Supplementary Figures to Jilma-Stohlwetz et al. “A dose ranging phase I/II trial of the von Willebrand Factor (VWF) inhibiting aptamer ARC1779 in patients with congenital thrombotic thrombocytopenic purpura (TTP)” (Thromb Haemost 2011; 106.3)

Online Suppl. Figure S1: Cyclic variations in platelet counts in a patient (F2) with congenital thrombotic thrombocytopenic purpura before and after plasma transfusion. The patient received plasma transfusions approximately every three weeks (vertical arrows). Each cross represents one blood sample.
Online Suppl. Figure S2: Excessive shear induced platelet function occurs even after plasma transfusion. Patient F2 received three units of S/D plasma during the period of sc. ARC1779 injection. Collagen adenosine diphosphate induced closure time (open stars) already normalised, when platelet counts (closed symbols) reached >50x10⁹/L and were pathologically short (the lower dashed line indicates the lower limit of the normal range for CADP-CT) when platelet counts exceeded >150x10⁹/L, i.e. the lower limit of the normal range.
Online Suppl. Figure S3: Shear induced platelet hyper-function in congenital thrombotic thrombocytopenic purpura. Despite moderate thrombocytopenia two out of three patients had collagen adenosine diphosphatse induced closure times (CADP-CT) within the normal range (indicated by dashed horizontal lines) at baseline, increased to a maximum of 300s during high dose ARC1779 infusion when platelet counts increased, and were pathologically shortened one week after plasma transfusion when platelet counts were still in the normal range. This reflects excessive von Willebrand activity between consecutive plasma transfusions. Normal ranges of CADP-CT are between dashed lines. Patient F1 (open circles) Patient F2 (black stars), patient M (black squares).
Online Suppl. Figure S4: Effects of ARC1779 on platelet counts in congenital thrombotic thrombocytopenic purpura. The male patient (M) received the anti von Willebrand factor aptamer ARC1779 intravenously at a high dose (4 mg/kg/min). Platelet counts were stable during ARC1779 infusion but dropped after discontinuation of the infusion. Transfusion of plasma produced the expected increase in platelet counts.