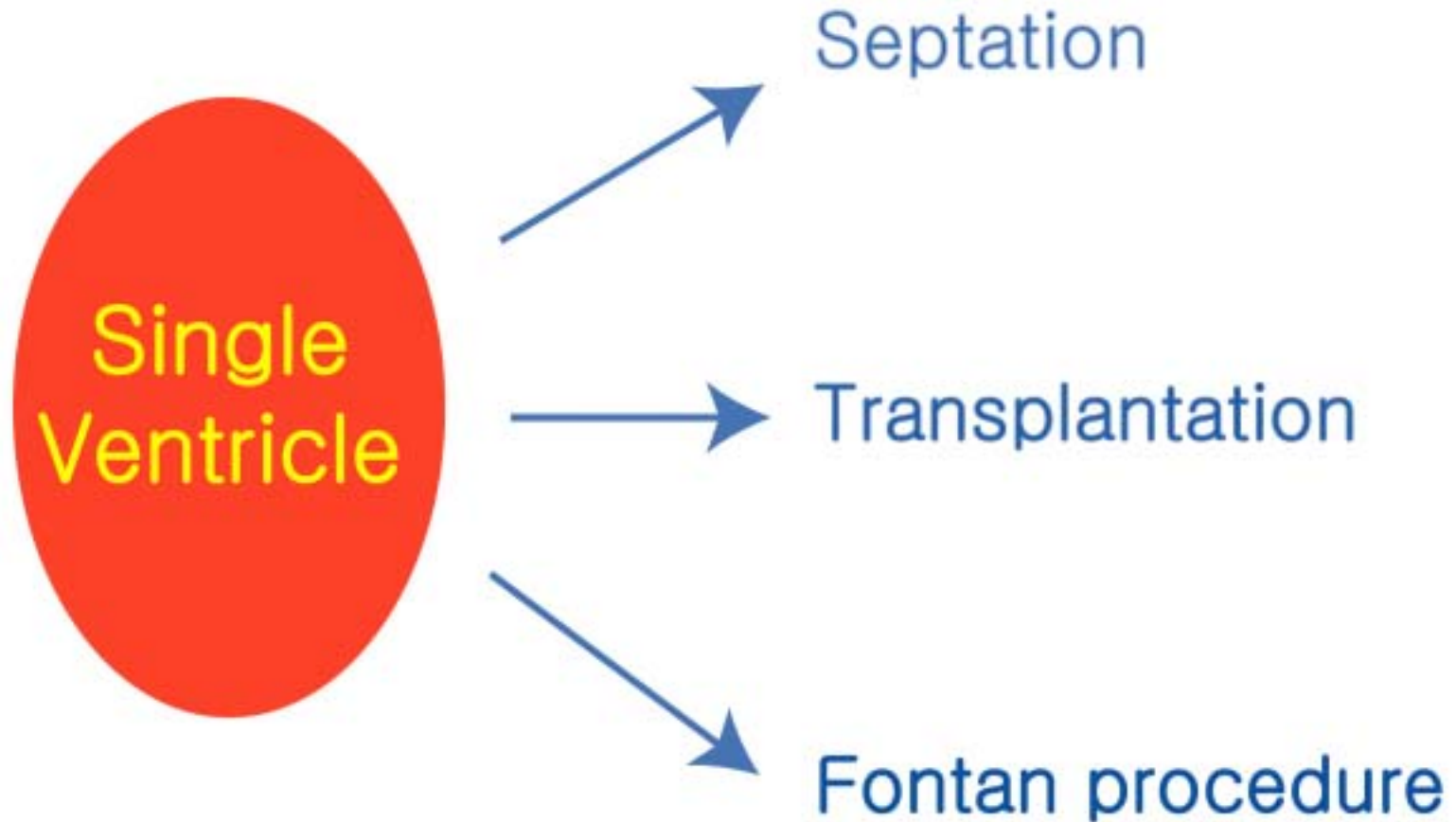


# Surgical Management of Patients with Single Ventricle

부천세종병원 흉부외과  
이 창 하

# Surgical Management



# History & Evolution

1950s 1958 1970 1971 1972 1973 1978



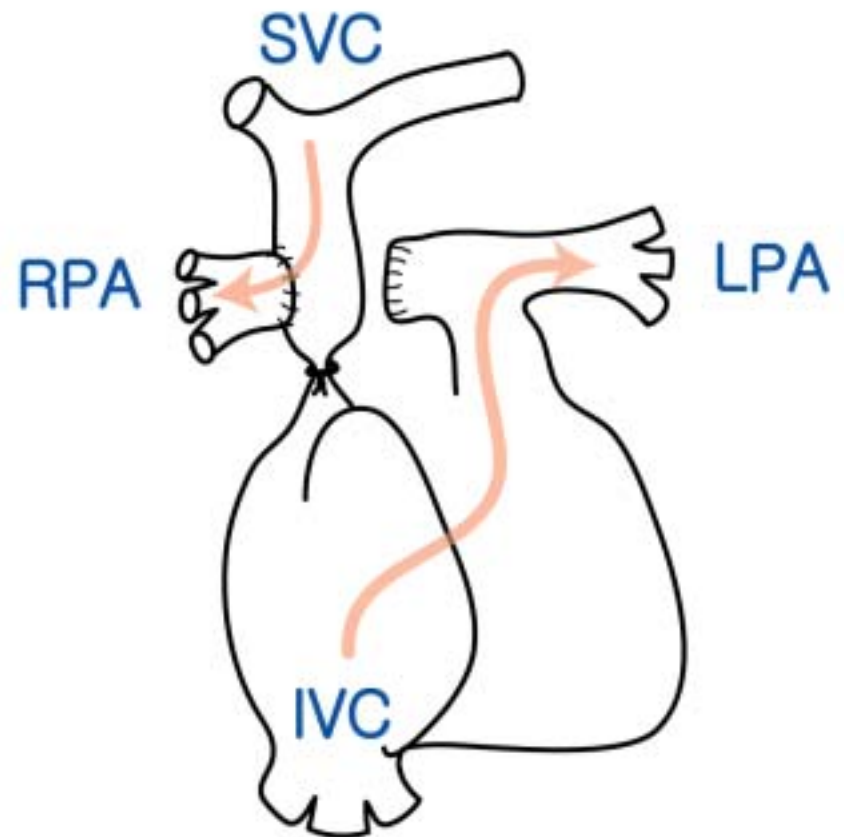
Experimental works into right heart bypass  
by a number of investigators

# History & Evolution

1950s 1958 1970 1971 1972 1973 1978

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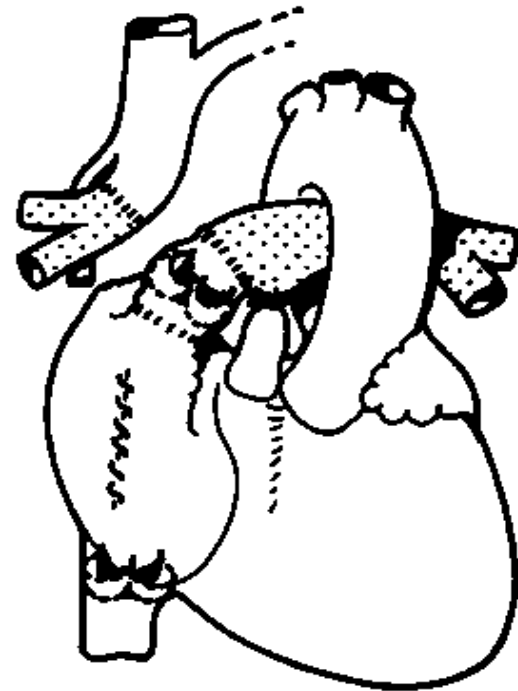
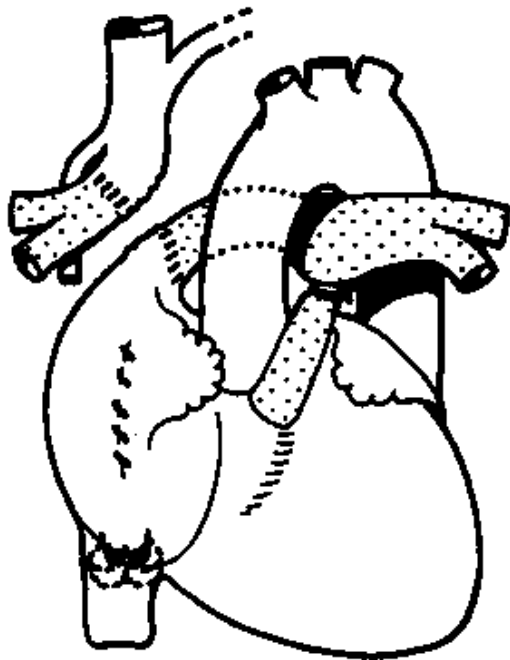
Glenn  
classic Glenn shunt



# History & Evolution

1950s 1958 1970 1971 1972 1973 1978

Fontan



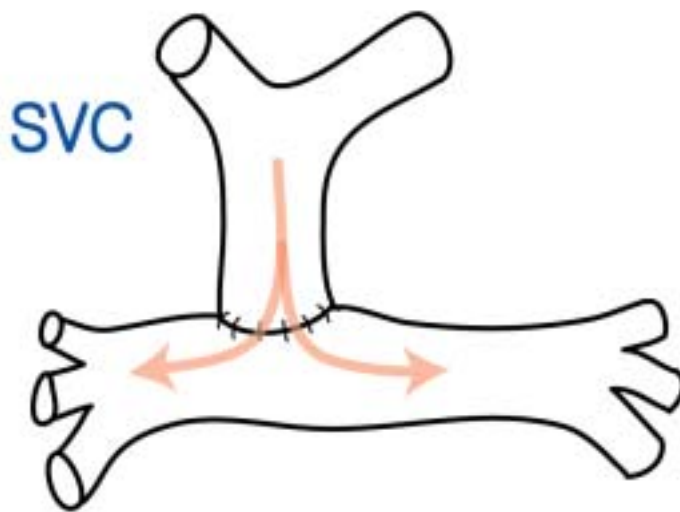
# History & Evolution

1950s 1958 1970 1971 1972 1973 1978

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## Azzolina

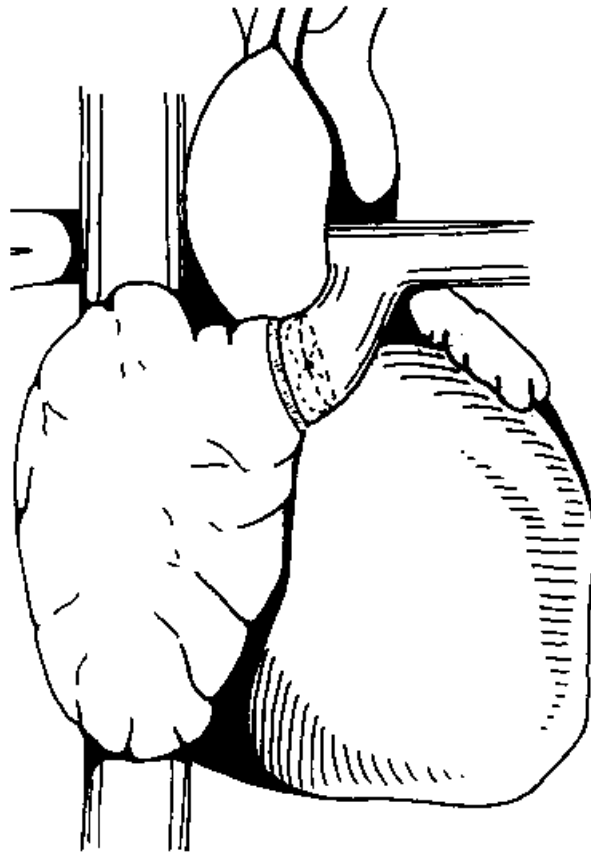
bidirectional Glenn shunt  
bidirectional cavopulmonary  
shunt / connection



# History & Evolution

1950s 1958 1970 1971 1972 1973 1978

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Kreutzer  
modified Fontan

# History & Evolution

1950s 1958 1970 1971 1972 1973 1978

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Choussat

Selection criteria for Fontan's procedure

## 'Ten Commandments'

1. minimum age 4 years
2. sinus rhythm
3. normal caval drainage
4. right atrium of normal value
5. mean PAP  $\leq 15$ mmHg
6. pulmonary alveolar resistance  $< 4$  units/m<sup>2</sup>
7. PA to aorta diameter ration  $\geq 0.75$
8. normal ventricular functions (EF  $\geq 0.6$ )
9. competent left AV valve
10. no impairing effects of previous shunt

# History & Evolution

1950s 1958 1970 1971 1972 1973 1978



## Atriopulmonary connection

RA dilation

Atrial arrhythmias

Atrial thromboembolism

Compression of PV by the giant atrium

# History & Evolution

1983

1984

1987

1988

Norwood

Physiologic repair of aortic atresia–hypoplastic left heart syndrome.

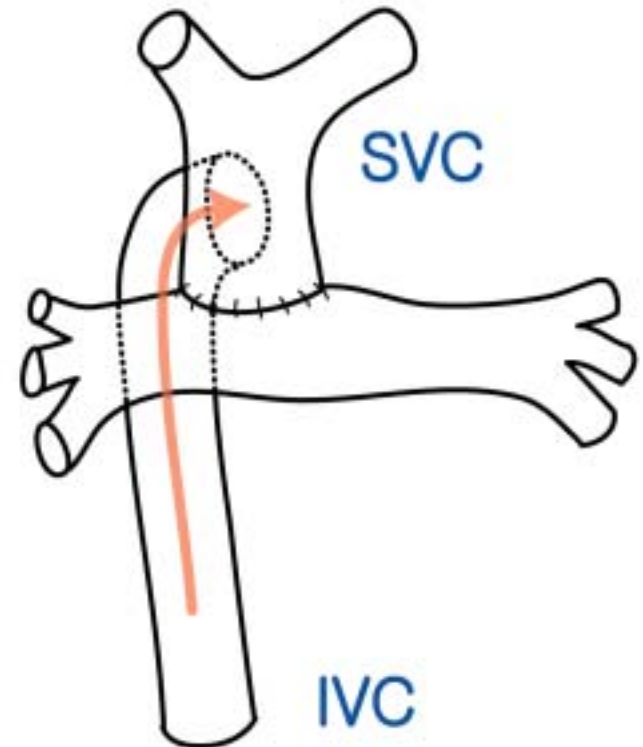
# History & Evolution

1983 1984

1987 1988

## Kawashima

total cavopulmonary shunt operation  
in complex cardiac anomalies.  
A new operation.



# History & Evolution

1983 1984

1987 1988

Puga

Modification of the Fontan operation applicable to the patients with left atrioventricular valve atresia or single atrioventricular valve.

# History & Evolution

1983 1984

1987 1988

Jonas

Modified Fontan procedure: Atrial baffle and systemic to pulmonary artery anastomotic techniques

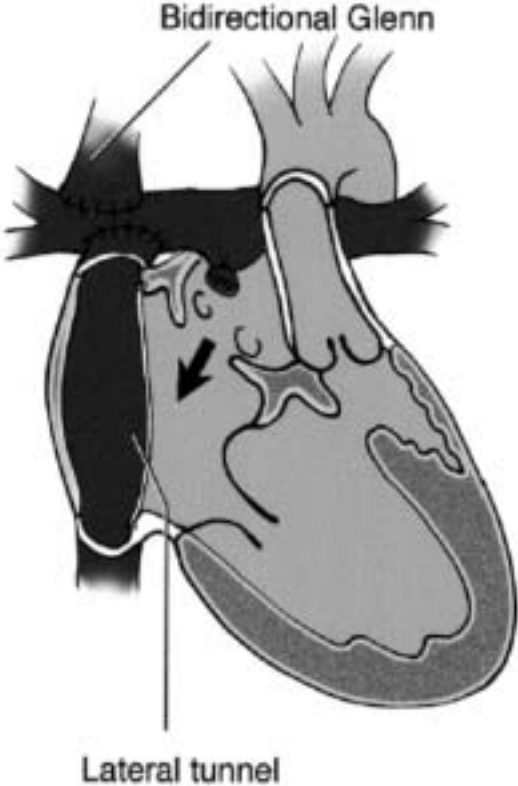
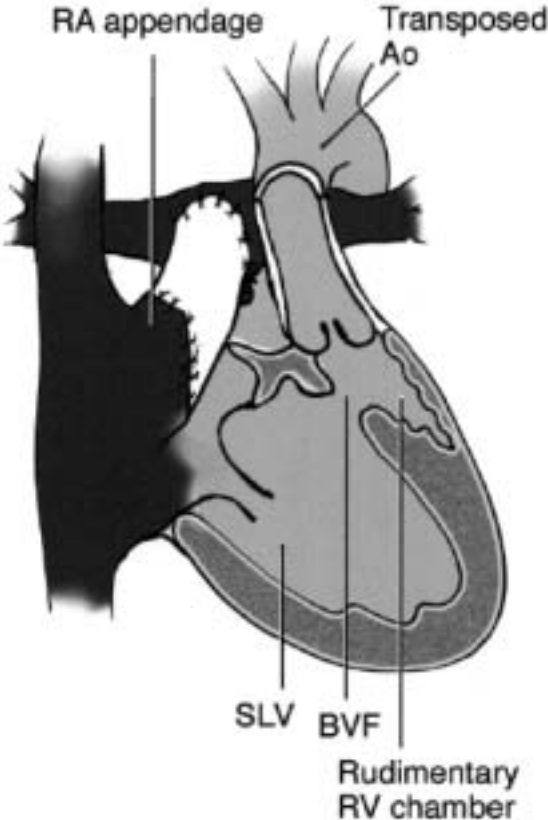
deLeval

Total cavopulmonary connection: a logical alternative to atriopulmonary connection for complex Fontan operations. Experimental studies and early clinical experience

# History & Evolution

early 1970s

late 1980s



# History & Evolution

early 1970s

late 1980s

## Total cavopulmonary connection

### Lateral tunnel

- (1) technical simple and reproducible regardless of type AV connection
- (2) avoidance of AV node injury and reduced risk of early or late arrhythmia because the amount of atrial tissue subjected to elevated venous pressure was minimized
- (3) reduction of turbulence, with subsequent diminution of energy loss and lessened risk of thrombosis

# Additional advancements

1989 1990

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## Mazzera

Bidirectional cavopulmonary shunt: clinical applications as staged or definitive palliation

## Bridges

Bidirectional cavopulmonary anastomosis as interim palliation for high-risk Fontan candidates: early results

# Additional advancements

1990

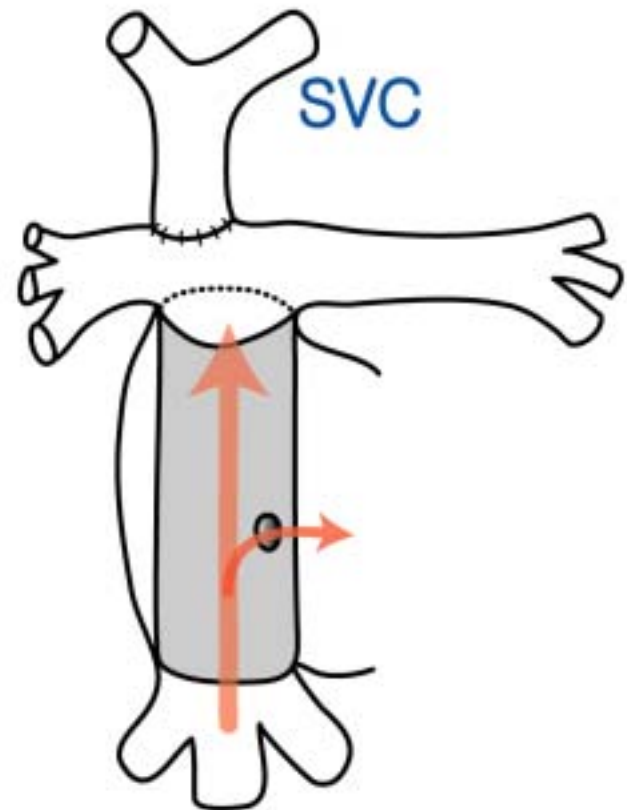
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## Bridges

the concept of adding a fenestration

providing a limited degree of right-to-left shunting

allowing for an increase in cardiac output at the expense of a tolerable degree of desaturation



# Modification

1990

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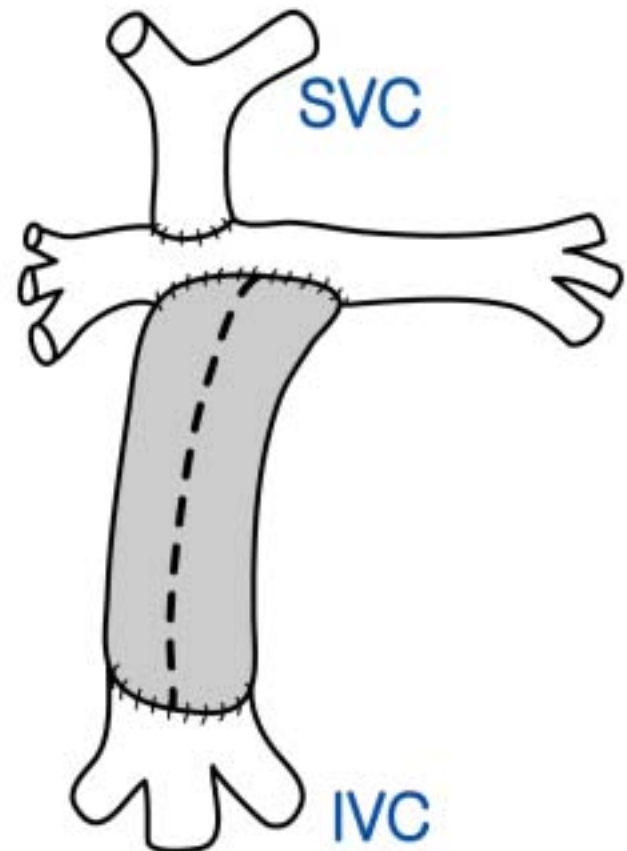
Marcelletti

IVC-PA extracardiac conduit. A new form of right heart bypass

providing a hydrodynamically efficient pathway for systemic venous flow.

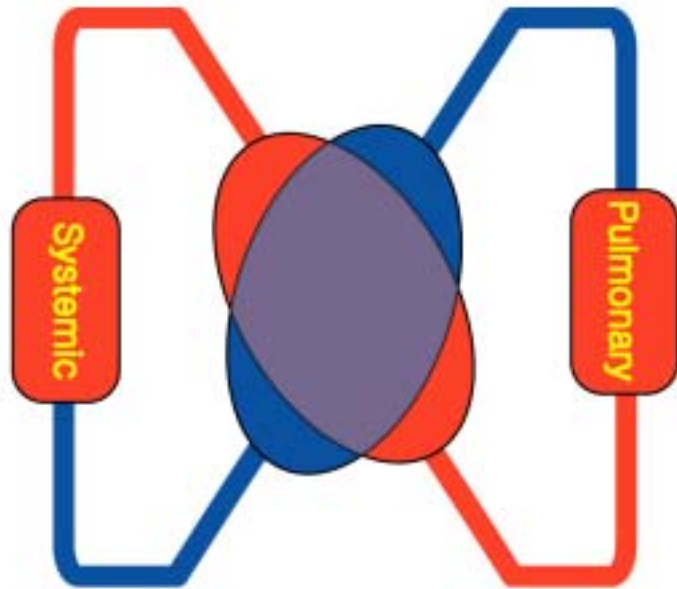
leaving atrium at low pressure  
minimizing atrial suture lines  
performing without aortic cross-clamping

**No growth potential**



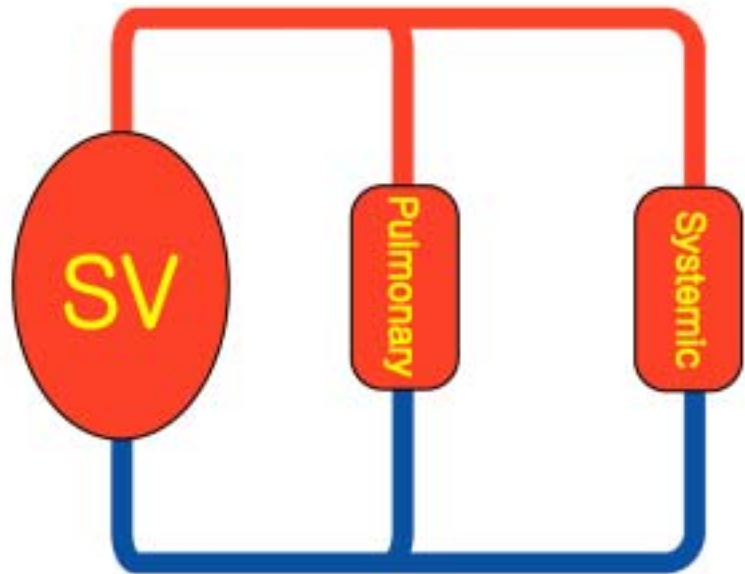
# Bi-ventricle vs. Single ventricle

2 pumps



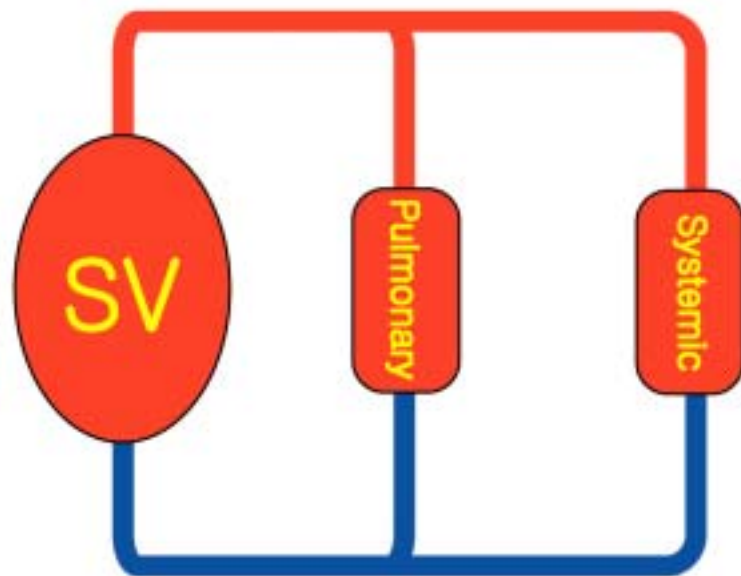
In series circulation

1 pump

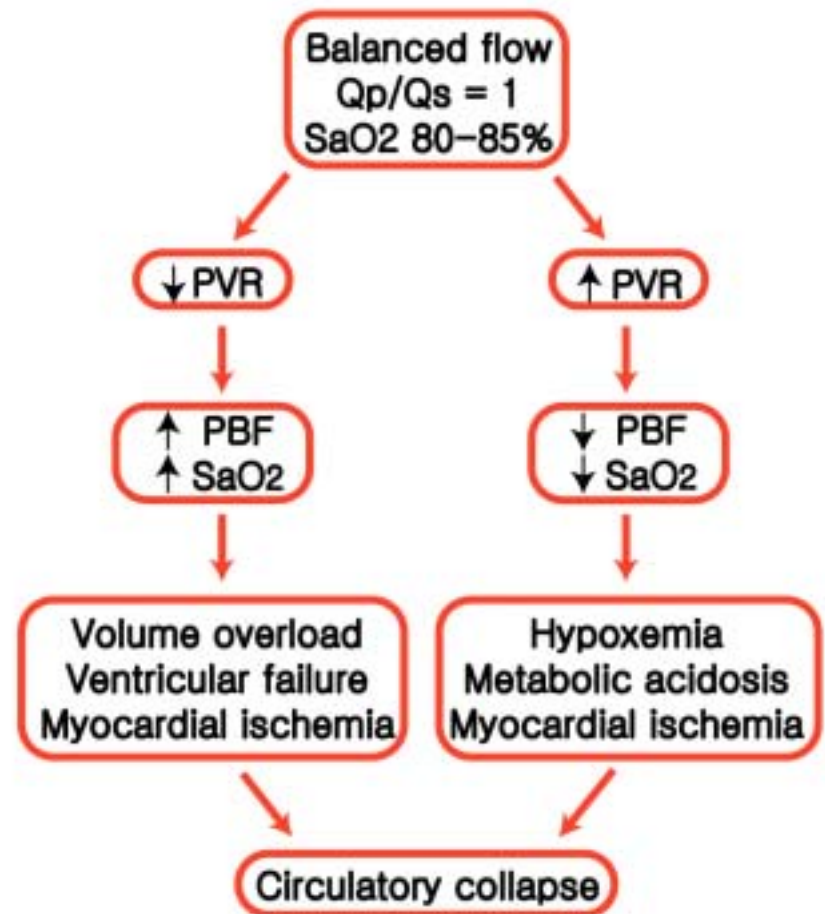


Parallel circulation

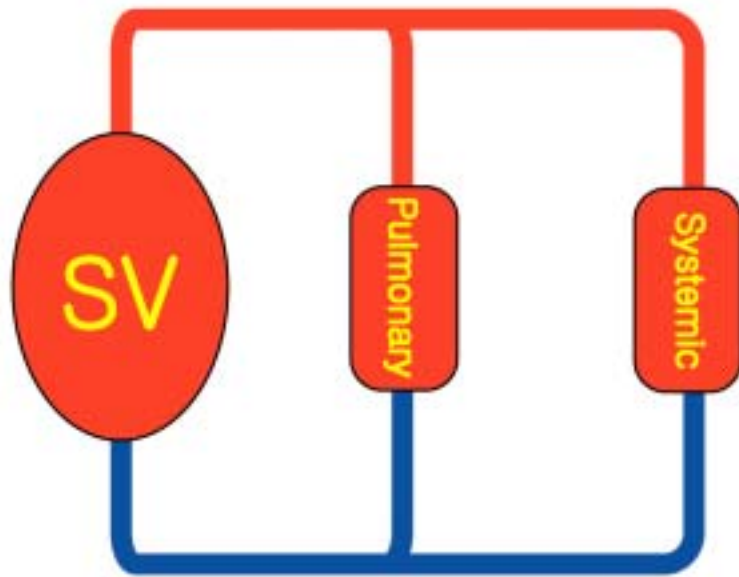
# Fontan procedure – Single ventricle



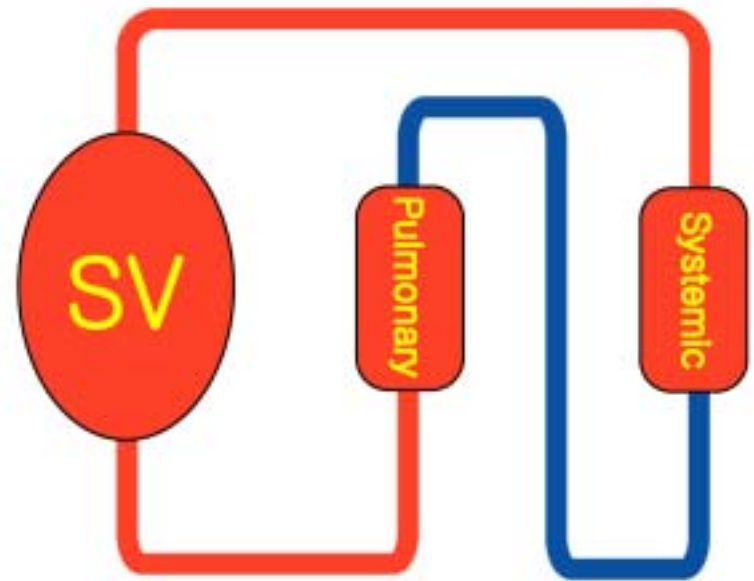
Parallel circulation



# Fontan procedure – Single ventricle



Parallel circulation



In series circulation

# Fontan procedure – Single ventricle



Stage I      Palliation  
Neonate

Stage II      Bidirectional Glenn shunt  
Hemi-Fontan  
2.5–6 mo

Stage III      Fontan  
2–3 yr

# Fontan procedure – Single ventricle



Stage I

Parallel circulation

Stage II

In series circulation  
relieving volume overload, but not  
cyanosis

Stage III

Total cavopulmonary connection

# Fontan procedure – Single ventricle



## Stage I

Qp/Qs balance

Stable PBF source (non-ductal)

Controlled PBF

- protection of pulmonary vascular bed from high pressures and flows

## Stage II

- keeping adequate PVR

Relief of any significant obstruction to systemic outflow or pulmonary venous return

## Stage III

# Fontan procedure – Single ventricle



**Stage I**    BT shunt  
                 PA banding  
                 Atrial septectomy  
                 DKS / Norwood

Stage II

Stage III

# Fontan procedure – Single ventricle

## Stage I

1. Inadequate pulmonary circulation  
(or ductal-dependent)

→ Blalock-Taussig shunt

## Stage II

2. Excessive pulmonary circulation  
without systemic outflow obstruction

→ PA banding

3. Excessive pulmonary circulation  
with systemic outflow obstruction


→ DKS / Norwood

## Stage III

4. Balanced pulmonary blood flow

→ Observation

# Fontan procedure – Single ventricle



**Stage I** Potential or mild systemic outflow obstruction

**Stage II** Neonatal DKS/Norwood repair  
distortion of pulmonary (neoaortic) valve caused by PAB  
rapid progression of ventricular hypertrophy – feedback loop

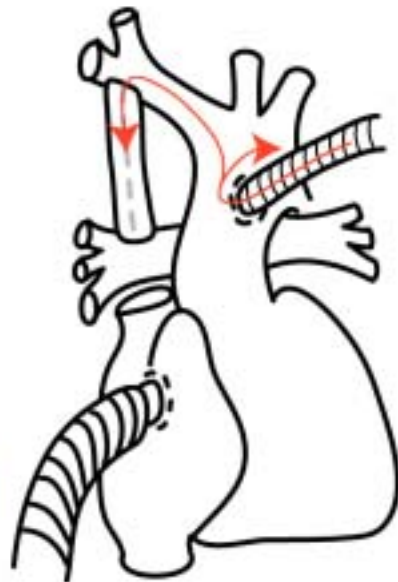
**Stage III** Delaying – early BCPS  
greater hemodynamic stability

# Fontan procedure – Single ventricle

**Stage I** Mechanical assistance in BT shunt lesion  
– Perioperative hemodynamic collapse  
'ECMO without the oxygenator'

Stage II

Stage III



# Fontan procedure – Single ventricle

Stage I

Inherent instability of parallel circulations

Interval  
period

- A progressive decrease in PVR
- Aggravation of a potential coarctation & obstruction to systemic outflow
- Outgrowing BT shunt or PAB

Stage II

catheterization with cineangiography  
cardiac MRI

Stage III

# Fontan procedure – Single ventricle

Stage I Bidirectional Glenn / Hemi-Fontan


Accessory PBF (Pulsatile)  
providing hepatic venous flow to  
pulmonary circulation

Stage II – precluding development of PAVM  
improving pulmonary artery growth

volume loading

Stage III  
an option as a definitive palliation

# Fontan procedure – Single ventricle



Stage I Bidirectional Glenn / Hemi-Fontan

Excising or oversewing the native  
pulmonary valve  
significant source of thromboemboli

Stage II

Stage III

# Fontan procedure – Single ventricle



Stage I Concomitant procedures

Stage II

pulmonary arterioplasty  
revision atrial septectomy  
repair of residual or recurrent aortic  
coarctation  
atrioventricular valvuloplasty

Stage III

# Fontan procedure – Single ventricle



Stage I    Fontan procedure

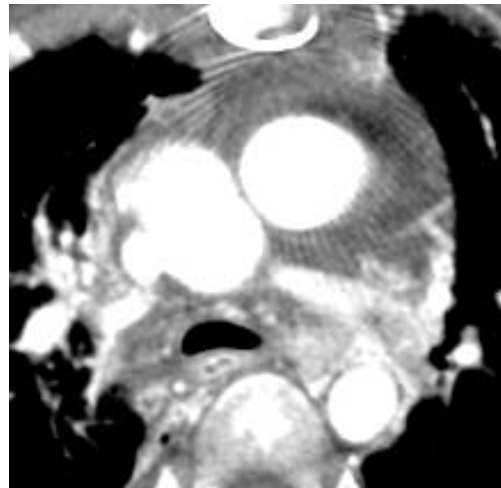
Extracardiac conduit  
conduit size  $\geq$  18mm

Stage II    Lateral tunnel

Stage III

# Fontan procedure – Single ventricle

Stage I Extracardiac Fontan  
PA distortion

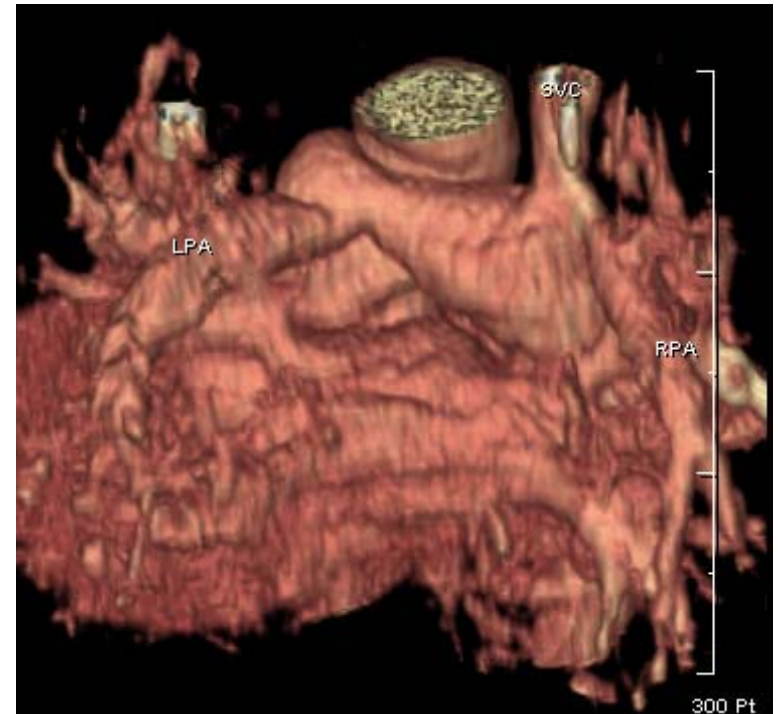


Stage II

Stage III

#1

#2



# Fontan procedure – Single ventricle

Stage I Fontan procedure – Fenestration

high risk candidates

$R_p > 2 \text{ WU}$

$\text{PAP} > 15 \text{ mmHg}$

Ventricular dysfunction

or  $\text{EDP} > 15 \text{ mmHg}$

PA distorsion or hypoplasia

moderate to severe AVVR

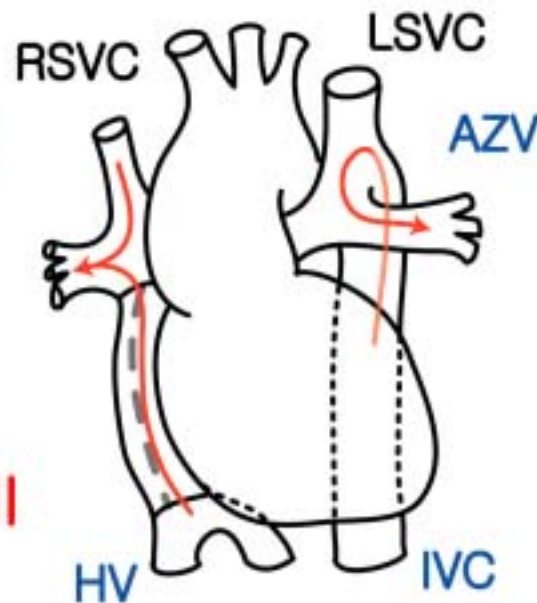
Stage III

intraop CVP  $> 18 \text{ mmHg}$

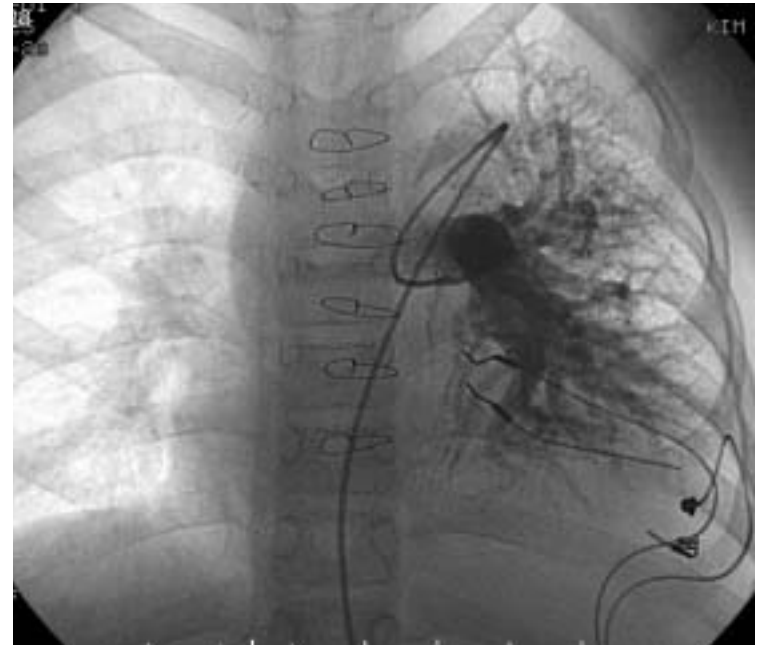
# Fontan procedure – Single ventricle

**Stage I** Pulmonary AV malformation/fistula  
s/p Kawashima & DKS  
s/p extracardiac conduit (HV to PA)

**Stage II**



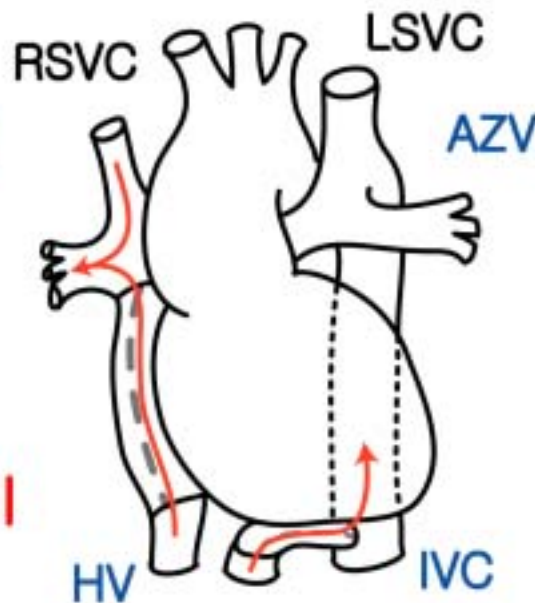
**Stage III**



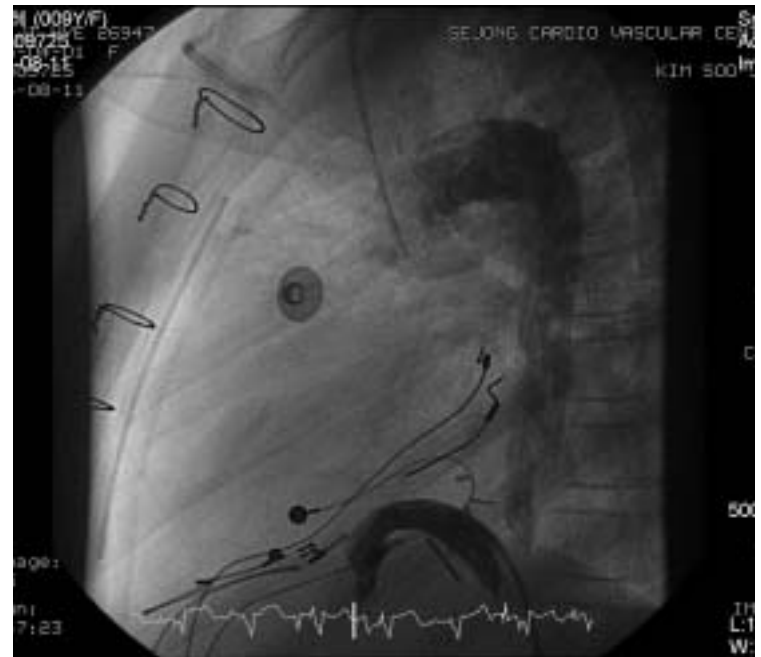
# Fontan procedure – Single ventricle

**Stage I** Pulmonary AV malformation/fistula  
s/p Kawashima & DKS  
s/p extracardiac conduit (HV to PA)

**Stage II**



**Stage III**



# Summary

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Continued close follow-up of Fontan patients will allow both surgeon and cardiologist to confidently determine if staged strategy provides for optimal long-term cardiovascular performance.