Hemodynamics and systolic and diastolic function in single ventricle

Department of Pediatrics, College of Medicine,
Pusan National University, Busan, Korea

Hyoung Doo Lee, M.D.

Single ventricle or univentricular heart

- Hearts for which division into two separate ventricles is deemed surgically infeasible

Clinical presentations of single ventricle patients

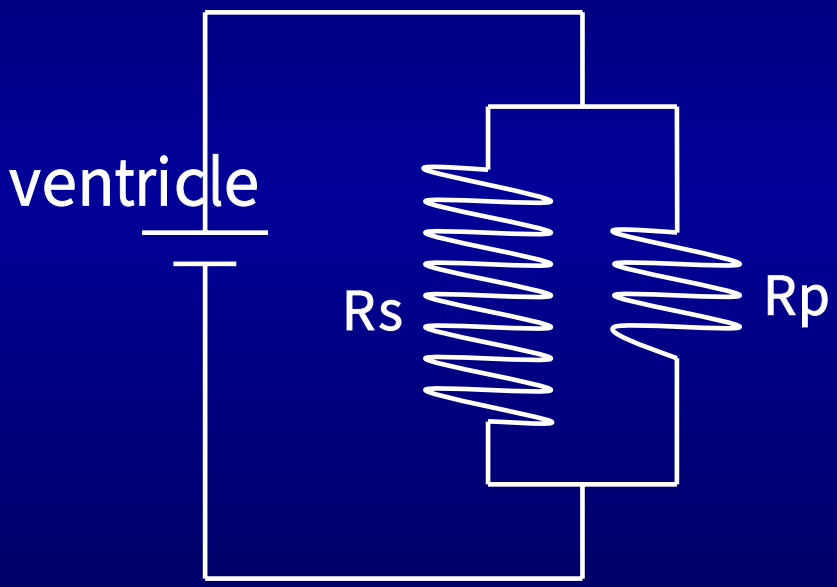
Clinical presentation	Anatomic example
1. Inadequate pulmonary circulation (or ductal-dependent)	–{S,D,S} Tricuspid atresia with pulmonary stenosis (IB) –Single right or left ventricle with pulmonary atresia
2. Excessive pulmonary circulation *without* systemic outflow obstruction	–{S,D,D} Tricuspid atresia, with transposition of the great arteries and unrestricted pulmonary blood flow (IIC)
3. Excessive pulmonary circulation *with* systemic outflow obstruction	–Hypoplastic left heart syndrome (MS/AS; MA/AS; MS/AA; MA/AA) –{S,D,D} tricuspid atresia with restrictive VSD –{S,L,L}Double outlet left ventricle with restrictive VSD
4. Balanced pulmonary blood flow	–{S,D,S} Tricuspid atresia with pulmonary stenosis (IB) –{S,D,D}DORV uncommitted VSD with pulmonary stenosis

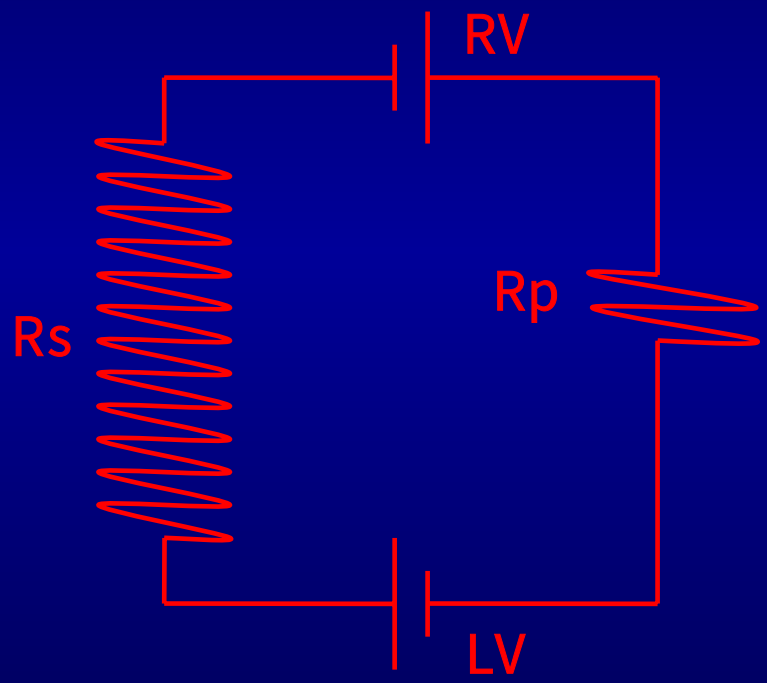
Goals of early managements

- Relief of outflow obstruction
- Optimized Qp/Qs
- Unimpeded pulmonary venous return
- Provision of adequate intracardiac mixing

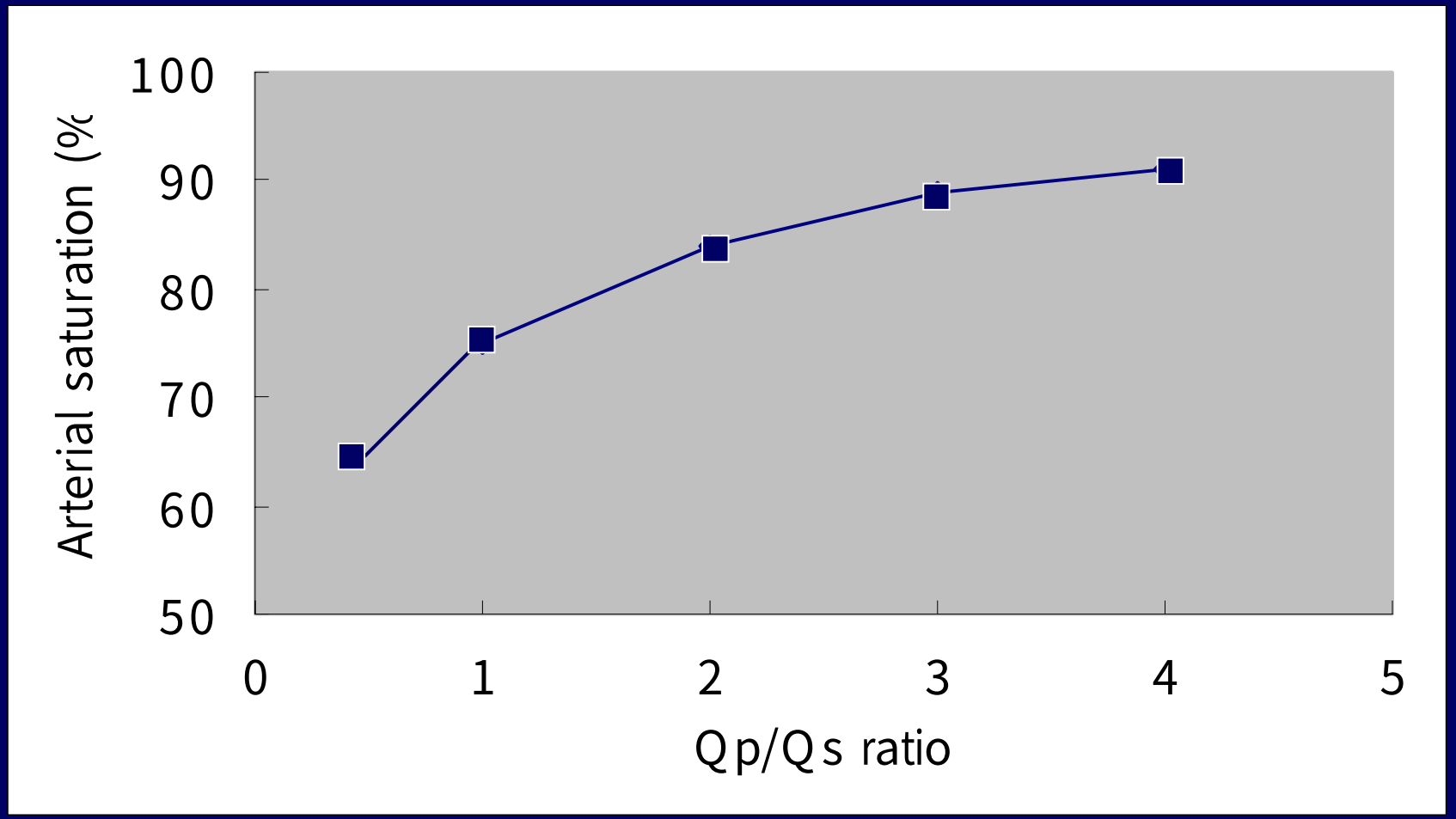
- Arch reconstruction, aortopulmonary anastomosis, correction of APVR, atrial septectomy, aortopulmonary shunt, PA banding

Circuits in parallel and in series





Arterial saturation and Qp/Qs in single ventricle



- Aortic saturations as plotted against Qp/Qs

Cardiol Young 2003;13:316-22

Arterial saturation and Qp/Qs in single ventricle

- SaO_2 depends on Qp/Qs
- To achieve SaO_2 90%, Qp/Qs at least > 3:1
- Qp ↑ to very high level → little gain in SaO_2
- At Qp/Qs 1:1~2:1, Qp ↑ → significant SaO_2 ↑

Arterial saturation and Qp/Qs in single ventricle

- SaO_2 depends on Qp
- Qp related to Rs

To increase Rp	To decrease Rp
Acidosis	Alkalosis
Inc PEEP	Nitric oxide
Vasopressor agents	Low MAP
Noradrenaline, adrenaline	High FiO_2
High $PaCO_2$, hypoxia, added nitrogen	Phosphodiesterase inhibitors, nitrates

Potential natural history of HLHS

- Ideal anatomy and physiology – Qp/Qs around 1; mild RV volume overload; SaO_2 around 80%; normal Qs
- Falling Rp – Qp/Qs ↑; sign of failure; SaO_2 around 85%
- Low Rp – Qp ↑↑; obvious heart failure with poor peripheral perfusion, metabolic acidosis but high SaO_2
- Closing arterial duct – very low Qs; shock with renal failure; high SaO_2

Systolic functions in palliated univentricular heart

- Result of palliation – volume overload
- SaO_2 > 85% ← Qp/Qs > 1.5:1 ← total ventricular output 2-3 times normal ← end-diastolic volume 200-300% normal
- Chronic volume overload – progressive ventricular dilation, more spherical ventricular shape

Systolic function after SCPA

- Persistent cyanosis with mean SaO_2 75%
- For adequate systemic O_2 delivery, CO↑↑
- End-diastolic volume↓ but still 125-200% normal
- Volume-load reduction, but not normalization

Systolic function after SCPA

- Age-related phenomenon
- SCPA at ＜3 yrs → ventricular volume ↓
- SCPA at ＞10 yrs → no change

  JACC 1996;28:1301-7
- Morpholgic single LV vs RV
- Prior aortopulmonary shunt vs PA band

Systolic function after TCPA

- Nearly total separation of systemic and pulmonary circulation
- SaO_2 90-95% or less, ↓ during exercise
- Prior to SCPA or Fontan, Rs 20% below normal ← vasorelaxation due to arterial desaturation and parallel relationship of two circulation

Systolic function after TCPA

- Acute rise in Rs
- Moving the pulmonary circulation into series→ cross sectional area ↓, total vascular length ↑→ Rs ↑
- Peripheral vasoconstriction and neurohumoral activation
- Net impact, 75% increase in Rs

Systolic function after TCPA

- After Fontan, dramatic fall in preload (35-50%, previous SCPA 15%)
- Afterload ↑ and preload ↓ ⇒ systolic function ↓
- Under the optimal circumstances, subsequent ventricular remodeling and somatic growth lead to normalization of ventricular size

Systolic function after TCPA

- Ventricular remodeling with improved ventricular contractility in Fontan < 10 yrs of age, no such recovery in Fontan at a later age

  Circulation 1992;86:1753-61

- Other factors related to late outcome
- Prolonged elevation of coronary sinus pressure
- Valve regurgitation
- Durability of systemic right ventricle

Diastolic function in palliated univentricular heart

- In tricuspis atresia
- Significant ventricular dilation, reduced mass/volume ratio, normal chamber stiffness

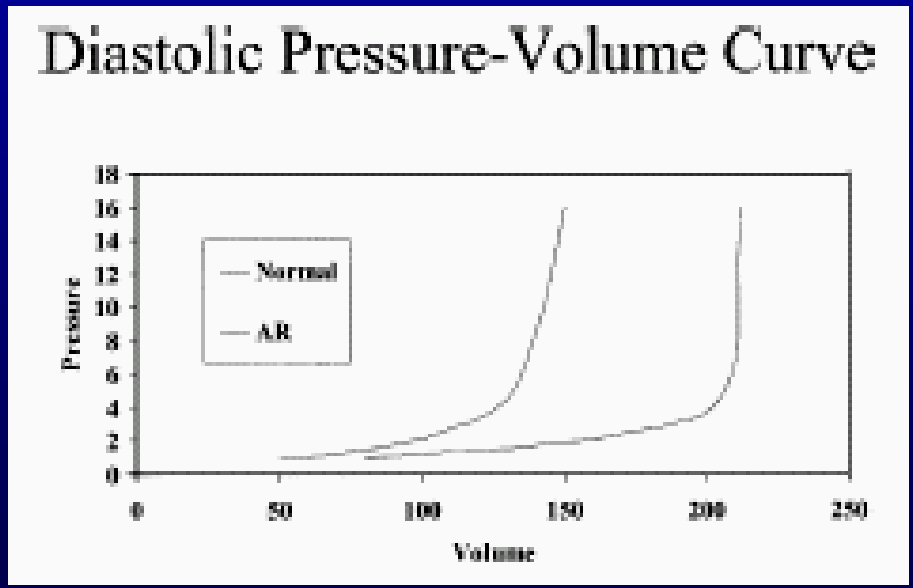
  Am J Cardiol 1994;73:292-7

Diastolic function after SCPA or Fontan

- Preload ↓ → end-diastolic volume ↓ → mass:volume ratio ↑
- Ventricular compliance ?
- Prolonged isovolumetric relaxation time
- Reduced early rapid filling

Ventricular compliance

- Chronic volume overload → rightward shift of diastolic pressure-volume relationship
- Increased chamber distensibility due to ventricular remodeling
- Chamber compliance is not impaired by acute fall in preload



Impact of ventricular hypertrophy

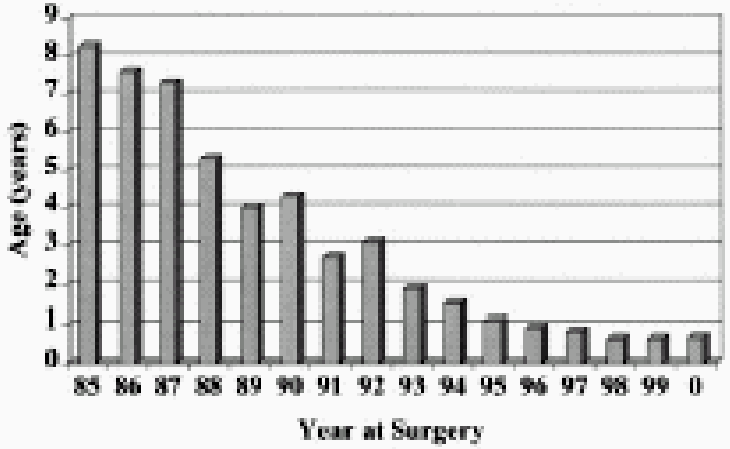
- *Circulation 1987;76(Suppl):III45-52*

  Reduced mass:volume ratio asso with reduced survival after TCPA
- *J Thorac Cardiovasc Surg 1986;92:1049-64*

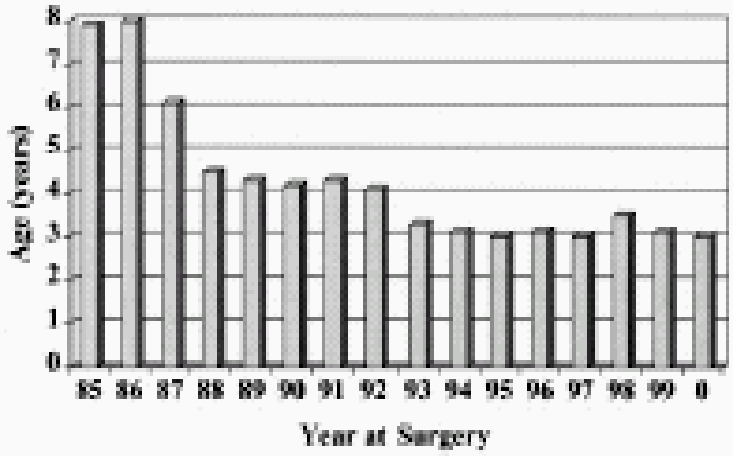
  Hypertrophy – longterm risk factor for death after Fontan
- *Herz 1992;17:220-7*

  Lower volume and higher mass:volume ratio in group with poor outcome

Median Age at "Hemi-Fontan" (Boston Children's Hospital)



Median Age at Fontan Operation (Boston Children's Hospital)



Ventricular relaxation

- Prolongation of isovolumetric relaxation time and lower E:A ratio after Fontan indicating impaired relaxation
- Incoordinate relaxation, inter- and intra-ventricular conduction defect, elevated coronary sinus pressure

Summary

- Palliated single ventricle is at risk for chronic volume overload induced myocardial injury, the severity of which is related to the duration and magnitude of volume overload.
- SCPA and Fontan procedures reduce the volume load on the ventricle, and performed early serve to protect the myocardium from irreversible injury.