Hypoplastic left heart syndrome (HLHS) patients may have associated extracardiac anomalies. Rarely requires surgical intervention prior to cardiac surgery. Myelomeningocele repairs performed on 2 HLHS at CHLA. Both proceeded with planned cardiac surgeries.

**Case Report**

- 3-kg male born at 36 6/7 weeks via cesarean section
- Fetal US & ECHO showed myelomeningocele & HLHS
- At birth, APGARS 7/8, umbilical arterial (UAC) & venous (UVC) lines placed
- Maintained on alprostadil in cardiothoracic intensive care unit
- On DCL #2, underwent myelomeningocele repair with VP shunt placement
- Standard ASA monitors, UAC, UVC & peripheral IV placed
- Inhalational induction with sevoflurane & vecuronium
- Uneventful endotracheal intubation via direct laryngoscopy
- FiO2 maintained at 0.21 through most of case
- Alprostadil continued & dopamine added at 10 mcg/kg/min
- Arterial blood gas (ABG) measurements taken every 30 min
- Fluid boluses given for worsening metabolic acidosis
- Estimated blood & CSF loss was 45 ml
- Returned intubated to CTICU after operation (~5 hrs)

**Introduction**

- HLHS occurs in 7.5% of patients with congenital heart defects
- Consists of a functional single ventricle with right ventricle supplying systemic & pulmonary circulation
- Maintained on alprostadil to keep ductus open prior to palliative staged repair [2]
- Myelomeningocele repairs on 2 HLHS children prior to cardiac surgeries completed at our institution without complications
- Recent article reports performing 8 general surgeries on unrepaired HLHS children with positive outcomes [3]

**Discussion**

- HLHS patients require a balance between pulmonary & systemic vascular resistance as well as a patent ductus due to their parallel circulation
- Meticulous attention to this balance may result in a positive outcome

**References**


**Table 1:** Select values from ABGs taken every 30 minutes

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