ABSTRACT
Hemophilia A is a hemorrhagic trend almost exclusively affecting males (X-related recessive disease). In 85% of cases, it is caused by factor VIII deficiency, called hemophilia A or classic hemophilia. Successful anesthetic management depends on the special care and a multidisciplinary team of health professionals informed about the disease, including qualified hematologist, surgeon, and anesthesiologist.

Key words: Anesthesia; hemophilia; ventriculoperitoneal shunt

Introduction
Hemophilia is a hemorrhagic trend almost exclusively affecting males (X-related recessive disease). In 85% of cases, it is caused by factor VIII deficiency, called hemophilia A or classic hemophilia. All the patients with hemophilia, regardless of the severity of the disease, are at risk of excessive bleeding during surgery. For any surgical intervention, it deserves special care and a multidisciplinary team of health professionals informed about the disease, including qualified hematologist, surgeon, and anesthesiologist. We report a case of child with hemophilia A undergoing an emergency ventriculoperitoneal shunt for large cerebral arachnoid cyst.

Case Report
A 3-month-old boy weighing 5.4 kg with diagnosed hemophilia A (American Society of Anesthesiologists-III E) was scheduled for ventriculoperitoneal shunt due to large cerebral arachnoid cyst [Figure 1]. He had a positive family history of hemophilia A. He was born full term by cesarean section with normal Apgar score and without neonatal intensive care unit admission. He was referred in emergency department with history of hydrocephalus. Glasgow coma scale was 15/15 and head circumference was 42 cm with full and nonbulging anterior fontanel. On airway examination, there was a receding mandible.

Baseline laboratory results showed hemoglobin (Hb) 11.3 g/dl, a prothrombin time (PT) of 17 s (control value, 10.9–13.3 s), an activated partial thromboplastin time of 137 s (control value, 25–34 s), and a platelet count of 130,000/mm³. His preoperative factor VIII activity was <4% of normal. The consulting hematologist prescribed factor VIII therapy at 50 U/kg 12 hourly. Recommendations also provided for at
least 4 days of additional postoperative therapy in an attempt to maintain a postoperative factor VIII activity of at least 50% of normal.

As scheduled, the patient received 270 U of factor VIII before surgery and tolerated the infusion well. Preoperative PT of 14 s, APTT of 55 s, and a platelet count of 140,000/mm$^3$ were observed. He was then transported to the operating room where an uneventful induction of anesthesia was performed. Endotracheal intubation was done with Glidescope to avoid airway trauma in view of possible difficult intubation. The entire operation lasted 1 hour. Estimated blood loss during surgery was minimal. The patient was monitored with electrocardiograph, noninvasive blood pressure, and pulse oximetry. All vital parameters were maintained well throughout the procedure.

Reversal of residual neuromuscular blockade was achieved satisfactorily with neostigmine and glycopyrrolate. Spontaneous ventilation was quickly established. After extubation of the trachea, the patient was given oxygen by facemask for 10 min in the operation theater and subsequently shifted to pediatric intensive care unit (PICU).

Postoperatively, the patient was given factor VIII 12 hourly for 4 days; on the forth postoperative day, the patient’s factor VIII activity was 60% with Hb of 10.7 g/dl. The patient was discharged on the forth postoperative day.

**Discussion**

Hemophilia is a hemorrhagic trend almost exclusively affecting males (X-related recessive disease). In 85% of cases, it is caused by factor VIII deficiency, called hemophilia A or classic hemophilia.$^{[1]}$ The incidence of hemophilia A is 1 in 5000 or 10,000 males and may be classified as mild, moderate, or severe.

Hemophilia severity is classified according to the baseline level of clotting factor activity. Factor VIII activity levels are reported in units, with 1 U/ml corresponding to 100% of the factor found in 1 ml of normal plasma. Normal plasma activity levels usually range between 0.5 U ml$^{-1}$ and 1.5 U ml$^{-1}$ (50–150%).$^{[1]}$ Severely affected patients have <1% of normal factor levels whereas those with moderate disease have 1–4% and with mild disease have 5–50%.$^{[2]}$ Those with <1% of normal factor VIII level are susceptible to spontaneous bleeding episodes such as hemarthrosis, soft tissue hematoma, and intracranial hemorrhage. Intracranial hemorrhage either extra- or intra-cerebral is common in severe disease.$^{[3]}$ All the patients with hemophilia, regardless of the severity of the disease, are at risk of excessive bleeding during surgery. Intramuscular premedication should be avoided. Vascular access itself does not cause excessive bleeding and should be appropriate for the proposed procedure. After induction of anesthesia, extra care should be taken in manipulation or intubation of the airway as it can cause submucosal hemorrhages, which can prove life-threatening. Nasal intubation should be avoided as it can prove traumatic and bleeding from the site can lead to aspiration. Care should be taken during positioning of the extremities, and pressure points should be padded to prevent intramuscular hematomas or hemarthrosis.$^{[4]}$ Hypertension and tachycardia during surgery in hemophiliacs can lead to increased surgical bleeding. Controlled hypotension techniques prevent hemostasis of small vessels but are not recommended. Hemodynamic conditions should be maintained near normal. Surgeon should give special attention to small vessel hemostasis, rather than trusting on hemostatic physiological mechanisms.$^{[5]}$ The patient was taken up for surgery at the earliest after the aptt was normalized and availability of sufficient quantities of clotting factor concentrates ensured before undertaking surgery.$^{[6]}$ As our patient needed emergent surgery, factor VIII assay and inhibitor screening were not feasible preoperatively. Postoperative PICU transfer is mandatory to monitor the signs and symptoms of intracranial hemorrhage. Postoperative monitoring for bleeding was done with Hb and AT levels along with factor VIII assay.

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**Conflicts of interest**
There are no conflicts of interest.

**References**

1. DiMichele D, Neufeld EJ. Hemophilia. A new approach to an old disease. Hematol Oncol Clin North Am 1998;12:1315-44.
2. Cahill MR, Colvin BT. Haemophilia. Postgrad Med J 1997;73:201-6.
3. William ED, Glass DD. Haematological diseases. In: Katz,
Khokhar, et al.: Hemophilia A and emergency ventriculoperitoneal shunting

Bemumof, Kadis, editors. Anesthesia and Uncommon Diseases. Philadelphia: W. B. Saunders Company; 1990. p. 378-436.

4. Carle G, Frank F. De Gruchy’s Clinical Haematology in Medical Practice, Coagulation Disorders. Boston: Oxford University Press; 1993. p. 406-36.

5. Flores RP, Bagatini A, Santos AT, Gomes CR, Fernandes MS, Molon RP. Hemophilia and anesthesia. Rev Bras Anestesiol 2004;54:865-71.

6. Bullock MR, Chesnut R, Ghajar J, Gordon D, Hartl R, Newell DW, et al. Surgical management of traumatic brain injury author group. Surgical management of acute subdural hematomas. Neurosurgery 2006;58(3 suppl):S16-24.

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