

# **FONTAN OPERATION IN SINGLE VENTRICLE TREATMENT**

Dr. Trần Vũ Hoàng  
Intensive care unit



Cardiac surgery was established in 2010.  
2010 – 2015 : 31 cases Glenn operation  
Fontan: not yet.

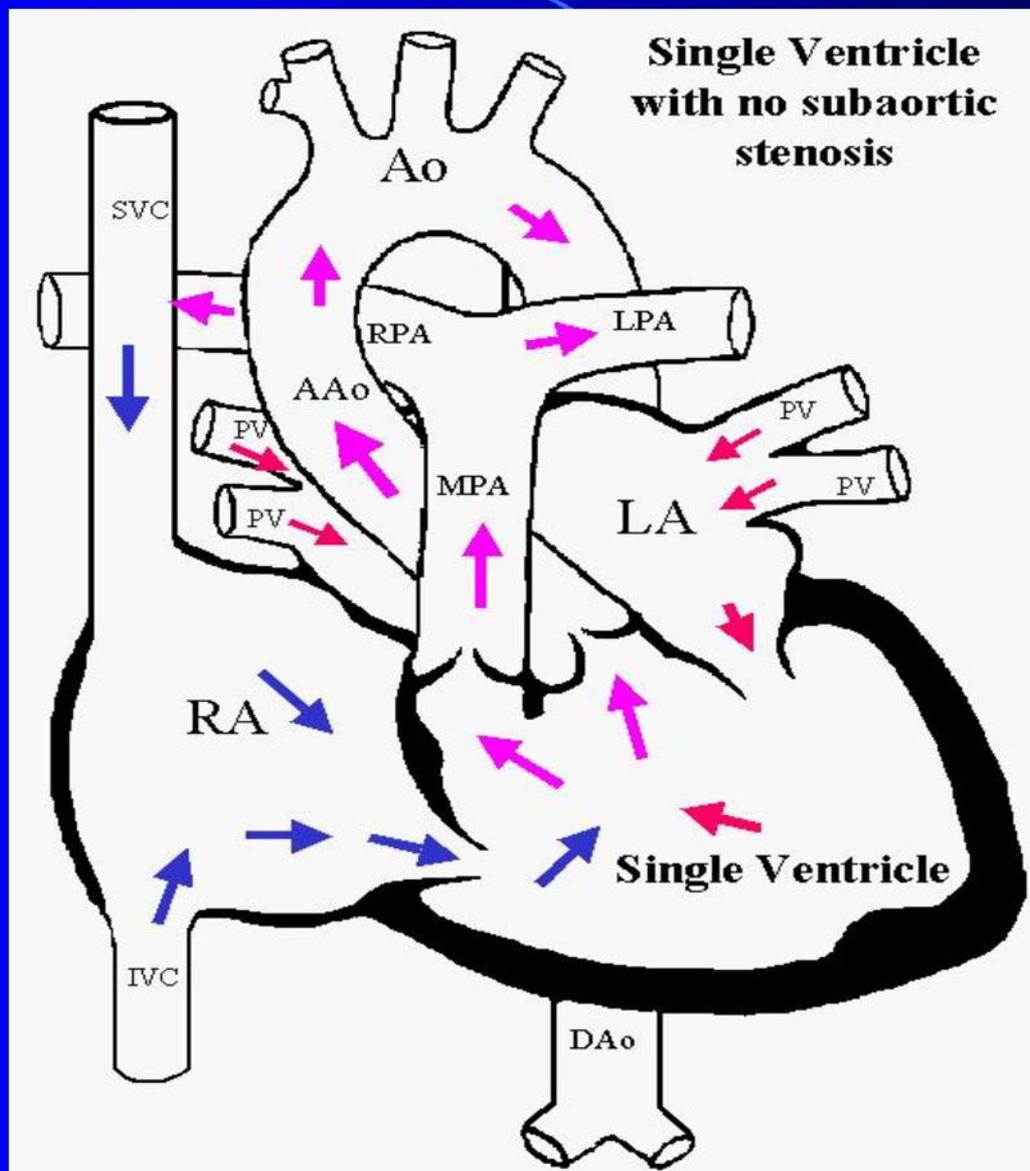
- **I. PATHOLOGY:**

- - Both AV valves are connected to a main -> single ventricular chamber.
- - 80% characteristics of the LV.
- D-TGA or L-TGA is present in 85% of cases.
- 75% : single ventricle is double – inlet LV + L-TGA

- **II.PATHOPHYSIOLOGY :**

- Because there is complete mixing in the single ventricle, the systemic arterial saturation is determined primarily by amount of PBF.
- With PS, PBF is decrease and cyanosis is present. With pulmonary atresia, cyanosis is intense at birth.
- With the pulmonaty valve is not stenotic, the PBF is large and signs of CHF develop within days or weeks without cyanosis.

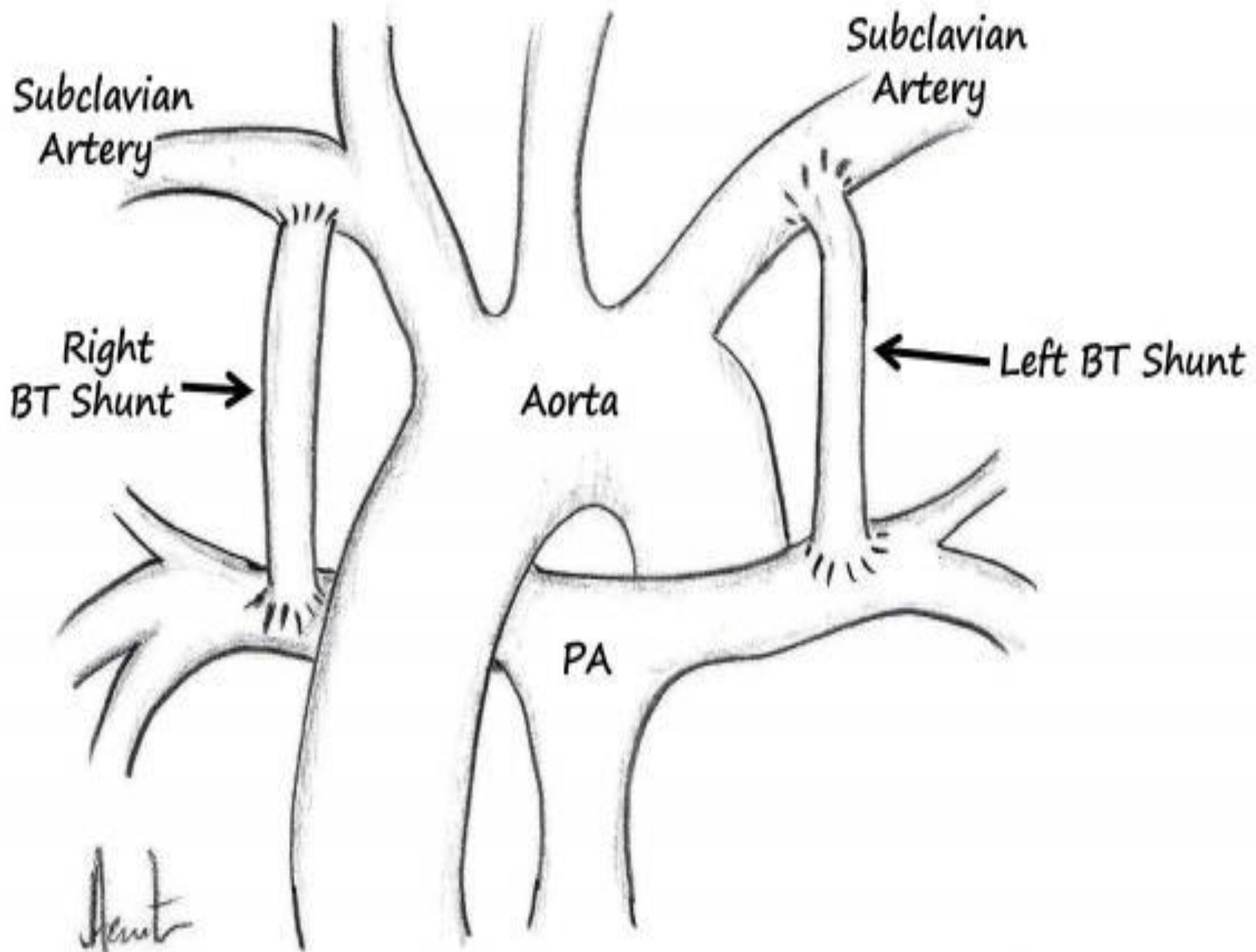
# Single ventricle



### **III.MANAGEMENT:**

- 1. Initial Medical Management :
  - Newborns with severe PS or pulmonary atresia and those with interrupted aortic arch or coarctation require PGE1 infusion and other supportive measures before surgery.
  - Anticongestive measures with digoxin and diuretics should be taken if CHF develops.

- 2. Surgery treatment:
- - Stage 1:
- B – T shunt ( Blalock – Taussig shunt) is necessary for cyanotic patients with PS or pulmonary atresia
- - PDA is ligated after placement of the shunt.



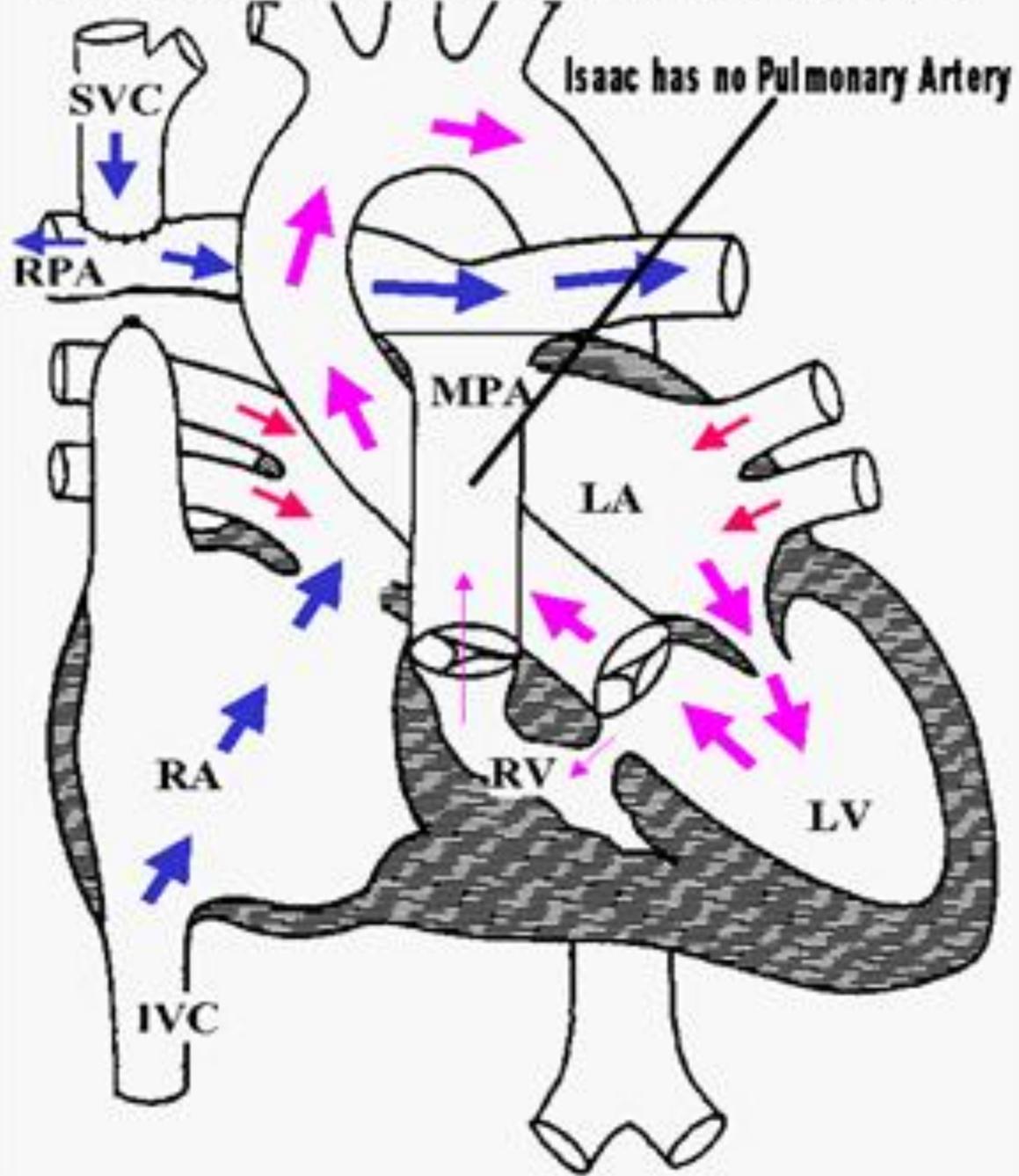
Spent  
2011

- PA banding is considered for infants with CHF and pulmonary edema resulting from increase PBF.

- - Stage 2:
- Glenn operation is carried out between the ages of 3 and 6 months, before proceeding Fontan operation.
- After this stage, the child needs to be followed up with attention to the O<sub>2</sub> saturation. Initial improvement in O<sub>2</sub> saturation ( 85%), but deterioration in O<sub>2</sub> saturation may occur in the months postoperatively .

- Cardiac catheterization is performed by 12 months.
- Low mean PA pressure ( $< 16-18$  mmHg), low PVR  $< 2$  units, and low end diastolic pressure less than 12mmHg.

# TRICUSPID ATRESIA - S/P GLENN SHUNT

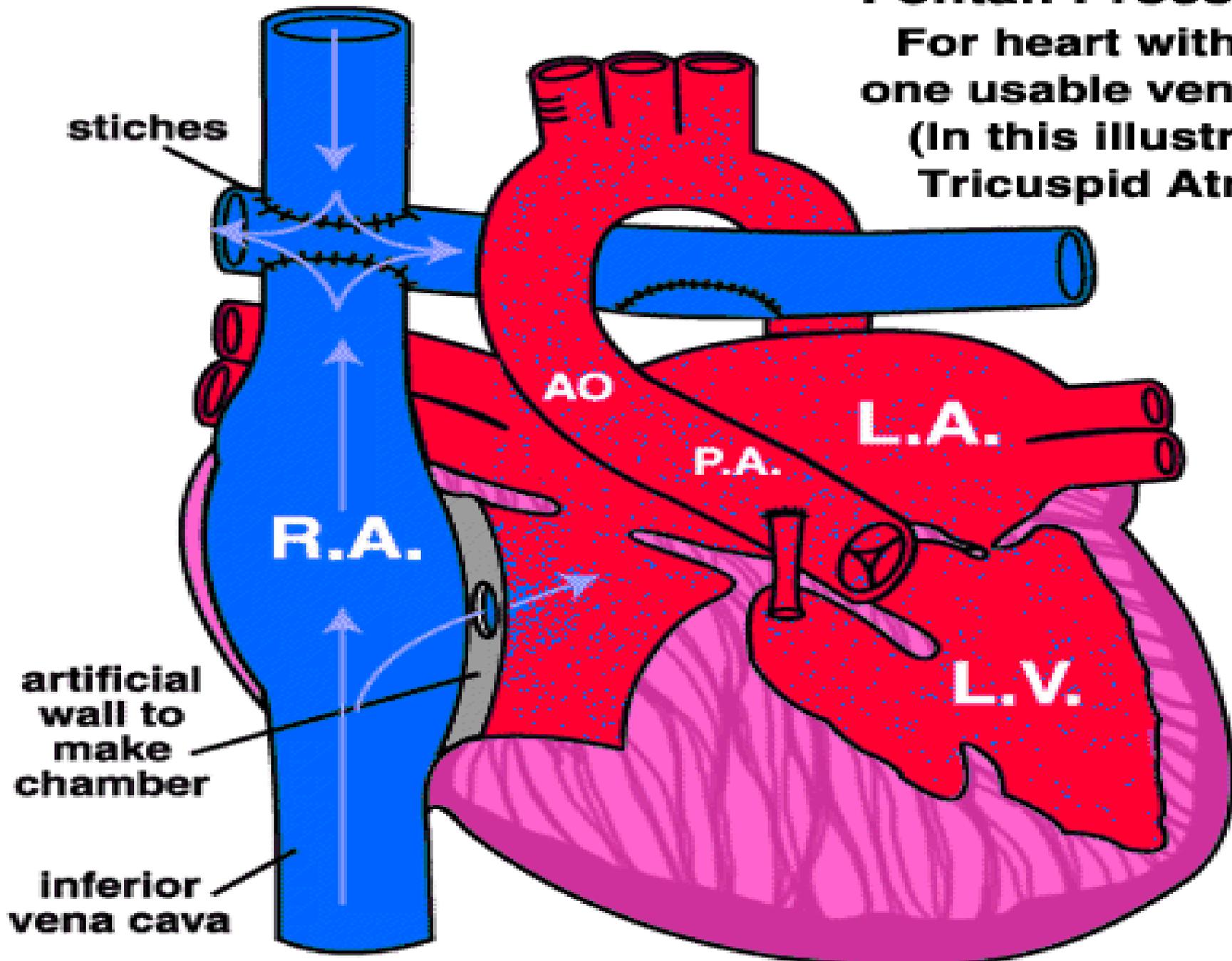


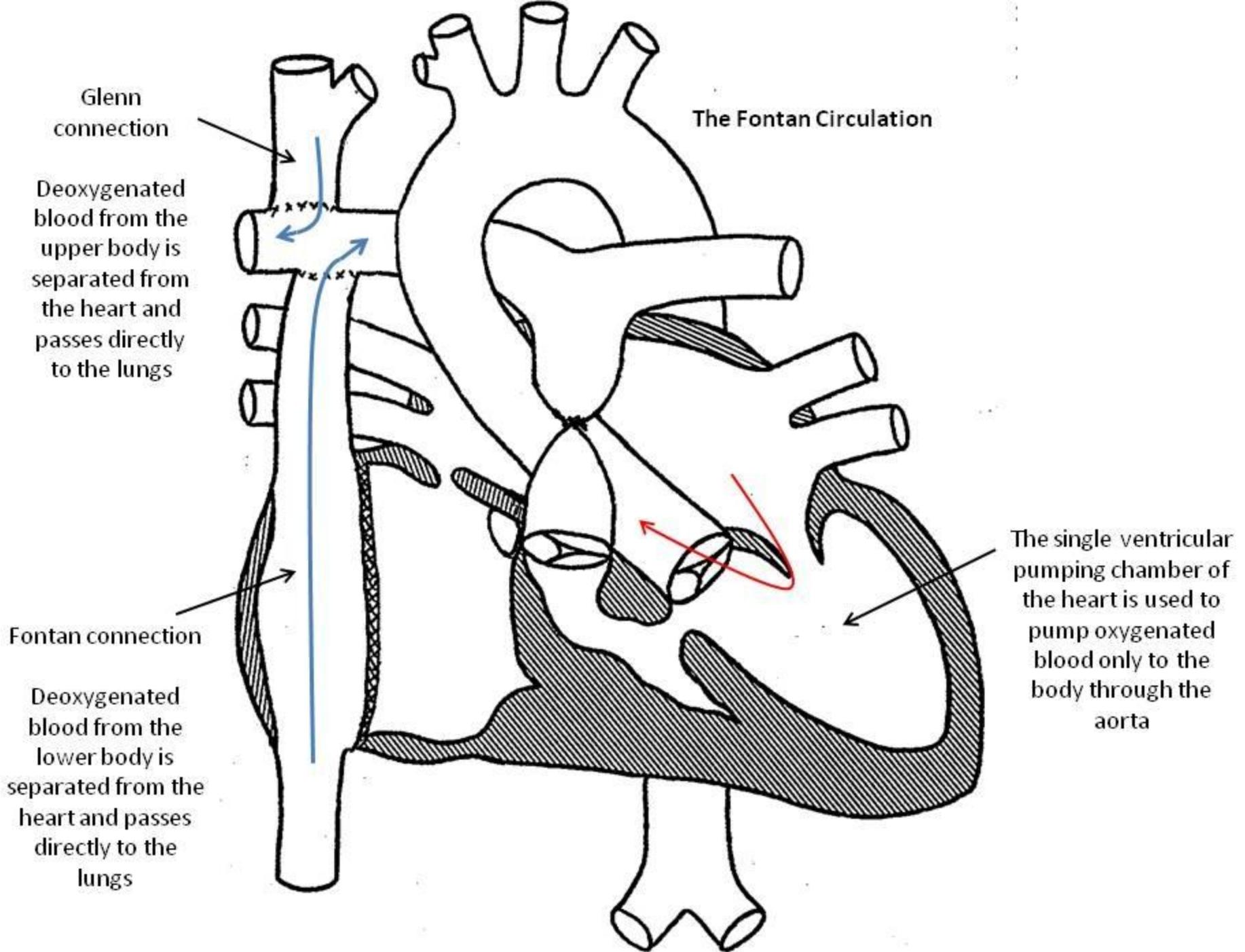
- - Stage 3:
- The Fontan operation is performed at 18 to 24 months of age
- There are risk factors for Fontan operation:  
two or more of these risk factors constitutes a high-risk situation:

- 1. High PVR ( $> 2\text{U/m}^2$ ) or high mean PA pressure ( $> 18\text{mmHg}$ ).
- 2. Distorted PAs secondary to previous shunt operations.
- 3. Poor systolic or diastolic ventricular function, with LV end – diastolic pressure greater than  $12\text{mmHg}$ .
- 4. AV valve regurgitation.

# Fontan Procedure

For heart with only one usable ventricle  
(In this illustration  
Tricuspid Atresia)



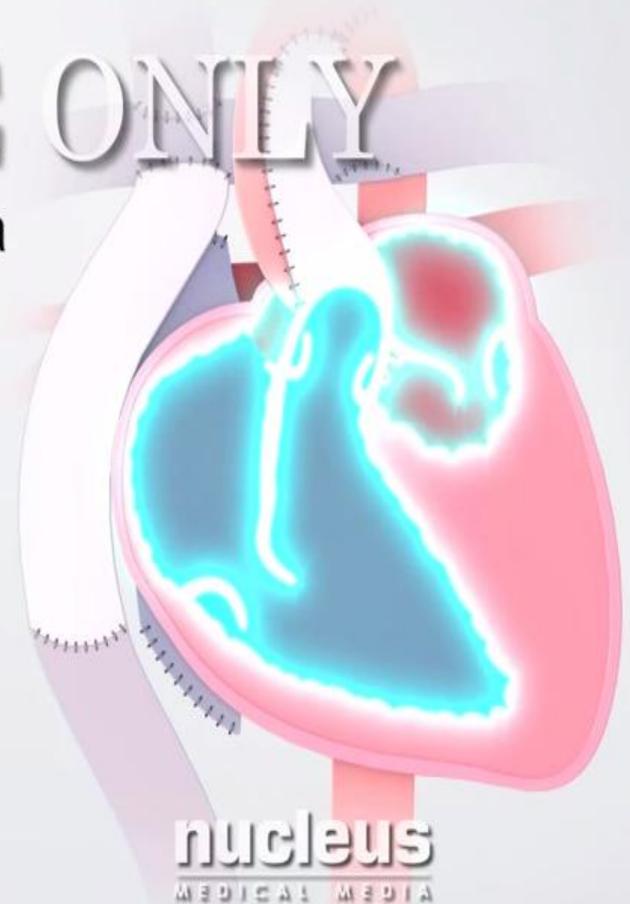


In 500 cases Fontan operation: successful rate 85% in first month, 80% in 5 year, 70% in 10 year.

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## *Goals of 3 operations:*

- Delivery of oxygen-rich blood to the body through a new, expanded aorta
- Creation of a new path for oxygen-poor blood to reach the lungs
- Prevention of mixing oxygen-rich blood and oxygen-poor blood



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## Editorials

### Fontan Operation After 3 Decades

#### What We Have Learned

Welton M. Gersony, MD

[Author Affiliations](#)

Correspondence to Welton M. Gersony, MD, Children's Hospital of New York, 3959 Broadway, 2 North, Room 263, New York, NY 10032. E-mail [wmgl@columbia.edu](mailto:wmgl@columbia.edu)

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management of patients with a single ventricle.

- Controversies
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## Mortality

Perioperative and early mortality after the Fontan operation have decreased markedly over the past 3 decades. Among early survivors, late mortality and modes of death can be assessed with the notion that predictors of intermediate or late mortality can be uncovered. In the report by Khairy et al,<sup>1</sup> 82.6% of the early survivors were alive and had not had a cardiac transplant 15 to 20 years later. It is of interest that this study found no significant difference in life expectancy beyond the postoperative period between the more obsolete, less efficient RA-PA connections and the direct caval-pulmonary artery connections. Among long-term survivors, the data from this series indicated that the 3 most common causes of late death were thromboembolism, heart failure, and sudden death.

Thromboembolic late death was found to be more common than previously documented.<sup>4</sup> The predictors of thromboembolic death by multivariate analysis were lack of aspirin/warfarin therapy and intracardiac thrombus. Five of the 6 deaths were in patients who had direct RA-PA connections, which perhaps indicates the modern total cavopulmonary connections are less likely to be associated with thromboembolism; however, less follow-up time is available for the latter group of patients.

The higher risk for heart failure related to the presence of a single morphological right as opposed to left ventricle is consistent with other studies that indicate the

## Fenestration

Provision of a surgical fenestration between the total cavopulmonary connection and the right atrium is a part of Fontan operation management at most centers. In the immediate postoperative period after the Fontan operation, a fenestration allows adequate cardiac output, albeit with lower than normal oxygen saturation. A number of reports have indicated a more uneventful postoperative course, with smaller pleural effusions and earlier discharge from the intensive care unit and the hospital. For many centers, all Fontan operations include a fenestration regardless of the preoperative risk assessment; however, more recently, external conduits have not always been fenestrated, given the more technical challenge of keeping the anastomosis open at an optimal size. If one accepts the advantages of fenestration for many patients in the early postoperative period, the question must be asked as to whether a persistent fenestration becomes a risk factor in the presence of late right-sided thrombus formation and possible paradoxical embolization. Most centers will close residual significant-sized fenestrations, as determined by arterial oxygen saturation, by percutaneous device placement. It is likely that low-risk Fontan patients can be managed surgically without fenestration initially, thereby eliminating the need to close the fenestration if it persists over many years.

## Protein Losing Enteropathy

The development of protein-losing enteropathy is associated with a poor clinical

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## Protein Losing Enteropathy

The development of protein-losing enteropathy is associated with a poor clinical course in patients after a Fontan operation. Despite all measures, mortality is 50% within the first 5 years after the diagnosis is made.<sup>8</sup> Multiple treatment modalities exist for protein-losing enteropathy, and this reflects the absence of definitive management principles based on its cause. Protein-losing enteropathy remains one of the greatest challenges in the management of post-Fontan single-ventricle patients.

## Anticoagulation Therapy

The present findings by Khairy and associates<sup>1</sup> address the controversial aspects of whether anticoagulation/antiplatelet therapy should be administered routinely to all Fontan patients. The significant incidence of thromboembolic death in this series suggests that morbidity from thromboembolism is higher than previously reported. It would seem prudent to institute daily antiplatelet therapy for most Fontan patients. There has been no consensus on the requirement for coumadin anticoagulation therapy. Most centers will provide anticoagulation therapy for patients whom they consider to be at higher risk (eg, RA-PA connection, external conduit, sluggish venous circulation, or low cardiac output).

## Fontan Conversion and Antiarrhythmic Surgical Intervention

As patients who had RA-PA connections in early life reach young or middle

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- **The main defects in this category are:**
- Tricuspid Atresia
- Pulmonary Atresia with underdeveloped right ventricle
- Double inlet left ventricle
- Double outlet right ventricle with associated defects that prevent repair
- Hypoplastic left heart syndrome
- Other defects that cannot be fixed by other types of surgery

