P3. EVOLUTION IMMUNOSUPPRESSIVE THERAPY WITH MONOCLONAL ANTIBODIES IN A PATIENT WITH SEVERE HEMOPHILIA A, FACTOR VIII INHIBITOR.

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Hemophilia A is an inherited blood disorder, because of mutation HEMA gene on the X chromosome, characterized by a deficiency of blood clotting protein, Factor VIII (FVIII), accompanied by hemorrhagic manifestations typical. Patients with severe hemophilia produce less than 1% of normal clotting factor affected the administration of FVIII are addicted to treat or prevent episodes of bleeding.They can inherite allo-anticorpiin after the administration of FVIII, a situation that requires special care because of the high risk of mortality increaseang bleeding complications develop.

This paper presents the case of a patient diagnosed at severe haemophilia A-form, with massive bleeding that despite administration of FVIII rFVII maintaince the bleeding active hemostatic serious (hemohidrotorax 2/3 lower left, retroperitoneal hematoma, retrogastric the fall in hemoglobin to 2.6 g%). It raises suspicion of the presence of factor VIII inhibitors, confirmed the investigation specialist (80 u Bethesda) needing treatment immunosuppressive monoclonal antibody and Ciclofosfamidasi desensitization treatment with high doses of FVIII.

The management of inhibitors of the hemofilici patients is an ongoing challenge that requires induction of immune tolerance by using a technique the most successful desensitization observed in patients with low titer inhibitors (<5 u Bethesda), which are treated immediately after detecting a allo-antibodies, and include the use of immunosuppressive therapy and also with repeated infusions of FVIII high titer with high results in studies to date.