**Title page**

**Platelets compensate for poor thrombin generation in type 3 von Willebrand disease**

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
|  | Peak thrombin in PPP | ETP in PPP | Peak thrombin in PRP | ETP in PRP |
| FVIII | **r=0.77**  **p=0.023** | r=0.33  p=0.38 | r=0.69  p=0.19 | r=0.60  p=0.19 |
| VWF:GPIbM | r=0.53  p=0.19 | r=0.03  p=0.96 | r=0.37  p=0.19 | r=0.37  p= 0.41 |
| VWF:Ag | r=0.50  p=0.17 | r=0.16  p=.69 | r=0.69  p=0.09 | r=0.66  p=0.48 |
| VWF:CB | **r=0.81**  **p=0.016** | r=0.48  p=0.19 | r=0.55  p=0.20 | p=0.65  p=0.23 |
| ADAMTS-13 | r=0.55  p=0.13 | r=0.37  p=0.37 | r=0.66  p=0.059 | r=0.68  p=0.11 |
| TFPI total/free | r=-0.40/-0.083  p=0.29 / 0.84 | r=-0.77/-0.083  p=0.067/0.071 | r=-0.071/-0.30  p=0.31/0.91 | r=-0.38/-0.05  p= p=0.39/0.91 |
| PS | r=0.27  p=0.49 | r=0.48  p=0.19 | r=0.13  p=0.74 | r=0.43  p=0.35 |
| F1+2 | r=-0.53  p=0.15 | r=-0.62  p=0.086 | r=-0.63  p=0.076 | r=-0.32  p=0.49 |
| P-selectin % (ADP 1 µM) | r=-0.45  p=0.26 | r=-0.52  p=0.27 | r=-0.29  p=0.50 | r=-0.31  p=0.56 |
| P-selectin % (ADP 2 µM) | r=-0.31  p=0.46 | r=-0.52  p=0.46 | r=-0.29  p=0.88 | r=-0.31  p=0.91 |
| P-selectin % (ADP 5 µM) | r=-0.33  p=0.43 | r=-0.33  p=0.43 | r=-0.14  p=0.75 | r=-0.089  p=0.91 |
| Annexin V % (CRP 0.5 mg/mL) | r=-0.43  p=0.30 | r=-0.21  p=0.61 | r=-0.24  p=0.58 | r=-0.26  p=0.66 |
| Annexin V % (CRP 7.5 mg/mL) | r=-0.41  p=0.33 | r=-0.29  p=0.50 | r=-0.17  p=0.70 | r=-0.20  p=0.71 |
| Annexin V % (CRP 15 mg/mL) | r=-0.41  p=0.33 | r=-0.29  p=0.50 | r=-0.17  p=0.70 | r=-0.20  p=0.71 |
| BAT | r=-0.18  p=0.65 | r=-0.38  p=0.32 | r=-0.05  p=0.90 | r=0.25  p=0.59 |

**Supplementary Table 1.** TG parameters peak thrombin and ETP in association to FVIII,VWF, ADAMTS-13, TFPI (free and total), PS, F1+2, flow cytometry analysis and BAT by Spearman’s correlation. Only peak thrombin in PPP showed correlation to FVII and VWF:CB. None of the TG parameters showed association to ADAMTS-13, TFPI or platelet activation status in flow cytometry.

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|  | **EXTEM**  **CT**  38-79 s | **EXTEM**  **CFT** 34-159 s | **EXTEM**  α 63-83 ° | **EXTEM**  **MCF** 50-72 mm | **INTEM**  **CT** 100-240 s | **INTEM**  **CFT** 30-110 s | **INTEM**  **α**  70-83 ° | **INTEM**  **MCF** 50-72 mm | **FIBTEM**  **MCF** 9-25 mm |
| All  n=9 | **49**  44-52 | **69**  51-106 | **79**  71-85 | **68**  62-72 | **186**  162-207 | **65**  41-105 | **77**  70-81 | **70**  61-73 | **21**  12-27 |
| PRE  n=6 | **49**  47-52 | **69**  51-106 | **78**  71-82 | **69**  62-72 | **184**  172-207 | **68**  41-105 | **76**  70-81 | **69**  61-73 | **20**  12-27 |
| POST  n=6 | **48**  43-62 | **84**  51-136 | **78**  64-83 | **68**  62-72 | **190**  160-235 | **77**  65-81 | **75**  71-80 | **67**  63-73 | **19**  13-26 |

**Supplementary Table 2.** Thromboelastometry (ROTEM) (median and range) in all patients, also given before and after treatment. No significant change was detected between pre and post samples in any parameter (p>0.05). Reference ranges provided by the manufacturer given in the top column.