

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

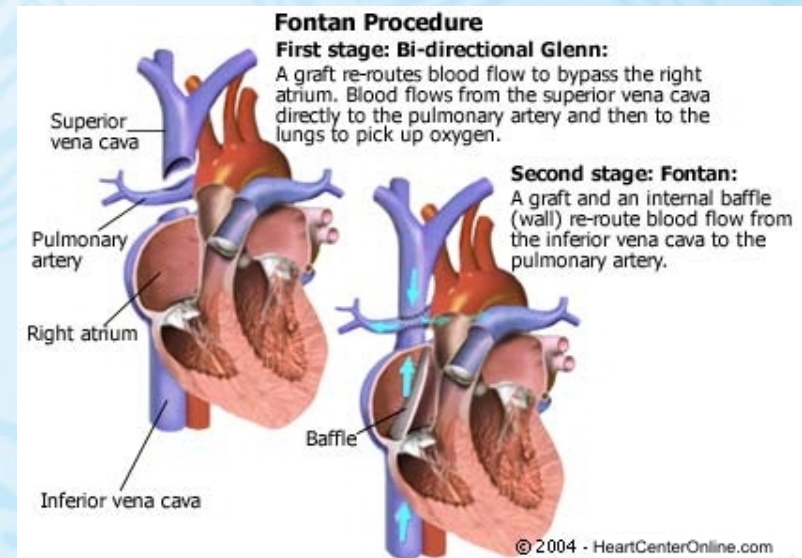
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## Introduction

- Series of cardiac shunt procedures for an anatomical or physiological single ventricle
- Creates total cavopulmonary circulation pumping venous blood in to 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%)  
Hedequest et al (2006)
- 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.

Physiological parameters  
assessed at steps 3-7.



## Post-Operative Procedure

1. Transfer to ITU.
2. Early extubation.
3. Restart warfarin anticoagulation when appropriate.

## 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)

Case	Cardiac diagnosis	Scoliosis aetiology	Age at index spinal procedure	Weight (kg)
<b>Cavopulmonary shunt (Pre-Fontan)</b>				
1	Double outlet left ventricle	Thoracogenic	8	20.8
2	Hypoplastic Left Heart Syndrome	Hemivertebrae T6 and L1	6	13.1
<b>Fontan circulation completed</b>				
3	Hypoplastic Left Heart Syndrome	Thoracogenic	11	34
4	Tricuspid atresia	Thoracogenic	16	51
5	Atrio-ventricular septal defect	Thoracogenic	11	25.9
6	Atrio-ventricular septal defect	Thoracogenic	15	46

## 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

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

## 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%)

Case	Index Spinal Procedure	Length of Follow-Up (months)	Pre-Op Cobb Angle	Final Cobb Angle	Further Spinal Procedures
<b>Cavopulmonary shunt (Pre-Fontan)</b>					
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
<b>Fontan circulation completed</b>					
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

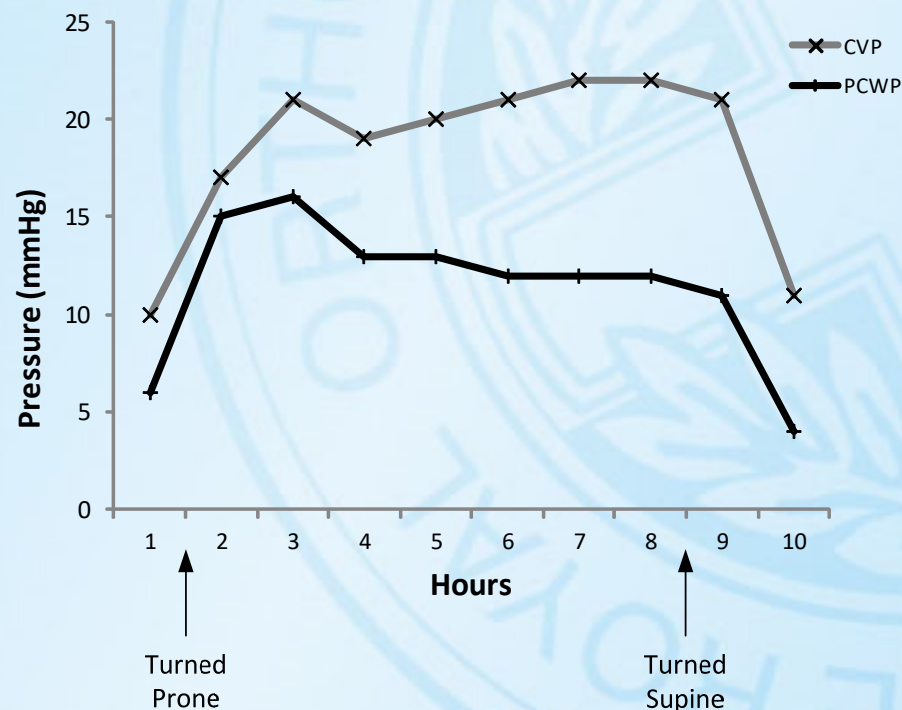


## Discussion

- 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



