

Introduction to the Fontan circulation

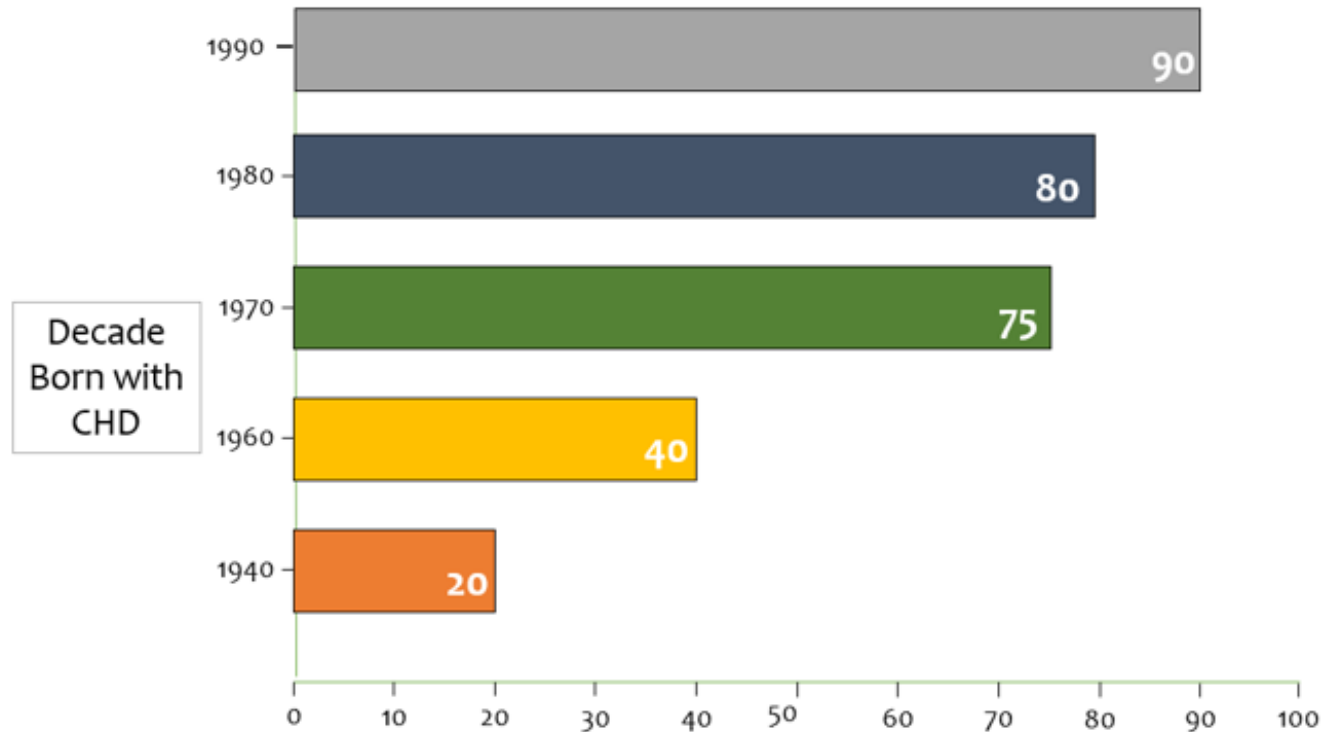
September 2020

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Newcastle upon Tyne

Improved survival of patients with congenital heart disease

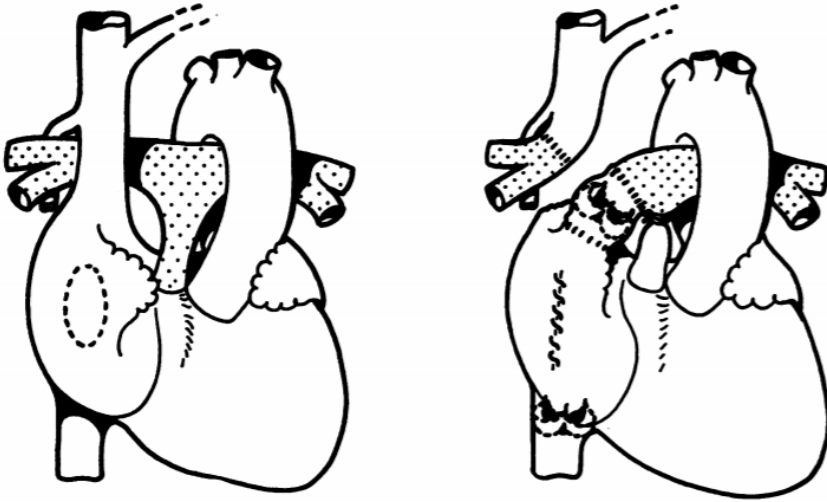


The Fontan-Kreutzer procedure

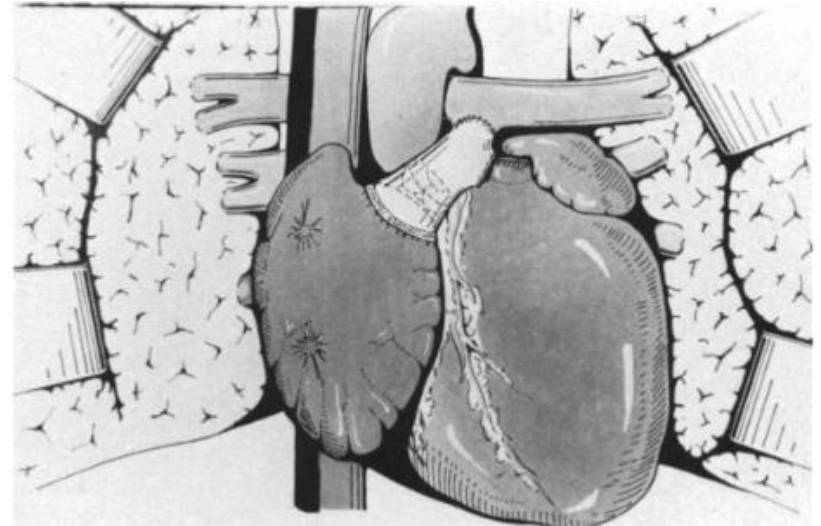


Francis Fontan (left) and Guillermo Kreutzer (right) in 2009

“Surgical repair of tricuspid atresia”



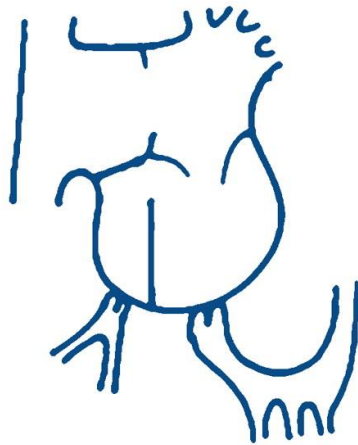
Fontan F, Baudet E, *Surgical repair of tricuspid atresia. Thorax 1971;26;240*



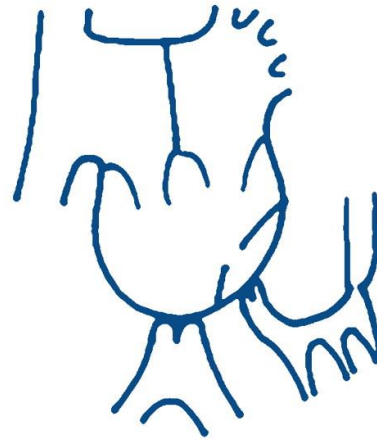
Kreutzer G, et al., *An operation for the correction of tricuspid atresia. JTCVS 1973;66:613-21*

Single ventricle physiology – multitude of diagnosis

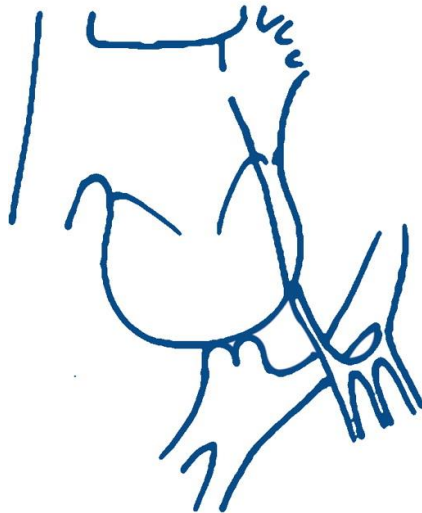
Tricuspid atresia



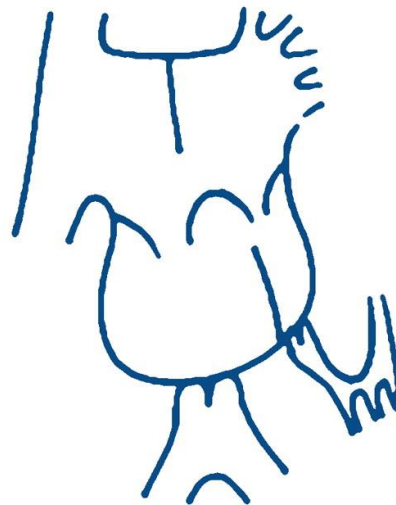
Double inlet left ventricle



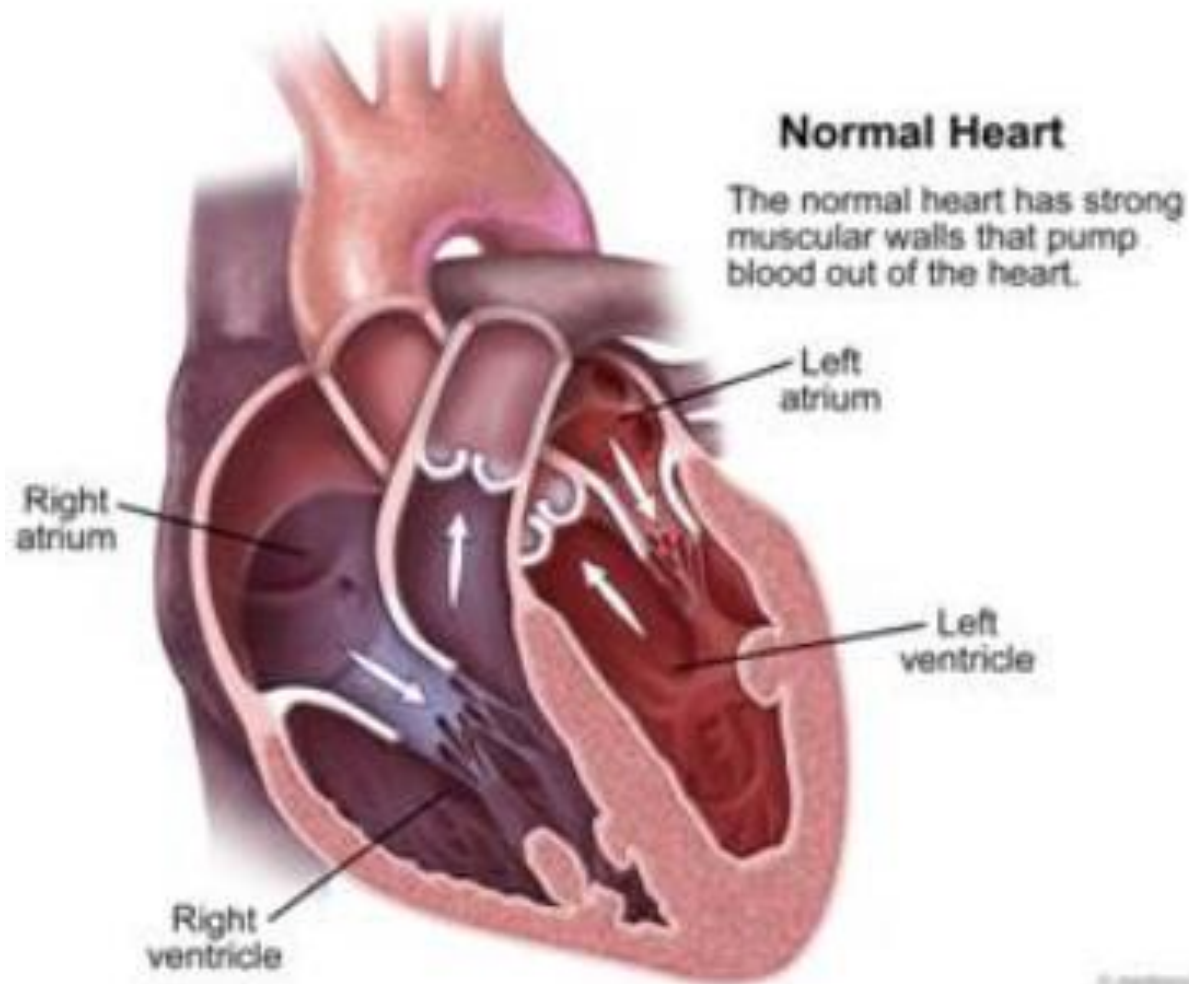
Hypoplastic left heart syndrome



Unbalanced AVSD



Biventricular physiology



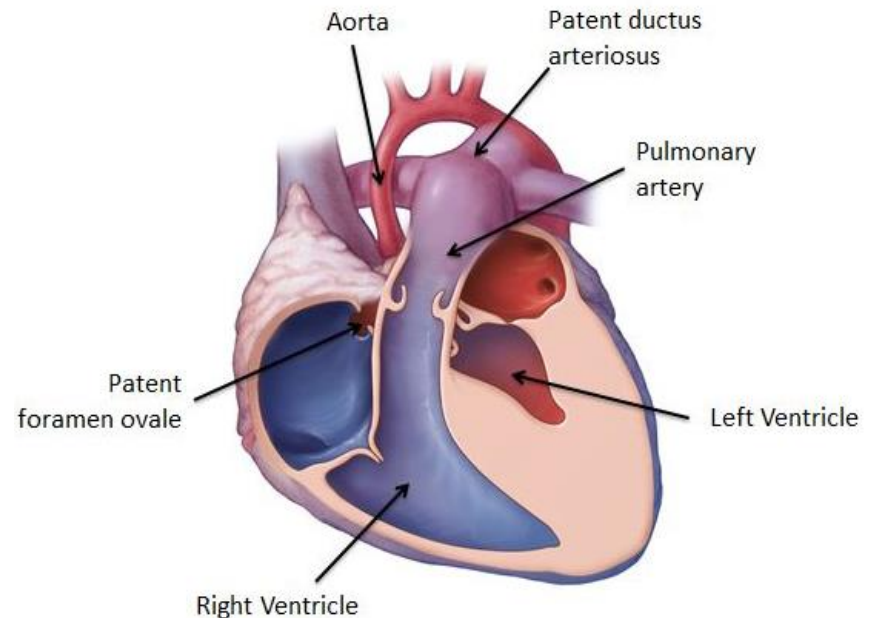
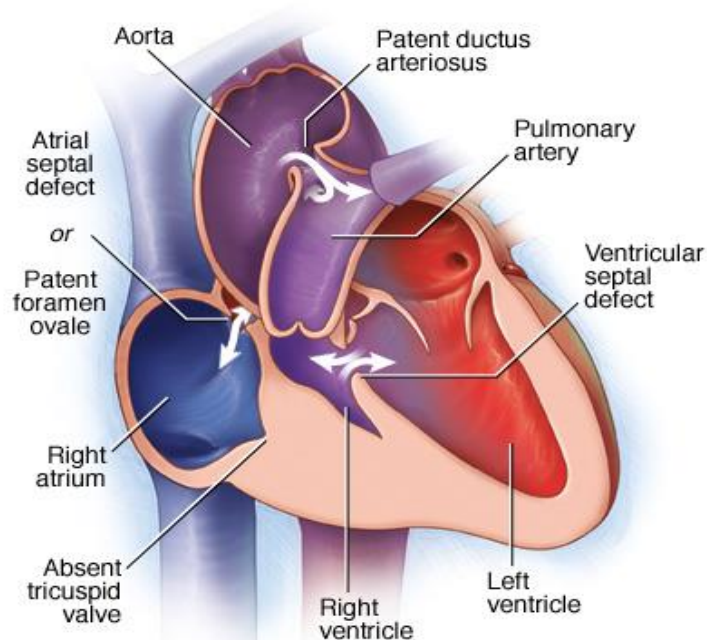
Single ventricle physiology

- One functional ventricle
- Supplying both systemic and pulmonary blood circulation – not connected in series but parallel

Disadvantages:

1/ Arterial desaturation

2/ Chronic volume overload to single ventricle



Aim of Fontan procedure

- Separate pulmonary and circulatory blood flow
 - Stop volume overload to SV
 - Improve Oxygen saturations

Fontan - Staged approach

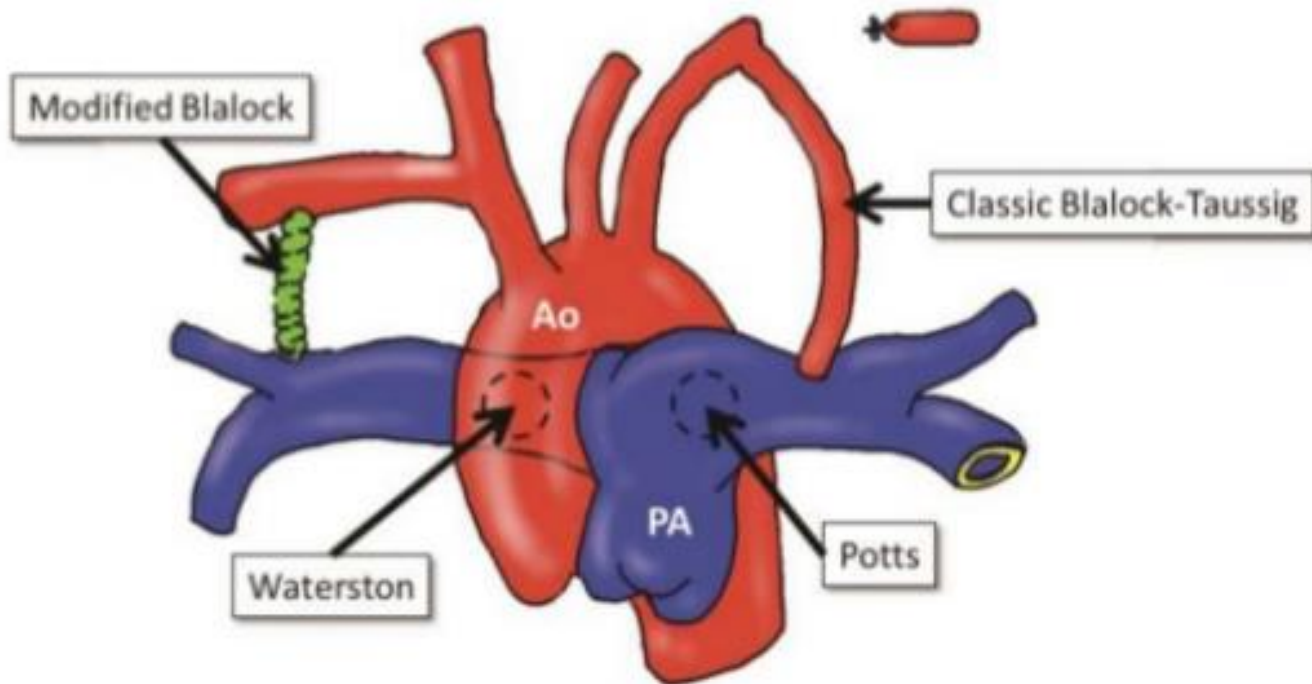
- A Fontan circulation is contra-indicated in the neonatal period:
 - relatively high (physiological) PVR
 - PAs and SVC/IVC too small
- A staged approach allows progressive adaptation of the heart and lungs and reduces the overall perioperative morbidity and mortality

Neonatal period

- Well balanced limited flow to lungs
 - If required:
 - Pulmonary artery band
 - Systemic to pulmonary shunt (Blalock Taussig shunt)
- Unrestricted flow from heart to aorta
 - If required:
 - Norwood (HLHS)
 - Damus Kaye Stansel
 - Coartectomy
- Unrestricted return from blood to ventricle
 - If required:
 - Rashkind balloon septostomy

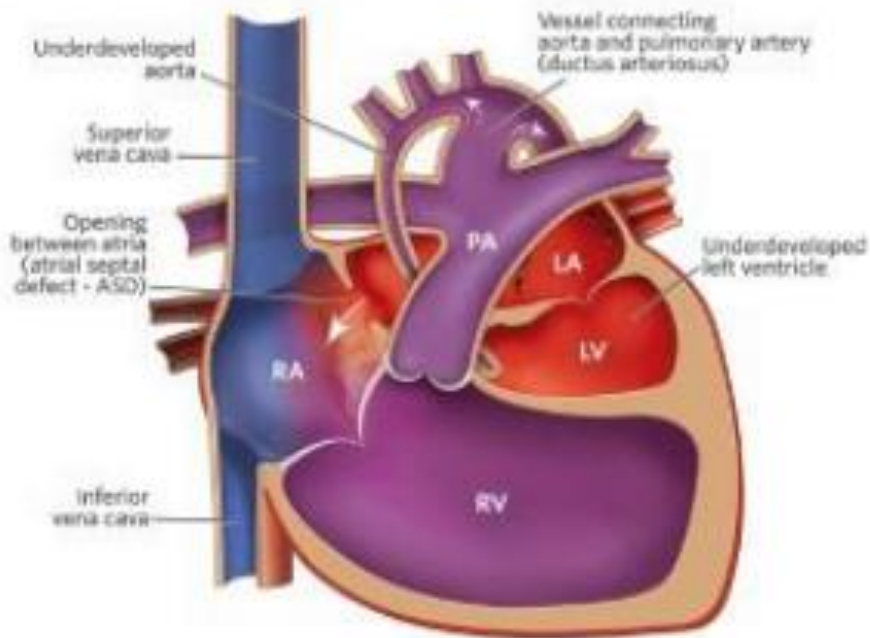
Systemic to pulmonary shunt

Blalock-Taussig shunt

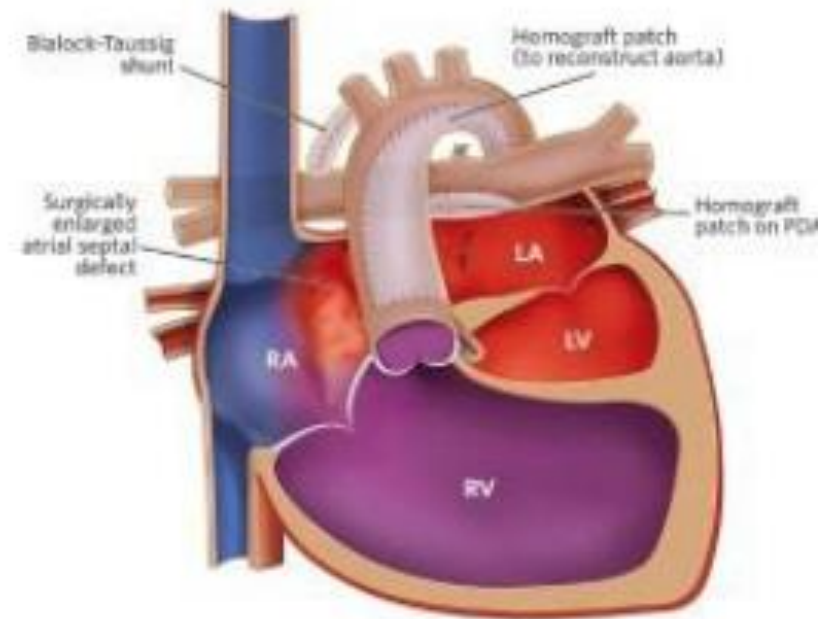


Norwood procedure (stage 1)

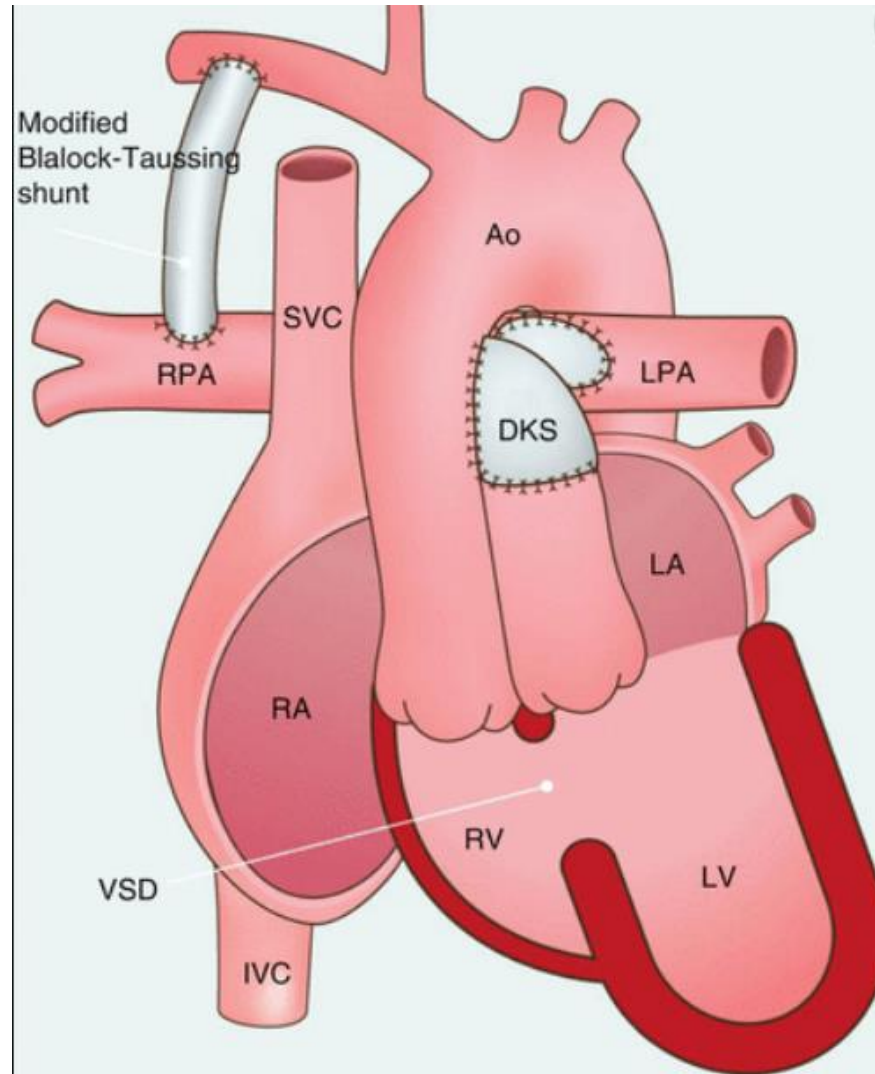
Hypoplastic Left Heart Syndrome (HLHS)



Hypoplastic Left Heart Syndrome (HLHS)
Stage 1 - Norwood

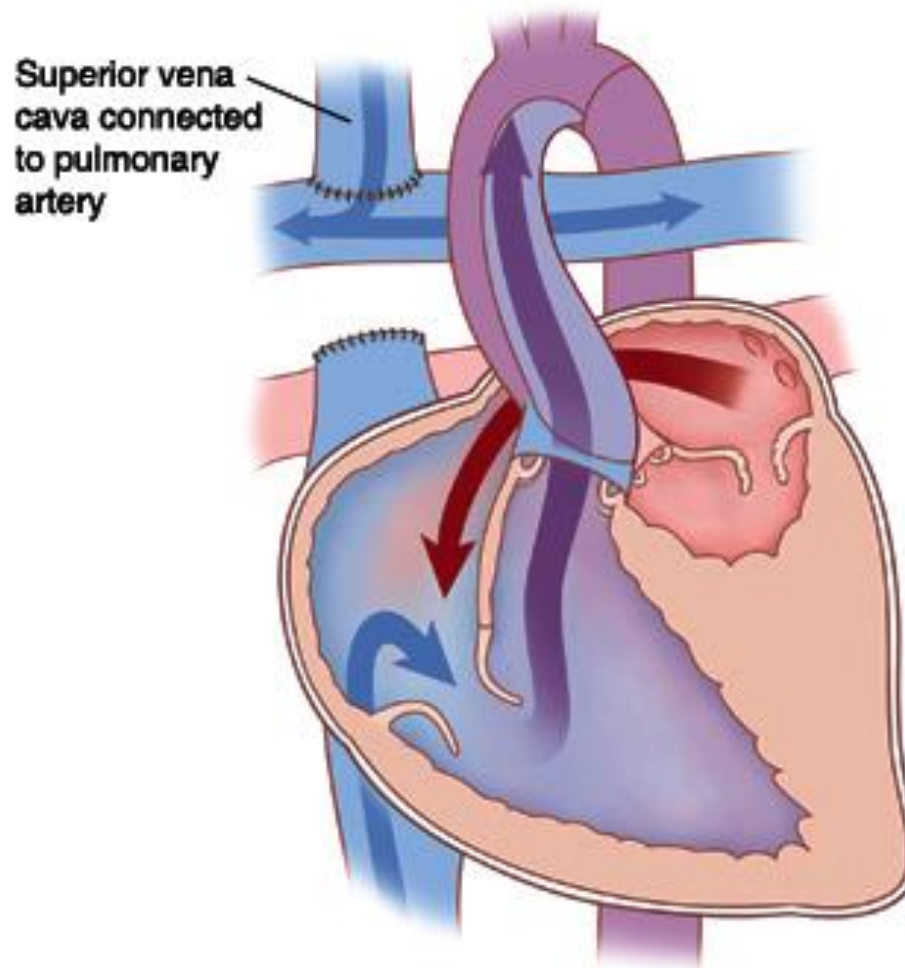


Damus Kaye Stansel

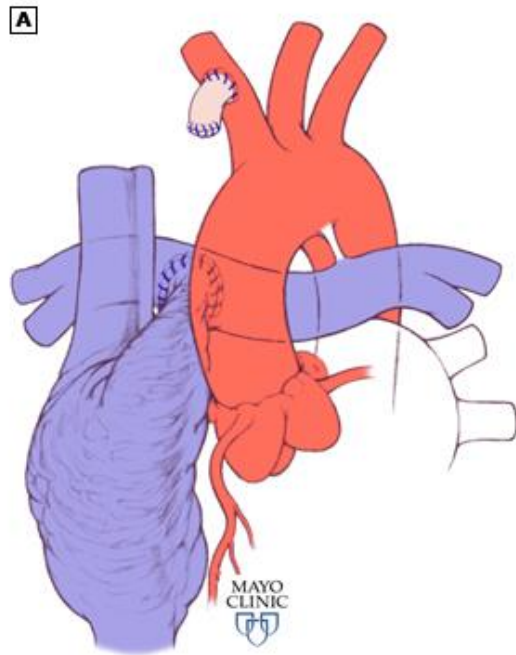


Age 4 to 12 months – bidirectional Glenn shunt (stage 2)

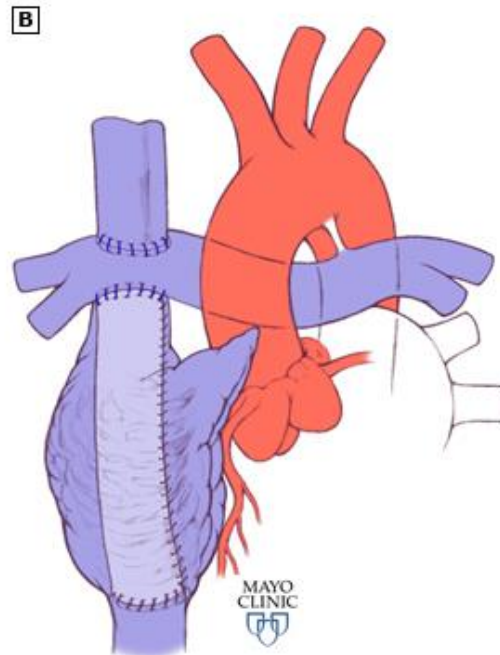
Bidirectional Glenn for HLHS



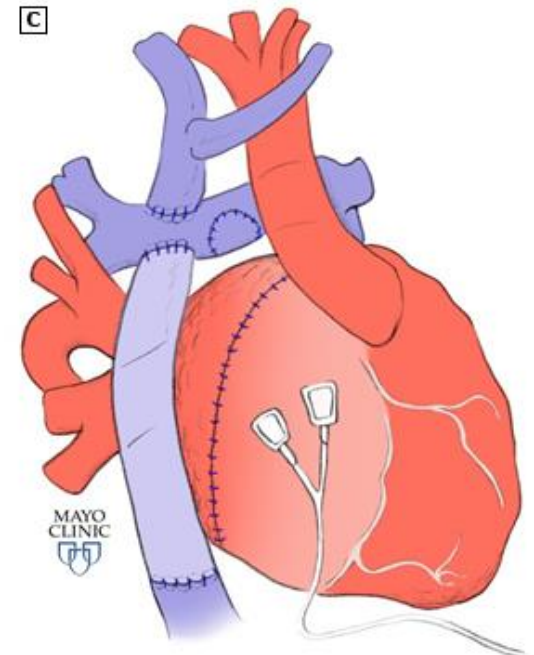
At age 1 to 5 years - Fontan completion (Stage 3)



“Classic”
atriopulmonary
connection



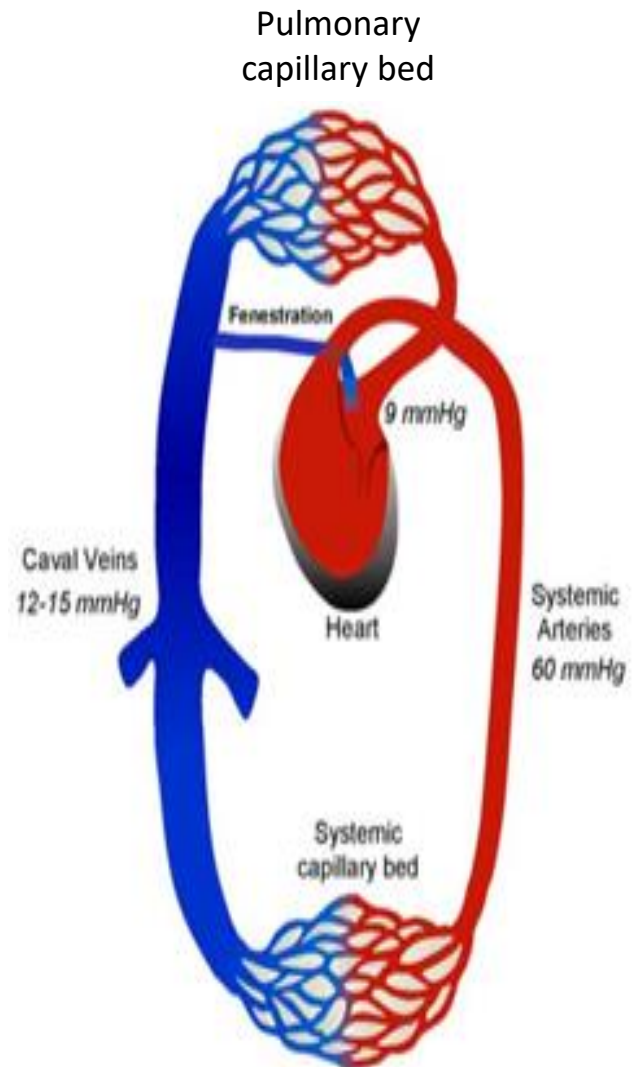
Intra-atrial total
cavo-pulmonary
connection (TCPC)



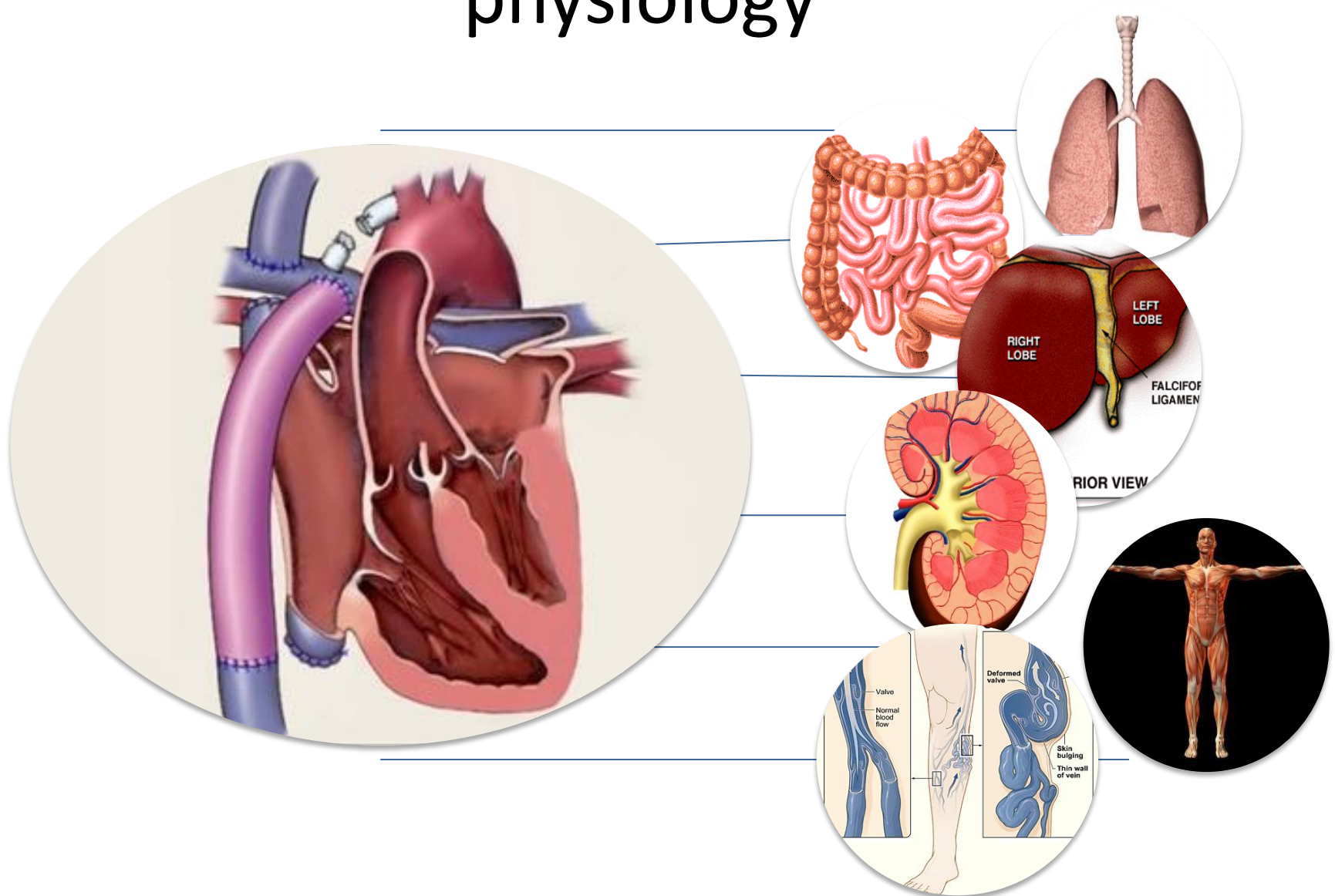
Extra-cardiac total
cavo-pulmonary
connection (TCPC)

Fontan physiology

- ***Systemic venous hypertension and congestion***
- ***Low cardiac output*** (ventricular preload deficiency)
 - CO not determined by heart but by transpulmonary flow
- Increased afterload
- Non-pulsatile pulmonary blood flow → endothelial dysfunction



Profound implications for organs physiology



Determinants of well functioning Fontan



Sinus rhythm



Unobstructed Fontan connections, pulmonary arteries, low pulmonary vascular resistance



Well functioning single ventricle



Well functioning AV valves and unobstructed outflow to aorta



**TIMELY DISCUSSION
WITH/REFERRAL TO ACHD-
TRANSPLANT TEAM**



Antiarrhythmic drugs, DCCV
EPS/ablation, pacing devices
TCPC conversion – careful patient selection



Avoid drugs/factors that increase PVR
Consider pulmonary vasodilators
Relief any obstruction, ? Fontan fenestration



Avoid drugs with negative inotropic effects
Consider classical heart failure drugs, ? Sildenafil
Consider coil embolisation AP collaterals



Surgical management – careful patient selection!
? Medical management
? Treatment of AP collaterals

Thank you