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A 2 year old female with a history of Tetralogy of Fallot with pulmonary atresia and mild factor VII deficiency underwent a third redo sternotomy. At 6 days of life, her first operation consisted of unifocalization of the left and right branch pulmonary arteries (PA) and BT shunt creation. At 3 months of age she underwent redo sternotomy for complete repair due to hypoxia and failure to thrive. Both surgeries required delayed closure of the sternum but neither reported any significant coagulopathy or hemostasis issues. At 15 months age, patient had worsening exertional dyspnea and underwent cardiac catheterization for stenting and angioplasty of the PAs that was complicated by pulmonary hemorrhage. A hematology consult revealed that the patient had a congenital factor VII deficiency. The patient continued to have worsening symptoms and the decision was made to proceed with cardiac surgery. The hematologist recommended a regimen of recombinant factor VIIa (rFVIIa). Given the risk of thrombosis on cardiopulmonary bypass (CPB), recommendation for low dose bolus followed by continuous infusion of rFVIIa was made. After multidisciplinary discussion, the patient underwent redo sternotomy with CPB. She only received a bolus dose of rFVIIa at 20mcg/kg prior to incision & immediately after termination of CPB without continuous infusion. Postoperatively the patient was found to have oozing from line sites in the intensive care unit, and a bolus dose of rFVIIa 20mcg/kg was given followed by a continuous infusion which was titrated to maintain a goal factor VII level of 50-75% normal. This ultimately aided in resolution of the bleeding at invasive line sites. There were no complications related to her surgery despite the low factor VII levels postoperatively.

Factor VII deficiency is more commonly acquired due to liver failure, vitamin K deficiency, or secondary to massive transfusion. It is a rare congenital factor deficiency that has an autosomal recessive inheritance pattern with a prevalence of 1 in 500,000. Deficiency in which levels are <1% of normal can lead to severe bleeding in the form of hemarthroses and crippling arthropathy.

Studies of the use of rFVIIa and CPB are limited to case reports and those used during massive hemorrhage protocols. Upon completion of this presentation, participants should be able to:

- Describe acquired versus congenital factor VII deficiencies.
- Recognize the clinical & laboratory signs and symptoms of factor VII deficiency
- Debate the role of rFVIIa during pediatric CPB.
- Identify strategies to reduce perioperative blood loss in patients with factor VII deficiency.

1. Emiroğlu, O. et al., 2010. Pediatric cardiac surgery under cardiopulmonary bypass in factor VII deficiency. *The Turkish journal of pediatrics*, 52(1), pp.101–3.
 2. Matsuhisa, H. et al., 2010. Open-heart surgery in an infant with heterozygous factor VII deficiency. *Interactive cardiovascular and thoracic surgery*, 10(6), pp.1037–8.
 3. Orkin, S.H. et al., 2008. Nathan and Oski's Hematology of Infancy and Childhood. In pp. 1525–32, 1591–1620.
 4. Witmer, C.M. et al., 2011. Off-label recombinant factor VIIa use and thrombosis in children: a multi-center cohort study. *The Journal of pediatrics*, 158(5), pp.820–825.
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