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# Anaesthesiologist's Role in Patient with Von Willebrand Disease for Elective Adenoidectomy: A Case Report

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## Abstract

The most common bleeding disorder is von Willebrand disease (vWD). It is however a complex disease to diagnose. Management of this disease is not well understood by many anaesthesiologists who may experience these patients in elective or even emergency settings. This is a case presentation of a six-year-old patient of Arabic ethnicity who was diagnosed with vWD during preoperative testing and was managed perioperatively with a multidisciplinary team for a good outcome.

**Keyword:** Von Willebrand disease, Von Willebrand factor, Adenoidectomy, Preoperative testing

## Introduction

The most common disorder of bleeding is von Willebrand disease (vWD) [1]. Bleeding may occur for the first-time during surgery or post-trauma. The aim of treatment for vWD is to correct the dual defect of hemostasis caused by the abnormal or reduced vWF and the concomitant deficiency of factor VIII. This case report is about a six-year-old male patient with vWD posted for adenoidectomy in whom the disease was diagnosed on routine pre-operative testing. vWD is classified into three major types. Type one is a partial quantitative deficiency of vWF, type two consists of qualitative abnormalities of vWF, and type three is a complete quantitative deficiency of vWF [2]. Diagnosis in vWD is challenging and requires several tests done repeatedly with a correct interpretation. The three tests most used to diagnose vWD are Von Willebrand antigen, vWF activity, and circulating factor VIII levels [3]. Anaesthesiologists need to understand the diagnosis and management of vWD and have a plan in place [4], as many times these may be encountered in emergency settings.

## Case Report

A six-year-old male child of Arabic ethnicity weighing 21 kilograms was posted for elective adenoidectomy. There was no past surgical history. The parents gave a history of phototherapy for one day for neonatal jaundice. There was no history of bleeding or chronic use of medication. Neither was there a history of anemia. There is no history of parental consanguinity. The patient had a single female sibling. During his routine pre-operative blood tests done in April of 2023, the aPTT was prolonged (54 seconds with an upper limit of 45 seconds) [5, 6]. The hematologist reviewed the patient and advised a series of tests, including complete blood count with WBC differential, transferrin saturation, vWF antigen, vWF multimers, factor VIII levels, factor IX levels, thrombin time, D Dimer, aPTT mixed studies, factor XII activity, hemoglobin fractionation, quantitation, and electrophoresis. aPTT was repeated and found to be prolonged (60 seconds with an upper limit of 45 seconds). On further testing, lupus anticoagulant [7] was elevated at 45 Micropulse Lidar Units (upper limit 35 MPL). Initial vWF was 54% (range of 50 to 200%), and vWF antigen was 31% (normal range of 50 to 160%). Factor VIII activity was reduced at 49% (57-163%). Repeating the lupus anticoagulant after two months was advised. The plan was to repeat the tests in 2-3 months. The patient still did not have any bleeding symptoms. He was not on any treatment except iron supplements. In fact, during this period, he had fallen once on his face while playing but did not have any bleeding. Repeat testing in July 2023 showed the lupus anticoagulant had resolved, but vWF antigen was 28% (50-160%), and vWF activity was 19.2% (50-200%). So, a diagnosis of progressive vWD was made. Lupus anticoagulant presence is usually transient in children [7]. Repeat aPTT in August 2023 had a normal result of 32 seconds (30-45 seconds). The patient was cleared for surgery by the hematologist. The perioperative plan was as follows: tranexamic acid orally 25 milligrams per kilogram three times a day to start one day before surgery and continue three days after the surgery. Secondly, on the day of surgery, 10 milligrams per kilogram of intravenous tranexamic acid was given eight hourly till discharge. Oral tranexamic acid to continue at home for the next three days. vWF -factor VIII product (Wilate) 25 International Units per kilogram was given preoperatively one hour before surgery (to be given 1-2 hours preoperatively). He was shifted to the operation theatre with factor VII recombinant (Novoseven) at bedside in case of bleeding (90 micrograms per kilogram to be given as starting dose if required).

In addition, on the day of surgery (28 August 2023), complete blood count, antibody screening, factor VIII, vWF activity, vWF antigen,

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and Von Willebrand multimers were sent.

Endoscopic coblation was used for the adenoidectomy. The duration of surgery was prolonged (95 minutes). However, there was no significant bleeding intraoperatively. Vitals were stable throughout the procedure.

He was in Postanaesthesia care unit for 2 hours with frequent checks for any bleeding and discharged to ward with Modified Aldrete score 10/10.There was no bleeding post-operatively either in the immediate or late postoperative period. The child was followed up both in the hematology clinic as well as otorhinology (ENT) clinic for two weeks postoperatively. His first postoperative visit to the hematology clinic was three days postoperative, as advised.

## Discussion

The most common disorder of bleeding is vWD. However, this disease is heterogeneous and complex to manage [8]. It isn't a static disease as the coagulation factors may be affected by other factors causing fluctuating values [8]. The different types of VWD must be treated with different approaches. The treatment has adverse effects that must be monitored and managed, especially in the pediatric age. Adenoidectomy is an airway surgery, considered a major surgery for bleeding disorders. Acquiring hemostasis and maintaining it for 7-14 days postoperatively is recommended. Regular follow up in hematology clinic after surgery for at least two weeks is as important as the preoperative management to ensure postoperative hemostasis.

In vWD, hemostatic levels are to be maintained for 3-5 days for minor and 7-14 days for major surgeries.

In case of elective surgeries, early vWF replacement will stabilize factor VIII. However, in emergency cases, coadministration of vWF and f VIII is required to ensure hemostasis.

In our hospital, all patients undergoing adenoidectomy with or without tonsillectomy undergo laboratory tests that include the complete blood count, including hemoglobin level and hematocrit, prothrombin time and partial thromboplastin time, and international normalized ratio.

Bleeding time is obtained only if a patient has a medical or surgical history of abnormal bleeding. If any tests are abnormal, the tests are repeated, and patients with persistent abnormalities will undergo further coagulation testing. vWF is a plasma glycoprotein [9] that plays a role in primary and secondary hemostasis. vWF is synthesized exclusively in endothelial cells and megakaryocytes. It mediates platelet adhesion at the site of vascular injury by binding to the connective tissue and the platelets, critical to hemostasis and thrombus formation. vWF also binds and stabilizes blood clotting factor VIII. Therefore, defects in vWF can cause bleeding with features typical of platelet dysfunction or features of mild to moderately severe hemophilia A or both [10].

To come to a diagnosis of vWD, the physician should ask questions about personal and family histories of bleeding. The doctor should also check for bruising or other signs of bleeding. They order tests to measure the blood clotting [11]. vWD diagnosis requires several tests to be done. Since the screening tests are usually normal in most patients with vWD, the diagnosis requires specific tests to be performed. Screening tests are often normal in vWD and more specific tests need to be performed. Screening tests include the complete blood count, activated partial thromboplastin time, prothrombin time, and fibrinogen time. Specific tests must be done repeatedly to diagnose the specific bleeding disorder. Sometimes, clotting factors are ordered to confirm the presence or absence of coexisting disorders.

Once the diagnosis of vWD is made, vWF antigen electrophoresis [10] is done to help diagnose the variant of vWD. Managing vWD includes treatment with antifibrinolytics like tranexamic acid, which is often helpful for mucosal bleeding, most commonly in the mouth., for epistaxis and menorrhagia [2]. Patients with mild to moderate type one vWD can be treated with desmopressin; it may occasionally be effective in type 2 vWD but never in type 3 vWD [8]. Desmopressin is generally not recommended in children less than three years of age due to documented reports of hyponatremia and seizures [10] and is also relatively contraindicated in children with previous seizure disorders. When required, preoperatively administer 30 minutes before the surgery. Daily dosing can be given up to three doses. After 48 hours, tachyphylaxis may be a concern [8]. vWF –Factor VIII plasma concentrate is also another modality of treatment for vWD. It may be required in type 1 vWD if there is severe bleeding or unresponsiveness to desmopressin. It treats patients with bleeding in type 2 and type 3 vWD. The recommended doses depend on the type of bleeding. Increased doses are required according to whether it is a gastrointestinal bleed, a central nervous system bleed, post-trauma, or a surgical bleed.

Thromboembolism is a possible complication of treatment for vWD. The overall rates of thromboembolism are very low (0.048% of all vWF replacement or 1.9% treated vWD). Caution must be exercised for those with high risk for thromboembolism including old age, obesity, or estrogen replacement.

## Conclusion

With the complete workup of vWD, its management can be planned best with a multidisciplinary team. It requires regular follow-up as well to have the best outcome. vWD must be diagnosed and managed by an experienced hematologist. The patient must be optimized preoperatively as best as possible and postoperative follow up depending on surgery, for a minimum of three days to upto two weeks. The plan must be shared with anaesthesiologist as they are responsible for the intraoperative and postoperative medical management of the patient. The patient's blood investigations may be repeated in consultation with a hematologist.

## Clinical message

Anaesthesiologists' role and scope of practice include pre-operative evaluation and optimization, intraoperative anaesthetic and medical management, and postoperative care. This consists of the management of perioperative bleeding. The perioperative management for our case was planned meticulously, and the surgery was managed successfully with the help of multidisciplinary care from hematology, anaesthesiology, and otorhinology (ENT) to have an uneventful outcome.

**Declaration of patient consent:** The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his/her consent for his/her images and other clinical information to be reported in the Journal. The patient understands that his/her name and initials will not be published, and due efforts will be made to conceal his/her identity, but anonymity cannot be guaranteed.

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