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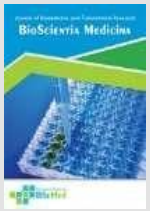

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Emergency Neuroanesthesia for Spontaneous Subdural Hematoma in a Pediatric Patient with Hemophilia A: A Protocol-Based Multidisciplinary Approach

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A B S T R A C T

Background: Spontaneous subdural hematoma (SDH) in pediatric patients with Hemophilia A is a rare, life-threatening emergency requiring a delicate balance between hemostatic correction and neuroprotective anesthesia. The mortality rate is high without immediate surgical decompression, yet the surgery itself poses catastrophic bleeding risks. **Case presentation:** We report the case of an 11-year-old male (25 kg) with severe Hemophilia A (Factor VIII <1%) who presented with a three-day history of headache and vomiting, culminating in a sudden loss of consciousness (GCS E2V2M5). Neuroimaging revealed a massive left frontotemporoparietal SDH (8 mm thickness) with a 12 mm midline shift and non-communicating hydrocephalus. The patient had discontinued prophylaxis five months prior. Management involved a strict multidisciplinary protocol. Preoperatively, aggressive Factor VIII replacement was initiated to achieve 100% activity. Intraoperatively, a total intravenous anesthesia (TIVA) strategy utilizing propofol, fentanyl, and dexmedetomidine was employed to maintain cerebral perfusion pressure (CPP) while strictly controlling intracranial pressure (ICP). Tranexamic acid was used as an adjunct. The patient underwent successful craniotomy and hematoma evacuation with minimal blood loss. Postoperative care focused on serial factor VIII replacement and neurological monitoring, resulting in a favorable discharge outcome. **Conclusion:** Successful management of spontaneous SDH in hemophilia requires a target-controlled approach to both hemostasis and hemodynamics. The integration of preoperative factor loading, neuroprotective anesthesia with dexmedetomidine, and postoperative vigilance is critical for survival.

1. Introduction

Hemophilia A is an X-linked recessive congenital bleeding disorder caused by a deficiency in coagulation factor VIII (FVIII), affecting approximately 1 in 5,000 male births globally.[1] The clinical phenotype correlates strongly with the residual factor activity; severe hemophilia is defined by factor VIII levels of less than 1%. While hemarthroses and intramuscular hematomas constitute the majority of bleeding episodes, intracranial hemorrhage (ICH) remains the most feared complication and the leading cause of mortality, with an incidence rate estimated between 3% and 10% in the severe hemophilia population.[2]

Among the subtypes of intracranial hemorrhage, spontaneous subdural hematoma (SDH) is particularly insidious in the pediatric population.[3] Unlike traumatic SDH, which results from high-velocity impact rupturing bridging veins, spontaneous SDH in hemophiliacs can arise from trivial physiological stress. The pathophysiology involves the rupture of fragile cortical vessels that, in the absence of the Tenase complex (Factor VIIIa-IXa), fail to form a stable platelet-fibrin plug. This leads to a slow but

progressive accumulation of blood in the subdural space, often presenting with non-specific prodromal symptoms such as headache or nausea before precipitating a catastrophic rise in intracranial pressure (ICP).[4]

The anesthetic management of a hemophilic child requiring emergency craniotomy presents a unique and complex physiological paradox for the anesthesiologist.[5] The primary neurosurgical goal is to reduce ICP and maintain cerebral perfusion pressure (CPP). However, the hematological goal is to prevent exsanguination and facilitate clot formation. Standard neuroanesthetic techniques used to lower ICP, such as induced hypocapnia or aggressive osmotic diuresis, must be carefully balanced against the risk of hemodynamic turbulence, which could dislodge fragile clots or induce re-bleeding.[6] Furthermore, airway instrumentation in a coagulopathic patient carries specific risks, including glottic hemorrhage and airway obstruction, necessitating a modified rapid sequence induction (RSI) approach.[7]

Despite the gravity of this condition, current medical literature often addresses the hematological and surgical management in isolation.[8] There is a paucity of data detailing the specific anesthetic strategy—specifically the synergy between hemostatic adjuncts, such as tranexamic acid, and neuroprotective agents, such as dexmedetomidine—in the pediatric hemophilia population. The integration of these pharmacological agents requires precise timing and dosage to ensure that the patient is not only anesthetized but also protected from secondary brain injury and coagulopathic collapse.[9,10]

This case report and review aim to bridge the gap between hematological guidelines and neuroanesthetic practice. We detail a comprehensive, protocol-based anesthetic management strategy applied to a pediatric patient with severe hemophilia A presenting with life-threatening SDH. We highlight the novel application of a multimodal neuroprotective regimen combining dexmedetomidine, total intravenous anesthesia (TIVA), and strict FVIII target-control. This report provides a reproducible clinical framework for perioperative physicians managing high-risk coagulopathic neurosurgery, emphasizing that time is brain must be coupled with time is blood in this specific demographic.

2. Case Presentation

Written informed consent was obtained from the patient's legal guardian (parents) for the publication of this case report and any accompanying images. The protocol was reviewed and aligned with the institutional ethical standards for clinical case reporting.

The case concerns an 11-year-old male patient, weighing 25 kg, who presented to the Emergency Department in a state of precipitous neurological decline. The clinical narrative began three days prior to admission, characterized by an insidious but progressive onset of symptoms. The patient's mother described a prodrome of intensifying cephalalgia and nausea, which culminated in episodes of projectile vomiting—a classic hallmark of rising intracranial pressure. Despite the severity of these symptoms, the presentation was devoid of any identifiable precipitating event; there was no history of significant head trauma, falls, high-velocity collisions, or sports-related injuries reported by either the family or the patient prior to his loss of consciousness.

The patient's medical background provided the critical context for this emergency. Diagnosed with severe Hemophilia A in infancy, he had a documented baseline factor VIII (FVIII) activity of less than 1%. Historically, his condition had been managed with on-demand factor VIII injections. However, the management history revealed a significant gap in care: due to logistical challenges and issues with medication access, his prophylactic regimen had been discontinued five months prior to this admission. This discontinuation left him in a vulnerable, unprotected state where spontaneous hemorrhage became a constant, latent threat. His past medical history was otherwise unremarkable, with no evidence of inhibitors, hypertension, diabetes mellitus, or congenital heart disease that might complicate

anesthetic management.

Upon admission to the Intensive Care Unit (ICU), the patient's condition was critical. He was stuporous, requiring immediate resuscitation and assessment. The primary survey revealed a Glasgow Coma Scale (GCS) of 9 (E2, V2, M5), indicating a moderate-to-severe traumatic brain injury classification with a threatened airway. The breakdown of the score—eyes opening to pain, incomprehensible sounds, and localizing to pain—suggested that while cortical function was severely depressed, gross motor pathways remained partially intact. Pupillary examination showed isocoric pupils (3 mm/3 mm) that were briskly reactive to light, offering a reassuring sign that brainstem reflexes were preserved despite the cerebral compromise. No lateralizing motor deficits were initially observed, and the patient demonstrated the ability to localize to painful stimuli, suggesting that the herniation syndrome had not yet progressed to uncal compression.

Vital signs reflected a physiologic stress response. The patient exhibited sinus tachycardia with a heart rate of 109 bpm, likely a compensatory mechanism for pain, hypovolemia from vomiting, or increased sympathetic tone associated with intracranial hypertension. His blood pressure was 119/75 mmHg, preserving cerebral perfusion pressure (CPP) in the face of rising intracranial pressure (ICP). Respiratory function was stable with a rate of 18 breaths per minute, maintaining an oxygen saturation of 100% on 15 L/min via a non-rebreather mask (NRBM). He was afebrile (36.5°C), ruling out an infectious etiology for his altered mental status.

The urgency of the clinical picture necessitated immediate neuroimaging. A non-contrast computed tomography (CT) scan of the head confirmed the working diagnosis of a catastrophic intracranial hemorrhage. The scan revealed a massive, hyperdense crescentic collection along the left cerebral convexity, the distinctive radiological signature of an acute subdural hematoma (SDH). The hematoma burden was substantial, measuring 8 mm in maximal thickness. The mass effect exerted by this collection was disproportionate to the volume, resulting in a significant midline shift of 12 mm to the right. This shift had caused complete effacement of the ipsilateral lateral ventricle and radiological evidence of subfalcine herniation, confirming that the intracranial compliance had been exhausted. Furthermore, secondary findings of generalized cerebral edema and non-communicating hydrocephalus underscored the critically high ICP, necessitating immediate surgical decompression to prevent irreversible brainstem ischemia.

The hematological profile confirmed the severity of the coagulopathy. The activated partial thromboplastin time (aPTT) was significantly prolonged at 98.4 seconds (Control: 32.1s), while the prothrombin time (PT) and INR remained within normal limits. This dissociation localized the defect to the intrinsic pathway of the coagulation cascade. A confirmatory factor VIII assay demonstrated activity levels of <1%, consistent with severe Hemophilia A. Additionally, the complete blood count revealed a hemoglobin of 10.2 g/dL with microcytic hypochromic indices, suggesting a chronic anemia likely secondary to occult bleeding or chronic disease, rather than acute blood loss alone.

Faced with radiological evidence of impending herniation and a profound coagulopathy, the multidisciplinary team—comprising Hematology, Neurosurgery, and Anesthesiology—activated a zero-delay protocol (Table 1). This protocol was designed to address the physiological paradox of the hemophilic neurosurgical patient: the need to lower ICP immediately while simultaneously correcting the inability to form a clot (Table 2). The cornerstone of the preoperative phase was the rapid correction of Factor VIII deficiency. The therapeutic target was strictly set at 100% activity to ensure hemostasis during the meningeal breach and to withstand the high shear stress of surgery. The required loading dose was calculated using the formula: Dose (IU) = 0.5 × Target Rise (%) × Body Weight (kg). With a baseline assumed to be negligible and a target rise of 80-100% for a 25 kg patient, the calculation yielded a requirement of

1000 IU. A loading dose of 1000 IU of factor VIII concentrate (Koate®) was administered intravenously over 10 minutes immediately upon diagnosis (09:00 AM, Day 1). This was followed by a maintenance regimen of 1000 IU every 12 hours to sustain trough levels. Recognizing the peak consumption of coagulation factors during tissue dissection, a pre-incision boost of an additional 1000 IU was scheduled for one hour prior to surgery (08:30 AM, Day 2). To further secure clot stability, Tranexamic Acid (TXA) was administered at a dose of 250 mg (10 mg/kg) IV. This antifibrinolytic agent was employed to inhibit the breakdown of the fragile platelet-fibrin plug in the hyper-fibrinolytic environment of intracranial surgery. Concurrently, aggressive neuro-resuscitation was initiated to buy time for the hemostatic agents to take effect. The patient's head was elevated to 30 degrees to facilitate venous drainage via the jugular system. Osmotherapy was initiated with Mannitol 20% (0.5 g/kg), administered as a 50 cc IV bolus over 20 minutes to draw edematous fluid from the cerebral parenchyma into the intravascular space. Seizure prophylaxis was established with a Phenytoin loading dose (15 mg/kg), followed by maintenance dosing, to prevent post-traumatic seizures, which would catastrophically increase cerebral metabolic demand ($CMRO_2$) and exacerbate intracranial pressure. This coordinated, parallel processing of hemostatic correction and neuro-protection allowed the patient to be safely transported to the operating theater for emergency decompressive craniotomy.

Table 1. Timeline of Clinical Events and Therapeutic Interventions (Zero-Delay Protocol)

TIMEPOINT	PHASE	STATUS / EVENT	THERAPEUTIC INTERVENTION	PROTOCOL TARGET
Day -3 to Day 0	PRODROME	Headache, progressive vomiting at home.	None (Prophylaxis discontinued 5 months prior).	*N/A*
Day 1, 08:00	ADMISSION	ED Arrival. GCS 9 (E2V2M5).	Airway assessment, IV Access established.	*Initial Resuscitation*
Day 1, 08:30	DIAGNOSTIC	CT Head: Left SDH, Midline shift. Labs: aPTT 98.4s, FVIII <1%.	Emergency Consultation (Neurosurgery & Hematology).	*Diagnosis Confirmed*
Day 1, 09:00	HEMOSTASIS	Pre-operative Stabilization.	**Factor VIII 1000 IU IV (Loading)**	***Target FVIII 100%***
Day 1, 09:30	ICP MGMT	ICU Admission.	Head up 30°, **Mannitol** 20% (0.5g/kg).	*Reduce Cerebral Edema*
Day 1, 21:00	MAINTENANCE	ICU Monitoring.	Factor VIII 1000 IU IV.	*Maintain FVIII >80%*
Day 2, 08:30	BOOST	1 Hour prior to Surgery.	**Factor VIII 1000 IU IV (Surgical Boost)**	*Cover Surgical Peak*
Day 2, 09:30	INDUCTION	Anesthesia Start.	**Tranexamic Acid** 250 mg IV. RSI (Propofol / Fentanyl / Atracurium).	*Hemodynamic Stability*
Day 2, 10:00	SURGERY	Decompressive Craniotomy & Evacuation.	TIVA: Propofol + **Dexmedetomidine** (0.3-0.5 mcg/kg/h).	*Maintain CPP / Reduce ICP*
Day 2, 21:00	POST-OP	ICU Recovery (Intubated).	Factor VIII 1000 IU IV.	*Prevent Re-bleeding*
Day 3	RECOVERY	Neurological Improvement. GCS E4V5M6.	Extubation.	*Wean Sedation*
Day 7	DISCHARGE	Stable. No deficits.	Discharged home.	*Resume Prophylaxis*

Table 2. Summary of Clinical Findings on Admission

Comprehensive assessment of neurological, hemodynamic, radiological, and laboratory parameters.

CATEGORY	PARAMETER	FINDING / VALUE	*CLINICAL INTERPRETATION*
Neurology	Glasgow Coma Scale (GCS)	9 (E2, V2, M5)	Moderate TBI; Stuporous; Threatened airway reflexes.
	Pupillary Response	3MM / 3MM (Reactive)	Brainstem reflexes intact; No uncal herniation yet.
	Motor Function	Localizes to pain; No lateralizing signs	Gross motor pathways preserved despite compression.
Vital Signs	Hemodynamics	BP: 119/75 mmHg HR: 109 BPM	Preserved CPP; Sinus Tachycardia (Stress/Pain/ICP response).
	Respiratory	RR: 18 bpm SpO2: 100% (15L NRBM)	Adequate oxygenation; No respiratory depression.
	Temperature	36.5°C	Afebrile; R/O infectious meningoencephalitis.
Radiology (CT Head Non-Contrast)	Lesion Characteristics	ACUTE SDH (Left)	Hyperdense crescentic collection; Frontotemporoparietal.
	Quantitative Metrics	Thickness: **8 mm** Midline Shift: 12 MM	Mass effect disproportionate to hematoma width.
	Associated Findings	Subfalcine Herniation Non-communicating Hydrocephalus	Critical intracranial hypertension; Imminent decompensation.
Laboratory	Coagulation Profile	aPTT: 98.4 SEC PT/INR: Normal	Isolated intrinsic pathway defect (Classic Hemophilia pattern).
	Factor Assay	Factor VIII: <1% ACTIVITY	Confirmed Severe Hemophilia A.
	Complete Blood Count	Hb: 10.2 g/dL (Microcytic) Plt: 250,000 /ML	Mild anemia (chronic/occult bleed); Primary hemostasis (platelets) intact.

Standard ASA monitors were applied (ECG, NIBP, SpO_2, capnography, temperature). Additionally, invasive arterial blood pressure monitoring was established via the right radial artery for beat-to-beat hemodynamic assessment. A urinary catheter was placed to monitor output given mannitol administration. A bispectral index (BIS) monitor was applied to titrate anesthetic depth, targeting a value between 40 and 60 to ensure hypnosis and suppress cerebral metabolic rate ($CMRO_2$).

The anesthetic management of a pediatric patient with severe Hemophilia A and acute intracranial hypertension demands a strategy that transcends standard pediatric protocols. The primary objective was to orchestrate a neuroprotective induction that balanced the competing needs of securing a threatened airway while preventing catastrophic spikes in intracranial pressure (ICP) or hemodynamic collapse (Figure 1). To achieve this, a modified rapid sequence induction (RSI) was utilized, prioritizing

hemodynamic stability over speed. The induction phase was designed to blunt the sympathetic surge associated with laryngoscopy, a stimulus known to potentially rupture fragile clots in the setting of spontaneous hemorrhage. Premedication involved a synergistic combination of Midazolam (1 mg) and Fentanyl (75 mcg or 3 mcg/kg) to provide anxiolysis and suppress the catecholamine release inherent to airway instrumentation. This was augmented by the administration of Lidocaine (30 mg or 1.5 mg/kg), a crucial adjunct employed not merely for analgesia, but to suppress the cough reflex and mitigate the reactive rise in ICP during tracheal intubation. The induction agent of choice was Propofol, titrated with extreme caution at a dose of 12.5 mg. This lower dosage reflects a clinical awareness of the patient's potential volume depletion following three days of vomiting; a standard bolus could have precipitated severe hypotension, compromising cerebral perfusion pressure (CPP). Neuromuscular blockade was achieved with Atracurium (12 mg or 0.5 mg/kg) to ensure complete paralysis and optimal intubating conditions. Airway management in a hemophilic child presents a dual risk: the standard risk of hypoxia and the specific risk of traumatic airway bleeding. Consequently, the trachea was intubated using a size 6.0 cuffed endotracheal tube facilitated by video laryngoscopy. This device was selected specifically to minimize cervical spine motion and reduce direct force on pharyngeal tissues, thereby preventing iatrogenic airway hematomas—a lethal complication in severe coagulopathy.

Following successful intubation, the anesthetic maintenance transitioned to a total intravenous anesthesia (TIVA) technique, deliberately avoiding volatile anesthetics. Volatile agents are known cerebral vasodilators that can uncouple cerebral blood flow (CBF) from metabolic demand, potentially increasing cerebral blood volume and ICP. In contrast, TIVA preserves cerebral autoregulation. A continuous infusion of Propofol (50–200 mcg/kg/min) was maintained to decrease the cerebral metabolic rate of oxygen ($CMRO_2$) and lower ICP. A distinctive feature of this protocol was the integration of Dexmedetomidine (0.3–0.5 mcg/kg/h). This alpha-2 agonist served as a multimodal neuroprotective agent, providing sympatholysis and reducing CBF without compromising oxygen delivery or causing ischemia. Furthermore, its opioid-sparing properties complemented the Fentanyl infusion (1 mcg/kg/h), ensuring profound analgesia with hemodynamic stability. Ventilation was strictly controlled using volume control settings to target a specific $PaCO_2$ window of 35-38 mmHg. This low-normal target was physiologically strategic: it induced mild cerebral vasoconstriction to reduce intracranial blood volume and pressure, yet avoided the profound hypocapnia that could lead to cerebral ischemia. This careful titration of carbon dioxide levels underscores the precision required when manipulating cerebral physiology in the presence of a space-occupying lesion.

A left frontotemporoparietal trauma flap craniotomy was performed. Upon dural opening, dark clotted blood was extruded under high pressure. The hematoma was meticulously evacuated. Hemostasis was achieved using bipolar cautery and oxidized cellulose. Crucially, no uncontrolled microvascular oozing was observed, confirming the efficacy of the preoperative factor VIII loading and maintenance protocol. Fluid therapy consisted of isotonic crystalloids (NaCl 0.9%) at a rate calculated to maintain euvolemia (1500 ml/24h equivalent). No intraoperative transfusion of packed red cells (PRC) or fresh frozen plasma (FFP) was required as hemodynamics remained stable (Mean Arterial Pressure > 65 mmHg) and the surgical field remained dry.

To strictly define minimal blood loss in this pediatric patient, we calculated the estimated total blood volume (EBV) and the maximum allowable blood loss (ABL) to guide transfusion decisions; Patient Weight: 25 kg; Pediatric EBV Factor: 75 mL/kg (Standard for 11-year-old male); Calculated EBV: 25 kg x 75 mL/kg = 1,875 mL. The patient's preoperative Hemoglobin (Hb) was 10.2 g/dL. We established a

restrictive transfusion trigger of Hb 8.0 g/dL (Hematocrit approx. 24%) to maintain oxygen-carrying capacity without risking viscosity-related stasis. Using the standard ABL formula: ABL = (EBV x (Hct_initial - Hct_target)) / Hct_average ABL = (1875 x (30.6 - 24.0)) / 27.3 = 453 mL. The intraoperative estimated blood loss was less than 100 mL (< 6% of EBV), which was significantly below the allowable limit. This quantitative assessment confirms the efficacy of the hemostatic protocol and justifies the decision to withhold blood transfusion.

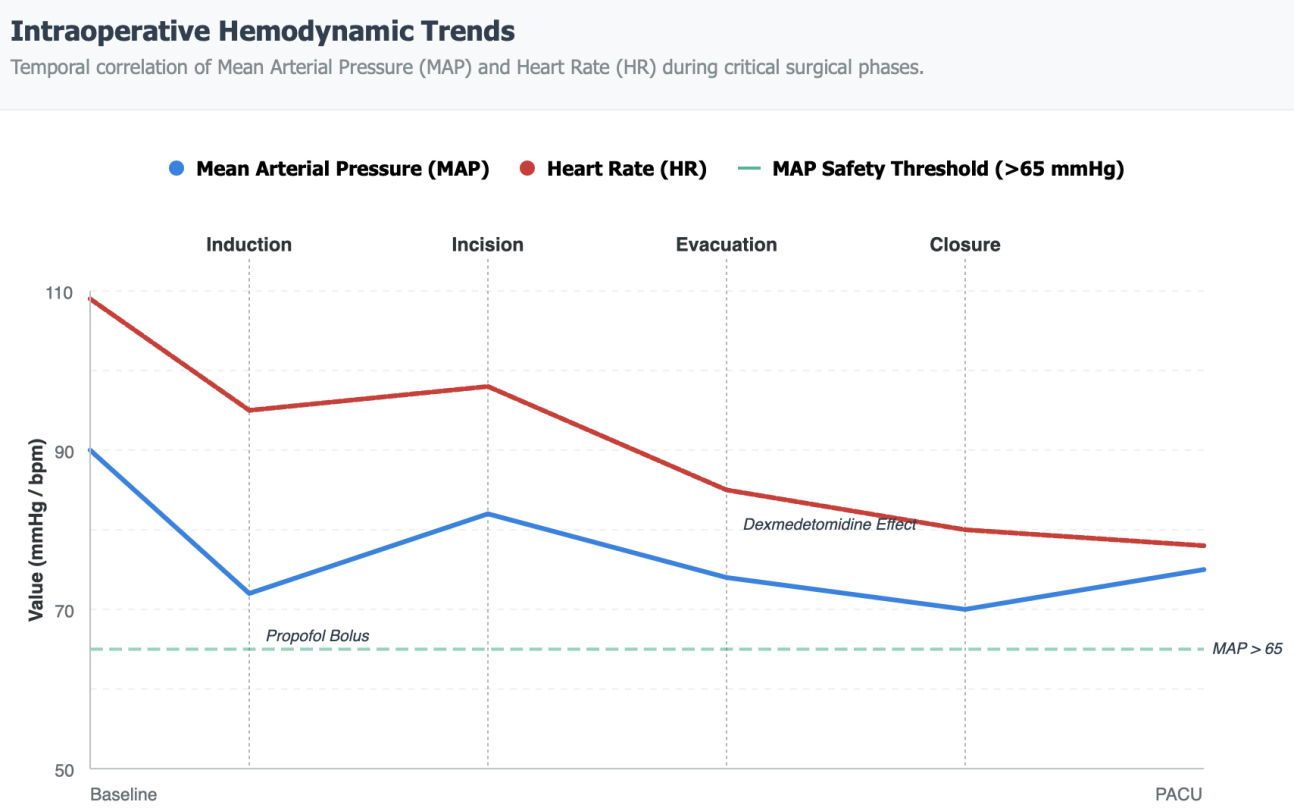

Figure 1. Intraoperative hemodynamic trends.

The patient was transferred to the ICU intubated and sedated for neuro-observation. Factor VIII (Koate) 1000 IU was continued every 12 hours (21:00 PM and 09:00 AM) to maintain trough levels >80%. Dexmedetomidine was continued for 24 hours to facilitate a smooth extubation. This allowed for the patient to be awakened without coughing or bucking (Valsalva maneuvers), which could spike ICP and risk re-bleeding. The patient was successfully extubated on Post-operative Day 1. The GCS improved to E4V5M6 (15/15) by Day 3. No re-bleeding or new neurological deficits were observed. The patient was discharged on Day 7 with a comprehensive plan for long-term hematology follow-up and resumption of prophylaxis.

3. Discussion

The successful management of the case presented herein underscores the critical, high-stakes intersection of hematology and neuroanesthesiology.

Spontaneous subdural hematoma (SDH) in the context of severe Hemophilia A represents a unique and perilous clinical entity that is physiologically distinct from the traumatic SDH typically encountered in pediatric trauma centers.[11] While the radiological appearance—a crescentic hyperdensity exerting mass effect—may be identical, the underlying pathophysiology dictates a fundamentally different surgical and anesthetic approach. In traumatic SDH, the pathology is often a mechanical disruption of a bridging vein due to acceleration-deceleration forces. In hemophilia, the bleeding source is frequently microvascular, diffuse, and relentless. Consequently, surgical hemostasis is not merely a matter of mechanical ligature or cautery; it is entirely dependent on the restoration of the patient's intrinsic coagulation factors. Without this hematological floor, the neurosurgeon is essentially operating on a patient who cannot seal the micro-breaches caused by the surgery itself.[12]

To understand the gravity of this case, one must appreciate the molecular failure at the heart of Hemophilia A. In patients with severe disease (Factor VIII <1%), the coagulation cascade is arrested at a critical juncture. Factor VIII acts as a cofactor for Factor IXa; together, they form the Tenase complex on the surface of activated platelets. This complex is the engine of thrombin generation, responsible for the propagation phase of coagulation that leads to a stable fibrin clot. In the absence of factor VIII, the initial platelet plug forms (primary hemostasis), but it lacks the fibrin meshwork required to stabilize it (secondary hemostasis). Consequently, the cerebral vasculature in these patients is exceptionally vulnerable to minor biomechanical stresses.[13] The spontaneous nature of these hemorrhages is often a misnomer. It is frequently the result of trivial trauma—physiological stressors as minor as the shear force generated by a coughing fit, sneezing, or, as seen in our patient, projectile vomiting. These events cause micro-tears in the fragile dural vessels that, in a healthy child, would be instantly sealed by the Tenase complex. In the hemophiliac, they result in uninhibited, slow-flow bleeding.[14]

This pathophysiology explains the distinct temporal profile observed in this case. Unlike the rapid, arterial accumulation of an epidural hematoma that leads to decompensation in hours, the spontaneous SDH in hemophilia is often insidious.[15] The bleeding is slow but progressive. The patient enters a symptom-free interval where the hematoma expands, utilizing the available intracranial reserve capacity. It is only when the intracranial volume reaches the critical threshold described by the Monro-Kellie Doctrine that the patient decompensates. Our patient's three-day prodrome of headache and vomiting represents the exhaustion of compensatory mechanisms (CSF displacement and venous efflux). The sudden loss of consciousness marked the point where the intracranial compliance curve became vertical, leading to the 12 mm midline shift and subfalcine herniation. Understanding this slow-motion crisis is vital for the clinician, as it emphasizes that by the time neurological deficits appear, the physiological margin for error is non-existent.

The management of this patient was anchored in a factor-first philosophy. Our protocol emphasized that in the hierarchy of resuscitation, correcting the coagulopathy must precede—or at least run parallel to—airway and hemodynamic management. Attempting invasive neurosurgical procedures, or even intubation, in a patient with <1% factor VIII activity invites catastrophic iatrogenic hemorrhage. The decision to target 100% factor VIII activity is supported by the most rigorous World Federation of Hemophilia (WFH) guidelines for life-threatening hemorrhage. While lower targets (50-80%) might suffice for hemarthrosis, the central nervous system (CNS) has zero tolerance for re-bleeding. The calculation utilized (0.5 x weight x target rise) proved robust and accurate. It is worth noting that we administered this as a rapid bolus. In the context of herniation, time is brain, and the pharmacological decompression provided by restoring the clotting ability is as vital as the surgical decompression.[16]

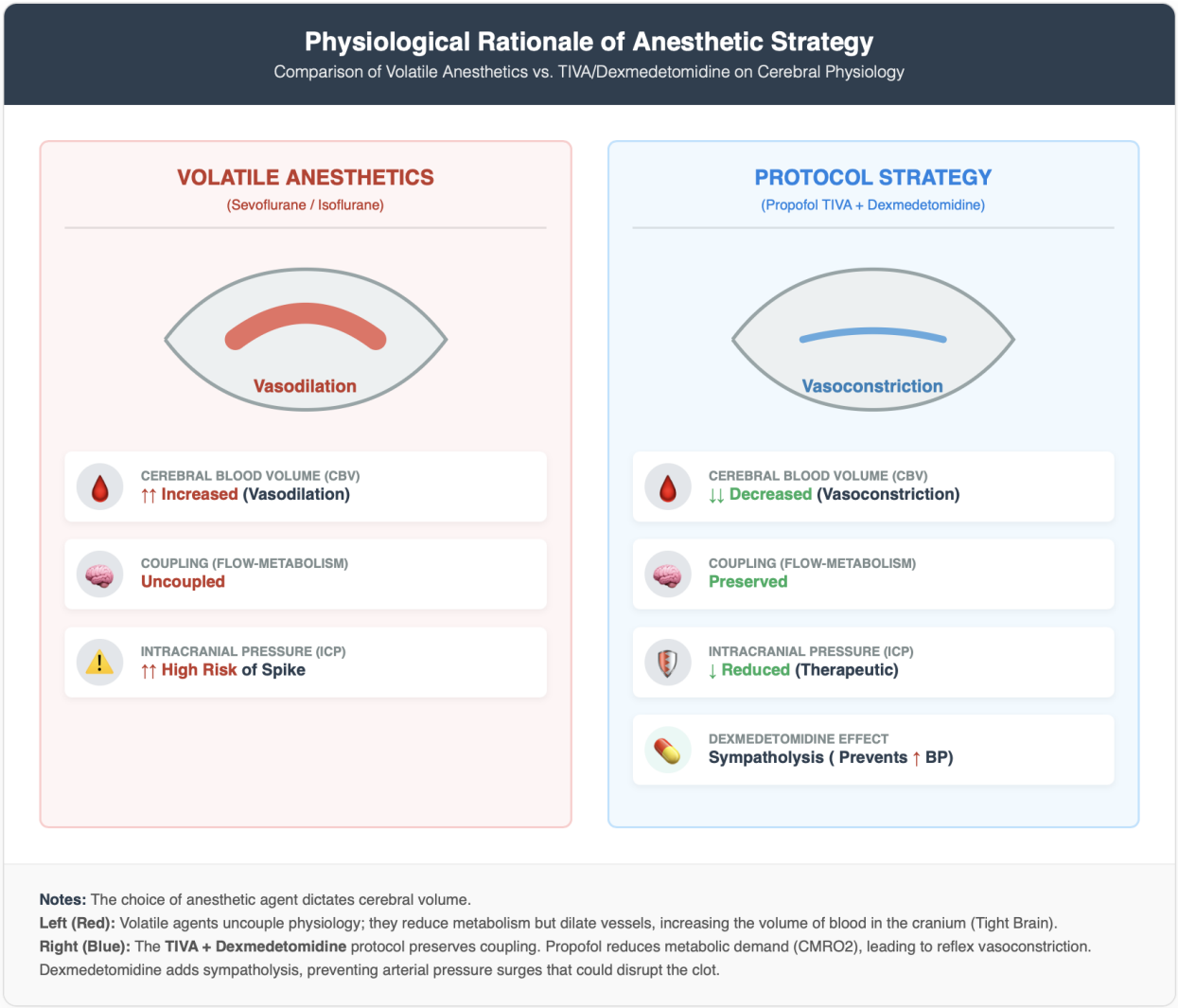

Figure 2. Physiological rationale of anesthetic strategy.

Furthermore, the inclusion of tranexamic acid (TXA) provided a critical biochemical safety net. The brain is an organ rich in thromboplastin and tissue plasminogen activator (tPA), making the surgical field inherently hyper-fibrinolytic. In a hemophiliac, any clot formed is structurally weak due to the delayed thrombin burst. If that weak clot is subjected to accelerated fibrinolysis, re-bleeding is inevitable.[17] While the CRASH-3 trial focused on traumatic brain injury in the general population, establishing TXA's survival benefit in mild-to-moderate TBI, we extrapolated these findings to our specific context. By inhibiting the conversion of plasminogen to plasmin, TXA stabilizes the fragile clot formed by the exogenous factor VIII, effectively locking in the hemostasis achieved pharmacologically. This dual-pronged approach—providing the building blocks of the clot (Factor VIII) and preventing its destruction (TXA)—was likely decisive in achieving the dry surgical field noted intraoperatively.

The anesthetic management of acute SDH is an exercise in physiological balancing: the anesthesiologist must provide profound hypnosis and analgesia while simultaneously shrinking the brain and maintaining perfusion.[18] The choice of anesthetic agents in this case was pivotal to the favorable

outcome (Figure 2). We deliberately employed a Total Intravenous Anesthesia (TIVA) technique, strictly avoiding volatile anesthetics such as Sevoflurane or Isoflurane. The rationale lies in the fundamental coupling of cerebral physiology. Volatile anesthetics are potent intrinsic cerebral vasodilators. In a dose-dependent manner, they uncouple cerebral blood flow (CBF) from the cerebral metabolic rate of oxygen ($CMRO_2$). This means that even as the brain's metabolism slows down, the blood vessels dilate, increasing cerebral blood volume (CBV). In a patient with a tight brain and a 12 mm midline shift, any increase in CBV translates directly to an exponential rise in ICP, potentially precipitating trans-tentorial herniation.

Conversely, propofol acts as a cerebral vasoconstrictor that preserves flow-metabolism coupling. It significantly reduces $CMRO_2$, which in turn leads to a reflex reduction in CBF and CBV, effectively relaxing the brain and lowering ICP. This pharmacological volume reduction facilitates surgical access and reduces the need for aggressive retractor pressure, which is particularly dangerous in coagulopathic tissue. However, the cornerstone of our neuroprotective strategy was the integration of Dexmedetomidine. Traditionally used as a sedative in the ICU, its intraoperative application in pediatric neurosurgery offers distinct advantages. Dexmedetomidine is a highly selective alpha-2 adrenergic agonist that acts on the locus coeruleus to provide sedation that mimics natural non-REM sleep.[19] Unlike opioids or benzodiazepines, Dexmedetomidine blunts the central sympathetic outflow. Surgery, particularly dural opening and cranial pinning, elicits a profound sympathetic surge. In a patient with a fresh clot, an arterial pressure spike could cause a hydrostatic blowout of the hemostatic seal. Dexmedetomidine attenuates this response, maintaining a stable hemodynamic corridor. Studies indicate that Dexmedetomidine reduces CBF and ICP without compromising cerebral oxygenation, making it an ideal adjunct to Propofol. Perhaps its most critical role was during extubation. Coughing, bucking, or straining (Valsalva maneuvers) during emergence can spike intracranial venous pressure to 40-50 mmHg. In a hemophiliac, this venous engorgement is a recipe for catastrophic re-bleeding. Dexmedetomidine allows for cooperative sedation—the patient can be awakened, follow commands, and be extubated while remaining calm and chemically sympatholyzed. This soft landing is essential for preserving the surgical result.[20]

The ventilatory strategy was equally rigorous. Historically, hyperventilation was the reflexive response to high ICP. However, we strictly avoided prophylactic hyperventilation. Reducing $PaCO_2$ below 30 mmHg causes severe hypocapnic vasoconstriction, which can reduce CBF to the point of ischemia. In the penumbra surrounding the hematoma, where perfusion is already compromised, this additional vasoconstriction can convert reversible injury into permanent infarction. Therefore, we targeted a tight Goldilocks window of normocapnia ($PaCO_2$ 35-38 mmHg). This induces just enough vasoconstriction to lower ICP and facilitate dural closure, but not enough to threaten oxygen delivery. Hemodynamically, the goal was to maintain cerebral perfusion pressure (CPP), defined as mean arterial pressure (MAP) minus intracranial pressure (ICP). In the setting of raised ICP, the MAP must be maintained at normal or high-normal levels to drive blood into the cranium. We utilized invasive arterial monitoring to ensure beat-to-beat management of MAP, targeting levels >65 mmHg (appropriate for age) to ensure adequate perfusion pressure without risking hydrostatic disruption of the clot.

While the outcome of this case was favorable, we must acknowledge the limitations of our approach. The primary limitation was the lack of viscoelastic point-of-care testing, such as Thromboelastography (TEG) or Rotational Thromboelastometry (ROTEM). We relied on standard coagulation assays (aPTT) and factor VIII levels. While these tell us that the blood can clot, they do not assess the quality, strength, or stability of the clot in real-time. TEG would have provided valuable data on the maximum amplitude (MA) of the clot and the degree of

fibrinolysis (Ly30), allowing for more precise titration of TXA and blood products. In resource-limited settings, the formula-based approach we used is necessary, but in resource-rich environments, goal-directed therapy guided by TEG should be the standard. Additionally, as a single case report, the generalizability of our findings is inherently limited. However, the physiologic principles applied—restoring the Tenase complex, preserving autoregulation, and preventing secondary injury—are universal. Future research should focus on the development of standardized massive hemorrhage protocols specifically designed for congenital bleeding disorders in neurosurgery. These protocols should move beyond simple factor replacement and incorporate a holistic hematology-anesthesia checklist that includes specific anesthetic agents, ventilatory targets, and emergence strategies.[19,20]

4. Conclusion

The management of spontaneous Subdural Hematoma in pediatric Hemophilia A is a clinical precipice where the margin for error is non-existent. It carries a high risk of mortality due to the dual and synergistic threats of uncontrolled hemorrhage and fulminant intracranial hypertension. This case serves as proof-of-concept that a rigorous, protocol-based approach can achieve favorable outcomes even in the most severe presentations involving herniation. The zero-delay protocol implemented here was not merely a sequence of orders, but a philosophy of parallel processing. The success of this case was not determined by the surgical skill alone, but by the meticulous physiological conditioning of the patient before, during, and after the incision. Key determinants of success identified in this protocol included: (1) Immediate Pharmacological Decompression: The recognition that Factor VIII replacement is the first step of resuscitation. Targeting 100% activity prior to surgical incision effectively normalized the patient's physiology, converting a high-risk hemophiliac into a standard trauma patient for the duration of the surgery; (2) Clot Stabilization: The proactive use of Tranexamic acid (TXA) as an adjunct to combat the hyper-fibrinolytic environment of the injured brain, ensuring that the hemostasis achieved was durable; (3) Neuroprotective anesthesia (TIVA + Dexmedetomidine): The rejection of volatile anesthetics in favor of a Propofol-based TIVA strategy preserved cerebral autoregulation and reduced metabolic demand. The addition of Dexmedetomidine provided a unique advantage by blunting the sympathetic surges of surgery and facilitating a smooth, cough-free emergence, thereby protecting the patient from the lethal complication of re-bleeding during the recovery phase. We aggressively advocate for the integration of this hematology-anesthesia protocol in emergency settings, managing congenital bleeding disorders. The collaboration between the anesthesiologist, neurosurgeon, and hematologist must be seamless. The traditional siloed approach—where the hematologist manages the factor, the surgeon manages the brain, and the anesthesiologist manages the sleep—is insufficient for this pathology. Only when hemostatic correction occurs in precise synchrony with airway control and neuro-resuscitation can we expect to alter the natural history of this devastating condition.

5. References

1. Scalia G, Porzio M, Costanzo R, Iacopino DG, Galvano G, Nicoletti GF, et al. Spontaneous spinal epidural hematoma in a pediatric hemophiliac. Surg Neurol Int. 2023; 14(69): 69.
2. Lin M-H, Kung C-H, Hsieh KL-C. Spontaneous intramural hematoma of the small bowel in a hemophilia A patient. Intern Emerg Med. 2024; 19(3): 863–5.
3. Martínez-García E, Roman-Chávez PD, Martínez-Canseco KA, Galván-Ortiz MA, Figueroa-Zariñana VH. Spontaneous retroperitoneal hematoma, an uncommon case of acute surgical abdomen in a patient with hemophilia: case report. Int J Res Med Sci. 2024; 12(6): 2106–9.

4. Oh SH, Shin J-W, Kee AW, Seo H-S, Kim H-S, Moon S-H, et al. Cervical epidural hematoma and cord compression in a hemophilia A patient with elevated factor VIII inhibitor levels after minor trauma: a case report. J Korean Soc Spine Surg. 2025; 32(2): 37.
5. Shinohara M, Konishi K, Kojima S, Kanda H, Yamaguchi T, Kaburagi T, et al. Small-intestinal obstruction due to intramural hematoma in a child with hemophilia A. J Pediatr Gastroenterol Nutr. 2025; 81(4): 1120–1.
6. Al Mustafa W, Aldossari SK, Afridi MA, Al-Dairi W. Profile and management of traumatic orbital hematoma in a child with hemophilia C: a case report. Cureus. 2025; 17(11): e97475.
7. Yanagihara K, Mochizuki Y, Katsube A, Kamio T, Akiyama M. Life-threatening pharyngolaryngeal hematoma in a patient with hemophilia A. Cureus. 2025; 17(4): e83147.
8. Marku L, Rajjoub H, Climov M, Uygur HS. Conservative management of a bullous hematoma in the case of von Willebrand disease and hemophilia A. Eplasty. 2025; 25: QA5.
9. Mistry T, Dogra N, Chauhan K, Shahani J. Perioperative considerations in a patient with hemophilia A: a case report and review of literature. Anesth Essays Res. 2017; 11(1): 243–5.
10. Chamos C, Yates M, Austin S, Bapat V. Minimally invasive aortic valve replacement in a patient with severe hemophilia A. J Cardiothorac Vasc Anesth. 2017; 31(4): 1378–80.
11. B F. Thoracic epidural for postoperative pain control after video-assisted thoracoscopic surgery (VATS) lobectomy in a patient with mild hemophilia A. Austin J Anesth Analg. 2017; 5(1).
12. Pérez-Moreno DP, Buitrago CL. Pain in hemophilia patients: Assessment and management in a fourth level hospital. Case series. Colomb J Anesthesiol. 2017; 45: 24–9.
13. Jimbo I, Uzawa K, Tokumine J, Mitsuda S, Watanabe K, Yorozu T. Ultrasonographic identification of the cricothyroid membrane in a patient with a difficult airway as a result of cervical hematoma caused by hemophilia: a case report. BMC Anesthesiol. 2019; 19(1): 124.
14. Isaacs J, Welsby IJ, Schroder JN, Onwuemene OA. Activated coagulation time and Hepcon protamine titration device to manage unfractionated heparin during cardiopulmonary bypass in a hemophilia A patient on emicizumab. J Cardiothorac Vasc Anesth. 2021; 35(11): 3299–302.
15. Ryoo S-H, Kwon D, Lee J-H, Seo K-S, Kim HJ, Karm M-H. An anesthetic management of head and neck cancer reconstructive surgery in a patient having hemophilia A: a case report. J Dent Anesth Pain Med. 2021; 21(3): 261–8.
16. Ogawa K, Honda Y, Kubota R, Hirota K, Yamazaki M. Perioperative management of emergent craniotomy in an acquired hemophilia A patient with remarkable coagulopathy: a case report. J Jpn Soc Clin Anesth. 2021; 41(5): 451–6.
17. Nurumbetova O, Tuç Ö, Çekmen N. An anesthetic management of patient with hemophilia A undergoing recurrent perianal abscesses. Bali J Anesthesiol. 2022; 6(4): 243–6.
18. Torfs A, Chardonnal L, Meunier S, Désage S, Henaine R, Lilot M. Perioperative hemostatic management of a newborn with hereditary hemophilia A undergoing emergent surgery for dextro-transposition of the great arteries. J Cardiothorac Vasc Anesth. 2022; 36(10): 3855–8.

19. Rajasekhar A, Arnaoutakis GJ, Janelle GM, Harris N, Wynn T, Anderson RD, et al. Multidisciplinary management of a hemophilia A patient requiring coronary artery bypass graft surgery. J Cardiothorac Vasc Anesth. 2022; 36(2): 534–8.
20. Lowell AE, Calgi MP, Caruso JJ, Man LM, McNeil JS. Perioperative management of hemophilia patients. Curr Anesthesiol Rep. 2024; 14(3): 354–65.