Hypoplastic left heart syndrome

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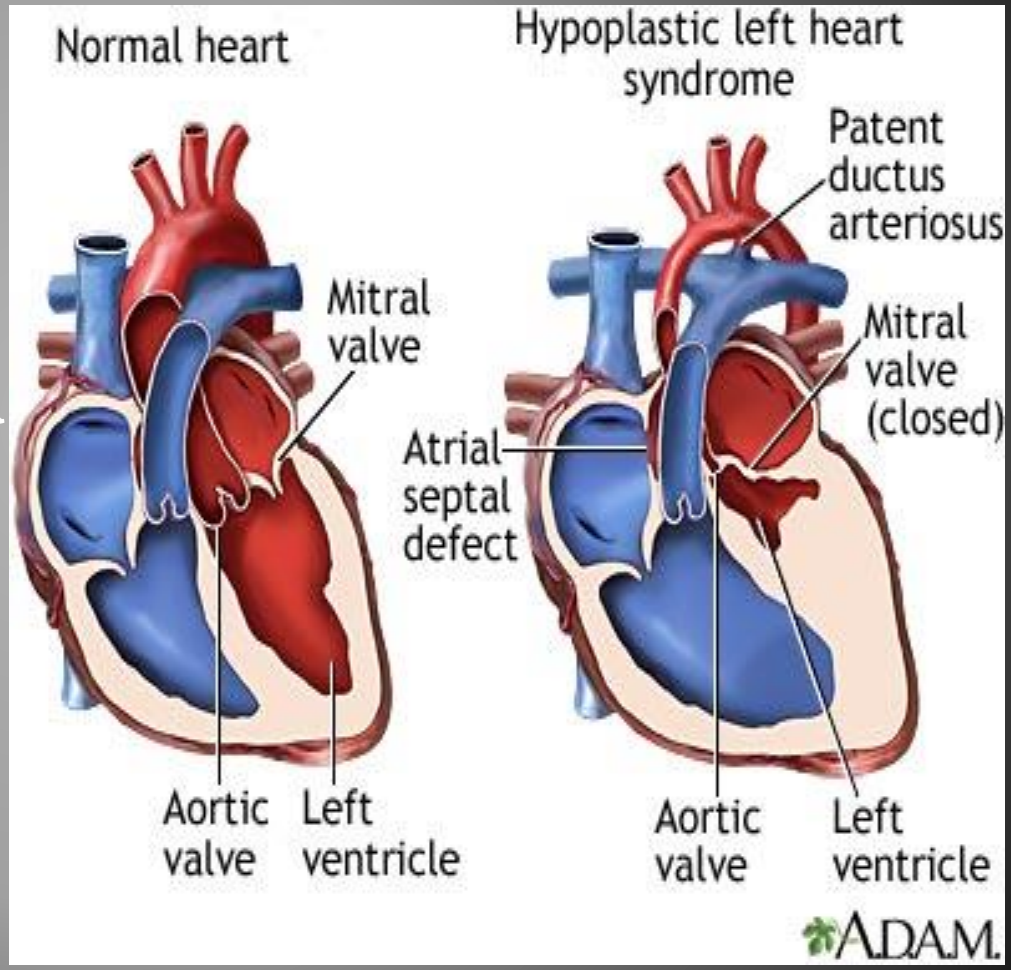
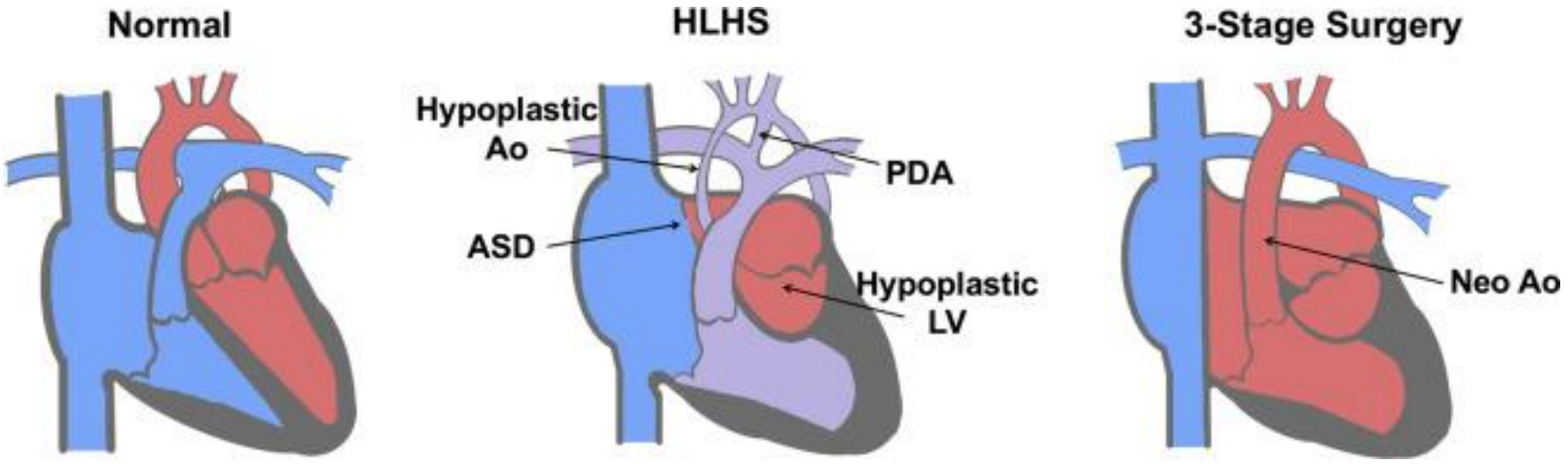
Anatomic features of HLHS

- Aortic valve stenosis/atresia
- Mitral valve stenosis/atresia
- Left ventricle – poor function; hypoplasia or dilation; endocardial fibroelastosis
- Aortic arch hypoplasia, coarctation, or interruption





Pre- and post-operative HLHS anatomy. Features of HLHS include: (1) hypoplastic ascending aorta (Ao); (2) hypoplastic LV; (3) large patent ductus arterious (PDA), supplying the only source of blood flow to the body; and (4) an atrial septal defect (ASD)

D. Woodrow Benson, Lisa J. Martin, Cecilia W. Lo

Genetics of Hypoplastic Left Heart Syndrome

The Journal of Pediatrics, Volume 173, 2016, 25–31

http://dx.doi.org/10.1016/j.jpeds.2016.02.052

Three stage palliation

Goals of stage one:

- Relieve systemic outflow tract obstruction
- Provide adequate coronary blood flow
- Provide pulmonary blood flow
- Create non-restrictive atrial level shunt

Goals of stage two:

- Eliminate the high pressure source (arterial or systemic ventricular) of pulmonary blood flow by connecting SVC to RPA
- Reduce pressure, volume, and work for single ventricle
- Improve circulatory efficiency – source of pulmonary blood flow more desaturated, higher arterial saturation

Third stage:

- Completes extracardiac pulmonary parallel circulation
- Directs remaining desaturated blood returning from IVC to the pulmonary arteries

Stage I Considerations

Goals of stage one:

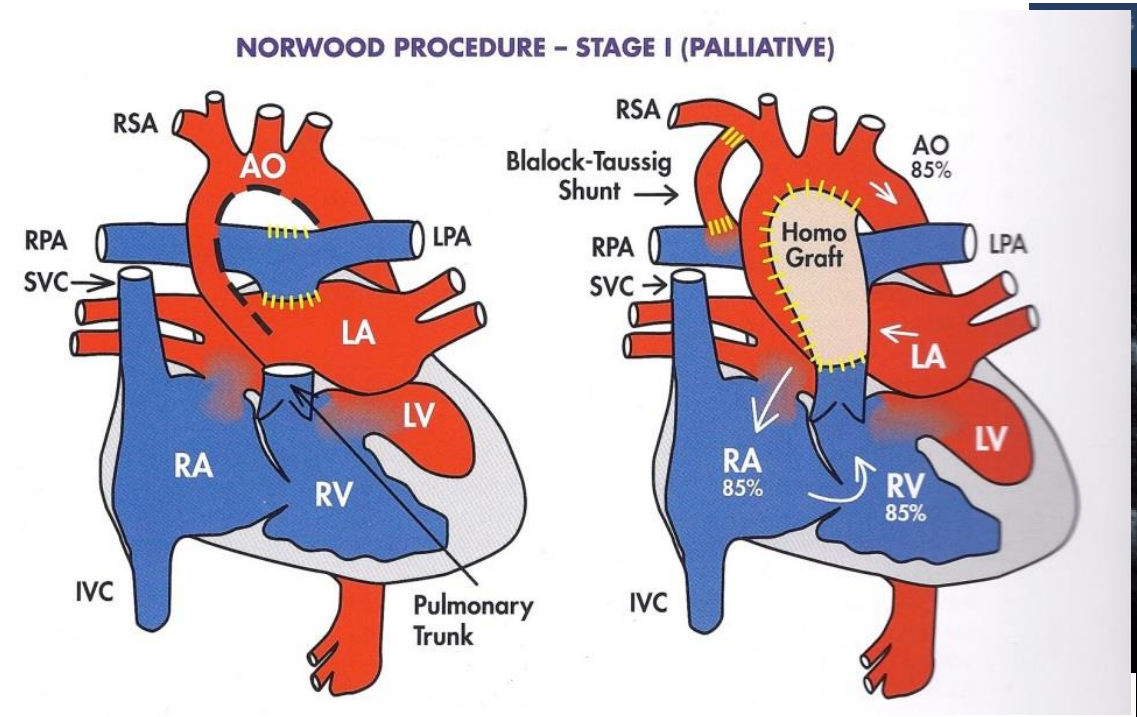
- Relieve systemic outflow tract obstruction
- Provide adequate coronary blood flow
- Provide pulmonary blood flow
- Create non-restrictive atrial level shunt

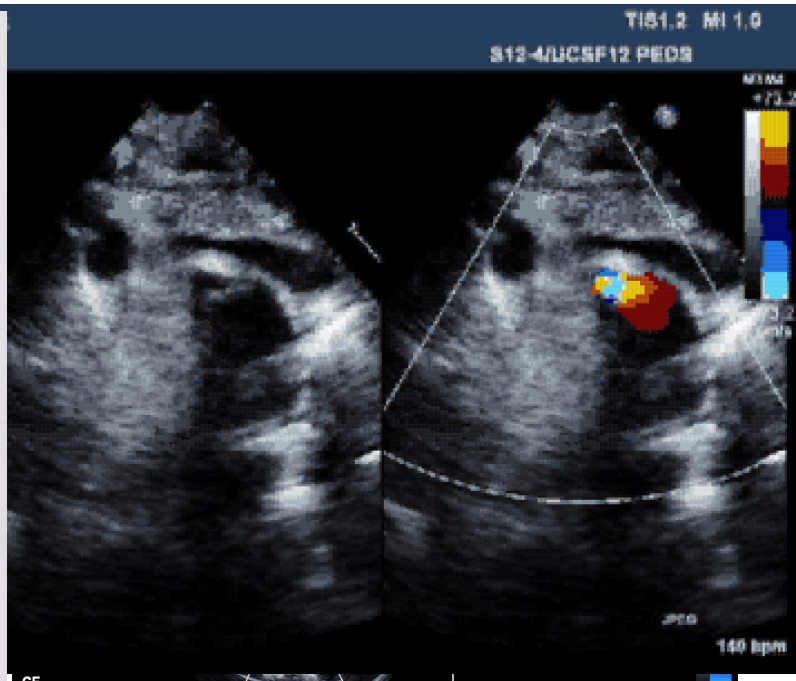
Three strategies:

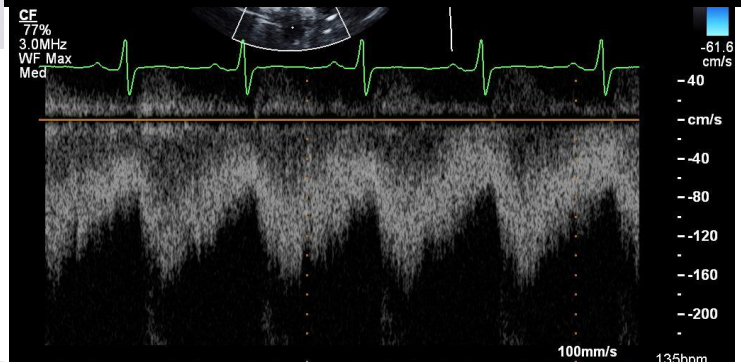
- Norwood procedure
  - With modified BT shunt
  - With RV-PA conduit
- Hybrid palliation
  - Surgical bilateral PA banding
  - Transcatheter ductal stenting
- Orthotopic heart transplantation

Norwood: modified BT shunt

Coronary “steal” due to continuous forward flow into BTS

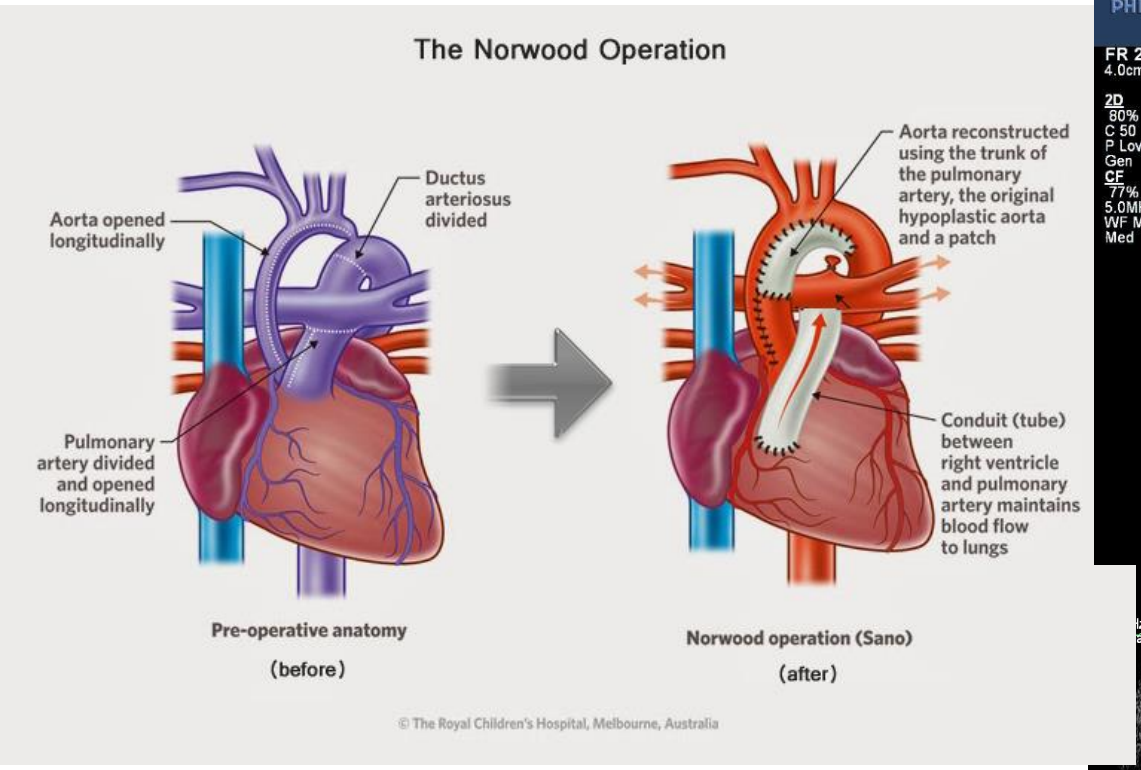


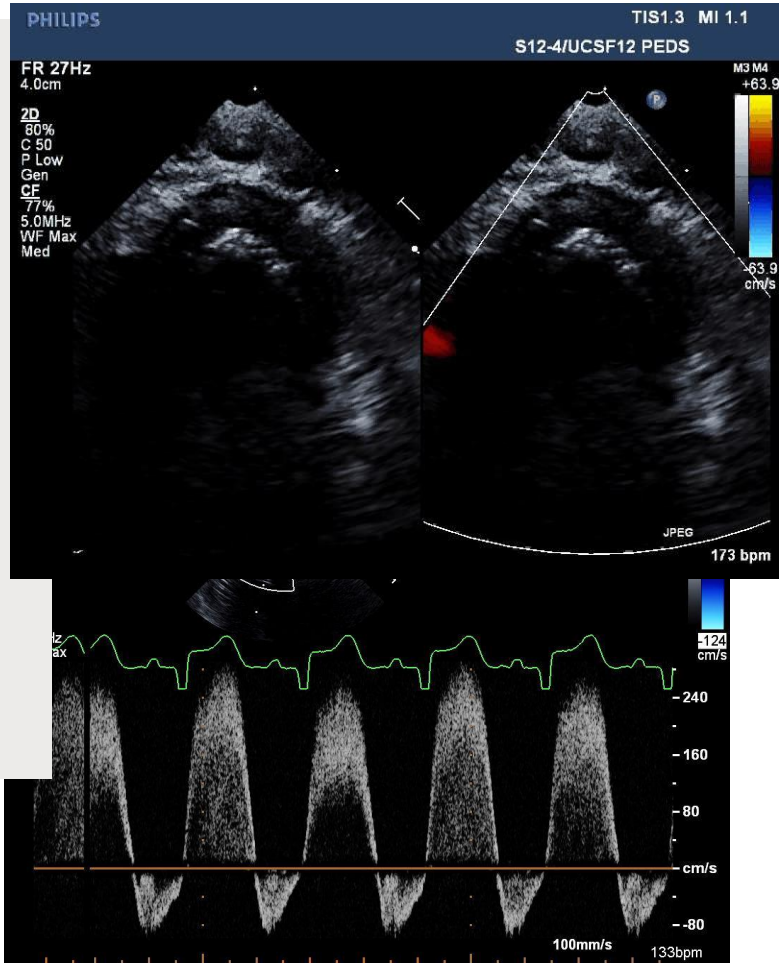




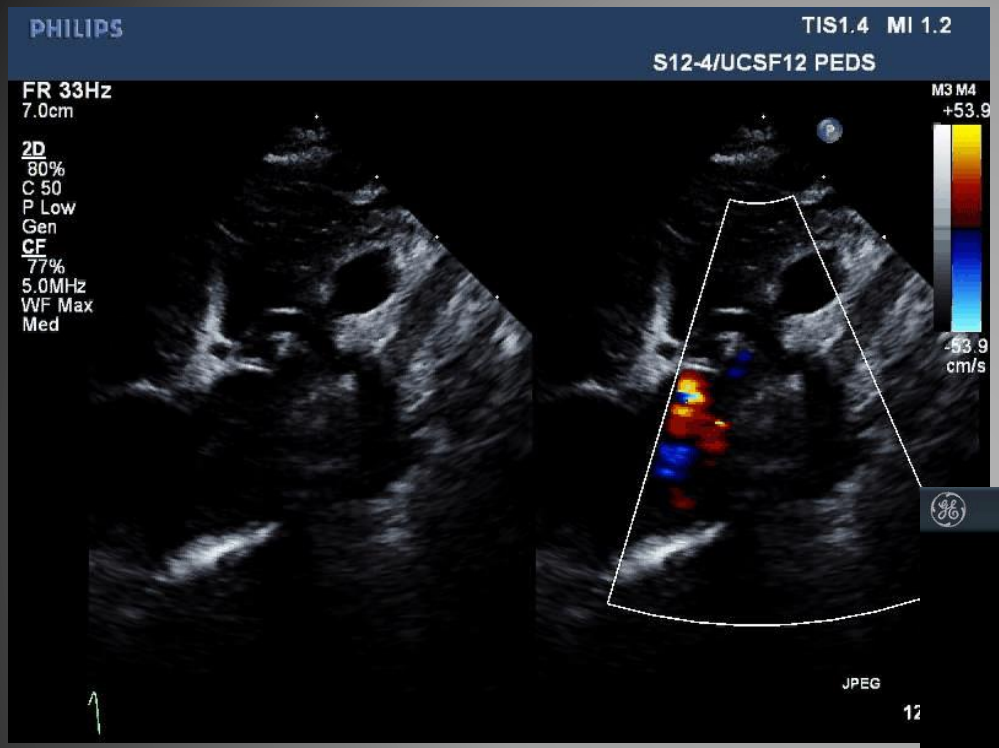
Norwood: RV-PA conduit

Ventriculotomy in systemic ventricle may cause diminished function and possibly arrhythmias





Norwood: DKS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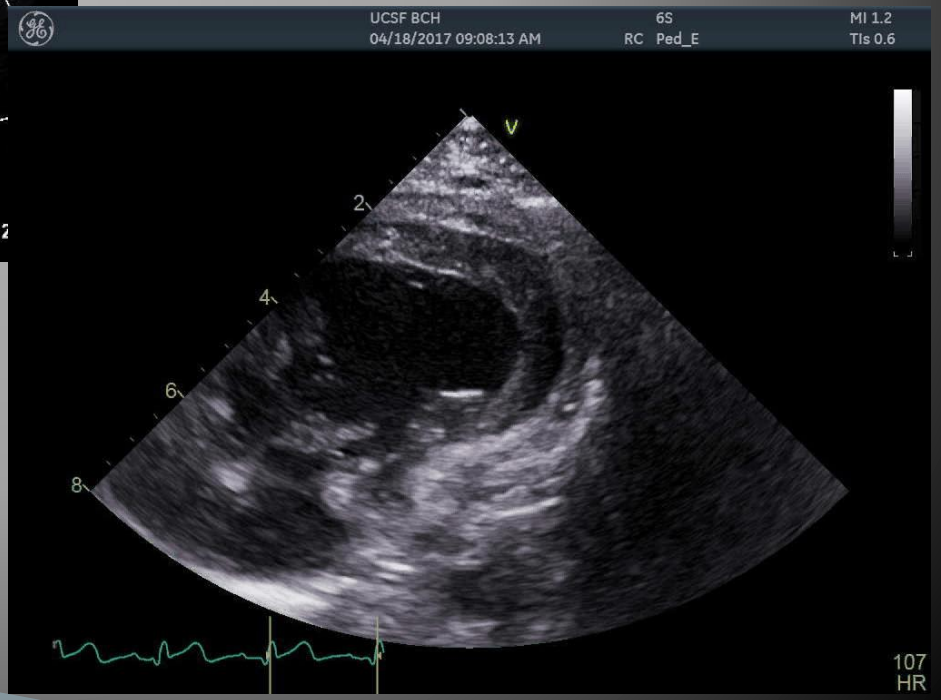
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Stages II & III Considerations

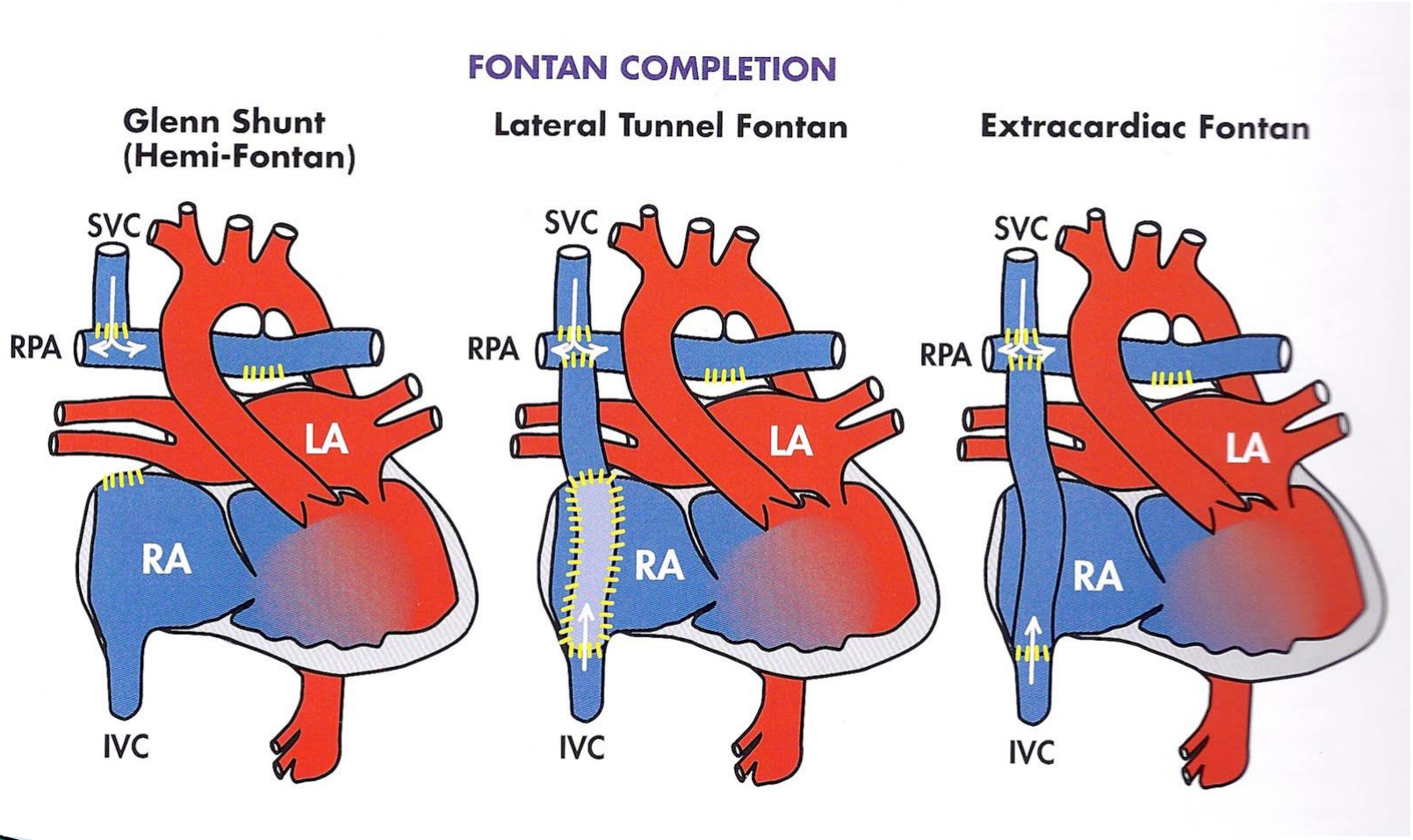
Goals of stage two:

- Eliminate the high pressure source (arterial or systemic ventricular) of pulmonary blood flow by connecting SVC to RPA
- Reduce pressure, volume, and work for single ventricle
- Improve circulatory efficiency – source of pulmonary blood flow more desaturated, higher arterial saturation

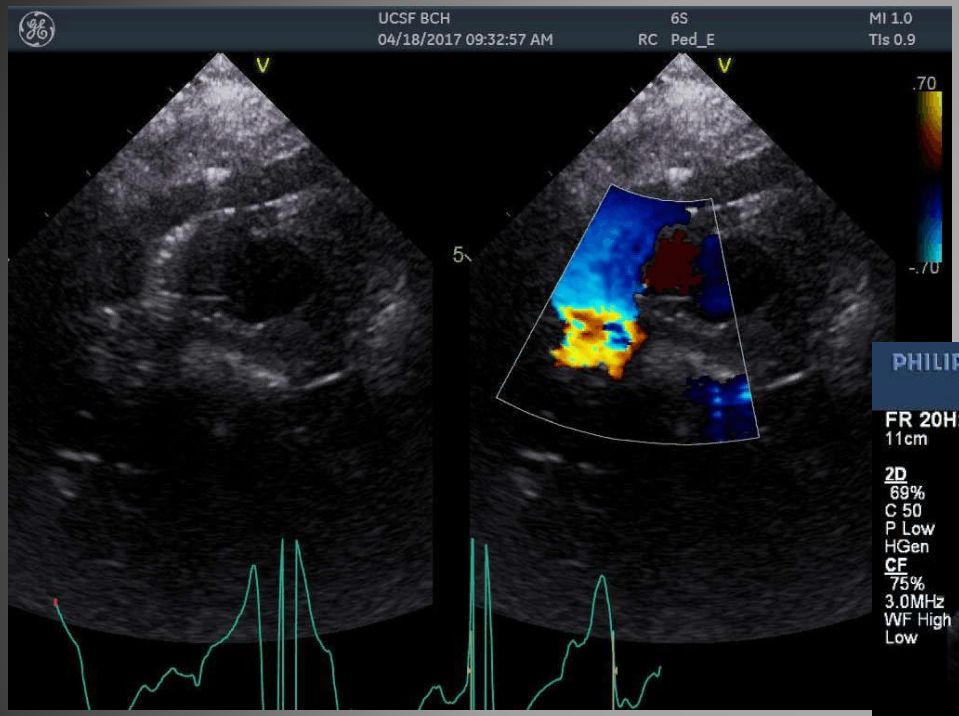
Third stage:

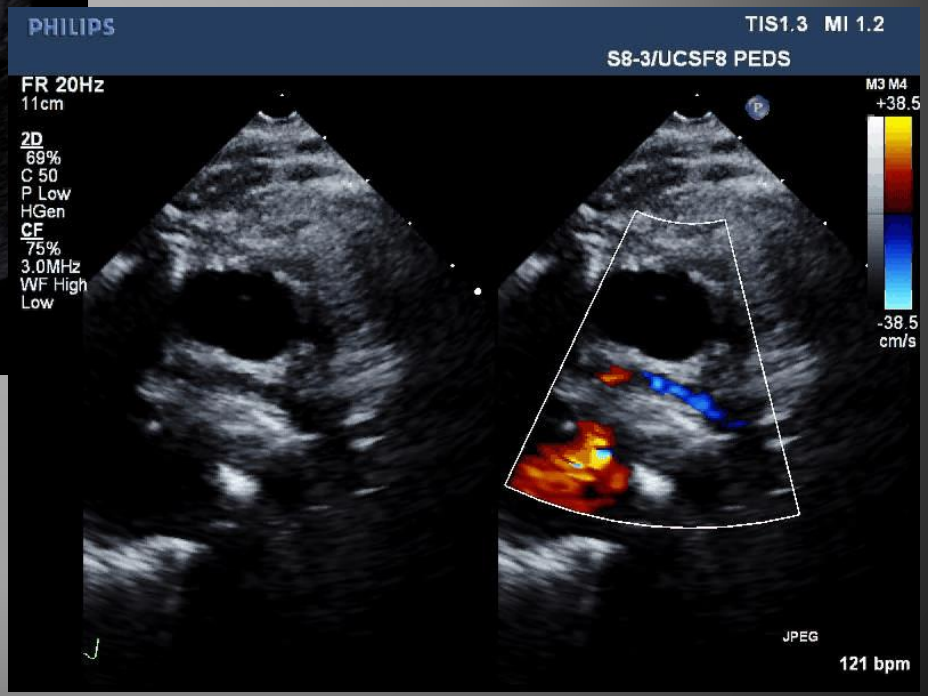
- Completes extracardiac pulmonary parallel circulation
- Directs remaining desaturated blood returning from IVC to the pulmonary arteries

Glenn and Fontan

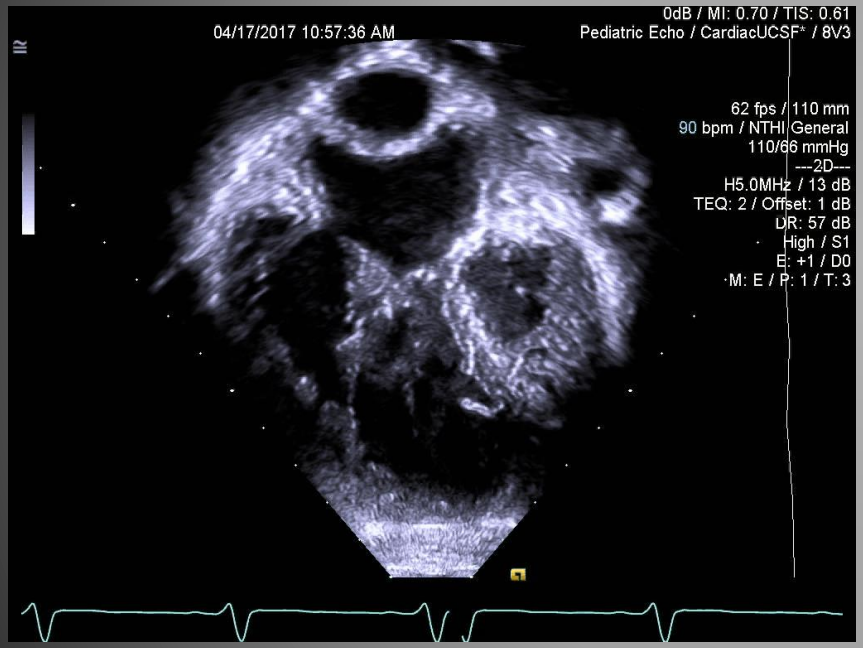


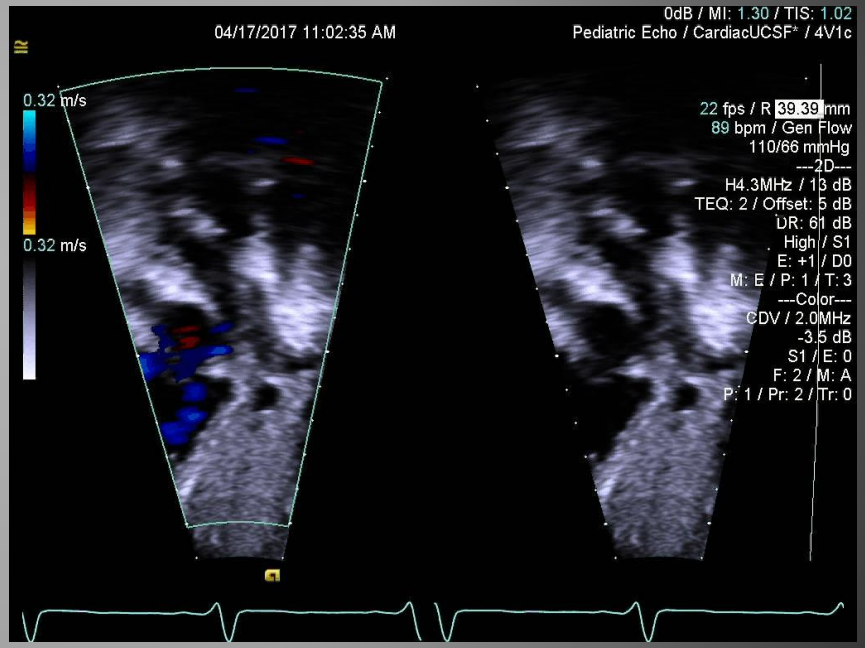
Glenn: superior cavopulmonary anastamosis





Fontan: completion of extracardiac pulmonary circulation





Etiology

- Heterogeneous disease; variety of theories
  - Mitral and/or aortic valve stenosis altering flow into the developing LV
  - Abnormalities of the atrial septum limit right to left flow, altering filling and subsequent hypoplasia of the LV
  - Strong genetic influence: Turner's syndrome; mutations associated with HLHS have been noted in genes *NKX2-5*, connexin 43, and *NOTCH1*

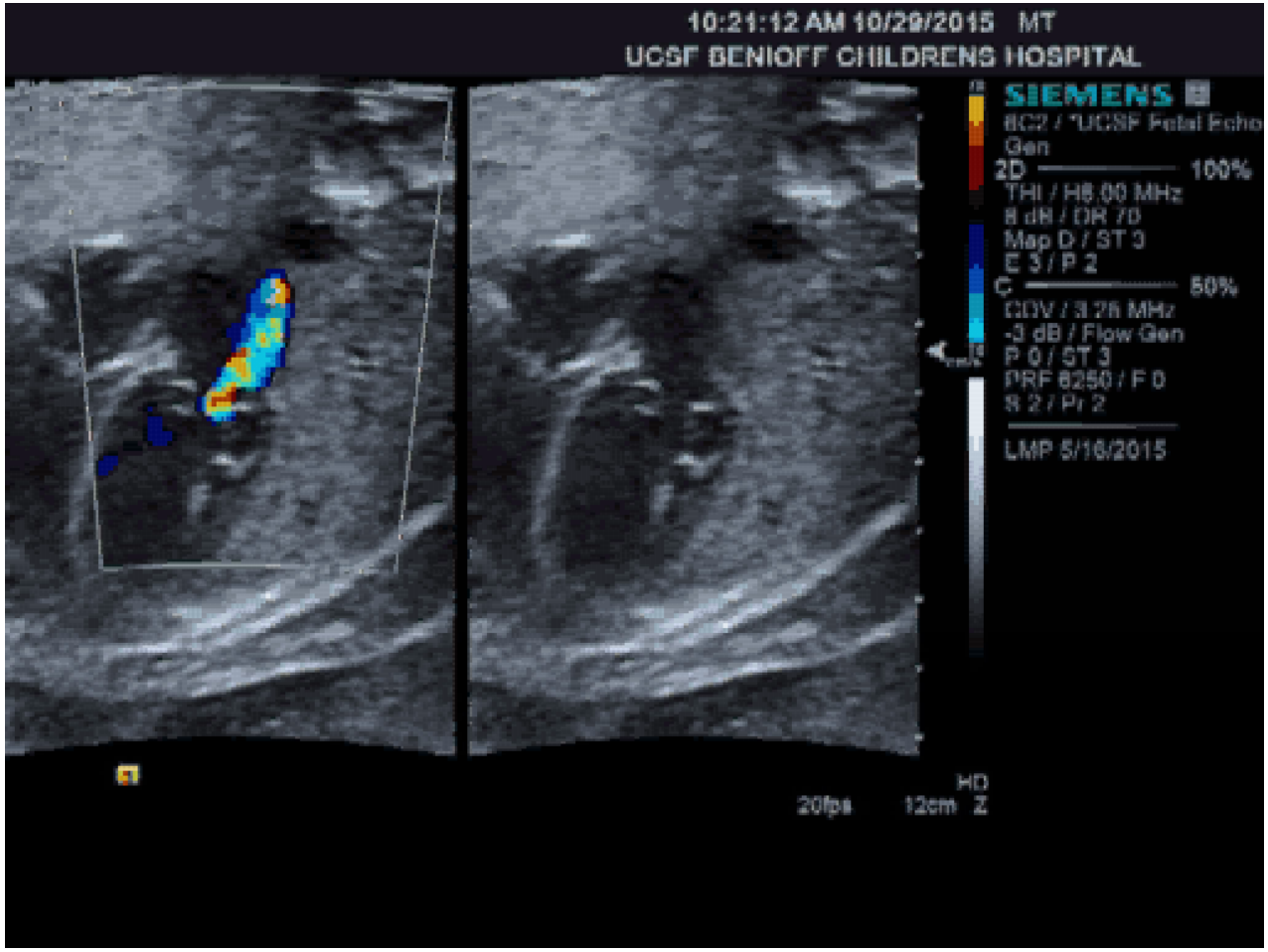
Incidence and associated anomalies

- Prevalence of HLHS is 2.6 per 10,000 births (approximately 2000 live births annually in the US)
- Despite the low prevalence, HLHS accounts for 4-8% of all CHD, and ~25% of deaths in infants with CHD
- Approximately 30% of fetuses with HLHS have genetic syndromes and/or the presence of extra-cardiac anomalies (e.g. Turner syndrome)
- Marked genetic heterogeneity in individuals with HLHS; genetic variants in *NOTCH1, NKX2.5, ERBB4 and HAND1*
- Strong familial clustering of HLHS with CHD in families; some type of CHD is found in 18% of first degree relatives in families with HLHS
- HLHS is genetically heterogeneous and inherited in a complex manner.

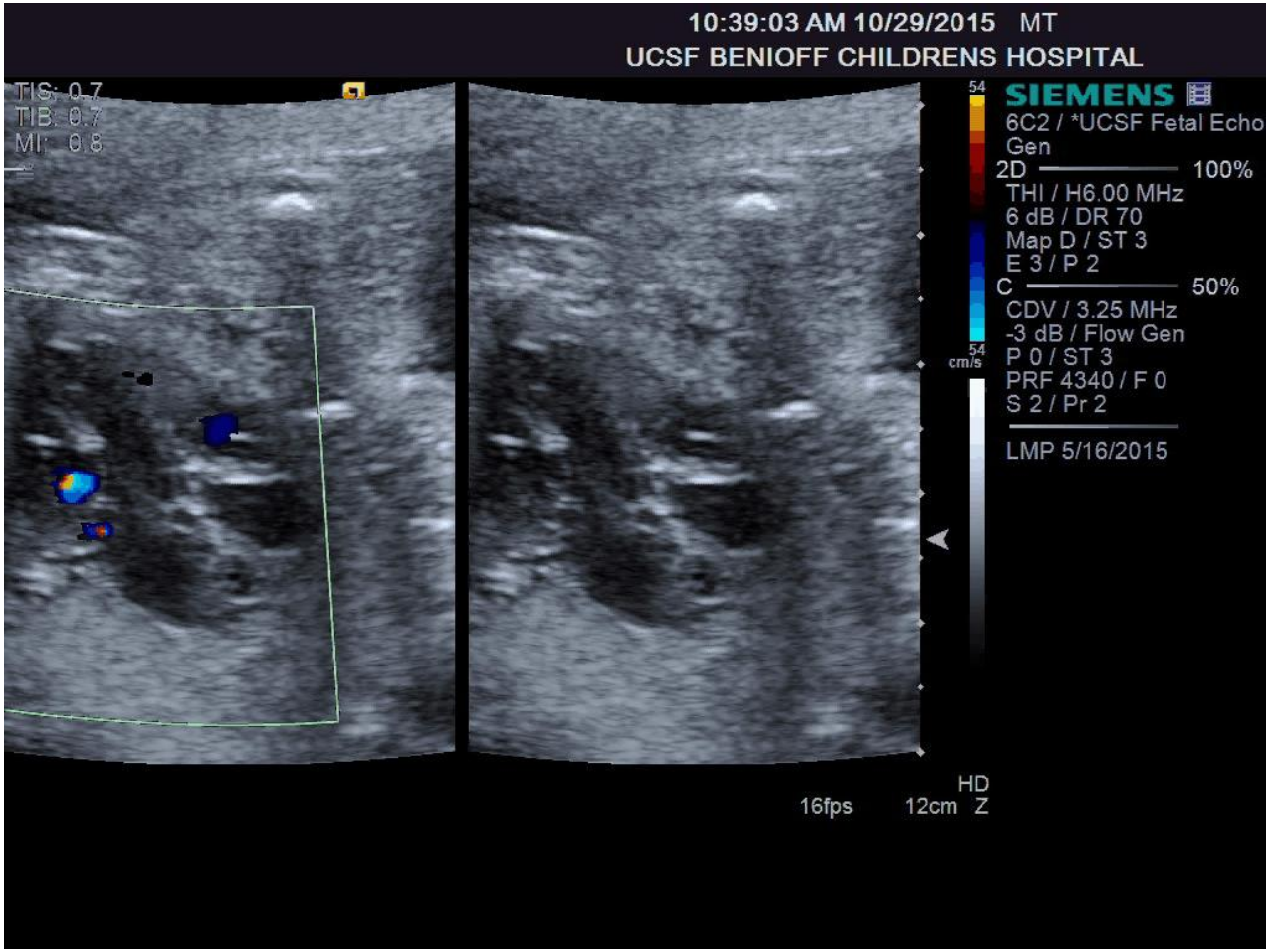
Same mom; two different fathers

- 11 y.o. boy—Interrupted aortic arch, VSD
  - Norwood and RV-PA conduit
  - Bidirectional superior cavopulmonary anastamosis
  - Yasui with baffle closure of VSD (both native aortic and pulmonic valves arise from left ventricle) with preservation of Glenn
- 15 month old boy—critical AS, MR, EFE
  - Fetal aortic balloon valvuloplasty
  - Post-natal aortic balloon valvuloplasty
  - Ross-Konno, MV annuloplasty suture, EFE resection
  - RV-PA conduit stent placement (RCA occlusion)
  - RCA unroofed, RV-PA conduit replacement

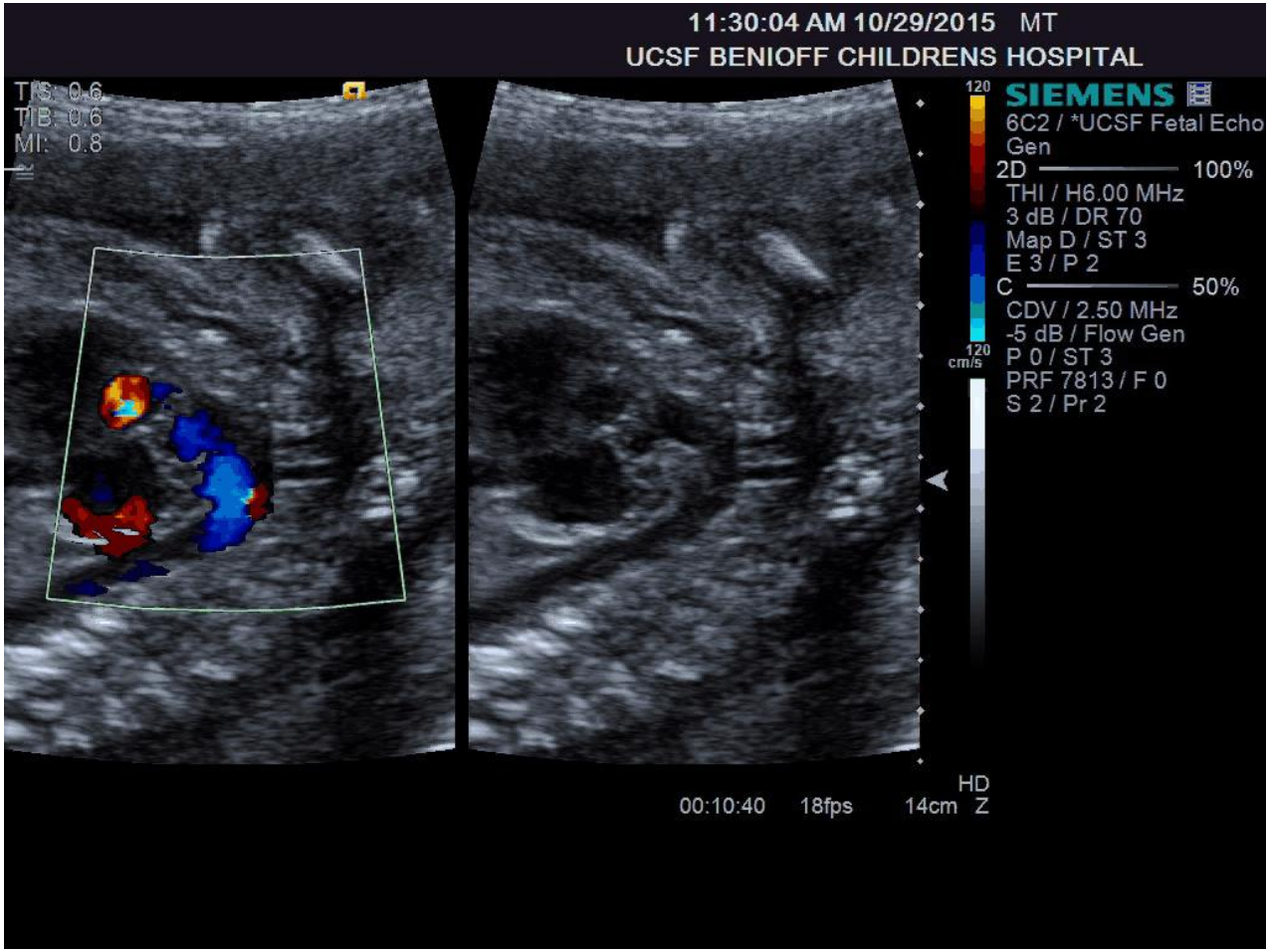
Evolving HLHS? Mitral regurgitation



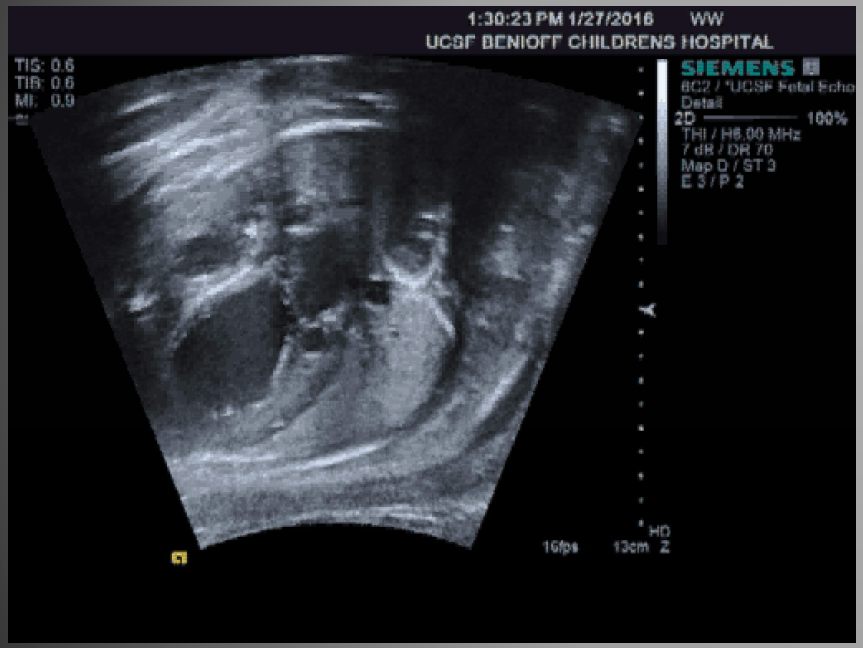
Aortic stenosis and insufficiency

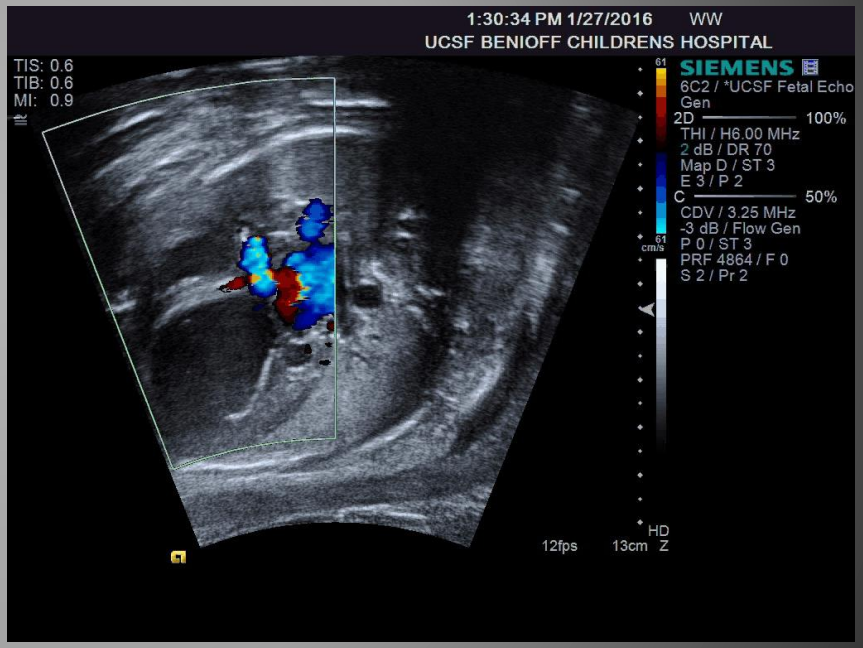


Coarctation of the aorta



After fetal aortic valvuloplasty





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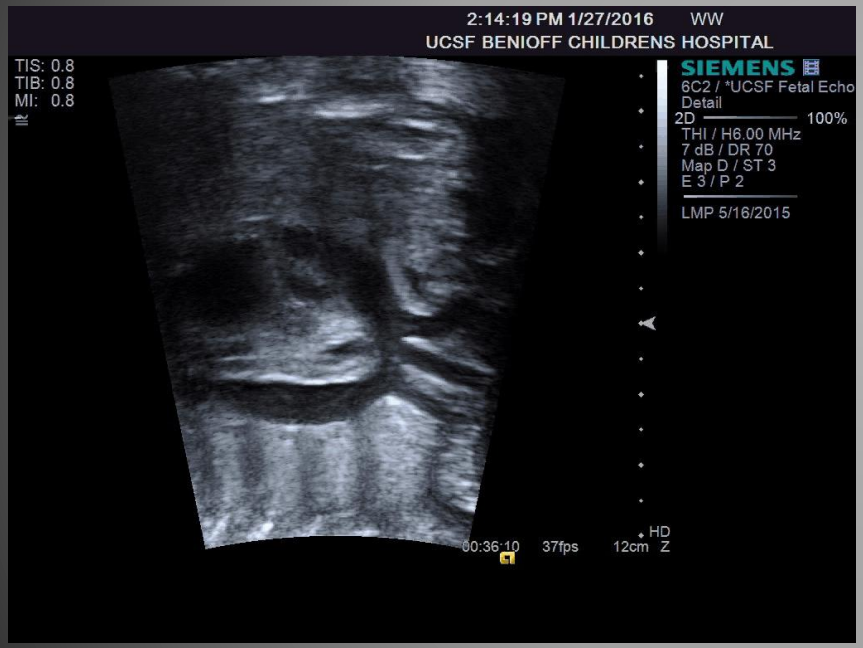
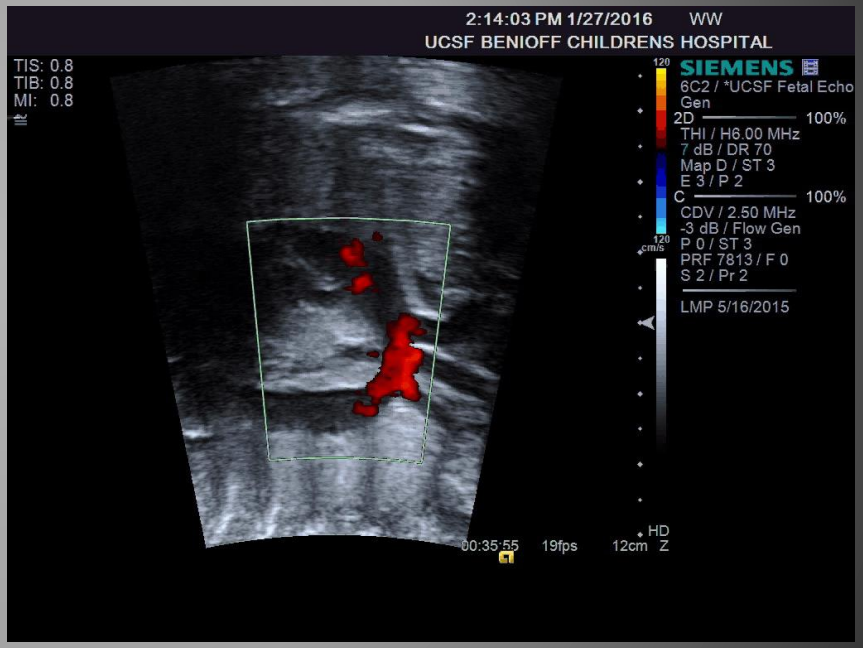
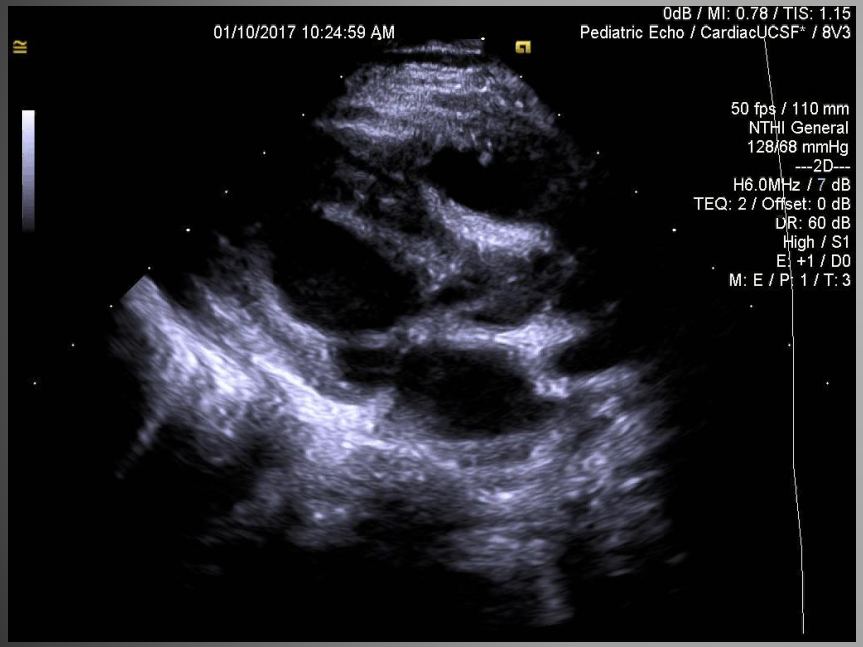
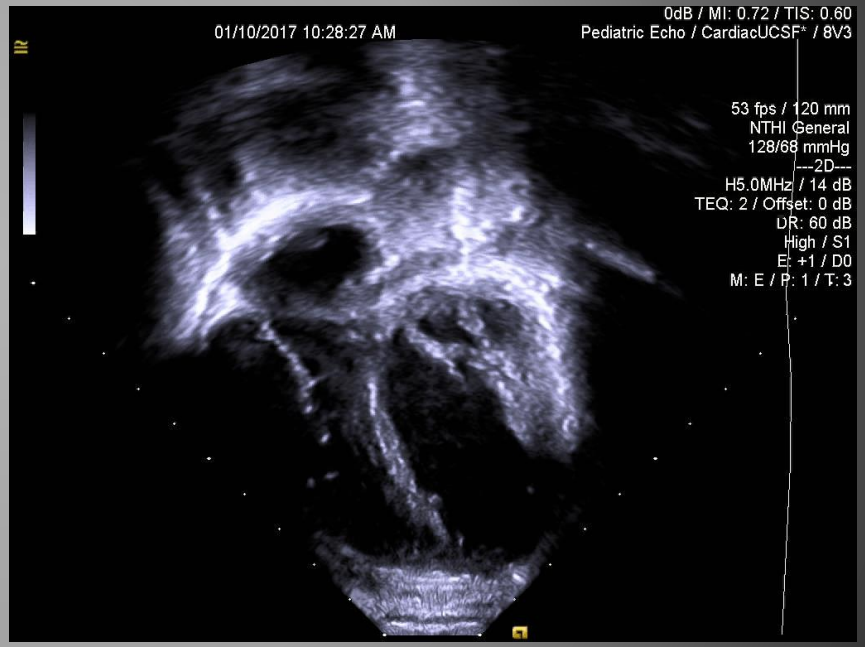




15 month old—biventricular repair





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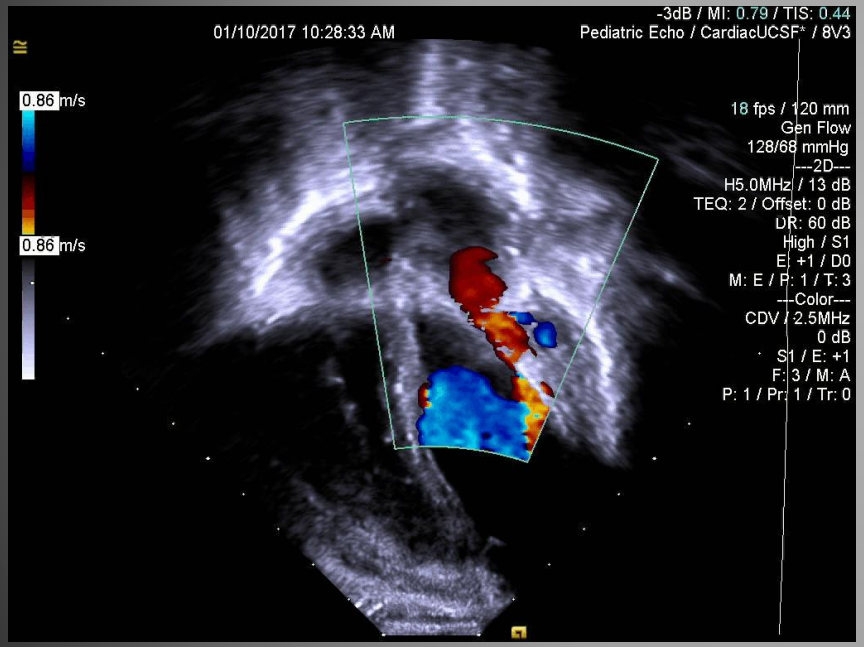

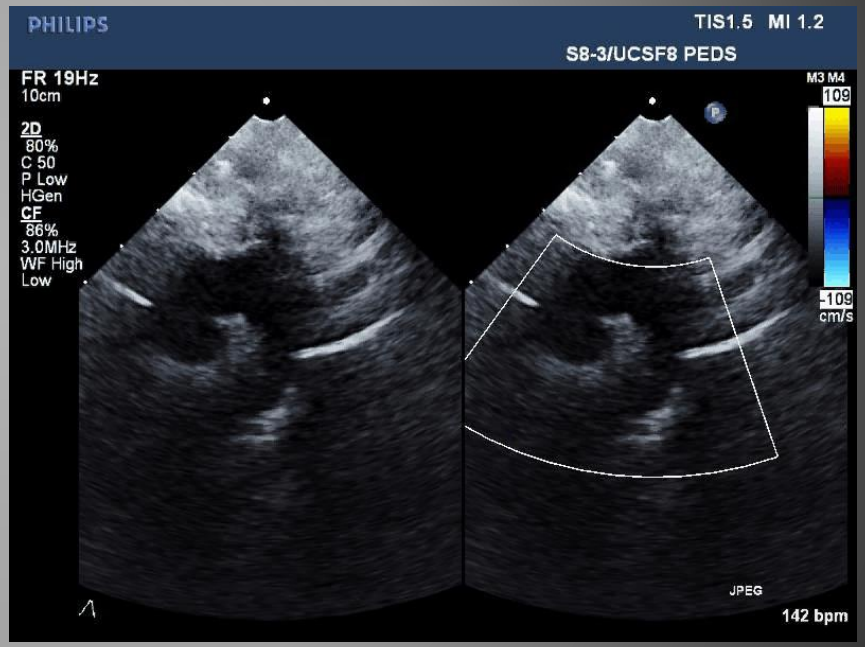

HLHS: beyond surgery

- Quality of life
  - Multiple hospitalizations
  - Psychosocial impact on the entire family
- Neurological development
  - Fetal circulation
  - Multiple cardiac bypass runs
- “Failing Fontan” physiology
  - PLE
  - Plastic bronchitis
  - Arrhythmias

J Am Coll Cardiol. 2012 Jan 3;59(1 Suppl):S1-42. doi: 10.1016/j.jacc.2011.09.022.

Hypoplastic left heart syndrome: current considerations and expectations.

Feinstein JA1, Benson DW, Dubin AM, Cohen MS, Maxey DM, Mahle WT, Pahl E, Villafañe J, Bhatt AB, Peng LF, Johnson BA, Marsden AL, Daniels CJ, Rudd NA, Caldarone CA, Mussatto KA, Morales DL, Ivy DD, Gaynor JW, Tweddell JS, Deal BJ, Furck AK, Rosenthal GL, Ohye RG, Ghanayem NS, Cheatham JP, Tworetzky W, Martin GR.

Erratum in
J Am Coll Cardiol. 2012 Jan 31;59(5):544.

Abstract

In the recent era, no congenital heart defect has undergone a more dramatic change in diagnostic approach, management, and outcomes than hypoplastic left heart syndrome (HLHS). During this time, survival to the age of 5 years (including Fontan) has ranged from 50% to 69%, but current expectations are that 70% of newborns born today with HLHS may reach adulthood. Although the 3-stage treatment approach to HLHS is now well founded, there is significant variation among centers. In this white paper, we present the current state of the art in our understanding and treatment of HLHS during the stages of care: 1) pre-Stage I: fetal and neonatal assessment and management; 2) Stage I: perioperative care, interstage monitoring, and management strategies; 3) Stage II: surgeries; 4) Stage III: Fontan surgery; and 5) long-term follow-up. Issues surrounding the genetics of HLHS, developmental outcomes, and quality of life are addressed in addition to the many other considerations for caring for this group of complex patients.