ensure privacy and enough space e.g. spacious residential homes with a low number of service users and a high staff to service users ratio, self-contained supported living flats/accommodation with dedicated one-to-one key workers who provide consistency and familiarity through the development of a trusting and friendly relationship.
- Emergency access to health care professionals including a single point of contact during out-of-hour periods to reduce the burden of care.
- Access to assistive technologies e.g. epilepsy bed sensor

- Regular review by adult ID psychiatrists for those presenting with challenging behaviour.

Families were also concerned about transition of children with sensory impairment into adulthood which can be extremely anxiety provoking for services users and their families. Care should be therefore taken to make this as smooth as possible through a coordinated approach among health care, education and social care organisations.

The charity organisation, Sense, has undertaken a project on the transition of those with multisensory impairment (including those with ID) which has resulted in a very comprehensive web-based package for families, young people and practitioners to help and guide them through the processes of transition. The project by Sense is called 'Getting a Result: The transition into adulthood' (http://www.sense.org.uk/content/getting-result-support-package).

A similar scoping study has been undertaken by Judy Bell in 2014 at the request of SeeAbility (https://www.seeability.org/) to identify and fill the gaps in transition of young people with visual impairment and complex needs into adulthood. At the time of writing this thesis the results had not yet been published (email correspondence with Judy Bell on 09.03.2015).

Similar views by carers have been recorded during another study at the Leicestershire adult ID service by Barrett (2014) when studying change of autistic traits in adults with ID over times.

8. DISCUSSION AND CONCLUSIONS

8.1. Summary of the study results, implications for specialist service development and contribution to the wider literature on this topic

The current research project showed a high prevalence of deafness, blindness and deaf-blindness in a cohort of adults with ID.

In stage 1 of the current study which was conducted on 3138 service users, 51 (1.6%) were identified with total deafness and 175 with partial deafness (5.6%). At this stage, the study also identified 114 service users with total blindness (3.6%) and 272 (8.7%) service users with partial blindness. There were 22 service users with deafblindness.

Service users with deafblindness (n=22), borderline IQ and blindness (n=9), borderline IQ and deafness (n=9), unilateral deafness (n=1) and unilateral blindness (n=6) were excluded from the data analyses. This stage of the study therefore included all service users with an acquired or congenital deafness (n=41) or blindness (n=99).

Results of the statistical analyses from stage 1 showed that both visual and hearing impairment (congenital and acquired) were significantly associated with degree of ID and age, but not with gender. People with visual impairment were more likely to have co-morbid epilepsy or Down syndrome. Approximately two-thirds (63.5%) of those with Down syndrome (n=270) wore spectacles. Hearing impairment was also commonly reported in service users with Down syndrome with approximately 1 in 10 people (n=41/425) with Down syndrome using hearing aids.

Furthermore, 17.2% of the service users qualified for a diagnosis of ASD as they had 4 or more autistic traits (threshold for diagnosing ASD based on the number of carer-reported autistic traits available on the LLDR database). However, there was no statistically significant association between ASD and

accompanying visual or hearing impairment at this stage of the study. Autistic traits were associated with a younger age, being male and having a more severe degree of ID. Both ASD and blindness (not deafness) were also associated with challenging behaviour.

Stage 2 of the study, which involved face-to-face interview and direct examination of the service users, made it possible to differentiate those with an acquired sensory impairment from those with congenital deafness or blindness. Therefore, at this stage an association of congenital blindness or deafness with ASD was subsequently explored. The study highlighted high rates of unmet complex needs among service users with congenital sensory impairment as a result of various physical and mental health co-morbidities, including challenging behaviour, mental health problems, epilepsy and ASD.

In contrast to stage 1, stage 2 revealed a statistically significant association between being congenitally blind and having ASD (OR=3.03; p=0.008) independent of other known risk factors. No such association was found between congenital deafness and ASD (OR=0.73; p=0.64). The identification of ASD based on three different assessment methods including (i) an objective assessment tool (PDD-MRS), (ii) clinical assessment and (iii) carers' report of autistic traits (available on the LLDR database) was compared between service users with sensory impairment and their controls. This revealed that the most accurate method for diagnosing ASD was using an objective assessment tool (PDD-MRS) in contrast to clinical assessment which was the least accurate method for identifying ASD in adults with ID and sensory impairment. Moreover, it showed that there was a poor agreement between diagnosing ASD by PDD-MRS and the two other assessment methods, using kappa statistics (kappa<0.4). Those with a diagnosis of ASD, regardless of whether they were in the blind, deaf or control group had a higher rate of presenting with challenging behaviour and being prescribed psychotropic medications.

Overall, the findings of the study contribute substantially to scientific knowledge with regard to understanding the increased prevalence of ASD in those with congenital blindness and ID (Box 8.1 & 8.2). These have significant implications

from a service improvement perspective as the results support the development of more expertise in this area, investment in training, raising awareness and making reasonable adjustments for this group of service users to be able to access generic and specialist services.

Box 8.1: What is already known on this topic?

- Sensory impairments are common in adults with ID and the risk increases with age and severity of ID.
- ASD is also commonly reported in adults with ID especially in those with more severe ID.
- There is a high chance of missing a sensory impairment and ASD in adults with ID if no objective assessment tools are used.
- Autistic traits, autistic-like features and autism have been commonly reported in people with congenital deafness and blindness, but increased rates of ASD in people with deafness or blindness have been mainly attributed to the brain damage.

ASD: Autism Spectrum Disorder; ID: intellectual disability

Box 8.2: How this research contributes to knowledge

- This study, in a cohort of adults with ID (i.e. brain damage), did not find an association between deafness and increased rates of ASD.
- However, for the first time, an independent and statistically significant association was found between blindness and ASD after adjusting for the main confounding variables: severity of ID (brain damage) and gender.
- This study highlights the complex needs of adults with ID, ASD and sensory impairment and advocates more expertise and service development in this area.

ASD: Autism Spectrum Disorder; ID: intellectual disability

8.2. Discussion/interpretation of the findings in the deaf subgroup

In contrast to the body of published literature in deaf children and deaf adults, findings from stage 2 of the study did not reveal an independent association between congenital deafness and ASD in a randomly selected and matched cohort of adults with ID. There might be several explanations as to why an association with ASD was not found in the subgroup with deafness. These are discussed in more detail below.

First, the sample size for stage 2 of this project may not have been large enough (lacking power) to detect statistically significant associations, but even in stage 1 of the project, with a relatively good sample size, no such association was detected.

Secondly, it is possible that there is not an independent association between congenital deafness and autism. Indeed, it has been reported that there is no particular feature of ASD that suggests that it might be associated with deafness (Roper *et al*. 2003) and that deaf children who have a diagnosis of ASD present similarly to those who are hearing and have ASD (Roper *et al*. 2003).

Thirdly, the relationship observed between ASD and deafness, which has been clinically seen and reported in studies of children (Jure *et al*. 1991), maybe being mediated through severity of brain damage, which would mean that the association is no longer seen when severity of brain damage is taken into account. Similarly, some of the literatures describing an association between ASD and deafness have been reported in the context of a genetic syndrome (Kiani *et al*. 2007) but this association is likely to be mediated by the presence of ID (O'Brian, 2006).

A number of previous studies describing the relationship between ASD and deafness have reported different results. For example, the 2009–2010 Annual Survey of Deaf and Hard of Hearing Children and Youth in US reported a comorbidity of ASD with hearing impairment at an approximate rate of 1.9%

(Szymanski *et al.* 2012). Kancherla *et al.* (2013) using data from the population-based Metropolitan Atlanta Developmental Disabilities Surveillance Program, reported a co-existing diagnosis of ASD in approximately 6-7% of 8 years old children with hearing and visual impairment.

Jure and colleagues (1991) studied audiological data of a group of 46 children diagnosed with both ASD and deafness. The authors concluded that while the severity of the ASD was related to the severity of ID, it was unrelated to the degree of the hearing impairment. They found that 21 of the children had delayed diagnosis of ASD for at least 4 years. This may be due to a lack of availability of standardised ASD assessment tools for children with deafness, though some have suggested that ADOS is suitable for this purpose, based on the children's language development and age (Edwards, 2004; Edwards & Crocker, 2008). On the other hand, another study found that children with severe language deficits received an ASD diagnosis on average 1.2 years earlier than other children (Mandell *et al.* 2005).

In 2004, Kielinen and colleagues carried out a population-based survey among 152,732 Finnish children and adolescents aged less than 16 years and found that 187 of them fulfilled DSM-IV criteria for a diagnosis of ASD. They reported that 8.6% of their autistic sample had a hearing impairment and 3.7% of children with ASD had severe accompanying visual impairment (Appendix 1). However, there was also high rate of comorbidity with other genetic syndromes and the association of sensory impairment with ASD may have been due to accompanying ID. In 1999, Rosenhall and colleagues also reported that 3.5% (n=7) of 199 children with ASD had profound hearing impairment.

A fourth explanation as to why a diagnosis of ASD has not been found to be associated with deafness in this study is that the current project was conducted in a specialist ID service (Leicestershire Adult ID service) and the majority of the service users (regardless of whether they were deaf or not) had been registered to receive support (either through social or health care services) because of challenging behaviour which can be a manifestation of ASD. It is reasonable to assume that the majority of the service users, referred to the services for

challenging behaviour, had an underlying ASD whether or not they were deaf or hearing; therefore no significant difference in the rate of ASD could be found between them. Finally, it can be assumed that the symptoms described clinically in deaf people are actually autistic-like features and, therefore, in formal assessments would not qualify for a formal diagnosis of ASD. Leekam *et al* (2011) reviewed literature and reported a repetitive and narrowed repertoire of interest and behaviour in conditions other than ASD, including ID and sensory impairment. Others based on their clinical experience point to the potential for false-positive diagnosis of ASD by professionals unfamiliar with working with deaf children, as deaf children with limited access to language are prone to have impaired communication and might show a strong adherence to routine, preference for predictability and rigidity that could be similar to ASD presentation (Gentili & Holwell, 2011).

Below is a summary of the literature that describes autistic-like symptoms in people with congenital deafness and how deficits in social skills and theory of mind are common in this population, without necessarily suggesting a diagnosis of ASD. These studies have been mainly carried out on children, as there is a dearth of literature on association of ASD and deafness in adults with ID. A literature review on autism and deafness can also be found on the website of the Association of University Centres on Disabilities: http://aucd.org/.

It has been found that congenital deafness causes deficits in theory of mind as a result of a deficit in communication experience, incidental learning and interaction between the deaf child and others; development of language is not only essential for development of communication skills but also for the development of theory of mind (Gould, 1997; Gentili & Holwell, 2011; Sessa & Sutherland, 2013). Development of language is crucial in the development and understanding of feelings and emotions (Lieberman *et al.* 2007; Hauser *et al.* 2006 & 2008; Gentili & Holwell, 2011). Therefore, language deprivation has an adverse effect on developing empathy, attachment and ability to maintain relationships (Dunn, 2004; Hauser *et al.* 2006 & 2008; Gentili & Holwell, 2011). It has been argued that human beings have an internal ability to communicate (either verbally through speech or spatially through pointing, signing and

gesturing), share interest and imitate through the functional role of mirror neurons (Rizzolatti & Craighero, 2004; Tomasello, 2008). There is research on the involvement of these neurons in understanding other people's emotion in people with ASD (Dapretto, 2006; Gentili & Holwell, 2011). The coupling between brains through wireless networks of non-verbal communication has also been described as 'inter-brain', e.g. in contagious yawning or smiling (Tantum, 2012). The Language Acquisition Support System is the process by which parents, carers and others help language and theory of mind to develop during a time when children start learning how to play (Peterson & Siegal, 1995 & 2000; Trevarthen & Aitken, 2001; Scheff, 2006; Tomasello, 2008; Gentili & Holwell, 2011).

Communication attempts such as pointing, gesturing and babies' making noises during the last few months of infancy develop further into words or signs between the first and second years of life (Carpenter *et al*. 1998). Delayed access to sign language in deaf children has negative impacts on language development and this, along with loss of access to incidental learning, are known to affect non-verbal cognitive abilities and social problem solving adversely (Mayberry & Locke, 2003; Edwards, 2004; Edwards & Crocker, 2008). In contrast, in deaf children who are born in deaf families where the parents are signing, or in those who are born to hearing families where parents have learned to sign, theory of mind and visual attention develop normally (Lederberg & Everhart, 1998; Spencer, 2000; Schick *et al*. 2007; Gentili & Holwell, 2011; Sessa & Sutherland, 2013). In children with ASD, it has been found that non-verbal cognitive skills and communication scores at an early age predict subsequent language and theory of mind development (Thurm *et al*. 2007). Children with deafness who are not exposed to a visual form of communication such as British Sign Language have delayed development of theory of mind (Lundy, 2002; Edwards, 2004; Edwards & Crocker, 2008; Gentili & Holwell, 2011; Sessa & Sutherland, 2013). In 1998, Russel and colleagues reported that deaf children's performance on theory of mind tests (aged 4 to 16 years) was more delayed and poorer than the performances of their younger hearing counterparts. This appears to be related to age and also to limited exposure to any method of communication, it being speech or sign language,

and restricted opportunities for learning about the mental state of others (Peterson & Siegal, 2000; Peterson, 2004), as a result of deaf children's parents' inability to communicate with them through sign language (Moeller & Schick, 2006). Not having any exposure to speech or sign language early in life results in a lack of incidental learning which might explain why ASD has been reported higher in congenitally deaf children (Jure *et al*. 1991; Newschaffer *et al*. 2007; Gentili & Holwell, 2011; Sessa & Sutherland, 2013).

In brief, children with congenital deafness may present with symptoms very similar to ASD e.g. a preference for predictability, structured routines, rigidity and inflexibility that can be mistaken with ASD (Roper *et al*. 2003; Edwards, 2004; Edwards & Crocker, 2008; Gentili & Holwell, 2011). Presence of other co-morbid conditions, such as brain damage and ID, also affect this equation; therefore, great care should be taken to avoid misdiagnosing either condition. In congenital deafness, the main psychopathology however is insufficient access to language exposure, while in classic ASD the main psychopathology seems to be genetically/neurologically determined (Baron-Cohen *et al*. 1993; Schick *et al*. 2007; Sessa & Sutherland, 2013).

Health care professionals should thus explore in detail signs and symptoms of ASD in people with congenital deafness, such as absence of a communicative intent, deficit in imagination, poor eye contact and facial expressions, qualitative impairment in language and mutual social interaction, presence of challenging behaviour (Hindley & Kitson, 2000; Austen & Jeffery, 2007; Collins & Carney, 2007; Gentili & Holwell, 2011), obsessively pursued interests and rigid adherence to routines at the expense of other activities and social interactions (Edwards, 2004; Edwards & Crocker, 2008; Szymanski & Brice, 2008). It is therefore paramount for those who embark on assessing ASD in congenitally deaf people to have a specialist knowledge of the normal development, communication skills, sign language and behavioural characteristics of this population so that in clinical practice misdiagnosis and inappropriate management strategies are avoided (Edwards, 2004; Edwards & Crocker, 2008; Gentili & Holwell, 2011; Sessa & Sutherland, 2013).

8.3. Discussion/interpretation of the findings in the blind subgroup

In the current study, information on the aetiology of ID in a considerable number of the patients in the blind subgroup was lacking in spite of having severe to profound ID. Possible reasons for this are: (i) unavailability of advanced genetic testing for older service users during their childhoods; (ii) a therapeutic nihilism on part of the health professionals, who might have felt that no more could be done, even if a cause was identified; (iii) service users moving from one geographical location to another resulting in a loss of case file information; (iv) priority being given to complex clinical presentations, such as epilepsy and challenging behaviour, rather than questioning the underlying condition; (v) service users' non-compliance and challenges of applying legal frameworks in non-consenting service users for investigating ID aetiology; (vi) reluctance from parents and families to pursue investigations to unravel the aetiology of the ID because of feelings of guilt, stigma, and also of the effect this might have on other family members, such as their children and grandchildren (e.g. mental health problems and suicide).

It is important to discuss the possibility of a genetic syndrome with the families as it not only sheds light on the aetiology but also provides information about the co-morbid conditions, that might present as the service user becomes older (e.g. Alzheimer's dementia in people with Down syndrome), and the prognosis. This knowledge will also help to plan and be prepared for the future and to quantify the risk to other family members through appropriate genetic counselling (de Villiers & Porteous, 2012). It might also help to justify the level of support required to look after service users' needs in the community when completing a decision support tool for continuing health care needs assessment applications; these help to secure funding from local commissioning panels for day services, respite/short break facilities, environmental adaptation, direct payment, and assistive technologies (such as epilepsy bed sensors or adjusted alarms).

Current study also showed that clinical assessment is the least accurate method in diagnosing ASD in people with ID and sensory impairment.

There may be a number of reasons for the low yield of identifying ASD clinically:

- Priority given to medical and psychiatric co-morbidities, such as challenging behaviour, mental ill-health and epilepsy.
- Diagnostic overshadowing, whereby symptoms of ASD were attributed to ID, institutionalisation, sensory impairment or the genetic syndrome such as Down syndrome.
- An inability to confirm presence of ASD due to lack of developmental information from a first degree relative.
- Lack of referral to relevant diagnostic services as the symptoms were manageable and did not require specialist input (for those who were not open to the specialist adult ID service).

In contrast to the general population, the prevalence of blindness and ASD is several times higher in the population of adults with ID. There, one would expect a higher prevalence of co-morbidity of these two conditions in this population. However, the findings from this aspect of the project showed that, not only was the co-morbidity high, blindness was an independent risk factor for the development of ASD i.e. the current study showed a statistically significant association between congenital blindness and ASD, independent of degree of ID and gender, an association which has been debated in the literature. The study finding is thus in line with those literature which are supporting such notion i.e. that a congenital blindness is an independent risk factor for the development of ASD. The association between blindness and ASD was not observed when using carer-reported traits as a proxy measure for ASD in stage 1 of the study, which suggests that observer's measures may not be that reliable in this population. The findings might also suggest that autistic traits present differently in people with ID who are blind, or that carers attribute the traits to the person's blindness, rather than to any other cause. The other explanation is that the studied population in stage 2 purely consisted of those with congenital blindness in contrast to stage 1 where there was a mixture of service users with congenital and acquired blindness (the latter is not known to be associated with ASD, hence masking any association that might be really present between a congenital blindness and ASD). In addition, in stage 2 of the study, assessment was completed by a trained professional with expertise in

diagnosing ASD using an objective assessment tool while in stage 1 a diagnosis of ASD was based on the carers' report of autistic traits.

Below a summary of literature, albeit with focus on congenitally blind children (as there is a general lack of evidence on this topic in adults with ID and blindness), arguing for and against the above association, is presented. For more detailed information on association of ASD and visual impairment, please look at Pring (2005) and also the following websites by Bell, Boyce & Hammond, Gense & Gense, Ingsholt and Pawletko & Rocissano respectively. Periodically, Ian Bell also issues a Newsletter concerning visual impairment and autism:

http://ianpbell.com/visual-impairment-autism/.
http://www.ssc.education.ed.ac.uk/resources/vi&multi/boyce.html
http://www.focusfamilies.org/focus/docs/blindnessandautism.pdf
http://icevi.org/publications/ICEVI-WC2002/papers/07-topic/07-ingsholt2.htm
http://www.tsbvi.edu/autism-in-the-visually-impaired-child

Previous literature in this area reveals that there is an interesting overlap between the developmental trajectories of blind children and sighted autistic children. For example, unusual facial expressions, deficits in eye contact, and poor skills in turn taking during conversation have been reported in people who have been born congenitally blind (Preisler, 1991; Mills, 1993; Perez-Pereira & Conti-Ramsden, 1999; Pring 2005). Other examples of autistic-like features seen in congenitally blind children during their development are limitation and deficit in spontaneous communication, symbolic and imaginative play, attempt at exploring the surrounding environment and social curiosity, a preference for being aloof and presence of repetitive speech and behaviours (Gense & Gense, 2002; Perez-Pereira & Conti-Ramsden, 2005; Pring, 2005).

Although previous research has shown an association between stereotypical behaviours and degree of ID and communication impairment (Bhaumik *et al.* 1997 & 2010), some of these behaviours are particularly common in blind children who do not have ID or obvious deficits in their language (Bak, 1999).

These behaviours tend to reduce as the child grows up (Troster *et al*. 1991; McHugh & Pyfer, 1999; McHugh & Lieberman, 2003; Perez-Pereira & Conti-Ramsden, 2005). Stereotypical repetitive behaviours have also been reported in people with severe ID in isolation, and without accompanying blindness or ASD (Bhaumik *et al.* 1997 & 2010; Frith, 2003), and there are strategies developed to reduce the impact of these on the children's lives (Estevis & Koenig, 1994).

In the majority of cases, however, blind children overcome these difficulties over time, albeit with delay, by compensating blindness with other senses such as hearing, touch, taste and smell (Perez-Pereira & Conti-Ramsden, 2005). However, in some children with blindness there are significant degree of qualitative impairments in social and language skills that might qualify for a diagnosis of ASD e.g. lack of empathy, not initiating conversation at all, continued use of echolalia or pronominal reversal even when they get older, and aloofness (Loftin, 1999; Gense & Gense, 2002; Pring, 2005).

For several decades there has been a focus on this issue when discussing the developmental path of children with congenital blindness (Fraiberg, 1977; Rogers & Newhart-Larson, 1989; Brambring and Troster, 1992; Gense & Gense, 1994 & 2005; Kekelis & Sacks, 1992; Brown *et al*. 1997; Recchia, 1997; D'Allura, 2002; Frith, 2003; Hobson & Bishop, 2003; Loots *et al*. 2003; Volkmar *et al*. 2005). For example, Hobson *et al.* (1999) found substantial overlaps but also subtle differences between the presentation of autistic blind children and that of sighted children with autism. Hobson *et al.* (1999) noted that in comparison to their sighted autistic counterpart, blind children with ASD presented differently and that they did not have many affect abnormalities. They questioned whether this could be a distinctive form of ASD, different from that described by Kanner in 1943.

Although some researchers (for example please look at Hobson & Bishop, 2003 and Jure *et al.* 2015) argue that a congenital lack of visual experience early on in life causes an autistic-like presentation that is qualitatively different from the neurologically determined ASD (or Kanner's autism) seen in sighted children, it is quite challenging to ascertain with certainty whether these features in blind

children are due to classical autism or are the result of another developmental trajectory (e.g. in the background of a pre-lingual visuo-social deprivation). One of the main reasons behind this controversy is the fact that any research on congenitally blind children is extremely difficult to carry out (Hobson *et al*. 1997). For example, Minter and colleagues (1998) conducted a study of theory of mind in blind children, and reported how challenging it had been to communicate with children while conducting the experiment to conclude if they had deficits in theory of mind.

But why do children with congenital blindness develop symptoms similar to ASD? Why do the symptoms they present with overlap with those of sighted autistic children (Andrews & Wyver, 2005; Pawletko & Rocissano, 2000; Pring, 2005)? To answer these questions, one needs to put oneself in the position of a child who has been borne congenitally blind, as he or she, in contrast to a sighted person, has not developed an internal picture of the world and everything that exists within it (Boyce & Hammond, 1996). The developmental trajectories of congenitally blind children are discussed in more detail below:

8.3.1. Developmental trajectories in children with congenital blindness

Congenitally blind children, very similar to those who are born deaf, miss out on incidental learning. Blind children have basic difficulties in knowing about the existence, nature and permanence of objects and their relationships (Boyce & Hammond, 1996). They cannot see others, therefore they face a challenge to internalise what is happening around them (e.g. social norms, empathy, facial expression, body language, expression of feeling), imitate others, understand the actions of others and their roles, show interest in any objects, what is expected of them or request to hold or to reach out for objects (Millar, 1983; Kekelis & Sacks, 1992; Warren, 1994; Boyce & Hammond, 1996; Perez-Pereira & Conti-Ramsden, 1999 & 2005; Pring, 2005; Lechelt & Hall, 2014).

Blind children are usually delayed in perceiving the holistic picture of the environment around them as they are limited to their hearing, touch, smell and taste to experience things from a close distance, whereas sighted children experience these things quickly from a distance without any need to learn. Blind

children, therefore, need to learn sequentially, slowly and step-by-step (Bishop, 1991; Boyce & Hammond, 1996; Sugden *et al*. 2012).

Blind children cannot see the carer's face or body and, therefore, can miss out on social cues that are seen in non-verbal communication such as a facial expression or body language (Pring, 2005; Botting, 2007; Akers, 2011). Lack of these skills can result in a feeling of isolation which, in turn, can cause emotional difficulties, mental health problems and challenging behaviour owing to a deficit in understanding the communication intent of other people and what is happening around them (Freeman *et al.* 1989; Buuljents *et al*., 2002; Hoff, 2005; Celeste, 2007; Roe, 2008; Akers, 2011).

8.3.2. Language development in congenitally blind children

It was initially thought that children with congenital blindness (without ID) were not at a disadvantage for language development (Paul, 2007; Akers, 2011) and that, ultimately, they would learn to speak, read and write (e.g. through Braille). However, more detailed studies revealed that the presence of a severe sensory deprivation early on in life could affect language acquisition significantly (Warren, 1994). For example, the limited range of experiences and lack of visual stimulation can first delay language acquisition and then psychosocial development (Tadic *et al*. 2010; Akers, 2011). Despite the above, blind children do have a good capacity to develop efficient verbal/language skills, unless they have another co-morbidity that can affect their language development further e.g. ID, ASD or deafness (McConachie & Moore, 1994; Perez-Pereira & Conti-Ramsden, 2005).

Studies of speech development in blind children and autistic sighted children have shown that both groups use highly imitative, repetitive and stereotypical (modelled) speech, known as echolalia (Frith, 1989; Mills, 1993; Webster & Roe, 1998; Perez-Pereira & Conti-Ramsden, 1999 & 2005; Tager-Flusberg, 1999; Ingsholt, 2002). However, in blind children without accompanying disabilities e.g. ID, this modelled speech seems to be more meaningful and positively related to language development in serving a communication purpose, e.g. as a way of initiating contact and making sense of the

environment around them (Perez-Periera & Conti-Ramsden 1999 & 2005; Kehoe, 2012).

Blind children tend not to refer to actions of others and their speech might appear self-centred because they are unable to observe and comment on other people's behaviour, body language and speech. They therefore have to use their own statements several times which might be labelled as repetitive and egocentric (Andersen *et al*. 1984 & 1993; Landau & Gleitman, 1985; Perez-Pereira & Castro, 1992; Perez-Pereira & Conti-Ramsden, 2005). Other examples of difficulties in speech development in blind children are problems in learning to use phonemes, which need sight to be learnt to utter e.g. "b", "m" and "f" (Mills, 1983; Akers, 2011).

Understanding words that convey a temporal, relative, size or geographical meaning, such as "here" and "there", ''now'' or ''later'', ''this and that'' and "big" and "small"' are quite difficult for a blind child. As a result, blind children might appear superficially fluent but yet still find it very hard to understand the total communication intent of others (Fraiberg, 1977; Boyce & Hammond, 1996; Gense & Gense, 2002; Frith, 2003; Akers, 2011).

Grammatically, there is also a delay in development of using "I" or "you" as a pronoun and auxiliary verbs such as "can" and "do", using gestures and deficits in the ability to point for requesting things (Fraiberg & Adelson, 1973; Landau & Gleitman, 1985; Andersen *et al*. 1984 & 1993; Iverson & Goldin-Meadow, 1997; Perez-Pereira & Conti-Ramsden, 1999 & 2005; Iverson *et al*. 2000; Akers, 2011).

Tadic and colleagues (2010) found that blind children had superior language skills but their skills in using language in social situations and mutual conversation (pragmatic language) was weak, i.e. appropriate use of language in a social and functional context such as turn taking, initiating or finishing a conversation, moving from one topic to another, incorporating social events happening around them in the conversation, and understanding other people's point of view.

A similar presentation is seen in sighted children with ASD who also have difficulties with pragmatic language (Bishop, 2000; Adams *et al*. 2002; James & Stojanovik, 2006; Botting, 2007). It has been shown that a pragmatic language difficulty can result in a deficit in socio-emotional development such as low self-esteem, lack of self-confidence, and an inability to develop friendships or a sense of identity (Huitt, 2008; Akers, 2011). Conversely, it has been found that there is a positive association between language development and abstract thinking/theory of mind development in children with visual impairment (Bigelow, 1990). It is therefore obvious to see how in congenitally blind children with additional co-morbidities such as ID, the challenges of surmounting the barriers become so complex that necessitates involvement of a speech and language therapist early on for further support (Goldbart & Caton, 2010; Akers, 2011).

8.3.3. Aetiology of ASD and deficits in theory of mind in congenitally blind children

Initially, researchers believed that a high rate of ASD in congenitally blind children was mediated through brain damage/ID (Keeler, 1958; Chess, 1971 & 1977; Rogers & Newhart-Larson, 1989; Ek *et al.* 1998; Ek, 2010). However, there have been other studies showing that a congenital blindness could also contribute to deficits in theory of mind and autistic symptomatology (please see below for further details). Deficit or lack of theory of mind has been explored in ASD through important research in the past few decades (Baron-Cohen *et al.* 1985). Whether or not a pre-lingual blindness can cause deficit in theory of mind or ASD independently, i.e. without a co-morbid central nervous system abnormality such as ID, has been a matter of debate (Perez-Pereira & Conti-Ramsden, 2005). This issue is further discussed below based on the outcome of different studies:

In 2014, Begeer *et al.* showed that performances of children with congenital ocular-plus blindness, i.e. those who had accompanying damage to their central nervous system (n=22), were delayed compared with children with congenital ocular blindness, i.e. those without an accompanying neurological deficit (n=9) and sighted children (n =103) on theory of mind tasks.

Mukaddes and colleagues (2007) also assessed the prevalence and associated risk factors of ASD in 257 visually impaired children and adolescents (age range: 7– 18 years) using a three-stage process. They used the Autism Behavior Checklist first and then directly observed the subjects in different settings. In the last stage, a final diagnosis of ASD (n=30), based on the 4th Diagnostic Statistical Manual of Mental Disorders (DSM-IV) criteria, was made after the carers' interview and clinical observation. The study showed that subjects with blindness and ASD had a greater chance of having neurological impairment and more severe visual impairment than the subjects with blindness only.

Previous research (Cass *et al*. 1994; Cass, 1996; Dale & Sonksen, 2002; Sonksen & Dale, 2002; Pring, 2005) revealed that for the study of psychological development in children with visual impairment, blindness could be categorised into 3 main groups, based on the site of the pathology: (i) those with accompanying neurological/central nervous system problems (e.g. congenital rubella syndrome); (ii) those who had impairment of the neurological components of the visual system (e.g. pathology in the optic nerve); and (iii) those who only had impairment of the peripheral visual system (e.g. problems with the cornea or lens). Research showed that only the third group had normal development during childhood, suggesting that development was impaired only in those who had blindness and neurological impairment.

Similarly, in 1998 Ek and colleagues reported that blind children with retinopathy of prematurity and brain damage were significantly more likely to have ASD than those with retinopathy alone (i.e. without any brain damage). They concluded that a diagnosis of ASD in the background of blindness was mediated by the brain damage. Similar findings have been reported by Bahar and colleagues (2003) who found that those blind children who had accompanying neurological damage (optic nerve hypoplasia or septo-optic nerve dysplasia) had abnormal developmental milestones in comparison to blind children without any neurological damage. ASD and social communication disorders have been further studied in this group of blind children with septo-optic nerve dysplasia (Parr *et al.* 2010). For more information on these studies

please refer to Appendix 1. On the other hand, some researchers argued that a pure blindness (i.e. without accompanying brain damage or ID) could be an independent risk factor for ASD. For example, in 1977, Fraiberg was one of the first researchers who suggested that some of the autistic symptoms seen in congenitally blind service users were as a result of gross impoverishment in the development of sensorimotor skills. Those in favour of this hypothesis argue that blind children's inability to see world and other people's interaction, faces and body language create challenges during their development which make their social and communication skills appear similar to those sighted children with ASD. Also, if not engaged in structured activities by parents/carers/teachers, children with blindness would isolate themselves and engage in stereotypical/repetitive behaviours which are commonly seen in sighted autistic children (Boyce & Hammond, 1996; Pring, 2005; Dale & Salt, 2008; Akers, 2011).

Hobson and Bishop (2003) & Hobson (2002 & 2005) explain these in more detail: it has been observed that if play materials are handed over to children who are blind, the game conducted by them appears to be less creative than one would expect from a sighted child. According to Hobson and Bishop (2003) & Hobson (2002 & 2005) there are several explanations as why this might be the case: one of the reasons is an inability to see the physical characteristics of the toys. The other reason is an inability to use toys symbolically in relation to other items or people in the environment because they do not have a mental picture of these. Blind children also struggle to have mutual conversations and reciprocal contact or play because of their lack of sight and therefore an inability to see and read non-verbal cues. They are unaware of events, items and other peoples' attitudes towards each other and themselves and how things are being experienced by others because it is impossible for them to see other people. This makes it very challenging for them during their development to learn how to relate to others and understand how other people's point of view could be different from their own. These all can result in deficits in social and communication skills which are similar to that seen in sighted autistic children (Hobson, 2002; Hobson & Bishop, 2003; Hobson, 2005).

Therefore, from studying congenital blindness, it might be possible to see heterogeneity in the aetiology of ASD (Hobson & Bishop, 2003; Pring, 2005). One possibility is that the cause of ASD is not in the child alone (not neurologically or inherently determined type of ASD i.e. Kanner autism), but rather in deficits of the system that Hobson and Bishop (2003) & Hobson (2002 & 2005) called "child-in-relation-to-others". Hobson (2002 & 2005) and Jure *et al.* (2015), therefore, argue that there may be several routes towards the pathogenesis of ASD, especially in children with congenital blindness and that an early and intensive intervention might, at least in some of the cases, improve the situation. Hobson draws parallels with Rutter and colleagues' study (1999) where Romanian orphans who suffered severe social deprivation presented with "quasi-autistic" symptoms. He explains that the disruption in the system of "child-in-relation-to-others" might occur due to a variety of reasons, either in isolation or together, e.g. a neurological abnormality as one can see in classical ASD, a severe psychosocial deprivation (e.g. in Romanian orphans) or a congenital blindness, all adversely affecting the child-in-relation-to-others system. This theory is clearly very different from the traditional top-down theory of ASD pathogenesis which is neurobiological and polygenic, as it searches at least for part of the problem beyond the genes and central nervous system deficits, i.e. deficits in the environment or sensory organs (a bottom-up theory of autism) which Hobson calls "the theory of inter-subjectivity", i.e. the difficulty in interaction between the affected individual and others as a core deficit in ASD in people with congenital blindness (Hobson, 1984; Hobson *et al*. 1997; Hobson, 2002 & 2005).

8.3 Limitations of the research project

The research carried out as part of this thesis is on a study population that is not representative of people with mild to moderate ID. The majority of service users in this project had severe to profound ID. In addition, the study population included only those adults who were registered on the LLDR and, therefore, they are not representative of most of people with mild to moderate ID in the community who are not yet diagnosed and those who are not in receipt of services.

Other ethnic groups, other than white people, were also underrepresented in the study population. This might be due to language and cultural barriers to accessing services or to the fact that many of these service users receive their care from their families and informal carers and are less likely to access specialist services.

The sample size in the deaf group was small owing to attrition (non-consenting, death and moving out of county) and also because many service users had partial or acquired deafness or deafness in conjunction with blindness and, therefore, could not be included in the deaf subgroup. For more information on the characteristic of the deaf-blind population and those who were excluded from the study, please refer to the Figures 7.1 & 7.2 and Appendix 4.

Although matching service users in the control groups with their sensory impairment peers at the designing stage of the project was intended to increase efficiency by eliminating the confounders (degree of ID and gender) on the association studies (Rose & van der Laan, 2009), this inevitably led to loss of data in a number of service users due to attrition.

Theoretically, although service users with a sensory impairment were matched on degree of ID with their controls, there still remains ‘’within group heterogeneity’’, degree of ID being a dimensional construct rather than a categorical one i.e. the IQ of service users placed in a category could vary between the minimum and the maximum scores (e.g. IQ of 20–35 for all service users within the severe ID category). In real life however, this is the best methodology that could be employed to achieve a near perfect matching.

8.4 Strengths of the research project

The main findings of this thesis (stage 2) are based on the first study of its kind to explore the relationship between sensory impairment and ASD in adults with ID in two randomly matched groups of people with underlying brain damage. In addition, in comparison to other published studies available, the current study compared the sensory impaired service users and their controls matched on

various degrees of ID (as well as gender) rather than just investigating those with IQ below 70.

Although not a population-based study, it could be argued with confidence that this study included a representative population of adults with severe to profound ID who were living in Leicestershire. Furthermore, as well as including service users registered with local ID services, the study also included service users who were not receiving input from these services, which suggests that the results can be generalised to all adults with severe and profound ID who live in the community. Another strength of the study is that the sample population consisted mainly of those with severe and profound ID who rarely receive screening for ASD in daily practice i.e. there are not much literature on the association of ASD and sensory impairment in this population.

The methodology used in this project was robust in that it used: (i) multiple sources to collect information i.e. face to face interview, LLDR database, medical case files or electronic data records of the service users available at the local adult ID service and primary care referral letters or summary clinical sheets, (ii) various objective assessment tools to collect data, both current and chronologically; (iii) randomisation; (iv) adjustment for confounders and matching; and (v) thorough and diverse statistical analyses to unravel any associations observed.

By administering visual and hearing impairment checklists, the study also succeeded in including those with only visual impairment or hearing impairment compared with controls without visual or hearing impairment. Furthermore, service users with unilateral, partial or acquired sensory impairments and those with dual sensory impairment were excluded from the final analysis.

The threshold for identifying ASD also was set high (PDD-MRS score of 10 and above or having 4 or more autistic traits in the LLDR database) to avoid false positive and type I error as autistic traits have been commonly reported in adults with ID. In the final analysis of the data, the project benefited from a triangulation method by which diagnoses of ASD using three different criteria

i.e. (i) carers' report of autistic traits (ii) ICD-10 clinical diagnosis (iii) an objective assessment tool (PDD-MRS) were compared using kappa agreement studies.

Although several people passed away or could not be contacted for a variety of reasons during the course of the study, the power calculation helped to achieve the minimum number of participants through multi-layered recruitment strategies with the help of the multi-disciplinary teams. In addition to answering the main study question, the project also managed to provide answers for other interesting queries with regard to co-morbidities in this particular population.

Finally, as this study was conducted in an adult population, the issue related to changes in autistic traits/a diagnosis of autism over time, which has been reported in children with blindness (Hobson & Lee, 2010), was not of a concern.

8.5 Benefits of diagnosing ASD in service users with sensory impairment

Although some families might feel stigmatised and carers may give up or develop a therapeutic nihilism that nothing more can be done, in most cases a diagnosis of ASD in the background of blindness/deafness will actually do more good than harm. For example, a diagnosis can facilitate access to funding for further support, including involvement from professionals with expertise in both ASD and blindness/deafness. It also raises the awareness to better adjust the environment based on the service user's needs (Brandsborg, 2002). It will provide alternative explanations for the service user's challenging behaviour and consequently reduce the guilt felt by the carer and stop others from blaming the service users (e.g. "he/she knows exactly what she/he is doing")'. Ultimately, when assessing a service user for ASD, their carers/families' views and those of the multidisciplinary teams should be taken into account so that they feel involved in decision making process (Steinberg *et al.* 2000; Fox *et al.* 2010; Salt, 2010).

8.6 Psychosocial, educational and family interventions for children and adults with sensory impairment and ASD

Help and support should be provided for parents and carers, ideally during the child's developmental stages to promote early language acquisition, social skill development and secure attachments (Bondy & Frost, 1994; Wallis *et al*. 2004; Edwards, 2004; Edwards & Crocker, 2008; Akers, 2011; Gentili & Holwell, 2011; Humphries *et al.* 2012; Sessa & Sutherland, 2013).

Early identification of sensory impairment and ASD and early intervention through different mediums such as alternative communication and educational strategies with any form of language, be it basic signing, deaf-blind manuals, Braille or through use of objective references, is the key to success. These are crucial in reducing a feeling of isolation and will eventually result in better communication and development of social skills (Akers, 2011; Gentili & Holwell, 2011; Sessa & Sutherland, 2013). For those who are deaf, this can be achieved through access to schools and colleges that allow them to mix with a more able group of deaf students who can sign fluently and who act as role models (Gentili & Holwell, 2011; Sessa & Sutherland, 2013). The National Deaf Children's Society provides more information on these issues (National Deaf Children's Society, 2012). For blind children, non-visual communications could be promoted to compensate for blindness and help them to develop joint attention, shared interest and social referencing skills (Moore & McConachie, 1994; Cass, 1996; Edlund *et al.* 2002; Loots *et al*. 2003; Clarke, 2010; Akers, 2011). Within schools and colleges, blind students could learn from reverse integration (Jordan, 2005), which means placing them with non-autistic and sighted children in specialist settings to make friends with, so that those with a disability can learn from their able counterparts before integration into mainstream education. It is also crucial to be aware of other co-morbid disabilities such as ID, dual sensory impairment, and epilepsy because they might slow down or prevent the normal development.

Some of the autistic-like features in the context of a congenital blindness, such as echolalia, pronominal reversal, resistance to change, limited interests and

adherence to routines, could improve over time as blind children grow older, provided that they can be offered more exposure and further enjoyable sensory and motor activities (Gense & Gense, 2002; Perez-Pereira & Conti-Ramsden, 2005). It is reported that with staff input, further exposure to various opportunities and appropriate sensory activities, blindism (autistic-like symptoms) could be reduced through therapeutic relationship (Adelson & Fraiberg, 1976; Perez-Pereira & Conti-Ramsden, 2005; Hagood, 2008; Barrett, 2014).

Both service users with deafness and blindness with similarities and differences in their communication and language development regardless of whether they have ASD or not, benefit from similar autism friendly approaches (e.g. the need for the same environment, predictable routines, structured activities and teaching, familiar one-to-one staff support, communication passport, consistency, repetition and enough time to process information) (Preisler, 1995). This will help them to gather the environmental information, step by step and sequentially, and convert them into a holistic manner and offer them an opportunity to learn from experiences (Howley & Preece, 2003; Gibbons, 2005; Macleod & Curtis, 2010; Stevens, 2010).

Multidisciplinary teams' overall knowledge of children's strengths, their communication and interaction skills, the areas that they need improvement and the presence of other physical and psychological co-morbidities (e.g. epilepsy, challenging behaviour) are immensely significant in coming up with various strategies to help them make up for their sensory impairment (O'Hare, 1996; Gense & Gense, 2002; Gibbons, 2005; Macleod & Curtis, 2010)

Although research shows that the mothers of visually impaired children elaborate more and make significantly more references to story's characters' mental states than mothers of sighted children (Tadic *et al*. 2013), these are not enough and support should be offered to them by carers and teachers in other settings.

Management plans for blind children should be individualised, client-centered and based on other communication strategies (e.g. through touch, taste, smell and hearing) to increase their engagement/involvement. They will benefit from meaningful feedbacks, rewards and positive reinforcements at every step during their development and information should be offered to them in short, concrete and simple terms (O'Hare, 1996; Gense & Gense, 2002; Perez-Pereira & Conti-Ramsden, 2005; Gibbons, 2005; Jordan, 2005; Macleod & Curtis, 2010; Akers, 2011). These will help blind children to participate and feel included in group activities, to express feelings, and to understand the world from the other peoples' point of view, which will lead to development of joint attention, shared interest, theory of mind and empathy in the long run (Buuljents *et al*. 2002; Gense & Gense, 2002; Perez-Pereira & Conti-Ramsden, 2005; Gibbons, 2005; Jordan, 2005; Roe, 2008; Akers, 2011).

Some researchers argue that the impact of congenital sensory impairment on socio-affective impairment might be amenable to modification in some people with autistic-like symptoms, if appropriate support is in place early on during their development (Brown *et al.* 1997; Hobson *et al.* 1999). It is therefore important to support parents and their children to develop an effective communication strategy and structured educational programme to prevent development of poor psychosocial skills, unsecure attachment, challenging behaviour and emotional disturbances (Vaccari & Marschark, 1997; Yoshinaga-Itano, 2000; Jamieson, 2004; Akers, 2011; Gentili & Holwell, 2011; Sessa & Sutherland, 2013). Teaching parents to incorporate facial expressions and gestures in their method of communication with their deaf children may help to alleviate some of the sensory/perceptual deficits that are commonly reported in the context of ASD, such as auditory and visual perceptual processing difficulties (Bonvillian *et al.* 1981; Chamberlain & Mayberry, 2000; Gentili & Holwell, 2011; Sessa & Sutherland, 2013).

Improvement in social skills, academic achievement, self-esteem and becoming part of a group at school seem to be related to a better quality of life in society as an adult (Walz & Bleuer, 1992; Akers, 2011). Therefore, by improving the social awareness of blind children, they can be helped to overcome loneliness

which, in the long term, prevents isolation, academic underachievement and mental health problems such as low self-esteem, depression and challenging behaviour (Sacks *et al*. 1992; Hurre & Aro, 1998; Sacks, 2006; Akers, 2011). In an interesting case study, Akers (2011) showed how it is extremely important for children with congenital blindness to develop awareness of social communication through other sensory modalities (such as touch) to help them to improve their relationships. This ultimately results in a better quality of life, development of problem solving/cognitive skills, happiness and healthy attachment, with the support of significant others (Urwin, 1983; Buuljents *et al*. 2002; Hoff, 2005; Roe, 2008; Akers, 2011). Similarly, Celeste (2007) suggests a social skills intervention plan consisting of role play, inclusion and practising to help improve social and communication skills of children with blindness which is crucial to their inclusion at school and for building friendships (MacCuspie, 2006; Wolffe, 2006; Akers, 2011). Improving communication of service users with autism and training their peers will also facilitate healthy social interaction among them (Kamps *et al*. 2002). This is paramount in reducing the challenging behaviour displayed by them (Samways & Bell, 2010; Bell, 2012). Discussing visual impairment and its effects with children who are blind might also help the above strategies through raising awareness. This can be sensitively done by the parents of children with blindness (Harrison & Crow, 1993; Akers, 2011).

In brief, a trans-disciplinary model of collaboration among carers, teachers, families and other specialists seems to be the way forward for the better education of children with congenital sensory impairment (Jordan, 2005). The National Autistic Society's (2003) recommendations for teaching children with ASD uses 5 principles: Structured, Positive, Empathic, Low arousal and Links (acronym of SPELL). This can be adjusted for children with sensory impairment (Gibbons, 2005). New technologies such as various computer software and iPad can all be used effectively in this regard.

Kendall *et al.* (2013) and Pilling *et al.* (2012) have summarised the NICE and SCIE guidance on management of ASD in children and young adults. These also apply to those who have additional disabilities such as sensory impairment or ID, albeit with some modifications.

To provide additional guidance for professionals, RNIB set up the Visual Impairment and Autism Project which ran from September 2008 to March 2011. More information about the Project is available at: http://ianpbell.com/visual-impairment-autism/. In May 2011, RNIB published the Project's resource pack which consisted of a CD-ROM and booklet. The material is now freely available at RNIB website: www.rnib.org.uk/autism. Bell (2013) provides more information about this project and also on communication issues in people with ASD and visual impairment at: http://ianpbell.com/communication-in-vi-children/.

More information on education of sensory impaired children and young people can be found on the website of the Scottish Sensory Centre: http://www.ssc.education.ed.ac.uk/library/publications/retrospective.pdf and the Texas school for the blind and visually impaired: http://www.tsbvi.edu/.

8.7 Service provision

Delayed diagnosis of medical or psychiatric conditions (e.g. ASD, sensory impairment) is part of the bigger picture of health inequalities that people with ID face. There are concerns that it is difficult for people with sensory impairment to access services such that many may be either slipping through the net or lost to follow up, which might lead to their coming to the attention of services in crisis (Beresford *et al*. 2008; Wright, 2011; Sessa & Sutherland, 2013). For deaf people (both children and adults) in the UK, several accessible specialist mental health services have been developed which employ staff who have expertise in both sign language and assessment and management of mental health problems. These services seem to be cost-effective, as they aim to reduce the cost of untreated or misdiagnosed ASD, challenging behaviour and other mental health problems which can potentially present in crisis in future (Sessa & Sutherland, 2013). Although for blind people there is no need to develop specialist mental health services, these service users will still benefit from input from staff who have expertise on assessment and management of ASD and mental health problems in blind individuals.

Nationally there are not many specialist care homes that have expertise in dealing with people with ID who also have ASD and profound sensory impairment. Each person will, therefore, require the development of a suitable care package tailored to their individual needs for an appropriate placement. It is important that staff receive training in how to communicate with people with sensory impairment when placed in such homes. The most suitable type of placement however seems to be the one in which staff have expertise in dealing with sensory impairment so that additional training can be put in place to address the complex needs of those with ASD through familiarity, consistency, adjusted communication strategies, environmental adaptation and structured routines (Jure *et al.* 1991; Gibbons, 2005; Jordan, 2005; Pring, 2005). In the long term, appropriate policies and procedures consistent with '*Valuing People' Now* (Department of Health, 2009) should facilitate access to generic health services for people with ID and sensory impairment (Alborz *et al.* 2005), through the mediums of annual health checks, health action plans, communication passports, reasonable adjustments and health facilitation.

In addition, the 2005 Mental Capacity Act provides a legal framework to enable the clinicians to treat those vulnerable service users who lack capacity to consent to appropriate health interventions in hospital settings. The issue of prejudice and institutional discrimination towards people with ID must also be actively recognised and addressed through appropriate legislation and training to change negative attitudes that unfortunately still continue to affect the lives of people with ID. Similarly, developing local and national databases, involving service users and their carers in service planning, and mandatory incorporation of ID teaching in undergraduate and postgraduate clinical training curricula can equip future healthcare professionals with the skill sets to address the complex needs of people with ID, ASD and sensory impairment in various settings. Specialist services for people with ID and sensory impairment should not be limited to the medical or health teams as local social services can provide specialist care managers, social workers, rehabilitation workers, technical officers, mobility officers, support workers, interpreters, guide communicators and communication support workers for deaf and blind people to help improve their quality of life (Butler, 2004). This can help with social exclusion,

unemployment or underemployment which affects a large proportion of adults with mild ID, sensory impairment and ASD (Barnard *et al*. 2001).

Butler (2004) has highlighted some of the other legislations, guidance and standards, which could be referred to in order to eliminate discrimination, reduce social exclusion and make services and employment more accessible when securing/developing services locally for people with sensory impairment:

- National Assistance Act;
- Chronically Sick and Disabled Persons Act;
- National Health Service and Community Care Act;
- Equality Act (previously Disability Discrimination Act);
- Human Rights Act;
- Copyright (Visually Impaired Persons) Act;
- National Service Framework;
- National Minimum Standards;
- Section 7 Guidance;
- RNIB accreditation;
- Best Practice Standards.

8.8 Conclusions and recommendations

The findings of the current study are in support of the argument in favour of an independent association between congenital blindness, but not deafness, and ASD in a cohort of adults with ID. Understanding the pathogensis of ASD in congenitally blind children would therefore be invaluable in understanding the pathogenesis of childhood autism (Hobson, 2005; Tager-Flusberg, 2005; Hobson & Lee, 2010; Jure *et al.* 2015). A distinction of a service user being deaf/blind/deaf-blind and autistic or just deaf/blind/deaf-blind with autistic-like symptoms appears however to be of academic and theoretical interest only, as there is a need to focus on practical strategies to help and support them to fulfil their potentials to relate positively with others and the surrounding environment (Carvill & Mraston, 2002; Pring, 2005).

In practice what clinically is important is that ASD and sensory impairments are common conditions in people with ID and, therefore, if missed, service users suffer in terms of their quality of life and the services to which they are entitled.

The likelihood of missing ASD and sensory impairment in people with ID rises if standardised assessment tools are not used and if there is no input from the skilled multidisciplinary team professionals. There are no standardised assessment tools for diagnosing ASD specifically in blind or deaf service users and the presence of deafness/blindness usually delays the diagnosis of ASD (Jure *et al.* 1991; Edwards, 2004; Edwards & Crocker, 2008; Pring, 2005; Absoud *et al.* 2011). Assessment should therefore be multidisciplinary and multiagency (e.g. involving doctors, carers, psychologists, teachers, families, social services, occupational therapists and speech and language therapists) and carried out by those with expertise in both ASD and sensory impairment (Edwards, 2004; Edwards & Crocker, 2008; Pring, 2005; Gentili & Holwell, 2011; Sessa & Sutherland, 2013). It is essential that full involvement of the families and carers is sought so that their views and concerns are taken into consideration (Steinberg *et al.* 2000; Fox *et al.* 2010; Macleod & Curtis, 2010; Salt, 2010). While a small number of carers/parents feel stigmatised with another label (e.g. ASD) for their sensory impaired child with ID, the diagnosis of ASD comes as a relief for most, as it helps them to understand why their loved ones behave the way they do and provides an explanation and support to adjust the environment and their communication with them (Brandsborg, 2002).

Raising awareness and training should, therefore, be prioritised and carers' views should be taken into consideration to make the management plan a success. Training will ensure awareness of sensory impairment for early detection and for any intervention required. Domokos (2000) found that the RNIB training course in multiple disability improved staff practice in the care of people with profound and multiple learning disabilities and that this was very beneficial for the service users. Training also facilitates the effective development of the communication passport and will help ascertain the level of support and how these could be secured through social or health care services.

8.9 Further work

Research projects have found different results when investigating the relationship between ASD and sensory impairments. This is likely due to the heterogeneity of those with ASD and sensory impairment e.g. their language skills, support they receive early on during their development from the significant others, their IQ and the degree and nature of brain damage (Pring, 2005). The research methodology should be therefore appropriate when assessing the ID service users with sensory impairment i.e. they need to be given adequate time, experience and opportunity (Perez-Pereira & Conti-Ramsden, 2005) so that they can show their abilities and that reliable conclusions can be drawn from the results: as Lewis and Collis (2005) put it, the research methods should be fit for purpose.

Another interesting research project on the topics relevant to this thesis which could be considered in the future, is the development of a new assessment tool for diagnosing ASD for people with severe to profound ID as, in spite of its high prevalence, ASD is often overlooked in this population in clinical practice. Improving diagnosis facilitates early access to services (Department of Health, 2010) which has been advocated by the National Institute for Health and Care Excellence (NICE, 2012).

Research shows that autistic traits change overtime and if service users' parents/cares are more engaged in therapeutic works and receive an autism friendly approach, some of the traits and maladaptive behaviours, might improve (Frith, 2003; Hobson and Lee 2010; Anderson *et al.* 2011). Improvement also seems to be related to service users' verbal IQ, adaptive functioning and language development; the higher these are the better the chance of improvement as they get older (Baghdadli *et al.* 2007; Gotham *et al.* 2012). An unpublished MSc project at Leicester Frith hospital also confirmed this assertion in a sample of autistic adult population with ID (Barrett, 2014).

Observations from follow-up work by Hobson and Lee (2010) suggest that symptoms of ASD may improve in blind children. Similarly, Jure *et al.* (2015)

reported an improvement in the symptoms of autism in 4 of their 18 congenitally blind children with ASD over time to the point that they no longer fulfilled the criteria for ASD in spite of having a confirmed diagnosis at an earlier assessment (please look at Appendix 1 for more detail). It is therefore important to carry out follow-up studies on congenitally blind children with ASD to find out more about any differences in presentation over time during their adulthood.

The effect of carers'/parents' mediated interventions guided by a psychologist or other therapists is another important area of study for future (Diggle *et al*. 2003). Further work should focus on these areas to tease out the beneficial impact of different interventions such as psychosocial approaches highlighted above in various sections of this thesis.

Further research is also needed to explore any association between epilepsy in people with blindness, ASD and ID. Previous research suggests that the association found between epilepsy and ASD is a function of degree of ID (Amiet *et al*. 2008; Viscidi *et al.* 2013), but more research is needed to determine the nature and magnitude of this association. The current project found no association between epilepsy and deafness when the degree of ID was taken into consideration. However, it did find an association between blindness and epilepsy even when controlled for degree of ID. Thus it is possible that, in addition to severity of ID, there are other significant factors that can affect development of ASD and/or epilepsy, such as the nature of the brain damage and its aetiology, the size and location of the brain damage, number of lesions (as shown by Waugh *et al.* in 1998 in the study of developmental set-back in congenitally blind children and by Bolton & Griffiths in1997 in the study of tuberous sclerosis), and other significant areas of brain affected.

Finally, the association between deafness and ASD needs further exploration in future, as studies on this association have rendered different outcomes and also because there is generally a dearth of information on this topic in the population of adults with ID.

APPENDIX 1: SUMMARY OF LITERATURE

Publication	Number of original research participants	Study population, setting and methods	Assessment tools and diagnostic criteria	Comparison groups	Findings and discussion
Keeler (1958) (quoted in Carvill, 2001)	75	n=40 with Retrolental fibroplasia (n=5 children referred to the Hospital for sick children; n=35 registered with the Canadian National Institute for the Blind). n=18 with congenital blindness due to other causes and n=17 due to postnatal blindness.	Developmental history and behavioural assessment	Children with Retrolental fibroplasia compared with children with congenital blindness and children with postnatal blindness	Children with Retrolental fibroplasia had striking similarity to infantile autism. Those with congenital blindness due to other causes had degree of withdrawal but did not show the same degree of autistic behaviour. Those with postnatal blindness had less conspicuous symptomatology. Autism thought to be due to combination of brain damage, blindness and maternal deprivation.
Chess (1971)	243	Preschool children with rubella at the age of 2 and half years to 5 years	Behavioural history and direct examination (using Kanner's criteria for autism)	-	n=10 had Kanner's autism and n=8 had partial autism (n=18; 7.4%). Brain damage was considered as the cause of autism and partial autism.

Appendix 1: Summary of literature (continued)					
Chase (1972)	263	Children with Retrolental fibroplasia	Parents interview, professionals ratings, medical histories & Rimland checklist E-2	-	Gradients of autistic-like symptoms in the group but none had infantile autism. Strong association between autistic-like features and neurological deficits as well as other abnormalities.
Chess (1977) & Chess *et al.* (1978)	210	n=205 from original sample and n=5 new cases of congenital rubella syndrome. Follow up of n=10 cases with Kanner's autism and n=7 with partial autism at the ages of 8-9 years (one child from the original sample with partial autism refused to participate).	Behavioural history and examinations (Kanner's criteria for autism)	Comparison with the same children studied in 1971 by Chess	Several of those with autism in the original sample could not be followed up. In the new sample, n=13 children qualified for a diagnosis of Kanner's autism (6.2%): n=3 with previously diagnosed Kanner's autism had recovered and one had improved; n=3 with partial autism had recovered and n=4 had worsened. There were n=3 new instances of autism. It was hypothesised that the course of autism was of a chronic infection (congenital rubella syndrome) in which recovery, chronicity, improvement, worsening and delayed appearance of the autistic symptoms were all found.

Appendix 1: Summary of literature (continued)					
Fraiberg (1977)	27	Children with congenital blindness referred for a guidance service	Review of medical findings and birth history, behavioural assessment and examination	-	n=7 children had clinical presentations resembling autism and the rest had autistic patterns. None of the children had an accompanying brain damage. It was postulated that either a central impairment, social deprivation or sensori-motor impoverishment had caused autism.
Jan *et al.* (1977)	92	Children with congenital visual impairment (n=65 with partial visual impairment; n=27 with light perception or less)	History and medical case review	-	Only n=3 had psychosis (or autism). Authors did not think that autism was common in congenitally blind children and autism was only diagnosed if the assessor was unfamiliar with the child.
Rogers & Newhart-Larson (1989)	10	Children of pre-school age with congenital blindness	CARS, ABC, DSM-III criteria for infantile autism, Reynell-Zinkin scales of development	Comparison of n=5 children with Leber's congenital amaurosis with n=5 children with congenital blindness from other causes, matched developmentally	Those with Leber's congenital amaurosis had autism but the others did not have autism. It was postulated that the neurological abnormalities, especially from cerebellar origin was the cause of autism.

Appendix 1: Summary of literature (continued)					
Gillberg *et al.* (1990)	28	Children with autism under the age of 3 years	Social history, observation, parents interview, examination, ABC, DSM-III-R, Griffith's developmental scale, questionnaire on autism, neurobiological investigations.	-	6-month follow-up confirmed autism in n=20 children, of whom 6 (30%) had moderate to severe conductive hearing impairment. The authors concluded that autism could be diagnosed before the age of 6 months.
Jure *et al.* (1991)	1150	Children with hearing impairment on the computer database of the one of the authors: n=387 severe to profound hearing impairment (without ID) from the St Joseph's School of the Deaf. n=277 new referrals with hearing impairment (biased towards those with complex conditions) to the neurologists. n=486 children (biased towards those who were very young and had ID) referred to the Auditory Evoked Response lab for audiological testing	Medical case files review for social and developmental history and a neurologist assessment of cognitive ability and autism	-	n=46 (30 boys/16 girls) had autism (4%). Delay in diagnosing autism of 4 years for n=11 & delay of 6 years for n=5. Delay in deafness diagnosis for n=10 (several years after diagnosis of autism). n=37 needed special education due to severity of hearing impairment. n=9 had "disastrous" educational programme. No association between severity of hearing impairment and severity of autistic symptoms. Association between severity of ID and autistic symptoms. Authors reported that it was extremely challenging to do cognitive assessment in children with autism and hearing impairment.

Appendix 1: Summary of literature (continued)					
Steffenburg (1991)	52	n=35 Children with autism and n=17 children with autistic-like symptoms.	DSM-III-R, neurobiological investigations, neuropsychiatric assessments, IQ testing, ABC	-	n=8 children had neurogenic hearing deficits: n=5 children had mild to moderate hearing impairment, n=1 had moderate, n=1 had severe and n=1 had profound hearing impairment. Author discussed challenges of diagnosing hearing impairment in children with autism.
Cass *et al.* (1994)	102	Children with congenital visual impairment	Retrospective developmental observation between the age of 16 months to two and half years of age	Comparison based on different categories of visual impairment.	Those with severe visual impairment and brain pathology had neurodevelopmental set-back. Improvement in vision during this period resulted in an improvement in development. Improvement in the environment was associated with improvement in development. Behaviours blocking the learning (stereotypical behaviours) caused deterioration in skill development.

Appendix 1: Summary of literature (continued)					
Brown *et al.* (1997)	43	3 groups of children: n=24 Children (3 to 9 years old) with congenital blindness from 6 Schools for the Blind, n=10 sighted children from mainstream schools and n=9 autistic children with ID from an earlier screening process.	Teacher reports including information in medical files, Assessment of verbal IQ using British Picture Vocabulary Scale (BPVS), Wechsler Pre-School and Primary Scale Intelligence (WPPSI) and Wechsler Intelligence Scale for Children-Revised (WISC-R), Direct behavioural observation, Assessments using CARS, BCDP, DSM-III-R	n=15 blind children were compared with 10 sighted (all had IQ >70) n=9 blind children were compared with n=9 autistic children (all IQ<70) All matched for age and verbal ability	Blind children had more autistic features. n=10 congenitally blind children fulfilled DSM-III-R criteria for autism. Only n=2 in blind group had a diagnosis of Kanner's autism compared with n=9 in sighted group. CARS score for blind children showed a broad range rather than a bimodal distribution to make a clear distinction between autism and no autism. Quality of autistic symptoms was different in blind children. Authors concluded that blindness added to the presentation & might be amenable to modification strategies if early in life.
Ek *et al.* (1998)	41	Children with blindness	DSM-IV, CARS, IQ, Behavioural and developmental history of medical case file	n=27 children with retinopathy of prematurity were compared with n=14 children with hereditary congenital blindness.	n=15 children with retinopathy had autism and n=4 had autistic-like features. n=2 with hereditary blindness had autism. All who were diagnosed with autism had ID. It was concluded that autism was related to the brain damage and that blindness contributed and intensified autistic symptoms. 'Blindism' considered separately from autism.

Appendix 1: Summary of literature (continued)					
Hobson *et al.*(1999)	18	Blind children from schools for the Blind and sighted children	DSM-III-R completed through interview with teachers, other assessment used: BCDP, CARS, the play items for disordered pre-schoolers	n=9 children with congenital blindness and autism were compared with n=9 sighted children with autism matched on chronological age and verbal mental age	None of the children with blindness were classically autistic and majority had different presentation from sighted autistic children. Authors suggested that the severity of autistic symptoms may improve over time with early interventions in educational or psychosocial domains.
Rosenhall *et al.* (1999)	199	Children and adolescents with autism (n=153 boys; n=46 girls)	IQ assessment, audiometry, autism assessment by a child psychiatrist/neurologist and a child psychologist (with excellent inter-rater reliability=100%), comprehensive neuropsychiatric assessment, DSM-III-R	-	Majority had mild to severe ID (n=143). n=19 had sensory-neural hearing impairment: n=2 (1.6%) had unilateral hearing impairment, n=7 had profound hearing impairment (3.5%) and n=10 had mild to moderate hearing impairment. 18% of autistic children had hyperacusis. 23.5% had otitis media. 18.3% had an otitis media related conductive hearing loss. Most children needed more than one test to assess their hearing with n=17 children needing 3 tests for this purpose.

Appendix 1: Summary of literature (continued)					
Carvill & Marston (2002)	18	Referred adults (>18 years) from residential homes run by SENSE, Deaf-Blind UK and Rubella Association to an ID service in the UK (South Birmingham service) (n=12 males; n=6 females)	ICD-10 criteria for pervasive developmental disorder and level of ID, blood investigations, EEG, observations, history from carers and families, review of medical records	-	In n=12 (71%) the cause of sensory impairment was congenital rubella syndrome. n=2 had Leber's congenital amaurosis. The rest had Joubert syndrome, self-injury resulting in blindness, infection during infancy and Rhesus haemolytic anaemia as the cause of their blindness and ID. All n=18 adults had moderate to severe ID. n=15 had a diagnosis of pervasive developmental disorder (n=8 atypical autism due to lack of early developmental history or full symptomatology; n=7 autistic traits). n=3 had hearing impairment, of whom n=2 were diagnosed with autism. n=4 had visual impairment, of whom n=3 were diagnosed with autism. n=11 had both visual and hearing impairment of whom n=10 had autism.

Appendix 1: Summary of literature (continued)					
Dale & Sonksen (2002)	69	Children with 'potentially simple' congenital disorders of the peripheral visual system (n=40 males; n=29 females)	Developmental and visual assessments at 10 to 16 months (Time 1) and 27 to 54 months of age (Time 2). Developmental status was determined by using the Reynell-Zinkin scales.	Comparison of groups according to visual status: profound visual impairment (PVI), severe visual impairment (SVI)	Majority of the sample showed normal development on all subscales (62% Time 1, 57% Time 2). PVI were more developmentally vulnerable than SVI with a greater incidence of: Uneven developmental profile at Time 1 (48% PVI, 16% SVI); Global learning difficulties at Time 2 (37% PVI, 0% SVI); Delay on individual subscales at Time 2 ($p<0.02$ PVI versus SVI); Deceleration in skills (verbal comprehension 74% PVI, 24% SVI, sensorimotor understanding 70% PVI, 27% SVI); and Severe developmental set-back (33% PVI, 7% SVI).

Appendix 1: Summary of literature (continued)					
Kielinen *et al.* (2004)	152,732	Population-based survey of children and adolescents aged under 16 years living in northern Finland	Diagnoses and associated medical conditions derived from hospitals and institutional records of this area DSM-IV criteria used for identifying those with autism	-	n=187 had autism. Associated medical disorders or associated disorders of known or suspected genetic origin were found in 12.3 percent. Other comorbidities were CP, epilepsy, etc. Hearing impairments were identified in 8.6 percent and severe vision impairment in 3.7 percent of those with autism. The authors concluded that comorbid medical disorders seemed to have a special impact on the genesis of autism.
Mukaddes *et al.* (2007)	257	Blind children, aged 7-18 years, were assessed in various settings through a 3-stage assessment process	Direct observation, using ABC & DSM-IV criteria for diagnosing autism Identifying various comorbidities in all children	Comparing those with a diagnosis of autism with those without a diagnosis of autism as regards comorbidities	n=30 children had a diagnosis of autism. Children with blindness and autism had greater chance of neurological impairment e.g. ID, CP. Children with autism had more severe visual impairment than the subjects without autism.

Appendix 1: Summary of literature (continued)					
Hoevenaars-van den Boom *et al.* (2009)	95	Both children and adults with deaf-blindness and ID recruited from a deaf-blind institution	Ophthalmology/ orthoptist assessment, audiology assessment, IQ assessment by a psychologist, an expert panel assessment of autism (consisting of a psychologist, a psychiatrist and an expert in deaf-blindness) by observation and video recording for comments by the panel. Instruments used: O-ADB (based on ADOS-R), ASIEP, ADI-R and Hands-on assessment	n=10 people met the inclusion criteria for the final stage of the project whose carers consented to participate: n=5 with autism (diagnosed based DSM-IV criteria) were compared with n=5 without autism. All had profound ID.	Aetiology of deaf-blindness and ID were as follows: congenital rubella syndrome (n=3), CHARGE syndrome (n=2), Zellweger syndrome (n=1), West syndrome (n=1), Goldenhar syndrome (n=1), Trisomie 22 (n=1) Prematurity (n=1) All with deaf-blindness (n=10) showed impairment in social interaction, communication and language. Those with autism (n=5) showed significantly more impairment on their skills. Two groups were not different regarding stereotypical behaviours, problem solving strategies and play/exploratory skills.
Hobson & Lee (2010)	16	Follow up of n=9 congenitally blind and n=7 sighted children who had been diagnosed with autism several years ago (please look at Hobson *et al.* 1999).	Discussion with teachers, observation, DSM-IV criteria of autism, CARS, BCDP	Comparison of these two groups of children matched on chronological age and verbal ability	n=8 out of n=9 blind children originally diagnosed with autism no longer met the diagnostic criteria for autism based on BCDP and CARS. All n=7 sighted children still fulfilled autism diagnostic criteria.

Appendix 1: Summary of literature (continued)					
Parr *et al.* (2010)	83	n=45 females and n=38 males (10 months to 6 years 10 months old) from a specialist developmental vision service	Retrospective case-note study of clinic records of children using standardized assessments of vision, development and clinician judgements about social, communication, and repetitive/restrictive behavioural (SCRR) difficulties & clinical assessment of autism by a multidisciplinary team.	Comparing various groups based on the aetiology and degree of blindness e.g. children with optic nerve hypoplasia (ONH), septo-optic dysplasia (SOD), profound visual impairment (PVI) or severe visual impairment (SVI)	58% of children had at least one SCRR difficulty, and 31% had a clinical diagnosis of autism. The prevalence of autism was higher in children with SOD than in children with ONH (36% vs 26%). This was also slightly more frequent in children with PVI than in children with SVI (36% vs 27%). The prevalence of SCRR difficulties was higher in children with PVI than in children with SVI (p=0.003).
Kancherla *et al.* (2013)	230,973 of 8-year-olds children	Data from the population-based Metropolitan Atlanta Developmental Disabilities Surveillance Program among 8-year-olds in metropolitan Atlanta 2000–2008 Children identified from record review at nine public school systems and selected private and public health sources reviewed by a team of clinician reviewers to determine final case status	Period prevalence of autism (based on DSM-IV), birth and parental characteristics, presence and severity of other developmental disabilities and age of earliest identification of autism were determined for children with sensory impairment	Comparison of children based on the presence or absence of autism or hearing and visual impairment (HI, VI)	VI and HI prevalence were respectively 1.2 and 1.3 per 1000 of children aged 8-year-olds. Approximately 5.8% of children with HI had autism in comparison to 7.2% of those with VI who had autism. Those with autism and VI and HI had a higher chance of developmental disability than those without HI or VI. Autism had been diagnosed at a later age in those with VI than those without VI (median age of 79 months versus 56 months).

Appendix 1: Summary of literature (continued)					
Begeer *et al.* (2014)	152	n=18 children with blindness were excluded. Children with congenital ocular-plus blindness (n=22), congenital ocular blindness (n=9) and sighted children (n=103). Children were recruited from regular and special schools in two countries: Germany and the Netherlands. All children were below 9 years old	Questionnaire sent to teachers, families and professionals to ensure children did not have autism or ID. Assessment based on performance on ToM tasks designed for children with blindness (nine tactile and auditory first-order false-belief tasks), memory and language tests	Performances of children with congenital ocular-plus blindness and congenital ocular blindness were compared with sighted children	The ocular-plus blind group (n=22) included those with ROP (n=5), Leber's congenital amaurosis (n=13), Norrie disease (n=2), optic nerve hypoplasia (n=1), and optic nerve atrophy (n=1). The ocular blind group (n=9) included those with microphthalmus (n=6), congenital cataracts (n=1), and infantile glaucoma (n=2). ToM performance was delayed in children with ocular-plus blindness, but not in children with ocular blindness. The findings suggest a connection between optic neural pathway involvement and ToM development of children with ocular-plus blindness.

Appendix 1: Summary of literature (continued)					
Jure *et al*. (2015)	125	n=38 unselected congenitally blind children from a school for the blind	DSM-IV diagnostic criteria for autism, medical and neurological assessment, Administering Developmental, Dimensional and Diagnostic Interview for autism Assessment of the course of autism	-	n=20 children had retinopathy of prematurity. n=10 children had malformations (anophthalmia, microphthalmia, and optic nerve hypoplasia). n=8 with other aetiologies: n=1 vitreous and retinal degeneration, n=1 aniridia and vitreous dysgenesis, n=2 optic nerve atrophy following neonatal asphyxia, n=1 asphyxia at 4 months resulting in severe brain damage, cortical blindness, and mild optic atrophy. n=3 children had acquired blindness. n=19 children had autism. Autism was linked to total congenital blindness, not blindness aetiology, acquired or incomplete blindness, sex, overt brain damage, or socioeconomic status.

ABC: Autism Behaviour Checklist; ADI-R: Autism Diagnostic Interview- Revised; ADOS-R: Autism Diagnostic Observation Schedule- Revised; ASIEP: Autism Screening Instrument for Educational Planning; BCDP: Behaviour Checklist for Disordered Pre-schooler; CARS: Childhood Autism Rating Scale; CP: cerebral palsi; DSM-- III or IV- R: Diagnostic and Statistical Manual of Mental Disorders-version III or IV- Revised; EEG: electroencephalography; ICD-10: International Classification of Diseases-10th Revision; ID: intellectual disability; IQ: intelligence quotient; O-ADB: Observation of characteristics of Autism in persons with Deaf-Blindness; ROP: retinopathy of prematurity; ToM: theory of mind;

APPENDIX 2: ETHICAL APPROVAL

Nottingham Research Ethics Committee 1
1 Standard Court
Park Row
Nottingham
NG1 6GN

Telephone: 01159123344 Ext: 39368
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11 April 2008

Dr Abdolreza Ashtarikiani
Specialist registrar in learning disability
Leicestershrie Partnership NHS Trust
Learning disability services,
Mansion House
Leicester Frith Hospital, Leicester
LE3 9QF

Dear Dr Ashtarikiani,

Full title of study: **Sensory (visual/hearing) impairment, mental ill health and autistic spectrum disorder in people with intellectual disability**
REC reference number: **08/H0403/8**

Thank you for your letter of 02 April 2008, responding to the Committee's request for further information on the above research and submitting revised documentation.

The further information has been considered on behalf of the Committee by the Vice-Chair.

Confirmation of ethical opinion
On behalf of the Committee, I am pleased to confirm a favourable ethical opinion for the above research on the basis described in the application form, protocol and supporting documentation as revised.

Mental Capacity Act 2005
I confirm that the committee has approved this research project for the purposes of the Mental Capacity Act 2005. The committee is satisfied that the requirements of section 31 of the Act will be met in relation to research carried out as part of this project on, or in relation to, a person who lacks capacity to consent to taking part in the project.

Ethical review of research sites
The Committee has designated this study as exempt from site-specific assessment (SSA). There is no requirement for [other] Local Research Ethics Committees to be informed or for site-specific assessment to be carried out at each site.

Conditions of approval
The favourable opinion is given provided that you comply with the conditions set out in the attached document. You are advised to study the conditions carefully.

Approved documents
The final list of documents reviewed and approved by the Committee is as follows:

Document	*Version*	*Date*
Application	AB/117888/1	12 December 2007
Investigator CV	Student/CI	01 December 2007
Investigator CV	Supervisor	01 December 2007
Protocol	2	01 April 2008
Summary/Synopsis - Flowchart	1	01 December 2007
Letter from Sponsor		23 October 2007
Peer Review		29 October 2007
Statistician Comments		07 December 2007
Letter of invitation to participant - Carer/Participants	1	01 December 2007
GP/Consultant Information Sheets	2	01 April 2008
Participant Information Sheet: Research	2	01 April 2008

participants		
Participant Information Sheet: RMO	1	01 December 2007
Participant Consent Form: RMO	1	01 December 2007
Participant Consent Form: Participants with Learning Disability	2	01 April 2008
Participant Consent Form: Research participants	2	01 April 2008
Response to Request for Further Information		02 April 2008
Vision Checklist		
Checklist for hearing		
PDD-MRS		
Aberrant Behaviour Checklist (ABC) - Community		
Sensory impairment Performa	1	01 December 2007

R&D approval

All researchers and research collaborators who will be participating in the research at NHS sites should apply for R&D approval from the relevant care organisation, if they have not yet done so. R&D approval is required, whether or not the study is exempt from SSA. You should advise researchers and local collaborators accordingly.

Guidance on applying for R&D approval is available from http://www.rdforum.nhs.uk/rdform.htm.

Statement of compliance

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees (July 2001) and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

After ethical review

Now that you have completed the application process please visit the National Research Ethics Website > After Review

Here you will find links to the following

a) Providing feedback. You are invited to give your view of the service that you have received from the National Research Ethics Service on the

application procedure. If you wish to make your views known please use the feedback form available on the website.

b) Progress Reports. Please refer to the attached Standard conditions of approval by Research Ethics Committees.

c) Safety Reports. Please refer to the attached Standard conditions of approval by Research Ethics Committees.

d) Amendments. Please refer to the attached Standard conditions of approval by Research Ethics Committees.

e) End of Study/Project. Please refer to the attached Standard conditions of approval by Research Ethics Committees.

We would also like to inform you that we consult regularly with stakeholders to improve our service. If you would like to join our Reference Group please email referencegroup@nationalres.org.uk .

With the Committee's best wishes for the success of this project.

08/H0403/8	**Please quote this number on all correspondence**

Yours sincerely

Mr R Johnson / **Ms Trish Wheat**
Vice Chair / **Co-ordinator**

Email: trish.wheat@nottspct.nhs.uk

Enclosures: *Standard approval conditions*

Copy to: *R&D Department for NHS care organisation at lead site – LPT*
Dr S Bhaumik - Consultant Psychiatrist and Lead Clinician

APPENDIX 3: LEICESTERSHIRE LEARNING DISABILITY REGISTER INTERVIEW SCHEDULE

LEICESTERSHIRE LEARNING DISABILITIES REGISTER QUESTIONNAIRE

ID No: |__|__|__|__|__| **Date of interview:** |__|__| |__|__| |__|__|__|__|

Interview No: |__|__| **Interviewer Code:** |__|__|

Enter X if not to be computerised |__|

Principal Interviewee Client = 1 Carer = 2 Supporter = 3 |__|

Title (Mr Mrs Ms etc) |__|__|__|__|

Forename (or initials)

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Family Name

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Relationship (to client) |__|__|

SECTION I: CLIENT

Q.1.1. **Title** (Mr Mrs Ms etc) |__|__|__|__|

Q.1.2. **First name(s):**

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Q.1.3. **Preferred name(s):**

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Q.1.4. **Family Name:**

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Q.1.5. **Alternative Name:**

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Q.1.6. **Date of birth**: |__|__| |__|__| |__|__|__|__|

Q.1.7. **Sex:** M/F |__|

Or approximate age: ___________ years (use to estimate DOB)

Q.1.8. **Address of local main residency (and name of residency)**:

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

GF Ward Code (GF only)

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|
|__|__|__|__|

Town: |__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Postcode: |__|__|__|__| |__|__|__| Team Code: |__|__|__|__|

Out of county residential placement? Yes = 1 No = 2 |__|
(other than Leicestershire, Rutland or Leicester City)

Q.1.9. **Do you have a telephone?** Yes = 1 No = 2 |__|

If yes, Tel. No. |__|__|__|__|__|__|__|__|__|__|__|__|__|
(Enter full std code)

Ex-directory |__|

Q.1.9a. **Interviewee's e-mail address:**
……………………………………………………………

Q.1.10. **General Practitioner:**

Name |__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Name of Surgery |__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Q.1.10a. **Are you able to tell me your NHS number?** Yes = 1 No = 2 |__|

NHS number |__|__|__|__|__|__|__|__|__|__|

Q.1.11. **Marital status:**

Married = 1 Single = 2 Widowed = 3 Divorced/separated = 4 |__|

Q.1.11a **How long has s/he had a learning disability?** (Interviewee opinion) |__|

Around the time of birth (< 1 year) 1

Before school age (1-4 years)	2
Childhood (5-9 years)	3
Later childhood (10-14 years)	4
Adolescence (15-19 years)	5
Later	6
Not known how long s/he has had a learning disability	7
Not sure whether s/he has a learning disability	8
No learning disability.	9

Q.1.11b **Has s/he ever been diagnosed as having Down's Syndrome?**

Yes = 1 No = 2 |__|

Q.1.12. **Place of birth:**

Leicestershire/Rutland = 1 Other UK = 2 Outside UK = 3 Not known = 9 |__|

Q.1.12a **Was the mother resident in Leicestershire/Rutland at the time?**

Yes = 1 No = 2 N/K = 9 |__|

Q.1.13. **Year of migration to UK if appropriate:** |__|__|__|__|
(If not appropriate enter 8888)

Q.1.14. **What culture (religion) does s/he belong to?** |__|

Christian	1	Muslim	4	None	0
Hindu	2	Rastafarian	5	Other*	8
Sikh	3	Jewish	6	Not known	9

*Specify

_____

Q.1.15a. **Ethnic group of the client: (show card)**

White	
British	11
Irish	12
Other European	13
Other*	14
Asian	
Indian	21
Pakistani	22
Bangladeshi	23

Other*	24
Black	
Caribbean	31
African	32
Other*	33
Mixed	
White & Black Caribbean	41
White & Black African	42
White & Asian	43
Other*	44
Other	
Chinese	51
Middle East	52
Other*	53
Not known	99

|__|__|

*Specify _____

Q.1.16. **What is the client's main language that s/he speaks or understands?** |__|

English	= 1	Punjabi	= 6
Other European	= 2	Urdu	= 7
Bengali	= 3	Other*	= 8
Gujarati	= 4	Not known	= 9
Hindi	= 5	None	= 0

*Specify _____

Q.1.17. **Does s/he speak or understand any other languages?**

Use codes as before. (if none, enter 0000) |__|__|__|__|

Q.1.18. **Client lives in:**

Type of accommodation	**Code**
Alone	01
With parent(s)	02
With spouse/partner	03
With other relative(s)	04
Foster home/Family placement/Guardian	05
Other unstaffed home/Friends/Cohabitee	06
New (un)registered 'home' (<4)	07
Registered private residential 'home'	08
Voluntary staffed 'home'	09

Voluntary unstaffed① 'home'	29	
NHS staffed group 'home'/community unit	10	See coding manual
NHS hospital	11	for complete code
SS staffed hostel	12	
SS unstaffed① group 'home'	13	
SS 24-hour support	23	
Recognised Residential Home	28	
Housing association accommodation (specific LD)	14	
No fixed abode	16	
Other*	15	

Enter complete code |__|__| |_____|

① "unstaffed" means no staff are 'living in' the accommodation

* Other - specify type of accommodation _____

If code 1520 give details of address (i.e. residential out of county)

* Address _____

_____

_____

Q.1.18a **Does the funding come from an authority other than Leicestershire, Rutland or Leicester City?**

Yes = 1 No = 2 N/K = 9 |__|

SECTION II (a): HOUSEHOLD/FAMILY MEMBERS

(Complete for main Leicestershire residence)

Q.2.1. **Who <u>else</u> lives in the household?**

				Special needs ④		
Rel ①	**Sex**	**YOB ②**	**Emp ③**	**Other**	**PH**	**Carer**

						(b)	**(a)**	⑤
1.	|__|__|	|__|	|__|__|__|__|	|__|	|__|	|__|	|__|
2.	|__|__|	|__|	|__|__|__|__|	|__|	|__|	|__|	|__|
3.	|__|__|	|__|	|__|__|__|__|	|__|	|__|	|__|	|__|
4.	|__|__|	|__|	|__|__|__|__|	|__|	|__|	|__|	|__|
5.	|__|__|	|__|	|__|__|__|__|	|__|	|__|	|__|	|__|
6.	|__|__|	|__|	|__|__|__|__|	|__|	|__|	|__|	|__|
7.	|__|__|	|__|	|__|__|__|__|	|__|	|__|	|__|	|__|
8.	|__|__|	|__|	|__|__|__|__|	|__|	|__|	|__|	|__|
9.	|__|__|	|__|	|__|__|__|__|	|__|	|__|	|__|	|__|
10.	|__|__|	|__|	|__|__|__|__|	|__|	|__|	|__|	|__|

① Use coding manual for 2-digit code

② Estimate, if necessary

③ <u>Employment codes:</u>

Had a job last week	1	Student/Trainee	4	Day-centre/Sheltered	7
Retired	2	House-person	5	School	8
Long term sick/disabled	3	Unemployed last week	6	Others	9
				Not known	0

④ <u>Special needs:</u> Ask - are they able to look after themselves? i.e. Do any of them need support in any way? Did they go to a special school?
Yes = 1 No = 0

For: a) <u>people with a physical disability</u>
e.g. problems with walking, vision, amputee, very frail elderly etc – see background notes for more details

and: b) <u>other</u>
e.g. learning disability. Long term emotional or behavioural problems, general impression of "something seriously amiss".

⑤Main carer=1 Other significant carer(s)=2 Main supporter=3 All other(s)= 0.

Q.2.3. **Main carer/supporter**. Is s/he:

Carer = 1 Supporter = 2 or: If there is no carer = 3 |__|

IF NO CARER/SUPPORTER, GO TO Q.2.11

Q.2.3a. **Residence of main carer**

Resident in household = 1 Non-resident member of family = 2 |__|

Non-resident other = 3

Q.2.4. **Complete all questions on main carer/supporter where appropriate. Main Carer/Supporter:**

Title (Mr,Mrs,Ms etc) |__|__|__|__|

Forename

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Surname

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Relationship to client |__|__|

Date of birth: |__|__| |__|__| |__|__|__|__| Sex: M/F |__|

If non-resident i.e. '2' or '3' above complete the following:

Address:

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Town

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Postcode |__|__|__|__| |__|__|__|

Contact Tel No |__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Q.2.5. **What is the main carer's/supporter's main language that s/he speaks or understands?** |__|

English	1	Punjabi	6
Other European	2	Urdu	7
Bengali	3	Other*	8
Gujarati	4	None	0
Hindi	5	Not known	9

* Specify

_____

Q.2.6. **Does the main carer/supporter speak or understand any other language?**

Enter codes as in Q.2.5 (if none, enter 0000)
|__|__|__|__|

Q.2.7. **If appropriate i.e. English not spoken fluently by main carer, is there an**

Interpreter readily available? |__|

Resident in household	1	None	3
Non-resident family/Other	2	Not applicable	8

Q.2.8. **If the main carer/supporter were ill/had to go into hospital for a relatively short time e.g. 2-3 weeks, who would take over?** |__|

In household	1
No-one in household (need hospital/other accommodation)	2
No-one in household (other person available)	3
No-one in household (capable and looks after self)	4
Care service needed in client's own home	5

HOUSE

Q.2.11. **Tenure: How do you and your household occupy your accommodation?** |__|

As an owner-occupier	1
By renting, rent-free or lease	2
In some other way	0
Not applicable (e.g. no fixed abode)	8
Not known	9

Q.2.12. **Type of House:** |__|

Ordinary 2/3 storey house	1
Bungalow	2
Flat/maisonette (including multi-storey)	3
Mobile home	4
Homeless/temporary accommodation e.g. salvation army/host.	5
Other	6

Q.2.13. **Access to front or back door:** |__|

Flight of stairs, no lift	1
Steps only	2
Lift	3
None/one step only	4

Q.2.14. **Inside stairs:** __|

Flight of stairs	1
Steps only	2
Chair/Lift	3
None	4

Q.2.15. **Amenities:** Yes = 1, No = 2

a) WC on ground floor |__|
inside |__|
WC outside |__|
WC upstairs

b) Bathroom on ground |__|
floor |__|
Bathroom upstairs

Q.2.16. **Do you have any form of central heating, including electric storage heaters, in your (part of the) accommodation?**

Yes = 1 No = 2 |__|

Q.2.18. **Does the family feel they need help with re-housing or to move?** |__|

Required (<1 year)	1	Long-term need (>2 years)	3
Non-urgent need (1-2 years)	2	No foreseeable need	4

MOTOR VEHICLES

Q.2.19. **Do you have a car or van available? If so, how many?** |__|

None = 0 One = 1 Two = 2 Three or more = 3

(include any car or vehicle provided by employer)

SECTION II (b): RESIDENTIAL HOMES

Q.2.26. **Date of admission to the residential home or similar accommodation:**

To be completed by all people <u>except</u> Leicester Frith Hospital and Gorse Hill Hospital residents

|__|__| |__|__|

|__|__|__|__|

Q.2.27. **Most recent <u>admission</u> date to any GF home or hospital**
|__|__| |__|__|
|__|__|__|__|

Q.2.33. **How many other long-term residents are there in this home/hostel/ward**
|__|__|

SECTION II (c): OTHER

Complete as appropriate

INFORMAL SUPPORT

This is informal (<u>unpaid</u>) support. Only appropriate if there is no main carer.

Q.2.24a. (i) **Is there any active family support/other background figure?**

Yes = 1 No = 2 |__|

Relationship to client |__|__|

(ii) **Can this person be contacted for any service queries?**

Yes = 1 No = 2 |__|

If appropriate complete the following:

Title (Mr,Mrs,Ms etc) |__|__|__|__|

Forename

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Surname

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Date of Birth |__|__| |__|__| |__|__|__|__|

Estimate from: approx. age _____

Sex: M/F |__|

Address

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Town

|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|__|

Postcode |__|__|__|__| |__|__|__|

Contact Tel No |__|__|__|__|__|__|__|__|__|__|__|__|

Q.2.24d. **Approximate number of hours per week provided by main informal carer**

|__|__|__|

Q.2.24e.**Is there any extra informal care?** (number of hours)
|__|__|__|

Q.2.24f. **Does this include overnight support?** Yes = 1 No = 2 |__|

FORMAL SUPPORT

Usually appropriate for clients living alone or in unstaffed accommodation with other disabled people only, whether in a registered home or not.

This is formal <u>(paid)</u> support

Q.2.24b. **Is there any <u>outside</u> supporter (formal)?** Yes = 1 No = 2 |__|

If yes, complete the following:

Relationship to client |__|__|

Q.2.24g. **Approximate number of hours per week provided by main formal carer**

|__|__|__|

Q.2.24h. **Is there any extra formal care – (number of hours)?** |__|__|__|

Q.2.24i. **Does this include overnight support?** Yes = 1 No = 2 |__|

Q 2.24j **Organisation**

_____

Q 2.24k **Contact Phone No.** _____

SECTION II (d): ALL

Appropriate for household/family members and residential care.

ADAPTATIONS AND MODIFICATIONS

Q.2.17a. **Are any modifications needed to present housing/accommodation on behalf of the client?**

Yes = 1 No = 2 |__|

b. **What modifications are needed?**

Complete for up to 6 modifications using the following codes.

bathroom/interior/aids	21	Plumbing	31
installation of bathroom	22	Doors	32
toilet/interior/aids	23	Windows	33
installation of a toilet	24	Roofs	34
installation of a shower	25	Floor	35
stair aids/rail	26	Ramp	36
other rails	27	Damp	37
modification/ other room installation	28	garden/gate	38
kitchen interior/aids	29	re-housing	39
heating	30	automatic/computerised control	40
misc/other	99		

If the client is living in a household/with family members, also state whether the modifications are:

for the client = 1
for someone else = 2
for both = 3

Modification	Who for *Household only*	Modification	Who for *Household only*
1. |__|__|	|__|	4. |__|__|	|__|
2. |__|__|	|__|	5. |__|__|	|__|
3. |__|__|	|__|	6. |__|__|	|__|

SECTION III: ALL

VISION

Q.3.2. **Does s/he have spectacles or contact lenses?** (Do not assess whether worn)

Yes = 1 No = 2 |__|

Q.3.1. **Is s/he able to see normally?** (Rate with spectacles or contact lenses if worn) |__|

1 Blind or almost
2 Poor/partially sighted
3 Normal

HEARING

Q.3.4. **Does s/he have a hearing aid?** (Do not assess whether worn)

Yes = 1 No = 2 |__|

Q.3.3. **How is his/her hearing?** (Rate with hearing aid if worn) |__|

1 Deaf
2 Poor
3 Normal

SPEECH OR GESTURING

Q.3.5. **Can s/he speak or gesture? How does s/he let you know what s/he wants?**

(Rate with aid if used) |__|

1 Little or nothing, or meaningless echolalia
2 A few sounds or concrete gestures such as pulling you by the hand, or pointing
3 Mostly gestures or signs
4 Mixture of speech and gesture
5 Can make her/himself understood by speech alone

Q.3.5a. **Does s/he have a speaking aid?** Yes = 1 No = 2 |__|

Q.3.5b. **If yes, what is it?** (Enter code) |__|__|

N/A = 88

UNDERSTANDING

Q.3.6. **Is s/he able to understand instructions? Does s/he understand if you ask her/him about things s/he has done?** |__|

1 Little or nothing
2 Understands a few simple commands (e.g. come here, sit down)
3 Understands a fair range of instructions related to practical needs
4 Understands comments, questions and instructions related to personal needs and experiences (eg. did you enjoy the trip to the seaside?)
5 Understands information about things outside her/his immediate experience (e.g. major items of current news)
6 5 and can make an informed decision (e.g. about using a service such as the LD register)

LEVEL OF COMMUNICATION

Speech/Gesture/Drawing/Communication aid

Q.3.7. **Can s/he ask for things he wants?**
Can s/he talk about things he has done? |__|

1 Little or nothing, or meaningless echolalia
2 Uses a few words or signs (e.g. hello, bye-bye, drink)
3 Uses words or signs for practical needs - variety of needs
4 Uses words or signs to comment on her/his own personal experience (e.g. tells people s/he has new clothes that s/he has been on an outing, that someone has done something wrong etc)
5 Can converse, in words or signs, about things outside his/her own personal experience (e.g. makes spontaneous comments about things such as TV programmes)
6 5 and can communicate a decision (e.g. about using a service such as the LD register)

PRONUNCIATION

Q.3.8. **How clear is her/his speech? How easy is it to understand?**
Rate on spontaneous speech not meaningless echolalia. |__|

0 Not enough spontaneous speech to rate, or only meaningless echolalia
1 Difficult to understand even by close acquaintances. Impossible for strangers
2 Easily understood by close acquaintances. Difficult for strangers
3 Clear enough to be understood by anyone

SIGNING

Q.3.9. **Does s/he use signing? How clear is her/his signing?** |__|

0 Not enough spontaneous signing to rate
1 Difficult to understand even by close acquaintances. Impossible for strangers
2 Easily understood by close acquaintances. Difficult for strangers
3 Clear enough to be understood by anyone fluent in use of sign language

5 Don't know how clearly he/she signs

READING

Q.3.10**. Can s/he read?** |__|

1 Unable to recognize any writing
2 Recognizes own name written down
3 Can read and act appropriately to signs giving directions in shops or in the street
4 Can read and follow a series of written instructions
5 Can read newspapers and books

WRITING

Q.3.11. **Can s/he write? – using pen/keyboard** |__|

1 Unable to write
2 Writes name and/or copy writing
3 Writes full name and address without help
4 Can write short notes, e.g. shopping lists
5 Can write own simple correspondence
6 5 and can write own complex correspondence

FEEDING AND DRINKING

Q.3.12. **Does s/he feed himself?** |__|

1 Not at all
2 With help
3 Supervision/prompting only
4 Without help

PREPARING FOOD

Q.3.13. **Does s/he prepare his food?** |__|

1 Needs all food prepared for her/him/no opportunity and not known
2 With supervision, can prepare simple foods
3 Makes up food which does not require cooking or with which s/he is familiar - cereals, teas, sandwiches
4 Prepares simple hot food without supervision - boils eggs, warms soups.
5 Prepares an adequate variety of meals without supervision
6 5 and prepares a **wide** variety of meals without supervision

WASHING AND BATHING

Q.3.14. **Does s/he wash himself?** |__|

1 Not at all
2 With help on a daily basis
3 Help with shaving only
4 Help with menstruation only
5 Supervision/prompting only (including minimal help with hair washing + bath only e.g. running the bath)
6 Without help

DRESSING

Q.3.15. **Does s/he dress himself?** |__|

1 Not at all
2 With help
3 With supervision/prompting/help with zips and buttons
4 Without help - and can manage zips and buttons

TOILETING

Q.3.16. **Does s/he go to the toilet by her/himself?** |__|

1 Dependant on physical assistance
2 May ask to go but requires help
3 Requires some prompting/verbal assistance/minimal help
4 Goes alone – independent

CONTINENCE

Q.3.17a. **Is s/he clean and dry?** (Assess resultant continence on usual toileting regime)

How often does s/he wet/soil? (regardless of use of aids)

Wetting:

Night: 1 = At least once per night |__|
2 = At least once per week
3 = At least once per month
4 = At least once per year
5 = No problem

Day: 1 = At least once per day |__|
2 = At least once per week
3 = At least once per month
4 = At least once per year
5 = No problem

Comment: .....code |__|__|

(or leave blank)

Q.3.18a. **Soiling**:

Night: 1 = At least once per night |__|
2 = At least once per week

3 = At least once per month
4 = At least once per year
5 = No problem

Day: 1 = At least once per day |__|
2 = At least once per week
3 = At least once per month
4 = At least once per year
5 = No problem

Comment: .....code |__|__|
(or leave blank)

Q.3.19. **If incontinent of urine, has it developed in the last year?**

Yes = 1 No = 2 N/A = 8 |__|

CONTINENCE AIDS

Q.3.20. **Does s/he use and/or need any pads/pants/appliances for continence?**

Code each item using:	Use	Sufficient	Code
	Yes	Sufficient	1
	Yes	Insufficient	2
	No	OK without	3
	No	In need	4

1. Pads/pants/nappies etc |__|
2. Catheter/tubing |__|
3. Sheets/rolls |__|
4. Commode |__|
5. Other/unspecified |__|

MOBILITY

Q.3.21. **Is s/he able to walk by himself?** |__|

1 Non-mobile
2 Fully mobile in wheelchair
3 Needs help/walking frame on flat
4 Needs help up stairs but unaided on flat - usually for short distances
5 May be able to walk unaided for distances up to half mile/ perhaps extremely
6 slow*
7 Walks unaided everywhere
Mobility and balance normal. Restricted for other reasons* e.g. refuses to walk or runs away persistently.

* Comment .....code |__|__|

(or leave blank)

MOBILITY AIDS

Q.3.22. Does s/he use any mobility aids?
(Enter up to 4 codes)
|__|__|__|__|

0	None	3	Walking frame	6	Buggy
1	Wheelchair	4	Callipers	7	Artificial Limb
2	Sticks	5	Special shoes	8	More than 4 aids
				9	Other

DOMESTIC SKILLS - AROUND THE HOUSE

Q.3.23. **Does s/he give any help with clearing up or tidying up?** |__|
Does s/he do anything useful for you?

1 Unable to do any household jobs
2 Attempts simple household jobs (e.g. setting the table, dries dishes) but cannot do them properly
3 Able to do these simple repetitive jobs properly
4 Attempts most jobs but needs supervision and help to complete the job properly
5 Capable of doing most jobs around the house without supervision - makes bed, washes and dries dishes, cleans floor etc
6 5 and capable of doing all jobs around the house without supervision - files papers, put away household goods

ORIENTATION

Q.3.24. **How far can s/he find her/his way around without help? *** |__|

1 Cannot find way around, even inside residence
2 Can find way around residence only
3 Can find way around residence and immediate environs (garden, grounds)
4 Can find way to local shops (or equivalent e.g. post box)
5 Can find way to town
6 Can travel longer distances on own
7 6 and can cope with getting lost

(* If insufficient road sense code 3)

MONEY

Q.3.25. **Does s/he understand money?** |__|

1 No understanding of money
2 Picks out coins by name, e.g. 50p, 10p etc
3 Estimates roughly what different amounts might buy, e.g. if given 50p has some idea of what he/she could get for that
4 Can select the money appropriate to stated price of article
5 No difficulty in coping with everyday money transactions: giving right amount and checking change but unable to use money fully responsibly.
6 Able to use money responsibly

TIME

Q.3.26. **Does s/he understand time?** |__|

1 Seems to have no idea of time
2 Shows by behaviour that s/he can anticipate some events of the day, e.g. start of TV programme
3 Knows what hour it is by the clock
4 Able to keep a sense of time
5 Regularly uses watch or clock to check timing of activities

MEMORY

Q.3.26b. **Is s/he often forgetful? Can s/he recall what happened yesterday or last week - is s/he easily reminded?** |__|

1 No memory or only remembers isolated events and not easily reminded;
- little idea of what happened yesterday or last week
2 Forgets what he/she has done or where things were left but easily reminded;
- can recall some of what happened yesterday or last week
3 Not often forgetful and doesn't usually need reminding;
- has good recall for what happened yesterday or last week

USE OF AMENITIES

Q.3.27. **Can the following be used <u>independently</u>?**

Yes = 1 No = 2 With help = 3

1. Domestic telephone (dial out) |__|
2. Public telephone - local calls/mobile |__|
3. Doctor, dentist |__|
4. Postage for letters |__|
5. Welfare rights (unemployment benefit, pension) |__|
6. Savings (post office/bank-deposits/withdrawals) |__|
7. Public entertainment (pub, cafe) |__|
8. The shops (includes being dropped off in town centre) |__|
9. A local shop |__|
10. Public convenience |__|

QUALITY OF SOCIAL INTERACTION - OWN ASSESSMENT AND CARER

Q.3.28. **Choose one of the following ratings which best describes the person** |__|

(N.B. If unable to assess, code as 0)

Questions:
- Is s/he friendly with his age peers?
- Does s/he make the first approach?
- Does s/he join in actively or passively?
- If s/he is left alone does s/he seek out company, or is s/he quite content to be by her/himself, even for very long periods of time?
- Is s/he friendly to staff?
- Does s/he go to people he knows but ignore strangers?

a) **Aloof**

1 Does not interact - mainly aloof, indifferent and bizarre.
2 Interacts to obtain needs - otherwise indifferent.

b) **Passive**

3 Responds to and may initiate physical contact only.
4 Does not initiate social contact, but responds passively if other people make approaches.

c) **Active but odd**

5 'Unwarm'.
Does make social approaches, but these are peculiar, naive or even bizarre. The person does not modify his behaviour in the light of responses, needs or interests of those whom he approaches. The interaction is one-sided and dominated by the person being rated.
6 Some warm qualities in addition to 5.

d) **Sociable**

7 People of very low level of development who enjoy having others around - would be unhappy without company. Smile and show positive response to interaction - make eye contact with people who speak to them. The person himself initiates contact by means of his eye, arm, hand or body movements etc. (unlike Group 9)
8 Is shy, but interaction appropriate once shyness is overcome (as for description given under 9 below. Do not confuse shyness with aloofness).

OR

Interacts sometimes in an appropriate way, but in general is not gregarious – prefers her/his own company (also use for people who are withdrawn as a result of temporary illness but who are usually sociable).

9 Makes social approaches to other people. S/he looks up with interest and smiles when approached by others. S/he enjoys social contact with her/his companions in the ward, centre, school, as well as with staff. S/he responds to other people's ideas and interests, as well as contributing to the interaction to the best of her/his ability.

EMPATHY [SYMBOLIC ACTIVITIES]

Q.3.29. **Is s/he aware and concerned about people's feelings?** |__|

Does s/he like people, know that they have feelings and show concern for their welfare? For example, if someone in the household became ill, would they notice and be concerned for them?

1 No such empathy
2 Limited empathy only
3 Has a range of such empathies

SIMPLE STEREOTYPIES

Q.3.30. **Does she have any simple repetitive activities e.g. rocking, flicking fingers etc?** |__|

Constant

1 This behaviour is marked, especially when unoccupied, although may be controlled by close supervision or being kept fully occupied - often a constant feature, present each day.

Sporadic

2 Present, but minor aspect of behaviour pattern
3 Minimal or none

ELABORATE ROUTINES/OBSESSIONAL BEHAVIOUR

Q.3.31. **Does s/he have any repetitive activities requiring some skill?** |__|

1 Has elaborate routines of the kind and intensity found in early childhood autism.
2 Has minor routines, or obsessional behaviour such as hand washing. Also use for tendency to repetitive behaviour seen in old people with early dementia, excessive tidiness in personal possessions, refusal to be parted from shopping bag, day or night, etc.
3 Minimal or none

REPETITIVE SPEECH

Q.3.32. **Does s/he go on talking about the same things, or asking the same questions over and over, even if you give her/him an answer?**

0 Not enough conversational speech to rate
1 Marked repetitive speech

2 Some tendency but can be distracted to other topics (include here the tendency for some elderly people to return to the same memories of the past).
3 Little or none

MOOD/PERSONALITY

Q.3.33. **Can you tell me if any of the following have been present recently?**

	Major	Minor	None	N/K	
1. frustration	2	3	4	9	|__|
2. unhappiness/upset/crying	2	3	4	9	|__|
3. withdrawn	2	3	4	9	|__|
4. anxiousness/phobias/irrational fears	2	3	4	9	|__|
5. mood swings	2	3	4	9	|__|
6. imagines voices/images	2	3	4	9	|__|
7. feels things always set against them	2	3	4	9	|__|
8. lethargic	2	3	4	9	|__|

BEHAVIOUR PROBLEMS

Q.3.34. **Does s/he have any behaviour problems? How do you manage when this behaviour occurs?**
How often does it happen?

Codes for items:

1 Severe behaviour problem and frequent occurrence (more than three times a week).
2 Less severe behaviour problem but frequent occurrence (more than three times a week).
3 Severe behaviour problem but infrequent occurrence (three times a week or less).
4 Lesser management problem.
5 Does not occur.
6 Previous severe problem currently controlled in present environment.

i) Physically aggressive to others |__|

ii) Destructive - paper, furniture, clothing, windows etc. |__|

iii) Excessive activity - paces up and down, does not sit still |__|

iv) Seeks attention - pesters staff or others |__|

v) Self-injury - head banging, picking sores, biting etc. |__|

vi) Wanders or runs away if unsupervised |__|

vii) Excessive noise - screams or makes other disturbing noises - shouts, grunts, uncontrollable laughter etc. |__|

viii) Temper tantrums or verbal abuse |__|

ix) Disturbs others at night/early morning |__|

x) Scatters or throws objects around - creates chaos aimlessly|__|

xi) Anti-social, delinquent - steals, lies, bullies, incites others etc.|__|

xii) Sexual behaviour which puts them at risk of getting into trouble with the legal system or at risk of abuse. If no social awareness code under difficult/offensive personal habits (xv) |__|

xiii) Repeated untruthfulness likely to cause problems |__|

xiv) Uncooperative |__|

xv) Difficult or offensive personal habits:

a) Spits |__|

b) Smears |__|

c) Self induced vomiting |__|

d) Eats non-food items |__|

e) Continuous eating/drinking |__|

f) Inappropriate swearing |__|

g) Inappropriate sexual behaviour |__|

h) Hoards rubbish |__|

i) Difficult/offensive habit with menstruation |__|

j) Other difficult or offensive personal habits* |__|

* Specify _____code |__|__|

(or leave blank)

xvi) Other behaviour problems* |__|

* Specify _____ code |__|__|

(or leave blank)

Complete Q.35 - 37 <u>if there are any behaviour problems</u>
(i.e. if question 3.34 not all coded 5).

Q.3.35. **How is/are the behaviour problems managed?**

a) i) Major environmental adjustment Yes=1 No=2|__|
e.g. rooms cleared, doors/windows locked

ii) Lesser environmental adjustment Yes=1 No=2|__|
e.g. avoidance of provocation/isolation

b) i) Constant and intensive supervision Yes=1 No=2|__|

ii) Lesser close supervision Yes=1 No=2|__|

c) i) Drugs, additional medication used to control outbursts/impending continuous behaviour Yes=1 No=2|__|

ii) Verbal persuasion/counselling Yes=1 No=2|__|

d) Other means of management* Yes=1 No=2|__|

* Specify _____ code |__|__|
(or leave blank)

Q.3.36. **During the last 2 years has the behaviour resulted in any physical hurt or damage to buildings or contents?**

a) i) Physical hurt to others requ. medical/ nursing attention Yes=1 No=2 |__|

ii) Physical hurt to others of lesser severity Yes=1No=2 |__|

b) i) Physical hurt to self requiring med./nursing attention Yes=1 No=2 |__|

ii) Physical hurt to self requiring lesser attention Yes=1 No=2 |__|

c) i) Major damage to building fabric (windows/doors broken) Yes=1 No=2 |__|

ii) Lesser damage to building fabric (decor damage) Yes 1 No=2 |__|

d) i) Major damage to furniture/fittings/belongings (chairs broken/things torn to shreds) Yes=1 No=2 |__|

ii) Lesser damage to furniture/fittings/belongings (torn material) Yes=1 No=2 |__|

e) i) Other _____Severe Yes=1No=2 |__|

ii) Other _____Lesser Yes=1No=2 |__|

Specify _____code |__|__|
(or leave blank)

Q.3.37. **During the last 2 years has the behaviour problem resulted in X being excluded from care or activities, or caused you to feel desperate about coping or involved the police?**

a) i) Client excluded permanently from day care Yes=1No=2|__|

ii) Client excluded temporarily from day care Yes=1No=2|__|

b) i) Client refused short break (respite care) placement Yes=1 No=2|__|

ii) Client excluded from other opportunity e.g. club/holiday/transport Yes=1 No=2|__|

c) i) Client's carers sought urgent short break (respite care) Yes=1 No=2|__|

ii) Client's carers felt unable to cope Yes=1 No=2|__|

d) Police threatened and/or took formal action Yes=1No=2|__|

e) i) Other _____Severe Yes=1 No=2|__|

ii) Other _____Lesser Yes=1 No=2|__|

Specify _____code |__|__|
(or leave blank)

ASK ALL: HEALTH

Q.3.39. **Does s/he suffer from epilepsy?**
Yes=1 No=2 Not sure=3 |__|

Q.3.40. **Does s/he experience fits?**

Once or more per month = 1 Occasionally = 2 None = 3 |__|

Q.3.40a. **Has a Doctor ever told you that s/he has diabetes?**

Yes=1 No=2 Not sure=3 |__|

Q.3.41. **<u>If known</u>: does s/he take medication to prevent:** (if not known enter 9)

Epileptic fits Yes=1 No=2 |__|

Behaviour problems Yes=1 No=2 |__|

Sleep problems Yes=1 No=2 |__|

Anxiety/depression problems Yes=1 No=2 |__|

Q.3.42. **Does s/he have any problems taking medicines prescribed for any reason?**

Prescribed and takes by self	1	|__|
Prescribed and takes with supervision	2	
Prescribed - but problems taking	3	
Not prescribed	4	

Q.3.43. **What form of long-term medication does s/he take?**
(If none enter N/A = 8, for all)

1.	Medicine by mouth	Yes=1	No=2	N/A=8	|__|
2.	Medicine by injection	Yes=1	No=2	N/A=8	|__|
3.	Inhalation	Yes=1	No=2	N/A=8	|__|
4.	Suppositories/other e.g. rectal valium	Yes=1	No=2	N/A=8	|__|
5.	Cream on skin	Yes=1	No=2	N/A=8	|__|
6.	Drops	Yes=1	No=2	N/A=8	|__|
7.	Other (including enemas)	Yes=1	No=2	N/A=8	|__|

Q.3.44a. **How would you rate the client's physical health?**

Poor = 1	Fair = 2	Good =3	Very good =4	Excellent = 5	|__|

Q.3.44. **Would you describe it as….?** \|__\|
(Ignore transient + trivial illness)

Steady	1	Rapidly declining	3
Slowly declining	2	Improving	4

ASK ALL: LIFESTYLE

Q.3.45 **Smoking**

a. Has the client ever smoked? Yes=1 No=2 |__|

(If No go to Q.3.46)

b. Does s/he smoke at all now? Yes=1 No=2 |__|

c. Does s/he smoke cigarettes? Yes=1 No=2 |__|

d. How many does s/he usually smoke per day (number) |__|__|

HEIGHT/WEIGHT

Q.3.46. **How tall is the client?**

feet _____ inches _____ |__|__| (in inches)

(_____ cms)

Q.3.47. **How much does the client weigh at present?**

stones _____ lbs _____ |__|__|__| (in lbs)

(_____ kg)

Q.3.48. **Body shape**

Which of these figures would you say describes his or her body shape?
(Select from "Body Shape" figures on back page)

Choose options 1-9 |__|
If no answer, i.e. they can't or won't answer, code as 0

Q.3.49. **Physical activity**

Compared with other people of his/her age would you describe him/her as:

|__|

1 Not at all physically active
2 Less physically active
3 About the same
4 More (i.e. very or fairly) physically active

Q.3.50. **How often does s/he drink a glass of fizzy soft drinks such as coca cola or lemonade?**
Estimate on average during the last 12 months.

1 4 or more times a day |__|
2 1-3 times a day
3 2-6 times a week
4 Once a week
5 Once a month or less

Q.3.51. **How often does s/he drink a half pint of beer, lager or cider?**
Estimate on average during the last 12 months

1 4 or more times a day |__|

2 1-3 times a day
3 2-6 times a week
4 Once a week
5 Once a month or less

Q.3.52. **How many <u>times a week</u> does s/he eat the following foods (medium serving) - estimate on average during the last 12 months?**

1 Chips |__|__|
2 Potatoes – boiled, mashed, instant, baked |__|__|
3 Fish and fish products |__|__|
4 Chicken or other poultry e.g. turkey |__|__|
5 Bread, white brown and wholemeal * |__|__|
6. Fruits and fruit products, not including fruit juice |__|__|
7. Vegetables† and salads, not including potatoes |__|__|

* a medium serving is 1 slice of bread or equivalent amount of flat bread (e.g. Indian nan)

† consider a tomato to be a vegetable for this purpose

HEALTH ACTION PLANS

Q.3.53 Do you know about Health Action Plans? Yes=1 No=2 |__|

If **yes** go to Q 3.54

If **no** enter '8' in Q 3.54 and go to Q 3.55

Q.3.54 Have you got someone to help you (health facilitator)?

Yes=1 No=2 N/A = 8 N/K = 9 |__|

If yes: Do you have a Health Action Plan?

Yes=1 No=2 N/A = 8 N/K = 9 |__|

Ask all:

Q.3.55 Would you like more* information about HAPs?

Yes=1 No=2 |__|

Q.3.56 Do you need further help with the process (of developing a HAP)?

Yes=1 No=2 N/K = 9 |__|

Tick if referral made to: City HAP Facilitator |__|

County HAP Facilitator |__|

LD Team |__|

Q.3.57 Have you been to your surgery to see your doctor or a practice nurse in the last year?

Yes=1 No=2 N/K = 9 |__|

SECTION IV: USE AND NEED FOR SERVICES

FORMAL DAYTIME ACTIVITIES

Q.4.1. **What are the client's <u>formal</u> daytime activities in a <u>typical</u> week?**
[daytime + organised by someone + scheduled]

b) Enter number of sessions <u>organised</u> in appropriate box i.e. allocated

PROVIDER → **↓ ACTIVITY**	**CLIENT/ CARER** **A**	**SS LD DAY CENTRE** **B**	**SS OTHER** **C**	**NHS** **D**	**EDUCATION** **E**	**VOL** **F**	**PRIVATE** **G**	**OTHER** **H**	**CARER DOES NOT KNOW** **I**
<u>WORK</u> FORMAL/'NORMAL' **01**									
VOLUNTARY **02**									
SHELTERED **03**									
TRAINING **04**									
<u>EDUCATION</u> **05**									
<u>AT DAY CENTRE</u> **06**									
<u>ELSEWHERE</u> **07**									
<u>HOME</u> **08**									
The bottom 2 rows									

have no (known) provider. The column should therefore reflect the type of accommodation.									
CARER DOES NOT KNOW 09									
NOTHING FORMAL 10									

[Fill in 10 half-day sessions]

a) Who is the prime instigator?

1 = Social Services
2 = Health Services
3 = GP
4 = Other
5 = Out of county active daycare (go to
6 = Carer
8 = N/A
9= Not known

|__|

Q4.4)

c) **Type of day centre(s) with number of allocated half-day sessions a week.**

Type of Day Centre (DC)			**Number of Sessions**
Main DC:	i. |__|	ii. |__|	|__|__|
Other DC:	i. |__|	ii. |__|	|__|__|

1 = Learning disability	2 = Autism	3 = Physical disability	4 = Mental illness
5 = Older persons	6 = Other	8 = N/A	9 = Not known

d) **How many formal half-day sessions does he/she typically attend each week?** |__|__|

e) **If something formal is not organised on a full time basis, what is the** [main] **reason?** |__|__|

01 Excluded by centre
02 Insufficient sessions available
03 Choice of client and carer
04 Choice of carer - client disagrees
05 Choice of client - carer disagrees
06 Choice of client (living without carer)
07 Choice of carer (client unable to voice an opinion)
08 Nothing suitable
09 Other

10 Varying amounts from week to week
88 Not applicable
99 Not known

Q.4.2. **Overall, do you feel that s/he is appropriately placed for her/his level of dependency?** |__|

1 Client has potential for greater independence/could do more than is possible in present circumstances
2 Client is appropriately placed for level of dependency but dissatisfied with the content of one or more activities
3 Client is appropriately placed for level of dependency and satisfied with the activity content
4 Client is more dependent than can be properly managed in present circumstances
5 Carer says spontaneously that he/she does not know enough about what goes on to judge

Comment _____ code |__|__|

IF NO FORMAL ACTIVITIES GO TO Q.4.3a.

Q.4.2a. **What formal daytime activities, if any, are unsatisfactory/causing problems?**
[Use grid letter and number e g A09, B07, D10 etc]
Otherwise use: None selected = 0 N/K = 9 in first box

|__|__|__| |__|__|__| |__|__|__|

Q.4.2b. **What formal daytime activities, if any, are particularly beneficial?**
[Use grid letter and number e g A09, B07, D10 etc]
Otherwise use: None selected = 0 N/K = 9 in first box

|__|__|__| |__|__|__| |__|__|__|

Q.4.3a. **Do you feel that the client is allocated an appropriate number of formal daytime sessions a week?**

How many more or fewer sessions are needed?+/- |__| No.|__|__|

01 02 03 04 05 06 07 08 09 10 0 00 = No change 9 99 = Don't know

LEISURE AND RECREATION / WEEKENDS/ EVENINGS

Q.4.4. **What does s/he usually do if allowed to choose his/her own activities?**

|__|

Nothing constructive	1	(includes watching TV without real interest and stereotyped behaviour)
Partially constructive	2	(constructive + stereotyped)
	3	(constructive + nothing)

Usually constructive 4 (domestic work, looking at books, talking to others, listening to radio, watching TV with some interest, knitting +/- snoozing)

Q.4.5. **What form of private recreation or leisure activities does s/he take part in?**

Codes

No = 2 Regularly = 3 Occasionally = 4

i)	Attends social club(s)	|__|
ii)	Attends sports	|__|
iii)	Participates in sports	|__|
iv)	Exercises considerable choice across a range	|__|
v)	Other/family arrangements	|__|
vi)	Education classes	|__|

Q.4.7. **Does s/he need to take part in some form of organised recreation more often?**

Yes = 1 No = 2 |__|

Q.4.8. **Does s/he need any help to develop/continue with her/his leisure time?**

Yes = 1 No = 2 |__|

If yes, are any of the following needed/or a problem?

a)	Broader range of leisure opportunities	Yes=1No=2 N/A=8	|__|
b)	Needs accompaniment/befriender	Yes=1 No=2 N/A=8	|__|
c)	Needs transport	Yes=1 No=2 N/A=8	|__|
d)	Motivation	Yes=1 No=2 N/A=8	|__|
e)	Refuses to go for any reason	Yes=1 No=2 N/A=8	|__|
f)	Finance/resources	Yes=1 No=2 N/A=8	|__|
g)	Other*	Yes=1 No=2 N/A=8	|__|

* Comment

_____

code |__|__|

HOLIDAYS

Q.4.9. **Does s/he usually go on holiday?**

Yes = 1 No = 2 |__|

Q.4.10. **Are there any special problems with taking X away on holiday?**

Yes = 1 No = 2 |__|

Comment _____code |__|__|

Code main problem if more than one (or leave blank)

Q.4.11. **How does s/he cope with the day centre (or other) holiday period?** |__|

1 No problem
2 Some problems
3 Severe problems
8 Not appropriate

Q.4.12. **Do you need any help with organising activities during the holiday period?**

Yes=1 No=2 N/A=8 |__|

SHORT BREAKS

Complete as appropriate.

Q.4.13. **Establish that the respondent knows what is meant by the term 'Short Breaks' by asking whether s/he has ever received it and if not, by describing it in simple terms. Then indicate whether they knew that Short Breaks (Respite Care) was available.**

1 Knew about Short Breaks |__|
2 Did not know about Short Breaks
8 N/A (Categories 8+) go to Q.4.23

Q.4.14. **During the last twelve months have you received any Short Breaks (Respite Care)?**

If YES: Have you felt you needed more during this time?

Yes = 3 No = 1 Not needed = 4

If NO: Have you felt you needed some during the last twelve months?

Yes = 2 No = 0 (Enter 0, 1, 2, 3 or 4) |__|

4 Received but not required
3 Received and additional required
1 Received and no more required
2 Not received but required
0 Not received and not required

Q.4.15. **Explore with the carer the <u>approximate</u> number of days of Short Break (Respite Care) used in the last twelve months.**

<u>Code</u> <u>Number of days</u>

5	100+
4	50 - 99
3	20 – 49
2	5 – 19
1	Less than 5
0	None
9	Not known

For example

a) a person using 2 days a week each week uses 100 or more days (Code 5)

b) a person using 1 week for holiday + 1 week emergency is using between 5-19 days (Code 2)

Q.4.16. **How much extra or new short breaks do they need?** (use above codes)

|__|

Q.4.17. **Urgency**

If new or additional requested by the carer: |__|

1 Prefer this care (<1 year)
2 Non-urgent need (1-2 years)
3 Long-term need (>2 years)

If no additional requested:

4 No foreseeable need

Q.4.18. **Special Requirements:**

Yes = 1 No = 2 |__|

e.g. Asian Culture/diabetic diet

Comment

_____

code |__|__|

(or leave blank)

Q.4.19. **Why received/needed**

3 Received and additional required
1 Received and no more required
2 Not received but required
0 Not received and not required

Use the above codes:

When	Holidays (annual)	|__|
	Regular support	|__|
	Other (inc. client request)	|__|
Type	Ordinary short breaks (respite care)*	|__|

Family placement scheme |__|

In own home |__|

* Where (if received): _____code |__|__|__|__|__| (or leave blank)

Q.4.19a.**Under present circumstances would there be a need for short break (respite care) in an emergency?**

Yes = 1 No = 2 |__|

Q.4.20. **Ask those who know about short breaks (respite care):**

How do you feel about short breaks - are you pleased or dissatisfied with what is available? |__|

1 Pleased
2 Dissatisfied
3 Neither pleased nor dissatisfied

Q.4.21. **Ask everybody whether they feel they** |__|

1 Need advice
2 Uncertain but some interest
3 Do not need advice

Q.4.22. **Any comment on Short Breaks:-**

_____

_____

code |__|__| (or leave blank)

INDEPENDENT LIVING: (Degree of supervision)

Complete for ALL individuals.

Q.4.23. **Can s/he be left alone safely? How long could you go out for and leave her/him alone?**

a. **What degree of supervision is <u>given</u> during the day?** |__|

6 Constant intensive supervision
5 Constant background supervision
4 Virtually constant background supervision but can be left alone for 2-3 hours
3 Supervision needed at peak periods eg.meal/bedtimes
2 Once or twice per day
1 Less than once per day
0 Less than once per week
-1 No supervision needed

d. **What degree of supervision is <u>needed</u> during the day?**
Use codes from (a) |__|

b. **Does s/he <u>need</u> attention during the night?** |__|

e.g. toileting, epileptic attack, turning, behaviour

5 Several times per night
4 Once per night
3 Less than once per night
2 Less than once per week
1 No

Q.4.24. **Do you think s/he:** |__|

1 has potential for (and needs) greater independence than possible in present circumstances.
2 is placed appropriate to level of dependency
3 is more dependent than can properly be managed in present circumstances

Comment code |__|__| (or leave blank)

Q.4.25. **We are concerned with long-term planning for the needs of people with a learning disability, particularly in relation to future living arrangements.**

Do you foresee a time when you will need to change his/her living arrangements?
Have you given any thought as to when this might be needed?

<u>Urgency:</u>

1 <1 year
2 1-2 years
3 3-4 years
4 5-10 years
5 No need for change in foreseeable future

Q.4.25a. **Planning for the future:**

Ask everybody whether they feel they: |__|

1 Need information
2 Uncertain but some interest
3 Do not need information

Q.4.25b. **Discuss relevant sources of information:**

Information or leaflet given Yes=1 No=2 N/A=8 |__|

Interviewer note/reminder: _____

Q.4.25c. Referral made regarding future needs in living arrangements

Yes=1 No=2 N/A=8 |__|

Interviewer note/reminder: _____

Q.4.26a. **Do you receive/need any of the following on behalf of the client?**

Receipt and need of service codes		Interviewer Action Codes	
0	Not received and not needed		
1	Satisfied with amount and quality.	1	Referral requested
2	Not received but required	2	Referral derived
3	Received and additional required / services received could be better	4	Carer/client given specific advice
8	Using 'out of county services' (i.e. out of Leicester, Leicestershire and Rutland).	5	No Action

Use the above codes:	**Receipt and need of service**	**Interviewer action**
Home (help) care	|__|	|__|
Sitting service	|__|	|__|
Medical advice from GP	|__|	|__|
Health Action Plan	|__|	|__|
Help with transport	|__|	|__|
Bus pass	|__|	|__|
Social Worker	|__|	|__|
Community Nurse	|__|	|__|
Speech Therapy	|__|	|__|
Physiotherapy	|__|	|__|
Occupational Therapy	|__|	|__|
Chiropodist	|__|	|__|
Specialist Dentist	|__|	|__|
Psychologist/Psychiatrist	|__|	|__|
Support worker/Key worker	|__|	|__|

General information about services	|__|	|__|
Financial advice	|__|	|__|
Other	|__|	|__|

Comment code |__|__|

(or leave blank)

BENEFITS

Q 4.27a. CLIENT
What benefits do you receive on behalf of the client?

i)	Do you receive **any** benefits?		Yes=1	No=2	N/K=9		|__|
	Do you receive:						
ii)	Income support		Yes=1	No=2	N/K=9	N/A = 8	|__|
iii)	Job Seekers' allowance		Yes=1	No=2	N/K=9	N/A = 8	|__|
iv)	Incapacity benefit / EAS		Yes=1	No=2	N/K=9	N/A = 8	|__|
v)	DLA		Yes=1	No=2	N/K=9	N/A = 8	|__|
	If yes, do you receive:						
vi)	Care component		Yes=1	No=2	N/K=9	N/A = 8	|__|
	If yes, please rate:	Higher=1	Middle=2	Lower=3	N/K=9	N/A = 8	|__|
vii)	Mobility component		Yes=1	No=2	N/K=9	N/A = 8	|__|
	If yes, please rate:	Higher=1	Lower=2		N/K=9	N/A = 8	|__|
	Do you receive:						
viii)	Motability allowance		Yes=1	No=2	N/K=9	N/A = 8	|__|
ix)	Attendance allowance		Yes=1	No=2	N/K=9	N/A = 8	|__|
x)	Housing benefit/allowance of any kind		Yes=1	No=2	N/K=9	N/A = 8	|__|
xi)	Any other source of income		Yes=1	No=2	N/K=9	N/A = 8	|__|

If yes, code (if more than one, code main source) _____ code |__|__|

Q 4.27b. CARER
What benefits do you receive as a carer? (because of the care they are giving)

i)	Do you receive **any** benefits?	Yes=1	No=2	N/K=9	|__|

Do you receive:

ii)	Carers' allowance	Yes=1	No=2	N/K=9	N/A = 8	|__|
iii)	Adult placement payment	Yes=1	No=2	N/K=9	N/A = 8	|__|
iv)	Housing benefit/allowance of any kind	Yes=1	No=2	N/K=9	N/A = 8	|__|
v)	Other benefits	Yes=1	No=2	N/K=9	N/A = 8	|__|

If yes, code (if more than one, code main source) _____code |__|__|

SECTION V: CARER

Notes for interviewers:

The following section (up to Q.5.18) is not applicable for persons living without a carer. i.e. ask Q. 5.18 onwards of all.

The section may be applicable for some main supporters.

This section always refers to the **main carer** even if the client or some other person is the interviewee.

Statement to read to interviewees:

It is important that carers and professionals understand one another's problems, if community care is going to work well. We are particularly concerned that carers don't shoulder more stresses and strains than they can manage without damaging their own health and that includes financial stresses. The next set of questions provide some indicators to help us monitor this situation

Q.5.2. **Number of <u>other</u> dependants** |__|__|

Q.5.1. **What kinds of things do you usually do for X - <u>over and above what you would normally do for someone of their age</u>:**

0 = No one 1 = Client 2 = Other dependents 3 = Both

a) Help with personal care. |__|
(e.g. dressing, bathing, washing, shaving, cutting nails, feeding, using toilet)

b) Physical help. |__|
(e.g. with walking, getting up and down stairs, getting into and out of bed)

c) Helping with paperwork/financial matters |__|
(e.g. writing letters, sending cards, filling in forms, dealing with bills, banking)

d) Other practical help. |__|
(e.g. preparing meals, doing his/her shopping, laundry, housework, gardening, decorating, household repairs, taking to doctors' or hospital)

e) Keeping her/him company. |__|
(e.g. visiting, sitting with, reading to, talking to, playing cards or games)

f) Taking out. |__|
(e.g. taking out for a walk or a drive, taking to see friends or relatives)

g) Giving medicines. |__|
(e.g. making sure s/he takes pills, giving injections, changing dressings)

h) Keeping an eye on her/him to see s/he is all right. |__|

i) Other help (SPECIFY). |__|

_____

code |__|__|

(or leave blank)

CARER

Q.5.3. **Do you have any health problems or disabilities?**

Yes = 1 No = 2

01 Problems or disabilities connected with: |__|
arms, legs, hands, feet, back or neck (include arthritis or rheumatism)

02 Difficulty in seeing |__|

03 Difficulty in hearing |__|

04 Skin conditions, allergies |__|

05 Chest or breathing problems, asthma, bronchitis |__|

06 Heart, blood pressure or blood circulation problems |__|

07 Stomach, liver, kidney or digestive problems |__|

08 Diabetes |__|

09 Depression, bad nerves |__|

10 Epilepsy |__|

11 Gynaecological/menopausal problems |__|

12 Other health problems or disabilities |__|

Give details _____

Q.5.4. **Priority code up to 3 health problems or disabilities, starting with the problem or disability that affects the person most:**

1. |__|__| 2. |__|__| 3. |__|__|

(Code as 88 if fewer than 3 problems)

Q.5.5. **Does this illness or disability limit your activities in any way?**

Yes=1 No=2 N/A=8 |__|

Q.5.6. **In general, how would you rate your health?**

Poor = 1 Fair =2 Good =3 Very good =4 Excellent = 5 |__|

SMOKING

We know from work done in other centres that carers can tend to cope with mounting pressures by some behaviours that damage their own health in the long run, for example smoking heavily. As part of our concern for your health it would be helpful to know:

Q.5.9. **Have you ever smoked?**

Yes = 1 No = 2 |__|

(If No go to Q.5.7)

Q.5.10. **Do you smoke at all now?**

Yes = 1 No = 2 |__|

If cigarettes:

Q.5.11. **How many do you usually smoke per day?** (number) |__|__|

HEIGHT/WEIGHT

Q.5.7. **How tall are you?**

feet _____ inches _____ |__|__| (in inches)

(_____ cms)

Q.5.8. **How much do you weigh at present?** (When not pregnant)

stones _____ lbs _____ |__|__|__| (in lbs)

(_____ kg)

Q. 5.8a **Which of these figures would you say describes your basic body shape?**
(Select from body shape figures on back page)

Choose options 1-9 |__|
If no answer, i.e. they can't or won't answer, code as 0

Q.5.12. **Do you feel able to provide basic care?** Yes = 1 No = 2 |__|

Q.5.13. **Do you feel particularly stressed with this situation?**
Is it affecting your health/well being?

	Carer				Interviewer			
	None	A little	A lot		None	A little	A lot	
Psychologically	1	2	3	|__|	1	2	3	|__|
Physically	1	2	3	|__|	1	2	3	|__|
Socially	1	2	3	|__|	1	2	3	|__|

Q.5.14. **How do you feel about providing this care?**

Content = 1 Resigned = 2 Dissatisfied = 3 |__|

Q.5.15. **Do you, <u>the carer</u>, get any support e.g. from your family or any groups?**

Yes = 1 No = 2 |__|

Who is the main support? |__|

None	0
Family	1
Friends	2
Self-help group	3
Other	4

Q.5.16. **Do you, <u>the carer</u>, feel you need additional long-term moral support?** (Prompt with Social Worker, nurse, other parent, support group).

Yes = 1 No = 2 |__|

INCOME

We are very concerned that you've got all you need to help support X. That includes having enough money, because it could be a source of worry.

Q.5.18. **Do you find it difficult to manage on your present income?**

Adequate=1 Difficult=2 No answer=3 |__|

Q.5.21. **Are there any problems we have not mentioned (or not recorded) elsewhere:**

Yes = 1 No = 2 |__|

If yes:

_____

_____

_____

_____

Q.5.22. **We would like to capture your views on anything concerning the support and care you need or receive. Would you like to comment on any aspects of this?**

Yes = 1 No = 2 |__|

If yes:

_____

_____

_____

_____

_____

Q.5.24. **General notes/comments (Carer or Interviewer):**

_____

_____

_____

Q.5.25 Persons living alone/with supporter:

Would you mind if I have a word with the specialist nurse/doctor or other background figure in case there are any problems we have over-looked?

OK = 1 Not OK = 2 NA = 8
|__|

SECTION VIa. OFFICE USE

Q.6. 1 **CONSENT/ASSENT GIVEN BY:-** (including verbal consent/assent) (See background note)

Carer = 1 Client = 2 Both = 3 Supporter = 4 |__|

Q.6. 2 **CONSENT/ASSENT FOR SHARING OF RECORDS (Having seen the leaflet)**

Consent/Assent given, no reservations	1	|__|
Consent/Assent not given for sharing	2	
Consent/Assent given, with reservations*	3	
Not Applicable	8	

*reservations
.....

Q.6.2a **Agreement to be contacted for research**

Occasionally doctors or other professionals do research or special studies and may like to contact you if they think you could help them. Would you be happy for them to get in touch?

Yes = 1 No = 2 |__|

Q.4.26b **Agreement to be sent information**

Sometimes other Learning Disability organisations ask for our help with sending out information. Are you happy for us to pass your name on if we feel the information may be of interest to you? It doesn't happen all that often.

Yes = 1 No = 2 |__|

Q.6.3a. **Did you communicate with the client?** Yes = 1 No = 2 |__|
(See background note)

Q.6.3. **Did the client take part in the interview?** Yes = 1 No = 2 |__|
(See background note)

Q.6.4. **Did anyone else take part in the interview?** Yes = 1 No = 2 |__|

Q.6.5a. **Apparent eligibility as assessed at time of interview:**
|__|

1 Definite
2 Probable
3 Possible
4 For review
5 Doubtful
6 Not LD

Q.6.7a. **Completeness of interview:**

1 = Complete 2 = Incomplete 3 = Abandoned |__|

Q.6.7b. **Accuracy of interview:**

1 = Satisfied 2 = Misgivings |__|

Q.6.8. **Welfare Rights**:

1 = Referral 2 = No referral 3 = Advice given |__|

Q.6.9. **Members of Community Services:**

1 = Drawn to attention 2 = Not drawn to attention |__|

Q.6.10. **Any referral made? (requested <u>or</u> derived)**

Yes = 1 No = 2 |__|

Q.5.23. **Any cause for immediate serious concern?** (including problems with supporters)

Yes = 1 No = 2 |__|

SECTION VIb. AMENDMENTS TO OTHER SCREENS

CORE:

From front page:

Care needed when visiting Yes = 1 Not known = 9 |__|

Noted on core screen (tick here) |__|

From Q.1.10a

NHS number: (NHS number given)
|__|__|__|__|__|__|__|__|__|__|

From Q.6.5

Eligibility: |__|

Definite	1
Probable	2
Possible	3
For review	4
Doubtful	5
Not LD	6

Noted on core screen (tick here) |__|

CARER:

Carer Deaths:

Details _____

_____

_____

Noted on core screen (tick here) |__|

Any Other Notes, including other people present (not for data entry):

Yes = 1 No = 2 |__|

_____

_____

_____

_____

Data entered on: |__|__| |__|__| |__|__|__|__|
By: (initials) |__|__|

Core data checked: |__|
Carer data checked: |__|

NOTES

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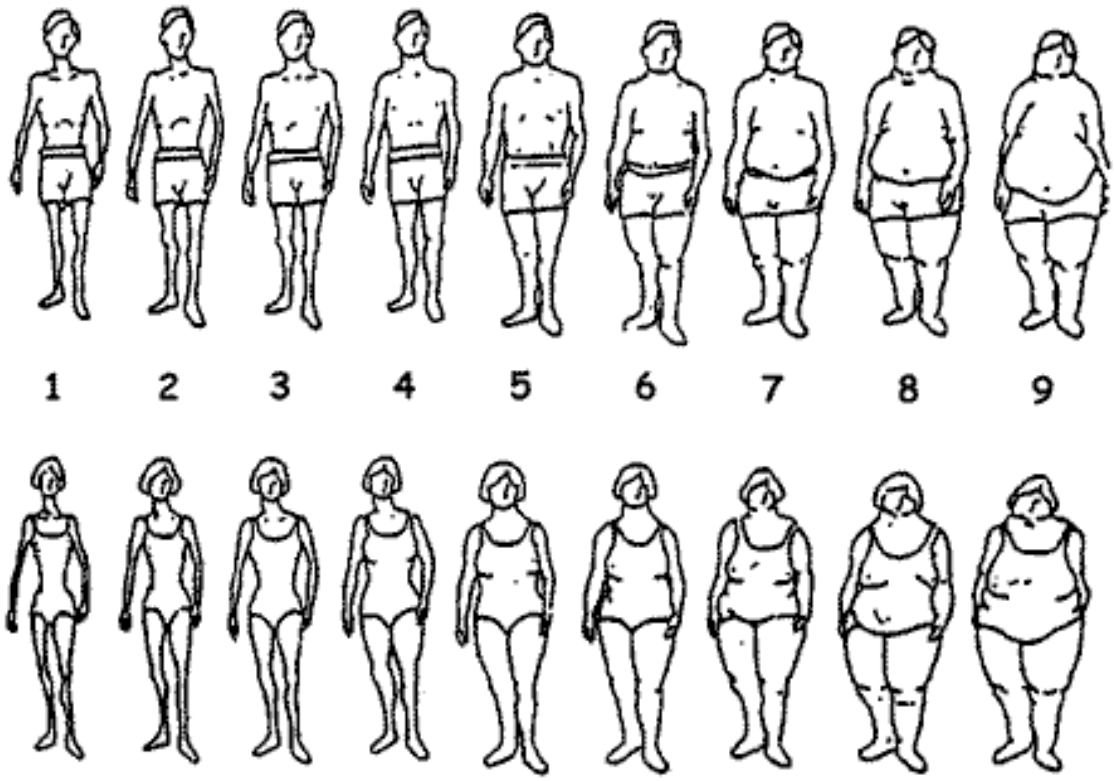
BODY SHAPE



APPENDIX 4: CHARACTERISTICS OF SERVICE USERS WHO WERE EXCLUDED

Characteristics of deaf people who were excluded

Nine adults had **no ID**; these were all assessed and did not have ASD. Five had congenital deafness of unknown aetiology and were all fluent signers:

- A 22 year old white man who had been referred for a suspected diagnosis of ADHD.
- A 48 year old South Asian woman who had been referred for assessment of mental health problems (depression).
- A 23 year old white man who had been referred for forensic reasons (sexually inappropriate behaviour).
- A 52 year old black Caribbean man who had been referred for assessment of his social needs.
- An 18 year old white woman who had been referred for management of challenging behaviour.

The remaining 4 service users with **no ID** had a known aetiology for their deafness; they comprised:

- A 50 year old white man with athetoid cerebral palsy who was a fluent signer and had been referred with aggressive challenging behaviour.
- A 44 year old white woman with Waardenburg syndrome who was a fluent signer and had been referred with aggressive challenging behaviour.
- A 20 year old white woman with deafness due to childhood meningitis, able to talk and lip read, who had been referred for vulnerability reasons and being bullied at college.
- A 52 year old South Asian woman who was employed and able to talk and lip read. She was deaf due to complications of meningo-encephalitis; she was referred for support regarding social isolation.

There were 11 service users with **acquired deafness** (occurring later in life). These comprised:

- A 65 year old white man with a diagnosis of Fragile X Syndrome and mild ID, diabetes, constipation and anxiety disorders who was non-compliant with hearing aids and needed blood tests and ECG investigations.
- A 48 year old white man with mild ID due to neurofibromatosis, migraine, depression, suicidal tendency and history of family abuse who needed referral to community nursing, physiotherapy and occupational therapy teams. He was compliant with hearing aids.
- A 50 year old white man with moderate ID, Down syndrome, depression and constipation who needed referrals to community nursing, speech and language therapy and outreach teams for management of his aggressive behaviour. He was compliant with hearing aids.
- A 56 year old white man with mild ID due to Down syndrome. He was verbally and physically able and compliant with hearing aids.
- A 58 year old white wheelchair user man with Down syndrome with moderate ID, cerebrovascular accident, vascular and Alzheimer's dementias, ischaemic heart disease, hypothyroidism and epilepsy. He had only developed basic speech and had swallowing difficulties.
- A 24 year old white man with Down syndrome and severe ID, mobility problems, chronic acne, constipation and ASD. He was non-compliant with hearing aids and needed referrals to acute liaison nursing team, optician and social services. The researcher also needed to request ECG, blood tests and a brain scan for the management of his complex health needs.
- A 29 year old white overweight woman with mild ID and diabetes who was referred to hearing services, physiotherapy/activity coordinator teams. She was compliant with hearing aids.
- A 51 year old white woman with mild ID, psychotic depression and asthma who needed referrals to social services and hearing services. She was non- compliant with hearing aids.

- A 74 year old white woman with mild ID of unknown aetiology, ischaemic heart disease, hypothyroidism, hyperlipidaemia and vitamin D deficiency. She was non-compliant with hearing aids.
- A 68 year old white man with mild ID of unknown aetiology, epilepsy, constipation and challenging behaviour who was non-compliant with hearing aids.
- A 86 year old white man with mild ID, anaemia, hypothyroidism and hypertension who was physically and verbally quite able.

Characteristics of blind people excluded from the final analysis

There were 9 service users with **no ID**:

- A 19 year old white man with Leber's congenital amaurosis, Down syndrome, hypothyroidism and constipation.
- A 70 year old white woman with unknown aetiology of blindness, hypertension, diabetes mellitus, anaemia and hyperlipidaemia.
- A 57 year old white woman with retinitis pigmentosa, genitourinary tract cancer; she needed referral to health facilitators.
- A 20 year old white woman with bilateral iris coloboma, microcephaly of unknown origin and Fallot's tetralogy, epilepsy and challenging behaviour; she needed referral to psychology.
- A 66 year old white man with congenital glaucoma, diabetes and hyperlipidaemia.
- A 19 year old South Asian man with retinopathy of prematurity.
- A 19 year old white man with retinitis pigmentosa and ADHD.
- A 20 year old South Asian man with anophthalmia.
- A 21 year old white man with congenital cataract, optic atrophy and albinism.

There were 24 service users with **acquired blindness** e.g. glaucoma, progressive retinitis pigmentosa (not causing blindness during childhood), retinal detachment, severe refractive error or complication of diabetes, head trauma:

- A 48 year old South Asian woman with mild ID of unknown aetiology who had depression, hypertension and insomnia. She needed blood tests and ECG.
- A 48 year old South Asian woman with moderate ID who needed referral to occupational therapy and the clinical genetics department. ECG and blood tests were also carried out for further monitoring.
- A 64 year old white woman with mild ID, schizophrenia and depression, who needed blood tests and ECG for further assessment.
- A 25 year old white woman with mild ID, personality disorder, psychosis, anaemia, constipation and incontinence.
- A 55 year old black woman with psychotic depression, insomnia and challenging behaviour. She had moderate ID of unknown aetiology.
- A 64 year old white woman with sever ID and ASD, who needed referral to the clinical genetics department after the researcher suspected a genetic syndrome. She was later diagnosed with PKU and therefore was referred to dietician to implement a PKU regime following multidisciplinary team meeting with the GP and family. She needed regular blood test monitoring.
- A 25 year old South Asian woman with cerebral palsy, treatment resistant epilepsy and ASD/challenging behaviour who needed blood tests, ECG and referral to social services, orthotic department and outreach ID nurses.
- A 30 year old white man with Down syndrome, mild ID and hypothyroidism.
- A 27 year old white woman with profound ID due to prematurity who needed blood tests and several referrals for the management of treatment resistant epilepsy, constipation and regular monitoring of PEG feeding and tracheostomy. She was also referred to optician.
- A 51 year old white man with mild ID and challenging behaviour who needed blood tests and ECG monitoring.
- A 47 year old white man with moderate ID and Down syndrome who was on antipsychotics due to Charles Bonnet syndrome (visual hallucinations in the context of acquired blindness). The aetiology of blindness was

complication of diabetes. He also suffered from ischaemic heart disease and hyperlipidaemia. He needed blood tests and ECG monitoring.

- A 42 year old South Asian woman who had blindness and moderate ID following a head trauma during childhood. She needed physiotherapy input.
- A 63 year old South Asian man who became blind following complication of cataract surgery. He had mild ID of unknown aetiology, anaemia and peptic ulcer disease. He needed referral to social services.
- A 22 year old white woman with profound ID of unknown aetiology, who was suffering from treatment resistant epilepsy, constipation and insomnia. He needed referral to optician.
- A 29 year old South Asian man with severe ID due to cerebral palsy, who also had epilepsy and hyperlipidaemia. He needed blood tests and referral to the ophthalmology services and optician.
- A 73 year old white woman with moderate ID due to Down syndrome, hypothyroidism and complication of cataract surgery who needed blood tests, ECG and referral to advocacy services (CLASP: carers' advocacy service and Mencap) and speech and language therapy.
- A 48 year old white man with moderate ID due to cerebral palsy who also had complication of diabetes, ASD, challenging behaviour, depression and epilepsy.
- A 54 year old South Asian man with polio, moderate ID, retinitis pigmentosa, hypothyroidism and psychosis who needed referrals for assessment of falls to occupational therapy, physiotherapy and also to clinical genetics department.
- A 35 year old white woman with unknown aetiology of mild ID and depression.
- A 28 year old white man with unknown aetiology of moderate ID and challenging behaviour.
- A 60 year old South Asian woman with mild ID of unknown aetiology, who needed support from acute liaison nurses for physical health problems.
- A 26 year old white woman who had depression and epilepsy.

- A 30 year old white man who had psychotic depression and ASD (blood investigations and ECG were requested).
- A 55 year old white woman who had depression.

Characteristics of people with unilateral sensory impairments whose data were not included in the final analysis

There were 6 people with **unilateral blindness** (with normal eyesight on the other side) who had been referred to the Leicestershire Adult ID Service.

Although these service users were fully assessed by the researcher and an optician to ensure that a bilateral blindness has not been missed, their data were not analysed as during the assessment process it became clear that none of them were blind bilaterally from childhood.

These were not registered blind with VISTA, neither did they have a CVI from the ophthalmology department at the Leicester Royal Infirmary as it had been determined that they had reasonable eye sight on the other side. For some of them who were compliant in using spectacles the refraction error had been corrected:

- A 34 year old white man with ASD and challenging behaviour, who needed referral to social services; his blindness was due to an accident (shot in the eye during play with air gun during childhood). He had mild ID and was verbally and physically able.
- A 51 year old white woman with severe ID, ASD, challenging behaviour and mood disorder, who needed referral to dentist/acute liaison nurses for procedure under general anaesthesia under the best interest; her blindness was due to self-injury (eye poking during childhood). She was non-verbal but was physically able.
- A 70 year old white woman with mild ID who was verbally and physically able with a history of fall in infancy resulting in blindness in one eye, diabetes, high blood pressure and hyperlipidaemia.

The following 3 service users had unilateral blindness as a result of post-operative complications in removing a cataract:

- A 47 year old Asian man with severe ID of unknown origin, ASD and challenging behaviour, who needed referral to dentist, optician and podiatry for visits at home.
- A 40 year old white man with severe ID of unknown origin, ASD and challenging behaviour, treatment resistant epilepsy and anaemia. Patient needed referral to occupational therapy.
- A 48 year old white man with severe ID due to birth complications, who had constipation and needed treatment. He also needed referrals to optician and social services for environmental adaptation.

There was only one person registered with **unilateral deafness** who was further assessed to ensure no bilateral deafness was missed (n=1, mild ID, white, verbally and physically able with no ASD). He had been receiving support for schizophrenia (paranoid type) and alcohol and cannabis abuse. He was on a depot antipsychotic medication administered once fortnightly by his community nurse.

Characteristics of service users with deaf-blindness who were not included in the final analysis

Service users who were found to have **deaf-blindness** were excluded from the cases as they were a heterogeneous group. These included those with:

A congenital profound deaf-blindness (n=6):

- A 28 year old white woman with Pseudo-Zellweger syndrome and epilepsy who was on PEG feeding. She had profound ID and was non-verbal and wheelchair bound. She needed referrals to community nurses, ENT department, epilepsy clinic and social services.
- A 22 year old white woman with profound ID due to complication of delivery, who was tetraplegic, non-verbal, and had treatment resistant epilepsy. She needed blood investigations at home as part of his annual monitoring.

- A 20 year old South Asian wheelchair user man with profound ID due to meningo-encephalitis. He was also suffering from treatment resistant epilepsy, asthma and constipation.
- A 30 year old white wheelchair user woman with profound ID, gastro-oesophageal reflux, constipation, dysplastic hip, dysmenorrhea and epilepsy due to extreme prematurity.
- A 48 year old white man with congenital rubella syndrome and severe ID, balance problems due to cerebellar atrophy, hypothyroidism, hyperlipidaemia, depression, epilepsy, ASD and challenging behaviour of self-injury.
- A 55 years old black man with congenital rubella syndrome and severe ID (non-verbal but physically able), hepatitis B positive carrier, ASD and challenging behaviour who needed referral to the hepatologist.

There were 10 service users with **partial deaf-blindness**:

- A 60 year old white man with severe ID due to complication of delivery, blindness following eye poking, ischaemic heart disease, scoliosis, swallowing difficulties, basic repetitive speech, challenging behaviour who had all his teeth removed under general anaesthetic. He needed ICU admission for suspected myocardial infarction.
- A 43 year old white woman with profound ID due to unknown aetiology who had PEG in-situ, challenging behaviour, hay fever and needed referral to dietician. She also needed blood and ECG investigations
- 21 year old white man with profound ID, due to meningo-encephalitis who was non-verbal and wheelchair bound. He had gastro-oesophageal reflux, treatment resistant epilepsy, PEG in-situ, obstructive sleep apnoea and asthma.
- A 22 year old white man with severe to profound ID, epilepsy, kypho-scoliosis, low platelet count, hay fever and sleep apnoea, due to Trigonocephaly C syndrome. He was also on PEG feeding.
- A 21 year old black man with mild to moderate ID due to congenital rubella syndrome, with cardiac malformation, who could sign but needed referral to social services for more support.

- A 20 year old white man with mild to moderate ID, growth hormone deficiency, eczema, keratoconus and optic atrophy due to Apert syndrome. He had ASD and vitamin D deficiency. He was also using hearing aids. He used type talk assistive technology to communicate and needed referral to occupational therapy team for sensory integration assessment.
- A 23 year old South Asian man with severe ID, due to birth complications. He also had epilepsy and autistic traits, social phobia. He needed referral to psychology, occupational therapy team and community nurses.
- A 36 year old white man with mild ID (verbally and physically able) due to Pseudo-hypoparathyroidism. He had depression, diabetes, vitamin D deficiency, anaemia and renal failure. He needed referral to physiotherapy team.
- A 73 year old white man with ocular albinism and mild ID (verbally and physically able). He had ischaemic heart disease, history of cerebrovascular accidents, asthma, peptic ulcer disease, diabetes and hyperlipidaemia.
- A 48 year old white woman with meningocele, spina bifida and moderate ID who could use Makaton. He used shoe calipers and a frame to walk.

There were 6 service users with **acquired deaf-blindness**:

- A 73 year old white man with moderate ID of unknown aetiology, bilateral deafness and unilateral blindness (the cause of blindness was aggression from others). Patient's deafness was suspected by the researcher at the time of the study. He had ischaemic heart disease, hypertension, epilepsy, depression, challenging behaviour and anaemia. Although he was non-verbal, he was physically quite able but needed referral to hearing services for assessment and prescription of hearing aids and to social service for environmental adaptation.
- A 58 year old white woman with moderate ID and Down syndrome, depression, epilepsy, hypotension, recurrent falls, ASD, dementia of Alzheimer's type, swallowing difficulties who needed referral to social

services, dietician, community nurses and speech and language therapist. She also needed referral to social services for carer's need assessment and allocation of day services.

- A 38 year old white woman with profound ID of unknown aetiology, who was wheelchair bound and non-verbal. She had lost one of her eyes because of infection following corneal transplant. She also was on PEG feeding and had treatment resistant epilepsy.
- A 72 year old white man with mild ID of unknown aetiology who had bilateral cataract, hearing impairment, high blood pressure, constipation, swallowing problems, ischaemic heart disease and depression.
- A 56 year old white man with moderate ID, Down syndrome, Alzheimer's dementia, hyperlipidaemia and insomnia.
- A 65 year old white woman with Down syndrome and moderate ID who became blind due to complication of corneal transplant. She suffered from epilepsy and Non Epileptic Attack Disorders, hypothyroidism, swallowing difficulties, diabetes, cerebrovascular accident, combined vascular and Alzheimer's dementia.

APPENDIX 5: LETTERS OF INVITATION

Leicestershire Partnership NHS
NHS Trust

A University Teaching Trust

Learning Disabilities Division
Mansion House
Leicester Frith Hospital
Groby Road
Leicester
LE3 9QF

Tel: 0116 225 5200
Fax: 0116 225 5202
www.leicspart.nhs.uk

Title of the study: Sensory impairment, mental ill health and autistic spectrum disorder in people with intellectual disability

Dear **Date:**

I am a doctor working in the Adult Learning Disability Service at Leicester Frith hospital. I am carrying out a study looking at visual and hearing impairment and their effect on the mental health of adults with intellectual disability.

Currently, not much is known about this topic, therefore this study will help us to learn more about it. It will also help people who are planning services to find out what kind of support people with intellectual disability are likely to need in the future. I enclose a copy of the study Information letter and would ask you to kindly take the time to read it.

The Leicestershire Learning Disability Register has provided me with details of people with intellectual disability. The person you care for is among this group. I intend to compare two groups of people with intellectual disability, one with sensory impairment and the other without such impairment with regard to their

mental wellbeing. The person you care for may or may not have a visual or hearing impairment. But I intend to screen for this by conducting an interview and a brief physical examination if possible. I will also ask questions about their mental well-being.

The session, which will be held at a place and time convenient for you, takes about one hour or two. In case of the service user becoming distressed at any time during the interview or examination, the process will be halted immediately.

If at the time of the interview, I suspect that a visual or hearing impairment has been missed out or not addressed properly; I would take the necessary steps and will refer the person you look after to a specialist (e.g. audiologist /ophthalmologist).

If you decide to come to a day centre or Leicester Frith hospital to take part in the research project, the travel expenses will be reimbursed at the start of the session.

I would therefore like to know whether you, and if appropriate the person you care for, would be willing to be contacted for this purpose. I enclose a copy of the accessible information leaflet and would be grateful if you could discuss it with the person you care for if possible. Please then complete and return the enclosed carer's consent form in the pre-paid envelope.

If you are happy to take part in the study, I will then get in touch with you directly. You do not have to agree to take part in the study and it will not affect the care provided to the person you look after in any way if you choose not to participate in the study. If you have any questions, please contact me at the above address.

Thank you very much for your help.

Dr R Kiani

(RNIB invitation letter)

Dear

RNIB College Loughborough has a working links with the Learning Disability Service at Leicestershire NHS Trust and we have been asked by Dr Reza Kiani, a consultant in the service, to take part in a research study regarding visual and hearing impairment in people with learning disabilities. As you will see from the enclosed information sheet, the long-term aim of the project is to improve services for people with sensory impairments and additional difficulties.

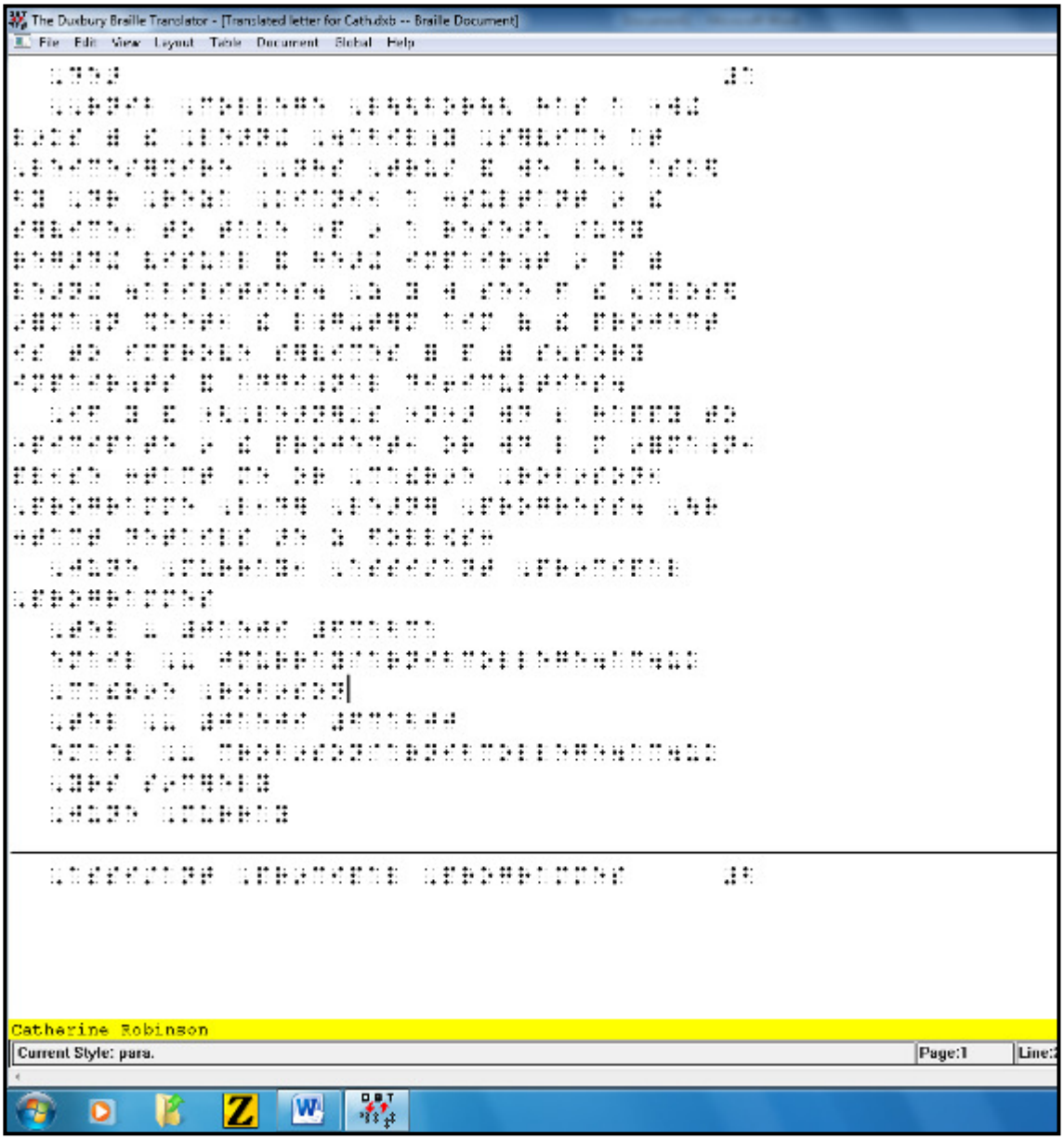
If you and (Learner's name) would be happy to participate in the project, or would like more information, please contact me or Catherine Robinson, Programme Leader Learner Progress. Our contact details are as follows:
June Murray, Assistant Principal Programmes
Tel - 01509 631231
email – jmurray@rnibcollege.ac.uk

Catherine Robinson
Tel – 01509 631200
email – crobinson@rnibcollege.ac.uk

Yours sincerely

June Murray
Assistant Principal Programmes

RNIB invitation letter in Braille



APPENDIX 6.1: INFORMATION LEAFLET FOR CARERS

Leicestershire Partnership NHS
NHS Trust

A University Teaching Trust

Learning Disabilities Division
Mansion House
Leicester Frith Hospital
Groby Road
Leicester
LE3 9QF

Tel: 0116 225 5200
Fax: 0116 225 5202
www.leicspart.nhs.uk

Title of the study: Sensory impairment, mental ill health and Autistic spectrum disorder in people with intellectual disability

Dear carer, **Date:**

You are being invited to take part in a research study regarding visual and hearing impairment in people with intellectual disability. Before you decide it is important for you to understand why the research is being done and what it will involve. Please take time to read the following information carefully and discuss it with others if you wish. Please contact me if there is anything that is not clear or if you would like more information.

1. What is the purpose of the study?

Visual and hearing impairment are very common in people with intellectual disability. The presentation of visual and hearing impairment in this population is not usually straightforward and often presents as a change in behaviour. It can occasionally be mistaken for mental illness. The study will look at visual and hearing impairment and their association with autism and mental illness in this population. This will help us to increase our knowledge about the issue and provide better services for people with these difficulties.

2. Why have you been chosen?

Information on adults with an intellectual disability has been provided by the Leicestershire Learning Disabilities Register. You have given consent to the Register to be contacted for research purposes. The service users may or may not have a visual or hearing impairment, however, it would be beneficial for them to be screened for these difficulties.

3. Do I have to take part?

It is up to you to decide whether or not to take part in the study. If you do decide to take part, you will be asked to sign a consent form and will be given a copy of both information sheet and the consent form to keep for your own record. If you decide to take part, you will still be free to withdraw from the project at any time and without giving a reason. A decision to withdraw at any time, or a decision not to take part, will not affect the standard of care that a service user is receiving.

4. What will happen to me if I take part?

I will interview you and the service user and do a brief physical examination on his/her ear and eyes at a time and place that is convenient for you. This should not last more than one and a half-hour. In case of the service user becoming distressed at any time during the interview or examination, the process will be halted immediately.

If you decide to come to a day centre or Leicester Frith hospital to take part in the research project, the travel expenses will be reimbursed at the start of the session.

5. What are the possible benefits of taking part in the study?

The study will help us in the future to plan and provide services that are better designed to meet the needs of people with intellectual disability and sensory impairment. In addition, if it is found that service users need any services that they are not already receiving, they will, with your permission, be referred to the appropriate clinical teams.

6. What if new information becomes available?

If new information becomes available during the course of the study I will make sure that you get to know about it either by post or by face-to-face meeting.

7. What if something goes wrong?

Medical research is covered for mishaps in the same way as for patients undergoing treatment in the NHS. Compensation is only available if negligence occurs.

8. Will my taking part in this study be kept confidential?

All information that you provide during the course of the study will be kept strictly confidential. Any information about the person you care for which leaves the hospital will have names and addresses removed so that he/she and you cannot be recognised from it. The person's Consultant (If they have one) and GP will be made aware of study participation in the study.

9. What will happen to the results of the research study?

The results will be anonymously published in medical journals and presented at conferences so that medical professionals working in this field will get to know about it and develop better services for adult with a learning disability. You will also receive a copy of the findings.

10. Contact for further information

Dr R Kiani (Above address)

APPENDIX 6.2: ACCESSIBLE INFORMATION LEAFLET

Leicestershire Partnership NHS Trust

Research Participant Consent Form

You are on the Learning Disability Register

You and your carer were asked a list of questions

Some times people may have difficulties with their eyes and ears. They may not be able to see or hear other people or things clearly.

We are doing a piece of work looking at how these difficulties might affect you and your health.

Would you help us?

? X

You can say no

Research Participant Consent Form

Who can I talk to about this?

Dr R kiani

Learning Disability Service

Leicester Frith Hospital

Groby Road

Leicester LE3 9QF

Telephone 0116 225 5275

I have talked about this study, in words I think are best for the person

Symbols used in this leaflet are Change Picture Bank and Somerset symbols.

Signed.....

Print .....

Date:

My name is Dr Kiani

Leicestershire Partnership NHS

Dr Reza Kiani

Consultant in Learning Disability

I want to visit you, ask you some questions to make sure your eyes and ears are working alright

about 90 minutes

The interview will be with you and your carer. It would take around 90 minutes.

I will keep your information safe and not tell anyone it is about you

I will tell you what we find out

If you say it is ok I will let your Doctor know you are helping me

Will you help?

No

If you do not want to do this it is ok

Yes

But if you do, please sign your name below and tick the box and return this form in the reply-paid envelope. Thank you.

You can tell my Doctor ☐

I am happy to be contacted for an interview ☐

Signed.....

Print .....

Date

APPENDIX 7.1: INFORMATION LEAFLET FOR GPS

Leicestershire Partnership NHS
NHS Trust

A University Teaching Trust

Learning Disabilities Division
Mansion House
Leicester Frith Hospital
Groby Road
Leicester
LE3 9QF

Tel: 0116 225 5200
Fax: 0116 225 5202
www.leicspart.nhs.uk

Title of the study: Sensory impairment, mental ill health and autism spectrum disorders in people with intellectual disability

Principal investigator	Dr R Kiani
Supervisors	Professor T Brugha and Dr S Bhaumik

Dear Doctor, **Date:**

I am one of the doctors working in the adult learning disability services at Leicester Frith hospital. I am writing to inform you that I am carrying out a study looking at visual and hearing impairment and their impact on the mental health of adults with intellectual disability.

This project has been granted ethical approval from the Nottinghamshire Research Ethics Committee. Some of these service users are registered at your surgery.

Currently, not much is known about this topic and therefore this study will help us to find out what kind of services people with intellectual disability and sensory impairment are going to need in future.

Anonymised information will be obtained on all individuals with visual or hearing impairment who have consented for their data to be accessed for research purposes from the Leicestershire Learning Disabilities Register.

Carers, and where appropriate, the patients themselves will then be contacted by letter and asked to give informed written consent before participating in the project which involves an interview and a brief physical examination.

This is a screening and non-invasive research project and will be beneficial for the service users in long term. If you do not wish your patients to participate in this study or if you wish to withdraw them from the study you may do so without justifying your decision. Please do not hesitate to contact me on the above address, should you require more information about the project.

Yours sincerely,

Dr R Kiani

APPENDIX 7.2: INFORMATION LEAFLET AND CONSENT FORM FOR RESPONSIBLE MEDICAL OFFICER

Leicestershire Partnership NHS
NHS Trust

A University Teaching Trust

Learning Disabilities Division
Mansion House
Leicester Frith Hospital
Groby Road
Leicester
LE3 9QF

Tel: 0116 225 5200
Fax: 0116 225 5202
www.leicspart.nhs.uk

Title of the study: Sensory impairment, mental ill health and autistic spectrum disorder in people with intellectual disability

Principal investigator	Dr R Kiani
Supervisors	Professor T Brugha, Dr S Bhaumik

1. What is the purpose of the study?

The study will look at visual and hearing impairment in adults with intellectual disabilities and its effect on their mental health and development of autism. The study aims both to further the knowledge base in this area and to provide data to assist with service planning and provision for this client group.

2. What will be involved if my patient takes part in the study?

Anonymised information will be obtained on all individuals on the Leicestershire Learning Disabilities Register with visual or hearing impairment. All individuals with total sensory impairment (deafness/blindness) will be allocated to case group. A control group without such impairment (matched on gender and degree of intellectual disability) will also be selected. Carers, and where appropriate, the patients themselves will then be contacted by letter and asked

to participate in an interview on the topic to ascertain further information. Interviews will be carried out at carer' and patient's convenience.

3. Will information in the study be confidential?

Any information obtained from the Register and interviews will be treated as confidential information. After the interview coded numbers will replace names on all documents containing patient-identifiable information so that anonymity is preserved.

4. What if my patient is harmed in the study?

Medical research is covered for mishaps in the same way as for patients undergoing treatment in the NHS. Compensation is only available if negligence occurs.

5. What happens if I do not wish my patient to participate in this study or wish to withdraw them from the study?

If you do not wish your patient to participate in this study or if you wish to withdraw them from the study you may do so without justifying your decision.

RMO Consent Form

Sensory impairment, mental ill health and autistic spectrum disorder in people with intellectual disability

Principal investigator Dr R Kiani
Supervisors Dr S Bhaumik and Professor T Brugha

Patient's Name ……

Patient's DOB ……

This form should be read in conjunction with the Information Leaflet

I agree for my patient to take part in the above study as described in the Information Leaflet.

I understand that I may withdraw consent at any time without justifying my decision.

I understand that medical research is covered for mishaps in the same way as for patients undergoing treatment in the NHS i.e. compensation is only available if negligence occurs.

I have read the Information Leaflet on the above study and have had the opportunity to discuss the details with Dr Kiani and ask any questions. The nature and purpose of the assessments to be undertaken have been explained to me and I understand what will be required in the study.

Signature of RMO ……
Date ……

Signature of Investigator ……
Date ……

APPENDIX 8: PARTICIPANT CONSENT FORM

Leicestershire Partnership NHS
NHS Trust

A University Teaching Trust

Learning Disabilities Division
Mansion House
Leicester Frith Hospital
Groby Road
Leicester
LE3 9QF

Tel: 0116 225 5200
Fax: 0116 225 5202
www.leicspart.nhs.uk

Sensory impairment, mental ill health and autistic spectrum disorder in people with intellectual disability

Researcher: Dr R Kiani

I am the carer for _____ (Please fill in the patient's name)

	Please initial
1. I confirm that I have read and understood the invitation letter.	
2. I confirm that I have read and understood the study information sheet for the above study and have had the opportunity to ask questions.	
3. I would / would not be willing to take part in an interview. (Please delete as applicable)	

Name of Carer	Date	Signature
Researcher	Date	Signature

1 copy for carer; 1 for researcher; 1 to be kept with hospital notes

APPENDIX 9: SENSORY IMPAIRMENT PROFORMA

Patient's ID

DOB

Age group

Marital status

Sex

Ethnicity

Employment

Paid
Voluntary

Type of Accommodation

Living with whom?

Who is the carer?

Has carer had assessment of their need?

Family history

Are family still in touch?

Degree of learning disability

Aetiology of learning disability including genetic syndrome (if present)

Co-morbid psychiatric diagnosis if already assessed, including

Mental illness (including dementia)
Autism and/or other neuro-developmental disorders (e.g. ADHD)
Challenging behaviour

Who has made this diagnosis?

What assessment tools have been used for diagnosing?

Mental illness
Autism
Challenging behaviour

Any psychiatric admission

On any psychotropic medication

On Mental Heath Act Section

On DOLS

On CPA

Forensic history

Alcohol and substance misuse

Smoking status

Sensory impairment

The degree
Nature and
Aetiology of the sensory impairment

This was diagnosed

By who and
When

History of any

Assessment,
Management or
Intervention for a sensory impairment

Compliance with hearing aids (if assessed and prescribed)

Compliance with eyeglasses (if assessed and prescribed)

Regular follow up and monitoring of SI

Hearing aids and spectacles are regularly maintained

Environmental adaptation

Other sensory aids

Communication skills

Comprehension
Expression

Makaton
Sign language
Others

Charity organisation involved

Staff received training in sensory impairment

Staff has the skill and confidence in communicating with the clients

Communication passport

Active epilepsy (on medication)

Type (s) of epilepsy (if recorded)
Well controlled
Relatively well controlled
Poorly controlled

Cerebral palsy

Incontinence

Type of incontinence

Motor problems

Problem with gait
Problem with coordination
Problems with hands and fine motor movement
Hemipelgia
Quadriplegia
Others (detail)

Swallowing problems

Other co-morbid physical illness

Health Action Plan

Health Facilitator

Regular review of physical health including physical examination and investigation through GP and Health Facilitator

GP appointment in previous 12 months

Please explain

Any hospital admission for a medical reason

On any physical medication

Life events in the past 12 months, if yes,

How many and
What

Person Centred Plan (PCP)

Access to services

Day centre
Respite care
Leisure activities
Home support
Community visit

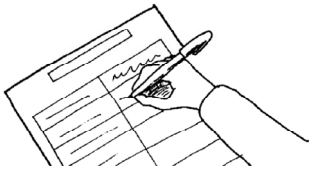
Direct payment

Other professionals involved in patient's care and for what reason:

Psychologist
Occupational Therapist
Speech and Language Therapist
Community nurse
Physiotherapist
Social worker
District nurse
Advocate
Probation service
Private support providers
Housing association
Other specialists

Any other issue that family/carers would like to discuss

APPENDIX 10: HEARING IMPAIRMENT CHECKLIST



Checklist for Hearing

Leicestershire Partnership Trust
Speech and Language Therapy

Appearance of ears	**Present**	**Comments**
What do the person's ears look like, (size, shape, any visible scars or damage)		
What the person does		
Does not speak		
Speaks very loudly or shouts		
Speaks very quietly or whispers		
Speaks in monotonous tone		
Speech hard to understand		
Unusual pronunciation of words		
Breathes through mouth not nose		
Frequent catarrh (blocked-up nose)		
Difficulty concentrating		
Poor balance		
On occasions seems confused		
Increased lack of co-operation at times/in certain places		
Can seems depressed for no apparent reason		
Watches people's faces closely		
Startled by people approaching who are not in their sight		
Can misunderstand instructions at times		
Responds better to some people's voices than others		
Can miss parts of conversation at times		
Takes time to 'tune in'		
Understands better with visual Prompts, e.g. objects, gestures		
Watches and follows the actions of others		
May have difficulties understanding when people change the subject in conversation		

Checklist for Hearing 2

Observation of behaviours may give an insight into what someone can hear. The person may do more or less of certain things in response to sounds. Record if the person does these things or not. If they do, comment on the situation, e.g. what's happening at the time.

What the person does	Present	Comments
Pokes or taps ears		
Moves ears in response to sound		
Slaps ears or side of head		
Covers ears with one or both hands		
Increases vocalisation in echoey places, e.g. bathroom, large rooms		
Removes self from noisy situations		
Makes sounds themselves, e.g. banging doors or tables		
Experiments with sounds, e.g. cups hand round their ear or shouting in the palm of their hand		
Is startled by loud noises		
Is woken by noises		
Obviously dislikes loud noises		
Watches others intently and copies their behaviour		
Has distinct musical preferences		
Sings/hums to tunes		
Moves spontaneously to music		
Enjoys using musical instruments		
Likes music to be loud		
Gets close to sounds, e.g. ear pressed to loudspeakers		
Switches music on by themselves		
Likes the vibrations rather than the		

sounds, e.g. sits on speakers, washing machines, spin dryers		
Can be distracted by sound when concentrating on something else		
Seeks sound source		
Copes well in a noisy environment		
Copes well in a quiet environment		
How the person interacts	**Present**	**Comments**
Turns or inclines head towards sounds		
Turns or inclines head towards speaker		
Copies any sounds used by others		
Invites others to shout in their ear		
Likes to listen to voices		
Turns to name being called		
Turns to name on both left and right		
Responds immediately Delay in response Responds to repetition		
Responds to shouting		
Responds to a whisper		
Difficulty listening to others speaking in a noisy environment		
Understands better in a quiet environment		

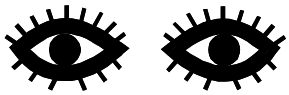
Checklist for Hearing 3

Environmental sounds. The following checklist is of a variety of environmental sounds. Observe . . . . . and note their response to them. They may do different things on different occasions. If there are any special circumstances make a note of these. For example, the person may respond to a sound without seeing the sound being made.

In the Home	**Response**
Television / radio music	
Television / radio talking	
Telephone ringing	
Door bell	
Door knocking	
Door closing	
Toilet flushing	
Paper tearing	
Vacuum	
In the Kitchen	**Response**
Toaster popping up	
Kettle boiling	
Washing dishes	
Washing machine	
Crockery rattling	
Cutlery rattling	
Stirring a spoon in a cup	
Chopping food on a board	
Opening a packet	
Water filling a sink	
Oven or microwave timer	
Pans rattling	

Frying pan sizzling	
Dropping cutlery / crockery	
Outside	**Response**
Car engine starting	
Car passing by	
Lorry passing by	
Motorbike passing by	
Siren (ambulance, fire, police)	
Aeroplane	
Car horn	
Footsteps on pavement	
Lawn mower	
Hammering	
Rain	
Wind	
Car radio	
Birds singing	
Dogs barking	
Human Sounds	**Response**
Talking	
Shouting	
Whispering	
Singing	
Laughing	
Sneezing	
Whistling	
Shhhh!	
Coughing	
Humming	

APPENDIX 11: VISUAL IMPAIRMENT CHECKLIST



Vision Checklist

Appearance of person's eyes	**L**	**R**	**Comments**
e.g. size, shape of pupil, noticeable squint, scarring/damage			
Movement of person's eyes			
Person's eyes - move slowly			
Person's eyes - move together			
Person's eyes - move independently of each other			
Any abnormal movement of eyes			
Person's behaviours	**Comments**		
Hand/finger flicking in front of eyes			
Enjoys flapping objects In front of eyes			
Eye poking or rubbing			
Head positioning, e.g. tipped To one side			
Rolling or shaking of head			
Does the person look at an object straight on or sideways?			
Does the person prefer to be approached from one particular side - if so, which?			
Does the person appear to ignore the presence of an individual or object on one particular side?			
Person's eye tracking	**Comments**		
Do the person's eyes follow movement?			
Does the person track specific objects, light or movement?			

Give examples	
Person's near and distant vision	**Comments**
Does the person hold objects very close to look at or at arm's length?	
Does the person recognise objects near to them or at a distance?	
Is the person able to recognise people in the distance (12 ft away)?	
Is the person able to recognise people close to (1 ft away)?	
Person's near and distant vision	**Comments**
Does the person see people wearing bright clothes better?	
Does the person fail to find small objects or place them accurately?	
Does the person grope to pick things up rather than reaching accurately?	
Does the person appear to see moving objects better than things that stay still?	
Explores people/items through touch?	
Does the person move very close to people/stares at people?	
Contrast	**Comments**
Does the person have a preference for bright objects/lights/colours?	
Does the person see bright objects on colour contrasting backgrounds better, e.g. a red plate on a white tablecloth? (specify which colours	

work best).	
Does the use of colours make any difference in the person's ability to complete tasks or join in?	
Can the person select a sweet from similar items, e.g. buttons, pebbles?	
Can the person select an object on a patterned surface, e.g. sweet on checked tablecloth?	
Person's mobility	**Comments**
Does the person bump into things, e.g. doors, furniture?	
Is the person anxious or unwilling to move independently - if so, what are the circumstances?	
Does the person find it difficult to judge distances? e.g. when using steps	
Is the person un-coordinated in any way?	
Does the person find it difficult to cope with changes in the environment? e.g. when furniture is moved	
Does the person walk confidently in well-lit conditions, but has difficulty in dimly lit conditions or stairwells?	
Does the person constantly look down? e.g. from a carpeted to tiled floor surface	
Does the person move their head from side to side to scan their surroundings when walking?	

Does the person appear to 'feel' their way around, not obviously using their sight? , e.g. sliding feet to find steps/using their hands along a wall for guidance	
Is the person reluctant to move in unfamiliar places?	
Does the person prefer to have their shoes off and walk around with bare feet?	
Does the person have any problems with balance?	
Does the person have any difficulties with steps or using stairs?	

Adapted from The Mulberry Trust
SALT/Communication Tools/Visionchecklist13.12.04

APPENDIX 12: PERVASIVE DEVELOPMENTAL DISORDER - MENTAL RETARDATION SCALE (PDD-MRS)

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South Birmingham NHS
Primary Care Trust

Birmingham Learning Disabilities Service
The Greenfields, Monyhull
P O Box 7041
BIRMINGHAM
B30 3QQ
Tel: 0121 465 8750 (Direct Line: 465 8763) Fax: 0121 465 8804

Date Typed: **7 December 2006**

MR/KC

Dr Kiani
Specialist Registrar to Dr Bhowmik
Learning Disabilities Services
Mansion House Firth Hospital
LEICESTER
LE3 9QF

Dear Dr Kiani

Please find enclosed a copy of the PDD-MRS and some further information. I know that following my discussions with him Dr Kraijer may have modified items 1 and 2 slightly. As a coincidence, I have just had an e-mail from him and I have told him about your project.

With kind regards

Yours sincerely

Katrina Cunningham

pp. **Dr Meera Roy, FRCPsych, FRANZCP**
Consultant Psychiatrist

Encs

APPENDIX 13: ABERRANT BEHAVIOUR CHECKLIST (ABC)

*APPENDIX 14: USING SAS TO MATCH CONTROLS TO CASES**

*Although stage 2 did not involve a case-control study, the same matching principle can be applied. For more detail please refer to Kawabata *et al.* (2004).

APPENDIX 15: GENETIC SYNDROMES AND OTHER AETIOLOGICAL CONDITIONS

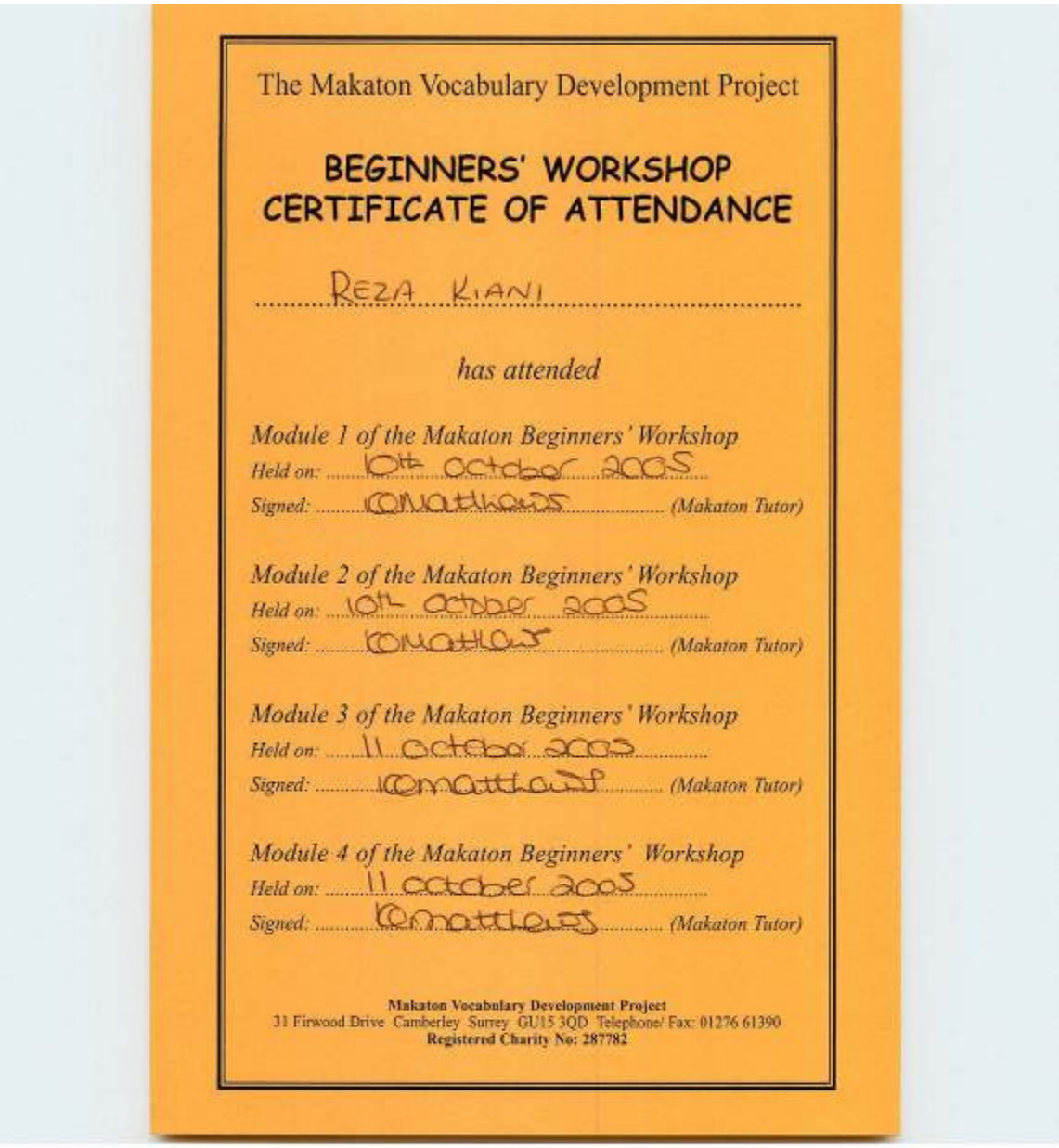
- 15q24 microdeletion
- 18q deletion
- 18q12.3 microdeletion
- Angelman syndrome
- Anophthalmia
- Batten disease
- Beta thalassemia
- Cerebral palsy and other causes of brain damage prenatally
- Coffin Lowry syndrome
- Cohen syndrome
- Coloboma
- Congenital cataract
- Cornelia de lange syndrome
- Cri du chat syndrome
- Down syndrome
- Fragile X syndrome
- Halt Oram syndrome
- Infantile spasm
- Joubert syndrome
- Klinefelter syndrome
- Laurence-Moon-Bardet-Biedl
- Leber's congenital amourosis
- Lennox Gastaut syndrome
- Meningo-encephalitis
- Metabolic (Renal Tubular Acidosis, hypoglycaemica, Congenital Adrenal Hyperplasia)
- Mitochondrial disease
- Mucopolysaccharidosis
- Norrie disease

- Ocular Albinism
- Osteogenesis imperfecta
- PEHO (Progressive Encephalopathy with Oedema, Hypsarrythmia and Optic atrophy)
- Prematurity
- Retinitis pigmentosa
- Rett syndrome
- Rhesus haemolytic disease
- Septo-optic dysplasia
- Sturge Weber syndrome
- Shaken baby syndrome
- TORCHES
- Tuberous sclerosis
- Waardenburg syndrome

APPENDIX 16: TRAINING COURSES AND ACCREDITATIONS

Summary of training courses and accreditations

- Makaton modules 1, 2, 3 and 4, Grantham, 2005
- Describing and presenting data, Leicester, 2006
- Analysing data and further data analysis using SPSS, Leicester, 2006
- Having confidence in data, Leicester, 2006
- Designing a questionnaire, Leicester, 2006
- The senses sight and sound, Bristol, 2006
- British Sign Language Level 1, 2, 3 and pre level 4 from the Council for the Advancement of Communication with Deaf People, CACDP, Lincoln, Leicester and Birmingham, 2006–2009.
- Deaf-blind manual and alphabet level 2 from Deaf blind UK, Peterborough, 2008.
- Diagnostic Interview for Social and Communication Disorders (DISCO), Lorna Wing centre, Bromley, Kent, 2007–2008.
- An introduction to visual impairments and autism spectrum conditions, RNIB/NAS, Birmingham, 2011.
- Autism Diagnostic Observation Schedule, 2nd edition, Spectrum Specialist Consultancy, Leicester, 2012.
- Better eye care for people with LD, Public Health England, Peterborough, 2013 (https://www.improvinghealthandlives.org.uk/)
- Mary Kitzinger Workshop on visual impairment, Institute of Child Health, UCL, 2012, 2013 & 2014.



The Makaton Vocabulary Development Project

BEGINNERS' WORKSHOP CERTIFICATE OF ATTENDANCE

REZA KIANI

has attended

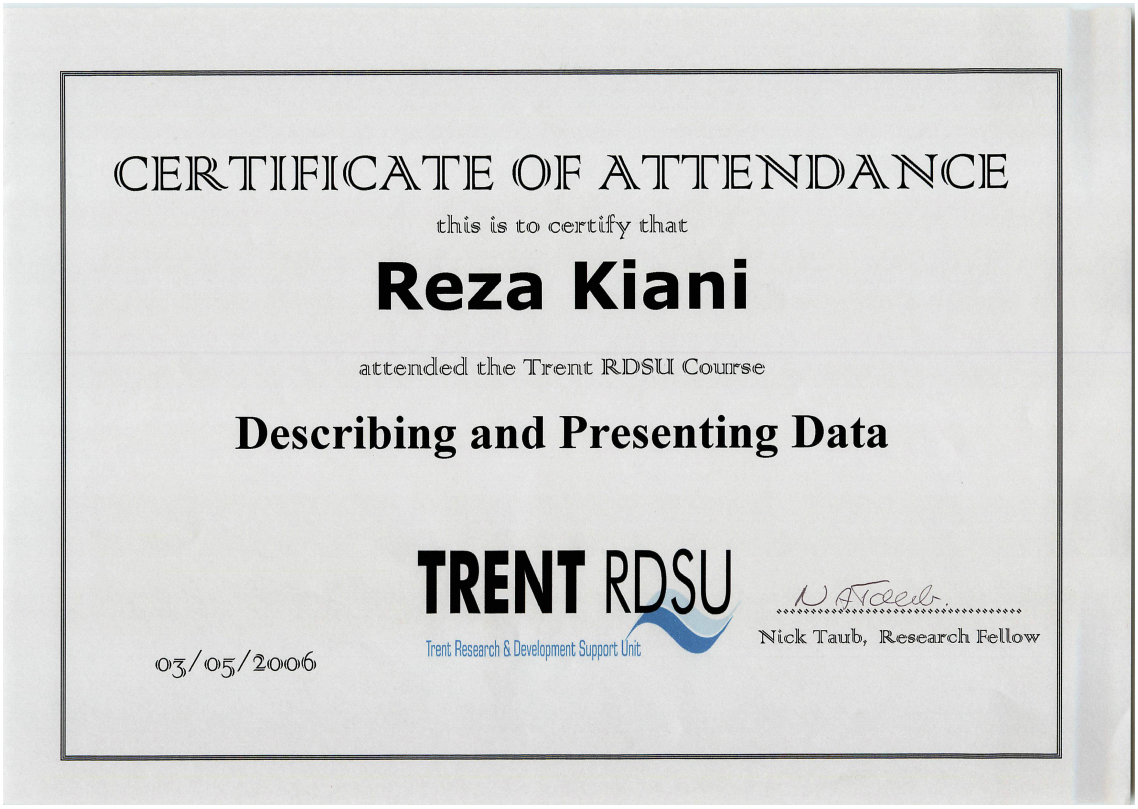
Module 1 of the Makaton Beginners' Workshop
Held on: 10th October 2005
Signed: K Matthews *(Makaton Tutor)*

Module 2 of the Makaton Beginners' Workshop
Held on: 10th October 2005
Signed: K Matthews *(Makaton Tutor)*

Module 3 of the Makaton Beginners' Workshop
Held on: 11 October 2005
Signed: K Matthews *(Makaton Tutor)*

Module 4 of the Makaton Beginners' Workshop
Held on: 11 October 2005
Signed: K Matthews *(Makaton Tutor)*

Makaton Vocabulary Development Project
31 Firwood Drive Camberley Surrey GU15 3QD Telephone/ Fax: 01276 61390
Registered Charity No: 287782



CERTIFICATE OF ATTENDANCE

this is to certify that

Reza Kiani

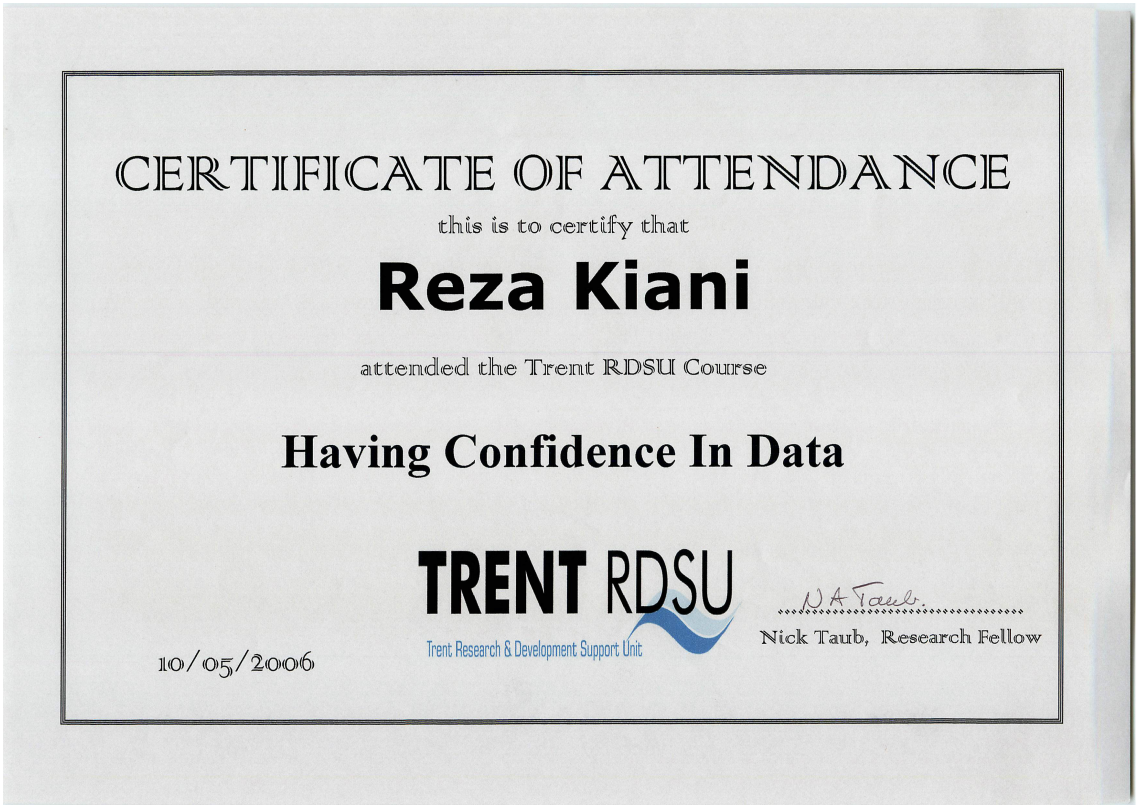
attended the Trent RDSU Course

Describing and Presenting Data

TRENT RDSU
Trent Research & Development Support Unit

Nick Taub, Research Fellow

03/05/2006



CERTIFICATE OF ATTENDANCE

this is to certify that

Reza Kiani

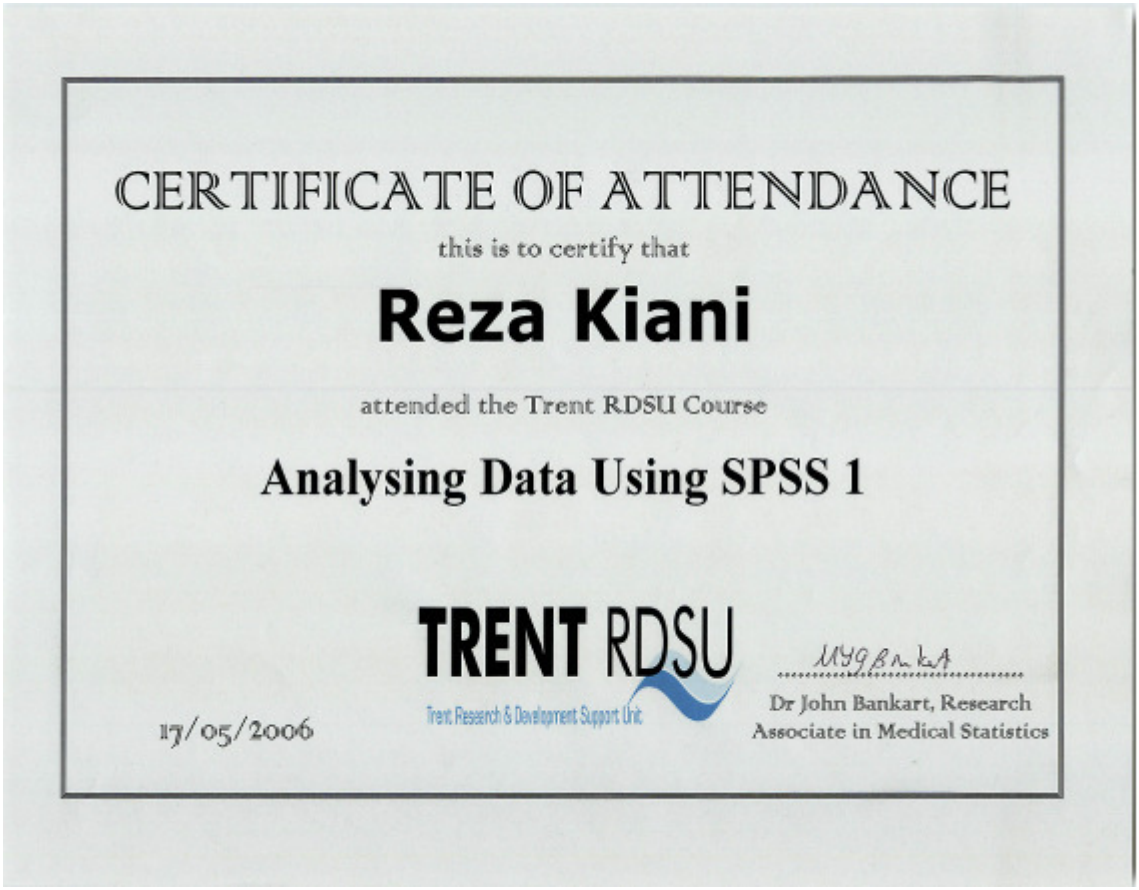
attended the Trent RDSU Course

Having Confidence In Data

TRENT RDSU
Trent Research & Development Support Unit

Nick Taub, Research Fellow

10/05/2006



CERTIFICATE OF ATTENDANCE

this is to certify that

Reza Kiani

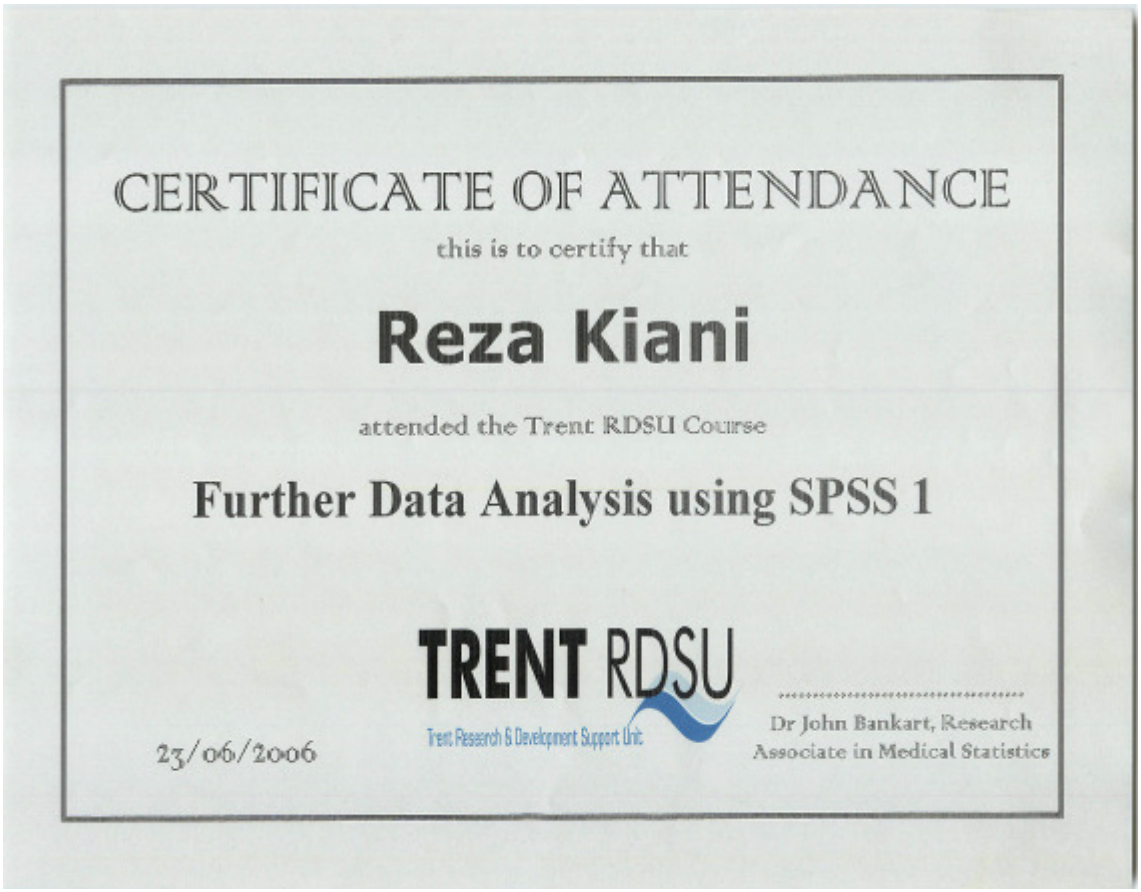
attended the Trent RDSU Course

Analysing Data Using SPSS 1

TRENT RDSU
Trent Research & Development Support Unit

17/05/2006

Dr John Bankart, Research Associate in Medical Statistics



CERTIFICATE OF ATTENDANCE

this is to certify that

Reza Kiani

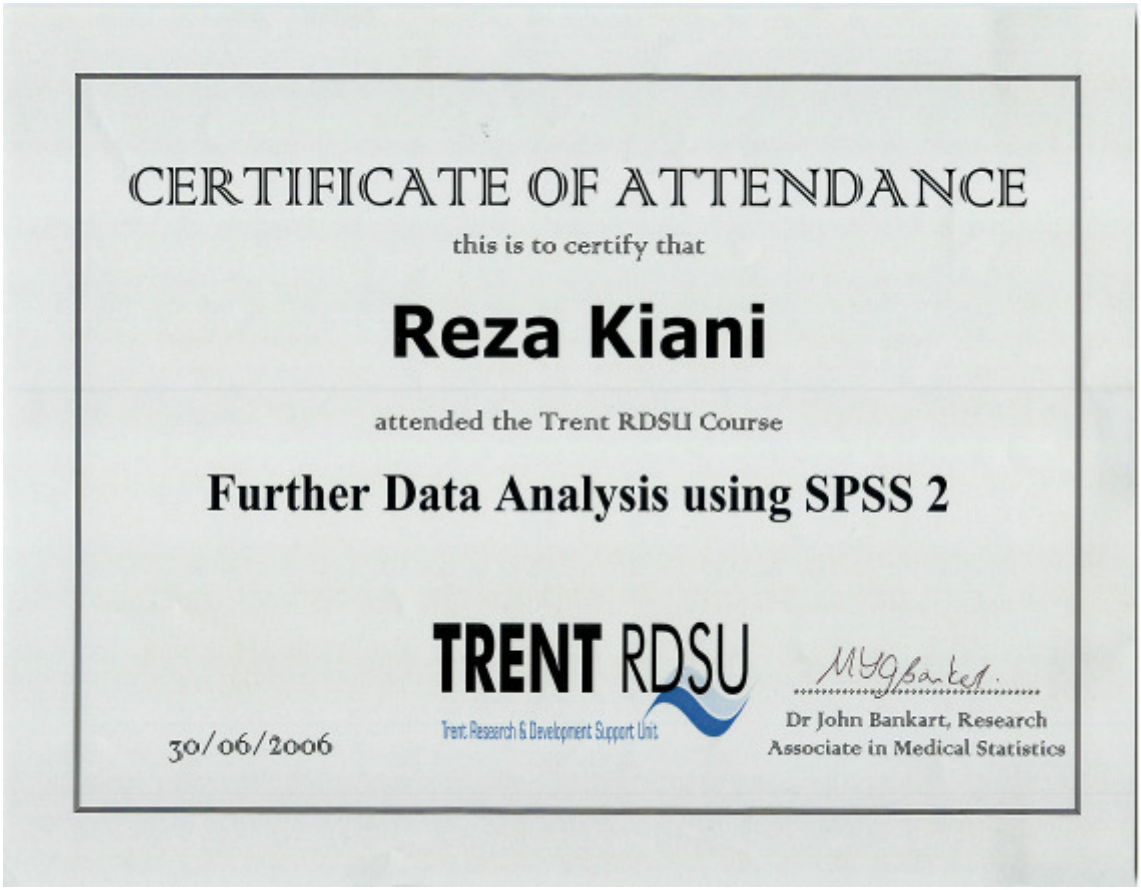
attended the Trent RDSU Course

Further Data Analysis using SPSS 1

TRENT RDSU
Trent Research & Development Support Unit

23/06/2006

Dr John Bankart, Research Associate in Medical Statistics



CERTIFICATE OF ATTENDANCE

this is to certify that

Reza Kiani

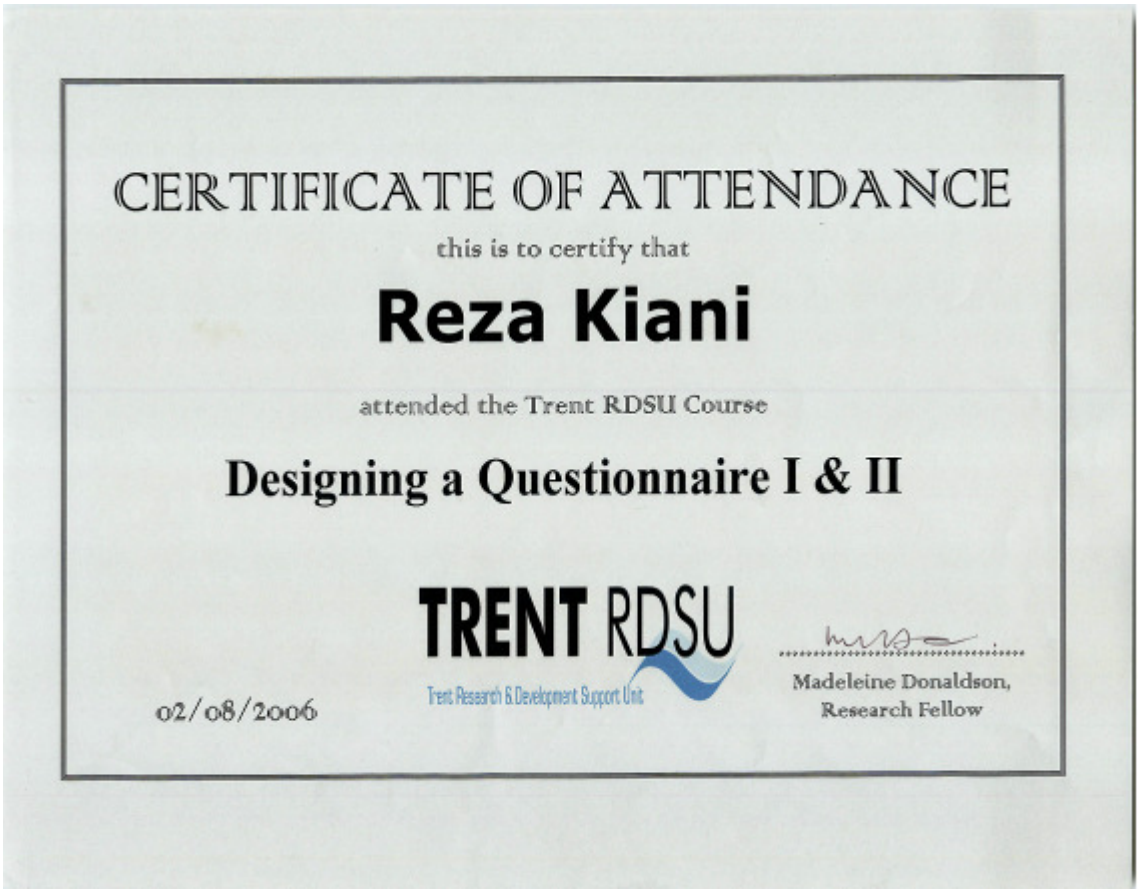
attended the Trent RDSU Course

Further Data Analysis using SPSS 2

TRENT RDSU
Trent Research & Development Support Unit

30/06/2006

Dr John Bankart, Research Associate in Medical Statistics



CERTIFICATE OF ATTENDANCE

this is to certify that

Reza Kiani

attended the Trent RDSU Course

Designing a Questionnaire I & II

TRENT RDSU
Trent Research & Development Support Unit

02/08/2006

Madeleine Donaldson, Research Fellow

Avon Learning Difficulties Education and Research Network

ACADEMIC PROGRAMME 2006

CERTIFICATE OF ATTENDANCE

Record of attendance at the

Multidisciplinary meeting entitled

"THE SENSES – SIGHT AND SOUND"

(3 external CPD units)

held on

WEDNESDAY 24th MAY 2006
at
New Friends Hall, Heath House Lane, Stapleton,
Bristol BS16 1EQ

Signed .....
(Attendee)

Signed .....
(Programme Director)

Lincolnshire
COUNTY COUNCIL

Lincoln Area Lifelong Learning
City Community Education Centre
Skellingthorpe Road
LINCOLN LN6 0EP
Tel: 01522 684577 Fax: 01522 872350
brenda.bainbridge@lincolnshire.gov.uk

August 2006

CACDP BSL LEVEL 1 EXAMINATION

Congratulations. I am pleased to tell you that we have received the result of your CACDP exam and you have passed. We have also received your Certificate.

We prefer not to post your Certificate in case it gets lost or damaged, as replacements are expensive. I should therefore be grateful if you would arrange to collect your Certificate. You can collect it from City Community Education Centre, Skellingthorpe Road, Lincoln (follow the signs for Adult Education when you come through the entrance to City of Lincoln Community College).

In the summer, the office is usually open Monday-Thursday, 9.30am-2.30pm; Friday 9.30am-12.30pm, though it is best to phone and check before you call in.

If you cannot collect your Certificate during these times, please phone to see if we can make alternative arrangements.

Yours sincerely

Brenda Bainbridge
Examinations Administrator

Exams\BSL1cert

THE COUNCIL FOR THE ADVANCEMENT OF COMMUNICATION WITH DEAF PEOPLE

Advancing Communication between Deaf and Hearing People

QCA Qualifications and Curriculum Authority

cea Rewarding Learning

accac Qualifications, Curriculum & Assessment Authority for Wales

This is to certify that

Abdolreza Ashtarikiani

has achieved the

Level 1 Certificate in British Sign Language

8 August 2006

National Accreditation No: 100/1521/8

CJ/50517/J4438

Chris Hughes CBE
Chairman

Miranda Pickersgill
Chief Executive

CACDP · Durham University Science Park · Block 4 · Stockton Road · Durham DH1 3UZ

135 Welford Road, Leicester LE2 6BE
Telephone: 0116 257 4800 Textphone: 0116 257 4850 Fax: 0116 257 4856
Email: enquiries@actiondeafness.org.uk Web: www.actiondeafness.org.uk
Chief Executive Officer: Craig A Crowley MBE

action DEAFNESS
independence equality & opportunity

Our reference: PS/BSL2/WEDNESDAY

4th September 2007

Dear Abdolereza

I am very pleased to send you your Level 2 British Sign Language Certificate and may I offer you my congratulations on your success.

The new prospectus is now out and we look forward to welcoming you back to Action Deafness in September should you be interested in enrolling onto our Level 3 BSL programme.

Should you have any queries or questions, please do not hesitate to contact Liz Brown, Learning Services Officer on lizbrown@actiondeafness.org.uk, who will be able to assist you.

We look forward to seeing you again in September!

Yours sincerely,

Liz Brown

Learning Services Officer

Advancing Communication between Deaf and Hearing People

Qualifications and Curriculum Authority

Llywodraeth Cynulliad Cymru Welsh Assembly Government

Rewarding Learning

Qualification Certificate

Abdolreza Ashtarikiani

has achieved the

Level 2 Certificate in British Sign Language

National Accreditation Number: 100/6195/2

BSL Receptive Skills
BSL201 - National Accreditation Number: Y/500/2761

BSL Productive Skills
BSL202 - National Accreditation Number: D/500/2762

BSL Conversational Skills
BSL203 - National Accreditation Number: H/500/2763

Date: 17 August 2007

017832/N3141

Chris Hughes CBE
Chairman

CJ Edwards
Chief Executive

CACDP · Durham University Science Park · Block 4 · Stockton Road · Durham DH1 3UZ

135 Welford Road, Leicester LE2 6BE
Telephone: 0116 257 4800 Textphone: 0116 257 4850 Fax: 0116 257 4856
Email: enquiries@actiondeafness.org.uk Web: www.actiondeafness.org.uk
Chief Executive Officer: Craig A Crowley MBE

action DEAFNESS
independence equality & opportunity

10th September 2008

Dear Action Deafness Student

Action Deafness CACDP Award Evening

I would like to congratulate you on your recent success in completing your British Sign Language Course at Action Deafness and in obtaining a qualification of your choice in Deaf Studies.

For the first time ever, Action Deafness is organising an Awards Evening for all our students who have successfully completed and passed their CACDP British Sign Language course at Action Deafness. This will be become an annual event to congratulate students on their success.

I would like to invite you to the Awards evening which will take place on Wednesday 1st October 2008 at 7pm till 9pm, here at Action Deafness, 135 Welford Road, Leicester.
Jim Edwards, Chief Executive Officer of CACDP and Craig Crowley, Chief Executive Officer of Action Deafness will present students with their certificates. The local media and press will be invited to attend the event as well.

Could you complete the attached slip below and send it back as soon as possible to Liz Brown, Action Deafness, 135 Welford Road, Leicester, LE2 6BE or alternatively you can email:lizbrown@actiondeafness.org.uk or telephone 0116 2574838 to state if you are able or unable to attend the evening.

If you are able to attend the Awards Ceremony, please bring your CACDP certificate so we can formally present it to you on the night.

I look forward to meeting you again.

Yours faithfully

Liz Brown
Deputy Manager of Learning Services

135 Welford Road, Leicester LE2 6BE
Telephone: 0116 257 4800 Textphone: 0116 257 4850 Fax: 0116 257 4856
Email: enquiries@actiondeafness.org.uk Web: www.actiondeafness.org.uk
Chief Executive Officer: Craig A Crowley MBE

action DEAFNESS
independence equality & opportunity

Abzolreza Ashtarikiani
23 Timberwood Drive
Groby
LE6 0YU

17th September 2008

Dear Abzolreza

CACDP Level 3 Certificate in British Sign Language

I am very pleased to send you your Level 3 British Sign Language BSL 301 and BSL 302 assessment units result slips.

May I offer you my congratulations on your success!

Yours sincerely,

Liz Brown

Deputy Manager of Learning Services

Encs. Unit BSL 301 and Unit BSL 302 result slips

This is to certify that

Abdolreza Ashtarikiani

has achieved the following qualification:

Level 3 Certificate in British Sign Language

QCA Accreditation Number: 100/6104/6

comprising the following units:

BSL301 Sign Language Receptive Skills
QCA Accreditation Number: Y/500/2033

BSL302 Sign Language Productive Skills
QCA Accreditation Number: D/500/2034

Date: 15/09/2008

0027967X/3141

Chris Hughes CBE
Chairman

CJ Edwards
Chief Executive

CACDP · Durham University Science Park · Block 4 · Stockton Road · Durham DH1 3UZ

sign solutions
nationwide sign language service

Thursday 12 March 2009

Mr A Kiani
23 Timberwood Drive
Groby
Leicester
LE3 9QF

Dear Reza,

Thank you for attending the Pre Level 4 BSL Initial Assessment at Sign Solutions.

Having considered your evidence carefully, we are pleased to inform you that we are able to offer you a place on the Pre Level 4 BSL course commencing 18 May 2009.

Enclosed is a feedback sheet which we trust will be useful to you as it will enable you to focus on identified areas prior to the beginning of your course here. We would encourage you to keep your feedback for future reference.

I would be grateful if you could provide the following information as soon as possible, the deadline being 27 March 2009.

- Confirmation that you wish to take up a place on our Pre Level 4 BSL Course commencing in May, with the course deposit of £500. If your employer/other third party are paying your fees, we require a letter from them confirming that they will be meeting the required costs and providing invoicing information. The remainder of your course fee is then due on 1 May 2009 .Cheques or Bacs payments should be made payable to Sign Solutions.
- One signed copy of the Learning Agreement, this must also be signed by your employer/other third party if you are not self-funded (the second copy is for your own records)

We look forward to hearing from you soon.

Yours sincerely

NVQ Training & Assessment Team

T005/05/15/01/09

Directors: Sean Nicholson MA, B.Sc(Hons), LASLI, MRSLI,MITI I Gloria Ogborn FASLI, MRSLI, MITI, MAPCI
Sign Solutions *(Sign Language Interpreting Agency)* Ltd
VAT NUMBER: 736 5367 12 REGISTERED No. WALES 4802772

Bordesley Hall I The Holloway I Alvechurch I Birmingham I B48 7QA
Tel: 0845 07700 41/42 I Fax: 0845 07700 43
Minicom: 0845 07700 41 I SMS: 07816217228
office@signsolutions.uk.com I www.signsolutions.uk.com

DEAFBLIND
·UK·

15 May 2008

Dr R Kiana
Learning Disability Services
Mansion House
Leicester Frith Hospital
Leicester
LE3 9QF

CACDP Candidate ID: 00609583

Dear Reza

CACDP Level 2 Certificate in Communication with Deafblind People (Manual) – February/March 2008

I have pleasure in enclosing your CACDP Result Slips to confirm that you successfully passed all three units of the above qualification. Well done!

Apologies for the lack of certificate: this will be forwarded as soon as we receive them from CACDP.

May I take this opportunity to thank you for your commitment during the course and of course congratulate you on your achievement.

Please do not hesitate to contact the Training Department if you have any questions.

Kind regards,

Julie Nettelfield
PA to Julie Brown, Training Officer

Enc

1928 2008
DEAFBLIND
·UK·
80 years of supporting deafblind people

FREE HELPLINE
0800 132 320

For deafblind people and those who support them

DEAFBLIND UK
National Centre for Deafblindness, John and Lucille van Geest Place, Cygnet Road, Hampton, Peterborough PE7 8FD.
Tel/Textphone: 01733 358100 Fax: 01733 358356
Email: info@deafblind.org.uk Website: www.deafblind.org.uk

Advancing Communication between Deaf and Hearing People

Qualifications and Curriculum Authority

Llywodraeth Cynulliad Cymru Welsh Assembly Government

Rewarding Learning

This is to certify that

Reza Kiani

has achieved the following qualification:

Level 2 Certificate in Communication with Deafblind People

QCA Accreditation Number: 500/1482/1

comprising the following units:

K202 Understanding Communication with Deafblind People
QCA Accreditation Number: R/500/2371

T201 Communicating with Deaf and Deafblind People (ACE)
QCA Accreditation Number: R/500/2368

T202 Deafblind Manual Communication and Guiding
QCA Accreditation Number: Y/500/2372

Date: 23/06/2008

00609583/2750

Chris Hughes CBE
Chairman

CJ Edwards
Chief Executive

C J Edwards

CACDP · Durham University Science Park · Block 4 · Stockton Road · Durham DH1 3UZ

NAS CENTRAL
REGIONAL TEAM

The National Autistic Society

SEMINAR
An Introduction to Visual Impairments and Autism Spectrum Conditions
Facilitated by Ian Bell

Whether you are a person on the Autism Spectrum, a family member, or support someone in a professional capacity, this seminar will be of interest to you.

Autism is more common in visually impaired children than in fully sighted children. In this seminar we will explore the nature of autism in visually impaired children, and discuss meeting the needs of children who have a visual impairment and autism. We will focus on the Resource Pack recently published by the Royal National Institute of Blind People (RNIB) the UK's leading charity offering information, support and advice to people with sight loss.

Thursday 3rd November 2011
Registration from 6.30pm, Seminar 7pm-9pm
The Hub, Hazelwell, 318 Vicarage Road, Kings Heath
Birmingham B14 7NH.

Ian taught children with severe learning difficulties for 12 years and then retrained as a speech and language therapist. As a therapist he has considerable experience with visually impaired children, especially those with additional complex needs, including autism spectrum conditions. Ian was Leader of the Visual Impairment and Autism Project, which finished in March 2011.

Cost:	
Professionals	**£25.00**
Parents & Carers	**£10.00**
Person with a diagnosis of Autism/Asperger Syndrome	**£5.00**

If you are interested in attending, please complete the booking form and return to:
Sarah Smith, Regional Team Administrator
The National Autistic Society
Castle Heights (6th Floor)
72 Maid Marian Way
Nottingham
NG1 6BJ
Email: sarah.smith@nas.org.uk
Tel: 0115 8473502

Please enclose payment by cheque with your booking form or tick the invoice option.

The Lorna Wing Centre for Autism

Elliot House, 113 Masons Hill, Bromley, Kent BR2 9HT
T: 020 8466 0098 F: 020 8466 0118 E: elliot.house@nas.org.uk
www.autism.org.uk Autism Helpline: 0845 070 4004

08 April 2008

Dr R Kiani
Psychiatrist
Adult Learning Disability Services
Mansion House
Leicester Frith Hospital
Leicester
LE3 9QF

Dear Reza

We have now been able to organise the printing and enclose your certificate. We do apologise for the long delay.

As mentioned on the course we informed you that we would require you to sign and return the enclosed Licence Agreement.

As you are now an accredited DISCO user your details are on our database. If you should move from your present base, could you please let us know so that we can keep you informed of any future developments.

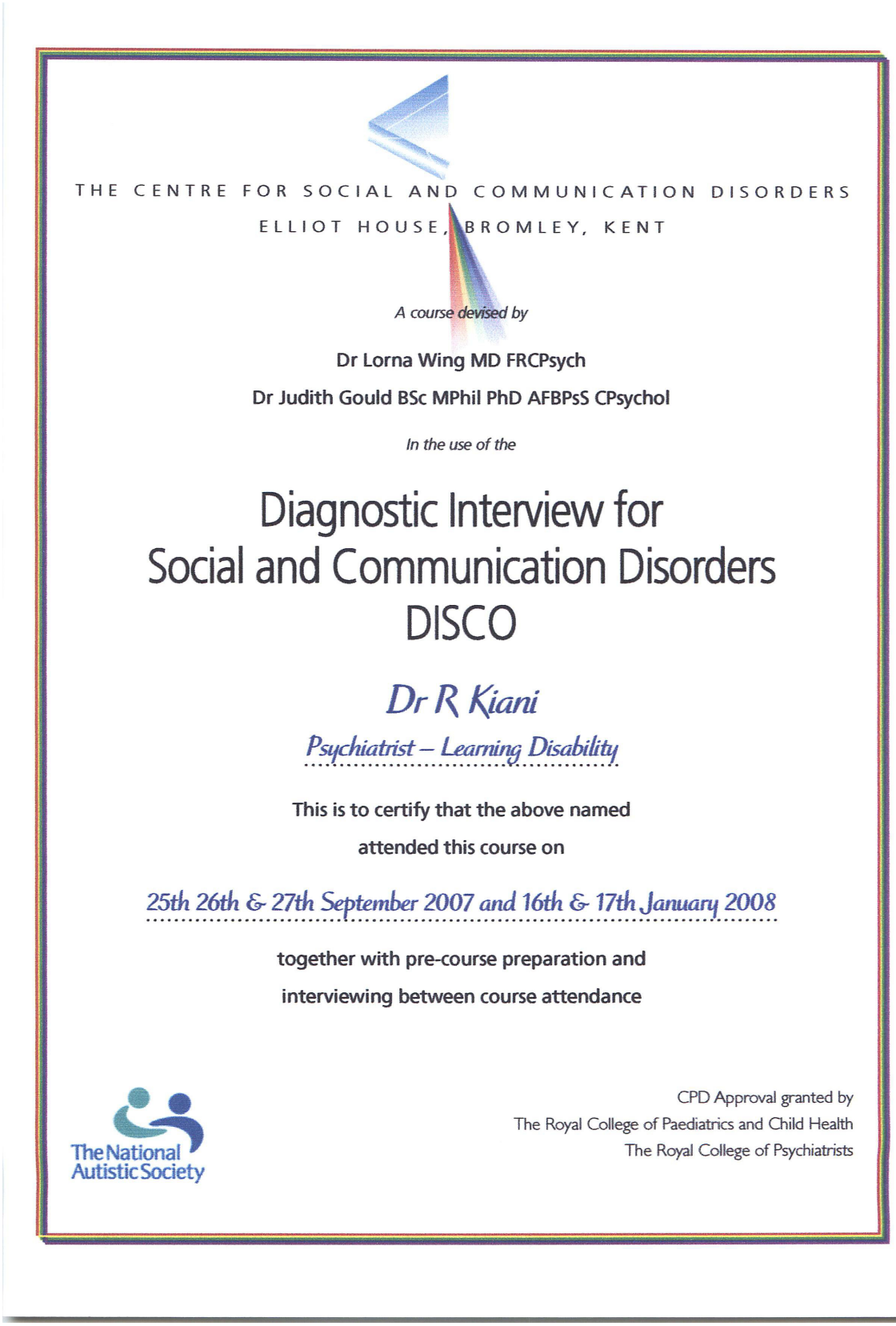
Best Wishes

Yours sincerely

Judith

Dr Judith Gould
Director – The NAS Lorna Wing Centre for Autism

Encl.



THE CENTRE FOR SOCIAL AND COMMUNICATION DISORDERS

ELLIOT HOUSE, BROMLEY, KENT

A course devised by

Dr Lorna Wing MD FRCPsych

Dr Judith Gould BSc MPhil PhD AFBPsS CPsychol

In the use of the

Diagnostic Interview for Social and Communication Disorders DISCO

Dr R Kiani

Psychiatrist – Learning Disability

This is to certify that the above named

attended this course on

25th 26th & 27th September 2007 and 16th & 17th January 2008

together with pre-course preparation and

interviewing between course attendance

The National Autistic Society

CPD Approval granted by

The Royal College of Paediatrics and Child Health

The Royal College of Psychiatrists

Spectrum Specialist Consultancy

Spectrum
Specialist Autism Consultancy & Training

Dr Fiona J Scott
Chartered Psychologist
HCPC Practitioner Psychologist
Autism Specialist
International ADOS/ADI-R Trainer

The British Psychological Society
Chartered Psychologist

Certificate of Attendance

This is to certify that

Dr Reza Kiani

attended a 5 day formal training course on the
Autism Diagnostic Observation Schedule 2nd Edition (ADOS-2)
and is qualified, where appropriate, to use all editions of the instrument in clinical practice

8-9 Oct & 11-13 Dec 2012
Date

Dr Fiona J Scott, ADOS/ADI-R Trainer

www.spectrumspecialistconsultancy.com

the Mary Kitzinger trust

Certificate of Attendance

This is to certify that

.....

attended the
Mary Kitzinger Trust Workshop
at the Institute of Child Health, UCL, London

"Adult-child interaction: Supporting communication, development and learning in the context of visual impairment"

On Saturday 17th May 2014
(10am-3.30pm)

Signed.....
Dr Elena Sakkalou, Workshop organiser
Dr Rebecca Greenaway, Workshop organiser

The Mary Kitzinger Trust (MKT), Registered Charity No 293473

http://www.marykitzingertrust.org/

the Mary Kitzinger trust

Certificate of Attendance

This is to certify that

REZA KIANI

attended the
Mary Kitzinger Trust Workshop
at the Institute of Child Health, UCL, London

Infant Observation and Research –
why it matters?
A tribute to Cathy Urwin

On Saturday 18th May 2013
(10am-3.25pm)

Signed.....
Dr Valerie Tadic, Workshop organiser
Dr Rebecca Greenaway, Workshop organiser

The Mary Kitzinger Trust (MKT), Registered Charity No 293473

http://www.marykitzingertrust.org/

the Mary Kitzinger trust

Certificate of Attendance

This is to certify that

Reza Kiani

attended the
Mary Kitzinger Trust Workshop
at the Institute of Child Health, UCL, London

Theme: 'Visual impairment and technology - supporting communication, education and activities of daily living'

On Saturday 26th May 2012
(10am-4pm)

Signed
Dr Valerie Tadic, Workshop organiser
Dr Rebecca Greenaway, Workshop organiser

The Mary Kitzinger Trust (MKT), Registered Charity No 293473

http://www.marykitzingertrust.org/

APPENDIX 17: PUBLICATIONS AND PRESENTATIONS ARISING FROM THIS THESIS

PUBLICATIONS

Kiani, R., Gandgadharan, S.K., Miller, H. (2007) Case report: Association of Waardenburg syndrome with intellectual disability, autistic spectrum disorder and unexplained aggressive outburst; a new behavioural phenotype? *British Journal of Developmental Disability,* 53 (104); 53-62.

Kiani, R., Miller, H., Gangadharan, S.K. (2008) Association of autism with sensory impairment. *British Journal of Developmental Disability*, 54; 59-61.

Miller, H. and **Kiani, R.** (2008) Inter-relationships between hearing impairment, learning disability services and mental health: are learning disability services 'deaf' to hearing impairments? *Advances in mental health and learning disabilities*, 2 (2), pp. 25-30.

Kiani, R. Miller, H. (2016) Sensory impairment and intellectual disability, CPD online module, The Royal College of Psychiatrists. http://www.psychiatrycpd.co.uk/

Kiani, R., Miller, H. (2010) Sensory impairment and intellectual disability. *Advances in Psychiatric Treatment.* 16; 228-235.

PRESENTATIONS

Kiani, R. Learning disability, Mental illness and Sensory Impairment, monthly academic meeting of adult learning disability service, Bristol, 2006

Kiani, R. Mental Illness in people with Learning Disability who have Visual and/or Hearing Impairment, Royal College of Psychiatrists' Faculty of Learning Disability Annual Residential Meeting, Prague, 2006

Kiani, R. (co-presented with Miller, H). Sensory impairment workshop, Royal College of Psychiatrists' Faculty of Learning Disability Annual Residential Meeting, Belfast, 2007

Kiani, R. (co-presented with Talbot, L). Sensory impairment in people with autism and learning disability, Royal College of Psychiatrists' Faculty of Learning Disability Annual Residential Meeting, Manchester, 2008

Kiani, R. Deafness, development and mental health, National Conference on deafness and mental health, Leicester, 2009

Kiani, R. (co-presented with Caldwell, P). Intensive Interaction in people with Intellectual Disability and autism, Therapeutics in learning disability symposium, Leicester, 2011

Kiani, R. (co-presented with Parker, R and Sinclair, R). Sensory integration for people with autism and learning disability, Learning Disability higher trainees national residential conference, Leicester, 2012

Kiani, R. Different applications of sensory integration, Evidence based practice in childhood mental health problems, Pune, India, 2012

Kiani, R. (co-presented with Sinclair, R). Sensory issues in offenders with intellectual disability, East midland low secure network conference, Nottingham, 2014

Kiani, R. (co-presented with Caldwell, P). Sensory interaction in people with autism and intellectual disability, Royal College of Psychiatrists' Faculty of Learning Disability Annual Residential Meeting, Birmingham, 2014

Royal College of Psychiatrists
Faculty of Psychiatry of Intellectual Disability
Annual Conference

Preliminary Programme

2-3 October 2014, Holiday Inn Hotel, Birmingham

Thursday **(Joint conference day with British Psychological Society)**

08:00-08:55	**Registration and Refreshments**
09:00-09:05	**Welcome and Introductory Comments** Dr Ashok Roy, Consultant Psychiatrist, Coventry and Warwickshire Partnership Trust & Chair, Faculty of Psychiatry of Intellectual Disability, Royal College of Psychiatrists
09:05-11:00	**Keynote Sessions** **Chair** – Dr Jane McCarthy, Consultant Psychiatrist, East London NHS Foundation Trust; Visiting Senior Lecturer, Department of Forensic and Neurodevelopmental Sciences (FANS), Institute of Psychiatry, King's College London; & Vice Chair, Faculty of Psychiatry of Intellectual Disability, Royal College of Psychiatrists
09:05-09:30	**Keynote 1** ***Assessment of Intellectual Disability in Adulthood - Updated Guidance*** Dr Theresa Joyce, Consultant Clinical Psychologist, Faculty of Intellectual Disabilities, British Psychological Society
09:35-10:00	**Keynote 2** ***Challenging Behaviour - A Unified Approach*** Dr Alick Bush, Consultant Clinical Psychologist, St Andrew's Healthcare & Chair of Learning Disabilities Professional Senate.
10:05-10:30	**Keynote 3** ***Intellectual Disabilities and International Human Rights – The New Challenges*** Professor Genevra Richardson, Chair of Law, Kings College London
10:35-11:00	**Keynote 4** ***Facial Gestalt Information from Ordinary Photos – Algorithm to Aid Diagnoses of Rare Diseases*** Dr Christoffer Nellåker, MRC Functional Genomics Unit, University of Oxford
11:05-11:25	**Refreshments and exhibition viewing and poster viewing**
11:30-13:00	**5 Concurrent Sessions**
	Session 1 – Forensic Issues in Intellectual Disabilities ***Where are we now? Services for people with intellectual disability who offend*** Dr Glyn Jones, Consultant Psychiatrist, Abertawe Bro Morgannwg University Health Board & Honorary Senior Lecturer, Department of Psychological Medicine & Neurology, Cardiff University

	"Distressing", "Frustrating" & "Utterly Hopeless": The legislation-clinical interface on inpatient services for people with learning disability and mental health or behavioural problems Dr Regi T Alexander, Consultant Psychiatrist, PiC LD Services, Norfolk & Honorary Senior Lecturer, Norwich Medical School, University of East Anglia
13:00-13:55	**Lunch and exhibition / poster viewing**
14:00-15:30	**Keynote Sessions** **Chair** - Dr Alick Bush, Consultant Clinical Psychologist, St Andrew's Healthcare; & Chair of Learning Disabilities Professional Senate
14.00-14.25	**Keynote 5** ***Behaviour that challenges: Evidence and action*** Prof Glynis Murphy, Co-Director and Professor of Clinical Psychology & Disability, The Tizard Centre, University of Kent
14.30-14.55	**Keynote 6** ***Proposals for the classification of disorders of intellectual development in ICD-11*** Dr Geoffrey Reed, Senior Project Officer, Revision of ICD-10 Mental and Behavioural Disorders, Department of Mental Health and Substance Abuse (MER/MSD), World Health Organization, Geneva
15.00-15.25	**Keynote 7** ***Psychotherapy and people who have intellectual disabilities*** Professor Nigel Beail, South West Yorkshire Partnership NHS FT & University of Sheffield
15:30-15:55	**Afternoon refreshments and exhibition viewing and poster viewing**
16:00 – 17:00	**5 Concurrent Sessions**
	Session1 – Intellectual Disabilities and Autism Spectrum Disorder ***Intensive interaction for people with intellectual disability and autism spectrum disorders*** Phoebe Caldwell, Practitioner in Intensive Interaction, London & Dr Reza Kiani, Consultant Psychiatrist, Leicestershire Partnership Trust
	Session 2 - Psychiatry of Intellectual Disability Curriculum based session ***Career in intellectual disability psychiatry film, 'Come and Work With Us Because We're Worth It' - attend this workshop to promote your speciality and to participate in research*** Dr Rupali Acharya, ST6 Psychiatry of Intellectual Disability, North Western Deanery; & Dr Ian Harwood, Consultant Psychiatrist, St Andrews Hospital, Northampton ***How to build your teaching portfolio*** Dr Zed Ahmed, Training Programme Director, Wales & Dr Geraldine Cassidy Training Programme Director, West Midlands
	Session 3 - Outcome measurement ***Building routine outcome measurement into psychological therapies for people with intellectual disability: Using the best available measure*** Dr Allan Skelly, Consultant Clinical Psychologist, Northumberland, Tyne & Wear NHS Foundation Trust

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REF: **RLW/HES/conference**

26th September 2014

Dr Reza Kiani
Mansion House
Leicester Frith Hospital Site
Groby Road
Leicester
LE3 9QF

Dear Reza

RE: ID Low Secure Network Conference

I am writing on behalf the ID Low Secure Network to thank you for speaking at the recent conference held on 19th September 2014. The conference was a great success and feedback thus far has been positive. We really do appreciate your continuing support of the network and are hopeful to do it all over again next year.

With Best Wishes
Yours sincerely

Richard Welfare

Dr R Lansdall-Welfare
Consultant Psychiatrist/Clinical Director
Intellectual and Developmental Disabilities Service

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SUNMINDS

TRAINING, EDUCATION AND DEVELOPMENT CENTRE

Sub: Invitation letter

Dear Dr. A Ashtarikiani,

Greetings from SUNMINDS Training, Education & Development Centre. SUNMINDS Training, Education & Development Centre and TDH Morris Centre, KEM Hospital, Pune in association with International Links, Royal College of Psychiatrists, U.K. present CME on "Evidence – Based Practice in Childhood Disorders".

It is with great pleasure that we extend an invitation to you and Prof. Bhaumik to be a part of this event and address the participants. Your topic will be "*Different Applications of Sensory Integration*". The program is scheduled on Sunday, 2nd December 2012. *You are required to give a brief presentation for 30 minutes only (12.00 p.m. – 12.30 p.m.) on the above topic.*

The panel discussion will be from 3.30 p.m. to 4.00 p.m. on the same day. Kindly confirm your attendance for the panel discussion.

We are happy to sponsor your accommodation for 1st of December 2012.

We would also like to mention that Maharashtra Medical Council has allocated 2 credit hours to Delegates for this activity.

Your presentation on the above mentioned topic will be extremely beneficial to the participants and it will be an enriching experience for them as well.

Thank you.

Yours Sincerely,

M. S. Tavawalla

Mrs.Masarrat Tavawalla

Dr. Sanyogita Nadkarni

Registered Address
Sunderji's Institutions
Bldg. No. 8, Vrindavan Society, Behind Mira Society, Shankarseth Road, Pune - 411 037.
Tel.: +91 20 2645 1425 / 6611 Helpline: 09096997373
Website: www.sunminds.net

Leicestershire Partnership NHS
NHS Trust

This is to certify that

_____

Attended one day

Intensive Interaction

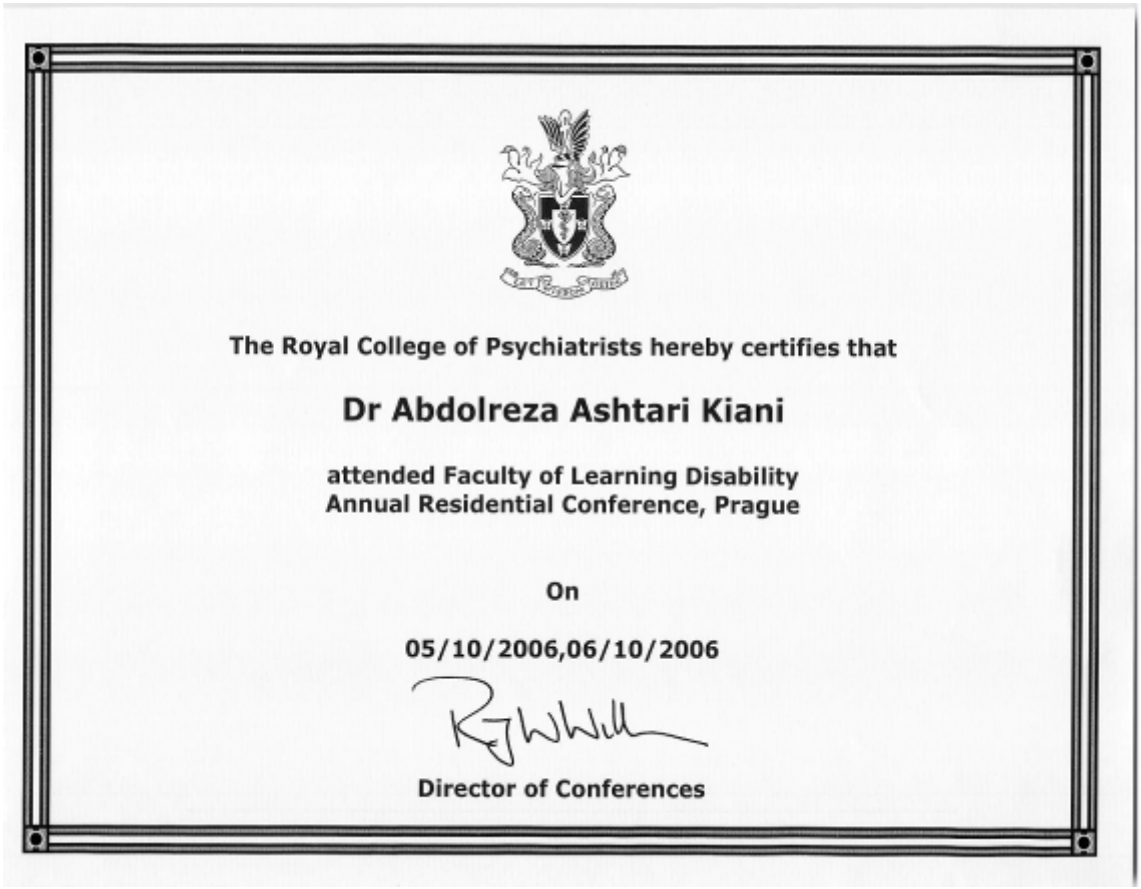
With Dr Phoebe Caldwell

On Friday, 27th April 2012
At Leicester Frith Hospital

This one day training event offers 6 CPD points

Phoebe Caldwell
Dr P Caldwell

Dr R Kiani



The Royal College of Psychiatrists hereby certifies that

Dr Abdolreza Ashtari Kiani

attended Faculty of Learning Disability
Annual Residential Conference, Prague

On

05/10/2006,06/10/2006

Director of Conferences

NHS

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24 November 2010

Dr R Kiani
Consultant in Learning Disability
Mansion House
Leicester Frith Hospital
Groby Road
Leicester
LE3 9QF

Dear Dr Kiani

Therapeutics Symposium at the Holiday Inn, Leicester, on 29th October 2010

I am writing to thank you for your presentation at our Therapeutics Symposium held at the Holiday Inn, Leicester, on 29th October 2010.

The feedback from the delegates was very positive, and the workshop that you ran was particularly appreciated.

Once again, thank you for making it a successful day.

Yours sincerely

Dr S Kumar
Consultant Psychiatrist
and Clinical Director

Dr J Devapriam
Consultant Psychiatrist

NHS

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Our Ref: SB/MEM

9th November 2009

Dr R Kiani
Consultant in Learning Disability
Mansion House
Leicester Frith Hospital
Groby Road
Leicester
LE3 9QF

Dear Reza

Re: Therapeutics Symposium at The Holiday Inn, Leicester, on 5th Nov 2009

I am writing to thank you for your participation in the workshops held at our conference last Thursday at The Holiday Inn, Leicester.

As you may know, the feedback from the delegates was very positive and encouraging, and your contribution towards making it a successful day is very much appreciated.

Yours sincerely

Dr S Bhaumik
Consultant Psychiatrist
and Lead Clinician

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NHS Trust

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Our Ref: SB/MEM

Dr R Kiani
Specialist Registrar
Mansion House
Leicester Frith Hospital

12th November 2008

Dear Reza

Re: Therapeutics Symposium at The Holiday Inn, Leicester, on 7th Nov 2008

I am writing to thank you for your participation in the workshops held at our conference held last Friday at The Holiday Inn, Leicester.

As you may know, the feedback from the delegates was very encouraging, and your contribution towards making it a successful day is very much appreciated.

Once again, thank you for your help in making it a successful symposium.

Yours sincerely

pp Ray Bhaumik

Dr S Bhaumik
Consultant Psychiatrist
and Lead Clinician

RC PSYCH
ROYAL COLLEGE OF PSYCHIATRISTS

23rd October 2008

Dr R Kiani,
Leicester Frith Hospital
Groby Road
LEICESTER LE3 9QF

Dear Dr Kiani,

The RCPsych Faculty of Learning Disability Annual Conference
Manchester 2nd – 3rd October 2008

Many thanks for your presentation which was very well received.
We have had excellent feedback from the delegates.

Yours sincerely

Dr Asit B Biswas
Academic Secretary to the LD Faculty

Royal College of Psychiatrists registered office: 17 Belgrave Square, London, SW1X 8PG, UK Tel: +44 (0)20 7235 2351 Fax: +44 (0)20 7245 1231
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Our Ref: SB/MEM

Dr R Kiani
Specialist Registrar
Leicestershire Partnership NHS Trust
Mansion House
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LE3 9QF

12th November 2007

Dear Reza

Re: Therapeutics Symposium at the Marriott Hotel, Leicester on 9th November 2007

I am writing to thank you for your presentation at our conference, which was held last Friday at the Leicester Marriott Hotel.

Feedback from the delegates was very positive, and the workshop that you carried out with Dr Gillott and Dr Barrett was appreciated by all who attended.

Thank you once again for helping us stage a successful symposium.

Yours sincerely

Dr S Bhaumik
Consultant Psychiatrist
and Lead Clinician

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