Echocardiography in Fontan Circulation

Jan Marek

Great Ormond Street Hospital & Institute of Child Health
London, United Kingdom

No disclosures
Fontan palliation

Atrio-ventricular & atrio- pulmonary connection using conduits (valved, non-valved)


• The functional capacity of the patients was less with longer the period of follow up

• No risk factors were found for the late decline in survival or the decline in functional status

“…….the Fontan operation is, therefore, palliative but not curative”

Fontan F, Circulation 1990
“Classic” Fontan operation

- Large atrial cavity
- Distension of atrial cavity and myocardium
- Absence of respiratory “sucking”
- Pro-coagulation environment
- Pro-arhythmogenic environment

Thrombosis, AFlu+AFib, lung and systemic embolisation, protein losing enteropathy, plastic bronchitis, right heart failure, low cardiac output
Total Cavo-Pulmonary Connection (TCPC)

Importance of streamlining
SVC-to-inferior caval conduit flow ratio was 40:60 with 70% of total flow directed to RPA

Hsia TY, Circulation 2000

deLeval MR, J Thorac Cardiovasc Surg 1988
Fontan Palliation

CLASSICAL
< 1990

LATERAL TUNNEL
1990-2000

EXTRACARDIAC COND.
> 2000

SVC
RPA
RA
IVC
Fontan circulation: long term survival


TCPC in 38.7%, Extracardiac in 1.1%

TCPC in 50%, Extracardiac in 16%

Atrio-Pulmonary Connection

Total Cavo-Pulmonary Connection
Fontan Physiology
**Normal:** Pulmonary and systemic circulation in series, RAp < LAp, enough energy to blood to pass PVR

**Univentricular heart:** Systemic and pulmonary circuits in parallel
Considerable volume overload to SV. Mixed systemic and pulmonary venous blood -> desaturation

**Fontan circulation:** Systemic and pulmonary circulations in series, Less volume overload to SV than expected for BSA
No more admixture of systemic and pulmonary venous blood, but CVP elevated.

*Gewillig M, Heart 2005*
Fontan Physiology

“Push” & “Pull” mechanism

• Elevated CVP drives blood into lungs
• Systemic ventricle & Inspiration is sucking blood from lungs
Fontan Physiology

SVCp 12 mmHg

SVCp 17 mmHg
Fenestrated TCPC
Increased pulmonary vascular resistance

- CI .............. 2.0 l/min/m²
- TO₂ .......... 355 ml/min/m²

- CI .............. 2.5 l/min/m²
- TO₂ .......... 400 ml/min/m²

Fontan Physiology

CVp 13 mmHg
LAp 5
TPG 8 mmHg

CVp 18 mmHg
LAp 12
TPG 6 mmHg
Non-LV Chamber

Supporting Systemic Circulation:
Morphological & Physiological Determinants
Fontan Circulation: Ventricular function

Cause of ventricular dysfunction:

- Architecture of myocardium
- Incoordination
- Abnormal loading condition
- Intrinsic myocardium abnormality
- Ischaemic dysfunction
- Rhythm disturbance
Fontan circulation

RV function in HLHS

- Precipitants of poor RV function
- Factors preserving RV function
- RV function crucial for decision making (TCPC vs Transplant)

RVEF may be the single most discriminatory Measure predicting success of Fontan circulation
Non-invasive Assessment of Myocardial Function in Non-LV Setting
Fontan Circulation

Ventricular function: good or bad?
Non-LV function
Echocardiographic challenges

• Eye balling
• Planimetry
• (T)APSE
• M-mode (AMM)
• +dP/dt (systemic AVVR)
• Tei index
• TDI (Strain, -SR)
• RT-3DE
• 2D/3D Strain
Non-LV function

Echocardiographic challenges

• Eye balling
• Planimetry
• (T)APSE
• M-mode (AMM)
• +dP/dt (systemic AVVR)
• Tei index
• TDI (Strain, -SR)
• RT-3DE
• 2D/3D Strain
Systemic ventricular function: 

\[ + \frac{dP}{dt} = \text{change in pressure during isovolumetric contraction; before semilunar valve opens when there is no significant change in atrial pressure} \]

\[
\frac{dP}{dT} \ [mmHg/s] = \frac{P_2-P_1}{T} \ [mmHg] \]

\[
T \quad \text{Time} \ [s]
\]

Oh JK, The Echo Manual. 2nd ed. 1999
+dP/dt: impact of loading conditions

Fontan pt. on V-A ECMO
(bridge to recovery)

Full flow ECMO

30% flow ECMO

Off ECMO
  +
  Adr.0.01, Mil.0.5

1,117 mmHg.s⁻¹
611 mmHg.s⁻¹
1,085 mmHg.s⁻¹
Myocardial deformation (longitudinal)

Shortening
Lengthening

SYS
DIAST

Strain Rate (sec$^{-1}$)
Strain (%)
IVA (cm / sec$^{-1}$)

Spectral TDI
Direct TDI

Courtesy P.Claus, Leuven
Myocardial Deformation Imaging in single ventricle morphology

All studies show reduced parameters of systolic and diastolic function (Strain, SR, IVA, velocities) in all patients with single ventricle morphology irrespective of procedure performed and pt`s clinical condition

Eicken A, Int J Cardiol 2007, Petko C, Int J Cardiol 2010
Mechanical dyssynchrony in single ventricle morphology
Temporary CRT in single ventricle

- Acute CRT in postoperative single-ventricle and narrow QRS (<130 ms)
- AV delay adjusted to achieve shortest QRS (fusion of intrinsic and paced activation)

Bacha EA et al., Ann Thorac Surg 2004
Systemic non-LV ventricular function: routine ECHO assessment

- **Eyeballing:** good, impaired, poor
- **M-mode (anatomic)**
  - Fractional shortening (FS 26-30%)
  - (T)APSE (>15cm)
- **CW Doppler**
  - +dP/dt (≥ 800 mmHg/s)
- **TDI**
  - For synchronicity if CRT considered

**All parameters used are load dependent!**

Apply for serial measurements information rather than for final decision making (with Tg/s)

For synchronicity if CRT considered
Fontan – Role of CMR

- Fontan connections
- Ventricular function
- AVVR quantification
- Ruling out pulmonary vein obstruction/compression
- Ruling out thrombus
- 3D “roadmap” for intervention
- Late gadolinium - ventricular and atrial scarring
Future of cardiac imaging:
Fusion of dynamic cardiac imaging modalities

- High resolution 4D imaging
- Blood flow dynamics
- Physiology at rest and exercise
- Metabolic dynamic tissue definition
- Education and modelling
Fusion of dynamic cardiac imaging