Hypoplastic Left Heart Syndrome(HLHS)

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HLHS

Hypoplastic left heart syndrome is a collection of various congenital heart anomalies in which the left ventricle is incapable of providing adequate perfusion.

May include abnormalities such as mitral stenosis or atresia,LV hypoplasia, aortic stenosis or atresia and ascending aorta hypoplasia

HLHS

Is one of the most severe forms of congenital heart anomalies and accounts for 20-25% of all mortalities in infants born with congenital heart disease

The incidence is 266 per 1 million live births

HLHS

Turner syndrome has been known to be associated with HLHS. Other syndromes include Smith-Lemi-Opitz,Holt-Oram,and Jacobsen.

Congenital brain anomalies have also been identified with HLHS including agenesis of the corpus callosum and a overall immature appearance

HLHS

The etiology of HLHS is still being researched and the precise cause is unknown. There are several theories as to how it develops.

One theory is altered flow into the developing LV due to aortic and mitral stenosis and/or hypoplasia

Another theory is abnormalities of the atrial septum that limits the amount of right to left shunting during fetal life leading to reduced filling of the LV

HLHS is a heritable disorder with a strong genetic influence

HLHS

HLHS can be divided into the following major anatomic groups

- mitral and aortic stenosis
- mitral stenosis and aortic atresia
- mitral and aortic atresia

And other anatomic variations exist

HLHS

Echo findings

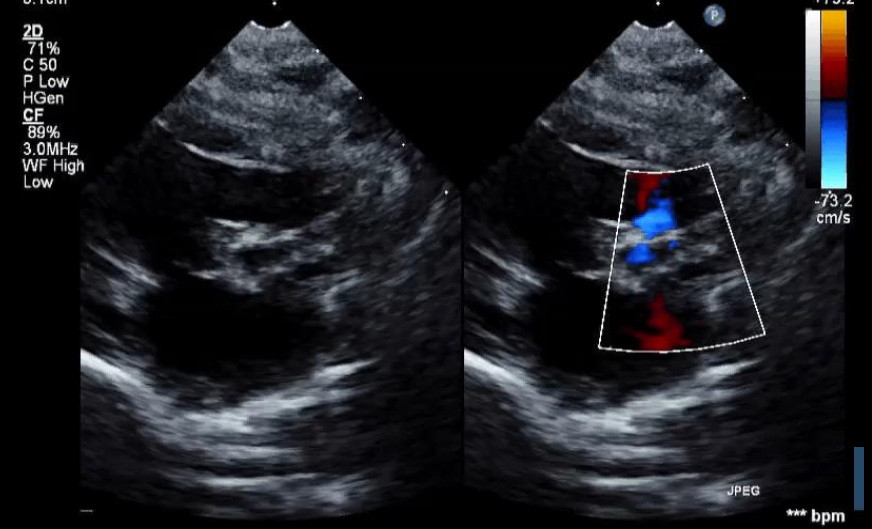
- The RSVC and IVC return to the right atrium normally with about 5% having a LSVC
- The right atrium is usually unaffected but could be dilated
- The left atrium is often small, but could be distended and hypertensive if the atrial septum in restrictve
- Abnormalities of the atrial septum are common for HLHS and may include primum and secundum atrial septal defects or abnormal postion of the atrial septum

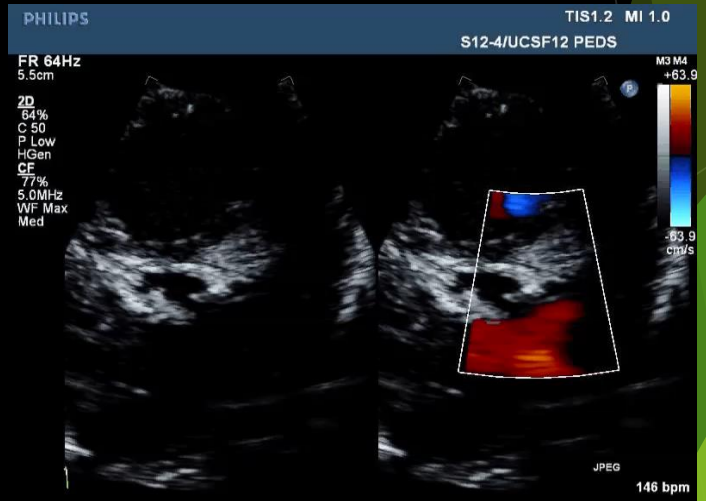
HLHS

Echo findings

- Pulmonary veins should be visualized to distinguish between anomalous pulmonary venous connection vs anomalous drainage (levoatriocardinal vein, decompressing vein)
- The mitral valve may be stenotic or atretic
- The left ventricle will contract poorly and may have endocardial fibroelastosis (EFE)
- PDA is vital for survival, during fetal life the PDA is the predominant avenue for systemic flow

- The ascending aorta varies in size and is related to the amount of antegrade flow, It is common that the ascending aorta is the size of a coronary artery 1-2mm
- Coarctation of the aorta is common and interrupted aortic arch has also been reported
- Coronary arteries in HLHS typically have normal ostia and arise normally from the aortic root and follow a normal course





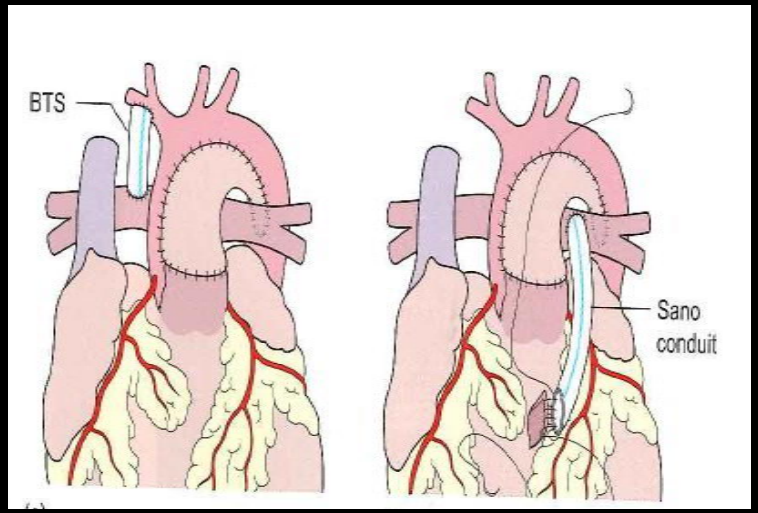
HLHS

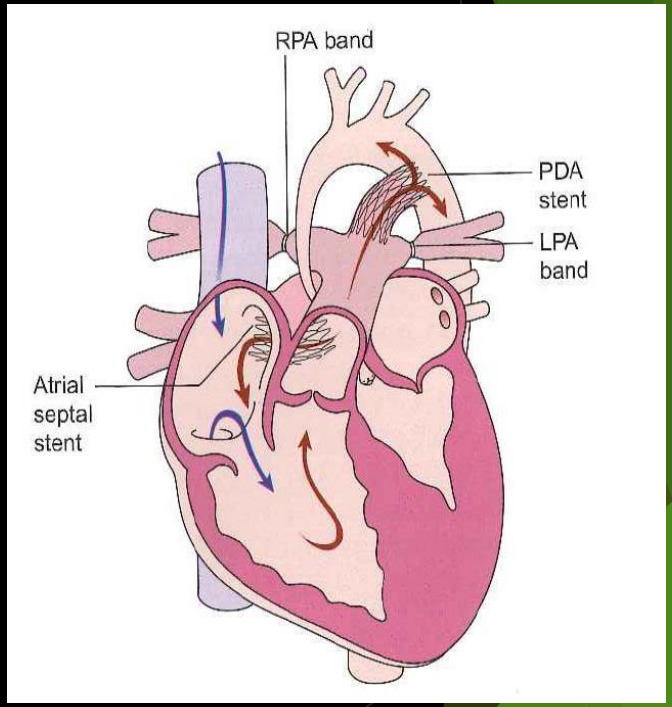
Pathophysiology

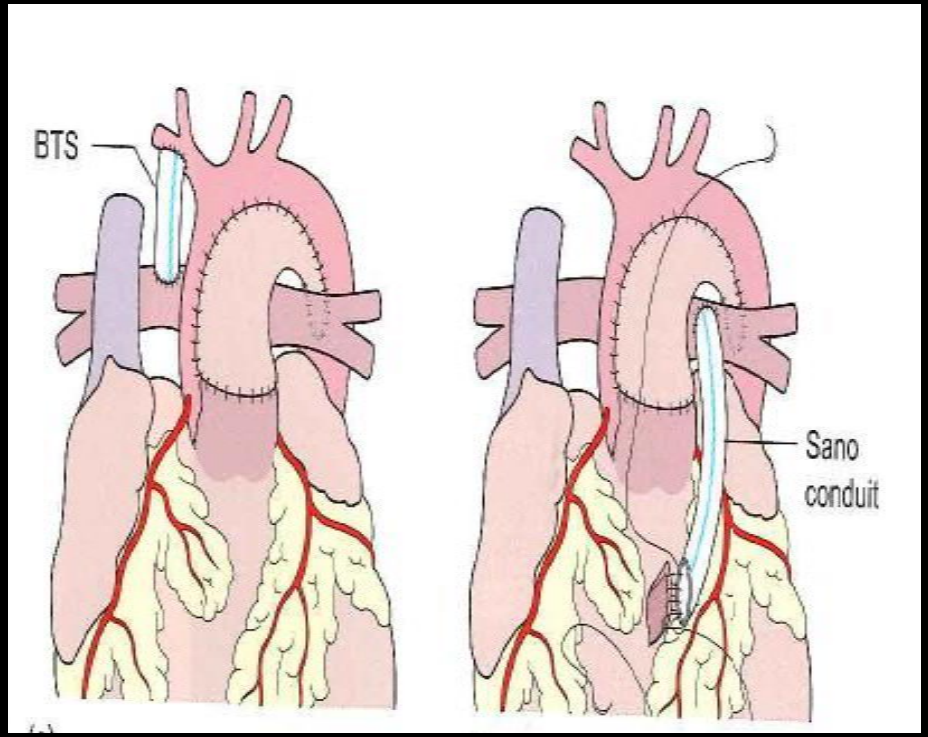
- Infants will typically manifest tachypnea and cyanosis at birth
- Peripheral pulses may be weak with cool extremities due to poor perfusion
- Chest x-ray will reveal a large heart with increased pulmonary vascular markings
- Postnatal survival depends upon patency of the PDA which supplies the systemic circulation
- A PDA supplies most, if not all, of the blood flow to the body
- Depending on the anatomic type of HLHS retrograde flow through the PDA into the aortic arch may also be the major or only source of blood supply to the upper extremities, head and coronary arteries

HLHS

Management

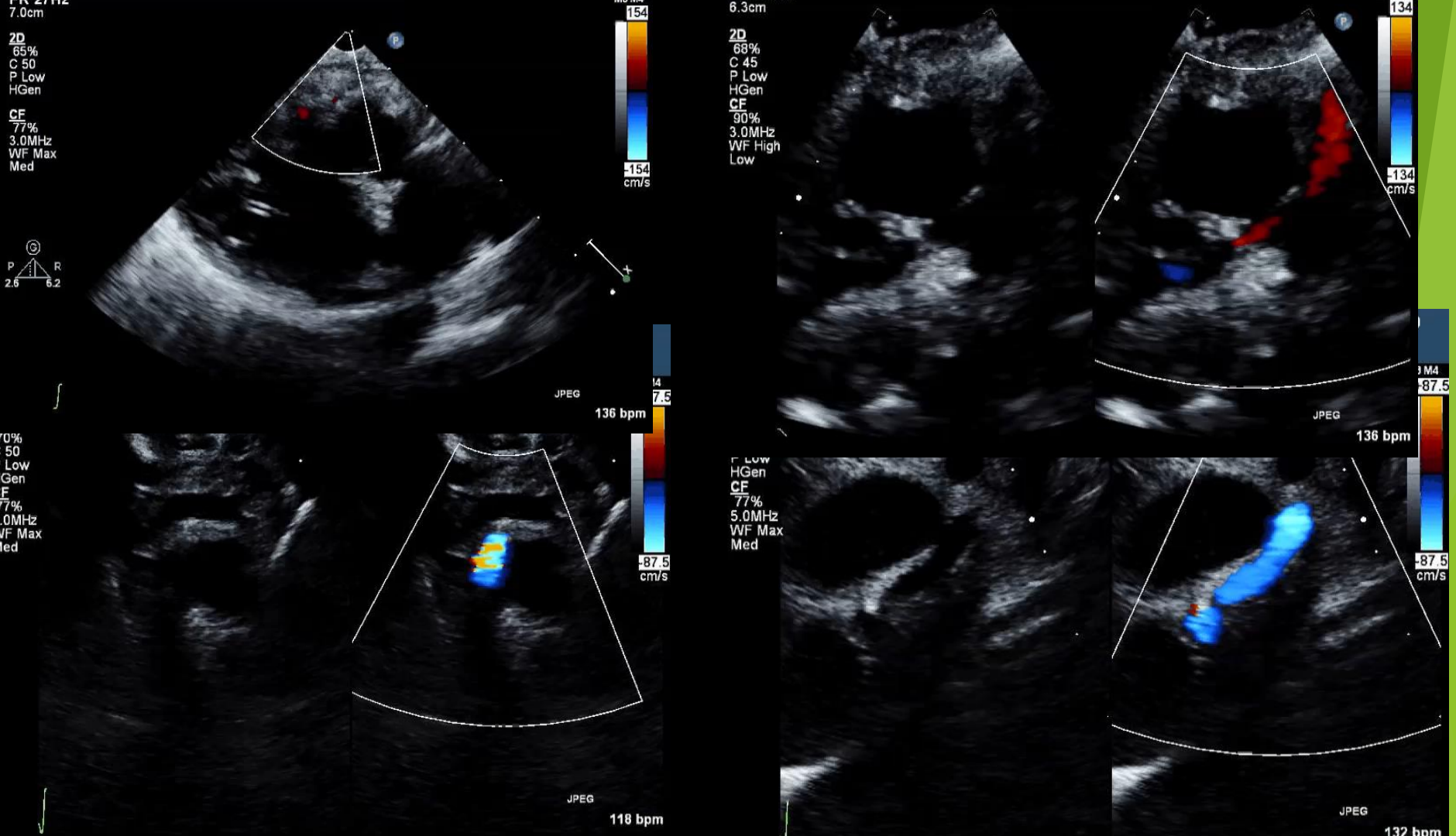






- There are a number of strategies available for neonates with HLHS
- Primary cardiac transplantation is practiced at a small number of institutions in the U.S. However, limited donor availability has decreased the wide spread use of this practice as a primary means of therapy
- Stage 1 reconstruction or Norwood procedure is the most common palliation for HLHS.

This operation consists of an atrial septectomy, transection of the pulmonary artery with oversewing of the distal stump, construction of a neoaorta using the incised native aorta augmented with a homograft patch sewn to the native proximal pulmonary artery, ligation of the PDA, and establishment of secure pulmonary blood flow using a modified BT shunt or Sano shunt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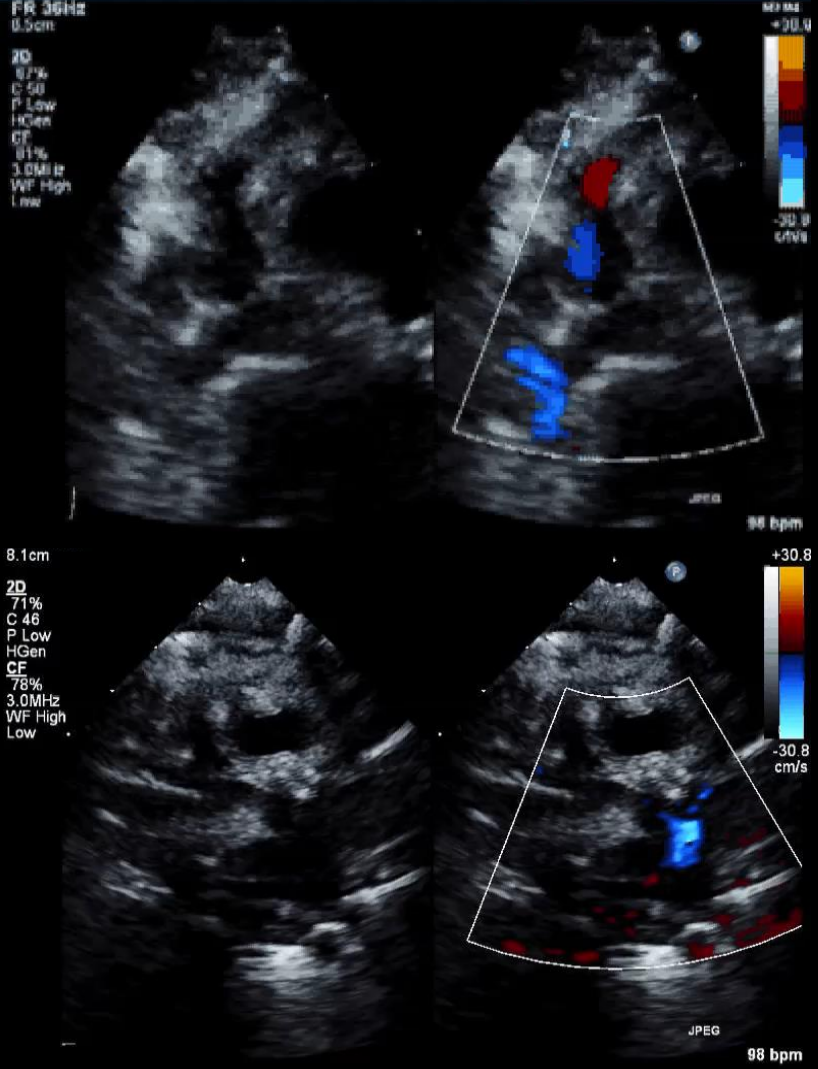
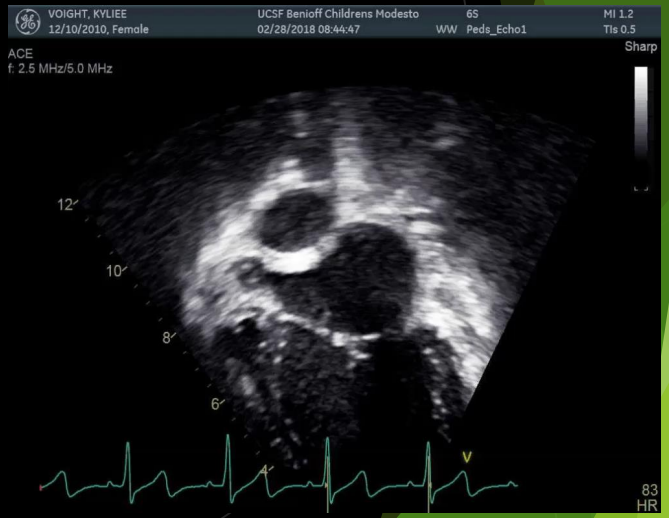


- In the second stage of palliation for HLHS, SVC blood is directed to the pulmonary arteries via a bidirectional Glenn
- The last stage is the Fontan procedure. IVC blood is directed into the pulmonary arteries bypassing the heart.

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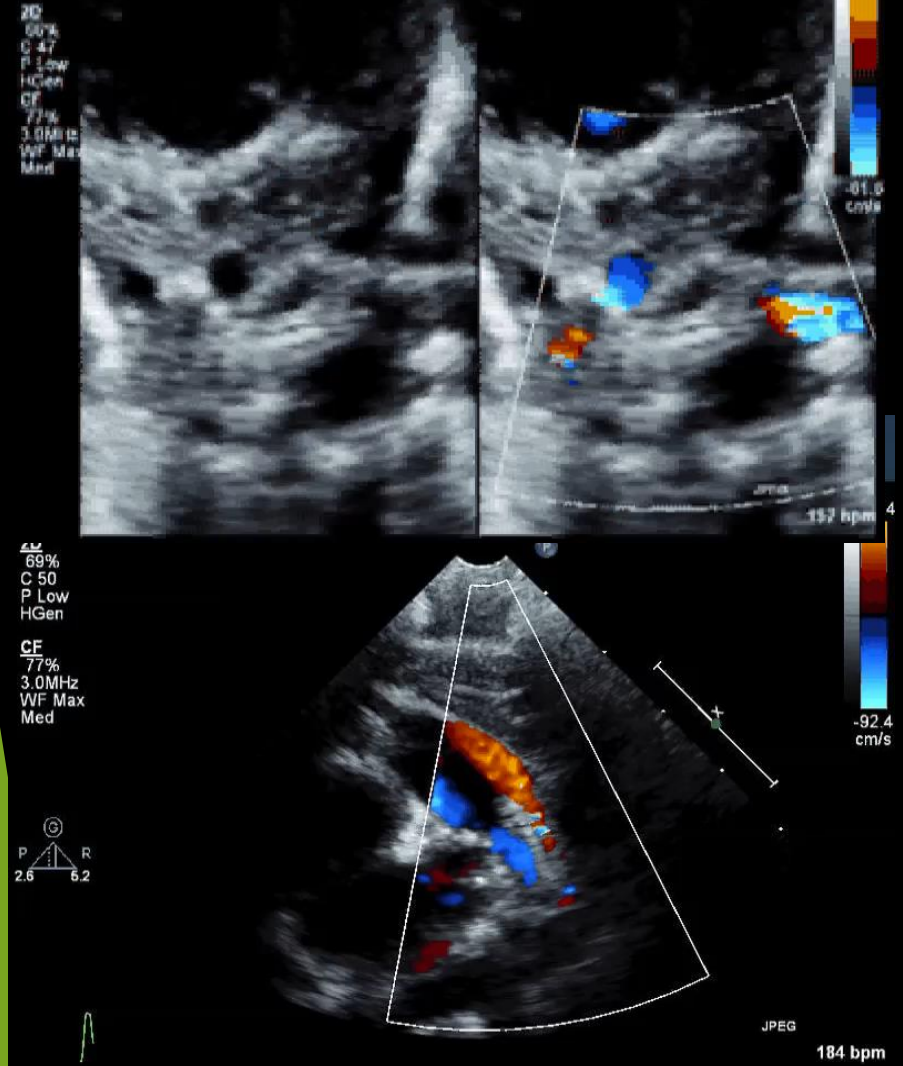
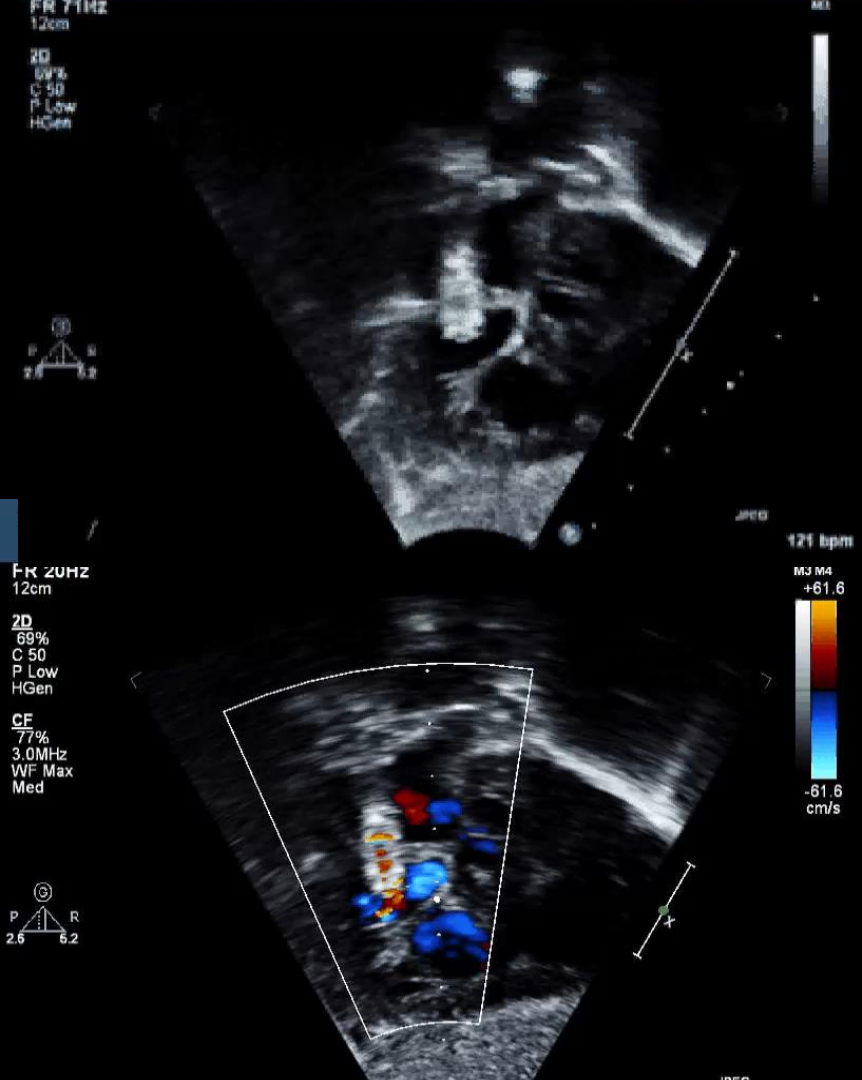


- Because mortality for neonatal Norwood reconstruction remains high in comparison with surgery for other forms of congenital heart disease, some have moved toward the development of strategies that avoid a major surgical operation in the neonatal period.

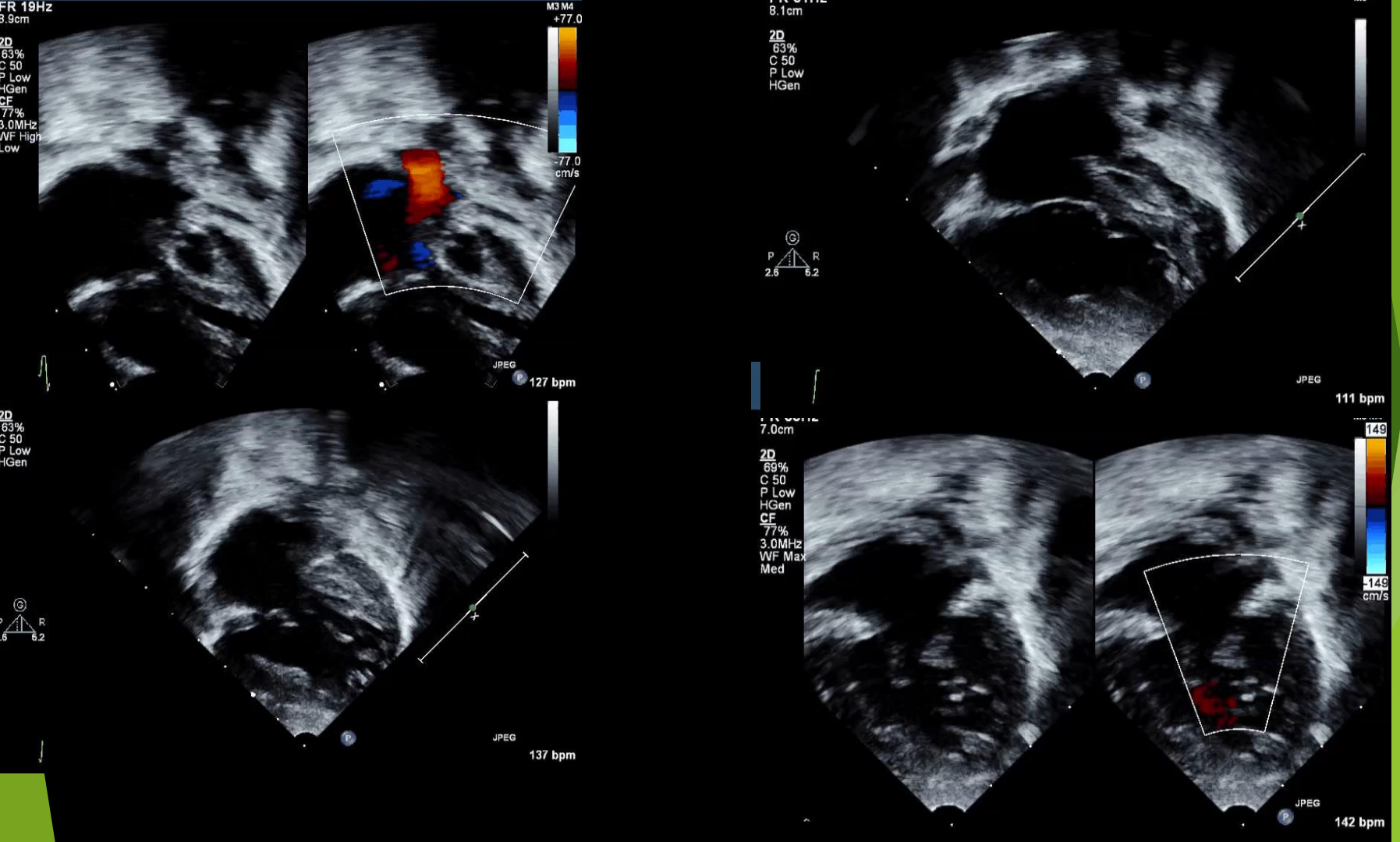
- The hybrid procedure

The hybrid procedure involves both cardiac catheter driven and surgical technique and achieves physiologic goals that are similar to the Norwood first stage reconstruction but without the performance of a challenging neonatal operation

By placing a stent in the PDA and banding the branch pulmonary arteries its possible of creating a physiologic situation where the RV supplies the systemic and pulmonary circulation creating unimpeded flow to the descending aorta and protecting the pulmonary vasculature.



- The atrial septum is also stented in the hybrid procedure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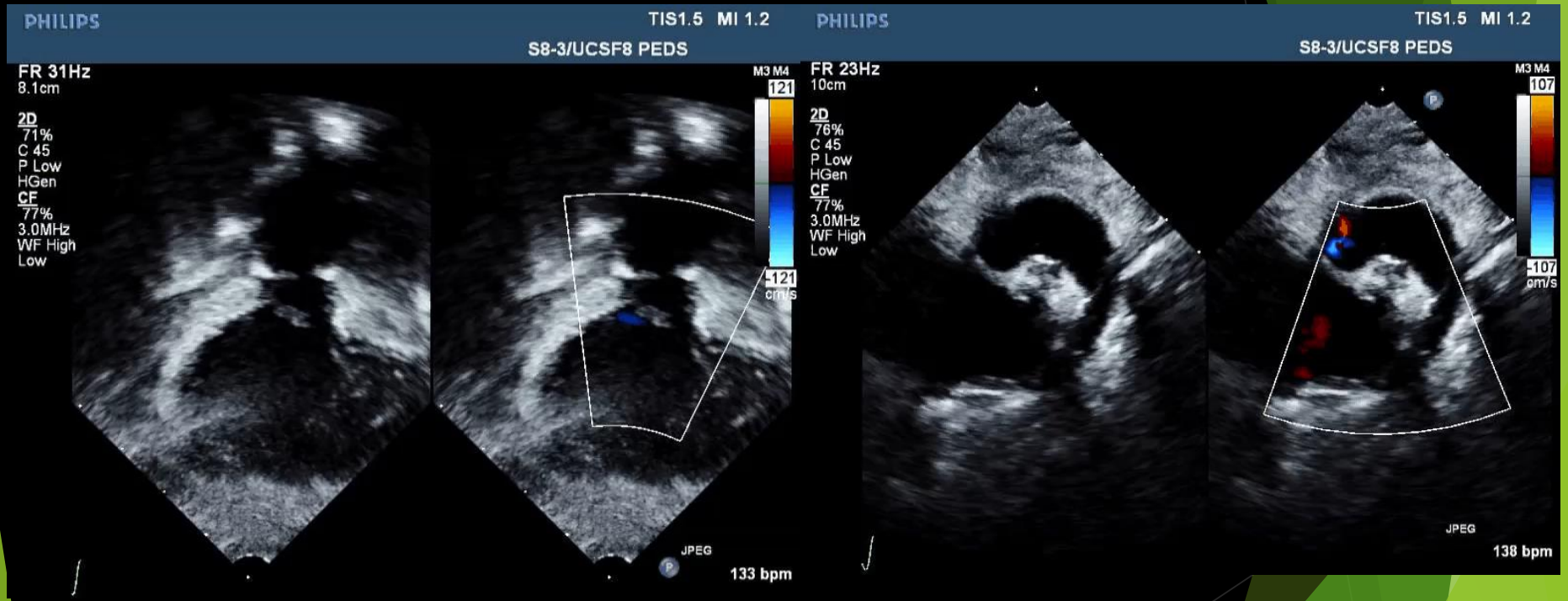
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THANK YOU