Successful resuscitation utilizing massive transfusion and recombinant human Factor VIIa in an extremely premature, VLBW infant with giant sacrococcygeal teratoma and hepatic rupture.

C. Hargrove, J. Farlo, F. Abdullah
Departments of Anesthesiology and Surgery, Children’s Hospital Los Angeles, Keck School of Medicine, The University of Southern California

Introduction: Fetal sacrococcygeal teratoma (SCT) associated with hydrops fetalis carries a poor prognosis. Very low birth weight (VLBW) and extreme prematurity are associated with significant morbidity and mortality. Hepatic rupture in an infant is frequently fatal. We report the case of a VLBW, extremely premature infant with SCT and hydrops fetalis who sustained a traumatic hepatic rupture during SCT resection and was successfully resuscitated with massive blood transfusion and recombinant human Factor VIIa (NOVO 7) without neurological sequelae.

Results: An 800g male infant with a pre-natally diagnosed giant sacrococcygeal teratoma was born by Caesarian section at 25 weeks gestation for evidence of hydrops fetalis. Initial Hgb was 8.3 gm/dL. Initial ABG revealed a metabolic acidosis with a pH of 6.99 and a BE of –14. The infant was stabilized in the NICU and underwent surgical PDA ligation on day five of life. Subsequently, the infant continued to demonstrate high-output cardiac failure, requiring hemodynamic support with dopamine infusion up to 20mcg/kg/min. His hematocrit decreased in association with increasing size of the teratoma. On day of life nine he underwent surgical resection of the teratoma via a combined anterior/posterior approach. Anesthesia was maintained with vecuronium and fentanyl infusion of 50mcg/kg/h. Hemodynamic stability was maintained with aggressive resuscitation with PRBCs, FFP and platelets as well as epinephrine and dopamine infusions. Near the end of the operation the patient experienced traumatic hepatic rupture. Following a 10 blood-volume transfusion, recombinant human Factor VIIa was administered with a decrease in bleeding. The right upper abdominal quadrant was packed with gauze and the abdomen closed. The patient received an additional 2 doses of Factor VIIa in the NICU along with additional blood products for evidence of ongoing bleeding. The coagulopathy and fibrinolysis improved and on POD #4 the patient was taken back to the OR for completion of the operation. Factor VIIa was again utilized in the treatment of surgical hemorrhage. The patient recovered and was discharged home at the age of three months on room air and PO feeds without evidence of intracranial hemorrhage, significant retinopathy or chronic lung disease.

Discussion: Neonatal liver hemorrhage is associated with very high mortality. There are few reports of survival after hepatic rupture in the newborn (1, 2). The neonatal liver is fragile and prone to rupture while the newborn coagulation system is immature and vulnerable to derangement (3). In our patient, this devastating event was successfully treated with abdominal packing and recombinant Factor VIIa to facilitate hemostasis.

Recombinant human coagulation Factor VIIa (rFVIIa, Novo Seven, Novo Nordisk, Copenhagen) is a genetically-engineered equivalent to plasma-derived Factor VIIa. It promotes hemostasis by activating the extrinsic pathway of the coagulation cascade (4), leading to thrombin generation and fibrin formation. Novo Seven is FDA-approved for the treatment of bleeding due to hemophilia. It has been used off-label, with anecdotally-described benefit, in a variety of clinical scenarios associated with bleeding. Prior use of Novo Seven in VLBW infants has been described (5, 6). To our knowledge this is the first report of its use for the successful treatment of hepatic rupture in a VLBW infant.

References: