The Fontan circulation

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What to expect?

Why a Fontan-circulation
  * Indications
  * How does it work

Types of Fontan circulation
  * Historical overview

Role of echocardiography
What to expect?

Why a Fontan-circulation
  Indications
  How does it work

Types of Fontan circulation
  Historical overview

Role of echocardiography
Why a Fontan circulation?

- No functional RV
- Volume overload LV
- Systemic venous return plus pulmonary venous return
- Desaturation systemic circulation - cyanosis

• How can this be solved?
Why a Fontan circulation?

- No functional RV
- Volume overload LV
- Systemic venous return plus pulmonary venous return
- Desaturation systemic circulation - cyanosis

Not by a biventricular repair

→ Univentricular repair
Indication for a Fontan

situations for which a biventricular is not possible
Indication for a Fontan

situations for which a biventricular is not possible

- HLHS
- TA
- MA
- DILV
- Unbalanced AVSD
- Ebstein
- PA – intact IVS
- etc
The essence of the Fontan circulation

- only 1 functional ventricle → systemic ventricle
The essence of the Fontan circulation

• only 1 functional ventricle → systemic ventricle
• no subpulmonary ventricle
The essence of the Fontan circulation

- only 1 functional ventricle $\rightarrow$ systemic ventricle
- no subpulmonary ventricle
- Systemic venous return flows, without a pumping ventricle, into the lungs
The essence of the Fontan circulation

- only 1 functional ventricle → systemic ventricle
- no subpulmonary ventricle
- Systemic venous return flows, without a pumping ventricle, into the lungs
- no desaturated blood in the systemic circulation
The essence of the Fontan circulation

- only 1 functional ventricle $\rightarrow$ systemic ventricle
- no subpulmonary ventricle
- Systemic venous return flows, without a pumping ventricle, into the lungs
- no desaturated blood in the systemic circulation
- No longer a volume-overloaded LV
How does it work?

How can an adequate pulmonary blood flow be achieved?

1. elevated systemic venous pressure
2. low pulmonary vascular resistance
3. Low LA pressures
How does it work?

Flow depends on pressure difference between systemic venous pressure and LA pressure and pulmonary vascular resistance.
Pressure difference VCS - LA

- **Push & pull** circulation:
  - *Elevated systemic venous pressure* = push
  - *Periodic lowering of LA pressure*
    - During systole
    - During rapid filling of ventricle
LA pressure curve = the “pull”
Anatomy (history) of the Fontan-operation
“the” patient with a Fontan circulation

- Does not exist
“the” Fontan circulation

- Mixed bag
- many different types of
  - Intracardiac anatomy
  - Type of surgical repair
Echocardiography & Fontan

- Know what the underlying cardiac defect is
- Know what type of Fontan repair is done
- Understand the Fontan circulation
Echocardiography & Fontan

• Know what the underlying cardiac defect is
• Know what type of Fontan repair is done
• Understand the Fontan circulation

• Read the surgical report!!
Echocardiography & Fontan

- Know what the underlying cardiac defect is
- Know what type of Fontan repair is done
- Understand the Fontan circulation

- If you do not know where you are looking for, you may not find it
Echocardiography & Fontan

• in univentricular hearts – **before surgery** – echo is important for:

  • *Intracardiac anatomy*
  
  • *Ventricular function*
  
  • *AV valve function*
  
  • *Biventricular - univentricular repair*
Echocardiography & Fontan

- Univentricular hearts after Fontan
  - (Intracardiac anatomy)

- Ventricular function

- AV valve function
Echocardiography & Fontan

• Univentricular hearts after Fontan
  • (Intracardiac anatomy)

• Ventricular function

• AV valve function

Anatomy & function of the Fontan circulation
Echocardiography & Fontan

- Univentricular hearts after Fontan
  - (Intracardiac anatomy)

Ventricular function

- AV valve function

Anatomy & function of the Fontan circulation
ventricular function & Fontan
ventricular & AV valve function
ventricular & AV valve function
ventricular & AV valve function
AV valve function

• Assessment of AV valve regurgitation = very important

Moderate to severe regurgitation
• → elevated LA pressure
• → decrease in transpulmonary gradient
• → decrease in transpulmonary flow
• → decrease in cardiac output
• → compensatory mechanism = fluid retention
• → elevated central venous pressure
• → transpulmonary gradient restored
• → cardiac output restored
AV valve function

- Assessment of AV valve regurgitation = very important
  
  Moderate to severe regurgitation
  
  - $\rightarrow$ elevated LA pressure
  
  - $\rightarrow$ decrease in transpulmonary gradient
  
  - $\rightarrow$ decrease in transpulmonary flow
  
  - $\rightarrow$ decrease in cardiac output
  
  - $\rightarrow$ compensatory mechanism = fluid retention
  
  - $\rightarrow$ elevated central venous pressure
  
  - $\rightarrow$ transpulmonary gradient restored
  
  - $\rightarrow$ cardiac output restored

- To a certain limit $\rightarrow$ decompensation
Fontan & AV valve function

• Assessment of AV valve regurgitation = very important

• If one sees development of AV valve regurgitation
  • Alarm signs
  • Worsening ventricular function

• How to assess AV valve regurgitation: beyond the scope of this talk
Fontan & ventricular function

How do we assess & measure function of the systemic ventricle?
Fontan & ventricular function

- EAE & ASE recommendations on chamber quantification
- LV = systemic ventricle
  - *Standardized cross sections*
    - 4Ch view
    - Parasternal long axis
  - *Normal values for sex and body size*
Fontan & ventricular function

- EAE & ASE recommendations on chamber quantification
  - *Standardized cross sections???
    - 4Ch view
    - Parasternal long axis
  - Normal values for sex and body size

- But embrace the idea behind it:
  - *Use reproducible cross section*
  - Anatomic landmarks
    - DILV ≠ HLFS ≠ unbalanced AVSD
Fontan & ventricular function

Eyeballing

Quantify what you can quantify, in a reproducible way

Essential for longitudinal follow-up

- M-mode
- 2D
- 3D
- Doppler (incl TDI)

Not for comparison with normal values; patient as his/her own control
2D & color Doppler
Tissue Doppler

FR 70Hz
19cm

2D
69%
C 35
P Low
HGen
TDI
89%
3.4 MHz

Med E' Vel 5.72 cm/s
E/Med E' 7.0 50%
3.6 MHz
SV5.0 mm
11.2 cm

75 mm/s
55 bpm
Ejection time, Tei-index

FR 26Hz
19cm

2D
53%
C 50
P Low
HPen

CF
75%
2.5MHz
WF High
Med

Time 313 ms
Slope 0.000 cm/s²

PW
25%
1.6MHz
WF 75Hz
SV 4.0mm
12.2cm

M3 M4
+69.4

-69.4 cm/s

-40
-80
-120
-160

75mm/s
56bpm
Relative duration systole in cardiac cycle (good functioning adult Fontan)
duration systole, incl IVCT & IVRT (poorly functioning adult Fontan)
duration systole – very short diastole (3 yr old with HLHS)
SD ratio in the RV

Friedberg, J Am Soc Echocardiogr 2007;20:749
duration systole – short diastole
(21 yr old criss-cross, TCPC with PLE)
CW dP/dt
PV flow pattern: very low S in absence MR
LA pressure curve

systole
Echo & Doppler measurements

- Absolute values difficult to interpret
- Change over time/ in different conditions may guide us:
  - *in clinical decision making*
  - *When to send a patient to the cathlab*
Echo & Doppler measurements

- Absolute values difficult to interpret
- Change over time/ in different conditions may guide us:
  - *in clinical decision making*
  - *When to send a patient to the cathlab*
- The essence of the Fontan circulation: \( V = \frac{P}{R} \) we cannot measure this with echo
Visualisation of Fontan anatomy
Visualisation of Fontan anatomy

- Segmental approach
  - Hepatic veins
  - VCI
  - Tunnel
    - Intracardiac
    - Extracardiac
  - RA
  - Connection with PA
  - VCS
  - Connection with PA
  - Pulmonary artery branches
  - Pulmonary veins
Visualisation of Fontan anatomy & function

- Segmental approach
  - Hepatic veins
  - VCI
  - Tunnel
    - Intracardiac
    - Extracardiac
  - RA
  - Connection with PA
  - VCS
  - Connection with PA
  - Pulmonary artery branches
  - Pulmonary veins

- Color Doppler patterns
- Pulsed Doppler flow patterns
- Shunts/leaks in the tunnel
- Thrombi
VCI & hepatic veins
VCI pulsed Doppler: Scale! Low velocity filter! Respiration!
VCI pulsed Doppler: Scale! Low velocity filter! Respiration!
TCPC – driving force = inspiration
no influence of cardiac cycle
Connection VCI – extracardiac lateral tunnel
Lateral, extracardiac tunnel
look for thrombi
Apical 4 Ch view intracardiac tunnel
Intra-atrial tunnel

- Initially a small diameter
- No PV obstruction

Apical scan; upside down
Intra-atrial tunnel; adult age

- can dilate in time,
- become very wide
- can give PV obstruction
- Looks like RA – PA
- (different flow patterns)
VCI Doppler in RA – PA conduit
effect of atrial contraction
Tricuspid atresia; RA – AP connectie
RA – PA conduit
Atrio – ventricular connection
Tricuspid atresia & RA – RV connectie
RA – RV conduit with stenosis
PA flow in RA – RV conduit:

Pulsatile systolic flow in PA!
Doppler patterns & type of Fontan

- Doppler patterns in different types of connection are different
  - *Doppler patterns in different patients with the same type of connections are different*

- registration of ECG – p-waves – and respiration curve

- Off-line studying on tracings:
  - *What explains flow patterns?*
  - *What explains phasic changes?*
intracardiac tunnel VCI - PA
Look for shunts/abnormal flows
Baffle leakage with R – L shunt
Baffle leakage with R – L shunt

![Echo image showing baffle leakage with R-L shunt](image)

- **Vmax**: 1.66 m/s
- **Vmean**: 1.04 m/s
- **Pmax**: 10.97 mmHg
- **Pmean**: 4.29 mmHg
- **Env.Ti**: 2425 ms
- **VTI**: 251.24 cm
- **HR**: 24.74 BPM

*Courtesy Jan Marek*
What to do with a baffle leak?

• Beyond the scope of this talk
Bidirectional Glenn anastomosis
VCS – Glenn from R supraclavicular
VCS – Glenn from R supraclavicular
Glenn anastomose

• VCS
APL from SSN; almost always obtainable
APL bij TCPC
From SSN: LPA
From SSN: LPA
APL in LA – AP connection
R upper pulmonary vein
PV Doppler

FR 28Hz
17cm

2D
55%
C 50
P Low
HPen
CF
66%
2.5MHz
WF High
Med

PW
35%
1.6MHz
WF 125Hz
SV 4.0mm
14.5cm

M3 M4

PW
35%
1.6MHz
WF 125Hz
SV 4.0mm
14.5cm
RUPV obstruction (RA – RV connection)

- AP4K
- PW
- Always look for the R pulmonary veins, especially if the IAS is bulging
Dilated RA in RA-AP connection
beware of R PV obstruction
RUPV obstruction

Pulsed wave Doppler ("pull-back")
Conclusion
Conclusion

Why a Fontan-circulation

*Indications*

*How does it work*

Types of Fontan circulation

*Historical overview ➔ essential for the understanding of echo & Doppler images*

Role of echocardiography in the follow-up

*Measuring function in a reproducible way for longitudinal follow up*
Fontan & echo = difficult & fascinating

You need to understand Fontan physiology, especially when you consider intervention (elective redo of “old” RA- PA to TCPC)

Echo is very helpful, but do not hesitate to send a patient to the cathlab when in doubt.

\[ V = P/R \]