Functionally Single Ventricle
Glenn and Fontan
echocardiographic assessment
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Echographic assessment in Fontan patients:
sequential segmental approach

1. Cavopulmonary connection
2. Branch PAs
3. Pulmonary veins
4. Atrial septum
5. Systemic outflow tract
6. Systemic ventricular function
7. Atrioventricular valves
8. Aortic arch

The Fontan operation
- First described in 1971
- Initially designed for tricuspid atresia
- Extension to all forms of functionally UVH

Choussat’s 10 commandments revisited
- Age > 4 years
- Sinus rhythm
- Normal systemic venous return
- Normal right atrial volume
- Mean pulmonary artery pressure < 15 mm Hg
- Pulmonary arteriolar resistance < 4 Wood units/m²
- Pulmonary artery–aorta ratio > 0.75
- Left-ventricular ejection fraction > 0.60
- Competent mitral valve
- Absence of pulmonary artery distortion

Goal of pre Fontan surgical management
- Balance aortic / pulmonary blood flow
- Optimize PA growth
- Protect from pulmonary vascular disease
- Surgical timing: limit the period of ventricular overload
- Keep in mind a simple logic

Early palliative approach:
- Systemic to pulmonary shunt or banding
  - First months of life
    - Always volume overload (by PA band or SPS)
    - Dilatation and spherical reconfiguration
    - Cardiac overgrowth
    - Eccentric hypertrophy
  - Fontan (TCPC)
    - Reduced preload (below normal)
    - Reduced compliance
    - Poor ventricular filling
SV with high pulmonary blood flow: PA banding
- SV-PA gradient: cw doppler (SAX/A4C)
- Pulmonary valve regurgitation: 2D/ color doppler
- Migration of PA banding: pulmonary branch distortion (RPA)

SV with low pulmonary blood flow: SPS
- First SPS: direct connection between SCA (CBTS)/ Ao (Waterston) and PA
  - Unpredictability of shunt flow
  - PA branch distortion

Systemic to pulmonary shunt
- Current modified BT shunt: prosthetic PTFE graft
- Innominate artery or SCA connected to ipsilateral PA branch

Hybrid procedure in HLHS

Systemic to pulmonary shunt
- Imaging: CW doppler: characteristic sawtooth doppler pattern
- Physiology: Peak Velocity (m/s) consistent with aortic to pulmonary pressure gradient and clinical findings (O2 sat)
- Anomaly: potential anomalies: distortion of inn Artery or PA branch, narrowing of prox or distal anastomosis (challenging)
The Fontan circulation: stage 1

- Staged approach
- Stage 1: BCPC – Glenn procedure
  - Re-routing SVC blood flow to pulmonary circulation
  - More desaturated blood shunted to the lungs
  - Diversion of 1/3 of the SVR to the lungs
  - Reduction of SV volume overload

Echocardiographic assessment

1. Cavopulmonary connection
2. Branch PA

Echocardiographic assessment of the Glenn anastomosis

- Suprasternal / high parasternal view
- Laminar flow of low velocity with respiratory variation (lower the Nyquist limit)
- Rule out stenosis at the anastomosis site

Pulmonary arteries

- Potential PA branch distortion following early palliative surgery
  - SPS / Banding
  - DKS (LPA hypoplasia or stenosis)
PA growth in (staged) Fontan palliation
- Old Fontan era: direct atripulmonary connection
- Single stage surgery
- Risk factor for late mortality
  - Mc Goon <1.8 (Fontan et al. Circ 1989)
  - Nakata < 250mm/m²
- New Fontan era: extracardiac conduit
- Staged palliation
- Early completion

PA growth after BDG
- PA index after Fontan completion
  - Fails to match the increase in BSA
  - Relation with decrease amount of blow flow
  - Absence of pulsatile flow

Multiparametric assessment
- Proximal PAs size
- Distal PAs size
- Local stenosis
- RVAP, PVR, cAVP

Abnormal cyanosis after BCPC
- Reopening of decompressing veins from SVC
- In the IVC: azygos vein
- In the atria: left SVC
- Diagnosis:
  - Substernal frontal view
  - Color doppler / saline contrast

Abnormal cyanosis after BCPC
- Pulmonary AV malformations
  - Heterotaxy syndrome
  - Lack of hepatic factor
  - Diagnosis: saline contrast

The Fontan circulation stage 2: TCPC
- SVC and Glenn anastomosis
- IVC to PA conduit assessment
- Conduit fenestration
- IVC and HV flow
- Thrombus in the Fontan pathway
Distal connexion high parasternal view

Fenestration assessment
- Right to left shunt
- Decompress the systemic venous pathway
- Maintain cardiac output

Hijazi et al. Circulation 1992

Transpulmonary gradient
- Mean gradient over several cardiac cycles
- A4C/ PW doppler

CVo= 14mmhg
TPG=5mmhg
LAp=9mmHg

Percutaneous fenestration guiding

TOE for conduit assessment

LA size
Conduit side
**IVC flow after TCPC**

- Normal: continuous anterograde flow of low velocity, respiratory variation

- Retrograde A wave:
  - failing Fontan (AVC, arrhythmia)

- Retrograde S wave:
  - AV regurgitation: IVC
  - Antegrade flow (pulm stenosis): SVC

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**Understand cardiopulmonary interaction**

Echo adulte
47.7±
13Hz
frez

RPM: 32.00
4.50

Systole

Antegrade flow (pulm stenosis): SVC

Retrograde S wave:
- AV regurgitation: IVC
- Antegrade flow (pulm stenosis): SVC

Arterial regurgitation: IVC

Failing Fontan (AVC, arrhythmia)

Continuous anterograde flow of low velocity, respiratory variation

Echocardiogram images showing various cardiac structures and flow patterns.

Images courtesy of Jan Marek.
**Thrombosis in TCPC**
- Systemic venous pathway (TCPC)
  - IVC, RA (atriopulmonary connection), lateral tunnel/ conduit, fenestration, PAs, SVC
- Intra cardiac chambers
  - Intracardiac: LA, LAA, SV (poor systolic function)
- Native PA trunk
  - PAs divided and pulm valve non sutured

**Persistent cyanosis after TCPC**
- Conduit fenestration
  - Balance between PVR and early diastolic function
- Systemic venous pressure > pulmonary vein pressure
  - VV collaterals to pulmonary veins or systemic atrium
    - origin: LSVC, RSVC, int Vein, hepatic veins
- Baffle leaks (intra cardiac type of connection)

**Superior veno venous collateral**
- Left « SVC » to the RA
Inferior veno venous collateral
- HLHS S/P TCPC. Persistant mild hypoxemia
- Shunt between HV and cardiac veins

Echographic assessment
- 3. Pulmonary veins
- 4. Atrial septum

Pulmonary veins and atrial septum
- PV compression
- Atriopulmonary connection
- Intracardiac tunnel
- Heterotaxy syndrome
- Restriction of interatrial shunt in HLHS
- Potential cause of elevated PA pressure

Echographic assessment
- SV AFTERLOAD
  - 6. Systemic outflow tract
  - 7. Aorta

Restrictive systemic outflow tract in DILV
- Proximal anastomosis
  - Restrictive bulboventricular foramen: DKS
Restrictive systemic outflow tract

- Proximal anastomosis
- Restrictive bulboventricular foramen: DKS

Anastomosis

Systemic outflow tract after Norwood procedure in HLHS

- Proximal anastomosis
- DKS stenosis
- Neoaortic valve regurgitation

Aortic arch assessment

- Distal anastomosis
  - Supra sternal sagittal view
  - Potential increased velocity (HLHS)
  - Funnel-like narrowing
  - Pitfall: potential absence of diastolic runoff pattern in coarctation
    - Change in Ao arch geometry: patch/mBTshunt
    - CoA index
    - Distal Ao root narrowing/ widened diam <0.7
    - Peak gradient > 30mmHg

Aorto-to-pulmonary collateral flow

- Present in most patients (80%)
- Suprasternal frontal and sagittal views
- Limited assessment with TTE
- Consider alternative imaging: MRI/Cath
  - MRI: quantification of aortic to pulmonary shunt flow
  - Cath: PAP, PVR measurements and percutaneous closure

Echographic assessment

- 5. Atrio-ventricular valves

Echographic assessment of AV valves

- Assessment of an heterogeneous group
  - Various anatomy
  - Different loading conditions/ different stage
  - Serial measurements++++

- Echo report
  - AVV diameter and function
  - Location of papillary muscles/chordal attachment
  - Difference between functional and anatomical regurgitation
LAVV anomalies in DILV

Unbalanced AVSD

TR in systemic right ventricle

Echographic assessment

Conclusion: echocardiography in Fontan circulation

- Easily accessible and cost-effective tool
- Good assessment of the Fontan pathway in children but may be limited in adults
- Best imaging modality for AVV assessment
- LVH systolic and diastolic function assessment remains challenging
- Consider alternative imaging modalities for extracardiac lesions (Ao-pulm collaterals, complex aortic arch stenosis)

Essential reading

- Marc R de Leval