

time [days]

- 100

50

10%

100

50

10%

% activity

40 50,00,00,00

30

% activity

9

700-

500

300

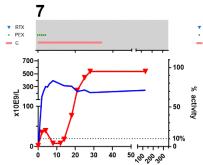
10

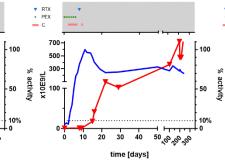
0<del>4</del> 0

10 20

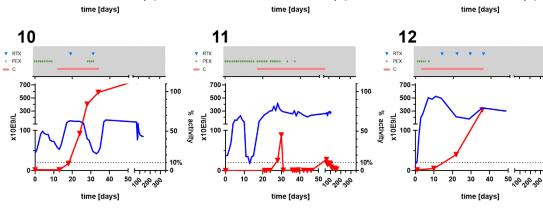
RTX
PEX
C

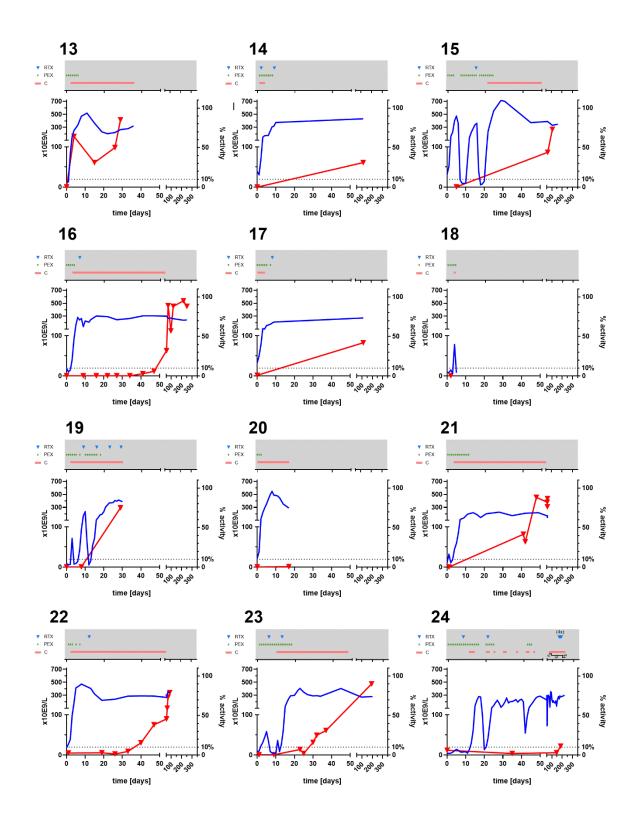
x10E9/L

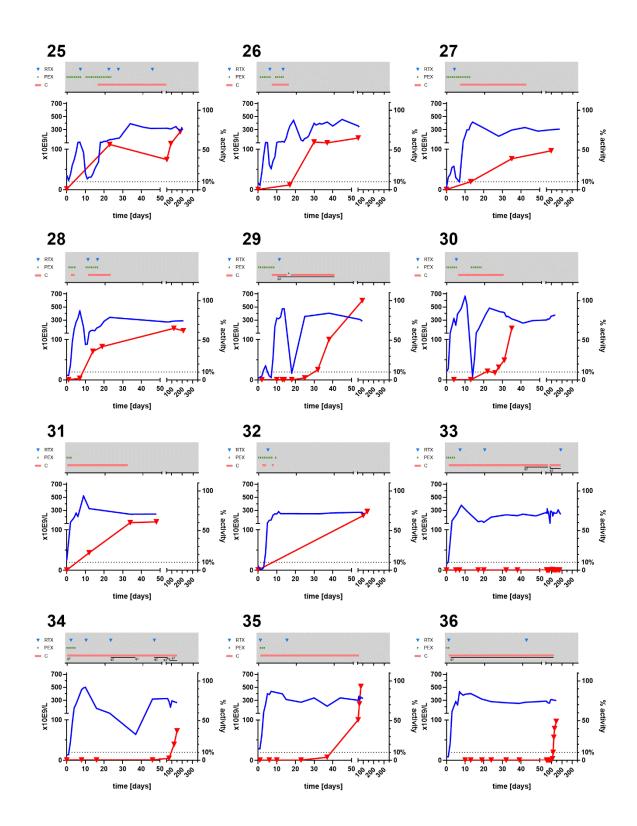


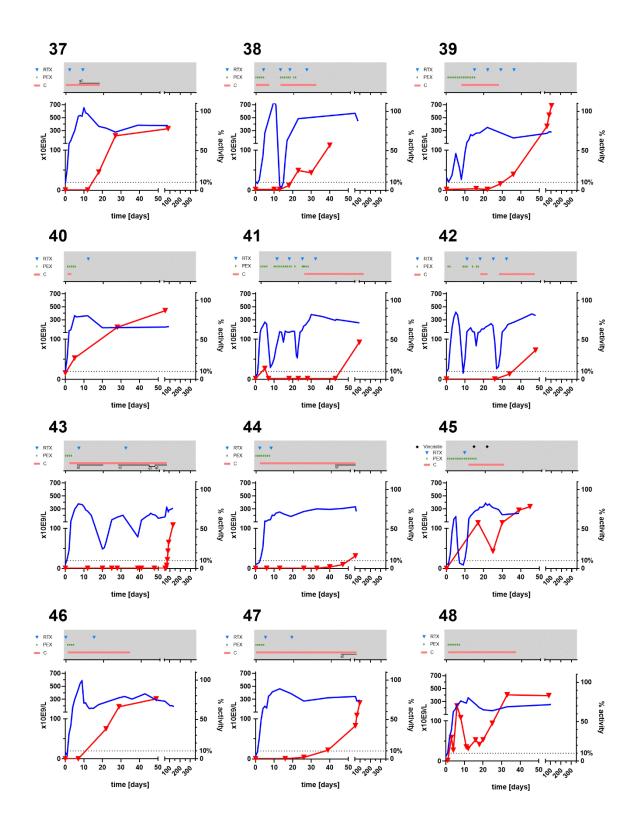


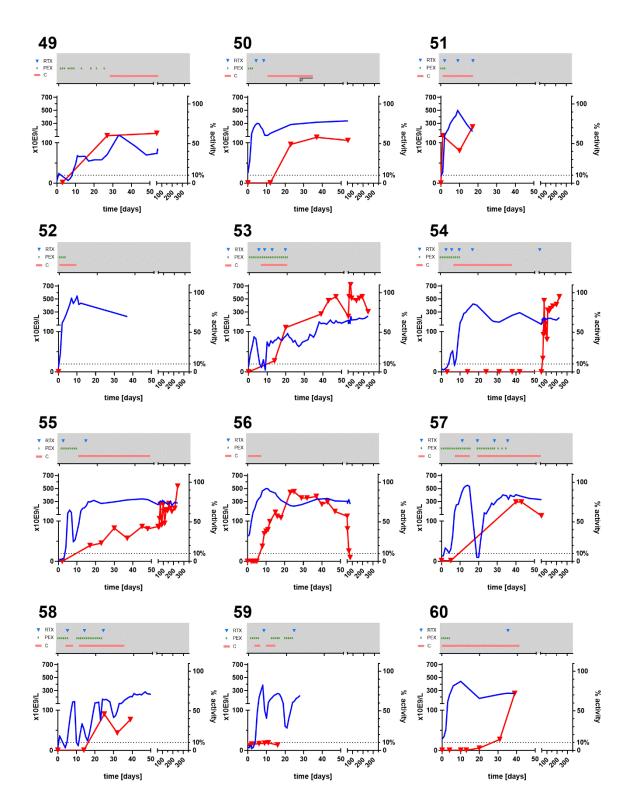
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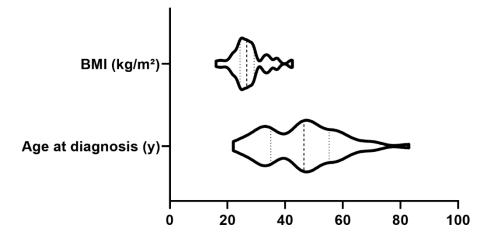








**Supplemental Figure 1** - Clinical courses of individual patients and treatment modalities. Day zero indicates day of diagnosis (see Methods). All patients except one (#21) were treated with variable doses of glucocorticoids. For purposes of clarity, glucocorticoid therapies were omitted from the diagrams. RTX – rituximab treatment. PEX – days of PEX treatment. For simplicity, specific doses of RTX or PEX are not indicated. C – caplacizumab. Black line indicates dosing intervals (i.e. no line – daily, q2 – alternate day, q3 – every third day, q4, every fourth day, q7 – once weekly). Thrombocytes are indicated by the blue line, ADAMTS13 activity is depicted in red triangles with connecting line. Dotted line marks ADAMTS13 activity cut-off of 10%. Star (\*) in clinical course #29 indicates missed caplacizumab dose.



**Supplemental Figure 2** - Violin blots depict the distribution of BMI and age diagnosis. Red lines indicate medians, blue lines indicate 75<sup>th</sup> and 25<sup>th</sup> interpercental quartile.