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

PRELOAD

3. Pulmonary veins
4. Atrial septum

FUNCTION

5. Systemic outflow tract
6. Systemic ventricular function
7. Atrioventricular valves
8. Aortic arch

AFTERLOAD
The Fontan operation

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

Choussat’s 10 commandments revisited

1971

Choussat’s ten commandments

- 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

2014

Choussat’s ten commandments REVISITED

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- Pulmonary arteriolar resistance<4 Wood units/m²
- Left-ventricular ejection fraction>0.60

Stern J. Pediatr Cardiol 2010
Kilner P. CITY 2005

www.chu-bordeaux.fr
**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

![Diagram](image-url)
**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

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**Fontan (TCPC)**
Reduced preload (below normal)
Reduced compliance
Poor ventricular filling

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![Graph showing the progression from birth to Fontan](image-url)
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
  - Unpredictibility 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
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)

- Anatomy: potential anomalies: distortion of inn Artery or PA branch, narrowing of prox or distal anastomosis (challenging)
Hybrid procedure in HLHS
The Fontan circulation: stage 1

- **Staged approach**

- **Stage 1: BCPC – Glenn procedure**
  - 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
Potential PA branch distortion following early palliative surgery

- SPS / Banding
- DKS (LPA hypoplasia or stenosis)
Pulmonary artery branches and BCPC

Risk of LPA stenosis post Norwood procedure

Mild stenosis

Distal LPA
PA growth in (staged) Fontan palliation

- Old Fontan era: direct atropulmonary connection
- Single stage surgery
- Risk factor for late mortality
  - Mc Goon <1.8 (Fontan et al. Circ 1989)
  - Nakata < 250 mm/m²

- New Fontan era: extracardiac conduit
- Staged palliation
- Early completion

McGoon Index: RPA/d (mm) + LPA/d (mm) / DMO (mm) > 1.5
Nakata Index: RPA (mm²) + LPA (mm²) / m² BSA > 150 mm²
PA growth after BDG

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

- **Multiparametric assessment**
  - Proximal PAs size
  - Distal PAs size
  - Focal stenosis
  - mPAP, PVR, cwp

Abnormal cyanosis after BCPC

- Reopening of decompressing veins from SVC

- In the IVC: azygos vein
- In the atria: left SVC
- Diagnosis:
  - Suprasternal frontal view
  - Color doppler/ saline contrast
Abnormal cyanosis after BCPC

- Pulmonary AV malformations
  - heterotaxy syndrome
  - Lack of hepatic factor
  - Diagnosis: saline contrast

[Image of a medical diagram and an X-ray with RSVC, showing abnormal vessels.]
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
TCPC

Subcostal sagittal view M mode

Subcostal sagittal view pulsed wave doppler

Proximal connection subcostal views
Distal connexion high parasternal view

Situs solitus

conduit

Situs inversus

conduit
Fenestration assessment

**Right to left shunt**

- Decompress the systemic venous pathway
- Maintain cardiac output

AO SaO2 84±6%
Qs:2.4±0.7 l1*min*m-2
SOT*425±154ml

AO SaO2 95±3%
Qs: to1.8+0.4l1*min*m-2
SOT: 366±112ml

* Systemic oxygen transportation

Hijazi et al. Circulation 1992
Fenestration assessment

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

CVp= 14mmhg
TPG=5mmhg
LAp=9mmHg
IVC-PA conduit

fenestration
TOE for conduit assessment
Percutaneous fenestration guiding

LA size

Conduit side

LA

Conduit
IVC flow after TCPC

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

- Retrograde A wave:
  - failing fontan (CVP-arrhythmia)

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

Courtesy Jan Marek
Understand cardiopulmonary interaction
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, inn Vein, hepatic veins

- **Baffle leaks (intra cardiac type of connection)**
Superior veno venous collateral

Left « SVC » to the RA
Inferior vena venous collateral

- HLHS S/P TCPC. Persistent mild hypoxemia
- Shunt between HV and cardiac veins
Echographic assessment

3. Pulmonary veins
4. Atrial septum

- PV compression
  - Atriopulmonary connection
  - Intracardiac tunnel
  - Heterotaxy syndrome

- Restriction of interatrial shunt in HLHS

- Potential cause of elevated PA pressure
Pulmonary veins and atrial septum
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
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:
    - Desc Ao ratio narrowest/ widest 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

- Restricted bulboventricular foramen
- Increased AFTERLOAD

- PA banding/MR
- Increased PRELOAD

- LV spherical remodeling: secondary MR worsening
Unbalanced AVSD
TR in systemic right ventricle
Echographic assessment

- Systolic and diastolic(++) function
  - No single morphology
  - Important confounding variables
    - Preload (surgical stage/ AVVR/ PVR/PA compliance)
    - Afterload (restrictive outflow trat, CoA)

- Eye balling/ EF (TM, 2D) / myocardial deformation

- Ongoing research on advanced parameters
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

- UVH 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