Hematological Management of a Pediatric Cerebral Palsy Patient with Factor VII Deficiency for Major Orthopedic Surgery
Ashka Shah, MD Jamey Eklund, MD
University of Illinois Hospital and Health Sciences Center, Chicago, IL

BACKGROUND
- Congenital Factor VII deficiency is rare
- Autosomal recessive, mutation on Chromosome 13
- 1/500,000 people
- Factor VII levels less than 65%
- Levels have a weak correlation with clinical bleeding
- Symptoms: minor bleeding, cutaneous hemosiderosis, hemarthrosis, intracerebral hemorrhages
- Limited data on perioperative hematology management
  - Factor VII < 10% and history of bleeding: transfuse recombinant Factor VII (NovoSeven)
  - Heterozygous Factor VII deficient patients: no guidelines
  - Factor VII levels 20-30%: theoretically adequate for hemostasis

PAST MEDICAL HISTORY
- 17 y/o, 55 kg, who has been in foster care system since age 1 month old, presents with sparse medical records.
- Heterozygous Factor VII deficiency
- Possible Factor V Leiden
- Patient reported undergoing evaluation for Factor V Leiden given strong PTH, but no records were available
- Premature birth with in-utero demise of twin
- Spastic cerebral palsy
- Congenital heart disease status-post “neonatal heart surgery”
- Asthma
- Seizure disorder
- Developmental delay
- Possible dorsal rhizotomy as child

CASE PRESENTATION
HPE: 17 year-old boy was scheduled for bilateral distal femoral extension derotational osteotomies, bilateral patellar distal advancement and internal fixation, bilateral distal tibial-fibular derotational osteotomies with internal fixation, and bilateral hip open myotomies.
- Pre-Op:
  - Pre-operative Factor VII level was 41%
  - Reported daily gingival bleeding with brushing
  - Hematology consultation obtained
  - Recommended “transfusion of NovoSeven for clinically significant bleeding”
  - Significant post-operative blood loss was anticipated after removal of tourniquets
  - Concern for hypercoagulability should NovoSeven be administered in context of possible Factor V Leiden & near normal Factor VII level
  - Type and Cross obtained
  - Platelets, cryoprecipitate and NovoSeven were immediately available
- Intra-Op:
  - Intubation was achieved with video laryngoscopy to minimize airway trauma and bleeding.
  - An arterial line was placed for continuous blood pressure monitoring and frequent blood tests.
    - CBC, ABG, fibrinogen and coagulation studies: obtained on hourly basis after surgical incision
  - Transfused weight-based dose of fresh frozen plasma (FFP) prior incision to increase Factor VII levels
  - Initiated low-dose bolus followed by low-dose infusion of tranexamic acid (TXA) to minimize bleeding
  - A second weight-based dose of FFP was transfused three hours later
  - Hemodynamically stable throughout the case
  - Estimated blood loss of 150 mL
  - Estabulated in the operating room
- Post-Op:
  - Transferred to the Intensive Care Unit for recovery
  - Remained hemodynamically stable
  - No apparent anesthetic complications

REFERENCES