대한흉부심장혈관외과학회 제12차 전공의 연수교육
May 23-25, 2019

Single Ventricle

Cheul Lee, MD

Pediatric and Congenital Heart Surgery
Seoul St. Mary's Hospital
College of Medicine, The Catholic University of Korea

Single Ventricle

- Broad category of hearts that lack two well-developed ventricles
- Functionally univentricular heart
- One of the most challenging congenital heart diseases

Congenital Heart Surgery Nomenclature and Database Project: Single Ventricle

Marshall L. Jacobs, MD, and John E. Mayer, Jr, MD

Section of Cardiothoracic Surgery, St. Christopher's Hospital for Children, Philadelphia, Pennsylvania, and Department of Cardiac Surgery, Children's Hospital, Boston, Massachusetts

The extant nomenclature for single ventricle (SV) hearts is reviewed for the purpose of establishing a unified reporting system. The subject was debated and reviewed by members of the STS-Congenital Heart Surgery Database Committee and representatives from the European Association for Cardiothoracic Surgery. Efforts were made to include all relevant nomenclature categories using synonyms where appropriate. Although many issues regarding single ventricle or univentricular hearts remain unresolved among anatomists and pathologists, a classification is proposed that is relevant to surgical therapy. A comprehensive database set is presented, which is based on a hierarchical scheme. Data are entered at various levels of complexity and detail, which can be determined by the clinician. These data can lay the foundation for comprehensive risk stratification analyses. A minimum data set is also presented that will allow for data sharing and would lend itself to basic interpretation of trends. Outcome tables relating diagnoses, procedures, and various risk factors are presented.

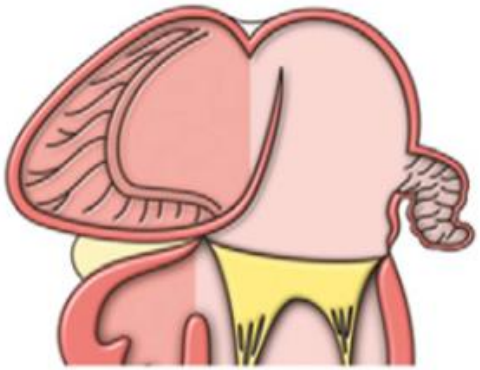
(Ann Thorac Surg 2000;69:S197–204)

Classification of Single Ventricle

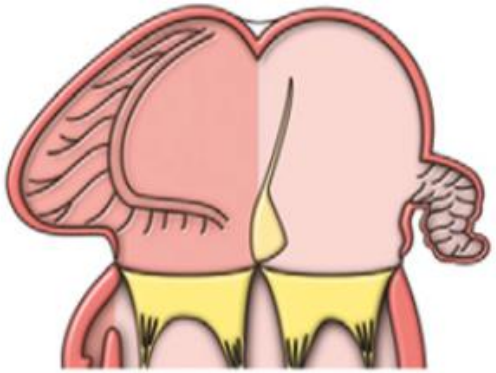
- Double inlet left ventricle
- Double inlet right ventricle
- Mitral atresia
- Tricuspid atresia
- Unbalanced atrioventricular canal defect
- Heterotaxia syndrome
- Other

Ann Thorac Surg 2000;69:S917-204

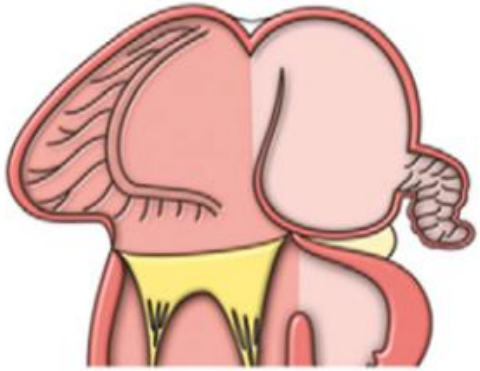
Univentricular Atrioventricular Connections



Absent right AV connection



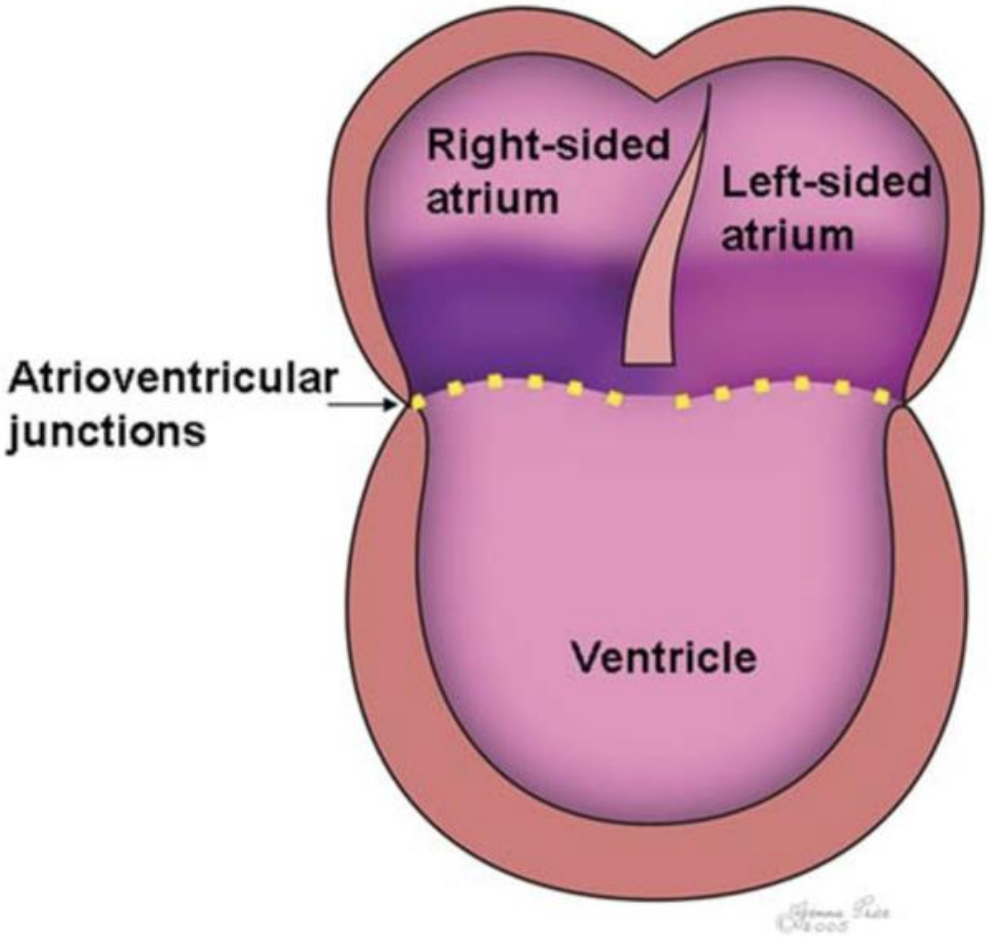
Double inlet ventricle



Absent left AV connection

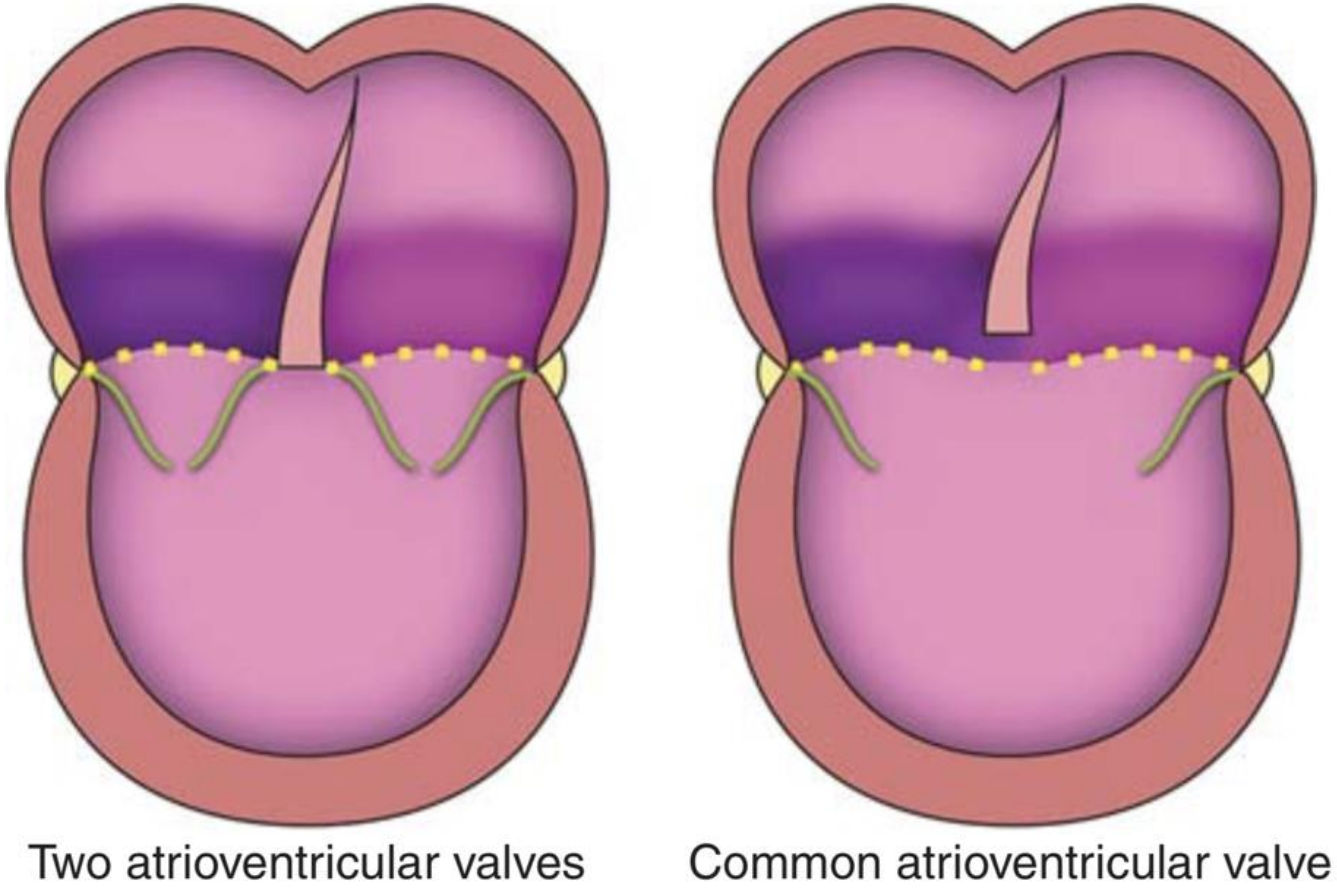
Anderson RH, et al. Paediatric Cardiology. 3rd ed.

Double Inlet Ventricle



Cardiol Young 2006;16 Suppl 1:22-6

Double Inlet Ventricle



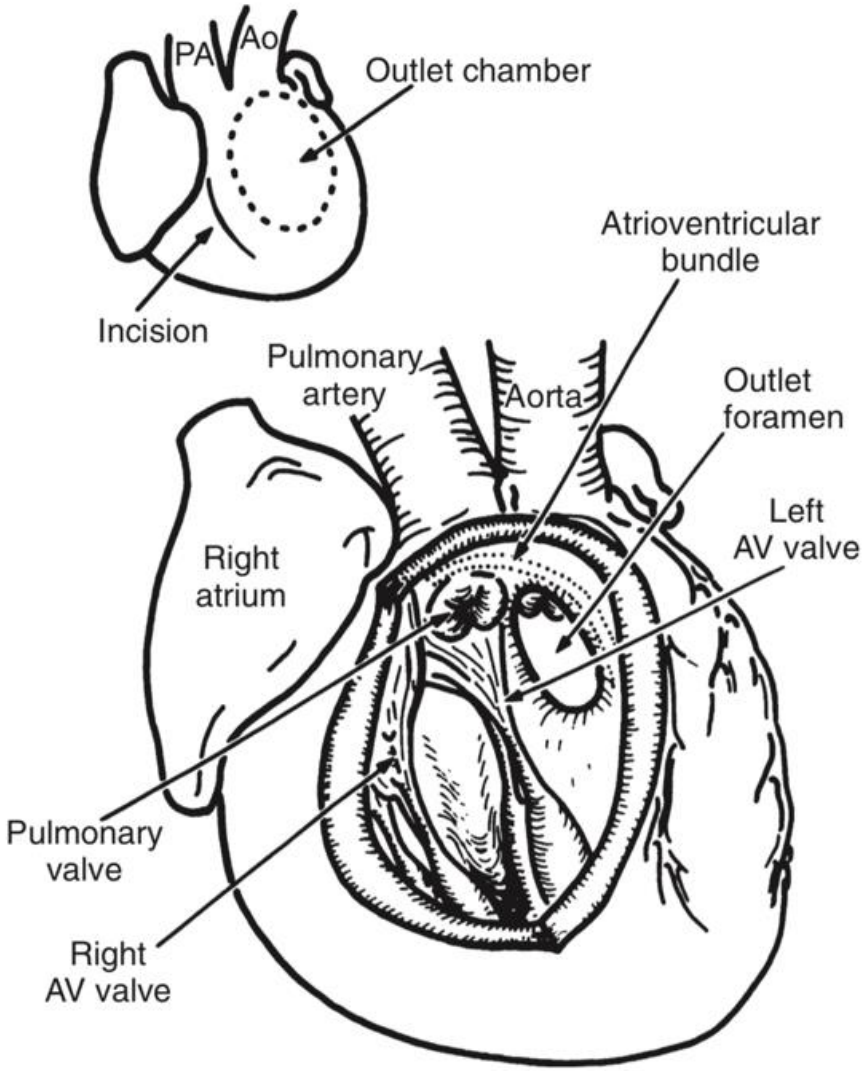
Cardiol Young 2006;16 Suppl 1:22-6

Double Inlet Left Ventricle (DILV)

- DILV, {S,L,L}
- DILV, {S,D,D}
- DILV, {S,D,N} (Holmes heart)
- DILV, DOLV
- DILV, DORV

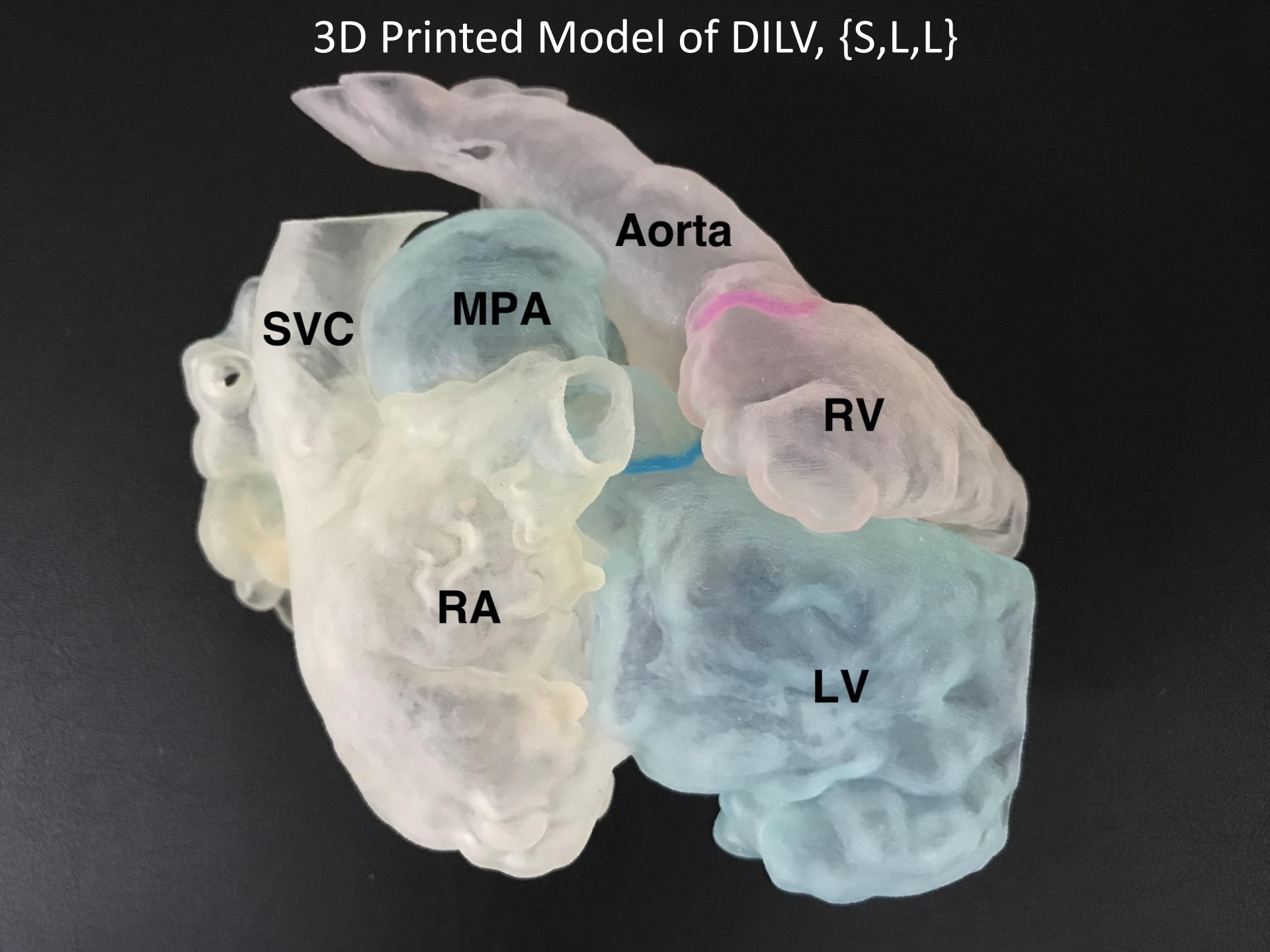
- Atrial situs is usually solitus.
- Ventriculo-arterial connection is usually discordant.

Double Inlet Left Ventricle, {S,L,L}

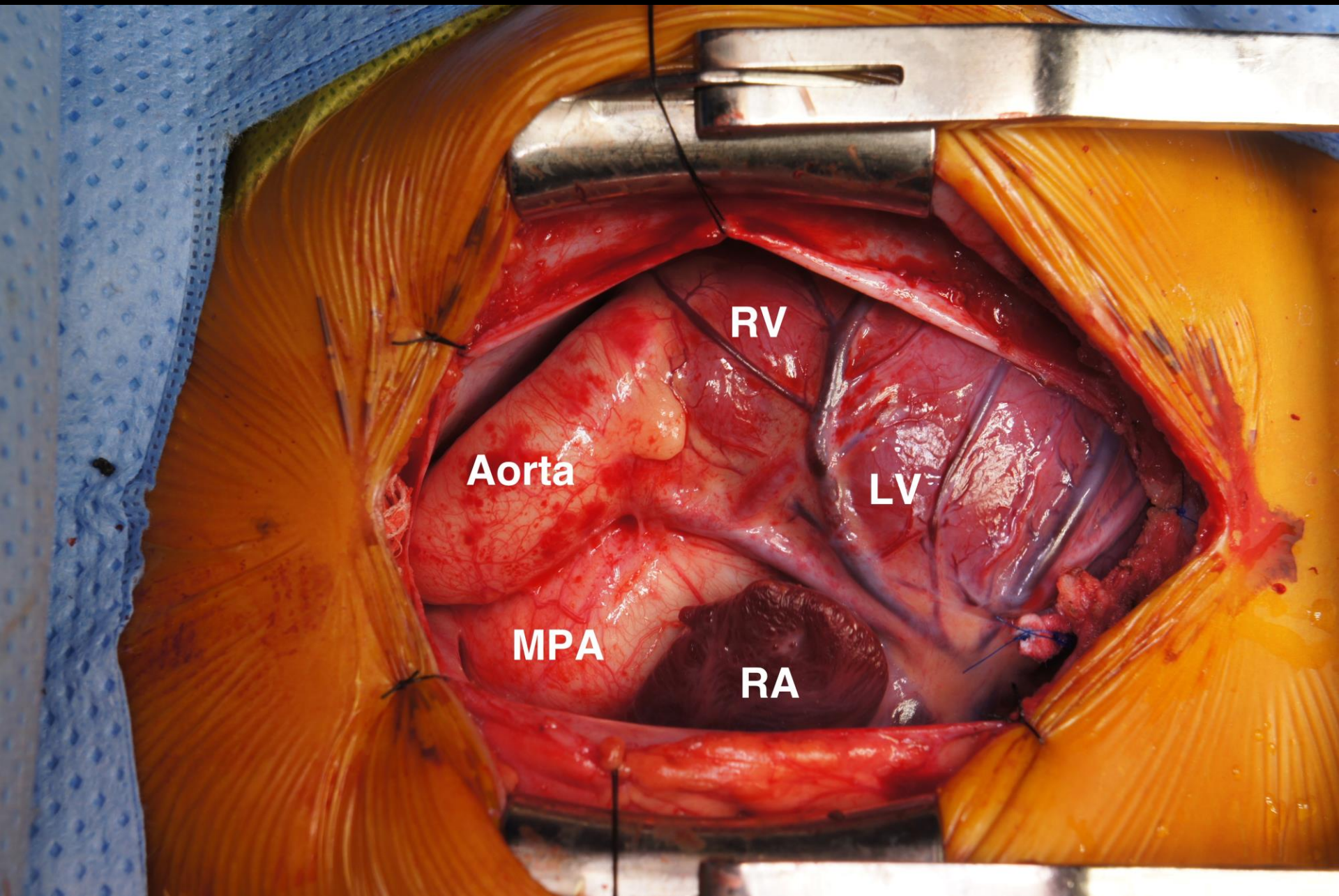


Mavroudis C, et al. Pediatric Cardiac Surgery. 4th ed.

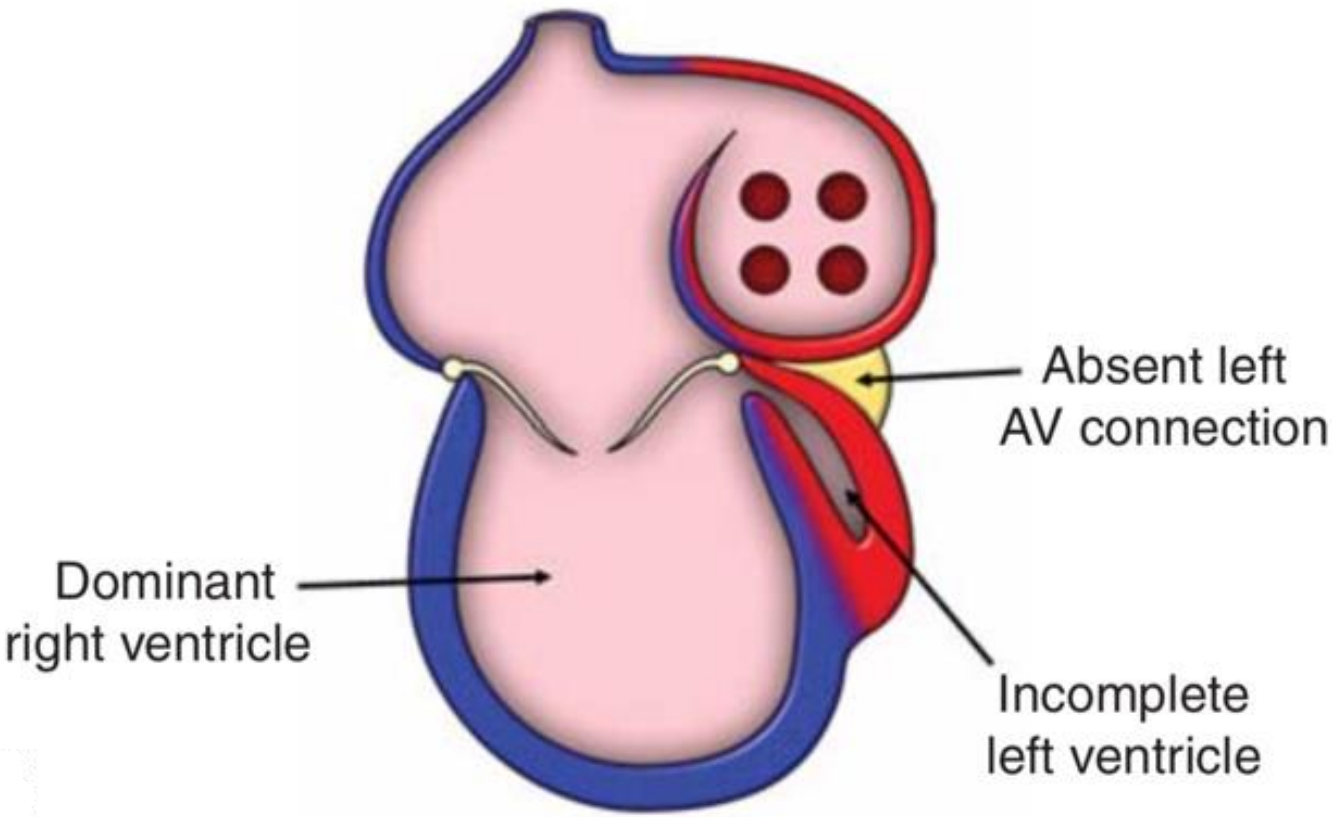
3D Printed Model of DILV, {S,L,L}



Double Inlet Left Ventricle, {S,L,L}

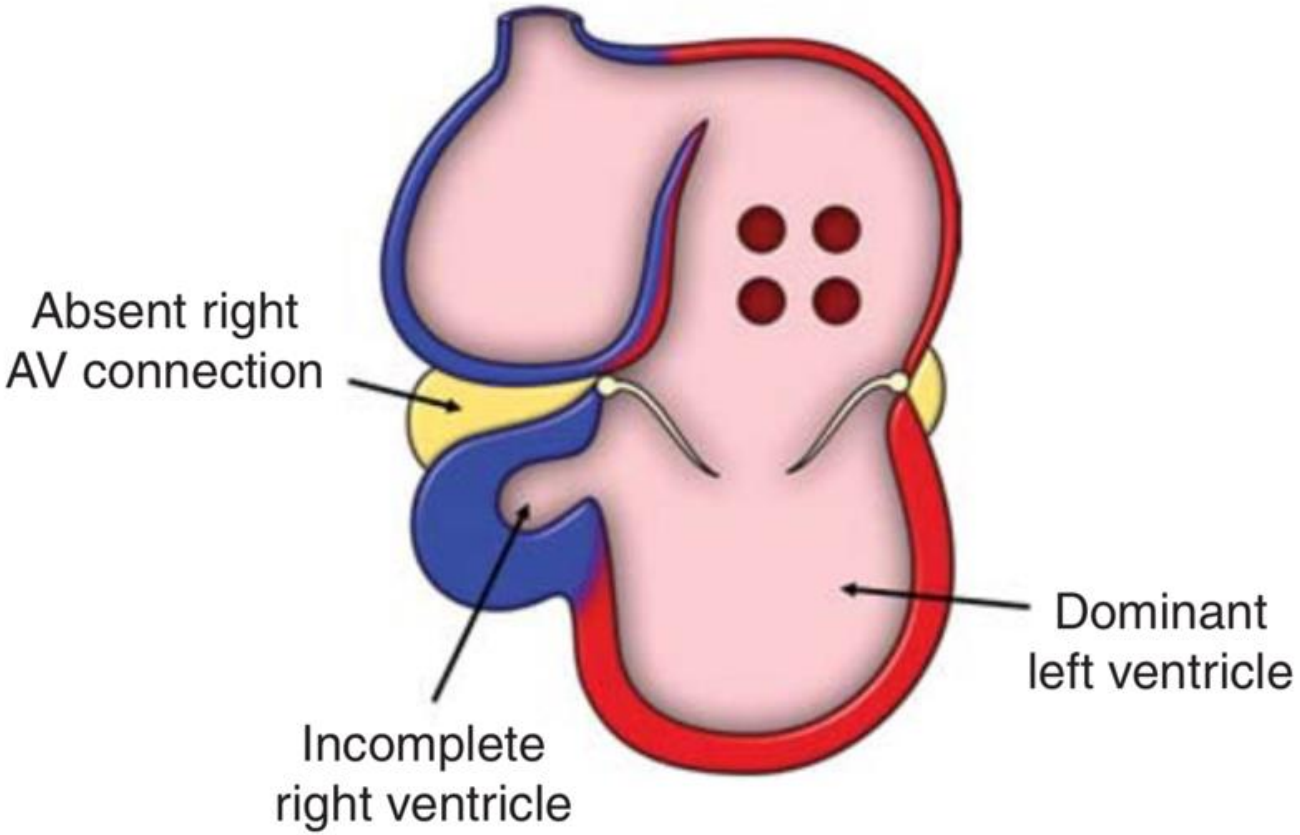


Mitral Atresia



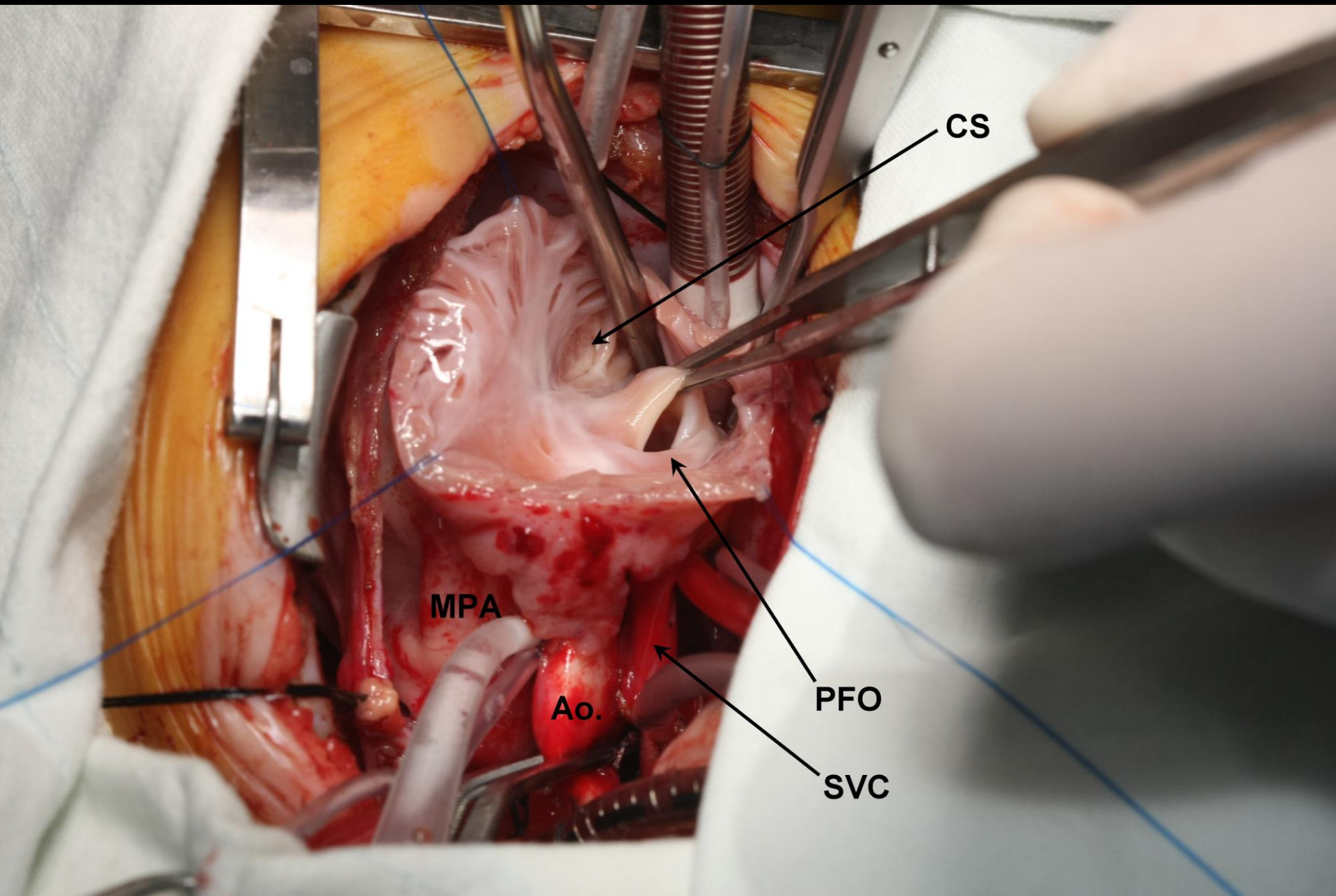
Cardiol Young 2006;16 Suppl 1:27-34

Tricuspid Atresia



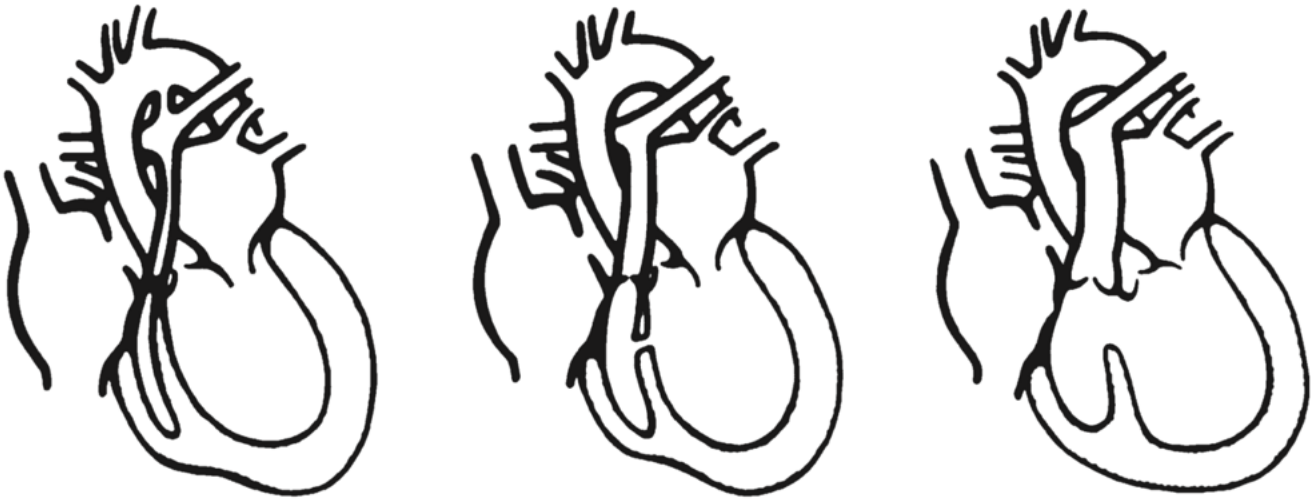
Cardiol Young 2006;16 Suppl 1:27-34

Tricuspid Atresia



Classification of Tricuspid Atresia

Tricuspid atresia with no transposition (69% - 83%)



I(a)
Pulmonary atresia

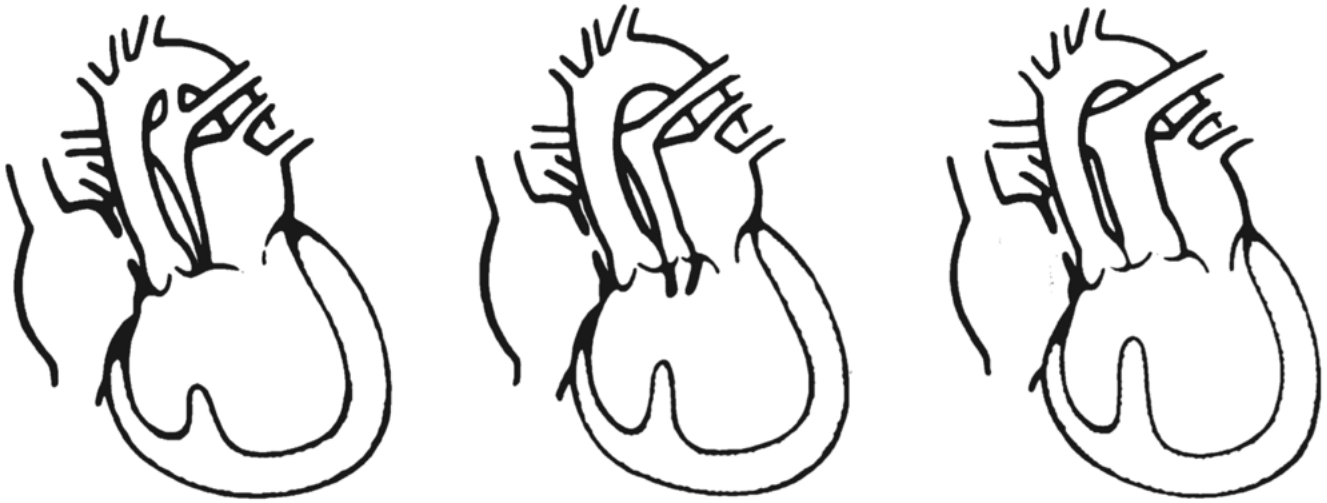
I(b)
Pulmonary hypoplasia, small ventricular septal defect

I(c)
No pulmonary hypoplasia, large ventricular septal defect

Mavroudis C, et al. Pediatric Cardiac Surgery. 4th ed.

Classification of Tricuspid Atresia

Tricuspid atresia with D-transposition (17% - 27%)



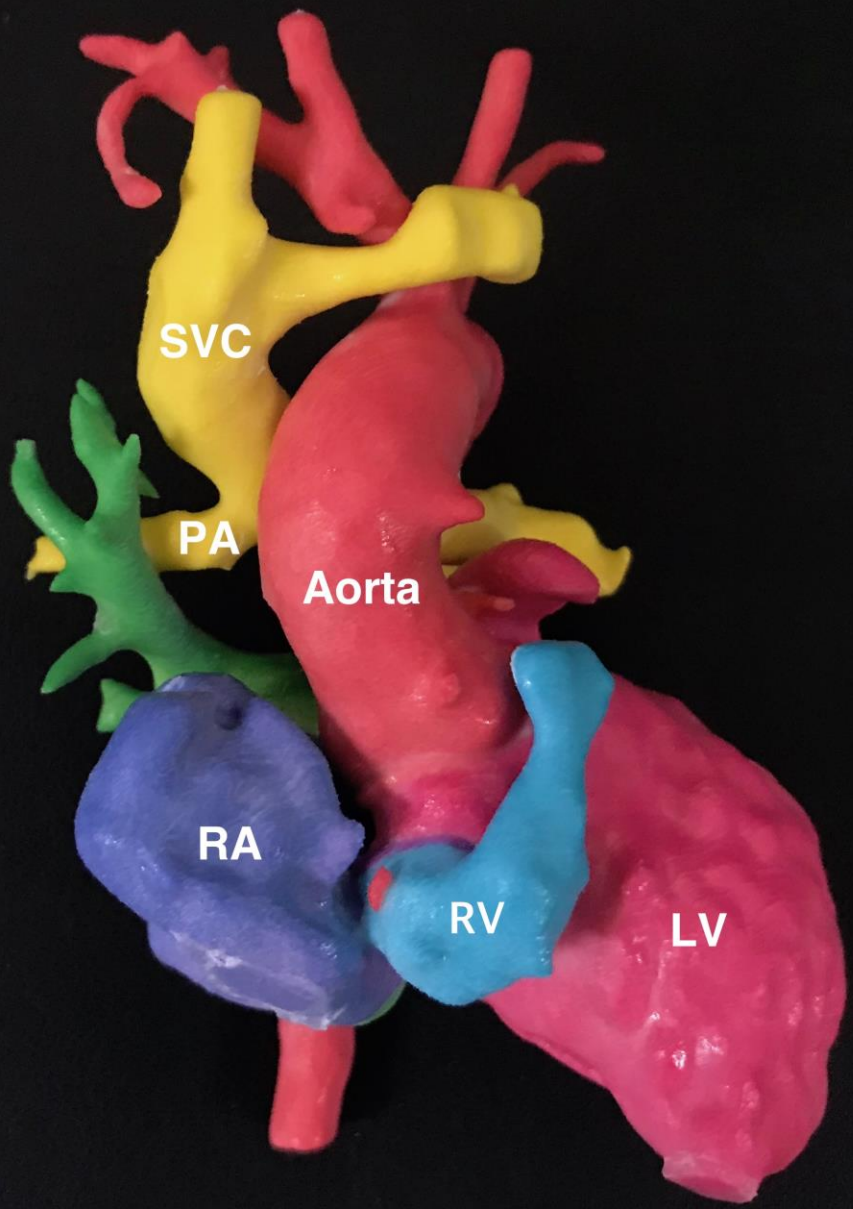
II(a)
Pulmonary atresia

II(b)
Pulmonary or subpulmonary stenosis

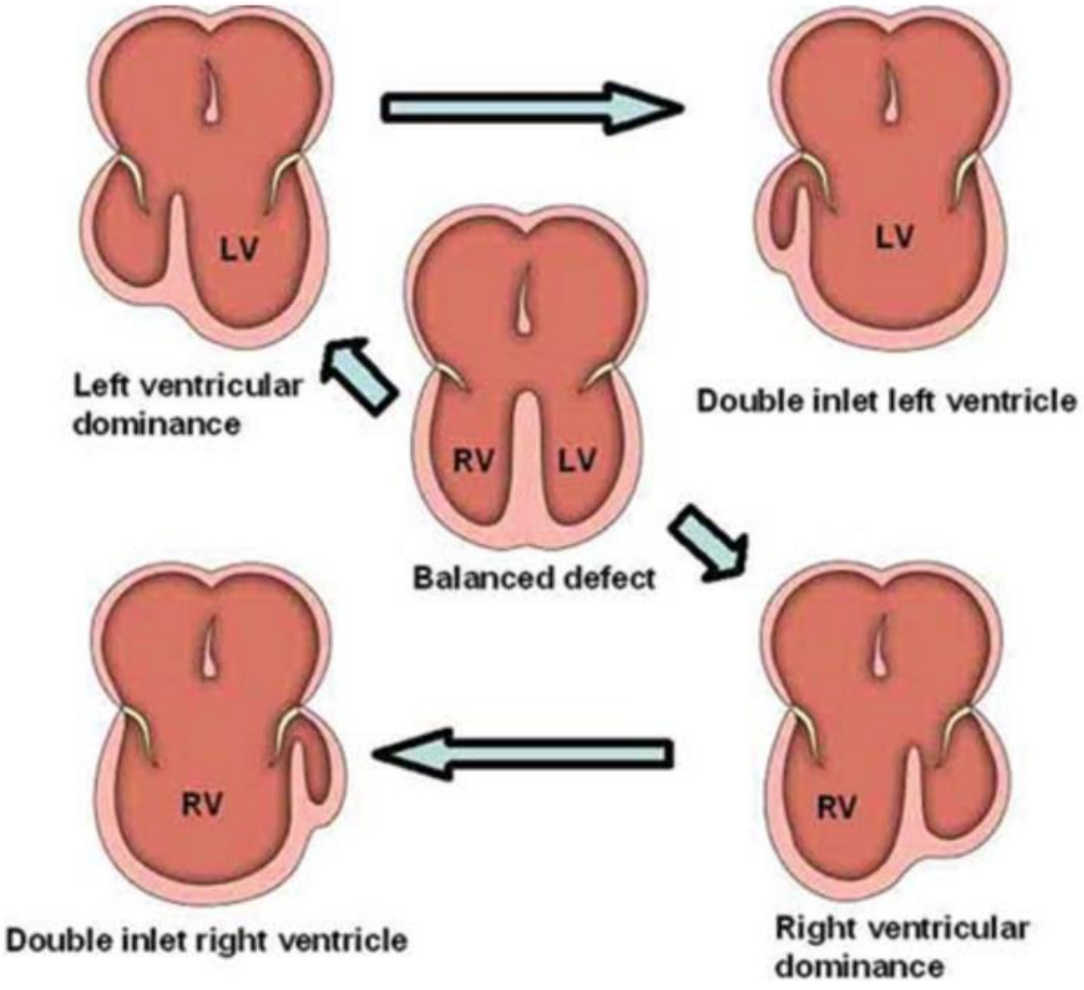
II(c)
Large pulmonary artery

Mavroudis C, et al. Pediatric Cardiac Surgery. 4th ed.

3D Printed Model of Tricuspid Atresia, Type 1b
S/P Bidirectional Cavopulmonary Anastomosis

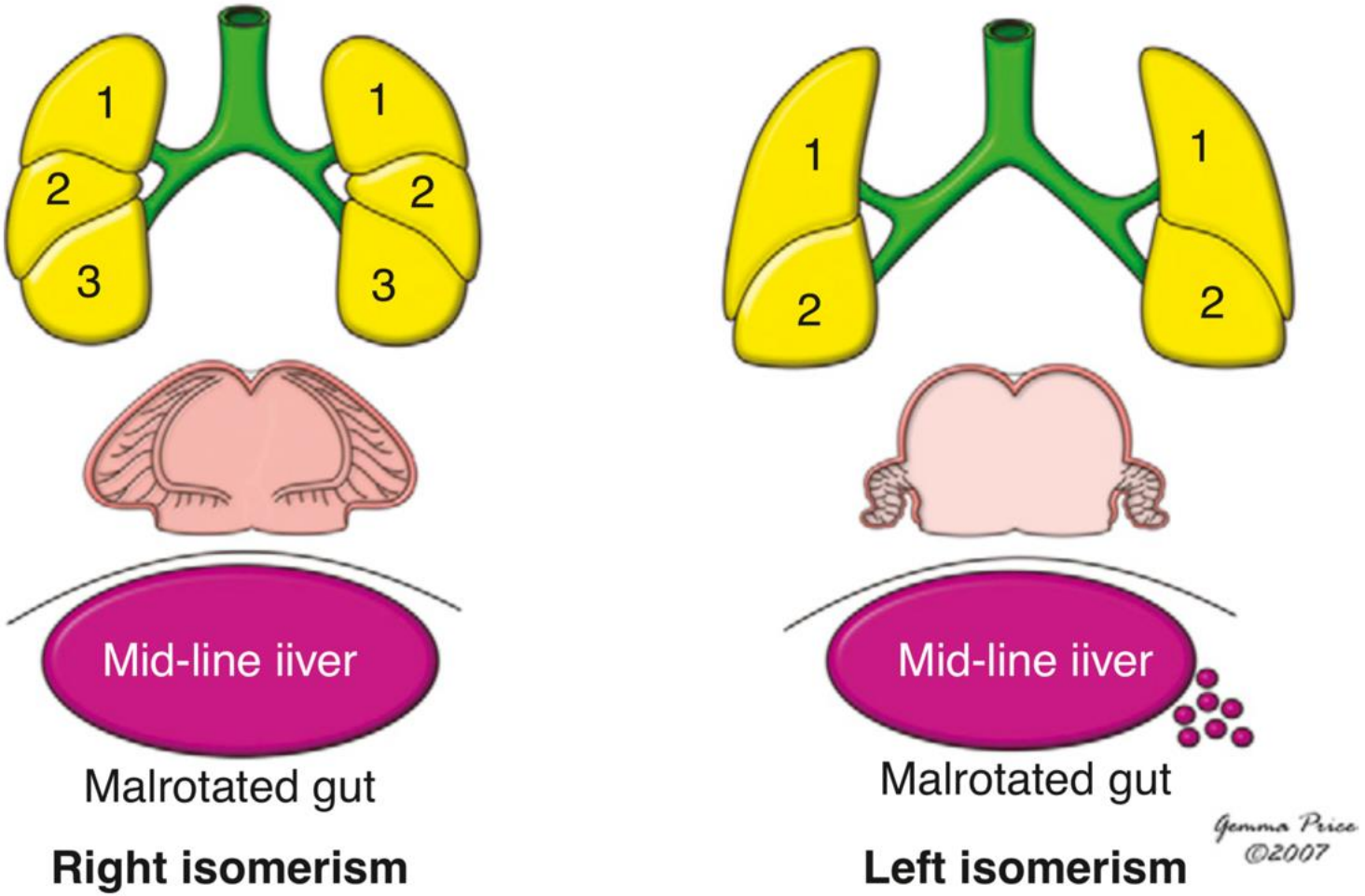


Unbalanced Atrioventricular Canal Defect



Cardiol Young 2006;16 Suppl 3:43-51

Heterotaxia Syndrome (Isomerism of the Atrial Appendages)



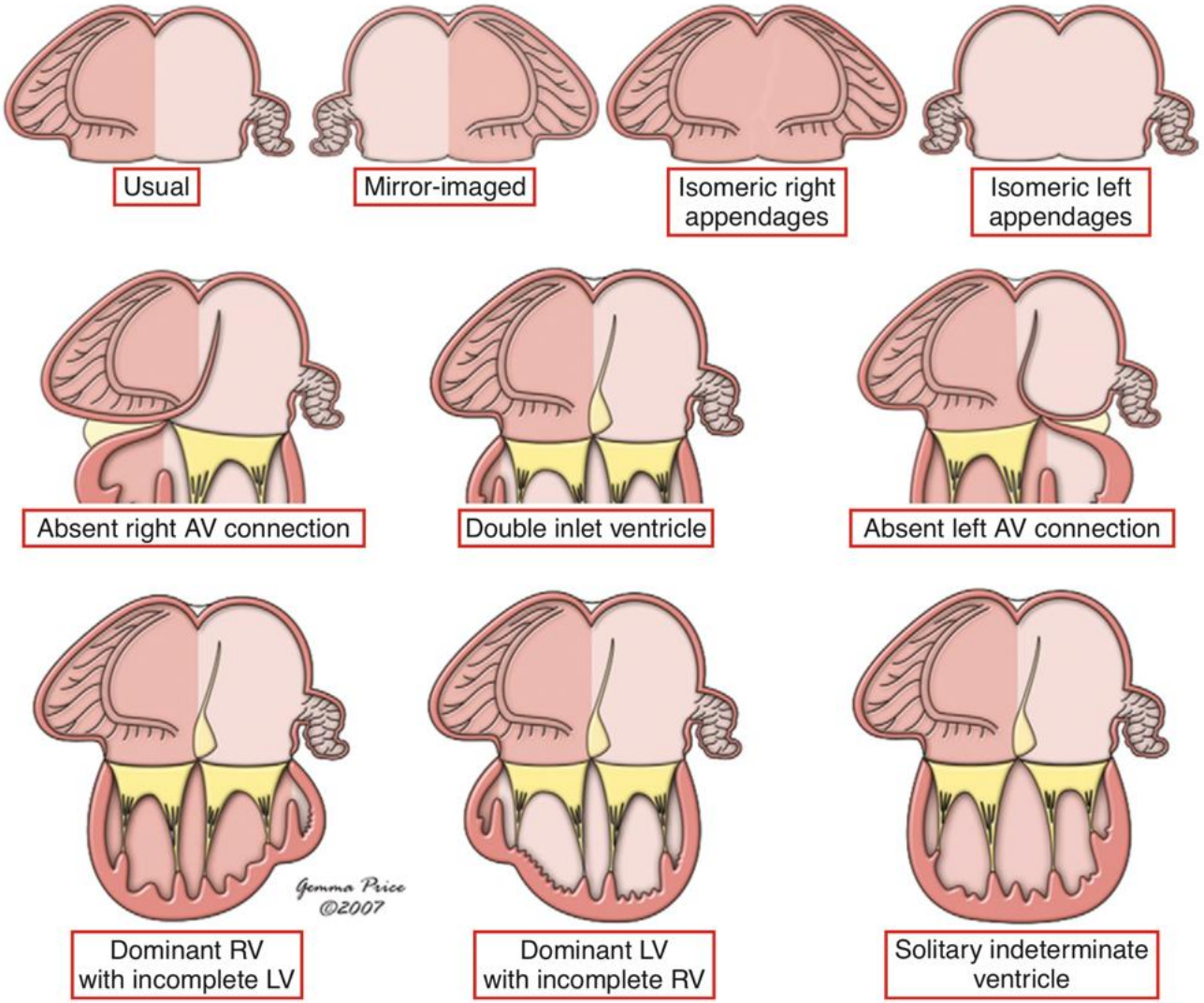
Anderson RH, et al. Paediatric Cardiology. 3rd ed.

Congenital Heart Surgery Nomenclature and Database Project: Single Ventricle

Single ventricle, Heterotaxia syndrome, **DORV, CAVC, Asplenia**
Single ventricle, Heterotaxia syndrome, **DORV, CAVC, Polysplenia**
Single ventricle, Heterotaxia syndrome, **Single LV**
Single ventricle, Heterotaxia syndrome, **Other**

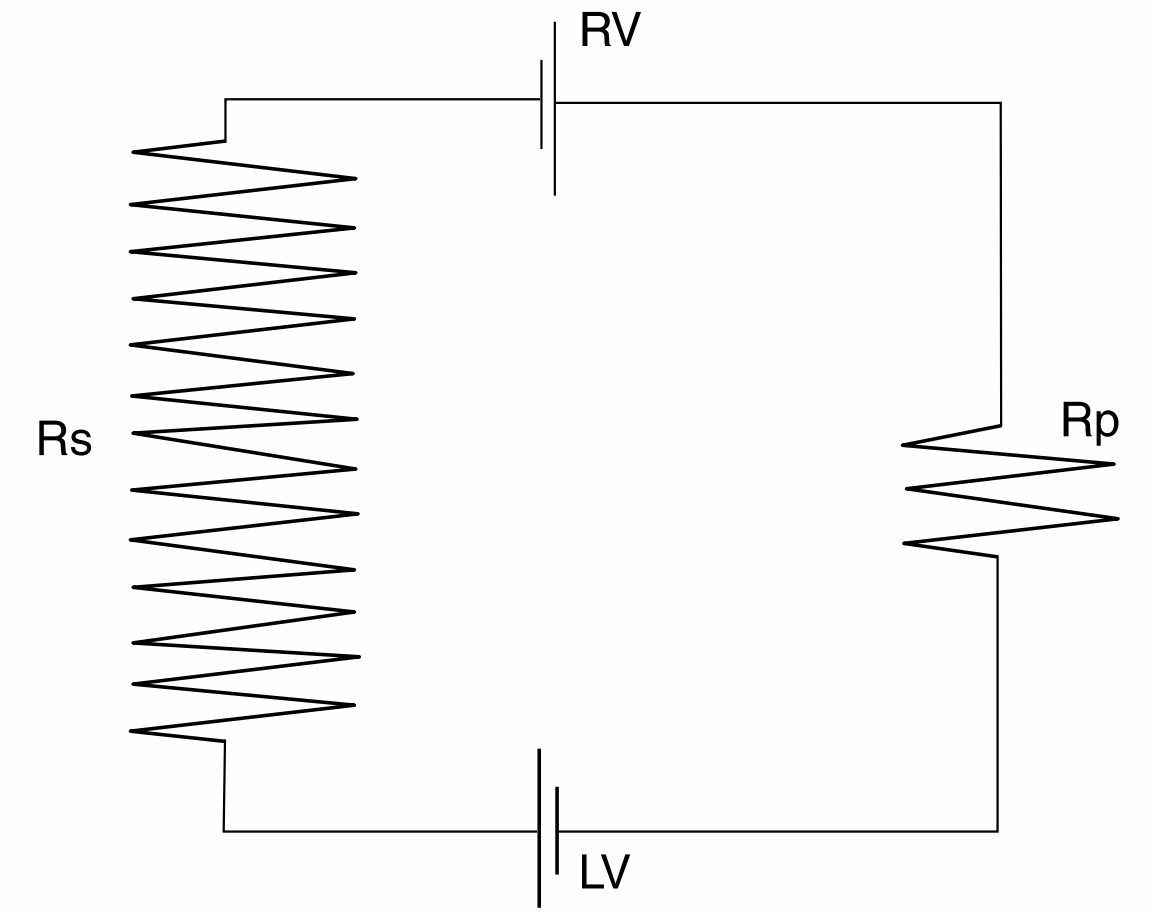
Ann Thorac Surg 2000;69:S197-204

Segmental Combinations Producing a Univentricular Atrioventricular Connection



Anderson RH, et al. Paediatric Cardiology. 3rd ed.

Normal Heart
A Serial Circuit



Cardiol Young 2003;13:316-22

Normal Heart

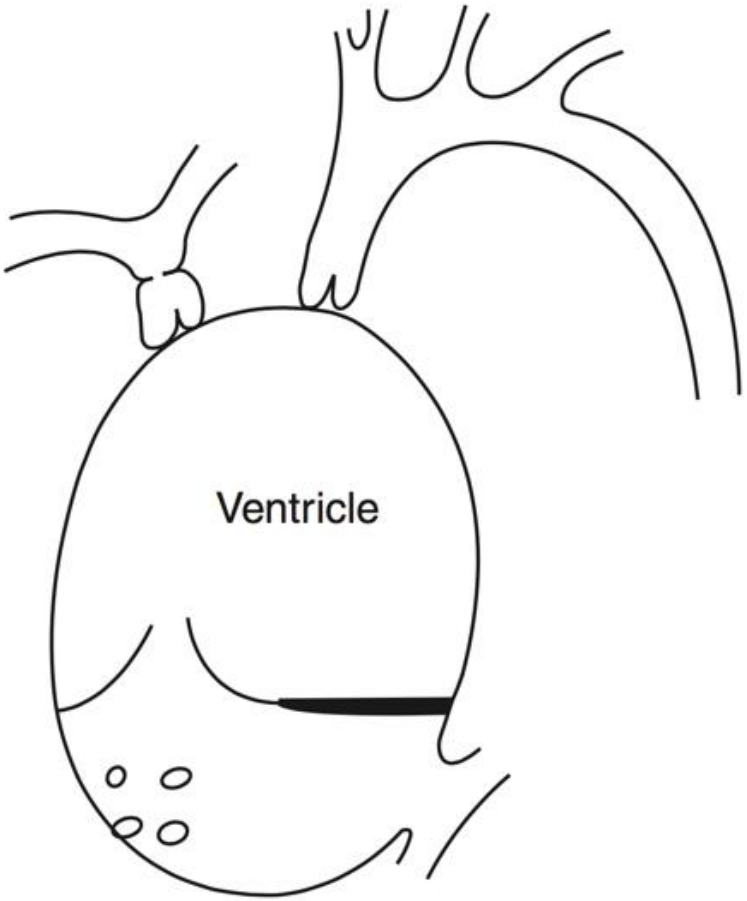
- Serial systemic and pulmonary circulations
- Different BP and O_2 saturation in each part
- Cardiac output = Qp = Qs (Qp/Qs = 1)

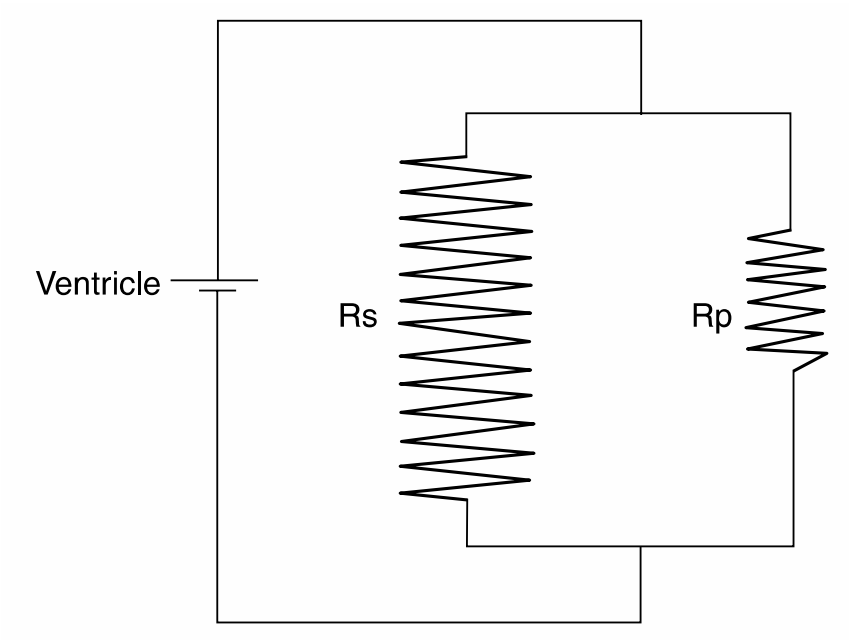
BP: blood pressure

Qp: pulmonary blood flow

Qs: systemic blood flow

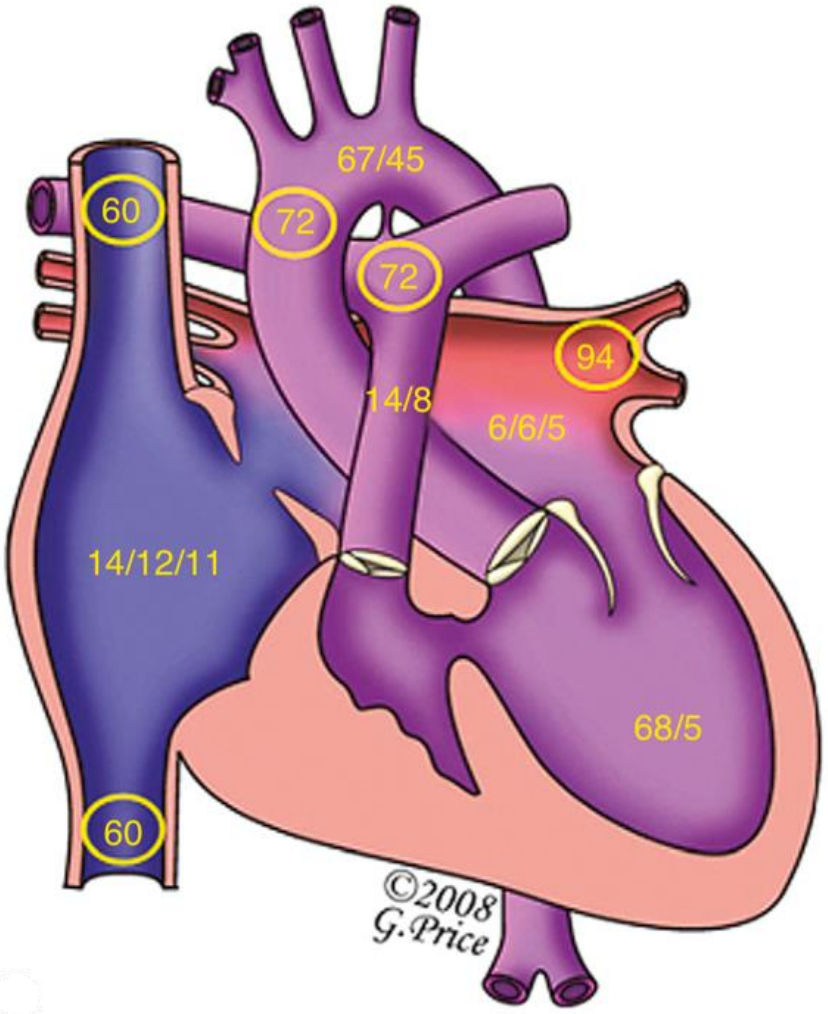
Single Ventricle
Parallel Circuits





Cardiol Young 2003;13:316-22

Parallel Circulations in Tricuspid Atresia



Anderson RH, et al. Paediatric Cardiology. 3rd ed.

Hemodynamics of Single Ventricle (1)

- Parallel systemic and pulmonary circulations
- BP in each part of the circulation is the same, if there is no obstruction to systemic and pulmonary outflow.
- O_2 saturation is the same in the aorta and the pulmonary arteries, if complete mixing of desaturated and saturated blood occurs within the single ventricle.

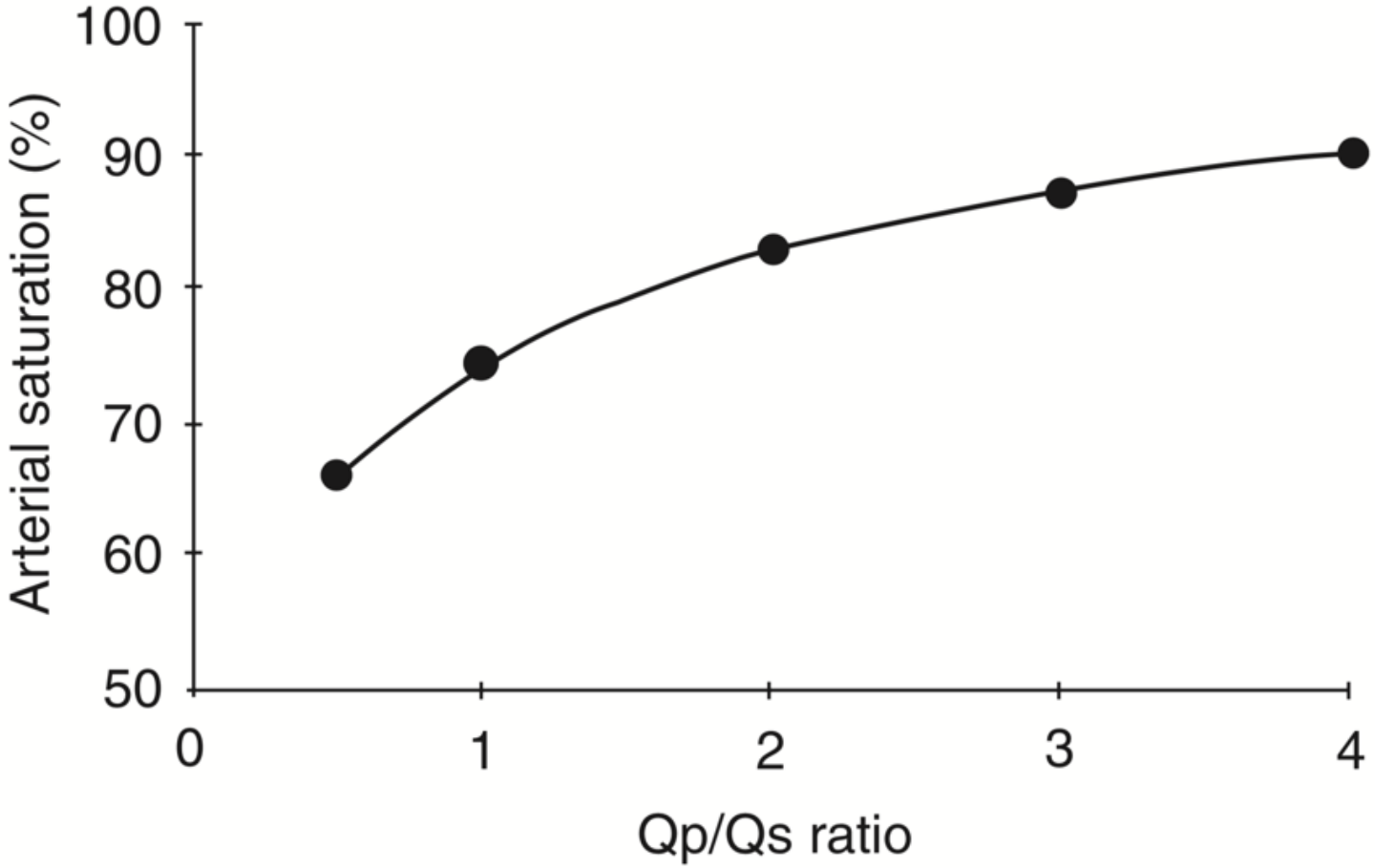
Hemodynamics of Single Ventricle (2)

- Cardiac output = Qp + Qs
- Qp/Qs = (BP/Rp)/(BP/Rs) = Rs/Rp
- Arterial O_2 saturation is determined by the ratio between the pulmonary blood flow and the systemic blood flow (Qp/Qs).

Rp: pulmonary vascular resistance

Rs: systemic vascular resistance

O_2 Saturation in Single Ventricle



Cardiol Young 2003;13:316-22

“Balanced” Single Ventricle

- Qp = Qs
- Needs natural obstruction to pulmonary blood flow
- Arterial O_2 saturation of approximately 80%
- Volume overloaded (2 × normal cardiac output)

Clinical Presentaion

- Determined by Qp/Qs and associated cardiac lesions
- Cyanosis (inadequate Qp)
- Congestive heart failure (excessive Qp)
- Asymptomatic with mild cyanosis (Qp = Qs)

Goal of Surgery for Single Ventricle

- Separation of systemic and pulmonary circulations, with the single ventricle connected to the systemic circulation (creation of serial systemic and pulmonary circulations)

- Best achieved by optimizing compliance of the single ventricle as well as by minimizing the total resistance between the systemic veins and the ventricular chamber

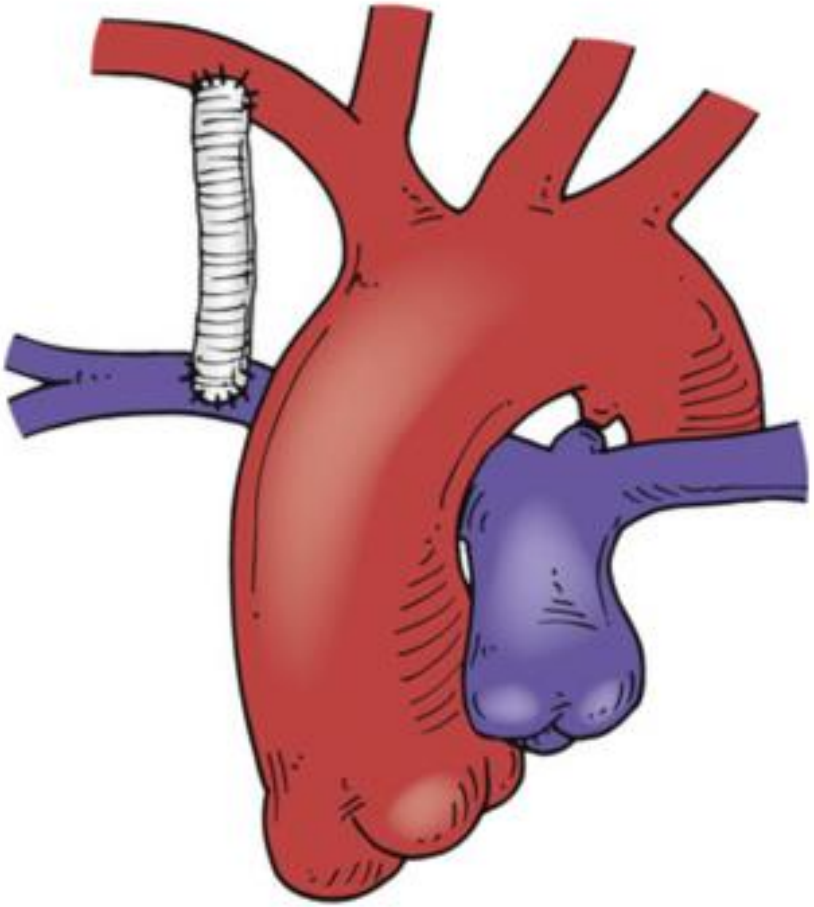
Three-Stage Surgical Management of Single Ventricle

1. First-stage palliation
2. Bidirectional cavopulmonary anastomosis
3. Fontan operation

First-Stage Palliation

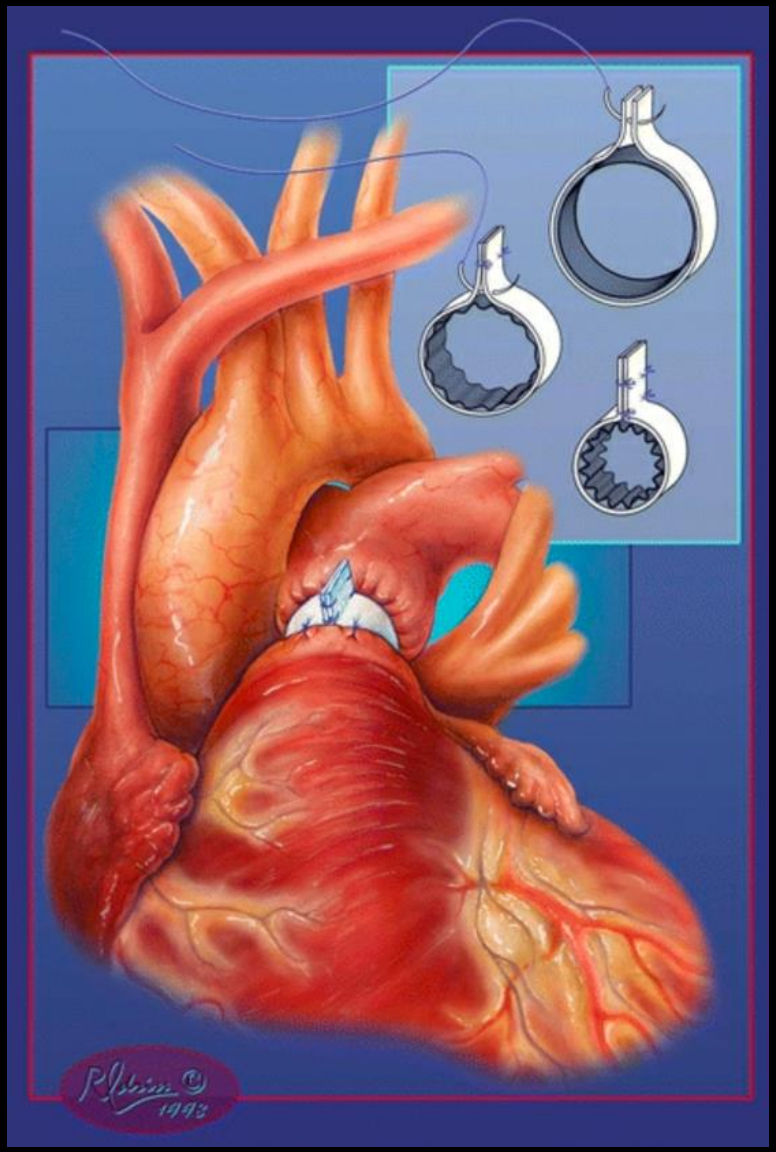
- Goals
  - ✓ Balanced systemic and pulmonary blood flow (Qp/Qs = 1)
  - ✓ Unobstructed mixing at the atrial level
  - ✓ Unobstructed systemic cardiac output
- Performed during neonatal or early infantile period
- The choice of procedure is determined to achieve the above-mentioned goals.

Modified Blalock-Taussig Shunt



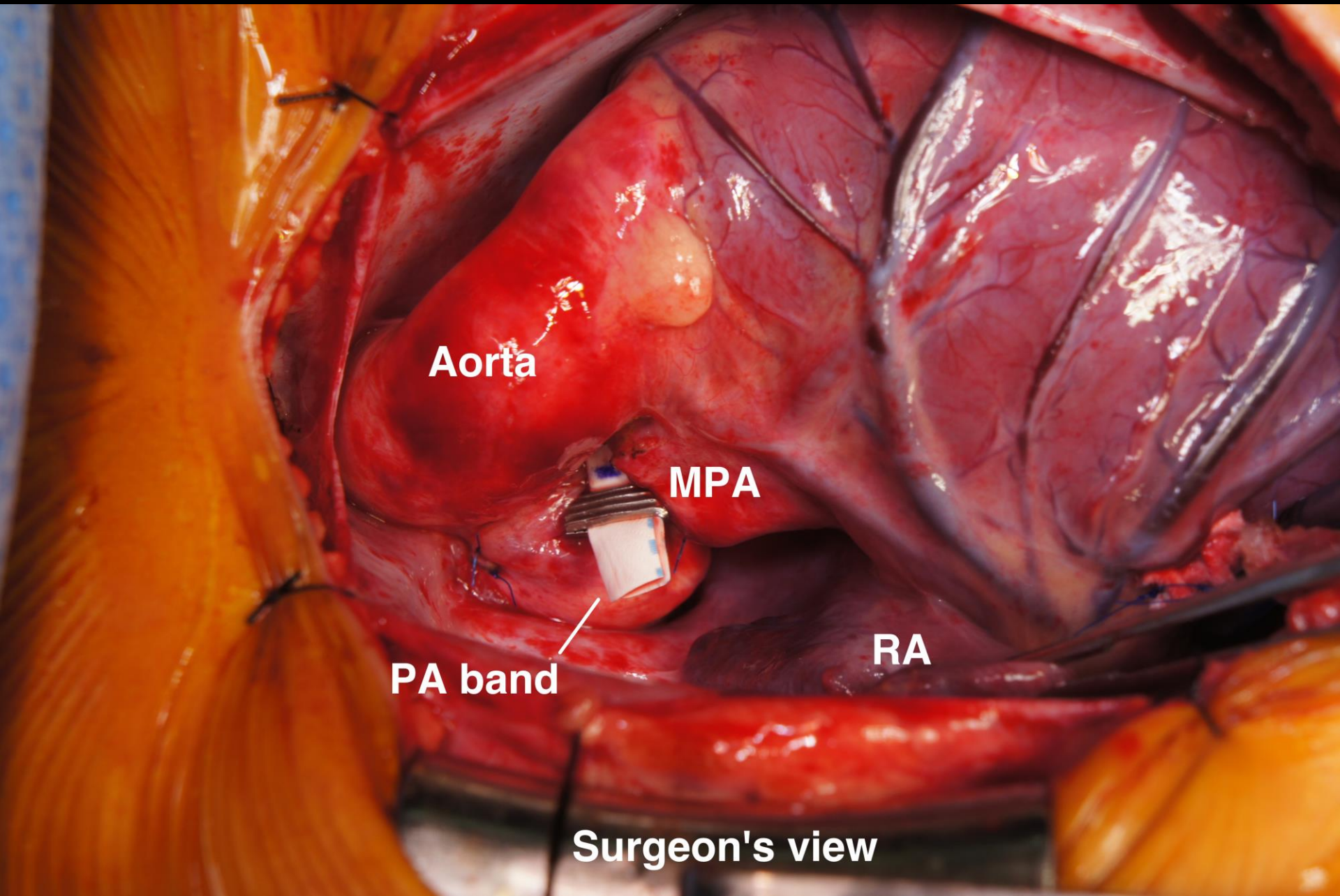
Ungerleider RM, et al. Critical Heart Disease in Infants and Children. 3rd ed.

Pulmonary Artery Banding



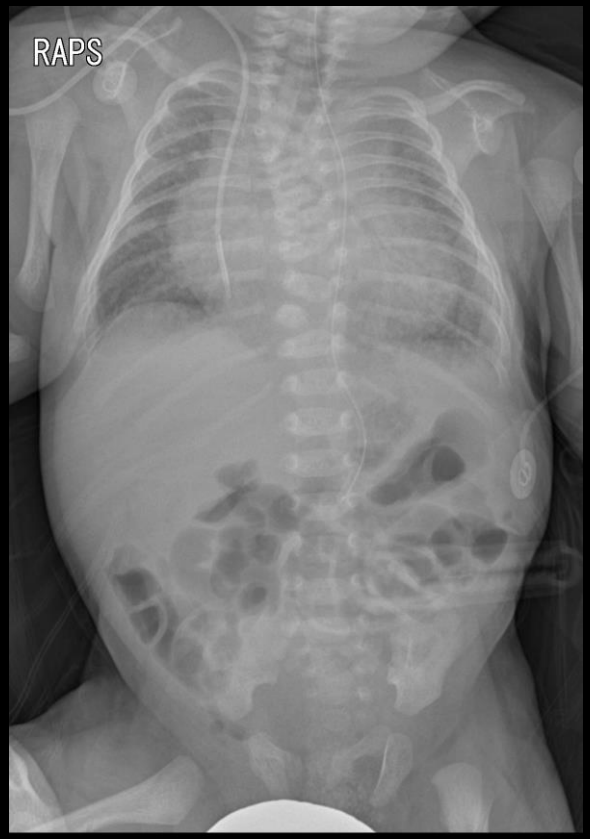
Mavroudis C, et al. Atlas of Pediatric Cardiac Surgery.

Pulmonary Artery Banding for {S,L,L} DILV

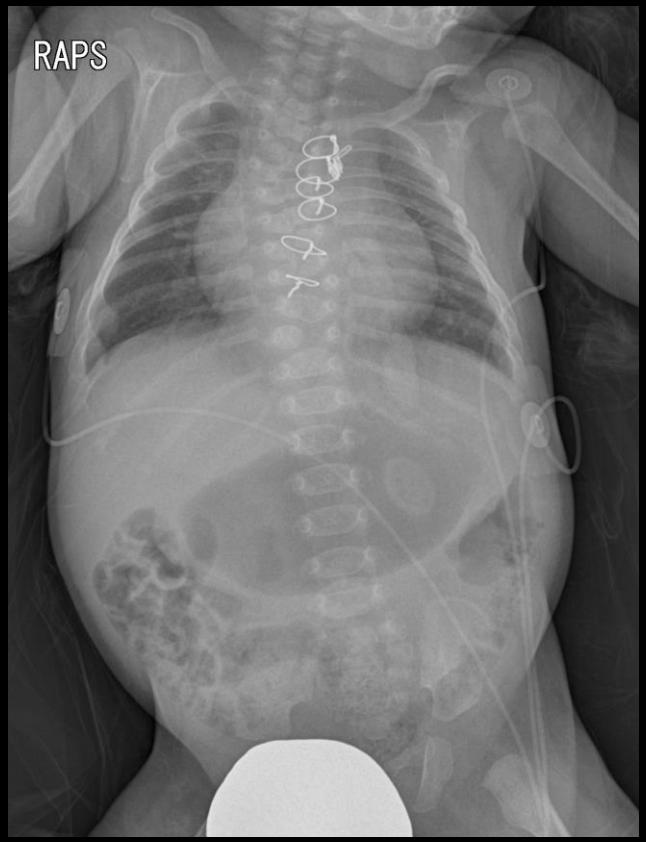


A Neonate with Mitral Atresia and DORV

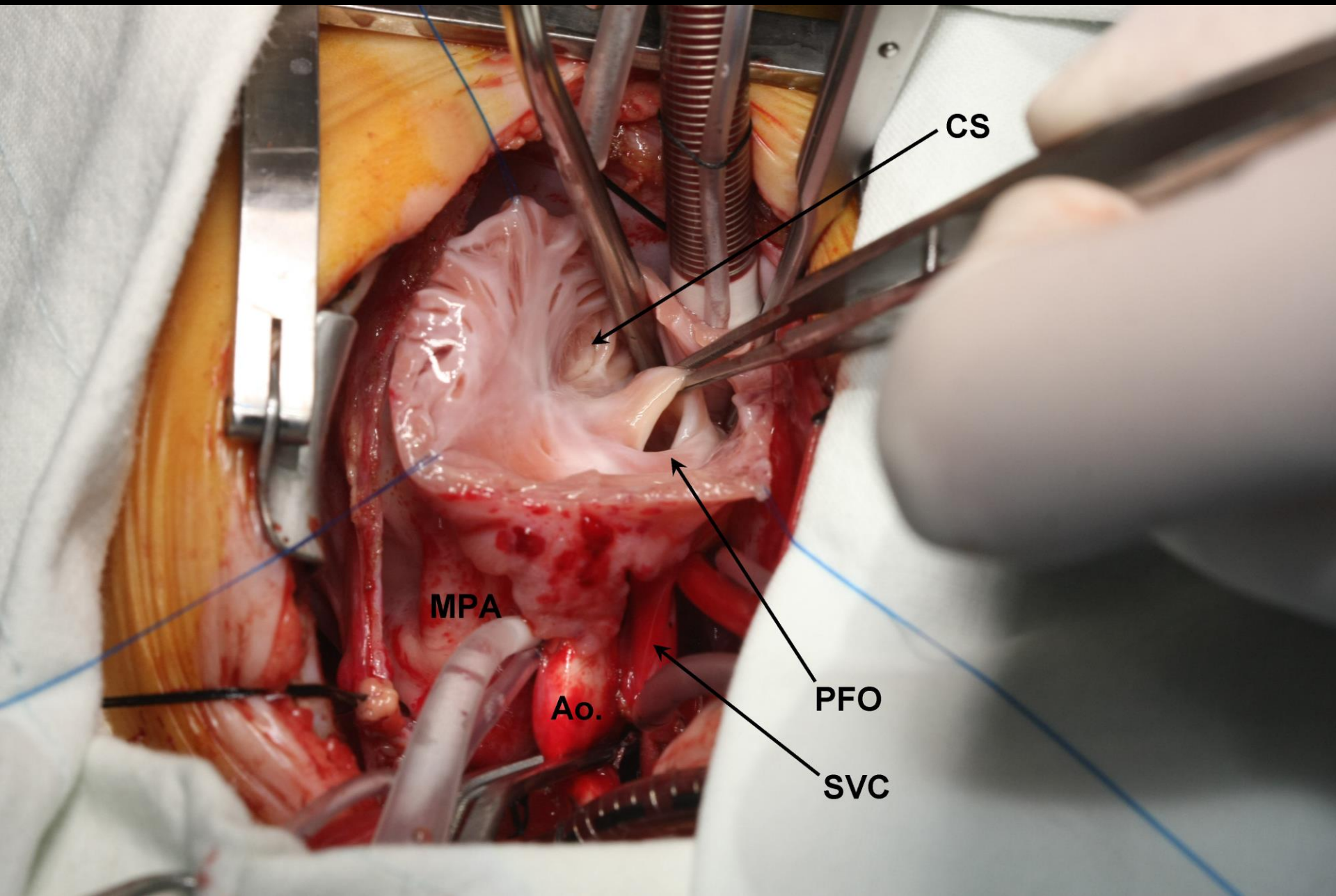
Pre-PA banding
SpO_2 around 95% at room air



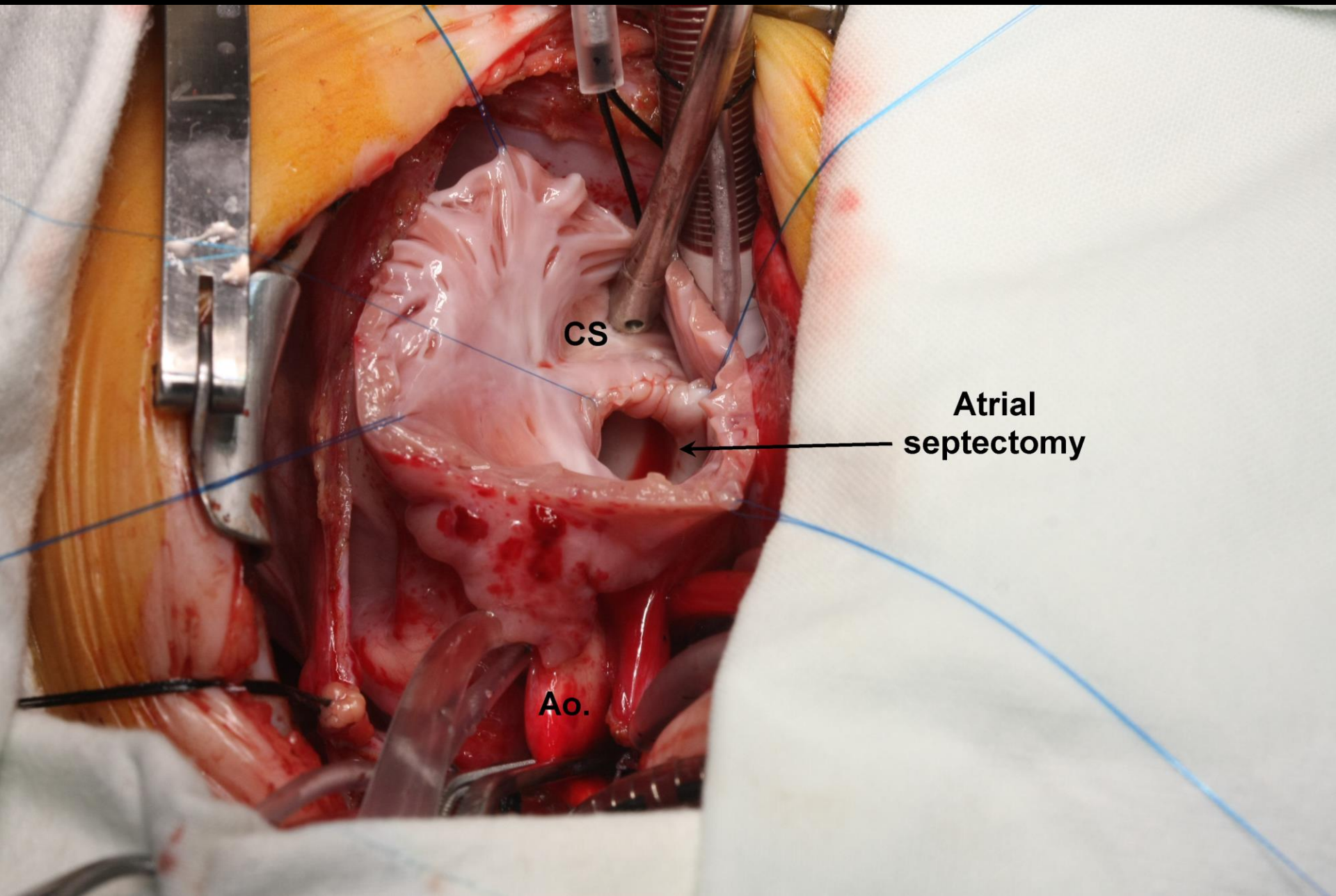
Post-PA banding
SpO_2 around 75% at room air



Tricuspid Atresia with a Restrictive ASD



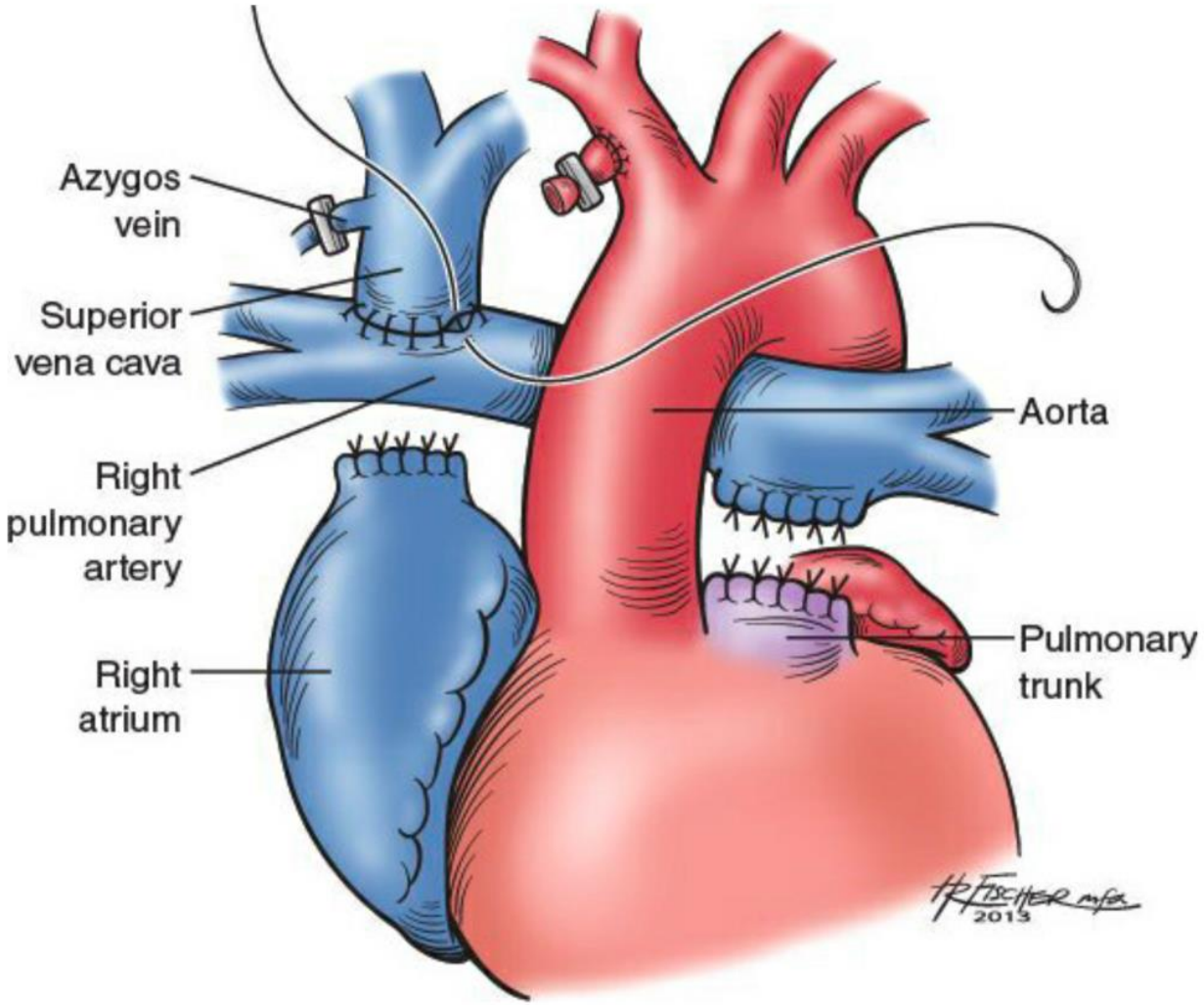
Atrial Septectomy for Tricuspid Atresia with a Restrictive ASD



Bidirectional Cavopulmonary Anastomosis

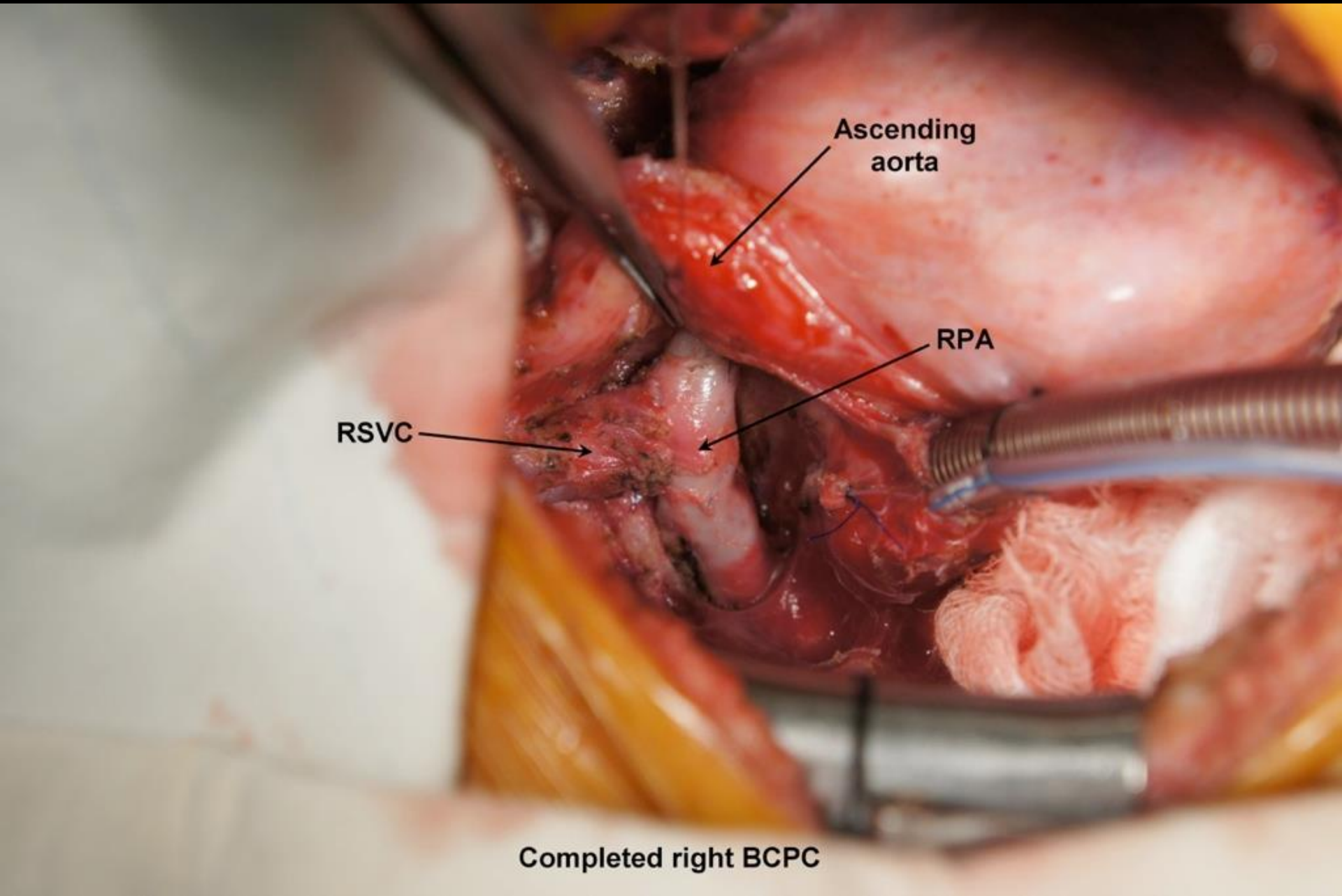
- Goals (Benefits)
  - ✓ Improvement in efficiency of gas exchange
  - ✓ Reduction in volume overload of the single ventricle
- Diversion of SVC blood into the pulmonary arteries
- Usually performed at 3-6 months of age

Bidirectional Cavopulmonary Anastomosis

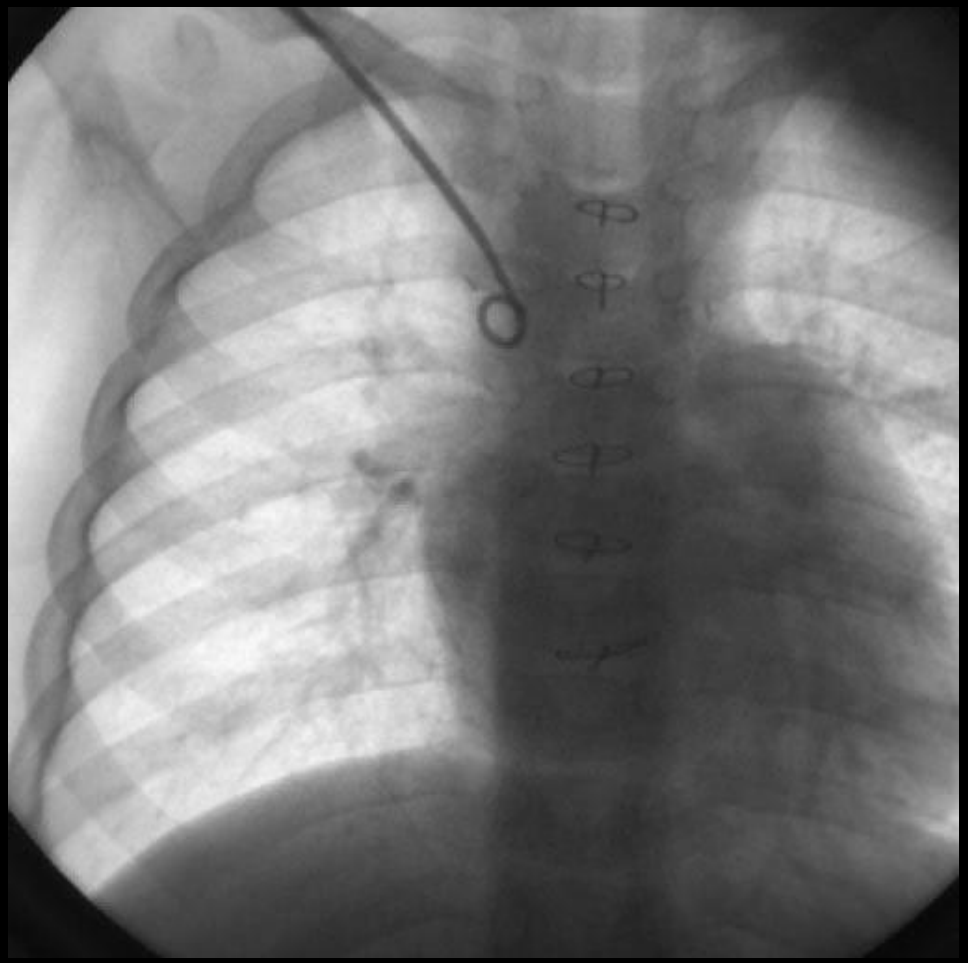


Kaiser LR, et al. Mastery of Cardiothoracic Surgery. 3rd ed.

Bidirectional Cavopulmonary Anastomosis



S/P Bidirectional Cavopulmonary Anastomosis



Fontan Operation

- Total cavopulmonary connection
- Separation of systemic and pulmonary circulations
- Usually performed at 2-3 years of age

Thorax (1971), **26**, 240.

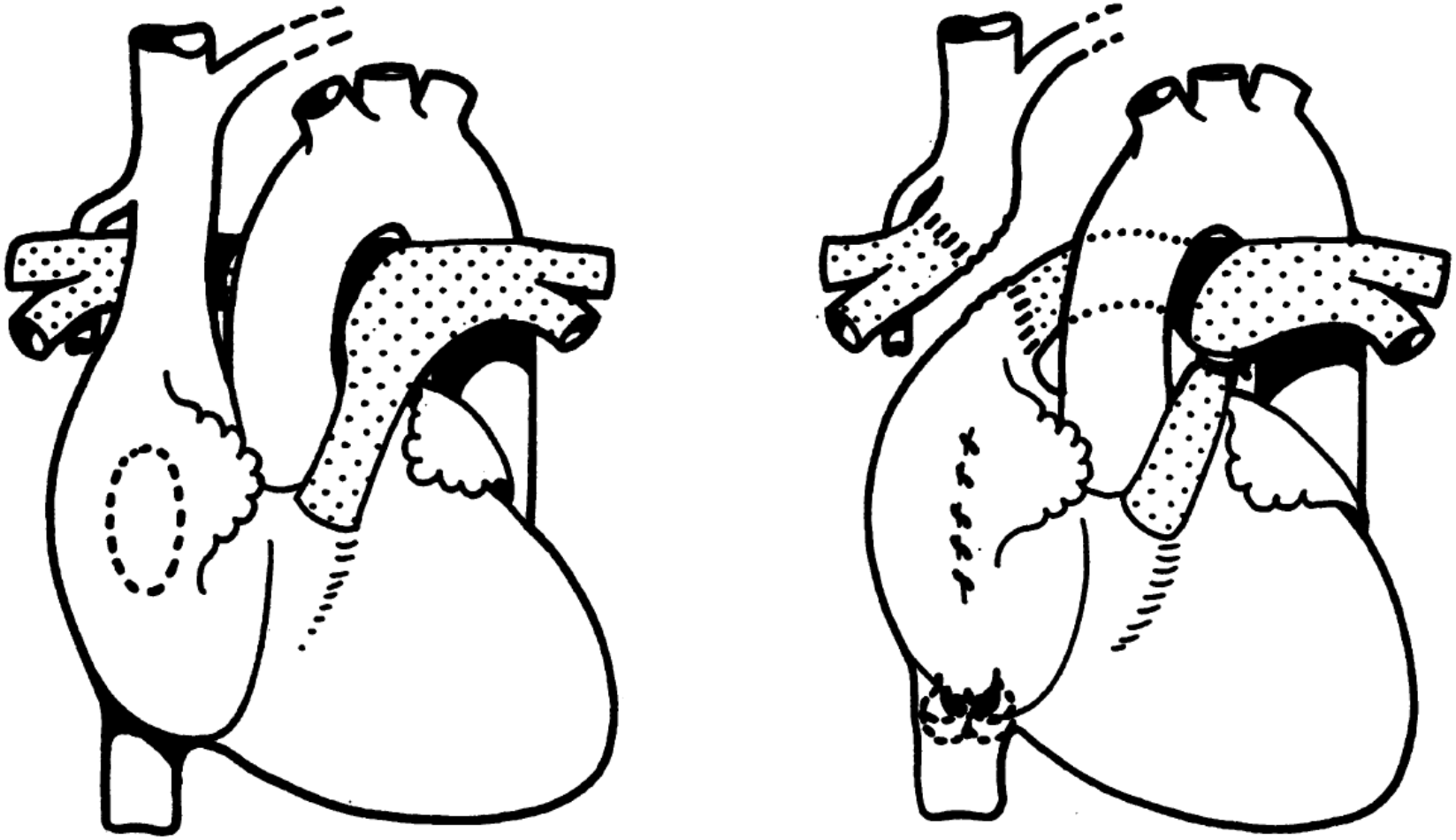
Surgical repair of tricuspid atresia

F. FONTAN and E. BAUDET

Centre de Cardiologie, Université de Bordeaux II, Hôpital du Tondu, Bordeaux, France

Surgical repair of tricuspid atresia has been carried out in three patients ; two of these operations have been successful. A new surgical procedure has been used which transmits the whole vena caval blood to the lungs, while only oxygenated blood returns to the left heart. The right atrium is, in this way, 'ventriclized', to direct the inferior vena caval blood to the left lung, the right pulmonary artery receiving the superior vena caval blood through a cava-pulmonary anastomosis. This technique depends on the size of the pulmonary arteries, which must be large enough and at sufficiently low pressure to allow a cava-pulmonary anastomosis. The indications for this procedure apply only to children sufficiently well developed. Younger children or those whose pulmonary arteries are too small should be treated by palliative surgical procedures.

Original Fontan Operation



Thorax 1971;26:240-8

With Professor Francis Fontan

3rd Scientific Meeting of the World Society for Pediatric and Congenital Heart Surgery

June 23-26, 2011, Istanbul, Turkey

Francis Fontan, MD (1929-2018)

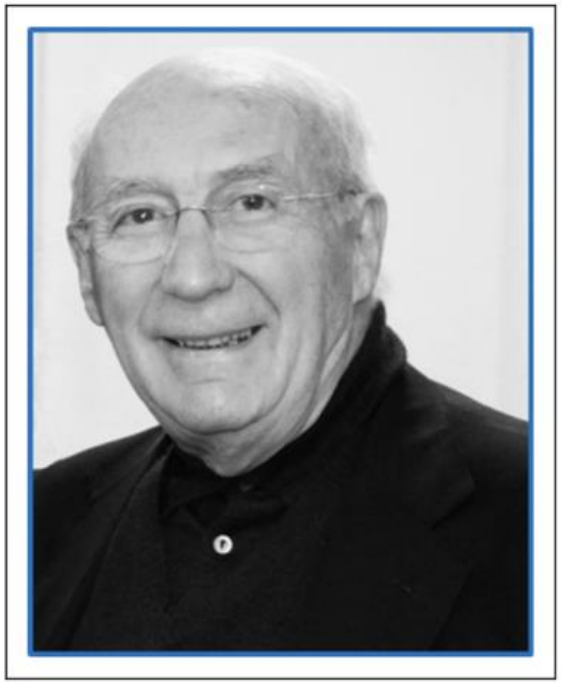
World Journal for Pediatric and
Congenital Heart Surgery
2018, Vol. 9(2) 129-130

DOI: 10.1177/2150135118759443
journals.sagepub.com/home/pch
SAGE

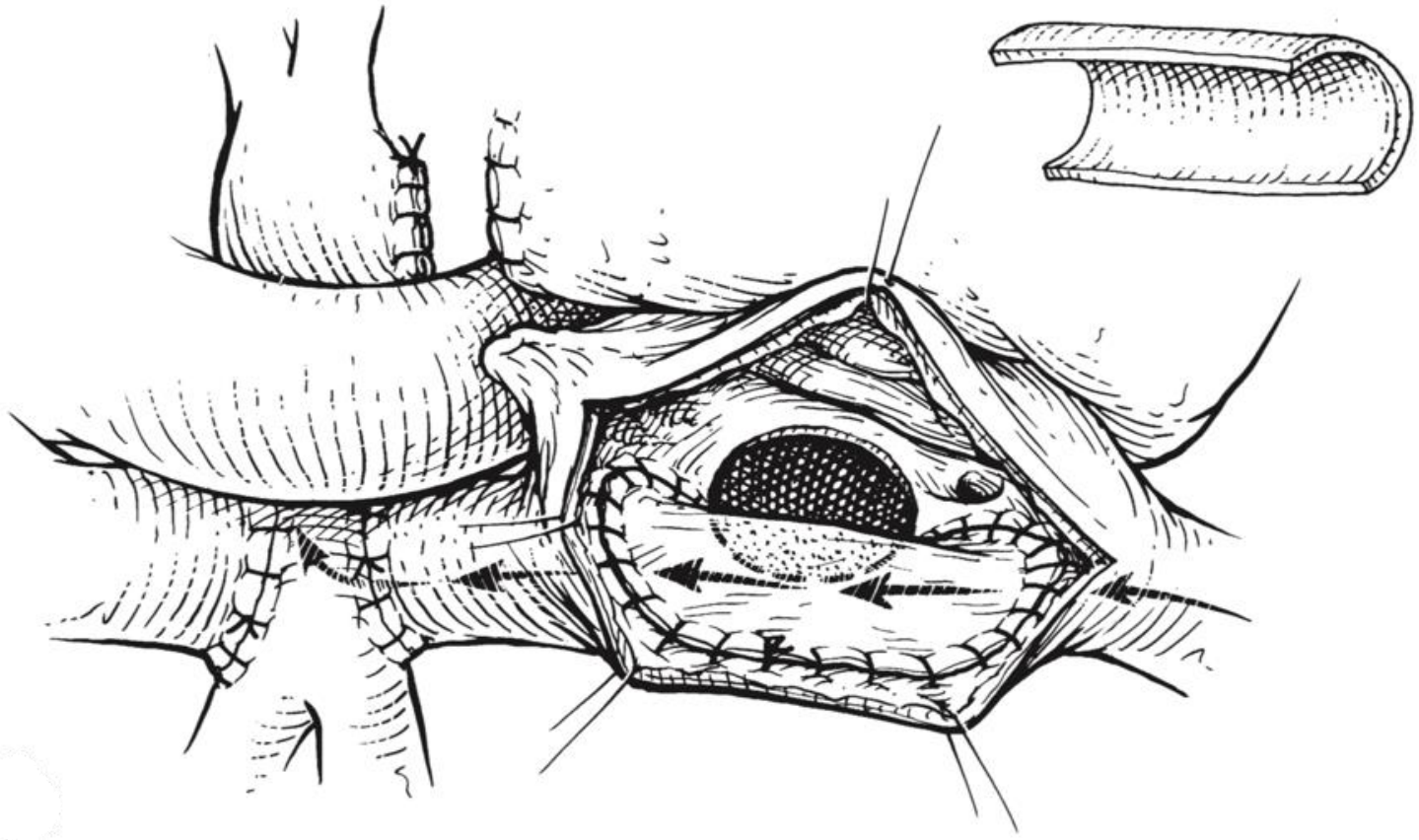
With the passing of Professor Francis Fontan on the 14th of January, this year, the community of congenital heart professionals lost one of its greatest leaders. He was a surgical innovator, a man of science, and a European visionary.

Fontan was born in the southwestern burgh of Ney, in a French province beside the Pyrénée foothills, called Bearn. He grew up in comfortable circumstances provided by his Tour de France, "King of the Mountains," cyclist father. The German occupation of Southern France did impact Fontan's early education at the local Lycée through loss of boarding facilities, which were appropriated to house troops, but he nonetheless achieved entry to the Faculté des Medicins in Bordeaux by 17 years of age.

After six years of medical studies and five of highly competitive "Internat des Hospitaux," Fontan embarked upon his chosen area of specialization in 1952, training in both surgery and cardiology in Bordeaux. This unusual path for an aspiring

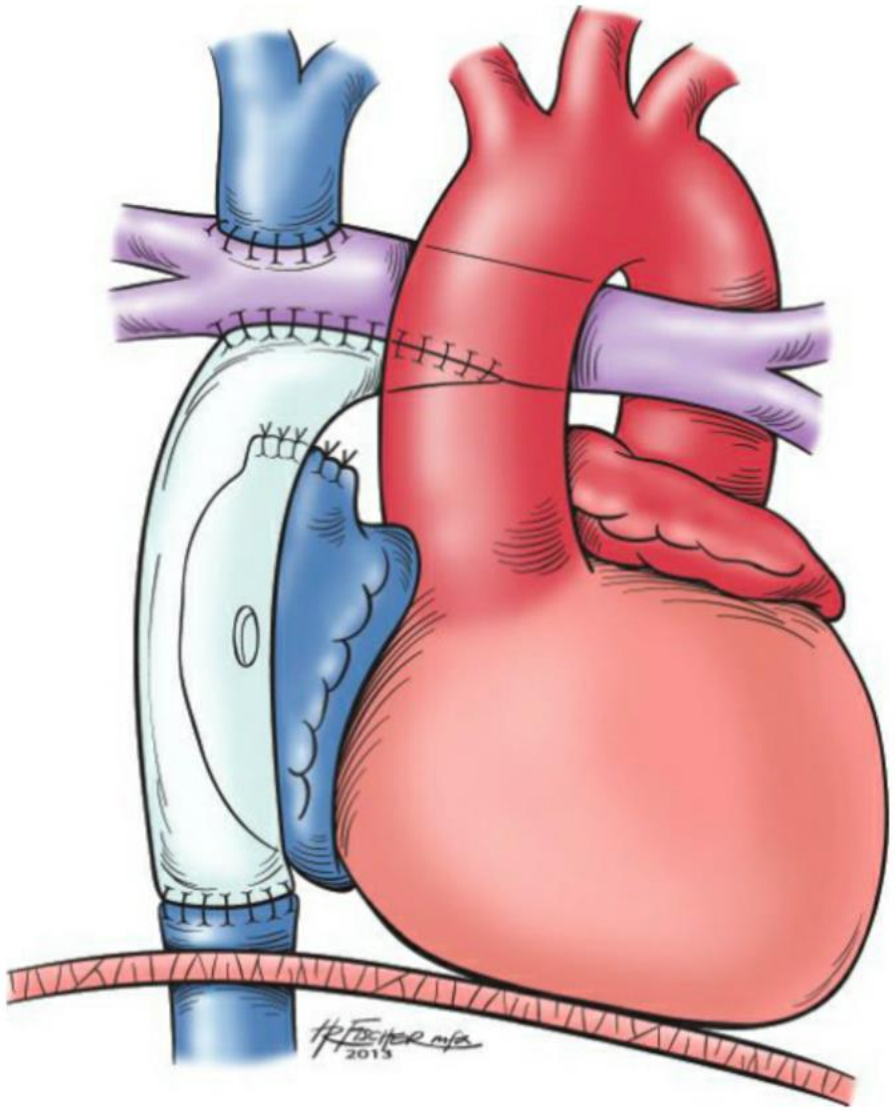


Lateral Tunnel Fontan Operation



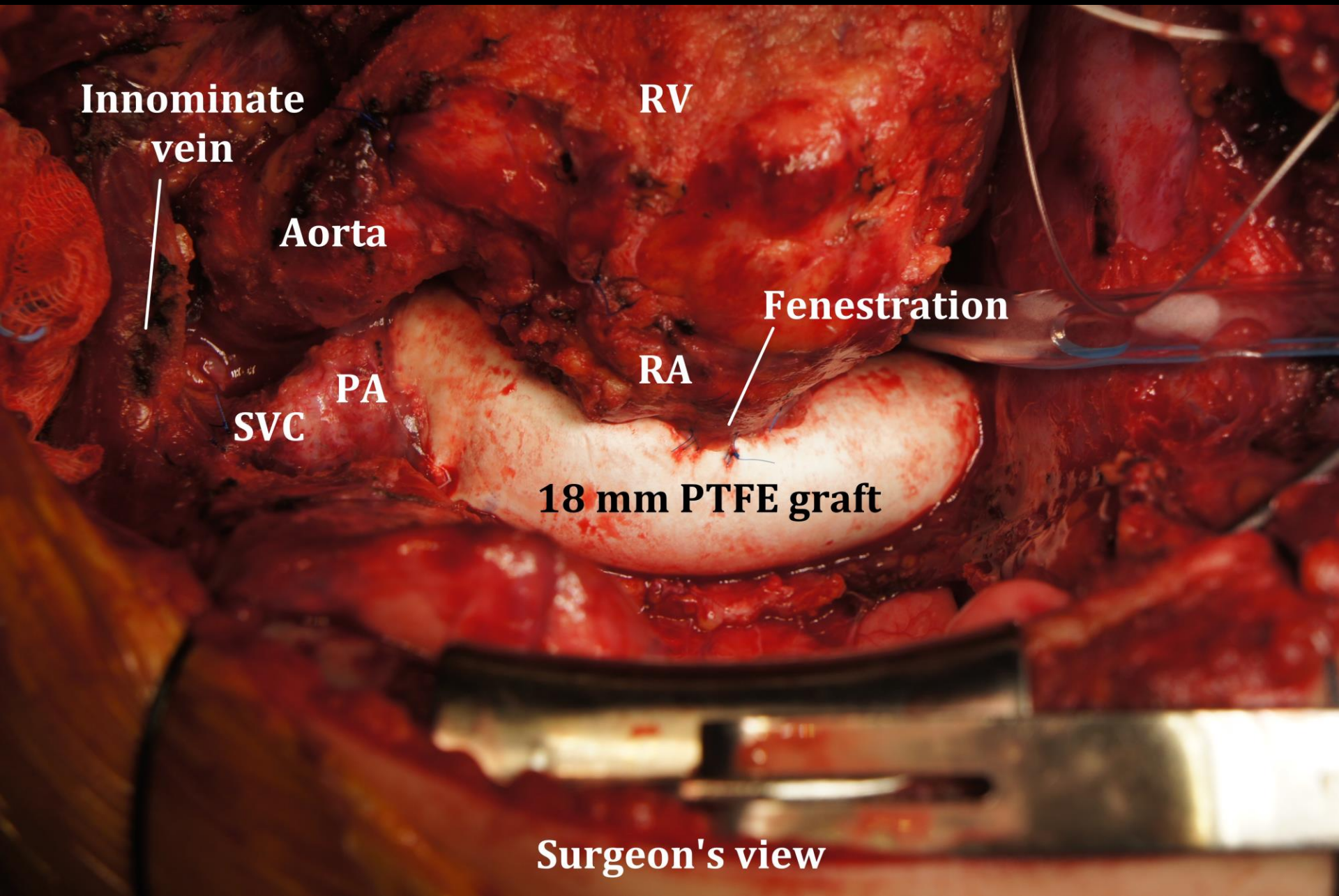
Kouchoukos NT, et al. Kirklin/Barratt-Boyes Cardiac Surgery. 4th ed.

Extracardiac Conduit Fontan Operation

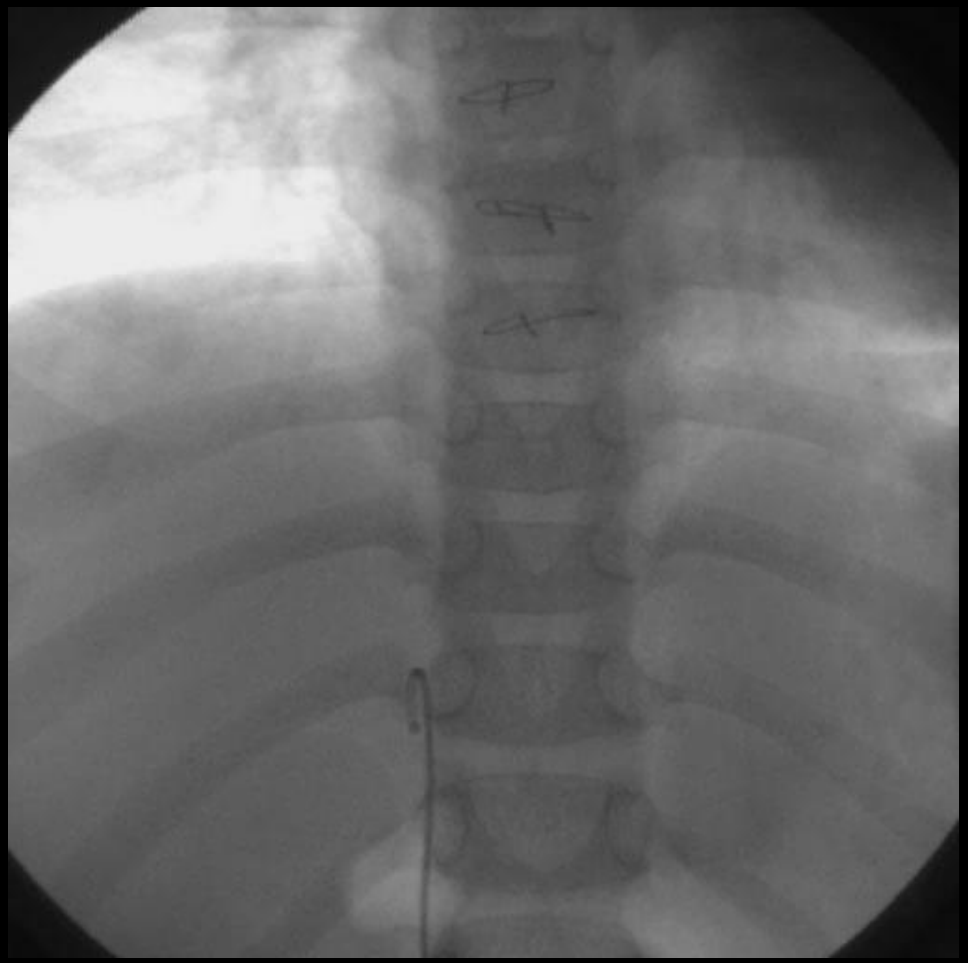


Kaiser LR, et al. Mastery of Cardiothoracic Surgery. 3rd ed.

Extracardiac Conduit Fontan Operation



S/P Extracardiac Conduit Fontan Operation



BOX 129-1 The "Ten Commandments" for Selection of Patients with Tricuspid Atresia for the Fontan Procedure

1. Minimum age 4 years
2. Sinus rhythm
3. Normal caval drainage
4. Right atrium of normal volume
5. Mean pulmonary artery pressure ≤15 mm Hg
6. Pulmonary arterial resistance <4 U/m^2
7. Pulmonary artery to aorta diameter ratio ≥0.75
8. Normal ventricular functions (ejection fraction >0.6)
9. Competent left atrioventricular valve
10. No impairing effects of previous shunts

Selke FW, et al. Sabiston & Spencer Surgery of the Chest. 9th ed.

Selection Criteria for Fontan Operation

- The pulmonary vasculature and ventricular function remains the most important selection criteria for successful outcome after the Fontan operation.

- Pulmonary vascular resistance < 4 WU·m^2
- Mean pulmonary artery pressure < 15-20 mmHg
- Ventricular end-diastolic pressure < 12-15 mmHg