Staged Hepatectomy for an Infant with Congenital Hepatoblastoma

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Introduction

Congenital hepatoblastoma is a rare tumor that requires early intervention including chemotherapy and surgery to improve survival. Previously thought to have very poor survival, a new surgical approach allows for significantly improved outcomes. Early involvement from the anesthesiology team, using the peripерioperative surgical home model, allows the anesthesiology team to guide patient care in the preoperative, intraoperative and postoperative periods. Below we present the youngest patient to undergo associated liver partition and portal vein ligation for staged hepatectomy (ALPPS).

Case

- 53 day-old male with large unresectable congenital hepatoblastoma diagnosed postnatally (Figure 1)
- Preoperative chemotherapy (Cisplatin, 5-FU and Vincristine) to decrease mass size.
- Future remnant liver (FRL) considered too small to be able to resect the tumor in one surgical procedure.
- Liver partition and portal vein ligation with two-stage hepatectomy attempted.

Anesthetic Challenges

- Staged procedure, 1 week apart, requiring multiple teams.
- Risk for major blood loss and hemorrhagic instability.
- Concern for insufficient liver function and coagulopathy with future small remnant liver.
- Anticipated prolonged post-operative intubation with delayed closure.
- Anticipated prolonged opioid requirement for analgesia requiring a weaning protocol.

Treatment Plan

- Multidisciplinary approach including: general surgery, surgical transplant team, anesthesiology, acute pain service, and radiology.
- ALPPS was initially performed. Using CT volumetric analysis, the liver growth was noted to have increased by 50% in 7 days, allowing for hepatoblastoma removal by partial hepatectomy (Table 1, Figure 2).

<table>
<thead>
<tr>
<th>Total liver volume (ml)</th>
<th>Before partition</th>
<th>After partition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tumor volume (ml)</td>
<td>67</td>
<td>73</td>
</tr>
<tr>
<td>Left lobe volume (ml)</td>
<td>85.8</td>
<td>129.2 (50% increase)</td>
</tr>
<tr>
<td>Segments 2/3 (ml)</td>
<td>45.7</td>
<td>87.3 (91% increase)</td>
</tr>
</tbody>
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Table 1. Volumetric studies based on CT scans obtained before and one week after hepatic partition and portal vein ligation for staged hepatectomy

Treatment Plan Cont.

- Anesthetic plan
  - Standard ASA monitors and NIRS.
  - Intravenous induction followed by endotracheal intubation.
  - Additional peripheral intravenous lines, arterial line and central venous line placement.
  - Available blood products with concern for massive transfusion.
  - Vaspressors and inotropic infusions.
  - Balanced anesthetic with sevoflurane, fentanyl, and desmedetomidine infusions.
  - CVP maintained on the low side to decrease blood loss and avoid liver congestion.
  - Delayed abdominal closure requiring postoperative mechanical ventilation.
  - An aggressive pain management model was used with anesthesia involvement in guiding the weaning of opioids and sedation with methadone and clonidine.

Discussion

- Early anesthesiology involvement (Perioperative Surgical Home Model) is important in complex surgical cases.
- ALPPS has emerged as an innovative two-stage hepatectomy with a short interval of 7-10 days between the first and second stages as compared to the classic interval of 8-12 weeks.
- The liver has the ability to regenerate and hypertrophy - 50% in 1 week.
- Liver resection is the first-line treatment for patients with malignancies, offering the best chance for long-term survival.
- Benefits of a shorter interval between the two procedures include fewer adhesions and therefore a less complicated second surgery, less time for tumor progression, and a single hospitalization.
- Meta-analysis showed ALPPS induces a greater degree of hypertrophy of FLR (future liver remnant) and has a greater likelihood of achieving complete tumor-free resection with acceptable margins.
- Three month follow-up: patient is doing well and gaining weight appropriately without signs of tumor recurrence.

Figure 1. MRI of tumor prior to chemotherapy at 3 days old.

Figure 2. CT images from which volumetric studies were obtained. Images A,B are obtained prior to hepatic partition and portal vein ligation. Images C,D are obtained 1 week later.

Figure 3. CT images with volumetric analysis for staged hepatectomy

References