

Disordini emorragici congeniti: aspetti diagnostici e clinici

PERIOPERATIVE BLEEDING IN CHILDREN WITH LOW VON WILLEBRAND FACTOR UNDERGOING ENT SURGERY: A 15-YEAR SINGLE-CENTRE EXPERIENCE

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Background and aims. Low von Willebrand factor is a borderline haemostatic condition with uncertain perioperative significance in children. ENT surgery represents a clinically relevant model, as bleeding risk may reflect both systemic haemostatic impairment and local surgical and fibrinolytic mechanisms. We aimed to evaluate the incidence, timing and predictors of perioperative bleeding in children with low VWF undergoing ENT surgery, and to compare outcomes with a large cohort of healthy paediatric controls. We conducted a retrospective single-centre study including 120 children with low VWF undergoing ENT surgery and 1,136 healthy paediatric controls.

Methods. Cohorts were derived from institutional databases collected over partially non-overlapping time periods.Collected variables included age, sex, type of surgery, personal and family bleeding history, ISTH-BAT score (available in a subset), VWF:RCo, VWF:Ag, FVIII, aPTT ratio and platelet function analyser (PFA) closure times.The primary outcome was intra-/post-operative bleeding, classified as early (0–48 h), late (G7–13) or not defined. Continuous variables were analysed using the Mann–Whitney U test and categorical variables using Fisher's exact test.

Results. The low VWF cohort (n=120) was comparable to controls for age and sex. Baseline clinical bleeding features were more prevalent in low VWF patients (personal bleeding 17.5% vs 3.9%, p<0.001; family history 17.5% vs 1.1%, p<0.001). Perioperative bleeding occurred in 25.0% of low VWF patients and 28.3% of controls (p=0.521). Major bleeding occurred in 5.8% vs 3.0% (p=0.103). Among classified events, major bleeding was more frequent in low VWF patients (26.9% vs 10.6%, p=0.023). Within the low VWF cohort, no baseline clinical or laboratory parameter predicted bleeding, including age, sex, type of surgery, personal bleeding history, family history, ISTH-BAT score, VWF:RCo, VWF:Ag, FVIII and PFA closure times (all p>0.1). The only parameter associated with bleeding was a lower aPTT ratio in bleeders (1.09 vs 1.15; p=0.014), with a stronger association observed in major bleeding, although its direction is not biologically intuitive. Bleeding events showed a bimodal temporal distribution: 70.0% occurred within 48 h, 23.3% between G7–13 and 6.7% were not defined.

Conclusions. Children with low VWF undergoing ENT surgery display a clinically relevant rate of perioperative bleeding; however, overall bleeding rates are not significantly different from those observed in healthy paediatric controls. Despite a higher prevalence of baseline bleeding phenotype, no clinical or laboratory parameter reliably predicted surgical bleeding, suggesting a multifactorial mechanism not captured by standard haemostatic assessment. A shift towards more severe bleeding among classified events and a bimodal temporal pattern support the coexistence of systemic and local mechanisms. Differences in cohort collection periods and potential heterogeneity in bleeding reporting should be considered when interpreting comparisons.

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