

The ADAMTS13-VWF axis is dysregulated in chronic thromboembolic pulmonary hypertension.

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Figures

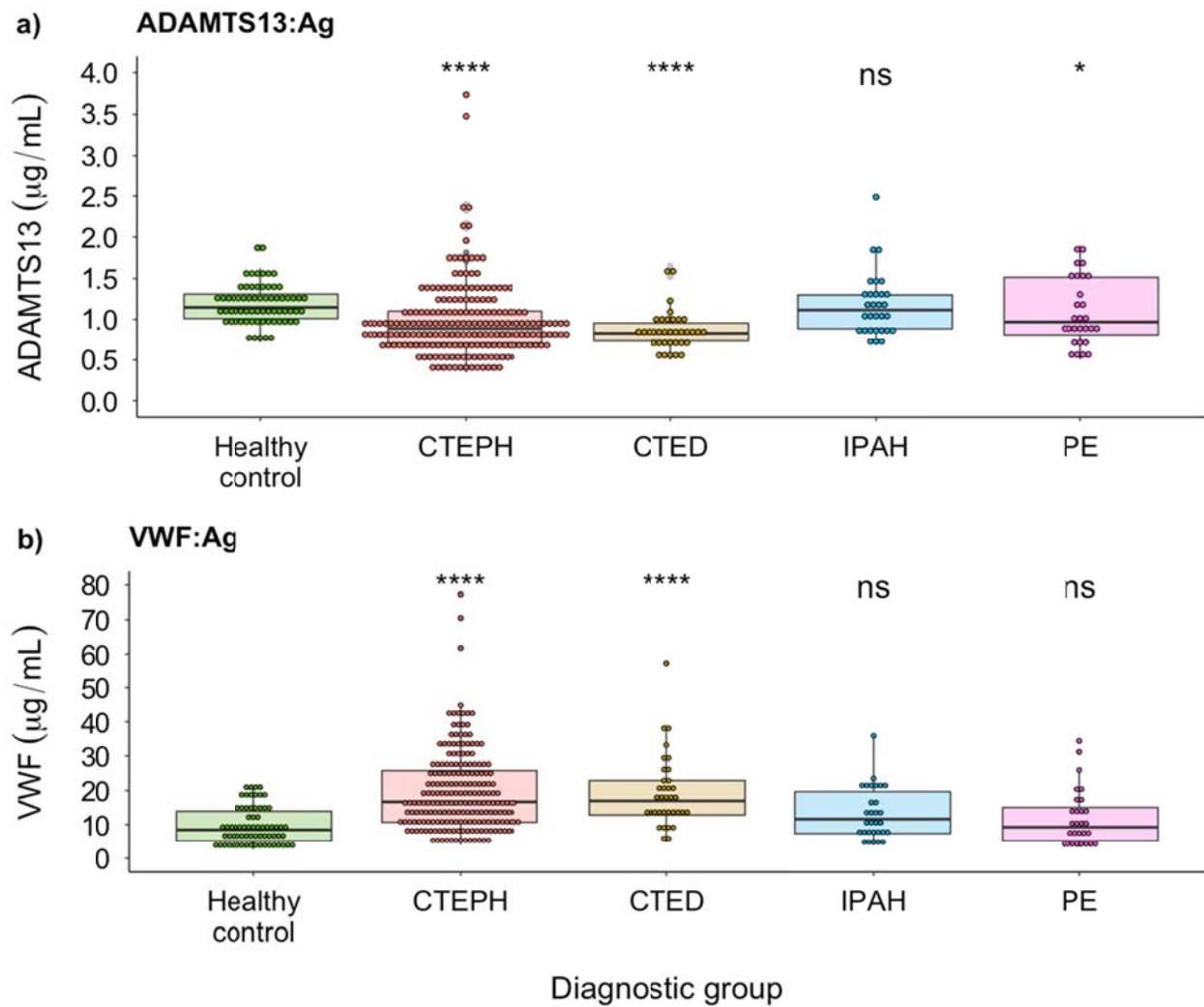


FIGURE 1

a) ADAMTS13 and b) VWF antigen (Ag) levels by diagnostic groups. Dunn's test with false discovery rate (FDR) adjustment was used to calculate p -values. ****: $p \leq 0.0001$, ***: $p \leq 0.001$, **: $p \leq 0.01$, *: $p \leq 0.05$, ns (not significant): $p > 0.05$. Healthy control (n=68), CTEPH (chronic thromboembolic pulmonary hypertension, n=208), CTED (chronic thromboembolic disease, n=35), IPAH (idiopathic pulmonary arterial hypertension, n=28), PE (pulmonary embolism, n=28).

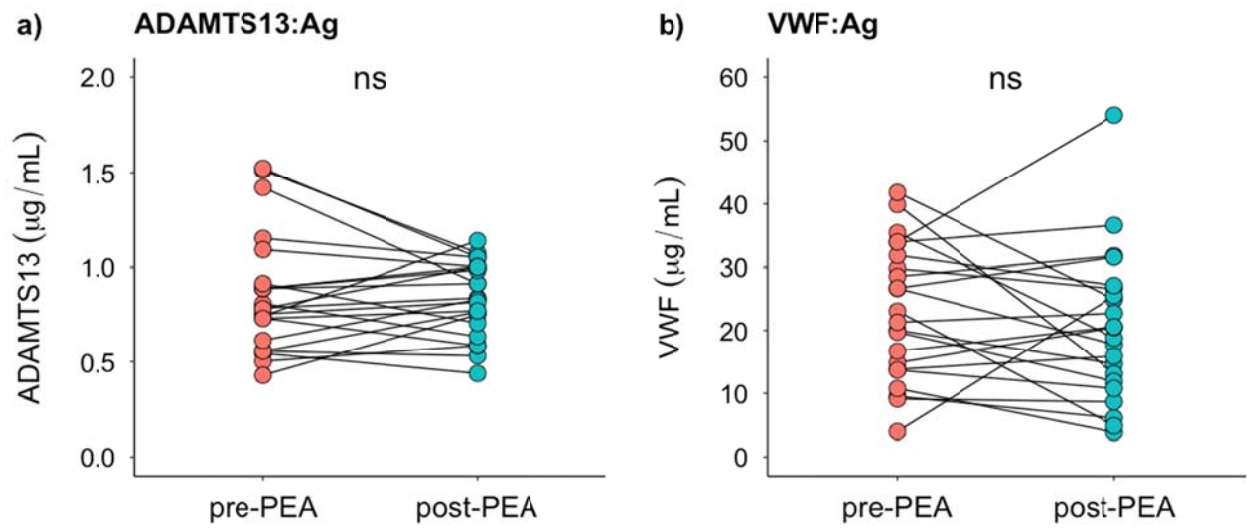


FIGURE 2

a) ADAMTS13 and b) VWF antigen levels pre- and post-pulmonary endarterectomy (n=22). Wilcoxon signed-rank test was used to calculate p -values.

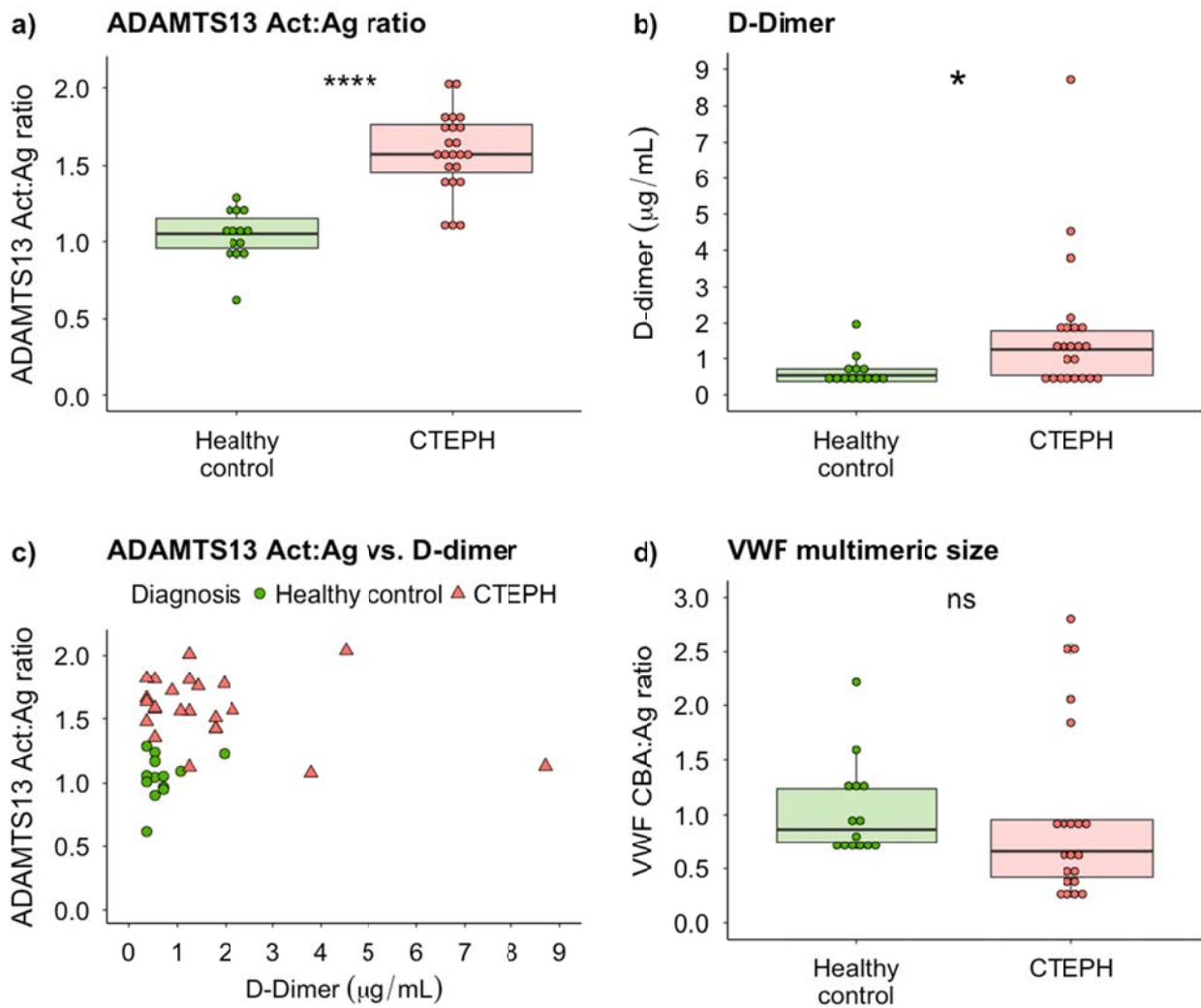


FIGURE 3

ADAMTS13 activity, D-dimer and VWF multimeric size in CTEPH and healthy controls. A subset of CTEPH patients ($n=23$) with the lowest ADAMTS13 antigen levels (below the first quartile of the CTEPH group) and healthy controls ($n=14$) were used for a-c). VWF multimeric size was measured in CTEPH ($n=21$) samples with the highest VWF antigen concentrations (above the third quartile of the CTEPH group) using the same healthy control subset and displayed in d). The Mann-Whitney U test was used to calculate group differences (a, b, d) and correlation was assessed with Spearman's rank correlation coefficients (c). A. Specific ADAMTS13 activity (Act:Ag ratio). b) D-dimer antigen levels. c) Specific ADAMTS13 activity and D-dimer antigen correlation. Healthy control correlation: $\rho=0.0938$, $p=0.761$;

CTEPH correlation: $\rho=-0.220$, $p=0.313$. d) VWF multimeric size (VWF Collagen binding assay:Antigen ratio).

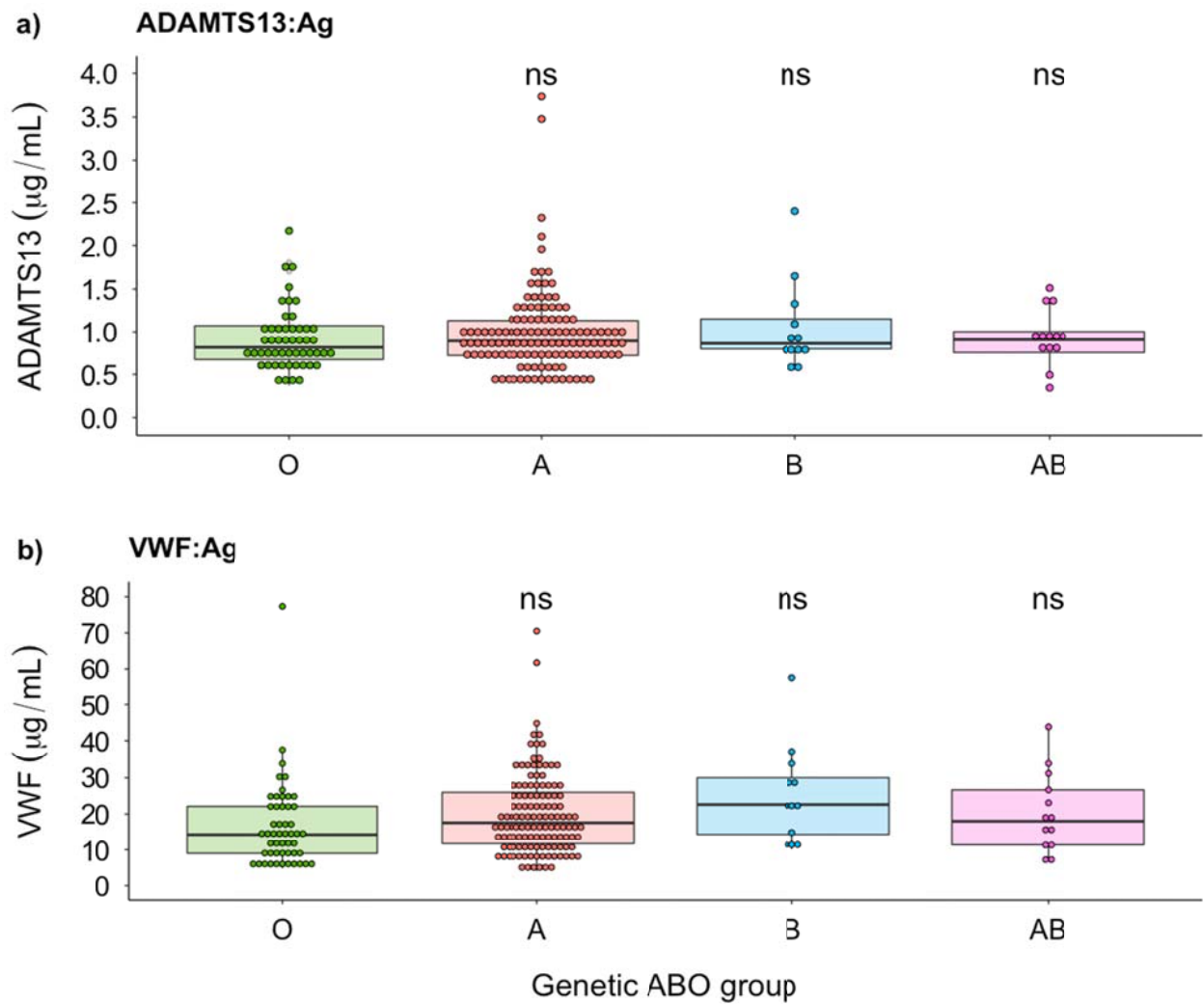


FIGURE 4

ADAMTS13 and VWF antigen levels by *ABO* genetic groups. CTEPH (n=182) and CTED (n=22) patients with genotypes and protein levels available (in n=3 a genetic *ABO* group could not be inferred) were included. Dunn's test with FDR adjustment was used to calculate *p*-values. Numbers in each group: O = 51, A = 128, B = 12, AB = 13.