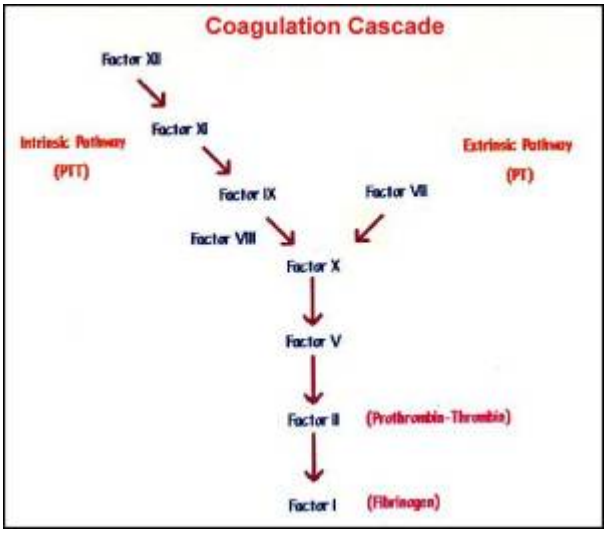
Factor V deficiency



Factor V deficiency is also known as Owren's disease or parahemophilia. It's a rare bleeding disorder that results in poor clotting after an injury or surgery. Factor V deficiency shouldn't be confused with factor V Leiden mutation, a much more common condition that causes excessive blood clotting.

Factor V deficiency may also occur at the same time as factor VIII deficiency, producing more severe bleeding problems. The combination of factor V and factor VIII deficiencies is considered to be a separate disorder.

FV inhibitors are a common complication of bovine thrombin exposure that can have devastating clinical consequences. Transfusion medicine specialists and hematologists can play a critical role in reducing the incidence of FV inhibitors by educating the medical community about safer alternative fibrin sealants [1].

Case reports

Meidert et al., report the case of an 82 year old woman with incidentally diagnosed severe factor V deficiency, who developed a symptomatic chronic subdural hematoma, requiring burrhole craniostomy. Successful management was achieved by a multidisciplinary approach. Preoperatively, factor V activity was increased from 2 % to 50 % by administration of 25 ml/kg body weight of fresh frozen plasma (FFP) over 30 minutes under close cardiopulmonary monitoring on ICU. Straight afterwards, the patient was transferred to the operating room where surgery was performed under general anesthesia. Burr-hole craniostomy could be performed without perioperative complications. In the postoperative days there was no relevant recurrence of the subdural hematoma in the follow-up CT scans under frequent control of coagulation parameters. However, despite further transfusion of FFP, factor V activity did not increase above 16%.

The patient was discharged without any neurological deficits. In a hemostaseologic follow-up two months after surgery, factor V activity below 1% was confirmed with evidence of a factor V inhibitor in the modified Bethesda assay. Most likely, the patient suffered from an acquired form of factor V

deficiency with preformed antibodies that had been boosted by the initial treatment with FFP.

They conclude that in this rare bleeding disorder, intracranial surgery was successfully managed due to a thoroughly planned perioperative therapeutic strategy. However, if there is time prior to surgery, a full check-up of the bleeding disorder is advisable [2].

Cavum vergae bleed in a term neonate with severe factor V deficiency [3].

Lee et al., reported a newborn infant girl, born to consanguineous parents, with recurrent intracranial hemorrhage secondary to congenital factor V deficiency with factor V inhibitor. Repeated transfusions of fresh-frozen plasma (FFP) and platelet concentrates, administrations of immunosuppressive therapy (prednisolone and cyclophosphamide), and intravenous immunoglobulin failed to normalize the coagulation profiles. Exchange transfusion followed-up by administrations of activated Prothrombin complex concentrate and transfusions of FFP and platelet concentrates caused a temporary normalization of coagulation profile, enabling an insertion of ventriculoperitoneal shunt for progressive hydrocephalus. The treatment was complicated by thrombosis of left brachial artery and ischemia of left middle finger. The child finally died from another episode of intracranial hemorrhage 10 days after insertion of the VP shunt. [4].

References

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