A Retrospective Analysis of Hydrocephalus Following the Bi-Directional Glenn Procedure for Single-Ventricle Congenital Heart Disease

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Introduction

Despite improvements in surgical management, cognitive development, and survival for patients with single-ventricle congenital cardiac disease (SVCHD), these children remain at high risk for poor quality of life, neurodevelopment, and low adult intelligence quotient (1-3). Modern surgical management often requires a palliative procedure as a newborn, with convergence toward the the final steps, the Bi-Directional Glenn (BDG) (Stage 2) and the Fontan (Stage 3). The BDG involves the anastomosis of the superior vena cava (SVC) to the pulmonary artery, resulting in an abruptly increased central venous pressure (CVP). We hypothesize that this increase in CVP triggers an acute neurologic insult, promoting the development of hydrocephalus in these developing infants.

Methods

Design: Retrospective Analysis in a Tertiary Care Children's Hospital

Patients: Between August 2006 and July 2013, 167 patients underwent the BDG. 24 patients had head imaging (CT, MRI, or ultrasound) performed both before and after the BDG.

Measurements: We measured the Frontal-Occipital Horn Ratio (FOR), a well-validated indicator of lateral ventricle volume (4-5). We assessed post-operative CVP at 12, 24, and 48 hours. Paired t-tests and linear regression were used. As this was a pilot study, statistical significance was set at p=0.10.

Results

Of 167 patients receiving the BDG shunt, 31 had post-operative imaging within 12 weeks of surgery:

- Median Age: 4.9 months (3.1-13.6 months)
- Median Weight & Length: 5.5 kg (3.6-9.4), 60.0 cm (50-71)
- 85% of these 31 patients had some post-operative intracranial abnormality noted
- 12 CVA, 10 Ventriculomegaly, 4 SDH, and 4 New Extra-Axial Fluid

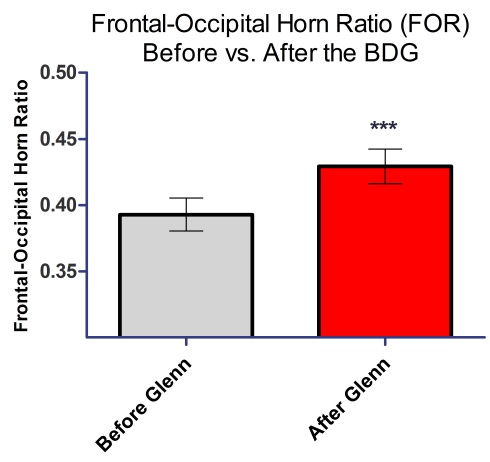
24 patients with both pre-operative and post-operative imaging:

- Pre-Operative Mean FOR: 0.397 (SD: 0.060)
- Post-Operative Mean FOR: 0.432 (SD: 0.066)
- Effectiveness of pairing was satisfactory (r=0.8230, p<0.0001)
- Paired analysis revealed a significant increase in lateral ventricle volume between pre-operative and post-operative imaging (p<0.0001, **Figure 1**)

Central Venous Pressures:

- Increasing change in FOR was associated with increased 12-hour (R2 = 0.369, p=0.003, **Figure 2**) post-operative CVP.

Figure 1

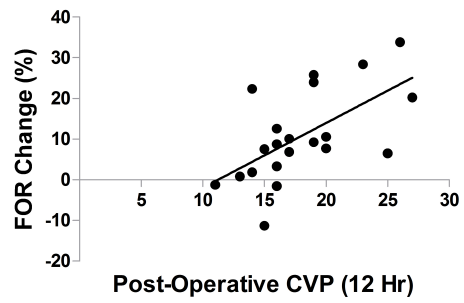


Two-tailed paired analysis demonstrates significant increase in FOR between pre-operative (M=0.397, SD=0.060) and post-operative (M=0.432, SD=0.066) head imaging. (***=p<0.0001)

Conclusions

- Our study is the first to demonstrate ventriculomegaly developing after the BDG.
- Physiologically, increasing CVP after the BDG was associated with greater change in lateral ventricle size. Increased SVC pressures inherent to the BDG may present an insult to the developing brain.
- Further prospective study is warranted to elucidate this association and to determine potential downstream impact on developmental outcome and areas where intervention may be useful.

Figure 2



Linear regression demonstrates that increasing change in FOR is significantly associated with increasing CVP at 12 hours following the BDG (R2 = 0.369, p=0.003)

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