Published in final edited form as:

Otolaryngol Head Neck Surg. 2010 May ; 142(5): 715–721. doi:10.1016/j.otohns.2010.01.029.

Post-tonsillectomy bleeding in children with von Willebrand disease: A single-institution experience

Kenneth D. Rodriguez, MD¹, Gordon H. Sun, MD², Francis Pike, MS³, Ellen M. Mandel, MD¹, Margaretha L. Casselbrant, MD, PhD¹, and David H. Chi, MD¹

¹University of Pittsburgh, Department of Otolaryngology-Head and Neck Surgery

²University of Cincinnati, Department of Otolaryngology- Head and Neck Surgery

³University of Pittsburgh, Department of Statistics

Abstract

Objectives—1) Compare rates of post-tonsillectomy bleeding in pediatric patients with and without von Willebrand disease (vWD). 2) Identify factors that may increase the risk for post-tonsillectomy bleeding in children with and without vWD.

Study Design—Historical cohort study.

Setting—Tertiary care, university-based pediatric hospital.

Subjects and Methods—Medical records were examined for 99 patients with vWD and 99 patients without vWD under the age of 18 years who underwent tonsillectomy with or without adenoidectomy from August 1997 to October 2005. Subjects were matched for age, year of surgery, type of surgery, and indication for surgery.

Results—Post-tonsillectomy hemorrhage occurred in 8 of 99 (8 percent) vWD patients and in 6 of 99 (6 percent) non-vWD patients ($P = 0.58$, Odds Ratio 1.36, 95% CI 0.45–4.08). A two-sample test of proportions demonstrated lower and upper limits of -0.051 and 0.092 . Four of 8 children with vWD and 2 of 6 non-vWD patients required surgical intervention for control of bleeding. Ninety-three of 99 vWD patients received desmopressin acetate (DDAVP) preoperatively. In patients with vWD who responded to DDAVP challenge, there was no increased likelihood of post-tonsillectomy bleeding compared to non-vWD patients. No significant difference in the number of bleeding events was noted based on demographics, preoperative laboratories, or use of aminocaproic acid.

Conclusions—Children with vWD undergoing tonsillectomy have a postoperative bleeding rate similar to that of a matched group. However, the sample size was not sufficient to eliminate the possibility of a clinically important difference between the two groups.

© 2010 American Academy of Otolaryngology Head and Neck Surgery Foundation, Inc. Published by Mosby, Inc. All rights reserved.

Corresponding Author: David H. Chi, MD 45th and Penn Ave Children's Hospital of Pittsburgh Pittsburgh, PA 15203. Presented in part at the American Academy of Otolaryngology Annual Meeting, Washington D.C. September 17, 2007.

Publisher's Disclaimer: This is a PDF file of an unedited manuscript that has been accepted for publication. As a service to our customers we are providing this early version of the manuscript. The manuscript will undergo copyediting, typesetting, and review of the resulting proof before it is published in its final citable form. Please note that during the production process errors may be discovered which could affect the content, and all legal disclaimers that apply to the journal pertain.

Introduction

von Willebrand disease (vWD) is the most common inherited bleeding disorder in Americans and Europeans, with a prevalence of 0.8 percent to 2 percent in the general population.^{1,2} The source of the coagulopathy is von Willebrand factor (vWF), a multimeric glycoprotein containing roughly 2800 amino acids, which is encoded by a gene on chromosome 12 and synthesized in vascular endothelial cells and megakaryocytes.³⁻⁵

Bleeding in vWD results from either a decrease in the concentration of vWF or functionally abnormal vWF. The vWF molecule serves two primary hemostatic functions. First, vWF mediates platelet adhesion to damaged endothelium by interacting with glycoprotein Ib and IIb/IIIa receptors on platelets.^{4,5} Second, vWF binds to coagulation factor VIII, a critical component of the hemostatic pathway, preventing its inactivation and clearance from plasma.^{4,5} vWD clinically presents as epistaxis, menorrhagia, gastrointestinal bleeding, excessive bleeding following soft tissue trauma, or other mucocutaneous bleeding.^{5,6} This disorder also commonly presents with disproportionate perioperative bleeding following otherwise benign surgical procedures, such as dental work.¹ In more severe cases, bleeding may be quite profuse, not unlike moderate or severe cases of hemophilia A, and may even result in death if inappropriately managed.^{4,7}

Because vWD is almost always inherited in an autosomal-dominant pattern, a detailed preoperative workup may reveal a family history of facile bleeding or ecchymosis. Currently, the most sensitive screening test for diagnosing vWD is the platelet function assay (PFA-100), which evaluates vWF-mediated formation of a platelet plug on a collagen and either ADP- or epinephrine-coated membrane.¹ In children with a confirmed diagnosis of vWD, the sensitivity of the PFA-100 approaches 100 percent, though both false positive and false negative test results have been encountered.¹ Definitive laboratory diagnosis of vWD relies heavily on a panel of four assays: vWF antigen levels, ristocetin cofactor activity, coagulation factor VIII activity, and vWF multimer analysis.^{1,4} However, because the level of plasma vWF itself is influenced by myriad factors, including age, acute inflammation, stress, the menstrual cycle, thyroid status, and various medications, establishing a clear diagnosis of vWD remains challenging.^{1,5,8-11} In pediatric patients, simply waiting in anticipation of phlebotomy may be enough of a stressor to cause a transient increase in vWF levels.⁵

Since the first case of von Willebrand disease was reported in 1926, six primary types of vWD have been described. Seventy to 80 percent of individuals with vWD are categorized as type 1, having an autosomal dominant-inherited abnormality that results in mildly to moderately decreased levels of normally functioning vWF in the plasma.^{5,6} Type 2 vWD, which is composed of four subtypes, comprises various qualitative defects in the vWF molecule.^{4,5} Type 2 vWD accounts for 10 percent to 30 percent of all cases of vWD and is inherited in an autosomal dominant pattern.⁴ Type 3 vWD, a variant seen in 1 percent to 5 percent of all cases, is usually inherited in an autosomal recessive manner and is characterized by nearly undetectable or absent levels of plasma vWF and below-normal levels of factor VIII.⁴ Of these categories, type 3 vWD is the most likely to cause severe, even life-threatening post-tonsillectomy hemorrhage.^{4,6}

Tonsillectomy is currently indicated for a variety of disorders, including recurrent tonsillitis, peritonsillar abscess, obstructive sleep apnea or sleep-disordered breathing, and tonsillar hypertrophy.¹²⁻¹⁴ In 1996, an estimated 287,000 children under the age of 15 years underwent tonsillectomy with or without adenoidectomy.¹² The risk of delayed hemorrhage, or bleeding beyond twenty-four hours after tonsillectomy, is between 2 percent and 4 percent regardless of existing co-morbidities.¹⁵ Underlying risk factors contributing to post-

tonsillectomy bleeding may include history of recurrent tonsillitis or infectious mononucleosis, advanced age, male gender, personal or family history of coagulopathy, and surgical technique, but nonetheless all remain controversial.^{7,15,16} vWD may remain asymptomatic in many patients until a significantly traumatic event occurs, such as adenotonsillar surgery.

We present results from a historical cohort study of 99 vWD patients undergoing adenotonsillar surgery compared with 99 matched non-vWD patients at the Children's Hospital of Pittsburgh, Pittsburgh, PA. We sought to determine if children known to have vWD were more likely to have post-tonsillectomy bleeding events than matched children without vWD, and if numbers were sufficient, whether other risk factors may be contributing to bleeding in both vWD and non-vWD patients following tonsillectomy with or without adenoidectomy.

Methods and Materials

This is a historical cohort study reviewing 99 vWD and 99 non-vWD patients who underwent tonsillectomy or adenotonsillectomy from August 1997 to October 2005. This project was approved by the Institutional Review Board of the University of Pittsburgh. An attending hematologist at Children's Hospital of Pittsburgh determined the diagnosis of vWD or the children presented with the diagnosis confirmed in the medical record. Primary exclusion criteria for all patients included the use of nonsteroidal anti-inflammatory drugs (NSAIDs), oral contraceptives, or anti-coagulation medications within two weeks of surgery, or a history of other coagulopathies. Surgical indications were limited to either obstructive sleep apnea, recurrent tonsillitis, or both. All procedures were performed by Children's Hospital of Pittsburgh staff otolaryngologists. The groups were matched as closely as possible using the following variables: age at the time of the operation (children 48 months or younger were matched within 6 months and children 48 months or greater within one year), year of procedure, type of surgery (tonsillectomy or adenotonsillectomy), and indication for surgery (obstructive sleep apnea, recurrent tonsillitis, or both). Electrocautery was used exclusively for both tonsillectomy and adenoidectomy in all patients in this study. Data on 500 non-vWD patients were used in the matching process. If multiple patients were found to be identical given the above criteria, then further matching for sex or ethnicity was used.

The following data were compiled after evaluation of paper and electronic medical records: the date of operation and postoperative follow up, age at operation, sex, race, history of bleeding disorders, concurrent surgical procedures, type of primary procedure and associated procedures, surgical indication, length of procedure, surgical technique, perioperative blood loss, total days in hospital, postoperative bleeding including needed interventions and postoperative day of bleed, coagulation studies, and pre- and post-operative medications.

Statistical analysis was performed with an exact Chi-Square test with a type 1 error of 5 percent. The secondary hypotheses were tested using exact logistic regression based model fitting and p-values. Confidence intervals were reported. All analyses were performed using SAS version 9.2.

The study was initially powered to detect a difference in proportions of 11 percent with 80 percent power and required samples sizes of 107 per group. The actual sample size available was 99 per group with an observed difference in proportions of 2 percent.

Results

Table 1 demonstrates the demographic composition of the study population. Ninety-seven patients (98 percent) in each group of 99 children underwent adenotonsillectomy, with the remaining 2 patients (2 percent) undergoing tonsillectomy only. Recurrent tonsillitis was the indication for 31 vWD patients (31 percent) and 36 non-vWD (36 percent), while obstructive sleep apnea was the indication for 51 vWD patients (52 percent) and 53 non-vWD (54 percent). No significant differences in demographic makeup were noted based on gender ($p = 0.63$, odds ratio 0.77, 95% CI 0.26-2.30) or race ($p = 0.35$, odds ratio 2.60, 95% CI 0.33-20.63). In addition there was no difference for surgical time ($p = 0.73$, odds ratio 0.99, 95% CI 0.12-0.74).

Post-tonsillectomy hemorrhage occurred in 8 of 99 (8 percent) vWD patients and in 6 of 99 (6 percent) non-vWD patients ($P = 0.58$ / Odds Ratio 1.36 / 95% CI 0.45-4.08). A two-sample test of proportions demonstrated lower and upper limits of -0.051 and 0.092. On average, bleeding occurred on postoperative day 8.6 in the vWD patient cohort and 7.8 in the non-vWD patient cohort. Four vWD patients required surgical intervention for control of hemorrhage, as did two of the non-vWD patients. The average length of hospital admission after post-tonsillectomy bleeding was 1.875 days for vWD patients and 1.0 day for non-vWD patients. No patients bled within the first 24 hours after surgery in either group. One non-vWD patient had two separate bleeding events (the first was observed and the second needed operative control). There were no differences in the severity of bleeds within the two groups. No children received blood products or required ICU admission from the blood loss.

Seventy-one of the 99 vWD patients (72 percent) were diagnosed with type 1, one was type 2B (1 percent), and one was type 2N (1 percent); 26 patients (26 percent) were of undetermined vWD type. Of the eight vWD patients that bled, six were type 1 (75 percent), one was type 2B (25 percent), and one was of unknown type (25 percent). Table 2 lists the medications given to the entire vWD cohort preoperatively and postoperatively and to those who bled or did not bleed postoperatively. Preoperatively, 93 of 99 vWD patients (94 percent) received DDAVP. Forty-six vWD patients (46 percent) received aminocaproic acid in the postoperative period, with three of these patients also being given concurrent DDAVP or factor VIII. The use of aminocaproic acid in these vWD patients did not significantly decrease the postoperative bleeding rate in comparison to vWD patients who did not receive the medication ($p = 0.11$, odds ratio 3.82, 95% CI 0.73-19.97). After post-tonsillectomy bleeding, five of the vWD patients received DDAVP and aminocaproic acid, one received DDAVP alone, and two did not receive DDAVP, aminocaproic acid, or factor VIII.

Seventy-one vWD patients in this study underwent a desmopressin challenge test prior to surgery as recorded in the charts. This consisted of infusing a single 0.3 $\mu\text{g/kg}$ dose of DDAVP in 50mL of isotonic sodium chloride solution, and obtaining activated partial thromboplastin time (aPTT), coagulation factor VIII (FVIII:C), von Willebrand factor (vWF), and ristocetin cofactor levels just before infusion and 30 minutes afterwards. The results are displayed in Table 3. Preoperative prothrombin time ($p = 0.17$, odds ratio 1.29, 95% CI 0.89-1.87) or partial thromboplastin time ($p = 0.64$, Odds Ratio 1.02, 95% CI 0.94-1.11) for non-vWD patients did not correlate with bleeding rates in non-vWD patients.

Discussion

We report our experience with 99 children diagnosed with vWD during an eight-year period, all of whom underwent tonsillectomy or adenotonsillectomy. The postoperative bleeding rate of 8 percent in this group is higher than the traditionally cited 2 percent to 4 percent post-tonsillectomy bleeding rate in a non-vWD population,¹⁵ yet in comparison to a cohort

matched using five variables (age, type of surgery, surgical technique, indication for surgery, and year of surgery) the difference in bleeding rates was not statistically significant (8% vs. 6% respectively). However, the 95% confidence interval of the odds ratio (0.45 to 4.08) was broad. With the current sample size and limited power of the study, we can not rule out the possibility of a difference in the two groups that is clinically relevant. No demographic factors or hematologic studies were identified as predictors for post-tonsillectomy bleeding in this study.

The specific variant of vWD dictates the treatment algorithm that is mostly likely to be effective. Desmopressin acetate (DDAVP), a synthetic analogue of antidiuretic hormone, is the treatment of choice for type 1 vWD and many patients with other forms of vWF/factor VIII-deficient vWD because it induces the secretion of autologous vWF and factor VIII from endothelial cell compartments into the plasma.^{4,6,17} This relatively inexpensive and widely available medication is diluted in 50 mL of isotonic saline solution and administered to children and adults at a dose of 0.3 µg/kg of body weight via intravenous infusion over 30 minutes, typically resulting in a transient increase in plasma vWF and factor VIII three to five times baseline levels.^{4,17} Because these elevated levels last for up to six to eight hours, DDAVP is frequently given just before surgery and for acute bleeding episodes, such as in post-tonsillectomy hemorrhage.^{4,18} Patients with vWD type 2B or 3, or anyone with vWD unresponsive to DDAVP or who may have contraindications for its use, can be treated perioperatively and in situations of acute blood loss with vWF/factor VIII replacement therapy.^{4,17} Two products that have been extensively studied for this purpose are Humate-P® (CSL Behring GmbH, Marburg, Germany) and Alphanate® (Grifols Biologicals Inc., Los Angeles, CA). Aminocaproic acid, or Amicar® (Xanodyne Pharmaceuticals Inc., Newport, KY), is a medication favored for control of acute mucocutaneous bleeds, such as those seen in the oropharynx, in patients with vWD. This drug directly inhibits fibrinolysis and inactivates salivary proteolytic enzymes, making it a potentially useful adjunctive therapy in patients with vWD.^{4,18} Aminocaproic acid can be given orally, topically, or intravenously, at a dose of 50 to 70 milligrams per kilogram every four to six hours.⁴

The results of this study indicate that response to the DDAVP challenge was not a good predictor of post-tonsillectomy bleeding. There was no difference in the pre- and post-DDAVP challenge of aPTT, factor VIII:c levels, vWF antigen levels, or ristocetin cofactor activity among bleeding and non-bleeding vWD patients, which may be due to our small bleeding rate. This marks a change in comparison to a prior study published in 1999 from this institution, in which initial factor VIII levels were lower in vWD patients who had post-tonsillectomy bleeding, and lower vWF antigen levels approached statistical significance.⁶ However, the data from this study do suggest that children with known vWD who responded well to the DDAVP challenge and were treated preoperatively with DDAVP and other appropriate medications have an overall post-tonsillectomy bleeding rate lower than expected in comparison to prior studies evaluating children with vWD who underwent tonsillectomy.^{6,19} Aminocaproic acid given postoperatively did not appear to decrease the likelihood of post-tonsillectomy bleeding in vWD patients. This suggests that the medication may not prevent secondary tonsillectomy hemorrhage and might be more effective as an adjunctive therapeutic agent (e.g. during an episode of acute post-tonsillectomy hemorrhage), consistent with recommendations on its use made in prior studies.^{4,17}

There is substantial debate in the otolaryngology literature regarding the most appropriate preoperative laboratory evaluation prior to tonsillectomy. Physicians are in favor of strategies, whether with surgical technique or with patient selection, to lower the potential risk of tonsillectomy hemorrhage. The advantage of routine screen is that it helps identify all children with potential bleeding disorders so that the appropriate evaluation and perioperative treatment may occur. Many physicians obtain preoperative blood work only if

a patient or family history suggests the presence of a bleeding disorder. However, inadequate history-taking or poor patient reliability or recall may lead to a failure to diagnose a potential coagulopathy.²⁰ Furthermore, several studies have indicated that post-tonsillectomy bleeding cannot always be predicted by patient history.^{6,16,20} Physicians also obtain routine screens based on personal anecdotal experience, group practice, or litigation concerns.

Although some studies in the literature support indiscriminate preoperative screening of all patients for vWD and other coagulopathies, there can be significant drawbacks to this strategy. The common tests for screening, prothrombin time (PT) and partial thromboplastin time (PTT), have low predictive value for postoperative bleeding.^{16,21} The diagnosis of vWD is often missed with these tests, and the PT and PTT may be normal in over 50% of individuals with vWD.⁵ Moreover, false positive initial tests may create unnecessary parental concerns or delay medical care. Routine screen also needs to factor the financial burden of laboratory tests. The yearly cost of routine preoperative coagulation laboratory screening for all adenotonsillectomy patients at a single institution has been estimated at \$27.5 to \$30 million, excluding the cost of the evaluation of patients who demonstrate false positive results.^{16,21} Therefore, it has been argued that routine preoperative coagulation blood work is not indicated or cost-effective unless there is a known personal or familial history of abnormal bleeding or bruising.^{19,20,21} In 1999, the American Academy of Otolaryngology- Head and Neck Surgery developed a consensus statement that recommended preoperative coagulation evaluation only when suspected or prior genetic evaluation was unavailable.²² Despite these guidelines, many physicians still obtain routine screens on all individuals regardless of history. A recent survey demonstrated that 21% of otolaryngologists obtain regular preoperative bloodwork prior to adenotonsillectomy.²³

This historical cohort study does not address the cost-effectiveness or practicality of performing routine preoperative screening for vWD and other coagulopathies. In addition, we did not investigate the most appropriate preoperative laboratory testing in diagnosing vWD in tonsillectomy patients. The main objective was to examine the post-tonsillectomy bleeding of children identified with vWD. Our patient population does demonstrate that vWD is diagnosed in children undergoing tonsillectomy with routine tests. It should be kept in mind that the present study compares the bleeding rates of children with vWD who were identified and treated preoperatively to a population of children without vWD; it does not compare the bleeding rates children with untreated laboratory identified vWD to those with vWD who received preoperative DDAVP. Therefore, this study does not support or refute routine preoperative hematologic screen before adenotonsillectomy.

The practice at the Children's Hospital of Pittsburgh during the study period was to obtain a complete blood count, prothrombin time and partial thromboplastin time on all patients undergoing tonsillectomy. A hematology consult is sought for abnormal laboratory values not corrected on repeat testing. Children who are identified with vWD undergo DDAVP challenge. An important noted potential side effect of DDAVP administration is hyponatremia. The current Children's Hospital of Pittsburgh protocol to manage perioperative DDAVP administration is outlined in Table 4. No adverse side effects of DDAVP were noted in this study. Specifically, no child had clinically significant hyponatremia during the time period of this study.

Of final note, the documented control bleeding rate of 6 percent is above the currently accepted bleeding rate of 2-4 percent.¹⁵ Potential reasons for this include the fact that we defined a post-tonsillectomy bleed as anyone presenting with a bleeding episode documented in the medical record and not necessarily a witnessed bleed by the medical staff. It is the current policy of Children's to admit these patients for observation overnight.

Also, given the small sample size, the value of 6 percent is within the range of what could be expected by random variability.

Conclusion

Children with vWD undergoing tonsillectomy have a postoperative bleeding rate similar to that of a matched group. However, the sample size was not sufficient to eliminate the possibility of a clinically important difference between the two groups. No significant demographic or hematologic measures were identified as predictors for postoperative bleeding. Acknowledgement: We thank Howard Rockette, PhD in the statistical review of the manuscript.

Acknowledgments

This study was supported in part by NIH 1 T32 GM074927-01

References

1. Brown DL. Congenital bleeding disorders. *Curr Probl Pediatr Adolesc Health Care*. 2005; 35(2):38–62. [PubMed: 15692557]
2. Rodeghiero F, Castaman G, Dini E. Epidemiological investigation of the prevalence of von Willebrand's disease. *Blood*. 1987; 69(2):454–9. [PubMed: 3492222]
3. Mancuso DJ, Tuley EA, Westfield LA, et al. Structure of the gene for human von Willebrand factor. *J Biol Chem*. 1989; 264(33):19514–27. [PubMed: 2584182]
4. Mannucci PM. Treatment of von Willebrand's disease. *N Engl J Med*. 2004; 351(7):683–94. [PubMed: 15306670]
5. Cox Gill J. Diagnosis and treatment of von Willebrand disease. *Hematol Oncol Clin N Am*. 2004; 18(6):1277–99. viii.
6. Allen GC, Armfield DR, Bontempo FA, et al. Adenotonsillectomy in children with von Willebrand disease. *Arch Otolaryngol Head Neck Surg*. 1999; 125(5):547–51. [PubMed: 10326812]
7. Windfuhr JP, Chen YS, Remmert S. Hemorrhage following tonsillectomy and adenoidectomy in 15,218 patients. *Otolaryngol Head Neck Surg*. 2005; 132(2):281–6. [PubMed: 15692542]
8. Cuianu M, Nussbaum A, Cristea A, et al. High levels of plasma von Willebrand factor in hyperthyroidism. *Med Interne*. 1987; 25(3):205–10. [PubMed: 3659809]
9. Rogers JS 2nd, Shane SR, Jencks FS. Factor VIII activity and thyroid function. *Ann Intern Med*. 1982; 97(5):713–6. Nov. [PubMed: 6814317]
10. Pottinger BE, Read RC, Paleolog EM, et al. von Willebrand factor is an acute phase reactant in man. *Thromb Res*. Feb 15; 1989 53(4):387–94. [PubMed: 2467404]
11. Werner EJ, Broxson EH, Tucker EL, et al. Prevalence of von Willebrand disease in children: a multiethnic study. *J Pediatr*. 1993; 123(6):893–8. [PubMed: 8229521]
12. Paradise JL, Bluestone CD, Colborn DK, et al. Tonsillectomy and adenotonsillectomy for recurrent throat infection in moderately affected children. *Pediatrics*. Jul; 2002 110(1 Pt 1):7–15. [PubMed: 12093941]
13. Mattila PS, Tahkokallio O, Tarkkanen J, et al. Causes of tonsillar disease and frequency of tonsillectomy operations. *Arch Otolaryngol Head Neck Surg*. 2001; 127(1):37–44. [PubMed: 11177012]
14. Rosenfeld RM, Green RP. Tonsillectomy and adenoidectomy: changing trends. *Ann Otol Rhinol Laryngol*. 1990; 99(3 Pt 1):187–91. [PubMed: 2178542]
15. Schmidt R, Herzog A, Cook S, et al. Complications of tonsillectomy: a comparison of techniques. *Arch Otolaryngol Head Neck Surg*. 2007; 133(9):925–8. [PubMed: 17875860]
16. Zwack GC, Derkay CS. The utility of preoperative hemostatic assessment in adenotonsillectomy. *Int J Pediatr Otorhinolaryngol*. 1997; 39(1):67–76. [PubMed: 9051441]

17. Federici AB, Mannucci PM. Management of inherited von Willebrand disease in 2007. *Ann Med.* 2007; 39(5):346–58. [PubMed: 17701477]
18. Shah SB, Lalwani AK, Koerper MA. Perioperative management of von Willebrand's disease in otolaryngologic surgery. *Laryngoscope.* 1998; 108(1 Pt 1):32–6. [PubMed: 9432063]
19. Derkay CS, Werner E, Plotnick E. Management of children with von Willebrand disease undergoing adenotonsillectomy. *Am J Otolaryngol.* 1996; 17(3):172–7. [PubMed: 8827276]
20. Bolger WE, Parsons DS, Potempa L. Preoperative hemostatic assessment of the adenotonsillectomy patient. *Otolaryngol Head Neck Surg.* Sep; 1990 103(3):396–405. [PubMed: 2122369]
21. Krishna P, Lee D. Post-tonsillectomy bleeding: a meta-analysis. *Laryngoscope.* 2001; 111(8): 1358–61. [PubMed: 11568568]
22. The Academy of Otolaryngology-Head and Neck Surgery. Clinical Indicators Compendium. American Academy of Otolaryngology- Head and Neck Surg; Alexandria, VA: 1999.
23. Wieland A, Belden L, Cunningham M. Preoperative coagulation screening for adenotonsillectomy: a review and comparison of current physician practices. *Otolaryngol- Head Neck Surg.* Apr.2009 140:542–547. [PubMed: 19328344]

Table 1

Characteristics of von Willebrand and Non-von Willebrand Patients.

	vWD (N=99)		Non-vWD (N=99)		Total (N=99) N
	Bleeders (N=8) N (%)	Non-Bleeders (N=91) N (%)	Bleeders (N=6) N (%)	Non-Bleeders (N=93) N (%)	
Age (yrs):					
<2	0 (0)	0 (0)	0 (0)	0 (0)	0
2-4	3 (37.5)	31 (34.1)	1 (16.6)	31 (33.3)	32
5-10	3 (37.5)	51 (56)	5 (83.3)	51 (54.8)	56
11-15	2 (25)	8 (8.8)	0 (0)	9 (9.7)	9
≥16	0 (0)	1 (1.1)	0 (0)	2 (2.2)	2
Male	4 (50)	50 (54.9)	2 (33.3)	40 (43)	42
Female	4 (50)	41 (45.1)	4 (67.7)	53 (57)	57
Race:					
Caucasian	7 (87.5)	88 (96.7)	6 (100)	72 (77.4)	78
Afro-American	0 (0)	0 (0)	0 (0)	0 (0)	18
Other	1 (12.5)	3 (3.3)	0 (0)	0 (0)	3
Procedure:					
T&A	7 (87.5)	90 (98.9)	6 (100)	91 (97.8)	97
T	1 (12.5)	1 (1.1)	0 (0)	2 (2.2)	2
Indication:					
Recurrent tonsillitis	2 (25)	29 (31.9)	3 (50)	33 (35.5)	36
OSA	4 (50)	47 (51.6)	3 (50)	50 (53.8)	53
Both	2 (25)	15 (16.5)	0 (0)	10 (10.8)	10
vWD type					
1	6	65	—	—	—
2B	1	0	—	—	—
2N	0	1	—	—	—
unknown	1	25	—	—	—

Table 2

Medications Administered to von Willebrand Disease Patients

vWD Patient Medications	Patients Receiving Medication Preoperatively			Patients Receiving Medication Postoperatively		
	Bleeders (N=8)	Non-Bleeders (N=91)	Total (N=99)	Bleeders (N=8)	Non-Bleeders (N=91)	Total (N=99)
DDAVP alone	6	80	86	0	1	1
DDAVP+Vit K	0	2	2	0	0	0
DDAVP+Amicar	1	4	5	2	0	2
Amicar + Factor VIII	1	0	1	1	0	1
Amicar alone	0	1	1	3	40	43
Factor VIII alone *	0	1	1	0	0	0
No DDAVP/Amicar/Factor VIII	0	3**	3**	2	50	52

* DDAVP non responder

** Includes unknown fields

Table 3

Laboratory Values Pre- and Post-DDAVP Challenge in von Willebrand Patients.

	Pre-DDAVP Challenge			Post-DDAVP Challenge		
	Bleeders	Non-bleeders	TOTAL GROUP	Bleeders	Non-bleeders	TOTAL GROUP
	N = 4	N= 67	N=71	N= 4	N= 67	N=71
aPTT						
mean	43.650	39.428	39.666	34.775	33.403	33.480
range	37.4-54.3	27.2-63.4	27.2-63.4	27.5-43.8	25.7-44.4	25.7-44.4
Factor VIII:C						
mean	1.160	0.855	0.872	1.958	1.889	1.893
range	.820-1.820	.350-1.480	.350-1.820	0.950-3.0	1.0-3.460	0.950-3.460
vWF Ag						
mean	0.753	0.982	0.969	1.553	1.788	1.775
range	.540-0.970	0.3-1.830	0.3-1.830	0.920-1.960	0.770-3.370	0.770-3.370
Ristocetin cofactor						
mean	0.768	0.721	0.723	2.193	2.057	2.065
range	0.430-1.280	0.210-2.840	0.210-2.840	0.990-2.690	0.4-4.470	0.400-4.470

Table 4

Current Children's Hospital of Pittsburgh Protocol for DDAVP

- DDAVP dose of 0.3 µg/kg of body weight via IV infusion over 30 minutes prior to surgery.
- All children who receive DDAVP are admitted.
- IV fluids consist of normal saline or lactated ringers with or without dextrose, administered at one-half the appropriate weight calculated rate.
- Strict input and output is measured.
- Serum sodium levels are checked every six hours.
- If sodium is less than 134,
 - IV fluids are stopped.
 - Sodium serum is checked every three hours along with urine osmolality.
- If sodium is less than 132,
 - IV fluids are stopped
 - Endocrinology consult is obtained.