

Bleeding risks associated with tonsillectomy in patients with Ehlers-Danlos syndrome

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Introduction: Ehlers-Danlos syndrome (EDS) is an inherited collagen disorder that, based on the subtype, comes with varying risks of life-threatening hemorrhage. The risk of post-operative adenotonsillectomy (T&A) hemorrhage should be scrutinized in patients with EDS. Better risk assessment will aid in medical optimization prior to surgery, counseling families of surgical risks, and may decrease admission costs in lower-risk EDS patients.

Objectives: Describe the post-T&A bleeding risk in patients with EDS compared to patients without EDS.

Methods: This was a retrospective study of children receiving a T&A with a diagnosis of EDS compared to a random selection out of over 25,000 patients without a diagnosis of EDS who received a T&A in the past 10 years at Children's Hospital of Colorado. Admission criteria via the clinical practice guidelines were analyzed. Bleeding risk in children with EDS was assessed by EDS subtype, prior echocardiograms, electrocardiograms, and laboratory data.

Results: Of 62 patients with EDS and 72 controls, those with EDS were more likely to be female ($p=0.033$), younger ($p=0.051$), and non-Hispanic white ($p=0.001$, $p=0.01$). Post-operative hemorrhage rates were unaffected by post-operative admission in both the controls and EDS patients ($p=0.609$). EDS patients had no differences in T&A blood loss ($p=0.669$) and no significant risk of a post-operative hemorrhage ($p=0.118$). Isolated bleeding disorders did not increase post-operative hemorrhage ($p=0.092$); however, EDS patients had more comorbid bleeding disorders ($p=0.001$). Controlling for type of EDS and comorbid bleeding risks swayed the p-value towards the null.

Conclusion: Patients with EDS had no significant increased risk of a post-T&A hemorrhage when compared to children without EDS. The combination of both EDS, prior hemorrhage events, and comorbid bleeding disorders may elevate the risk of post-T&A hemorrhage. Pre-operative optimization should be taken by a case-by-case basis. If low risk pre-operatively, patients with EDS may not pose a significant risk of hemorrhage.