제9차 전공의 연수교육
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Single Ventricle

Cheul Lee, MD

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

Contents

1. Classification
2. Pathophysiology
3. Surgical management
4. Outcomes of surgery

Terminology

- Single ventricle
- Functionally univentricular heart

- Broad category of hearts that lack two well developed ventricles

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.

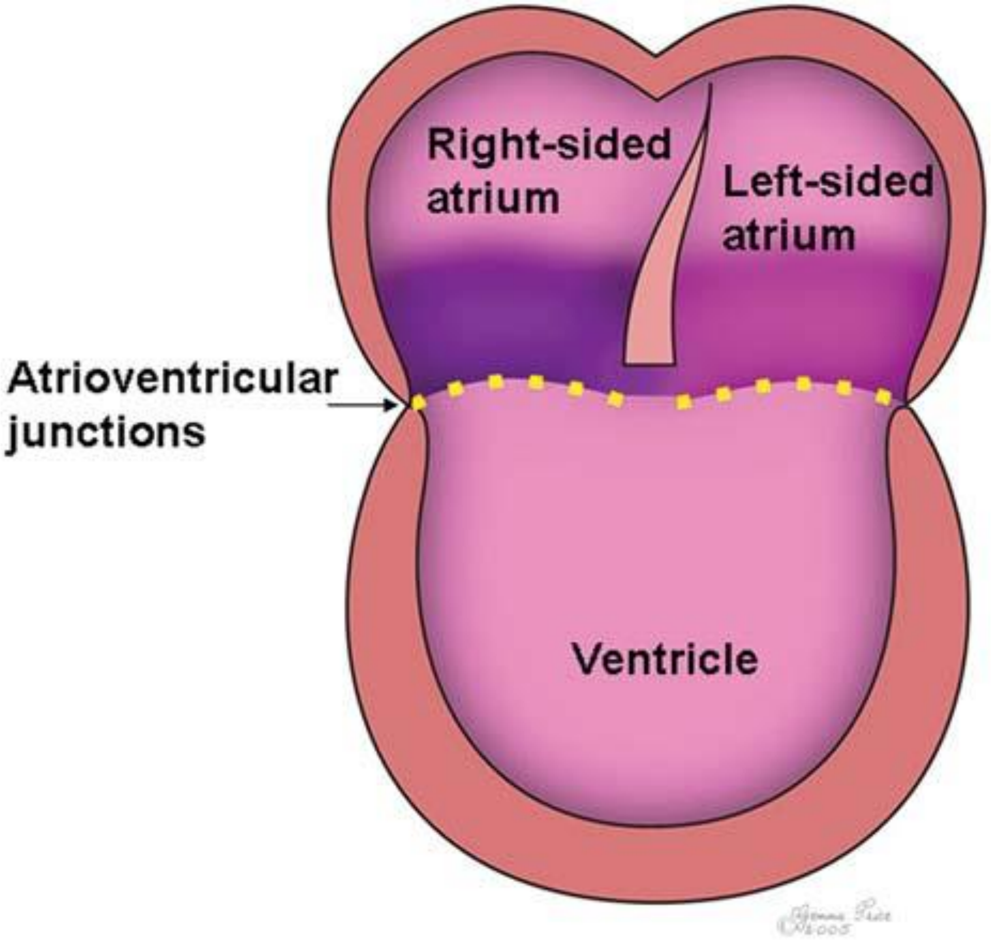
(Ann Thorac Surg 2000;69:S197–204)

Congenital Heart Surgery Nomenclature and Database Project: Single Ventricle

- Double inlet left ventricle (DILV)
- Double inlet right ventricle (DIRV)
- Mitral atresia
- Tricuspid atresia
- Unbalanced AV canal defect
- Heterotaxia syndrome
- Other

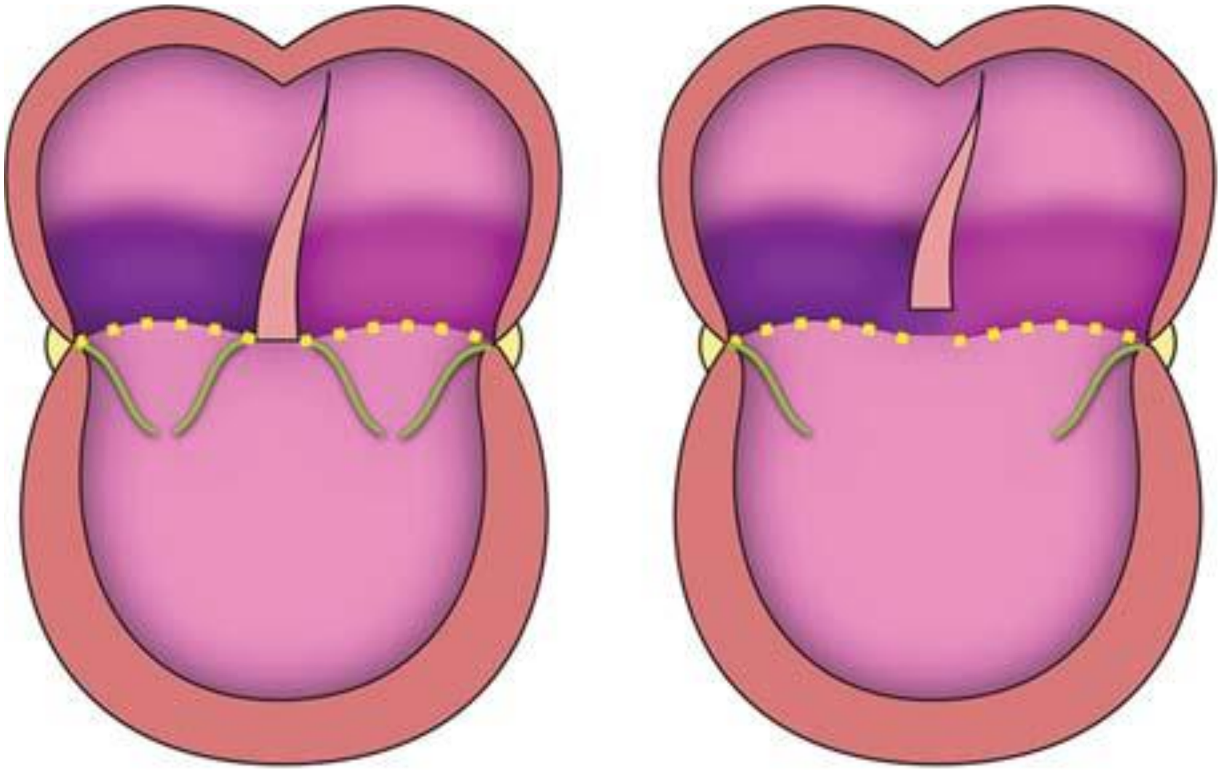
Ann Thorac Surg 2000;69:S197-204

Double Inlet Ventricle



Cardiol Young 2006;16 Suppl 1:22-6

Double Inlet Ventricle

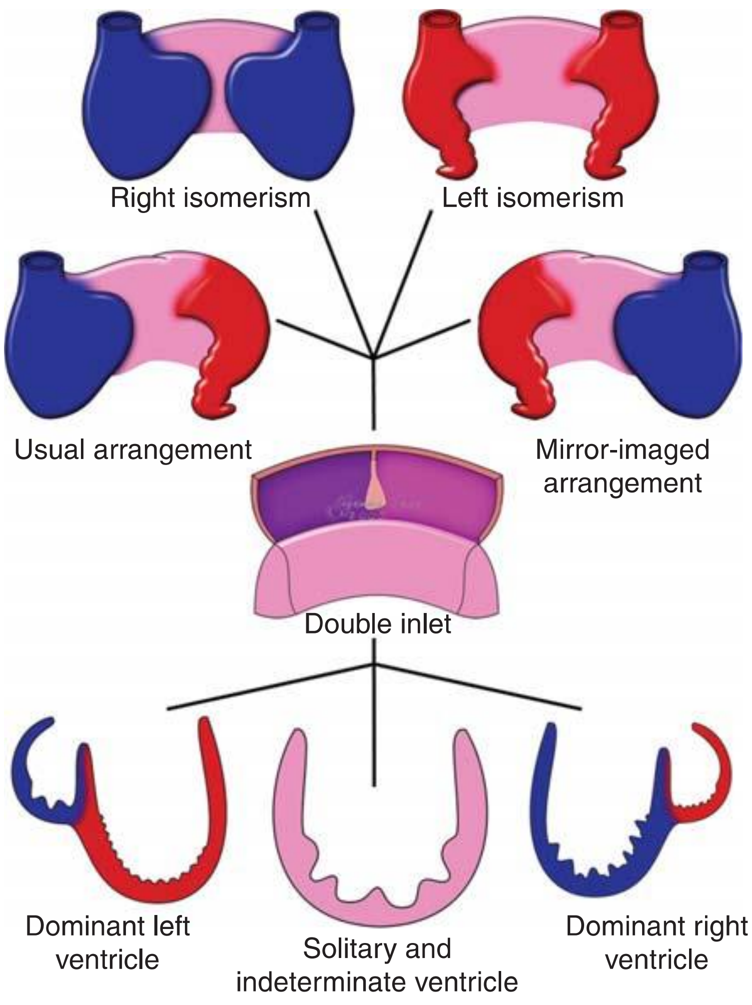


Two atrioventricular valves

Common atrioventricular valve

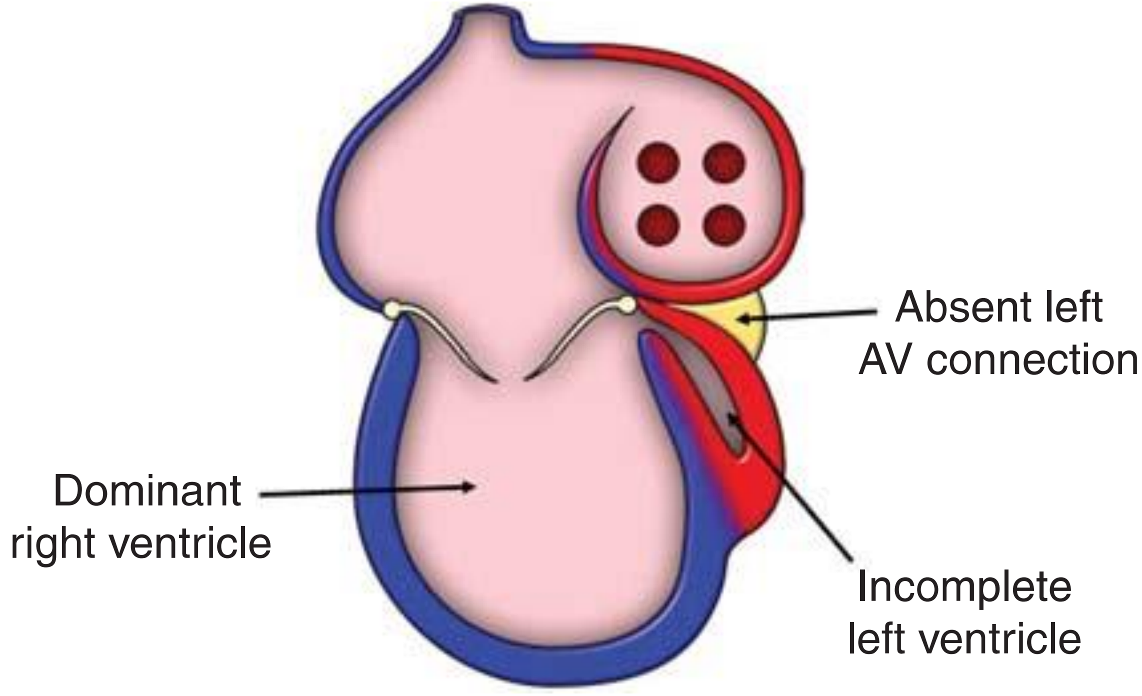
Cardiol Young 2006;16 Suppl 1:22-6

Double Inlet Ventricle



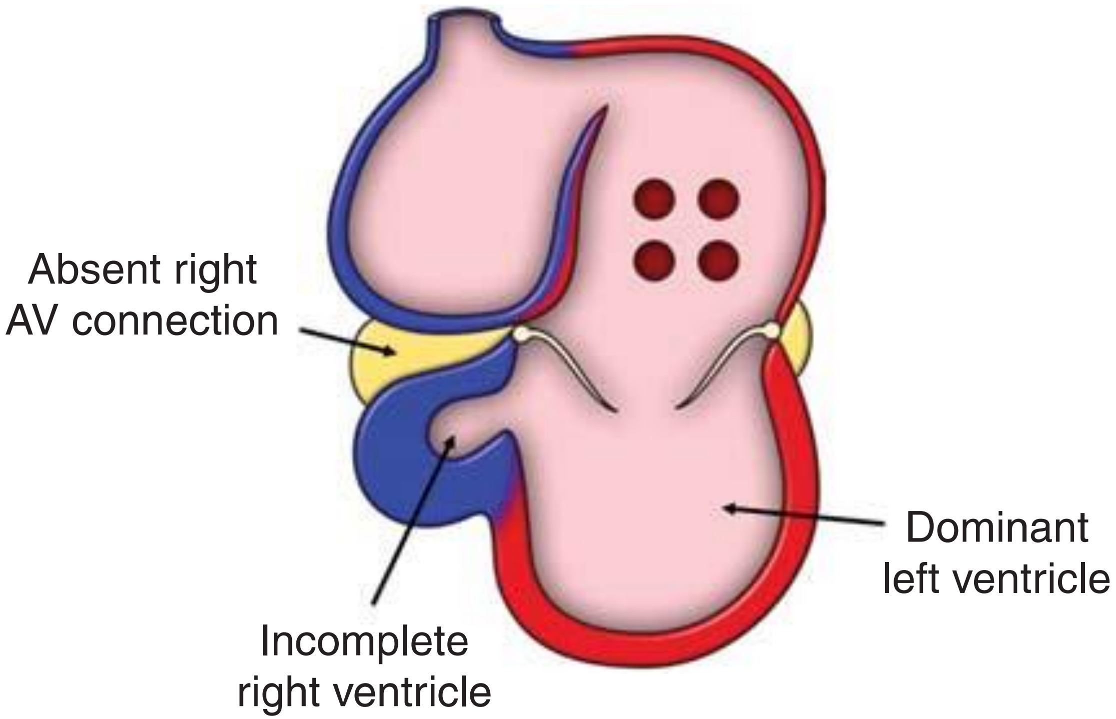
Cardiol Young 2006;16 Suppl 1:22-6

Mitral Atresia



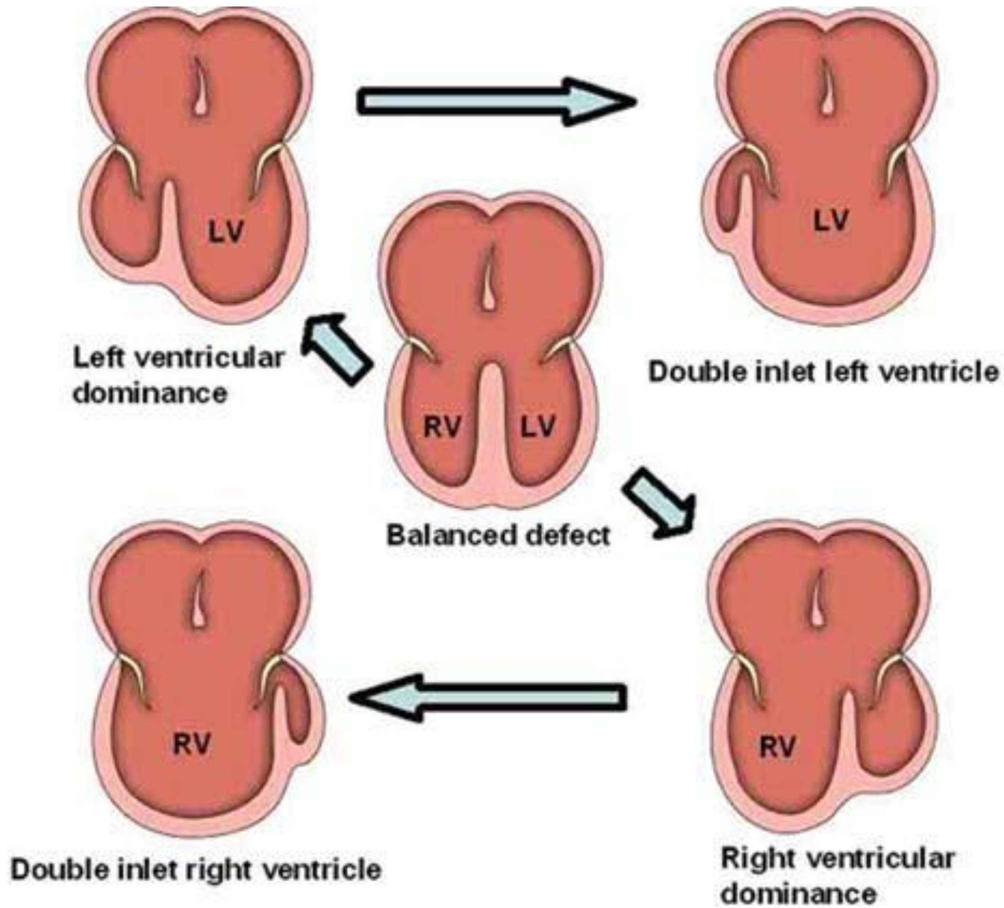
Cardiol Young 2006;16 Suppl 1:27-34

Tricuspid Atresia



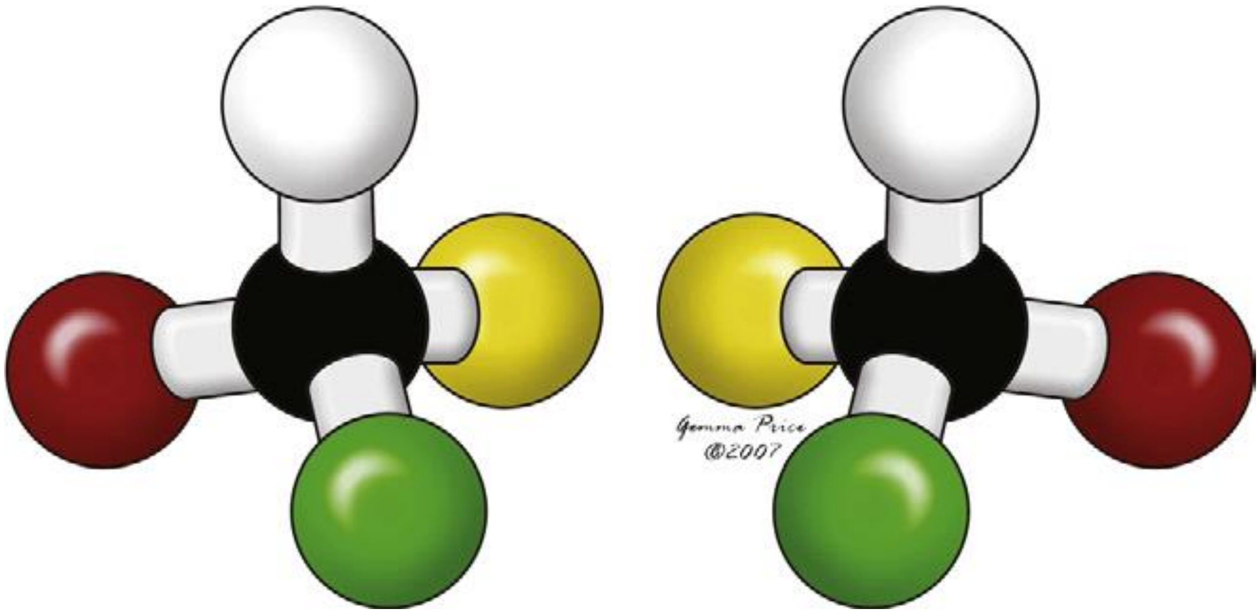
Cardiol Young 2006;16 Suppl 1:27-34

Unbalanced AV Canal Defect



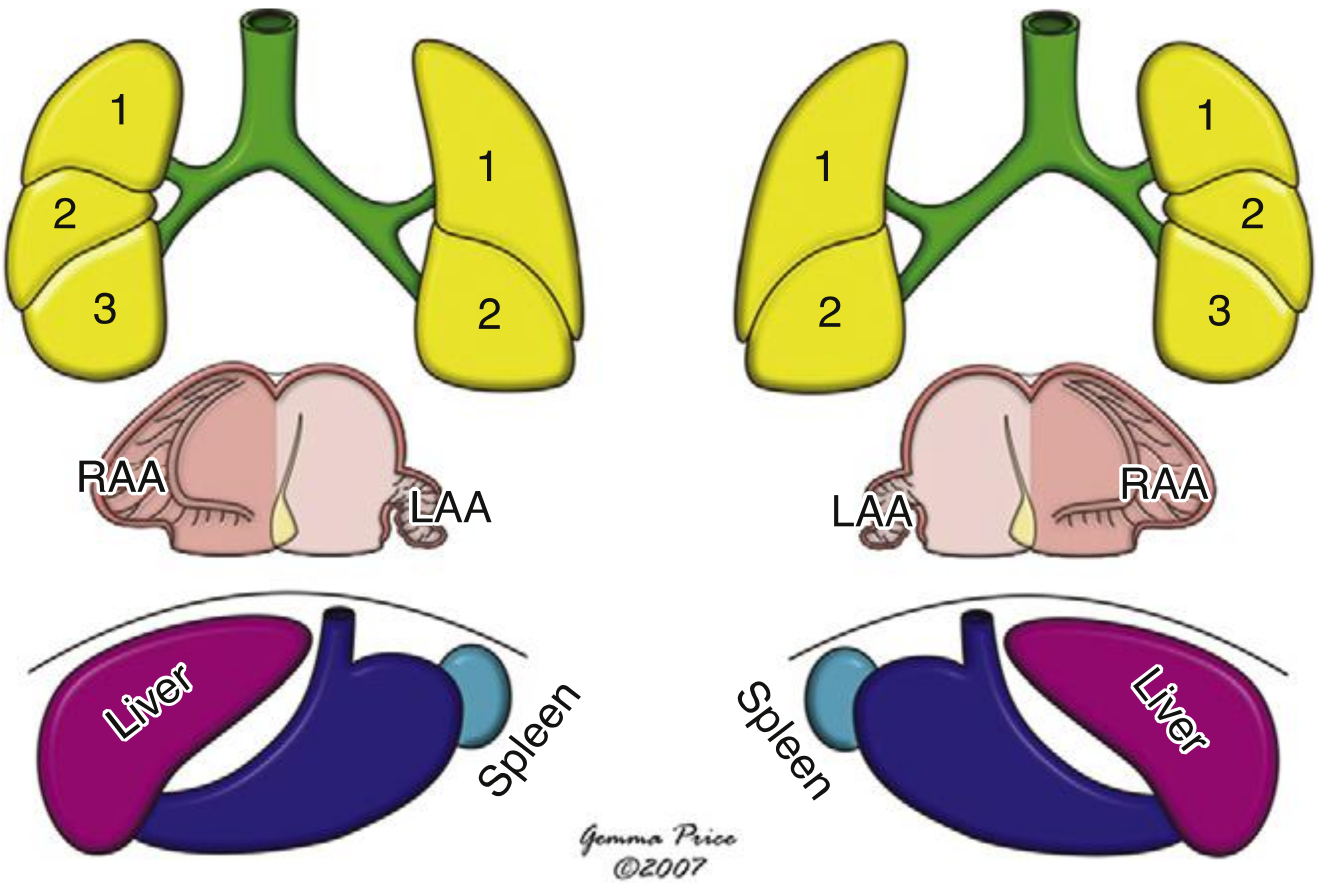
Cardiol Young 2006;16 Suppl 3:43-51

Isomerism



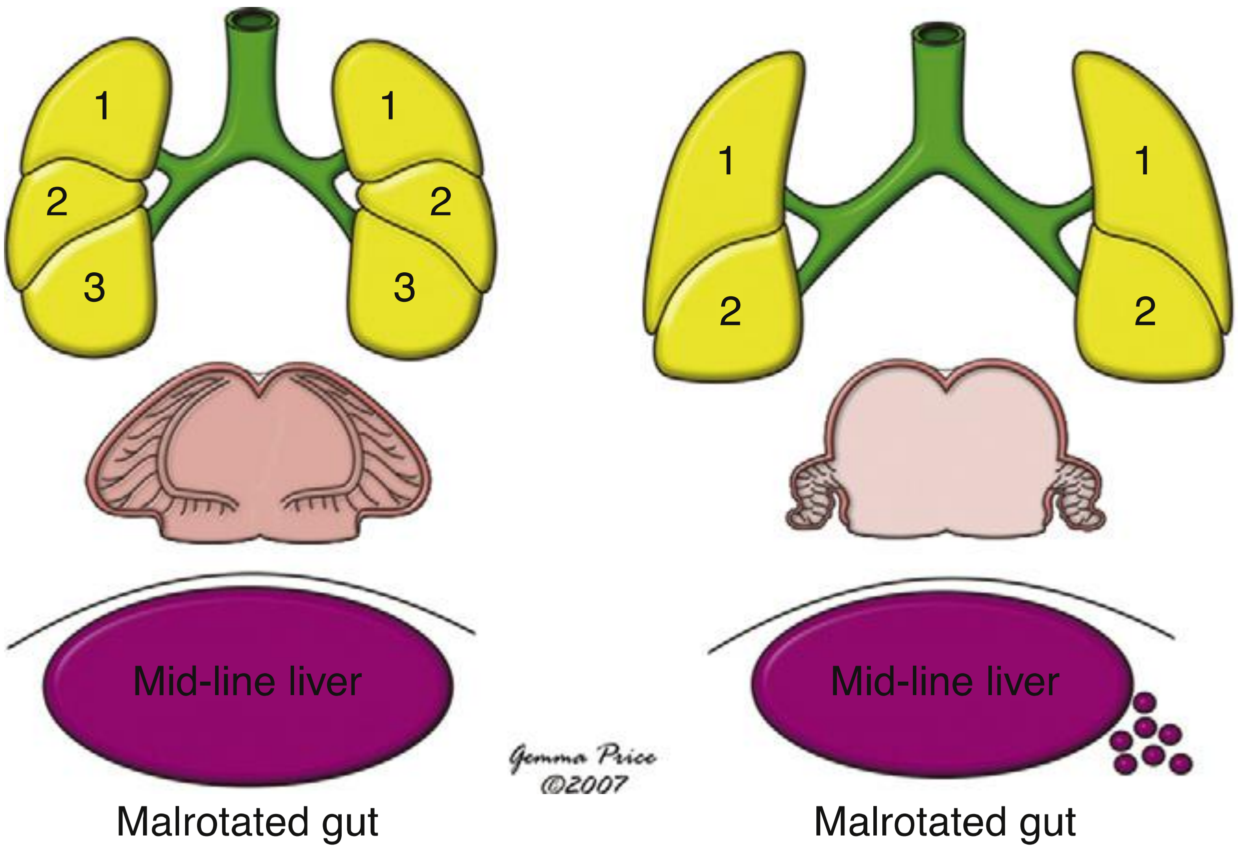
Anderson RH, et al. Paediatric Cardiology. 3rd ed.

Usual Body Arrangement and Its Mirror Image



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

Heterotaxia Syndrome (Isomerism of the Atrial Appendages)



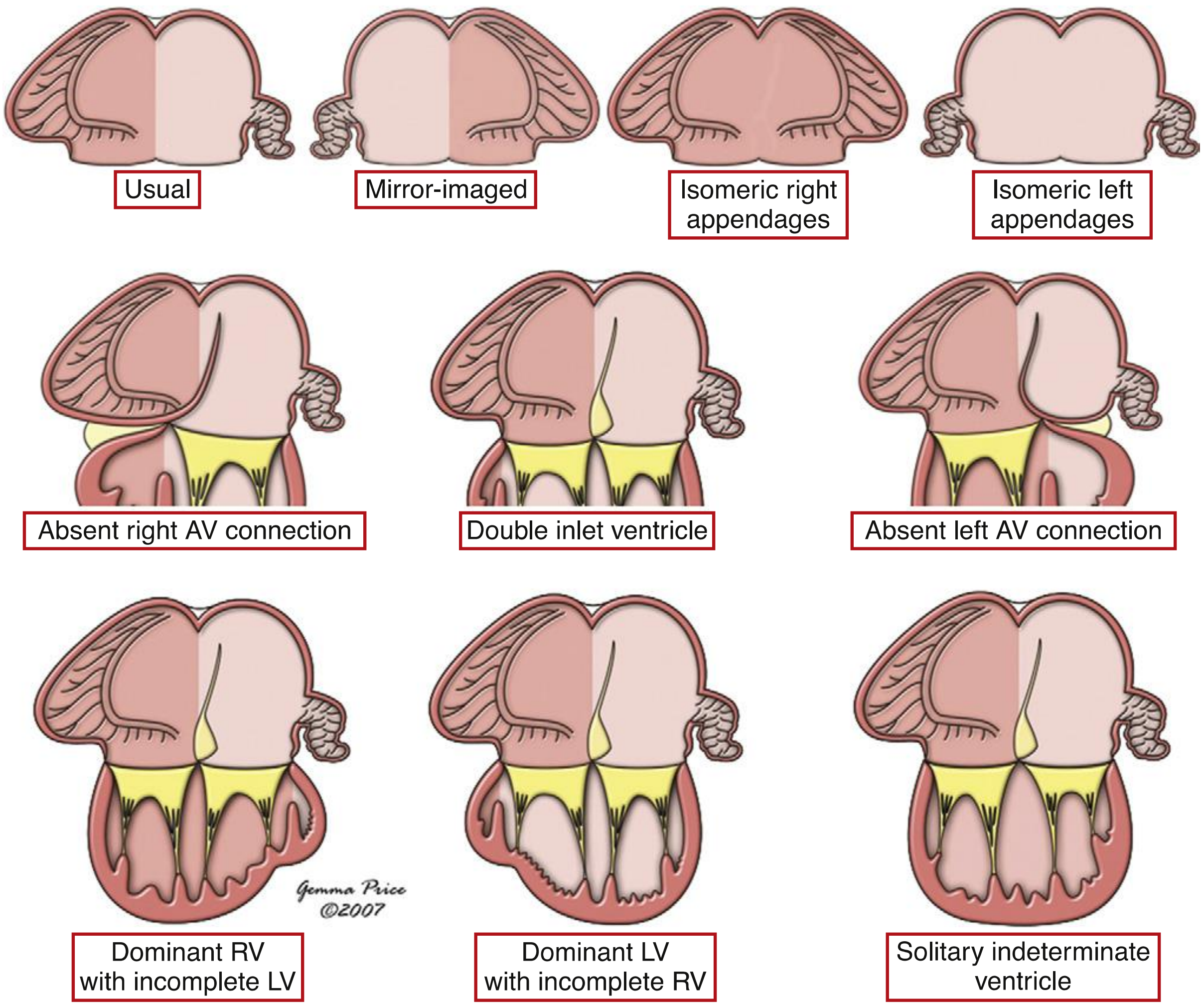
Anderson RH, et al. Paediatric Cardiology. 3rd ed.

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

Univentricular Atrioventricular Connections



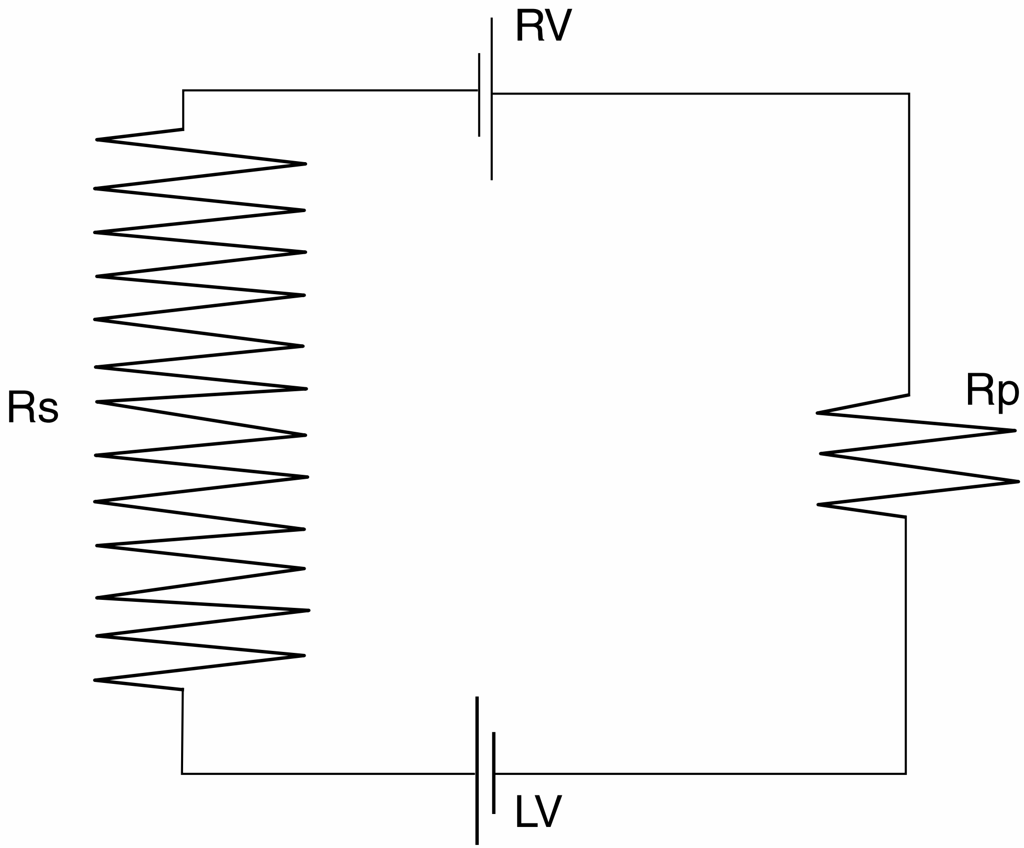
Anderson RH, et al. Paediatric Cardiology. 3rd ed.

Circulatory System vs Electrical Circuit

- $BP = Q \times R$

- $V = I \times R$

Normal Heart

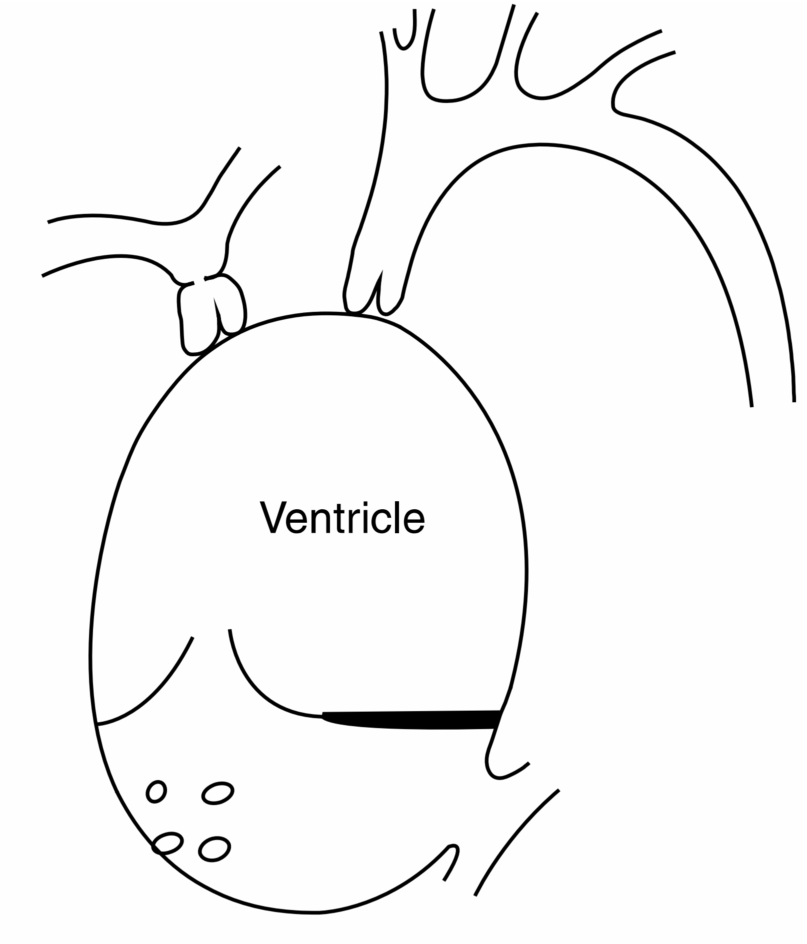


Cardiol Young 2003;13:316-22

Normal Heart

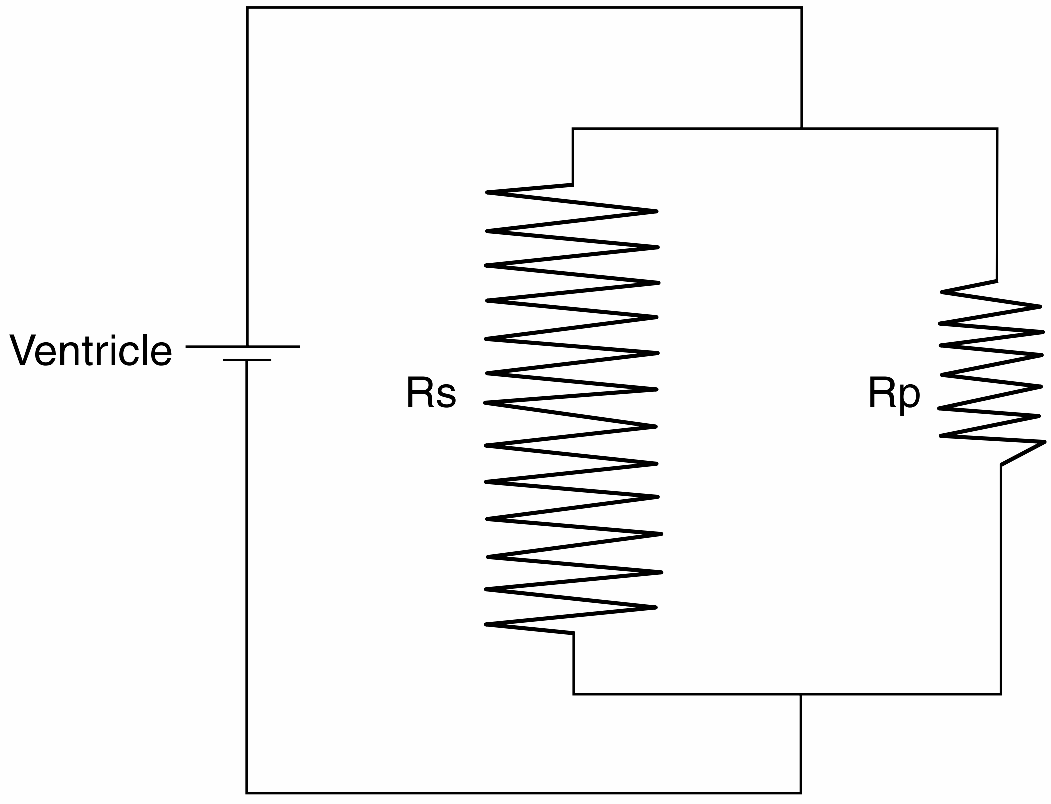
- Serial systemic and pulmonary circulations
- Different BP and O_2 saturation in each part
- CO = Qp = Qs (Qp/Qs = 1)

Single Ventricle



Cardiol Young 2003;13:316-22

Single Ventricle



Cardiol Young 2003;13:316-22

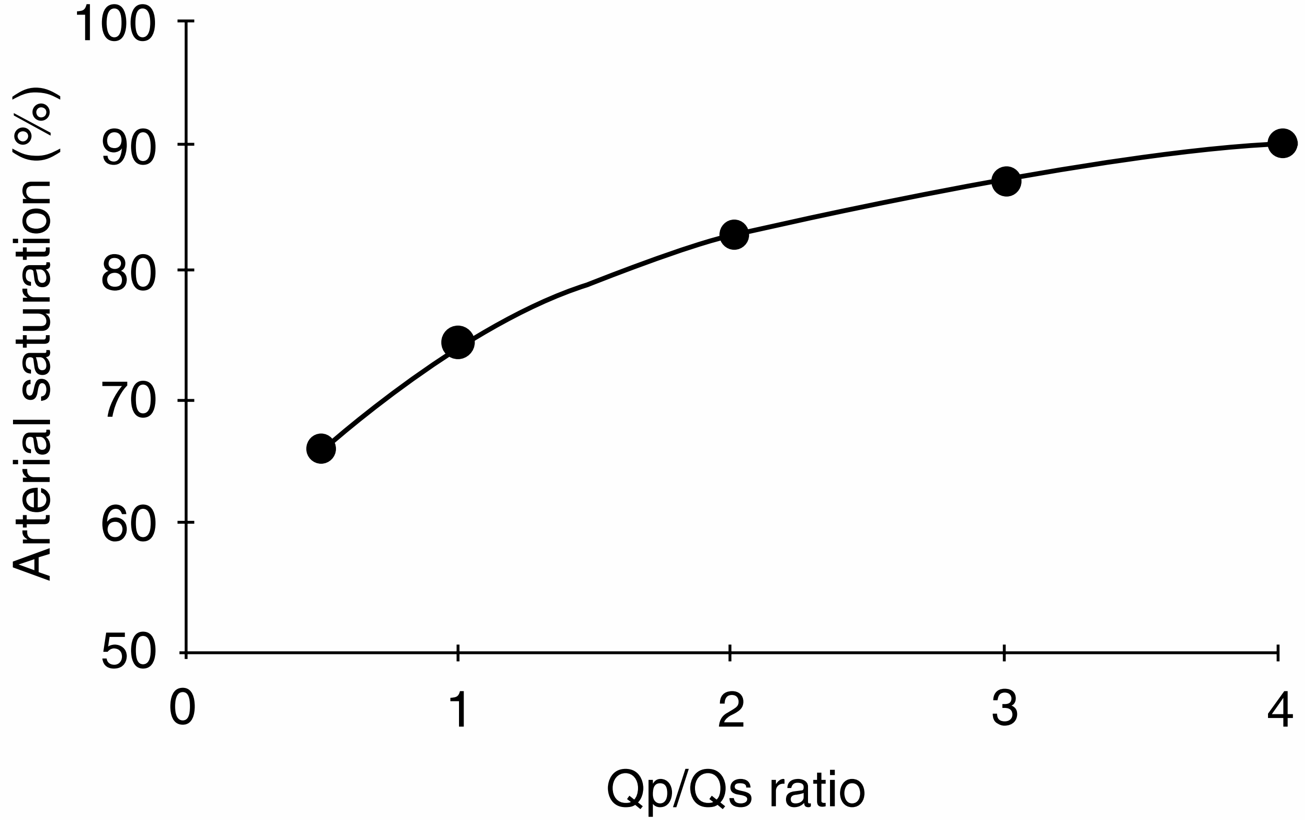
Pathophysiology of Single Ventricle (1)

- 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.

Pathophysiology of Single Ventricle (2)

- CO = Qp + Qs
- Because of the different vascular resistance in each component, flows are different.
- 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).

O_2 Saturation in Single Ventricle



Cardiol Young 2003;13:316-22

"Balanced" Single Ventricle

- Qp = Qs
- Natural obstruction to pulmonary blood flow
- Arterial O_2 saturation of approximately 80%
- Volume overloaded (double the normal CO)

Manipulation of Pulmonary Vascular Resistance

Manoeuvres that increase pulmonary resistance	Manoeuvres that decrease pulmonary resistance
Acidosis	Alkalosis
Increasing positive end expiratory pressure	Nitric oxide
Vasopressor agents (noradrenaline, adrenaline)	Isoproterenol
High arterial partial pressure of carbon dioxide hypoxia, added nitrogen	Lowering mean airway pressure
	High concentration of inspired oxygen
	Phosphodiesterase inhibitors, nitrates

Cardiol Young 2003;13:316-22

Clinical Presentaion

- Determined by Qp/Qs and the associated cardiac lesions
- Cyanosis
- Congestive heart faiure

Ultimate Goal of Surgery

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

Fontan Operation

- Final palliative surgery for single ventricle
- Total cavopulmonary connection
- Staged interventions are necessary to prepare for the successful Fontan operation.

Three-Stage Surgical Management

1. 1st stage palliation

2. Bidirectional cavopulmonary connection

3. Fontan operation

1st Stage Palliation

- Goal: balancing systemic and pulmonary blood flow (Qp/Qs = 1)
- Usually performed during neonatal or early infantile period
- The choice of procedure is determined by the amount of pulmonary blood flow and presence of systemic outflow obstruction.

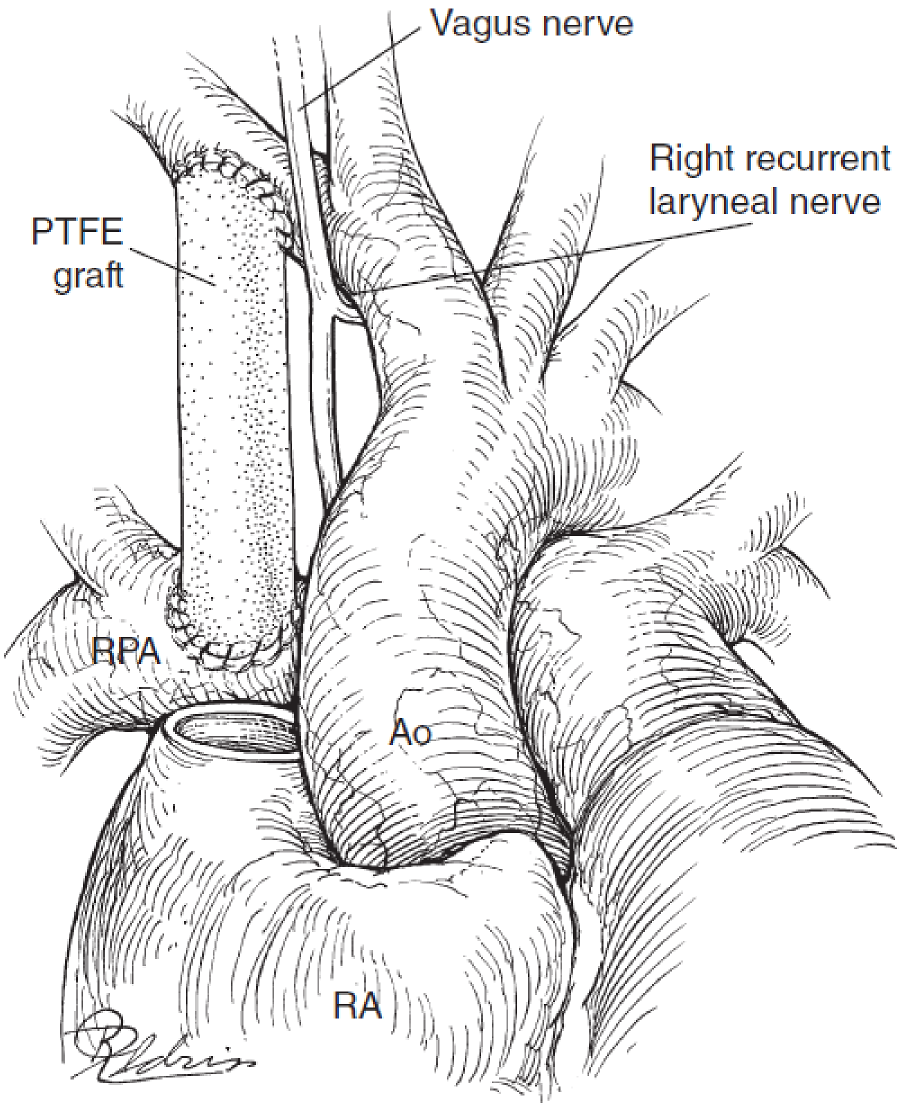
1st Stage Palliations for Inadequate Pulmonary Blood Flow

- Pulmonary outflow obstruction
  → Systemic-to-pulmonary arterial shunt

- Obstructed TAPVC
  → TAPVC repair

- Restrictive ASD
  → Atrial septectomy or balloon septostomy

Modified Blalock-Taussig Shunt

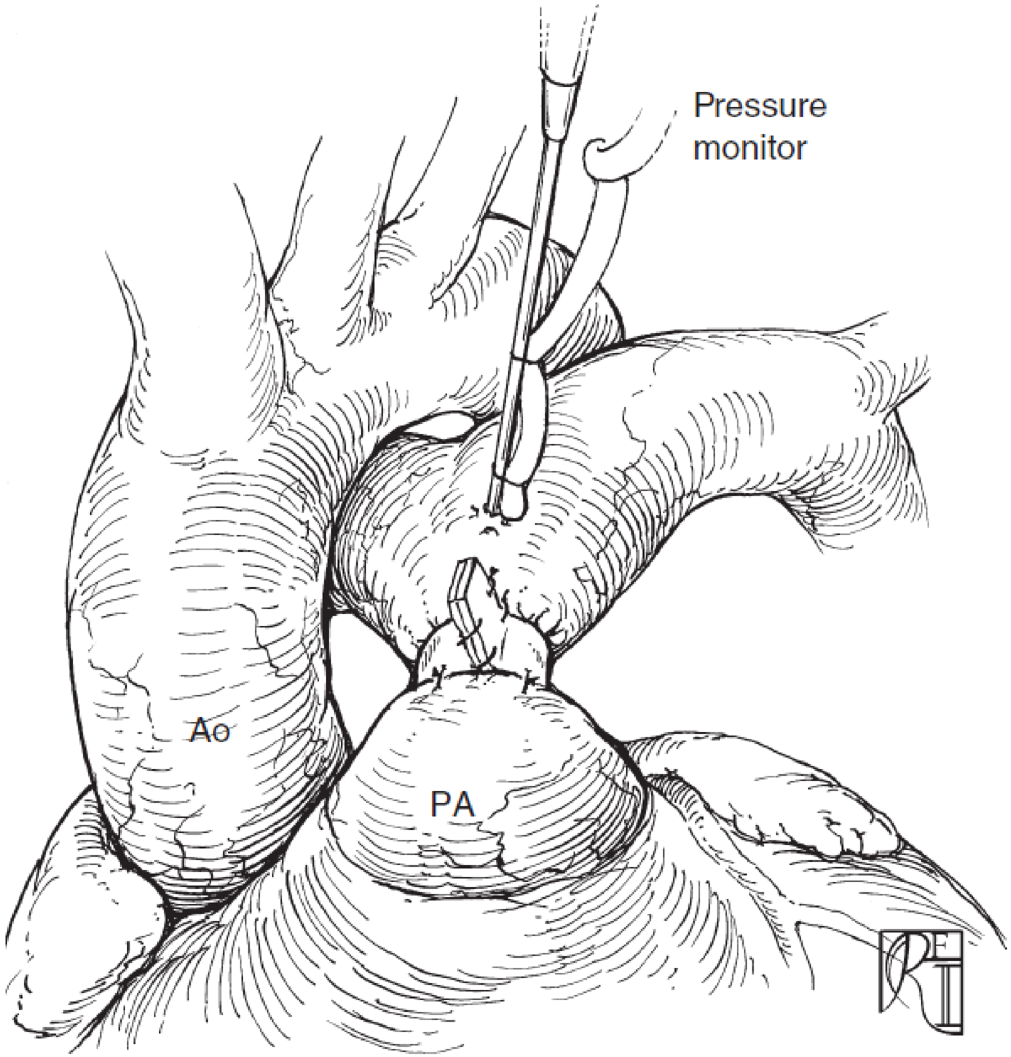


Mavroudis C, et al. Pediatric Cardiac Surgery. 4th ed.

1st Stage Palliation for Excessive Pulmonary Blood Flow

- No pulmonary outflow obstruction
  → Pulmonary artery banding

Pulmonary Artery Banding

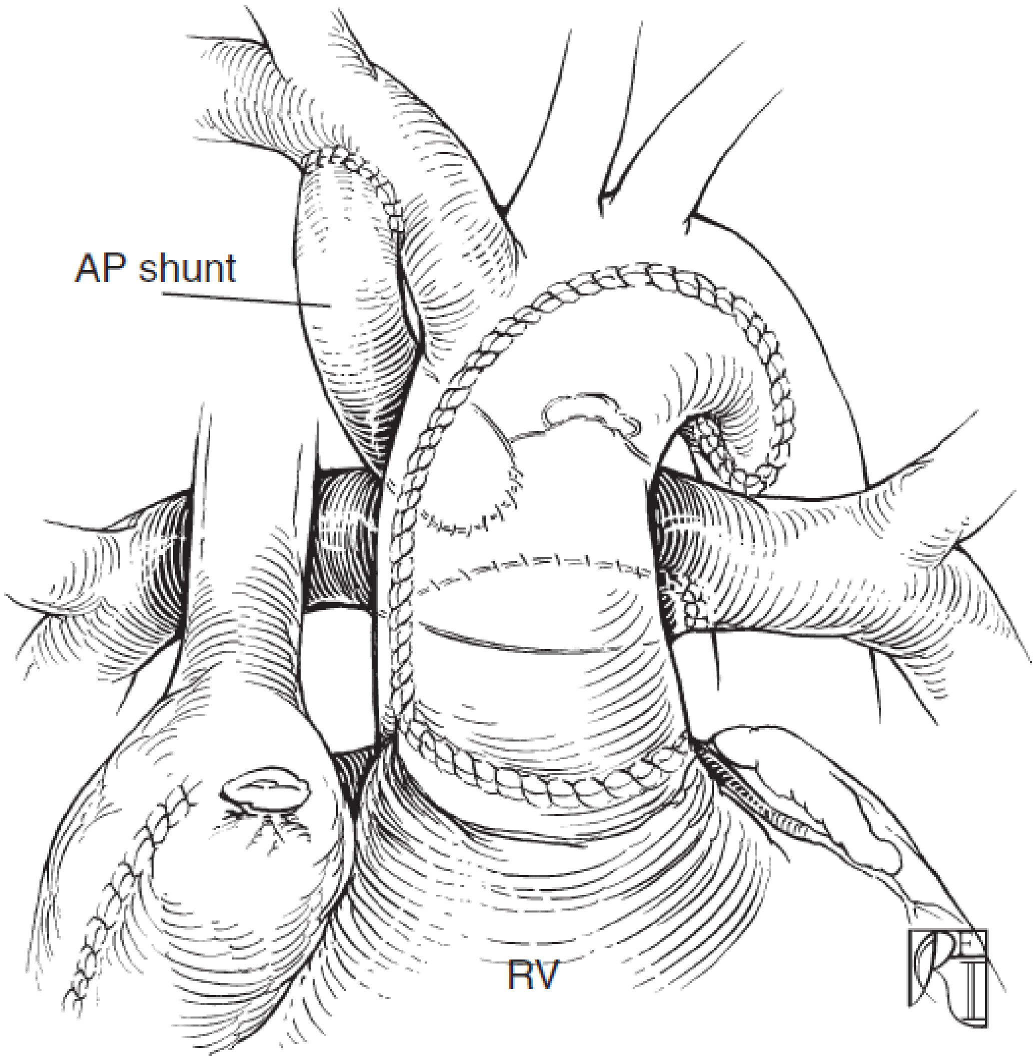


Mavroudis C, et al. Pediatric Cardiac Surgery. 4th ed.

1st Stage Palliations for Systemic Outflow Obstruction

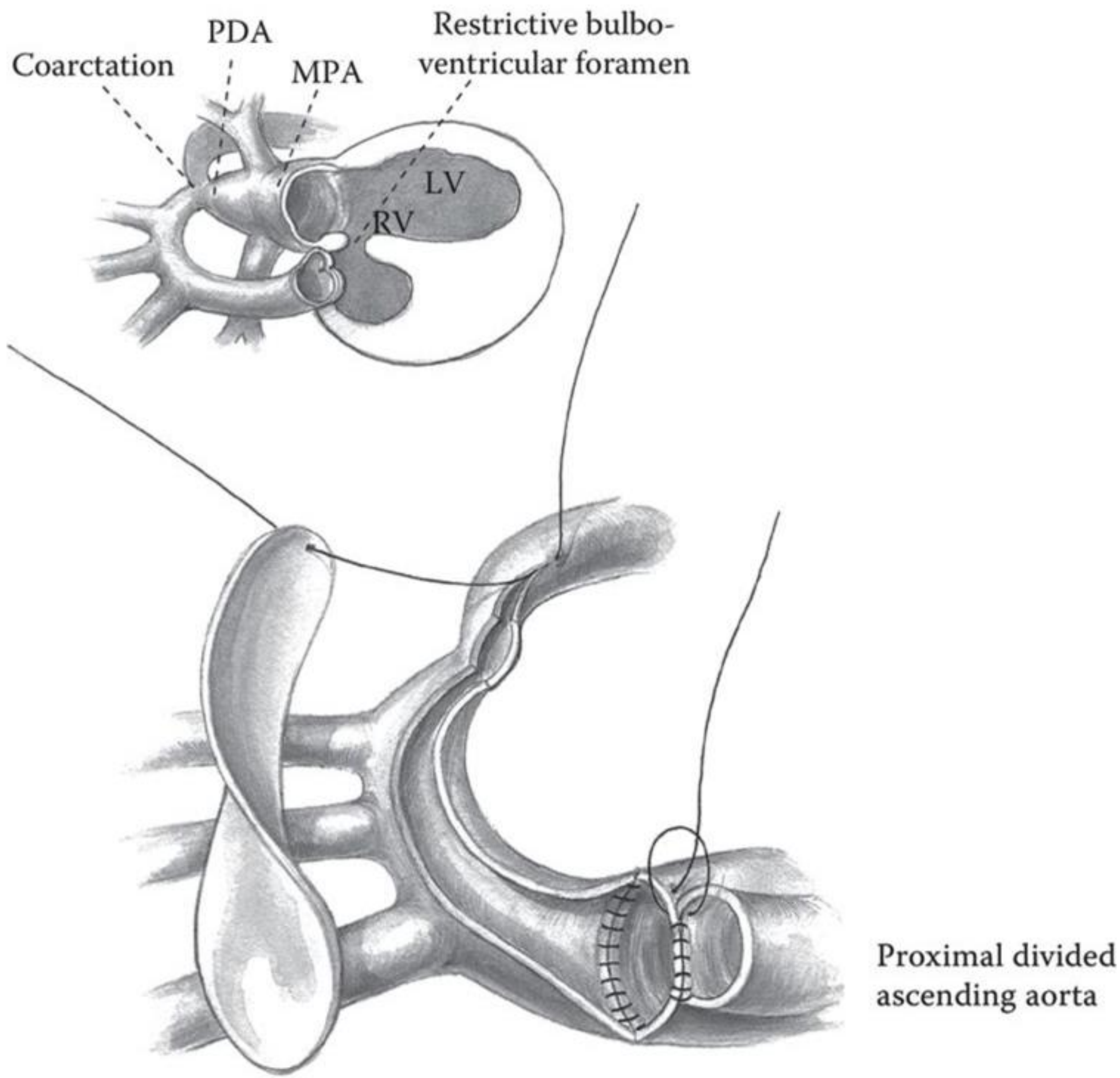
- Aortic and subaortic obstruction
  → Norwood or Damus-Kaye-Stansel procedure

Norwood Procedure



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

Damus-Kaye-Stansel Procedure

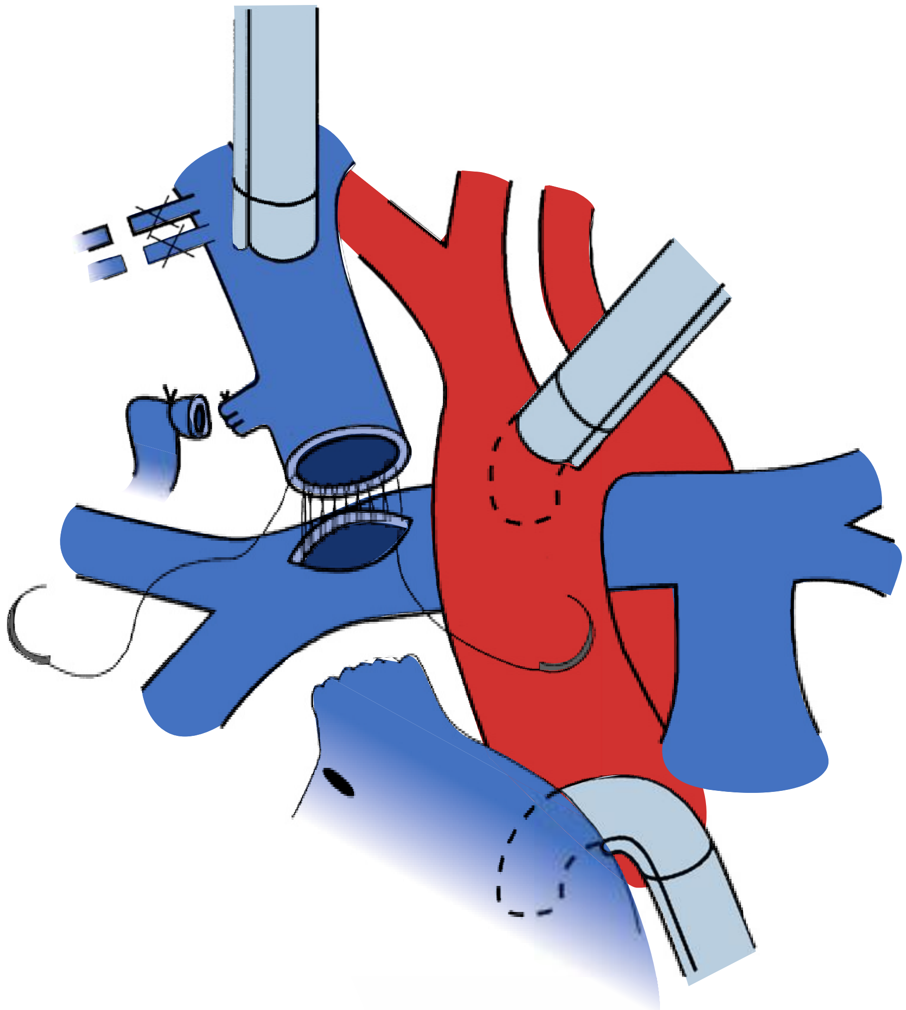


Jonas RA. Comprehensive Surgical Management of Congenital Heart Disease. 2nd ed.

Bidirectional Cavopulmonary Connection

- Also referred to as “bidirectional Glenn”
- 2nd stage palliation
- Usually performed at 3-6 months of age

Bidirectional Cavopulmonary Connection



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

Advantages of BCPC

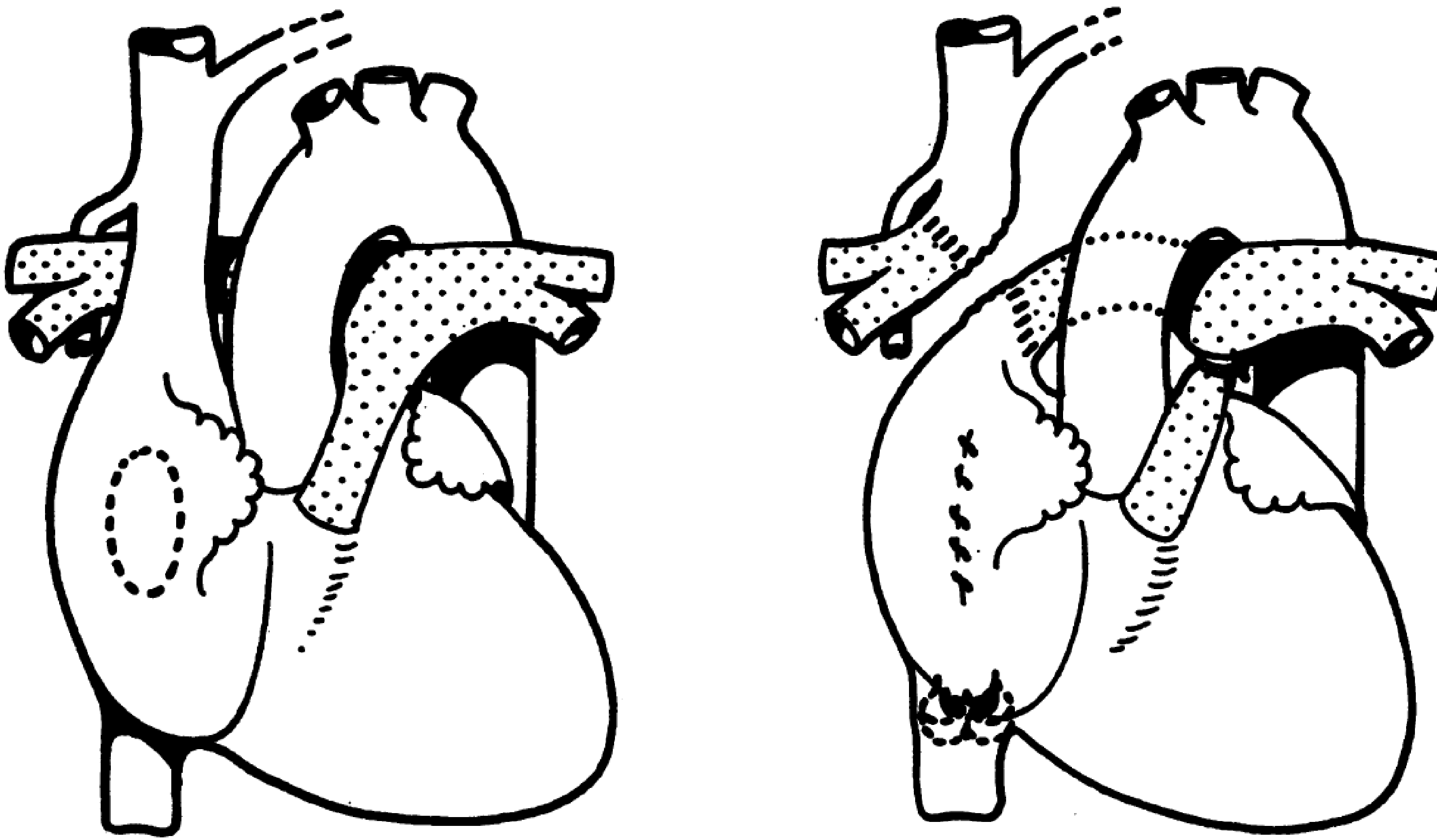
- More desaturated blood (systemic venous rather than arterial) is shunted to the lungs, with therefore a much greater efficacy on increments of arterial oxygen saturation.

- Diversion of approximately one-third of the systemic venous return to the lungs reduces the volume load on the heart, whereas an arterial shunt constitutes an additional ventricular volume and workload.

Fontan Operation

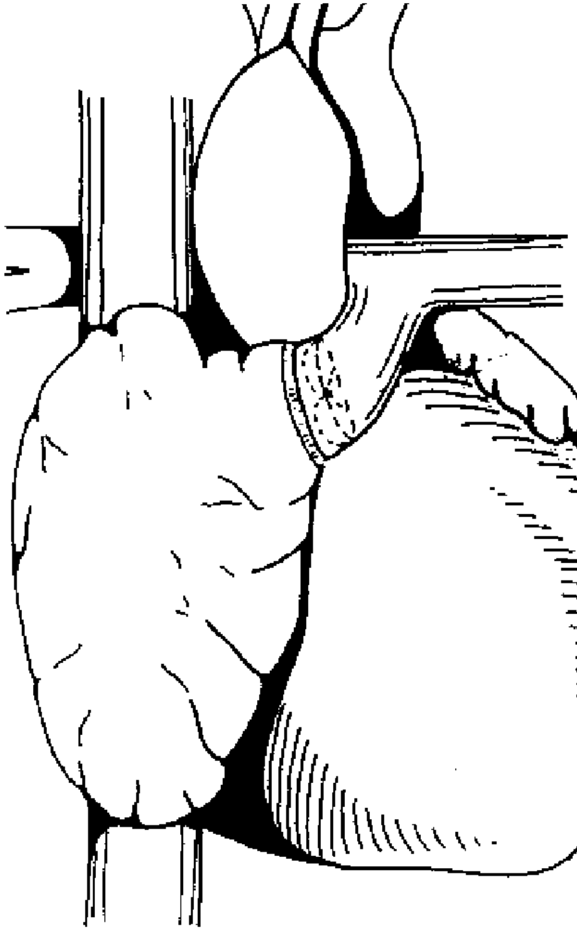
- 3rd stage (final) palliation
- Complete separation of systemic and pulmonary circulations
- Usually performed at 2-3 years of age

Original Fontan Operation



Thorax 1971;26:240-8

Kreutzer's Atriopulmonary Connection



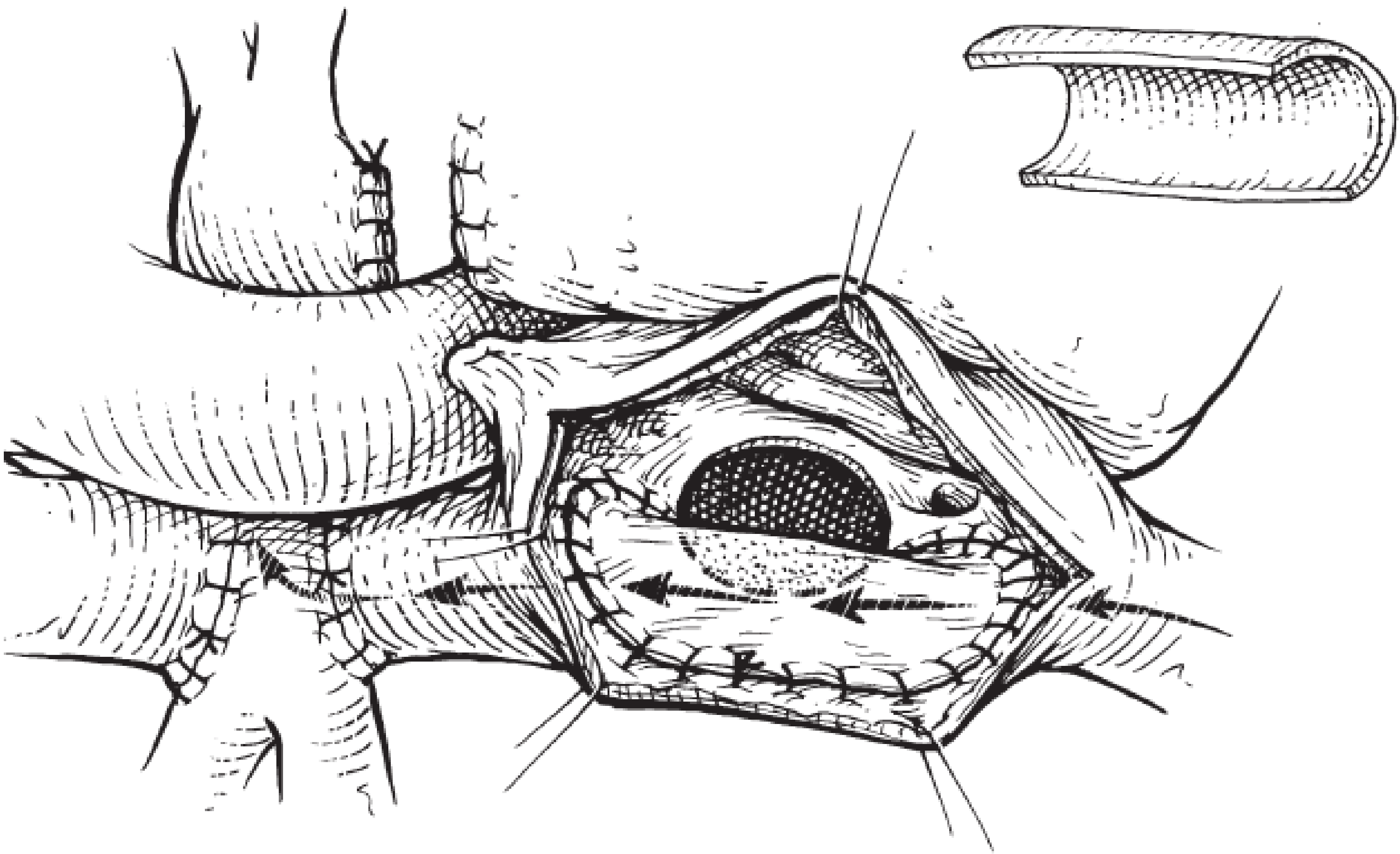
J Thorac Cardiovasc Surg 1973;66:613-21

Complications of AP Fontan

- Exposure of the RA to the high pressure of the Fontan circuit → huge dilatation of RA

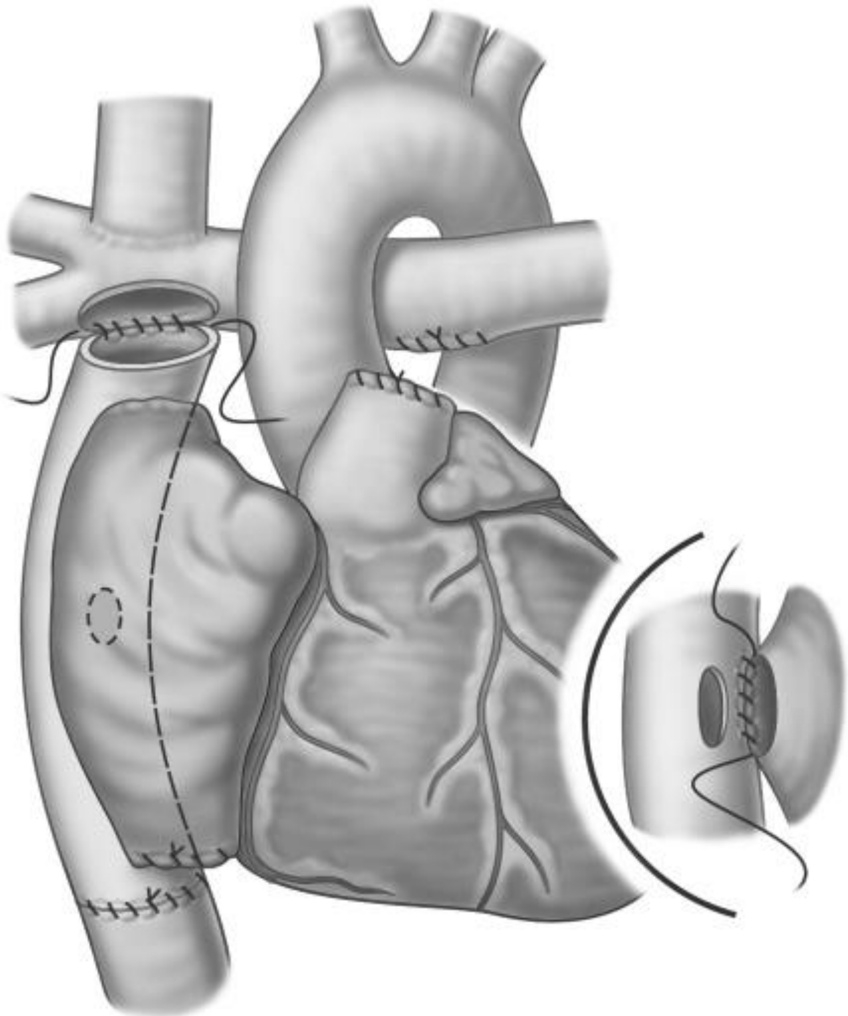
- Supraventricular tachycardia
- Pulmonary venous obstruction
- Atrial thrombi

Lateral Tunnel Fontan



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

Extracardiac Conduit Fontan



Stark JF, et al. Surgery for Congenital Heart Defects. 3rd ed.

Extracardiac Conduit vs Lateral Tunnel Fontan

Extracardiac conduit	**Lateral tunnel**
Technically simpler, more easily reproducible with variable patient morphology	Judgment required
"Minimizes" atrial suture lines	Heavier atrial suture burden
No atrial tissue at high pressure	Thin strip of atrial tissue at high pressure
Difficult to fenestrate	Easy to fenestrate
No catheter access to atrium	Catheter access to atrium available
No growth potential	Grows
Avoids CPB + cross-clamp	Short clamp time, CPB mandatory

CPB = cardiopulmonary bypass

Jonas RA. Comprehensive Surgical Management of Congenital Heart Disease. 2nd ed.

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.

European Journal of Cardio-thoracic Surgery 31 (2007) 344–353

EUROPEAN JOURNAL OF
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Factors influencing early and late outcome following the Fontan procedure in the current era. The 'Two Commandments'?☆

Riad B.M. Hosein [a], Andrew J.B. Clarke [a], Simon P. McGuirk [a], Massimo Griselli [a], Oliver Stumper [b], Joseph V. De Giovanni [b], David J. Barron [a], William J. Brawn [a,*]

[a] *Department of Cardiac Surgery, Birmingham Children's Hospital, United Kingdom*
[b] *Department of Cardiology, Birmingham Children's Hospital, United Kingdom*

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 U/m^2
- Mean pulmonary artery pressure: < 15 mm Hg
- Ventricular end-diastolic pressure: < 10-12 mmHg

Issues After Fontan Operation

- Atrial arrhythmias
- Venovenous collaterals
- Pulmonary arteriovenous fistulas
- Thromboembolism
- Protein-losing enteropathy
- Plastic bronchitis
- Fontan failure

Management of Failing Fontan

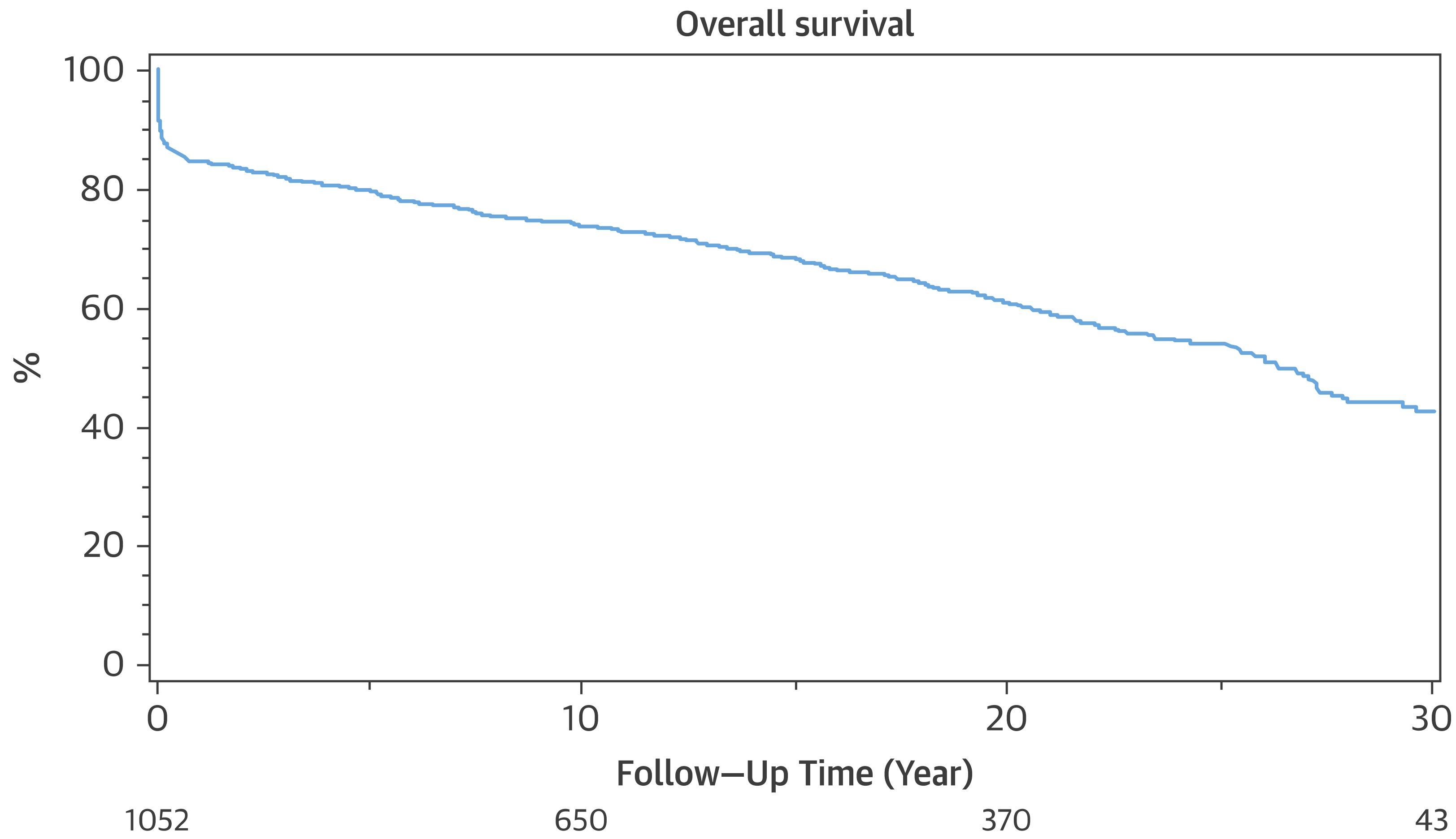
- Fontan conversion
- Heart transplantation
- Mechanical circulatory support

40-Year Follow-Up After the Fontan Operation

Long-Term Outcomes of 1,052 Patients

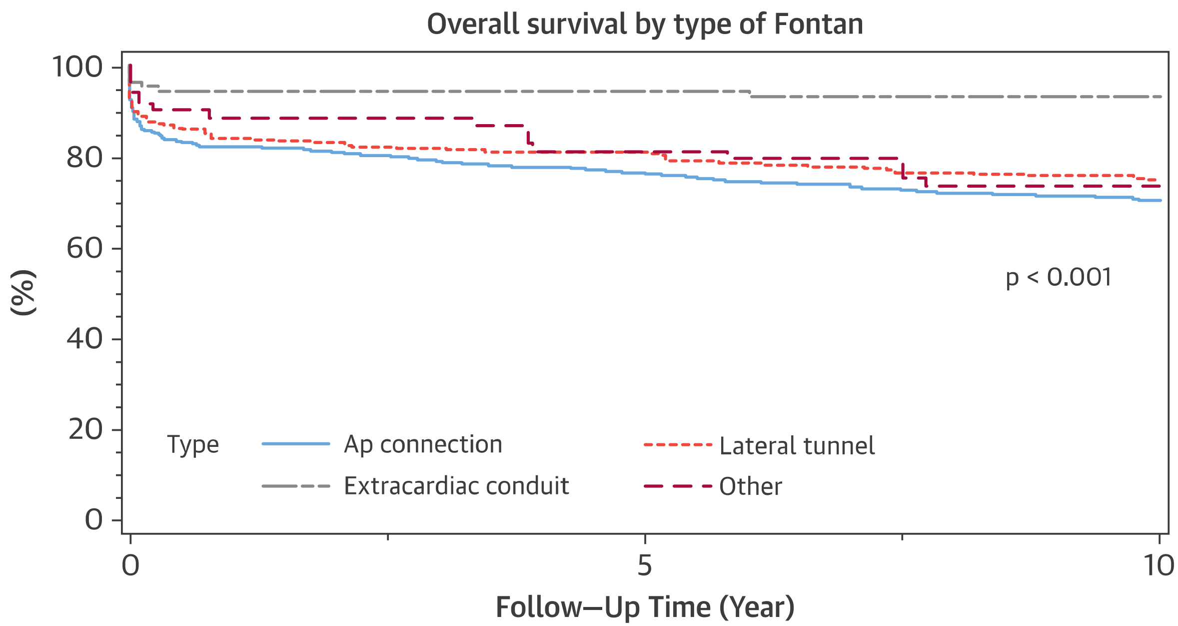
Kavitha N. Pundi, MD,* Jonathan N. Johnson, MD,*† Joseph A. Dearani, MD,‡ Krishna N. Pundi, BS,§ Zhuo Li, BS,||
Cynthia A. Hinck, RN, BSN,* Sonja H. Dahl, RN, DNP,* Bryan C. Cannon, MD,*† Patrick W. O'Leary, MD,*†
David J. Driscoll, MD,* Frank Cetta, MD*†

J Am Coll Cardiol 2015;66:1700-10



J Am Coll Cardiol 2015;66:1700-10

FIGURE 2 **Overall Survival by Type of Fontan Procedure**



Ap connection	616	462	401
Lateral tunnel	262	200	167
Extracardiac conduit	120	77	45
Other	54	44	37

The type of procedure performed greatly impacted survival; patients who had their Fontan with an extracardiac conduit had significantly less mortality. Ap = atriopulmonary.

J Am Coll Cardiol 2015;66:1700-10

European Journal of Cardio-Thoracic Surgery 48 (2015) 825–832
doi:10.1093/ejcts/ezv072 Advance Access publication 13 March 2015

ORIGINAL ARTICLE

Cite this article as: Nakano T, Kado H, Tatewaki H, Hinokiyama K, Oda S, Ushinohama H *et al*. Results of extracardiac conduit total cavopulmonary connection in 500 patients. Eur J Cardiothorac Surg 2015;48:825–32.

Results of extracardiac conduit total cavopulmonary connection in 500 patients[†]

Toshihide Nakano[a,*], Hideaki Kado[a], Hideki Tatewaki[a], Kazuhiro Hinokiyama[a], Shinichiro Oda[a], Hiroya Ushinohama[b], Koichi Sagawa[b], Makoto Nakamura[b], Naoki Fusazaki[c] and Shiro Ishikawa[b]

[a] Department of Cardiovascular Surgery, Fukuoka Children's Hospital, Fukuoka, Japan
[b] Department of Cardiology, Fukuoka Children's Hospital, Fukuoka, Japan
[c] Department of Neonatal Cardiology, Fukuoka Children's Hospital, Fukuoka, Japan

* Corresponding author. Department of Cardiovascular Surgery, Fukuoka Children's Hospital, 5-1-1, Kashii Teriha, Higashi-ku, Fukuoka 813-0017, Japan. Tel: +81-92-6827000; fax: +81-92-6827300; e-mail: nakano.to@fcho.jp. (T. Nakano).

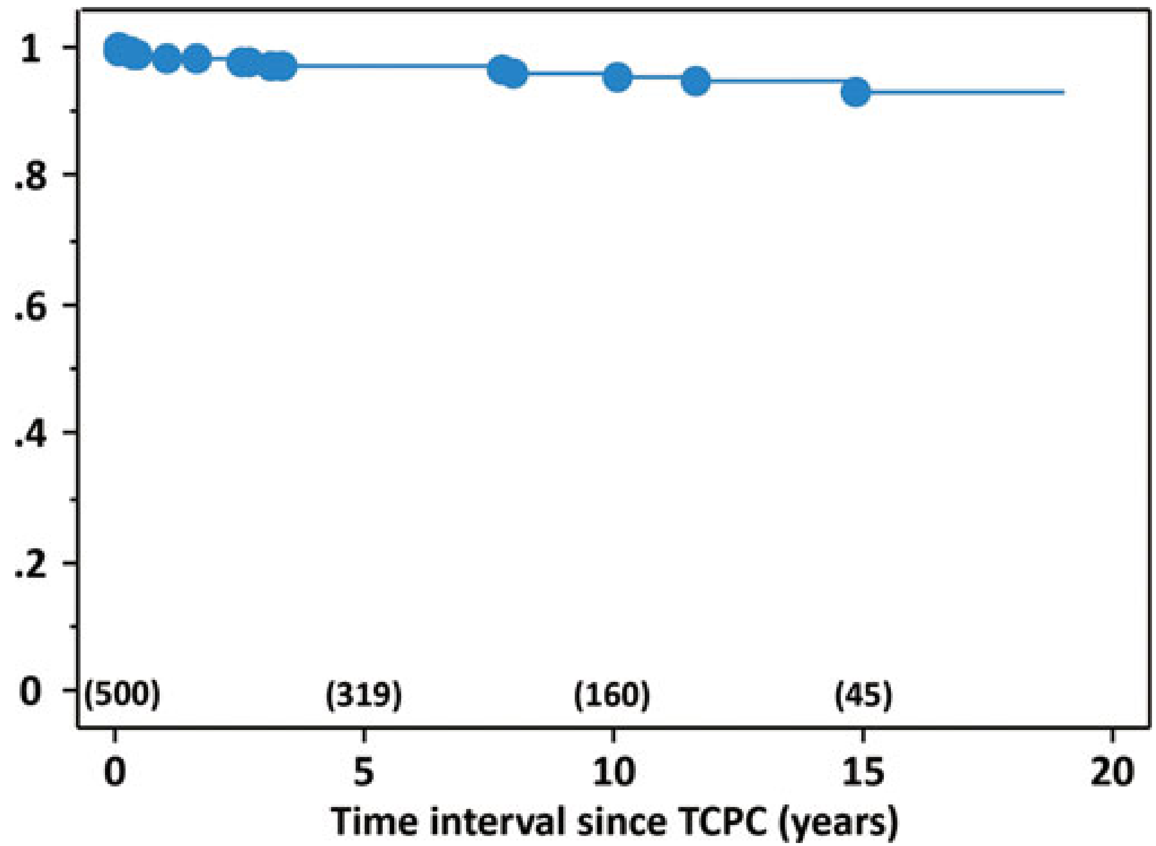


Figure 1: Actuarial survival. The Kaplan–Meier estimated actuarial survival rates after operation were 96.2% at 10 years and 92.8% at 15 years. The number of patients at risk are shown in parenthesis.

Eur J Cardiothorac Surg 2015;48:825-32

European Journal of Cardio-thoracic Surgery 31 (2007) 1008—1012

EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY

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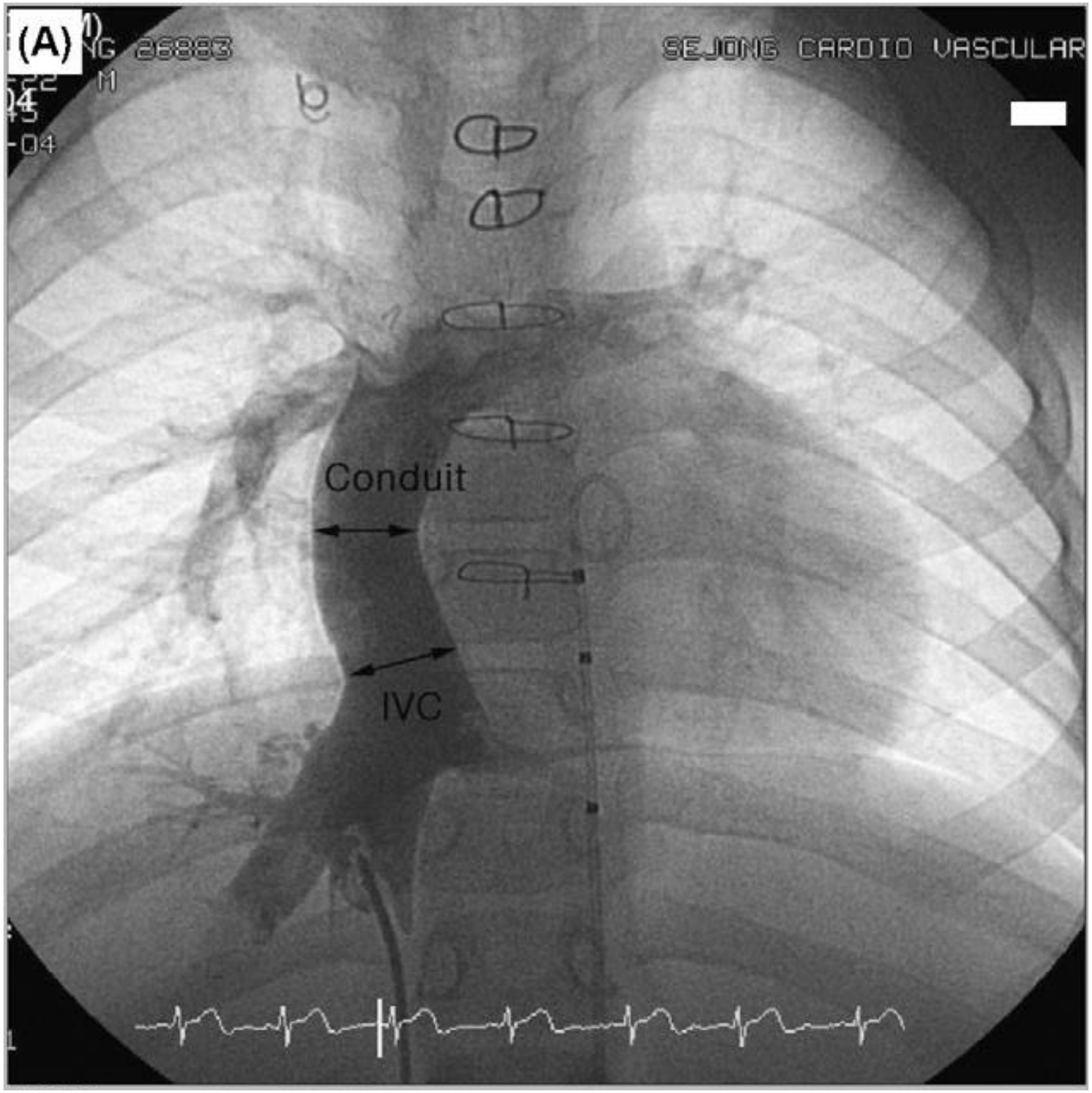
Midterm follow-up of the status of Gore-Tex graft after extracardiac conduit Fontan procedure☆

Cheul Lee [a], Chang-Ha Lee [a,*], Seong Wook Hwang [a], Hong Gook Lim [a], Soo-Jin Kim [b], Jae Young Lee [b], Woo-Sup Shim [b], Woong-Han Kim [c]

[a] *Department of Thoracic and Cardiovascular Surgery, Sejong Heart Institute, Sejong General Hospital, Bucheon, South Korea*
[b] *Pediatric Cardiology, Sejong Heart Institute, Sejong General Hospital, Bucheon, South Korea*
[c] *Department of Thoracic and Cardiovascular Surgery, Clinical Research Institute, Seoul National University, College of Medicine, Seoul National University Children's Hospital, Seoul, South Korea*



Eur J Cardiothorac Surg 2007;31:1008-12

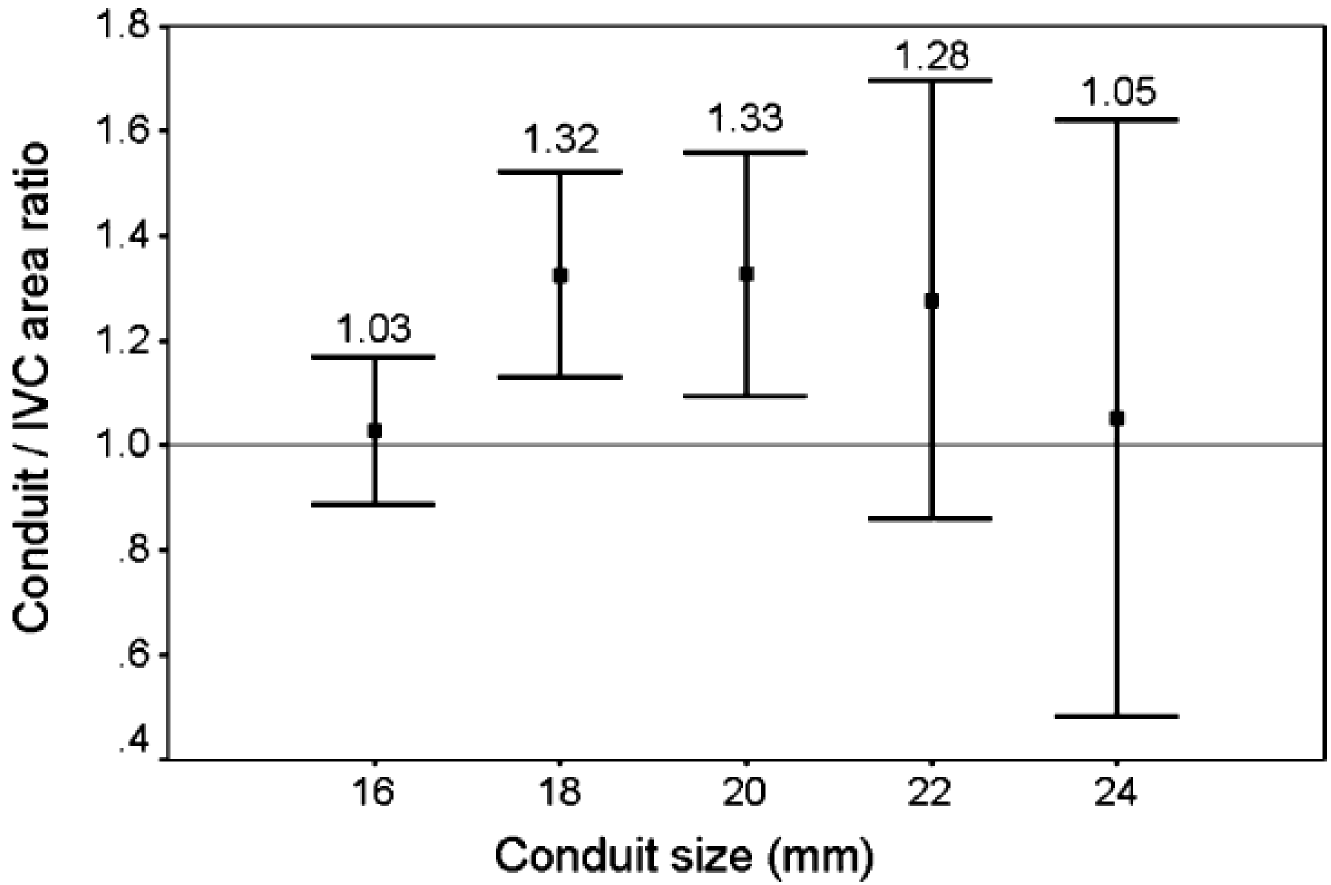


Fig. 5. Conduit-to-IVC cross-sectional area ratio according to conduit size. Filled squares denote mean values (numbers above error bars) and error bars represent 95% confidence interval for means.

Eur J Cardiothorac Surg 2007;31:1008-12