Emergent Bidirectional Glenn following Innominate Artery Injury and Considerations for Early Glenn Conversion

John P Scott, Robert A Niebler, Viktor Hraska, George M Hoffman

Medical College of Wisconsin, Children's Hospital of Wisconsin, Milwaukee, WI

BACKGROUND

Infants with Hypoplastic Left Heart Syndrome (HLHS) undergo staged surgical procedures to create a univentricular series circulation.

1. **Stage 1 (Norwood) –** Performed in 1st week of life. Neoaortic arch reconstruction with pulmonary blood flow (PBF) provided by a systemic artery to pulmonary artery shunt (modified Blalock Taussig (BT) shunt) or a ventricular to pulmonary artery conduit (Sano).
2. **Stage 2 (Bidirectional Glenn) –** Usually performed between 3-6 m/o. Converts Stage 1 parallel circulation to a partial series circulation with a superior cavopulmonary connection.
3. **Stage 3 (Fontan) –** Performed in toddlers (2-5 y/o). Redirects the inferior vena cava (IVC) blood flow to the pulmonary bed. Completes univentricular series circulation.

CASE REPORT

35-d/o male with HLHS s/p Norwood BT shunt who required early BDG following innominate artery injury with critically limited PBF.

- Cardiac Cath on DOL 34 for hypoxia - LPA stented with transiently improved oxygenation.
- DOL 35 acute hypoxia due to presumed stent thrombosis.
- Taken emergently for BT shunt revision, but innominate artery injury was found limiting viable positions for alternate BT shunt placement.

- From our database of early BDG in HLHS, we projected manageable oxygenation and pulmonary hemodynamics at his postnatal age, and therefore conversion to BDG circulation was performed.

- Post-BDG conversion
  - SpO_2 80-85%
  - Common atrial pressure 8-10 mmHg
  - Transpulmonary gradient 5-8 mmHg

- Mechanisms used to increase PBF and post-BDG SpO_2
  - Inhaled nitric oxide (iNO)
  - Permissive hypercapnea $PaCO_2$ 50-60 mmHg
  - Hematocrit maintained > 45%
  - Diuretics

- Extubated on POD 3 and supplemental oxygen was gradually weaned.
- Discharge on DOL 67 with SpO2 75-80 % on room air.

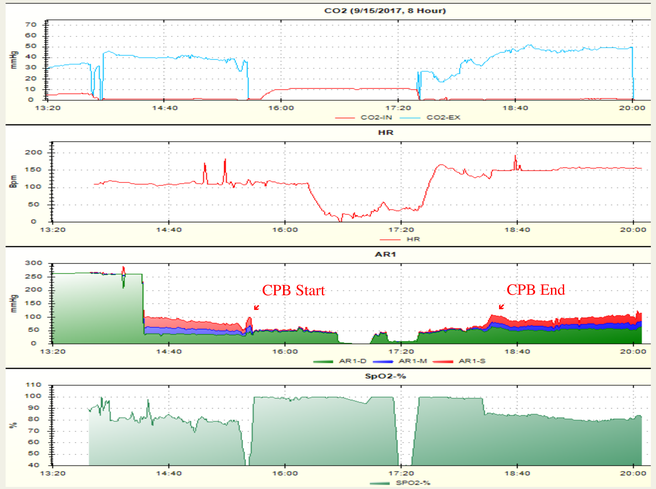

Figure 1. Key intraoperative vitals during BDG.

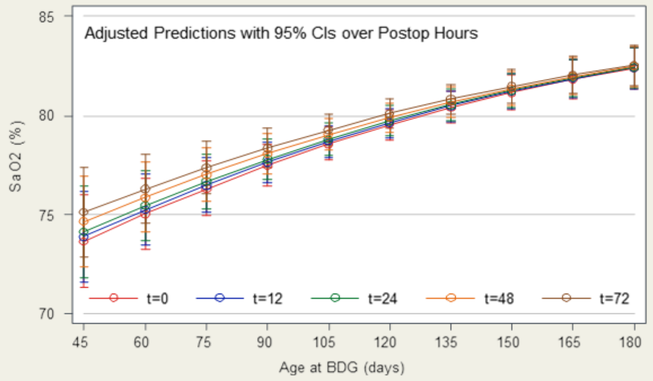

Figure 2. Predicted SpO_2 according to age at BDG.

DISCUSSION

The interstage period between the Norwood and BDG represents the time of greatest risk of mortality for the infant with HLHS. Optimal timing of BDG remains controversial.

Factors affecting decisions regarding timing of BDG conversion oxygen saturation:

1. Ventricular function
2. AV valve regurgitation
3. Pulmonary vascular resistance

Early BDG has been linked to:

1. Lower SpO_2
2. Prolonged pleural drainage
3. Hospital length of stay
4. **No increase in mortality**

Studies describing early BDG have generally described a lower age limit of approximately 2 m/o. Recent work suggests transplant free survival may be optimized when BDG was performed between 3-6 m/o. However, there are instances when BDG conversion must be performed earlier due to elevated risk of interstage mortality.

In this case we elected to perform BDG in an infant just beyond the neonatal period due to inadequate systemic to pulmonary artery shunt options.

We were able to successfully incorporate strategies to improve cerebral and pulmonary blood flow to reduce post-BDG hypoxia including permissive hypercapnea and iNO.

This case highlights the potential for selective BDG conversion in infants as early as 1 m/o of age when continued Stage 1 physiology represents a elevated risk of mortality.

REFERENCES

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