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# HIGH-RISK SURGERY IN SEVERE HAEMOPHILIA B: PERIOPERATIVE MANAGEMENT WITH ALBUTREPENONACOG ALFA

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**Background.** Perioperative management in severe haemophilia B remains challenging, particularly in patients undergoing surgery at high bleeding risk. In this setting, extended half-life recombinant factor IX products may offer effective haemostatic coverage while reducing treatment burden and allowing a more flexible dosing schedule. Clinical experience in complex surgical settings, however, is still limited.

**Case Report.** We report the case of a 72-year-old man with severe haemophilia B receiving prophylaxis with albutrepenonacog alfa 3000 IU weekly since November 2021, after previous treatment with standard half-life factor IX. His medical history included major craniofacial trauma approximately 40 years earlier requiring surgery, osteoarthritis, previous HCV infection. In late June 2024, he presented to the Emergency Department with painful left frontal swelling associated with local erythema, in the absence of fever. Brain computed tomography showed multiple defects involving the inner and outer tables of the left frontal bone, while the frontal sinus and anterior ethmoidal cells were partially occupied by hyperdense material. Initial antibiotic treatment with amoxicillin/clavulanate resulted in only partial improvement. Subsequent otolaryngological and neurosurgical evaluation, together with contrast-enhanced brain magnetic resonance imaging, confirmed inflammatory-infectious involvement of the left frontal region and anterior ethmoidal cells, communicating with an overlying subcutaneous collection, thus leading to the indication for combined surgical treatment. The patient was admitted on July 29, 2024 for a procedure considered at high haemorrhagic risk. Surgery was performed on August 1, 2024 and included uncinectomy, anterior ethmoidectomy with drainage of purulent material and thick grayish secretions suggestive of fungal infection, and frontal sinusotomy with surgical toilette extended to the most lateral and cranial compartment, with drainage of two mucocele cavities. Histological examination showed respiratory mucosa with inflammatory changes and aggregates of fungal hyphae. Perioperative haemostatic management was planned by the Haemostasis and Thrombosis Service. The last scheduled home dose of albutrepenonacog alfa had been administered on July 29. Preoperative laboratory testing showed factor IX activity around 50%. A replacement regimen with albutrepenonacog alfa 4000 IU intravenously approximately 30 minutes before surgery, followed by 3000 IU at 24, 48, and 72 hours, was initially planned and subsequently adjusted according to serial factor IX monitoring. Factor IX activity reached 107% at 24 hours after surgery, was 75.7% at 48 hours, 68% at 96 hours, 91.6% on postoperative day 5, and 60.2% on postoperative day 6, prompting additional 2000 IU doses when required (Fig. 1). No perioperative or postoperative bleeding complications occurred. Postoperative brain CT and MRI showed no evidence of haemorrhage, and the clinical course was uneventful.

**Conclusions.** This case highlights the practical usefulness of albutrepenonacog alfa in the perioperative management of severe haemophilia B undergoing high-bleeding-risk surgery. A tailored regimen based on serial factor IX assessment, within a multidisciplinary setting, allowed effective haemostatic control and safe completion of a complex surgical procedure without haemorrhagic complications.

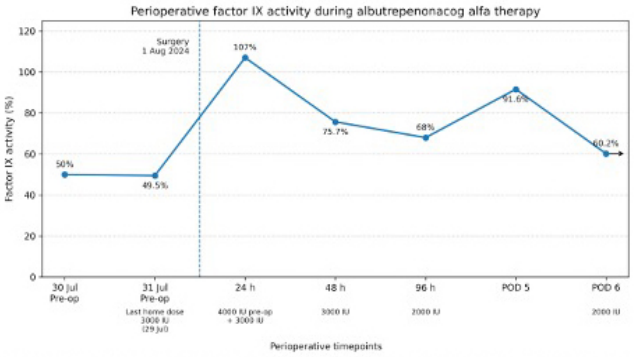



**Figure 1.** Perioperative trend of factor IX activity during replacement therapy with albutrepenonacog alfa. Following the preoperative dose, factor IX activity increased to 107% at 24 hours and remained within an adequate haemostatic range throughout the postoperative period, with further administrations guided by laboratory monitoring. No bleeding complications occurred.