**Título:** TO B OR NOT TO B – THE PERIOPERATIVE MANAGEMENT OF A PATIENT WITH HEMOPHILIA B UNDERGOING TOTAL KNEE ARTHROPLASTY

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**Área Terapêutica/Tema:** Transfusão e Hemostase (Transfusion and Haemostasis)

**Resumo:**

Hemophilia B (HB), termed Christmas disease, is a congenital deficiency of factor IX. Patients with factor IX <1% of the normal value (<0.01 IU/mL) are considered to have severe hemophilia, with the most common complication being hemarthrosis. As they often pose several anesthetic challenges (1), the goal of this case report is to discuss the perioperative management of a patient with this condition undergoing major orthopedic surgery (2).

A 32-year-old patient, ASA III, with severe HB presents for elective total knee arthroplasty due to severe hemophiliac arthropathy. The patient was monitored using standard ASA practices with the addition of an arterial line, bispectral index and train-of-four monitorization, as well as the placement of two large bore peripheral accesses. A rapid sequence induction followed by videolaryngoscopy was performed. Immediately after anesthetic induction, a loading dose of factor IX and tranexamic acid were administered before a tourniquet was placed. Intraoperatively, a severe pain multimodal analgesia regimen was prescribed, except for non-steroid anti-inflammatory drugs (NSAIDs). The surgical procedure underwent without any intraoperative intercurrences. Postoperative pain management was performed with a tramadol and droperidol perfusion associated with a femoral and sciatic single shot nerve block. Additionally, hemoglobin, coagulation and factor IX levels were monitored daily so that the necessary therapeutic adjustments could be made accordingly. Due to the development of postoperative anemia, ferric carboxymaltose was prescribed and red blood cells transfusions given until an 8g/dL or higher hemoglobin was achieved.

Throughout the entirety of the perioperative time, a multidisciplinary approach involving the immunohemotherapy department was carried out, ensuring the best medical practices.

This case report enhances the importance of a structured approach to hemophiliacs undergoing orthopedic surgery. These patients present with several potentially critical anesthetic moments (1), namely the necessary eviction of neuroaxial techniques and the likelihood for airway hemorrhage(2), a possible life-threatening condition. Furthermore, they also present analgesic limitations, (such as exclusion of all NSAIDs and any deep peripheral nerve blocks) which must be carefully managed in order to better approach pain control in these particular patients.

Overall, this disease and its restrictions pose an anesthetic puzzle from beginning to end.

Key words: hemophilia B; factor IX; total knee arthroplasty; perioperative management.

References:

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