Hemimegalencephaly is caused excess proliferation of neuronal tissue resulting in unilateral brain hypertrophy. It is characterized by medically refractory epilepsy resulting in significant developmental delay. The most effective treatment for this devastating disease is a hemispherectomy or functional hemispherotomy, which has significant peri-operative morbidity and mortality. Perioperative concerns include excessive blood loss, coagulopathy, hypothermia and hypokalemia. Surgery is delayed as long as possible and not attempted before 3-4 months of age.

32 4/7 gestation infant with prenatally diagnosed right hemispheric hemimegalencephaly and lissencephaly. Onset of seizures was noted shortly after birth and EEG showed status epilepticus. Multiple antiepileptics initiated and titrated however he remained with intractable epilepsy on 3 antiepileptics. He could not fixated on an object or track with his eyes. He could not feed. He moved bilateral arms and legs. Multiple family meetings held and family decided to pursue aggressive medical and surgical management. Gastrostomy tube placed to enable feeding. Decision made to pursue functional hemispherotomy when he reached a weight of 5 kg (44 weeks PGA). Surgery is delayed as long as possible and not attempted before 3-4 months of age.

**Intraoperative Management**
- Induction: Mask
- Lines: 2 PIVs, central line and arterial line
- Blood Products: pRBCs and FFP started pre-incision titrated to ABGs and a TEG
- Special medications: Tranexamic acid infusion
- Temperature management: warm room, betadine and cleaning solutions warmed, fluids warmed, Bair hugger.

**Postoperative Management & Follow up**
- Transported to ICU intubated
- Exubated POD#8, prolonged intubation secondary to airway edema, treated with steroids and diuretics
- Remains on preoperative antiepileptics (for 1 year) with a few post-operative seizures secondary to irritable brain
- Discharged POD #12
- Seizure free for 6 months, left neglect and hand weakness
  - Advancing to oral feeds
  - Tapering antiepileptics

**Discussion**
This extremely invasive surgery is challenging in a child of any age. At 44 week PGA and 5 kg our patient is one of the youngest reported to undergo a functional hemispherotomy. Hemodynamic stability was carefully maintained with meticulous surgical resection, titration of blood products, continuous infusion of TXA, temperature monitoring, and serial ABGs.

**References**

**Background**
Hemimegalencephaly is caused excess proliferation of neuronal tissue resulting in unilateral brain hypertrophy. It is characterized by medically refractory epilepsy resulting in significant developmental delay. The most effective treatment for this devastating disease is a hemispherectomy or functional hemispherotomy, which has significant peri-operative morbidity and mortality. Perioperative concerns include excessive blood loss, coagulopathy, hypothermia and hypokalemia. Surgery is delayed as long as possible and not attempted before 3-4 months of age.

**Case Presentation**
32 4/7 gestation infant with prenatally diagnosed right hemispheric hemimegalencephaly and lissencephaly. Onset of seizures was noted shortly after birth and EEG showed status epilepticus. Multiple antiepileptics initiated and titrated however he remained with intractable epilepsy on 3 antiepileptics. He could not fixated on an object or track with his eyes. He could not feed. He moved bilateral arms and legs. Multiple family meetings held and family decided to pursue aggressive medical and surgical management. Gastrostomy tube placed to enable feeding. Decision made to pursue functional hemispherotomy when he reached a weight of 5 kg (44 weeks PGA).

**Procedure**
Right anterior temporal lobectomy, amygdalohippocampectomy and modified peri-insular hemispherotomy (Villemure)
EBL 150 cc
Operative time 6.5 hours

**Case Illustration**

**Intraoperative Management**
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