Von Willebrand disease is the most common inherited blood disorder, occurring in about 1% of the population. It results from a deficiency in the quality or quantity of von Willebrand factor, which is necessary for adequate hemostasis. An evidenced-based approach is prudent when this derangement is coupled with a potentially fatal obstetric complication. This article examines the anesthetic management of a parturient with a known diagnosis of von Willebrand disease who presented to the labor and delivery unit in active labor and with a suspected uterine placental abruption.

Keywords: Abruption, anesthesia, placenta, uterus, Von Willebrand.
The mother and fetus were continuously monitored. The desmopressin administration was completed 30 minutes after the infusion was begun and 60 minutes after the decision was made to proceed to cesarean delivery.

Denitrogenation was accomplished using high-flow oxygen delivered via a tight-fitting face mask after the completion of the desmopressin infusion. The surgical site was prepared and draped, and the surgical team was fully prepared. A rapid-sequence induction with the Sellick maneuver was then performed. Induction was accomplished with propofol, 140 mg IV, and intubation was facilitated with rocuronium, 30 mg IV. The patient was intubated using direct laryngoscopy with a Macintosh No. 3 blade, and 7.0-mm oral endotracheal tube was placed into the trachea. After positive confirmation of intubation of the trachea via visualization of the endotracheal tube passing through the vocal cords, presence of end-tidal carbon dioxide, chest rise, and bilateral breath sounds, the obstetrician was prompted to begin the incision.

A 4.1-kg (9-lb) male infant was delivered 2 minutes after intubation. The anesthetic was maintained throughout the use of sevoflurane, at 2% in 100% oxygen. Fentanyl, 100 µg IV, and hydromorphone, 2 mg IV, were administered for analgesia. Oxytocin, 20 U in a liter of lactated Ringer’s solution, was initiated to assist with uterine contraction after delivery. The uterus was couvelaire in appearance. The placenta appeared to have detached prematurely, and there was some extravasation blood into the uterine musculature, but overall the anatomy of the uterus was normal. The placental abruption was estimated at 90%. Apgar scores were 1 at 1 minute, 3 at 5 minutes, and 6 at 10 minutes. The infant was transferred to the neonatal intensive care unit and remained there overnight. Neuromuscular muscle blockade was fully reversed using neostigmine, 3 mg IV, and glycopyrrolate, 0.4 mg IV. The endotracheal tube was removed when the patient was following commands. The estimated blood loss was 750 mL, and a total of 1,700 mL of lactated Ringer’s solution was infused during the procedure.

Initial postanesthesia recovery vital signs were a temperature of 37.5°C (99.4°F), blood pressure of 124/76 mm Hg, heart rate of 118/min, respiratory rate of 16/min, and oxygen saturation of 100% with 10 L/min of oxygen via a nonrebreathing face mask. The postoperative hematocrit was 24.9%. Vital signs before discharge from postanesthesia recovery were a temperature of 37.3°C (99.0°F), blood pressure of 120/76 mm Hg, heart rate of 95/min, respiratory rate of 18/min, and oxygen saturation of 99% while breathing room air. Lochia was minimal, and the patient was discharged to the postpartum unit, where a second dose of desmopressin (18 µg) was infused IV 12 hours after the initial dose. Both mother and infant were discharged home on postoperative day 3.

**Discussion**

- **Pathophysiology.** Von Willebrand disease, first described by Erik von Willebrand in 1926, is primarily an autosomal inherited disorder in which vWF is quantitatively or qualitatively deficient (Table 1). Four genetically linked types have been identified in addition to an acquired version. Von Willebrand factor is produced by endothelial cells and megakaryocytes, and can be found in these cells as well as in plasma and platelets. This factor is a carrier for factor VIII, helps prevent the destruction of other coagulation factors, and plays an essential role in hemostasis because it mediates adhesion of platelets to the endothelium in the presence of endothelial injury.

**Table. Von Willebrand Disease Types and Subtypes**

<table>
<thead>
<tr>
<th>Type</th>
<th>Prevalence</th>
<th>vWF levels</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type 1</td>
<td>60%-80%</td>
<td>Decreased</td>
<td>Quantitative deficiency of vWF</td>
</tr>
<tr>
<td>Type 2A</td>
<td>Normal</td>
<td>Normal</td>
<td>Qualitative deficiency of vWF; impaired binding between platelets and vWF</td>
</tr>
<tr>
<td>Type 2B</td>
<td>Normal</td>
<td>Normal</td>
<td>Qualitative deficiency of vWF; abnormal affinity between platelets and vWF</td>
</tr>
<tr>
<td>Type 2M</td>
<td>Normal</td>
<td>Normal</td>
<td>Qualitative deficiency of vWF; impaired binding between platelets and vWF</td>
</tr>
<tr>
<td>Type 2N</td>
<td>Normal</td>
<td>Normal</td>
<td>Qualitative deficiency of vWF; impaired binding between vWF and factor VIII</td>
</tr>
<tr>
<td>Type 3</td>
<td>5%-10%</td>
<td>Absent</td>
<td>Quantitative deficiency of vWF</td>
</tr>
<tr>
<td>Pseudo- or</td>
<td>Very rare</td>
<td>Decreased</td>
<td>Abnormal functioning platelet glycoprotein and impaired binding ability to vWF</td>
</tr>
<tr>
<td>platelet-type</td>
<td></td>
<td></td>
<td>removal of vWF multimers and platelets from circulation</td>
</tr>
<tr>
<td>Acquired type</td>
<td>Very rare</td>
<td>Decreased</td>
<td>Autoantibodies produced secondary to triggering mechanisms; decrease in vWF levels and functionality</td>
</tr>
</tbody>
</table>
deficiency of vWF, with levels ranging from 20% to 50% of normal values. Patients may be asymptomatic or have only mild manifestations, which include epistaxis or minor bleeding after dental procedures.

Type 2 is found in 15% to 30% of patients with vWD1 and results from a qualitative defect rather than a quantitative deficiency. Four subtypes (type 2A, type 2B, type 2M, type 2N) have been identified. Classification of the subtypes is based on the characteristics of vWF multimers or series of polymers. Symptoms may be mild such as epistaxis, but excessive bleeding after surgery can occur.

Type 3 is the rarest form of vWD and is found in only 5% to 10% of affected patients. Levels of vWF are undetectable in the plasma. Subsequently, levels of factor VIII are extremely low. Symptoms are usually severe and include spontaneous bleeding and bleeding into the joints and muscles.

Pseudo- or platelet-type vWD is the result of a genetic mutation and abnormal functioning of a glycoprotein found in platelets. Ultimately, the levels of vWF and platelets are reduced, and the susceptibility to bleeding is greatly increased. This type is extremely rare, with only 50 known cases in the world in 2011.

Acquired vWD is a very rare condition with many triggers. These include autoimmune disorders, heart disease, cancer, and medications including ciprofloxacin, griseofulvin, and valproic acid. Patients usually present with mucocutaneous bleeding.

Treatment of von Willebrand Disease. Treatment options for vWD include blood products such as cryoprecipitate and vWF-factor VIII concentrates, antifibrinolytics, and most commonly desmopressin. Desmopressin is a synthetic version of the natural hormone vasopressin or antidiuretic hormone. Most effective in patients with type 1 vWD, desmopressin increases the plasma levels of vWF and factor VIII by stimulating their release from endothelial cells. Side effects of its use include headache, nausea, gastrointestinal issues, flushing and (rarely) hyponatremia, and seizures. It is available in intravenous and nasal spray forms.

Uterine Placental Abruption. Placental abruption is a premature separation of the placenta from the uterine lining and occurs in 1% of all pregnancies. Treatment is dependent on the severity and location of the abruption as well as the gestational age. Bedrest and close monitoring are appropriate in mild cases, whereas an emergent cesarean delivery is performed in the most severe presentations. Induction of labor with vaginal delivery is preferred if cervical conditions are favorable and if there is no fetal distress. Placental abruptions have a fetal mortality rate of 15% to 25%.14

Obstetric and Anesthesia Considerations. There is a gradual increase in vWF and factor VIII in pregnancies without coagulopathies. These levels return to baseline in the postpartum period. Although these factors may increase in some patients with vWD as the pregnancy progresses, the levels in more severe cases increase minimally or not at all. The degree of increase in these factors is dependent on the type and severity of the disease. Assessment of coagulation in these patients may be difficult. Common laboratory tests assessing coagulation such as the activated partial thromboplastin time can yield normal results. The parturient is at increased risk of a number of perinatal complications, including miscarriage, antepartum hemorrhage, epidural and spinal hematoma in association with neuraxial anesthesia, and postpartum hemorrhage. Choices regarding mode of delivery can include vaginal delivery. However, cesarean delivery should be selected if prolonged labor or an instrumental delivery is anticipated. Although some sources report the successful use of neuraxial anesthesia in patients with type 1 vWD, they do not specifically address a scenario of vWD complicated with a confirmed uterine placental abruption. Overall, there is a paucity of published data regarding the provision and safety of neuraxial techniques in patients with bleeding diatheses. Therefore, each case should be evaluated individually and with a multidisciplinary approach to determine the best anesthetic plan.

This case presented a unique scenario in which a patient with a known diagnosis of vWD experienced a uterine placental abruption. If the situation had called for an immediate cesarean delivery, the plan was to administer desmopressin as soon as it was available and support the patient with blood products as needed. Fortunately, maternal vital signs and fetal heart tones were relatively stable. This stability allowed for adequate surgical and anesthesia preparation, including administration of desmopressin, which may have helped prevent excess blood loss during the perioperative period.

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