The Palliated Single Ventricle

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Single Ventricle Palliation

Learning objectives

• Echo evaluation of anatomic and physiologic features of the single ventricle heart at each stage of palliation

• Recognize echo features of complications at each stage
Types of Single Ventricle Defects

- Hypoplastic left Heart
  - HLHS
  - Unbalanced AVC
- Hypoplastic right Heart
  - Tri atresia
  - PA/IVS
  - Unbalanced AVC
- Univentricular AV connections
  - Double inlet
  - Single inlet
  - Common inlet

Systemic or pulmonary outflow obstruction
Surgical Palliation of SV

Goals:

• Ensure adequate unobstructed pulmonary and systemic blood flow
• Separate the systemic and pulmonary venous blood
Surgical Palliation of SV

Stage I
Norwood or BT

Stage II
Glenn

Stage III
Fontan

Khairey P et al. Circ 2007;115:800  Children’s Healthcare of Atlanta | Emory University
Imaging After Stage I Palliation for Single Ventricle
Stage I: Systemic Obstruction
Norwood Procedure

BT shunt
Sano

Images from uichildrens.org  Children's Healthcare of Atlanta | Emory University
Imaging After Stage I Palliation

- Unrestricted atrial septum
- AV valve function
- Ventricular function
- Unobstructed pulmonary blood flow
  - BT shunt, Sano, branch PAs
- Unobstructed systemic outflow
  - DKS, neo-aortic valve and arch
Imaging After Stage I Palliation

Imaging of the atrial septum
Imaging After Stage I Palliation

Tricuspid regurgitation

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Imaging After Stage I Palliation

Intracardiac lines and thrombi
Imaging After Stage I Palliation

Ventricular function

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Imaging After Stage I Palliation

Ventricular function
Ventricular Dysfunction

Menon et al. JASE 2011;24:826

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Imaging After Stage I Palliation

Imaging of the BT shunt
Imaging of Sano Conduit

Proximal Sano conduit
Imaging of Sano Conduit
Imaging of Sano Conduit - Branch PAs
Echo Imaging of Norwood Sano Conduit with Stenosis
Imaging of Sano Conduit Stent
Echo Imaging of Norwood Neo-aortic valve
Damus-Kaye-Stansel Anastomosis

Yang et al. KJTCVS 2014;47

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Echo Imaging of Norwood Neo-aorta

Size discrepancy and potential for coarctation
Echo Imaging of Norwood Arch-Doppler
Echo Imaging of Norwood Coarctation
Echo Imaging of Norwood Stent for Coarctation
Hybrid Procedure

Banding of LPA and RPA, Stenting of ductus

Image from uichildrens.org
Echo Imaging of Hybrid Branch PA Bands
Echo Imaging of Hybrid Branch PA Bands
Echo Imaging of Hybrid Ductal Stent
Imaging After Stage II Palliation for Single Ventricle
Stage 2: Glenn Procedure

SVC connection to RPA

Imaging goals After Glenn
SVC-PA connection
Branch PA stenosis
Function
AV valves
Arch

Image from uichildrens.org
Echo After Glenn Procedure
SVC-RPA Connection
Echo After Glenn Procedure
Doppler Assessment
Echo After Glenn Procedure
Evaluation of Central and Left PA
Doppler- LPA Stenosis

Mean LPA gradient
7 mmHg
Echo After Glenn Procedure
Evaluation of Left PA
Echo After Glenn Procedure
LPA Stent
Systemic Venous Anomalies

*Broad based hepatic venous drainage to the common atrium*
Echo After Glenn Procedure
Bilateral SVC

Echocardiography in Pediatrics and Adult CHD, 2nd Ed
Echo After Glenn Procedure
Neo-aorta Compressing the PA
Imaging After Stage III Palliation for Single Ventricle
Stage 3: Fontan Procedure

Atriopulmonary

Extracardiac conduit

Lateral tunnel

Images from Children’s Hospital Boston

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Echo Imaging of Fontan

- Patency of Fontan pathway
  - SVC-PA
  - IVC-Conduit
- Fontan fenestration
- Branch PAs
- AV valve and semilunar valve
- Ventricular function
- Aorta
Fontan Pathways

Echocardiography in Pediatrics and Adult CHD, 2nd Ed

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Echo After Fontan
IVC-Fontan Connection
Echo After Fontan
Fontan-PA Connection
Echo After a Fontan
SVC Stent
Fontan Fenestration

Fenestration
Mean = 6 mmHg
Echo After Fontan
Lateral Tunnel Baffle Leak
Echo After Fontan
Lateral Tunnel Baffle Leak
Clues to Fontan Dysfunction

- Progressive enlargement of IVC
- Spontaneous echo contrast
- Ventricular dilation and dysfunction
- Progressive atrial enlargement
- Ascites or pleural effusion
- Atrial arrhythmias
Echo After Fontan Rhythm Disturbances

Echocardiography in Pediatrics and Adult CHD, 2nd Ed

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Thrombus in Right Atrium
Thrombus in MPA

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Thrombus in Fontan Conduit
Single Ventricle Function

Quantitative Methods:
- Fractional Area Change
- M-mode unreliable
- Tissue Doppler Imaging
- Myocardial Deformation

Value of quantitative methods is in following them serially to outline trends in the same patient.
Ventricular Dysfunction
Failed Fontan
Single Ventricle Palliation

- Delineate the anatomy
- Assess SV physiology
- Recognize the complications