The surgical management of spinal deformity in children with a Fontan circulation: Development of a treatment algorithm

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Introduction

• Series of cardiac shunt procedures for an anatomical or physiological single ventricle
• Creates total cavopulmonary circulation pumping venous blood into pulmonary and systemic circulations simultaneously
• Many variations of procedure since original description
• Total cavopulmonary connection
• Performed between 5 and 7 years
• Requires life long anticoagulation
Current Treatment

• To date, all cases have utilised posterior instrumented fusion
• Associated with high complication rate (84%)
• Increasing use of growing rod instrumentation for deformity correction
Aim

• To compare growing rod instrumentation and posterior spinal fusion in the management of scoliosis in children who undergone cavopulmonary (CP) shunting or Fontan procedure

• To describe our treatment algorithm to manage these complex patients
Treatment algorithm

Pre Anaesthetic Assessment
1. Pre-operative MDT (Cardiology, Anaesthetics, Spinal Service).
2. Transthoracic and Transoesophageal ECHO.
3. Cardiac catheterisation +/- balloon dilatation of stenotic lesions.
4. 24 hour ECG
5. Conversion of Warfarin to IV Heparin.

In Anaesthetic Room
1. Establish invasive monitoring (CVP lines, arterial lines).
2. Transoesophageal echo or Pulmonary arterial wedge catheter placement.
3. Urinary catheter.
4. Spinal cord monitoring.

Intra-Operative Procedure
1. Preparation of emergency sterile bed.
2. Two experienced spinal surgeon operating.
3. Patient turned prone.
4. PA Chest Compression – Simulate surgery.
5. Spinal exposure.
6. Prior to instrumentation.
7. Prior to correction of deformity.

Post-Operative Procedure
1. Transfer to ITU.
2. Early extubation.
3. Restart warfarin anticoagulation when appropriate.

Physiological parameters assessed at steps 3-7.
**Results**

- 6 cases identified. Median age 11yrs (6-16 yrs)
  - 2 patients with CP shunts. Median age 7 (6-8 yrs)
  - 4 patients with completed Fontan procedure. Median age 13 (11-16 yrs)

<table>
<thead>
<tr>
<th>Case</th>
<th>Cardiac diagnosis</th>
<th>Scoliosis aetiology</th>
<th>Age at index spinal procedure</th>
<th>Weight (kg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Double outlet left ventricle</td>
<td>Thoracogenic</td>
<td>8</td>
<td>20.8</td>
</tr>
<tr>
<td>2</td>
<td>Hypoplastic Left Heart Syndrome</td>
<td>Hemivertebrae T6 and L1</td>
<td>6</td>
<td>13.1</td>
</tr>
<tr>
<td>3</td>
<td>Hypoplastic Left Heart Syndrome</td>
<td>Thoracogenic</td>
<td>11</td>
<td>34</td>
</tr>
<tr>
<td>4</td>
<td>Tricuspid atresia</td>
<td>Thoracogenic</td>
<td>16</td>
<td>51</td>
</tr>
<tr>
<td>5</td>
<td>Atrio-ventricular septal defect</td>
<td>Thoracogenic</td>
<td>11</td>
<td>25.9</td>
</tr>
<tr>
<td>6</td>
<td>Atrio-ventricular septal defect</td>
<td>Thoracogenic</td>
<td>15</td>
<td>46</td>
</tr>
</tbody>
</table>
Index Spinal Procedure

- Noted trend for greater blood loss in those pts who had their index spinal procedure after completion of their Fontan procedure
- 1 pt required intra-operative adrenaline, but no sequelae
- No significant post-operative complications (1x UTI)
- All patients discharged from ITU within 48 hours and discharged home within 10 days

<table>
<thead>
<tr>
<th>Case</th>
<th>Index Spinal Procedure</th>
<th>Time spent prone (hours)</th>
<th>Blood loss mls/kg (total mls)</th>
<th>Intra-operative event</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cavopulmonary shunt (Pre-Fontan)</td>
<td>Vertical Expandable Prosthetic Titanium Rib (VEPTR)</td>
<td>5</td>
<td>5 (100)</td>
<td>No</td>
</tr>
<tr>
<td>1</td>
<td>Limited growth arrest</td>
<td>Patient in lateral position</td>
<td>20 (260)</td>
<td>No</td>
</tr>
<tr>
<td>Fontan circulation completed</td>
<td>Paediatric ISOLA</td>
<td>3</td>
<td>46 (1200)</td>
<td>Significant hypotension requiring adrenaline bolus</td>
</tr>
<tr>
<td>4</td>
<td>Posterior fusion</td>
<td>3.5</td>
<td>37 (1900)</td>
<td>No</td>
</tr>
<tr>
<td>5</td>
<td>Paediatric ISOLA</td>
<td>4</td>
<td>16 (350)</td>
<td>No</td>
</tr>
<tr>
<td>6</td>
<td>Posterior fusion</td>
<td>8</td>
<td>65 (3000)</td>
<td>No</td>
</tr>
</tbody>
</table>
Outcome

- Median FU – 87.6 months (52-103 months). 1 pt died 24 months post procedure from underlying cardiac pathology
- Median pre-operative Cobb angle 64.5 degrees (37-90 degrees)
- Median post-operative Cobb angle 50.5 degrees (26-65 degrees)
- Deformity correction of 24.2% (13-37.7%)

<table>
<thead>
<tr>
<th>Case</th>
<th>Index Spinal Procedure</th>
<th>Length of Follow-Up (months)</th>
<th>Pre-Op Cobb Angle</th>
<th>Final Cobb Angle</th>
<th>Further Spinal Procedures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cavopulmonary shunt (Pre-Fontan)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
| 1 | VEPTR | 73.8 | 90 | 56 | (i) Paediatric Isola and Anterior Release (9/12 post index procedure)  
(ii) 5 x growing rod lengthenings  
(iii) Definitive posterior fusion aged 14. |
| 2 | Growth arrest | 89.9 | 37 | 26 | No |
| Fontan circulation completed | | | | | |
| 3 | Paediatric ISOLA | 52.0 | 80 | 65 | No |
| 4 | Posterior fusion | 24.0* | 75 | 63 | No |
| 5 | Paediatric ISOLA | 103.0 | 46 | 40 | (i) 6 x growing rod lengthenings  
(ii) Definitive posterior fusion aged 16.  
(iii) Revision fusion for pseudoarthrosis aged 17. |
| 6 | Posterior fusion | 87.6 | 54 | 36 | No |

*Pt died from underlying cardiac condition 24 months post index spinal procedure
Discussion

• Continued improvements in the treatment of complex congenital cardiac abnormalities results in increased life expectancy with the subsequent need to manage the spinal deformity into young adulthood

• The use of growing rod instrumentation can be successfully employed in this patient population with little morbidity. All patients successfully tolerated subsequent lengthenings without complication
Discussion

- Posterior instrumented fusion was associated with a trend towards increased blood loss, operation duration and resultant haemodynamic instability.

Central Venous Pressure (CVP) and Pulmonary Capillary Wedge Pressure (PCWP) in a patient undergoing posterior instrumented fusion with a Fontan circulation. Haemodynamic instability noted as reducing PCWP despite adequate CVP filling.
Conclusion

• Early surgical intervention with growing rod instrumentation systems allows staged correction of the spinal deformity and reduces the haemodynamic insult

• Due to the haemodynamic changes that occur with the completed Fontan circulation, the initial scoliosis surgery should ideally be undertaken when in the CP shunt stage

• Despite the technical difficulties, it is possible to manage these spinal deformities with an associated Fontan circulation