

**A.**

[illegible]

# B.

<i>GSD Ia severe</i> <i>Time-averaged Glucose Ra</i>	002_test 1	002_test 2	002_test 3	004_test 1	004_test 2	004_test 3	006_test 1	006_test 2	006_test 3	007_test 1	007_test 2	007_test 3
C1 (mM)	0.942142534	0.379372825	0.375897135	78.14014583	0.548961738	0.133179426	0.473321303	107.4064496	0.586824303	0.836295341	0.469393433	93.82053799
ka (min-1)	0.045269584	0.103157252	2.051897921	0.018072721	0.104494515	0.501906048	0.110396326	0.017628822	0.150506871	0.06602706	0.054598353	0.033810441
k2 (min-1)	0.026442194	0.018970588	0.023404117	0.017910074	0.023818256	0.007162696	0.010012328	0.01755021	0.0134756	0.033367113	0.016991877	0.033666605
td (min)	0.767329344	0.077047606	0.001977448	3.928687917	0.127391273	8.64334E-06	0.002506083	1.921500895	0.039489981	0.047369022	-0.025668726	1.410836491
Vol (mL kg-1)	176.7041173	223.6336392	186.3201972	136.7911666	226.9657663	732.7570558	197.6429719	177.6026865	159.2120175	139.2103737	178.116721	144.2813779
body weight (kg)	86	86	86	61.9	61.9	61.9	70	70	70	103.4	103.4	103.4
<i>GSD Ia severe</i> <i>Time course Glucose Ra</i>	002_test 1	004_test 1	006_test 1	007_test 1								
C1 (mM)	0.942142534	78.14014583	0.473321303	0.57237996								
ka (min-1)	0.045269584	0.018072721	0.110396326	0.078135905								
k2 (min-1)	0.026442194	0.017910074	0.010012328	0.028342063								
td (min)	0.767329344	3.928687917	0.002506083	0.062651456								
Vol (mL kg-1)	176.7041173	136.7911666	197.6429719	157.8757346								
body weight (kg)	86	61.9	70	103.4								

C.

<i>HV Time-averaged Glucose Ra</i>	003_test 1	003_test 2	003_test 3	005_test 1	005_test 2	005_test 3	008_test 1	008_test 2	008_test 3	010_test 1	010_test 2	010_test 3	011_test 1	011_test 2	011_test 3
C1 (mM)	0.574253639	0.902200768	0.619603441	0.560064383	0.384881378	0.64571005	0.20028235	68.43815824	86.21381069	0.39515158	0.615164156	178.4172943	0.574414988	12.01539019	0.134359808
ka (min-1)	0.17166814	0.035053676	0.086930346	0.231144846	0.224434654	0.22466467	0.117122116	0.063009348	0.031864263	0.051869973	0.04952357	0.019168891	0.243005358	0.0250337	0.083418799
k2 (min-1)	0.013737153	0.017933522	0.011944118	0.012813696	0.009725764	0.015767903	0.002261733	0.062793737	0.031700078	0.004892927	0.011513069	0.01907678	0.015932763	0.024404457	0.013313819
td (min)	0.040204376	-0.902689933	0.056190921	0.020583557	-0.087487542	-0.040627884	-0.16104617	0.254664693	1.165175103	-0.481114101	0.194005338	0.789757721	0.007919474	1.014358093	0.085956663
Vol (mL kg-1)	173.4005912	207.9000953	171.3989109	175.8716825	252.6843537	154.9640217	293.4749165	246.1397453	129.7592147	300.8754828	228.0539142	125.5929465	156.2472387	277.6874078	742.7329269
body weight (kg)	65	65	65	64	64	64	103.3	103.3	103.3	55.3	55.3	55.3	71	71	71
<i>HV Time-averaged Glucose Ra</i>	012_test 1	012_test 2	012_test 3	013_test 1	013_test 2	013_test 3	016_test 1	016_test 2	016_test 3	018_test 1	018_test 2	018_test 3	019_test 1	019_test 2	019_test 3
C1 (mM)	0.380033042	0.110181291	N.A.	0.577967935	0.276457321	97.28531223	0.39841741	0.325831758	0.38282549	0.404836047	0.623060848	0.411522738	53.62174217	-0.07627287	75.74439082
ka (min-1)	0.21612513	0.116736396	N.A.	2.040834883	0.08295424	0.026738162	2.055218046	0.247130111	0.077445831	0.127353101	0.086265192	0.136221914	0.019813355	-0.0004912	0.018161593
k2 (min-1)	0.013944621	0.004175088	N.A.	0.011759537	0.011421836	0.026460179	0.013413316	0.0169694	0.0083158	0.011211044	0.02913688	0.011699934	0.019599263	0.113523119	0.018066633
td (min)	0.010259889	-0.06575811	N.A.	0.003771977	0.10438508	2.118596607	0.00491664	0.004795889	0.506114797	0.123507925	0.002700815	0.117091526	1.023684846	-1.61857073	1.766207271
Vol (mL kg-1)	183.4506193	613.8805207	N.A.	164.4791059	396.4719451	93.44787568	232.5125754	303.2788454	269.3217964	175.688562	157.2013949	172.4287894	125.1755327	-4.09667405	183.1332051
body weight (kg)	91.3	91.3	N.A.	63	63	63	64.7	64.7	64.7	91.8	91.8	91.8	82.1	82.1	82.1
<i>HV Time course Glucose Ra</i>	003_test 1	005_test 1	008_test 1	010_test 1	011_test 1	012_test 1	013_test 1	016_test 1	018_test 1	019_test 1					
C1 (mM)	0.574253639	0.560064383	0.20028235	0.39515158	0.574414988	0.380033042	0.577967935	0.39841741	0.404836047	53.62174217					
ka (min-1)	0.17166814	0.231144846	0.117122116	0.051869973	0.243005358	0.21612513	2.040834883	2.055218046	0.127353101	0.019813355					
k2 (min-1)	0.013737153	0.012813696	0.002261733	0.004892927	0.015932763	0.013944621	0.011759537	0.013413316	0.011211044	0.019599263					
td (min)	0.040204376	0.020583557	-0.16104617	-0.481114101	0.007919474	0.010259889	0.003771977	0.00491664	0.123507925	1.023684846					
Vol (mL kg-1)	173.4005912	175.8716825	293.4749165	300.8754828	156.2472387	183.4506193	164.4791059	232.5125754	175.688562	125.1755327					
body weight (kg)	65	64	103.3	55.3	71	91.3	63	64.7	91.8	82.1					

**Supplemental file 3. Calculated model parameters in the study participants.** Data in italics were excluded from calculations. (A) GSDIa patients with attenuated phenotype. (B) GSDIa patients with a severe phenotype. (C) Healthy volunteers.

