

Fontan

Procedure

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Fontan Nurse Specialist



- 1- 10,000 births born with single ventricle physiology requiring the staged procedure
- Survival to adulthood is now expected
- Multisystem disorder
- Poses many challenges for patients/families and clinicians





Francis Fontan

02/07/1929 - 14/01/2018

Before the introduction of the Fontan procedure, most children with single ventricles would die within the first year of their life

The classic Fontan procedure was first performed in 1968

Multistage Fontan procedure introduced in the 1980s with an intermediate stage - Glenn or hemi-Fontan anastomosis

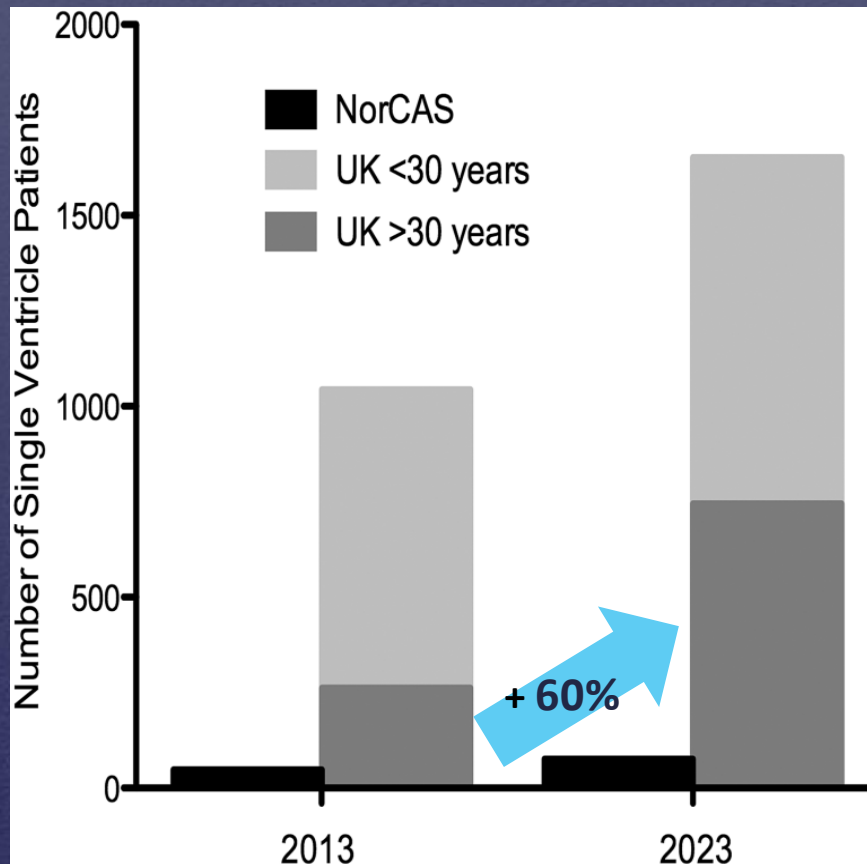


Staged Palliative Repair

- Norwood/shunt/stent
- Associated mortality
- Glenn approx 6 months – Total Caval Pulmonary Connection (TCPC) approx 4 years
- Full repair initially, causes too much pulmonary blood flow
- All patients individual, some do not tolerate



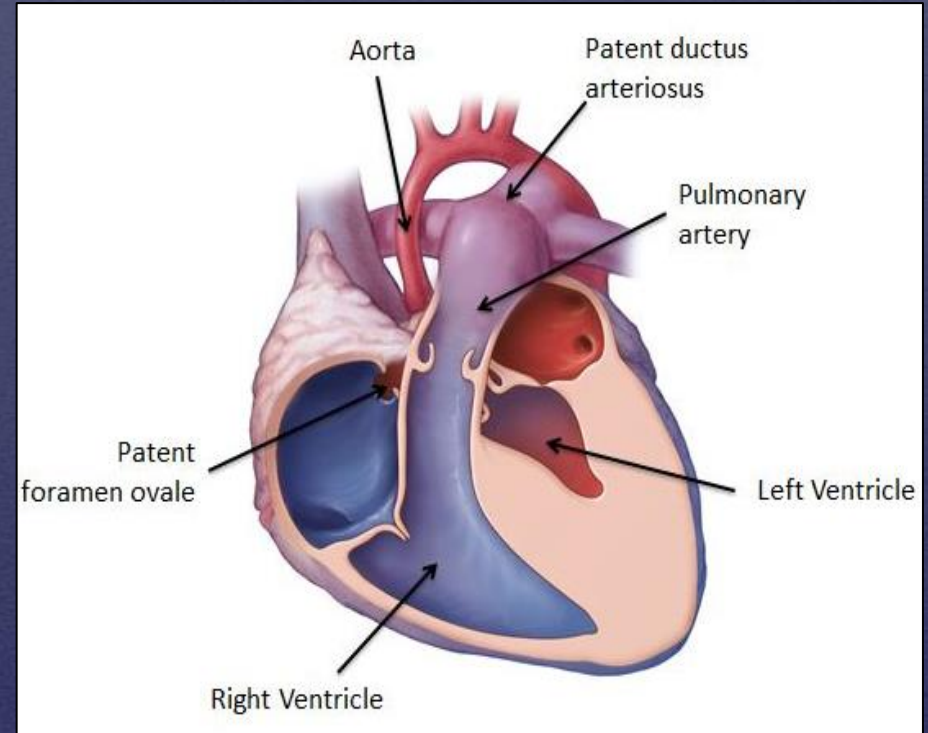
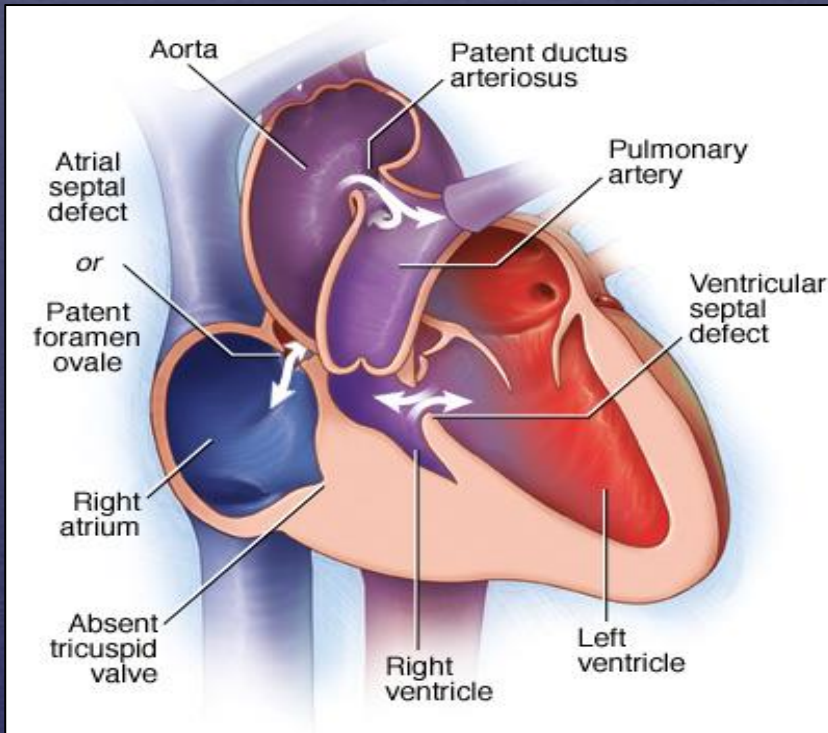
Population Growth



Coats L, et al. *Heart* (2014)

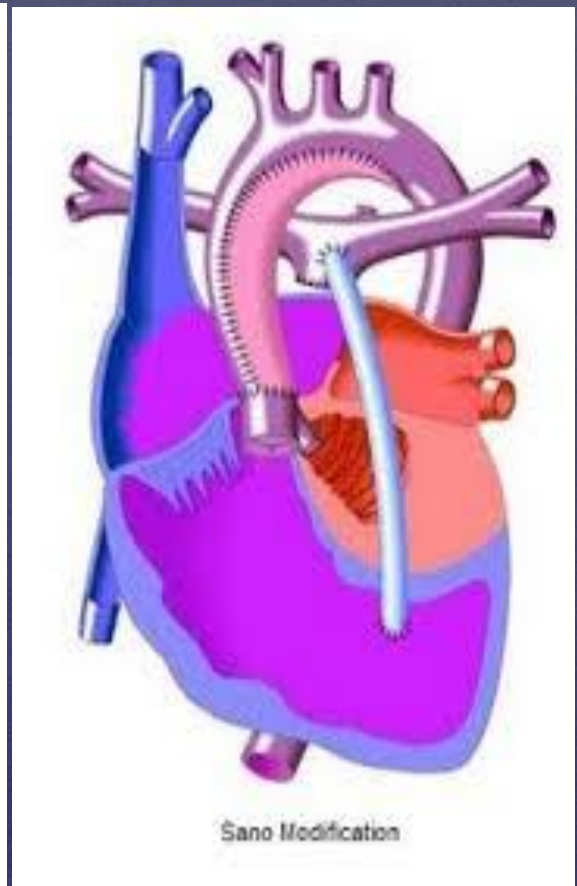
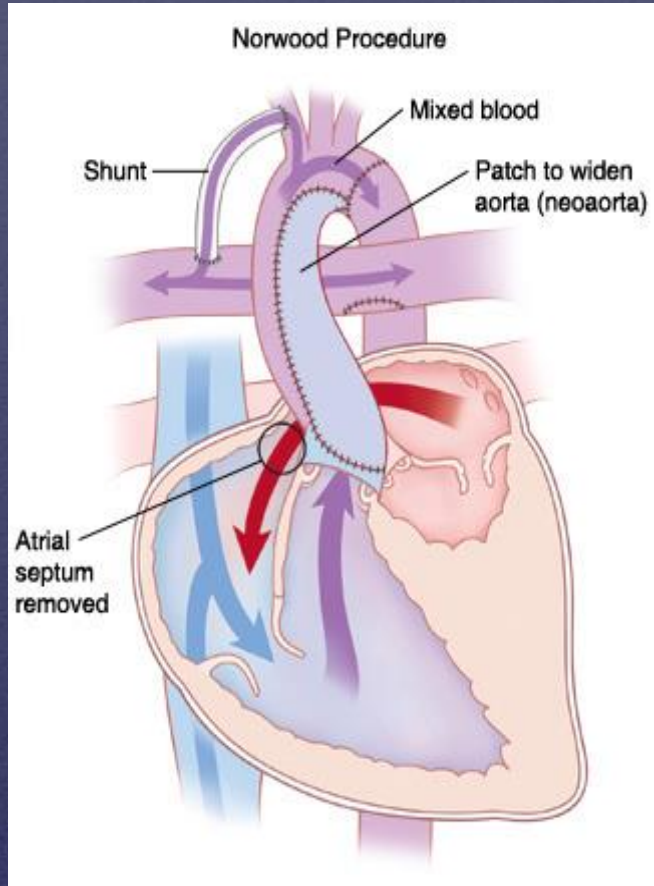
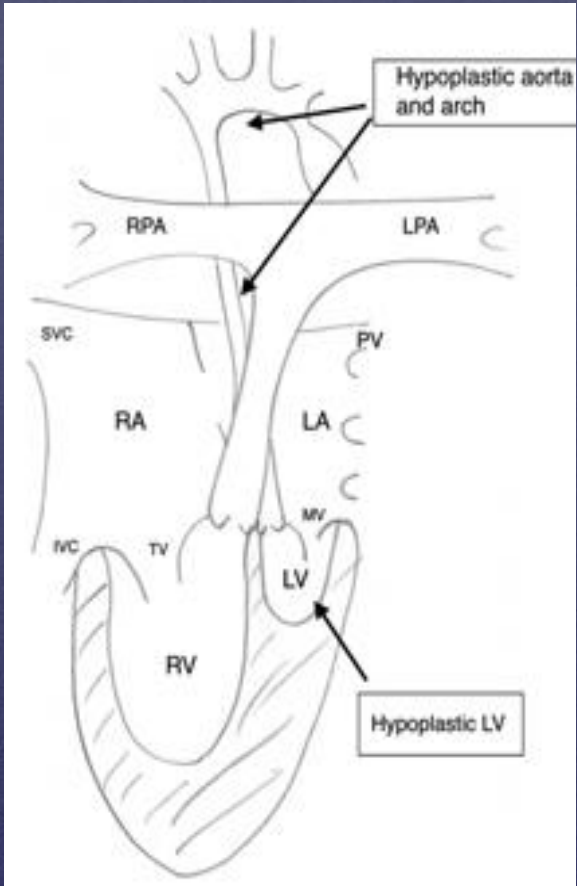


The Single Ventricle Patient A Multitude of Diagnoses!



Tricuspid Atresia, Double Inlet Left Ventricle, Unbalanced CAVSD, Hypoplastic Left Heart...with many more complexities



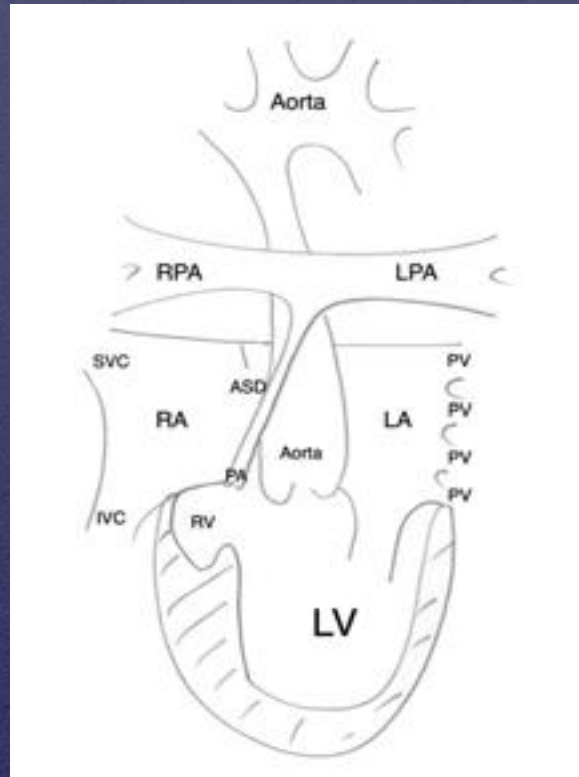


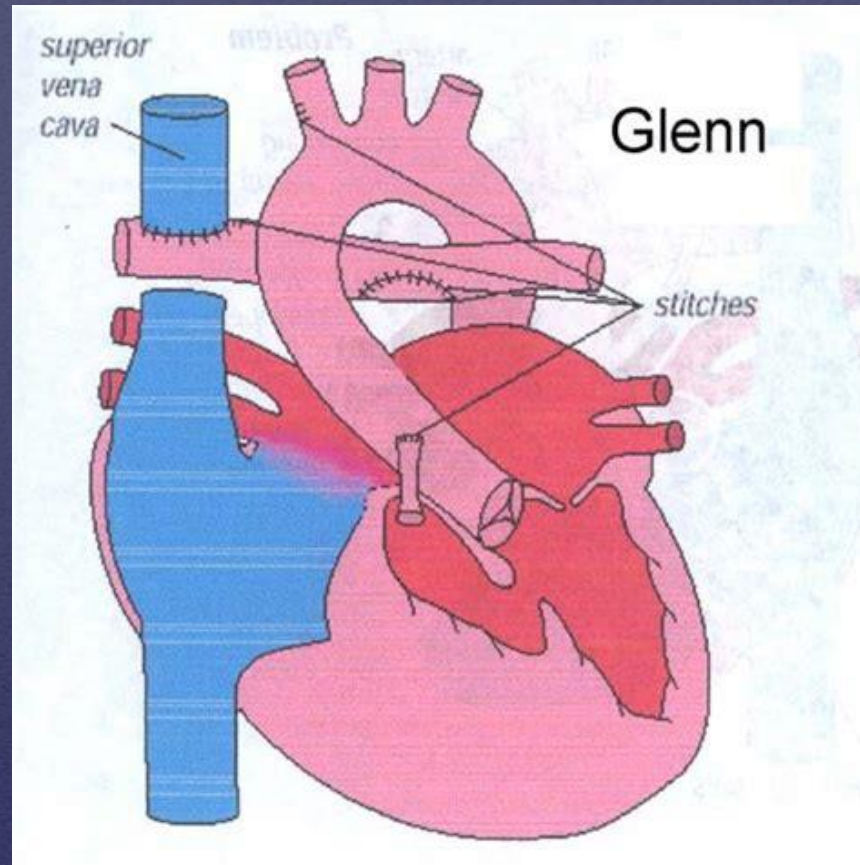
Hypoplastic LV



Hypoplastic RV

- Depending on how well balanced the circulation is, Norwood, banding or shunt may be needed prior to the Glenn

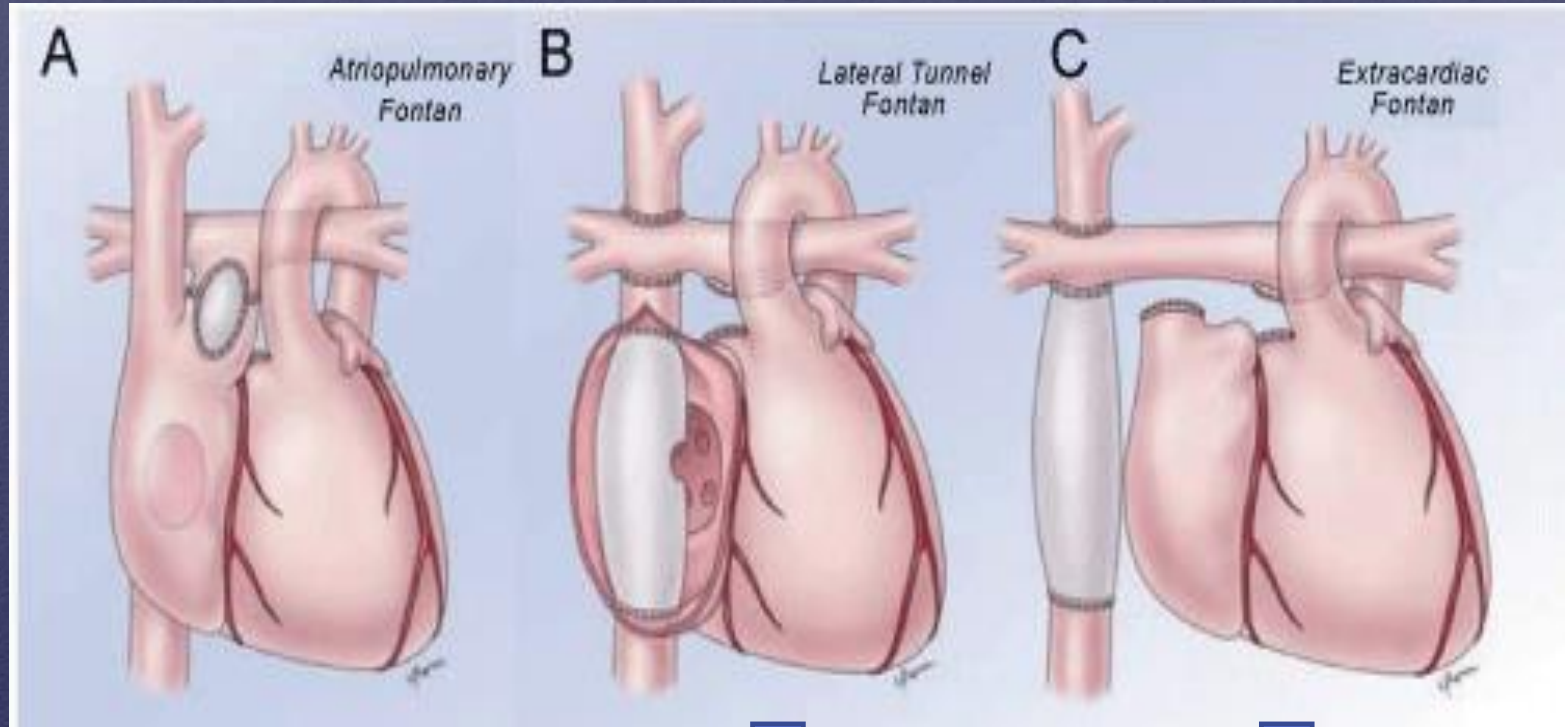




Glenn Shunt



Staged Series of Operations Performed as a Palliative Procedure for Uni-Ventricular Circulation



Classic Fontan first performed in 1968

Introduction of “multistage” Fontan procedures in the 1980s

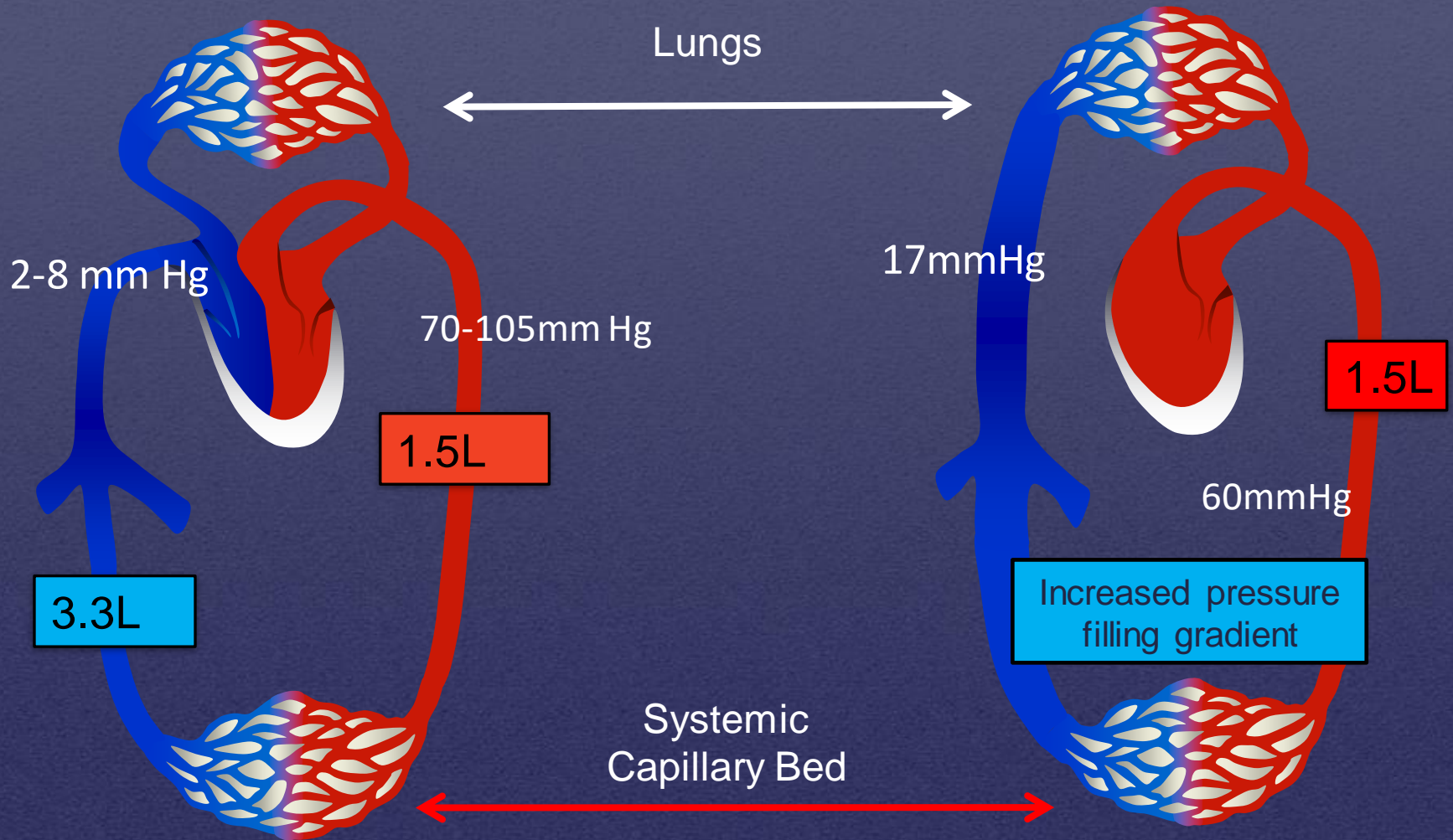


- The Fontan is Palliative:
Staged series of operations when a biventricular repair can not be done

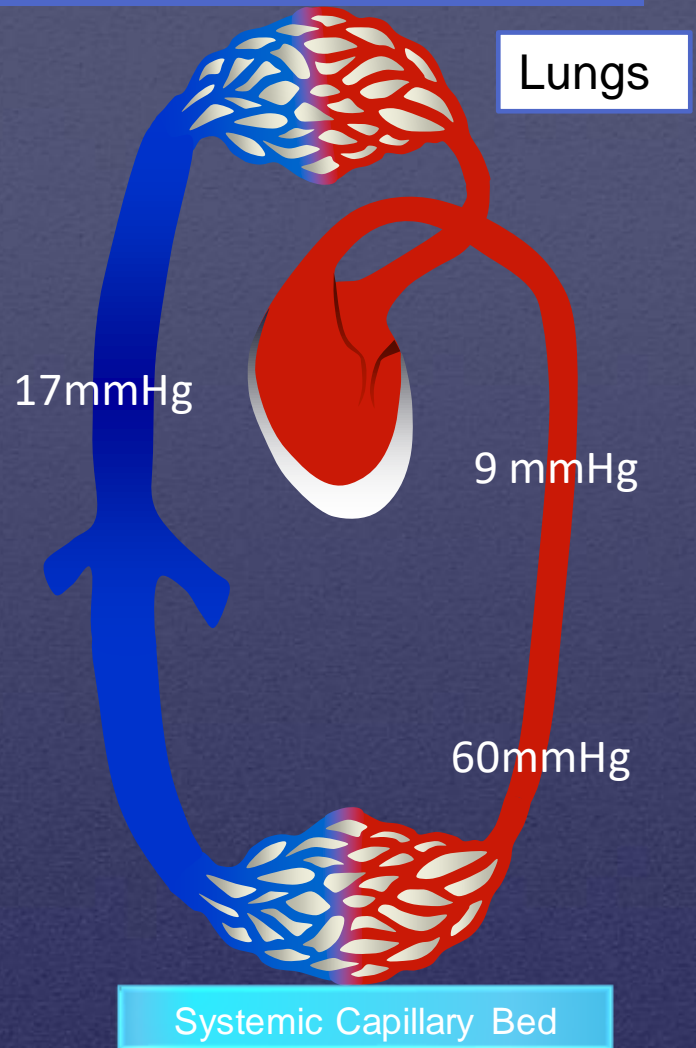
In Other Words



Normal vs Fontan Circulation



- Venous hypertension (Normal CVP 5-10)
- Increased resistance to venous return
- Lack of pulsatile flow - no pump on right
- Venous volume increased
Maintaining higher pressure gradient across lungs
- Systemic blood flow progressively decreases in older Fontans
- Low cardiac output state

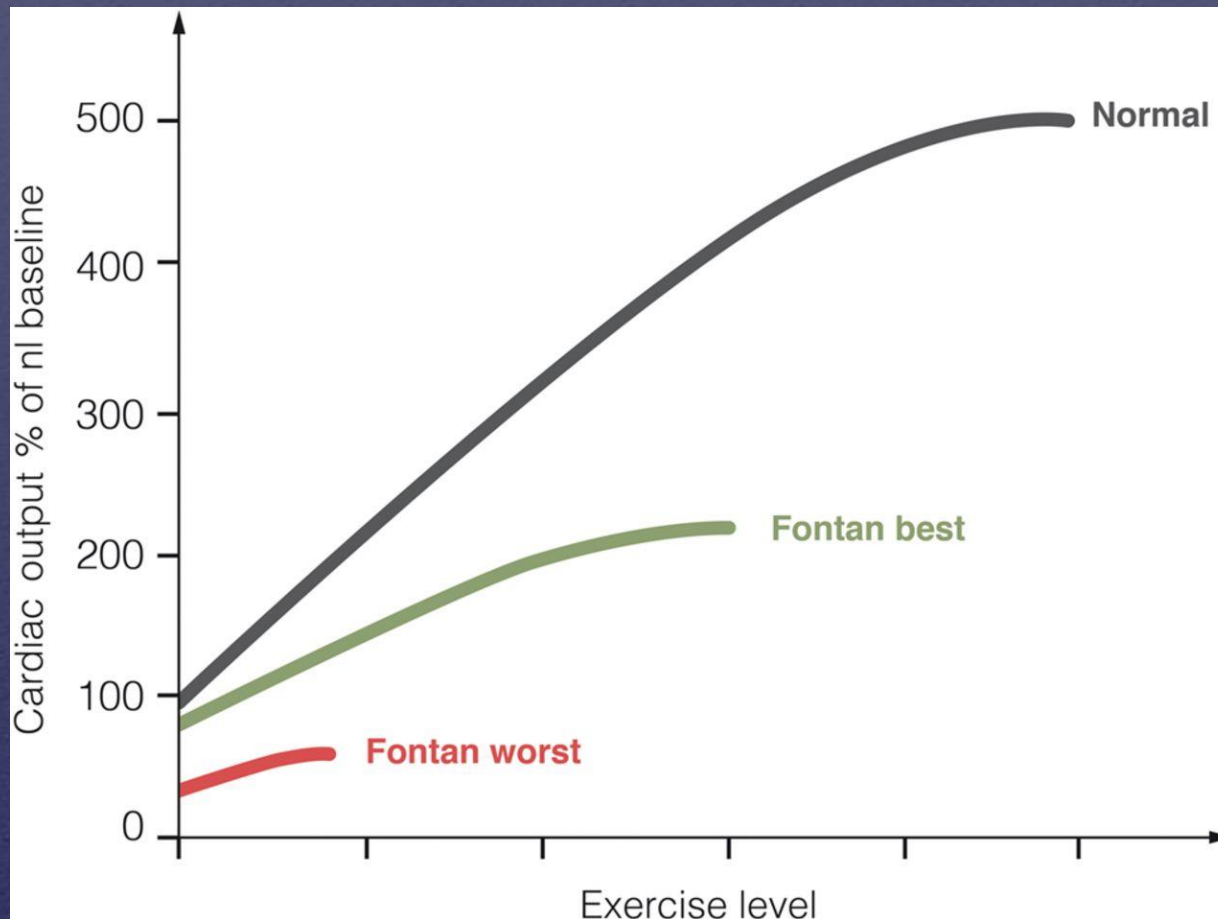


In Summary

Mace et al, Am J Physiol Heart Circ Physiol , 2000

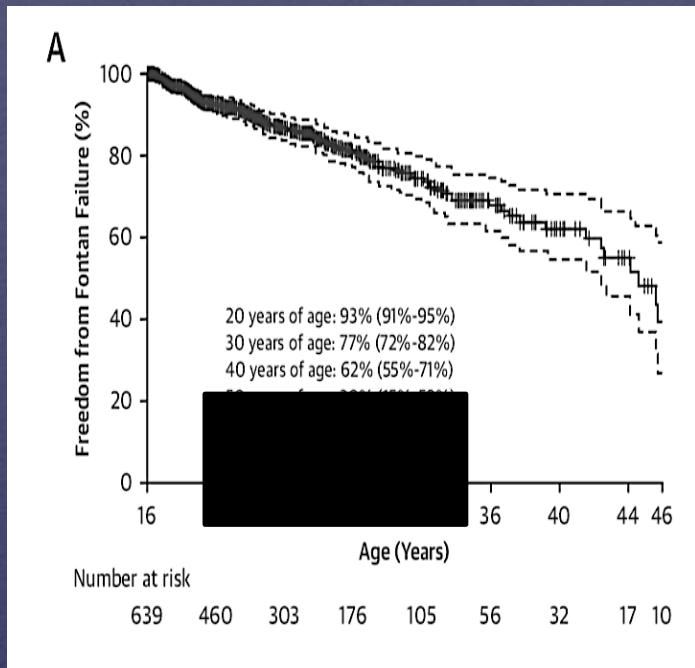


Exercise and Output: Normal V Fontan Circulation

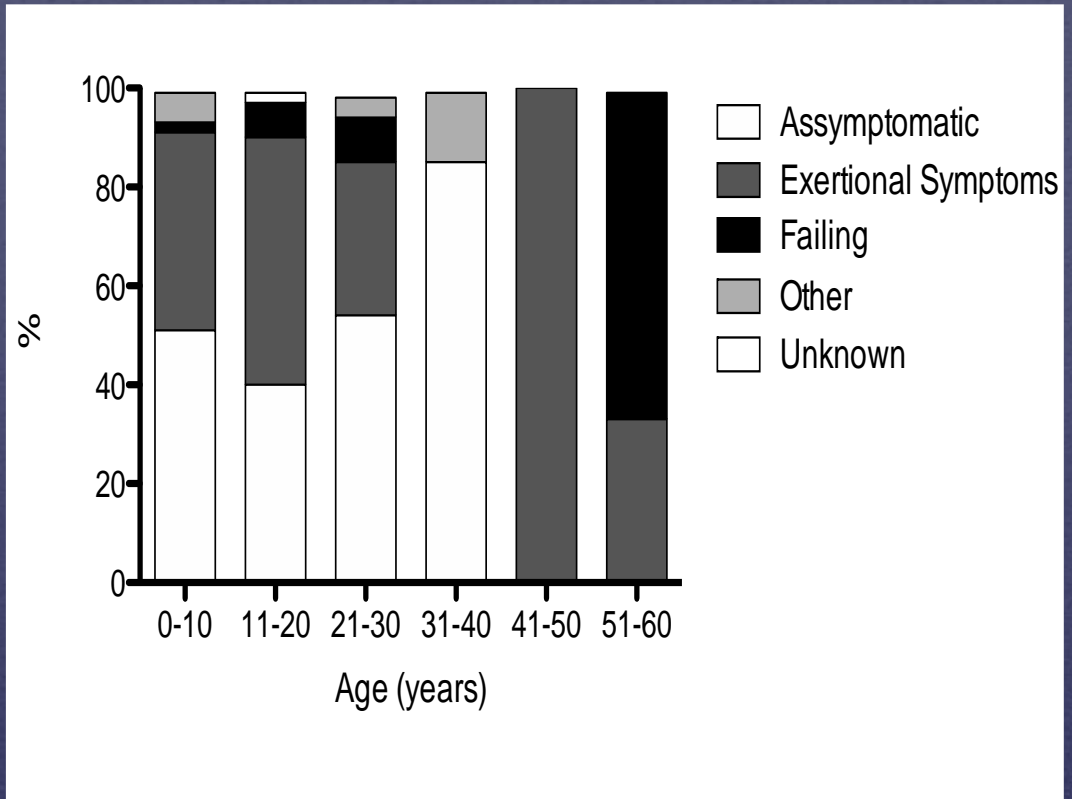


Marc Gewillig, and Stephen C Brown Heart
2016;102:1081-1086





Fontan failure



Dennis, Mark, et al. *Journal of the American College of Cardiology* (2018)



Symptoms and Age

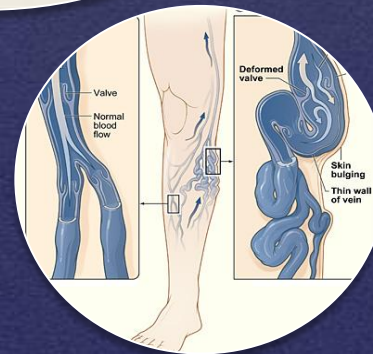
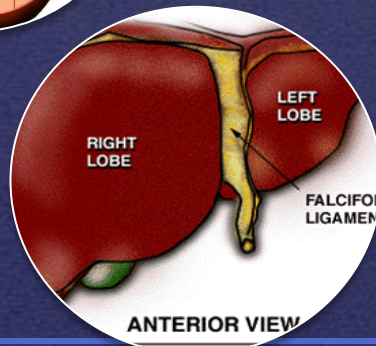
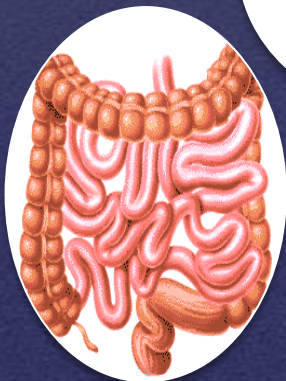
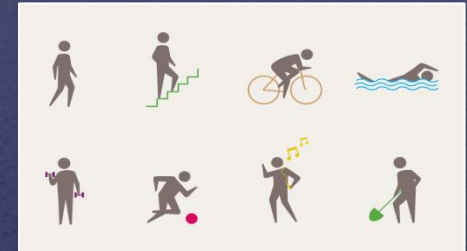
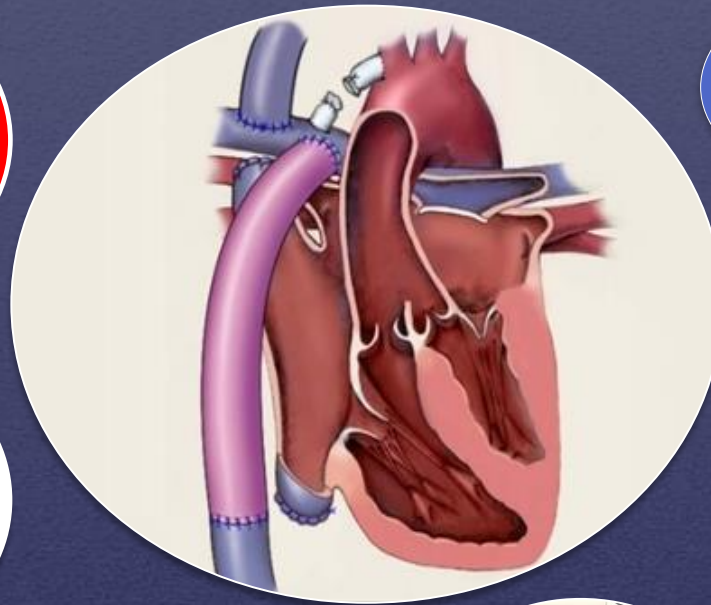


The Failing Fontan

A Multi-System Disorder

Ventricular failure
Arrhythmias
Valve incompetence
Blood Clots
Obstruction
Collaterals

PSYCHO- SOCIAL



When Will You See These Patients Decline in functional capacity

- Conventional surgery
- Electrophysiology
- Intervention - stents, coiling collaterals, fenestration
- Medical management
- Transplant/assessment
- Palliation



- Support complex group of patients
- Intervention at appropriate time
- Maintain Fontan circulation, optimally, for as long as possible:
 - Transplant not appropriate for all – anatomy etc
 - Lack of Donor organs
- Research:
 - Is there anything else that can be done?
 - Is what we are doing making a difference?
 - Are there alternatives to transplant?
 - What do the patients and families want?

Our Aim



- Thank You

Any Questions?