Double Outlet Right Ventricle

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Thoracic and cardiovascular surgery

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May 26 / 2017

Definition of DORV

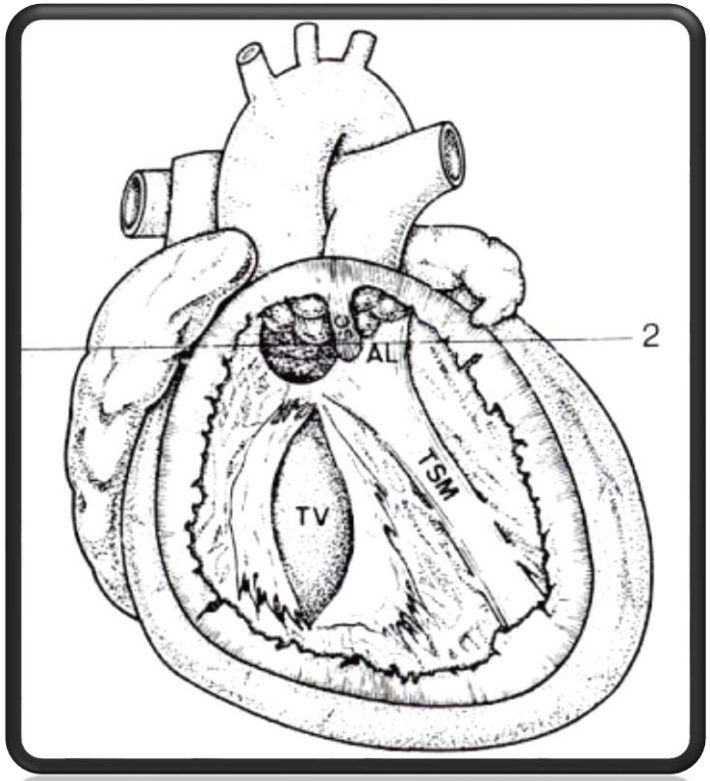
- A congenital anomaly in which both great arteries arise wholly or in large part from the right ventricle
- Presence of bilateral infundibulum
- Presence of atrioventricular valve-semilunar valve discontinuity (mitral aortic discontinuity)
- In the spectrum of TOF and TGA
- 50% rule: a heart is termed DORV if > 50% of both great arteries arise form the right ventricle

History of nomenclature

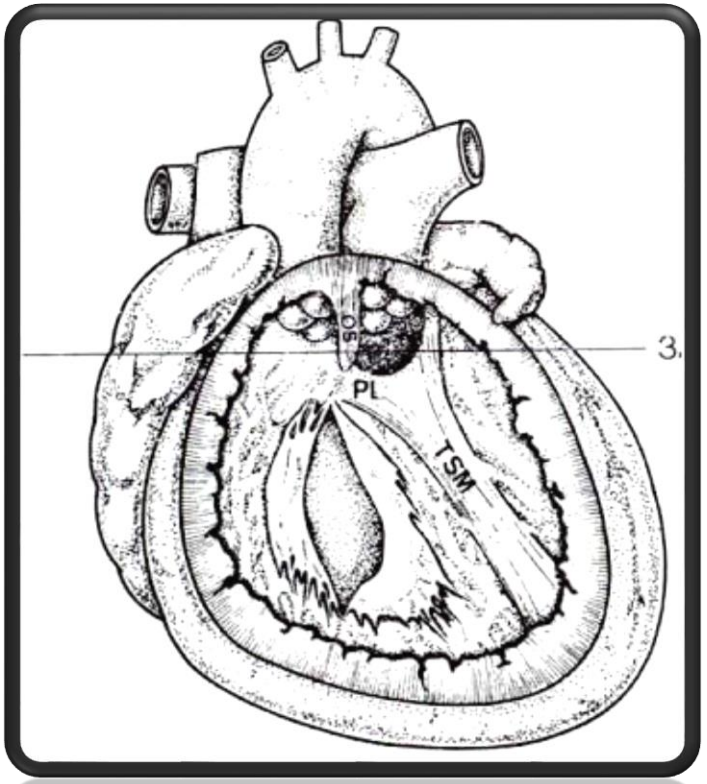
- 1898, Vierordt partial TGA
- 1923, Spitzer type II TGA (simple TGA)
- 1949, Taussig and Bing TGA with levoposition of PA
- 1950, Lev and Bolk Taussig-Bing heart
- 1952, Braun double outlet ventricle
- 1957, Witham DORV

- 1957, Kirklin at Mayo Clinic 1st operation
- 1967, Kirklin successful repair of Taussig-Bing
- 1968, McGoon
- 1971, Kawashima

Classification by Lev (1972)

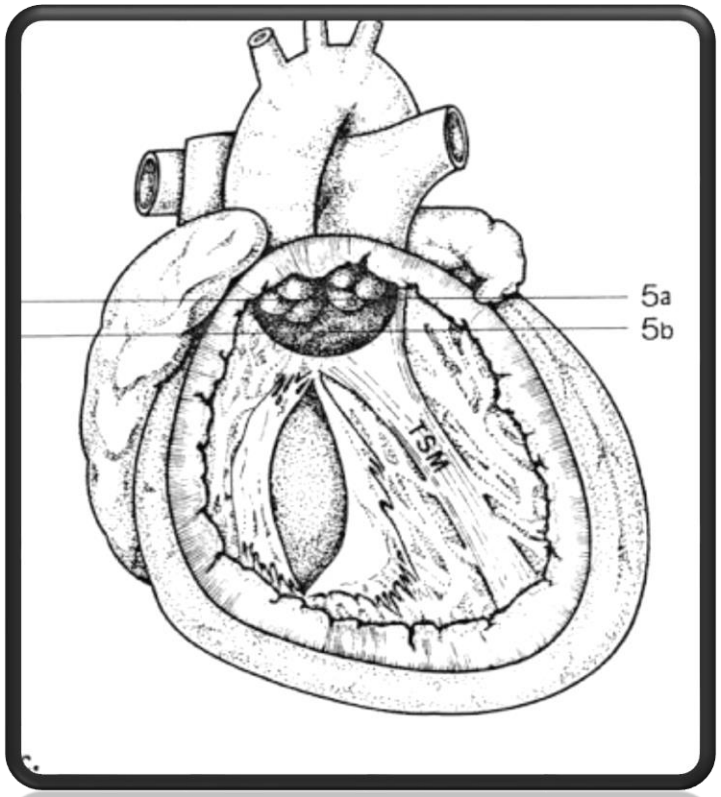


DORV with subaortic VSD

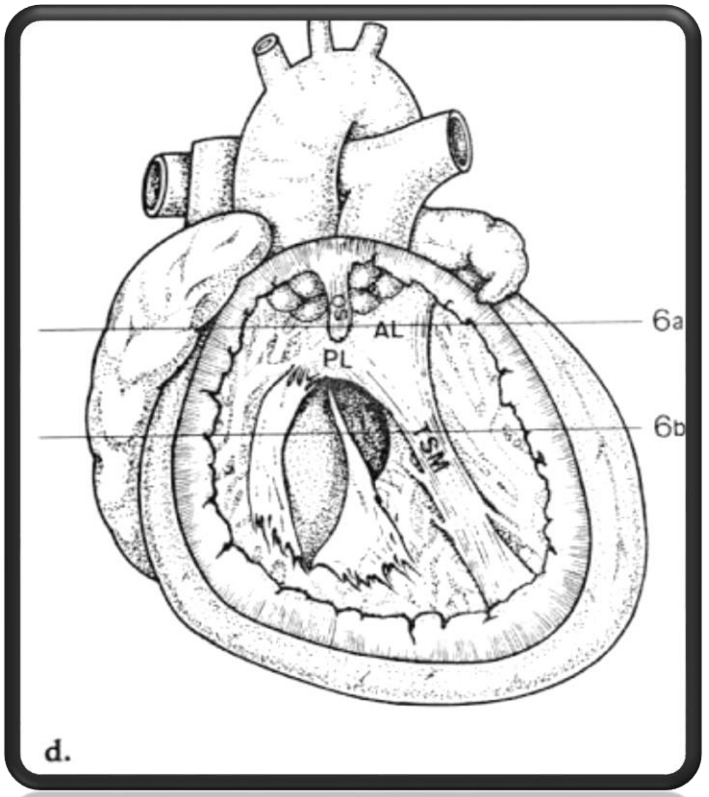


DORV with subpumonic VSD

Classification by Lev (1972)



DORV with doubly committed VSD



DORV with noncommitted VSD

CHSS classification

Congenital Heart Surgery Nomenclature and Database Project: Double Outlet Right Ventricle

Henry L. Walters III, MD, Constantine Mavroudis, MD, Christo I. Tchervenkov, MD, Jeffrey P. Jacobs, MD, François Lacour-Gayet, MD, and Marshall L. Jacobs, MD

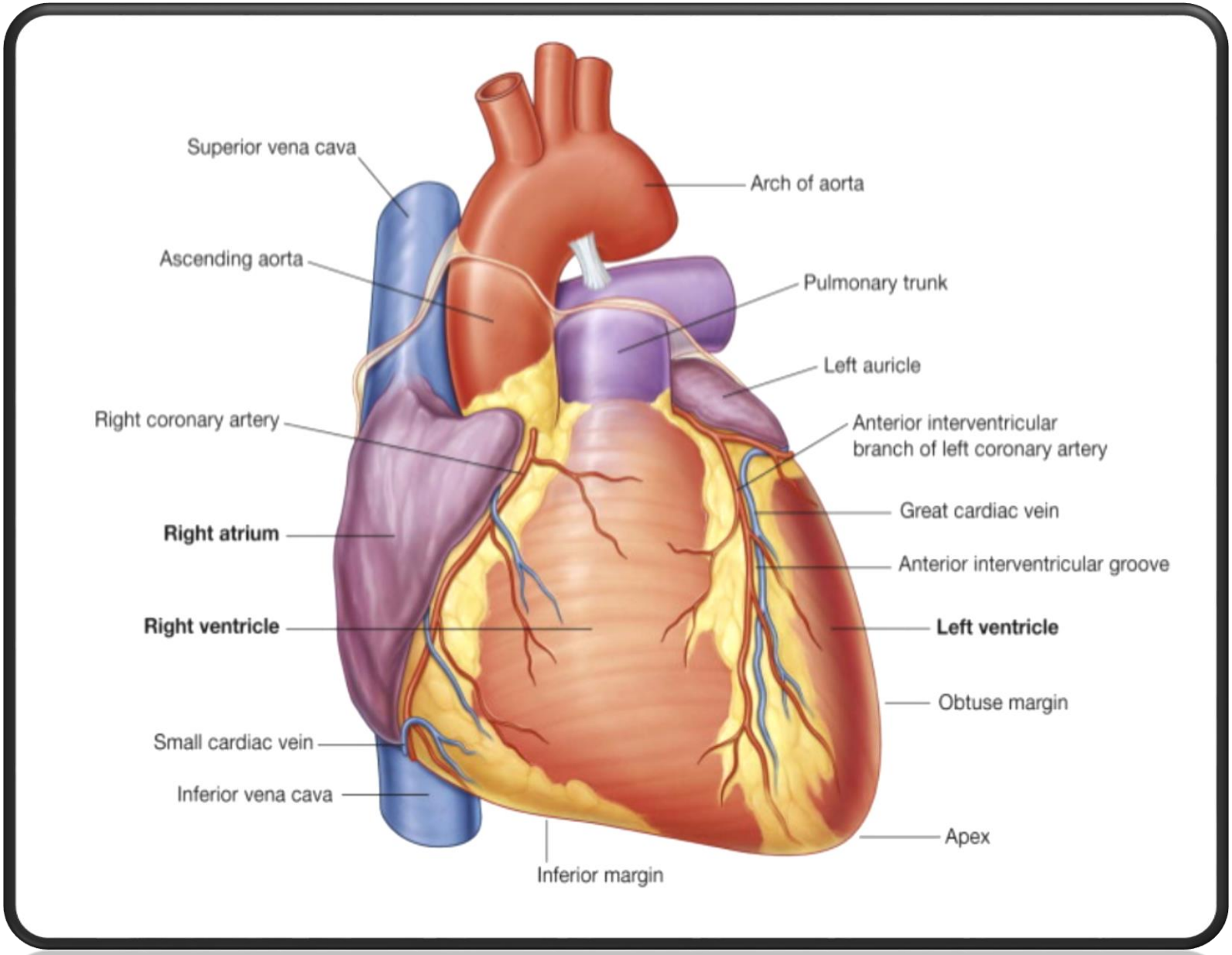
Departments of Surgery, Wayne State University School of Medicine, Children's Hospital of Michigan, Detroit, Michigan; Northwestern University School of Medicine, Children's Memorial Hospital, Chicago, Illinois; McGill University, The Montreal Children's Hospital, Montreal, Quebec, Canada; University of South Florida School of Medicine, All Children's Hospital, St. Petersburg, Florida; Marie Lannelongue Hospital, Paris, France; and Hanneman University School of Medicine, St. Christopher's Children's Hospital, Philadelphia, Pennsylvania

Double outlet right ventricle (DORV) is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. Although the presence of aortic-mitral discontinuity and bilateral coni are important descriptors, they should not serve as absolute prerequisites for the diagnosis of DORV. The morphology of DORV is encompassed by a careful description of the ventricular septal defect (VSD) with its relationship to the semilunar valves, the great artery relationships to each other, the coronary artery anatomy, the presence or absence of pulmonary outflow tract obstruction (POTO) and aortic outflow tract obstruction (AOTO), the tricuspid-pulmonary annular distance, and the presence or absence of associated cardiac lesions. The preferred surgical treatment involves the connection of the left ventricle to the systemic circulation by an intraventricular tunnel repair connecting the VSD to the systemic semilunar valve. This ideal surgical therapy is not always possible due to the presence of confounding anatomical barriers. A multitude of alternative surgical procedures has been devised to accommodate these more complex situations. A framework for the development of the DORV module for a pediatric cardiac surgical database is proposed.

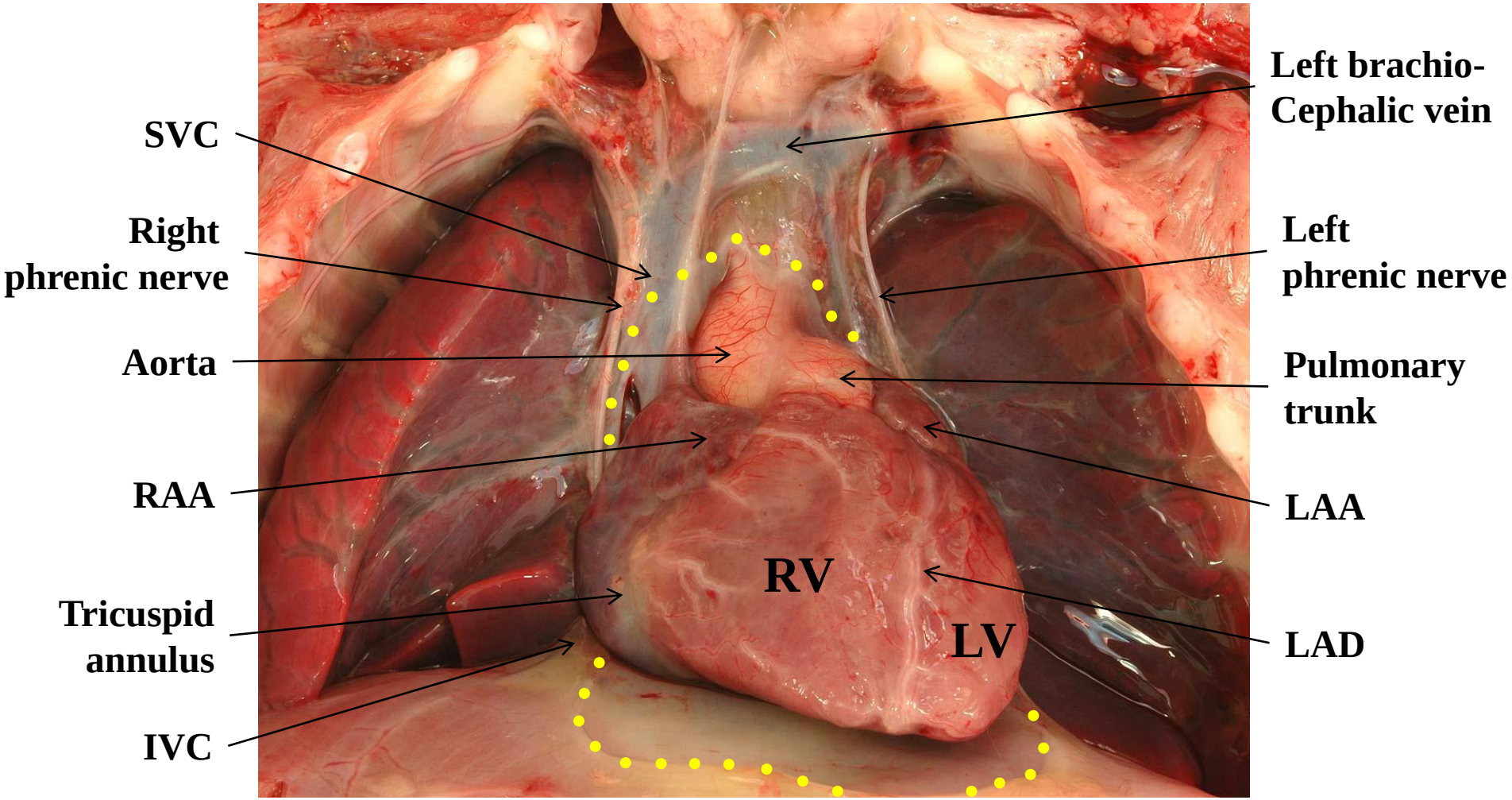
(Ann Thorac Surg 2000;69:S249–63)

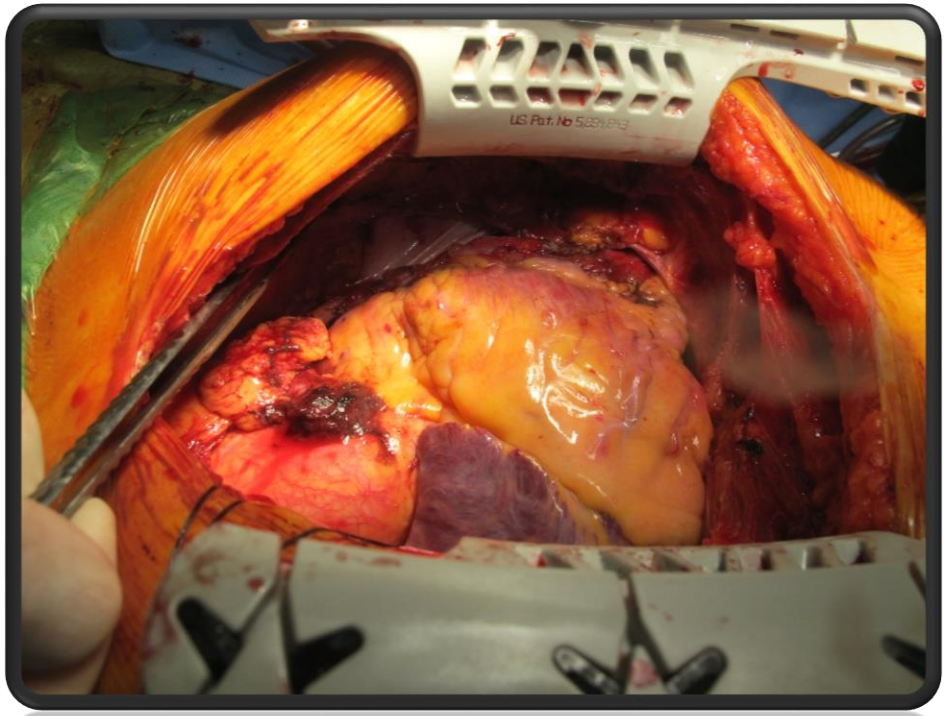
CHSS classification of DORV

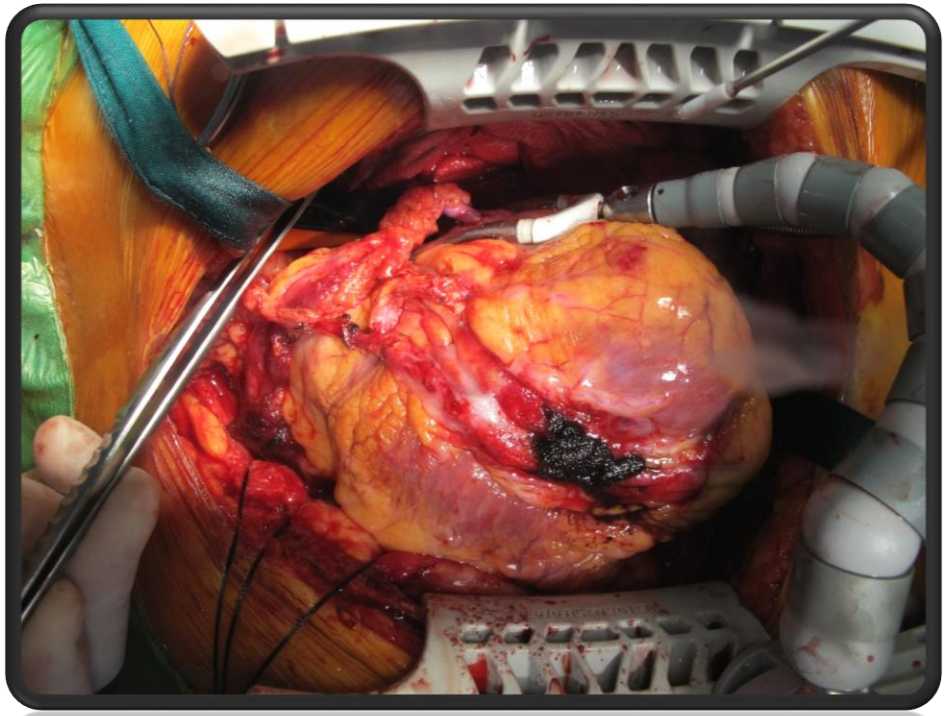
- DORV, VSD type
  - Subaortic or doubly committed, without RVOTO
- DORV, TOF type
  - Subaortic or doubly committed, with RVOTO
- DORV, TGA type
  - Subpulmonary without PS (Taussig-Bing) or with PS
- DORV, Remote type
  - Non committed, with or without RVOTO
- DORV, IVS


Superior vena cava
Arch of aorta
Ascending aorta
Pulmonary trunk
Left auricle
Right coronary artery
Anterior interventricular branch of left coronary artery
Great cardiac vein
Right atrium
Anterior interventricular groove
Right ventricle
Left ventricle
Obtuse margin
Small cardiac vein
Inferior vena cava
Apex
Inferior margin

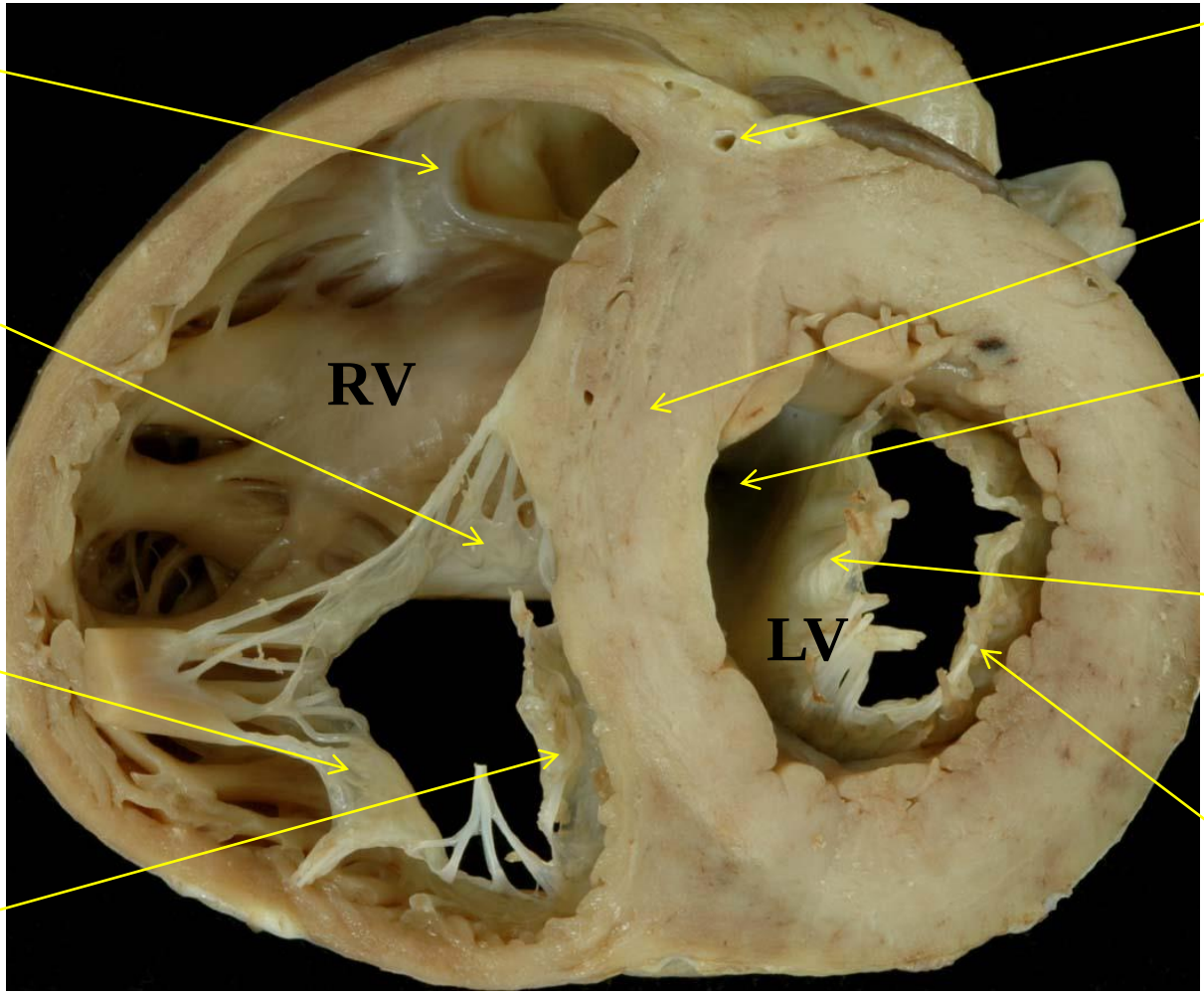
Heart In Situ

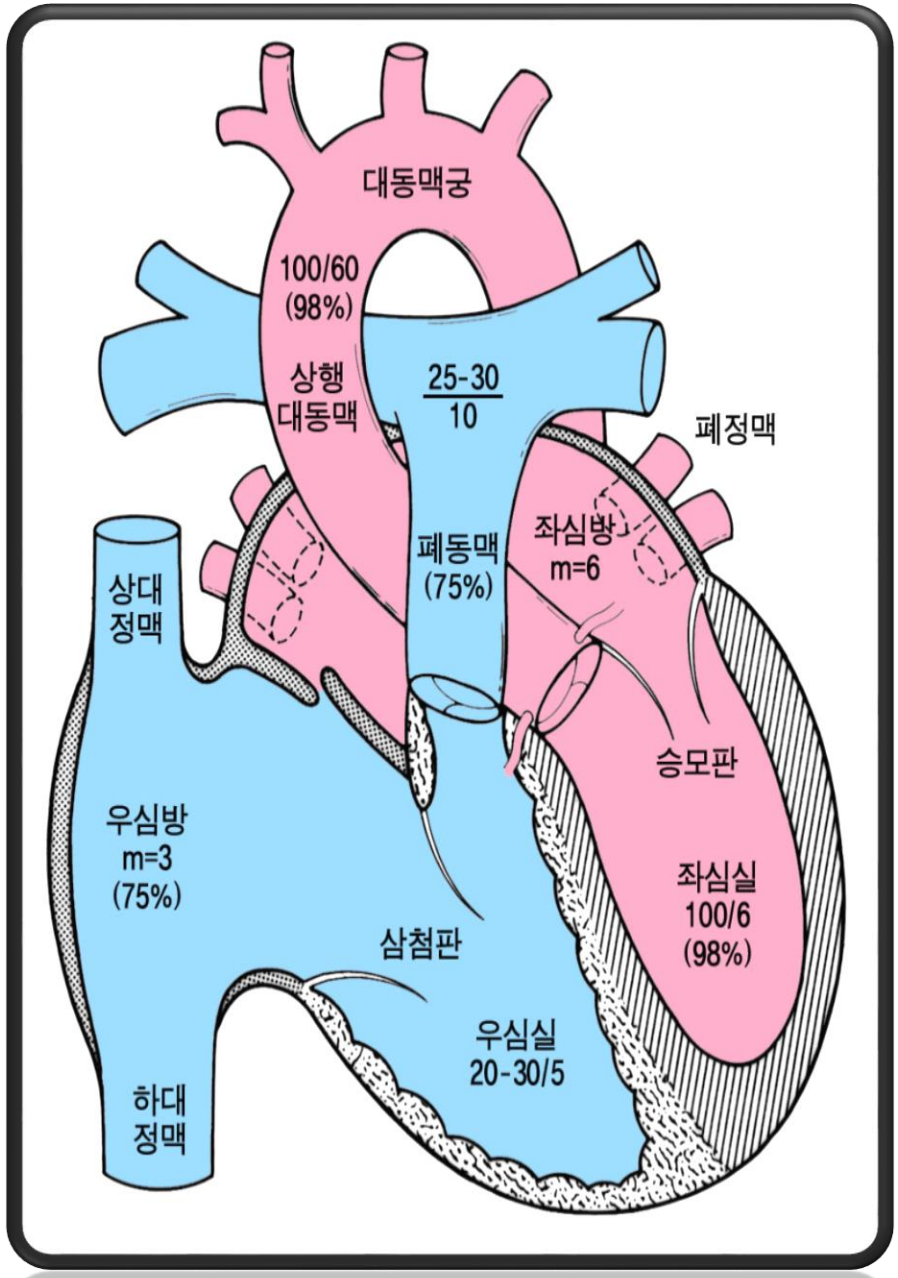


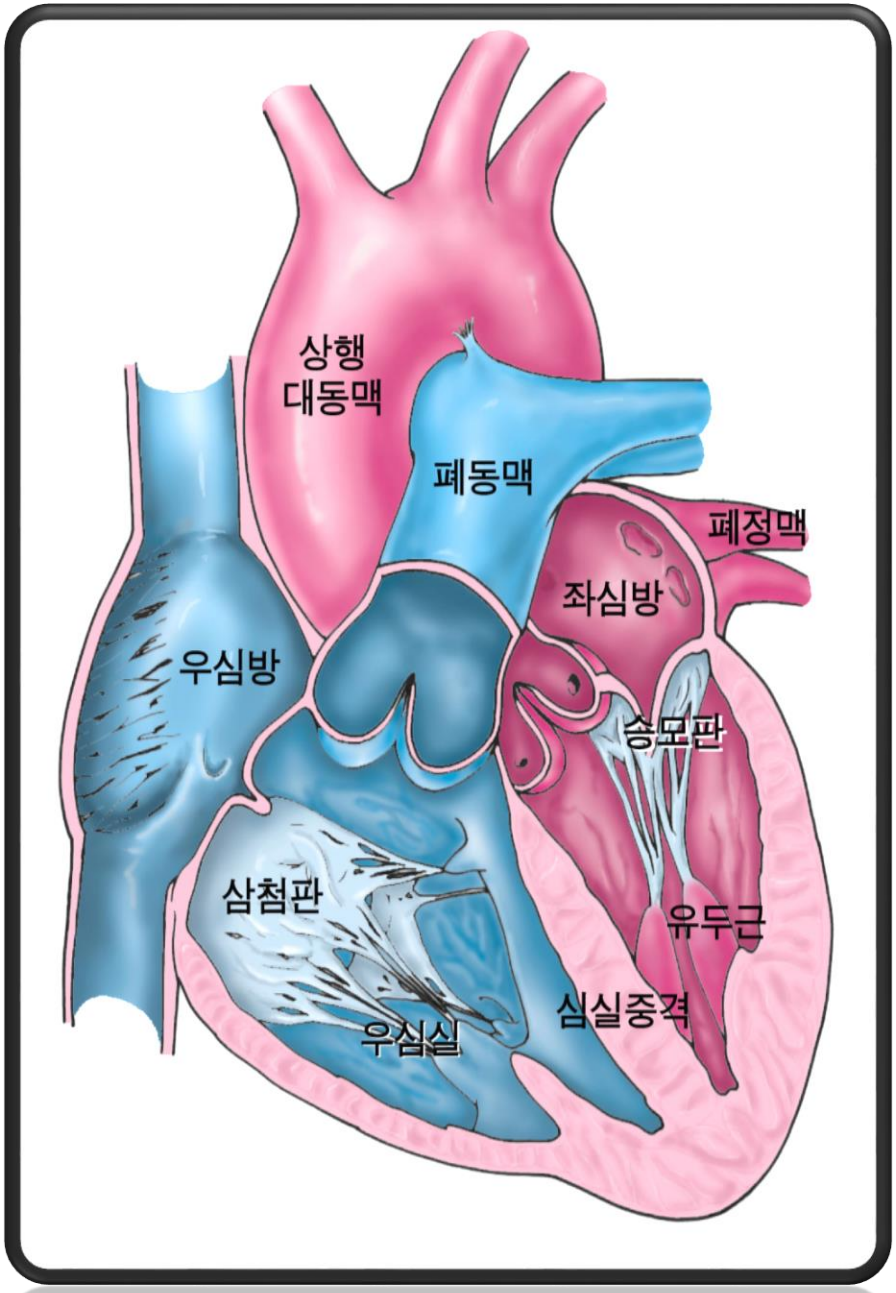


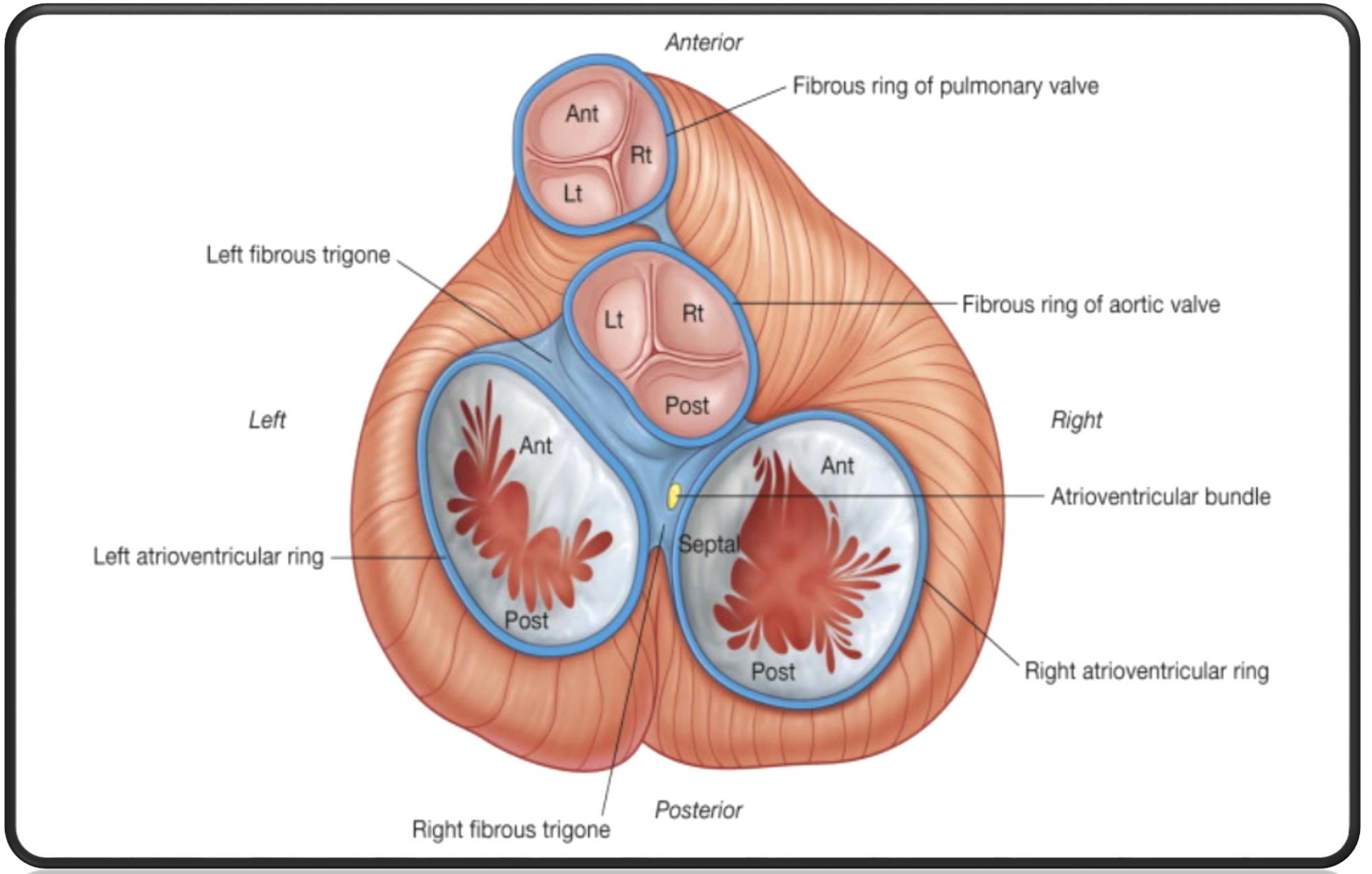


Short Axis View of the Ventricles




대동맥궁
100/60
(98%)
상행
대동맥
25-30
10
폐정맥
폐동맥
(75%)
좌심방
m=6
상대
정맥
승모판
우심방
m=3
(75%)
좌심실
100/6
(98%)
삼첨판
우심실
20-30/5
하대
정맥


상행
대동맥
폐동맥
폐정맥
좌심방
우심방
승모판
삼첨판
유두근
심실중격
우심실


Anterior
Fibrous ring of pulmonary valve
Ant
Rt
Lt
Left fibrous trigone
Lt
Rt
Fibrous ring of aortic valve
Post
Left
Right
Ant
Ant
Atrioventricular bundle
Septal
Left atrioventricular ring
Post
Post
Right atrioventricular ring
Posterior
Right fibrous trigone

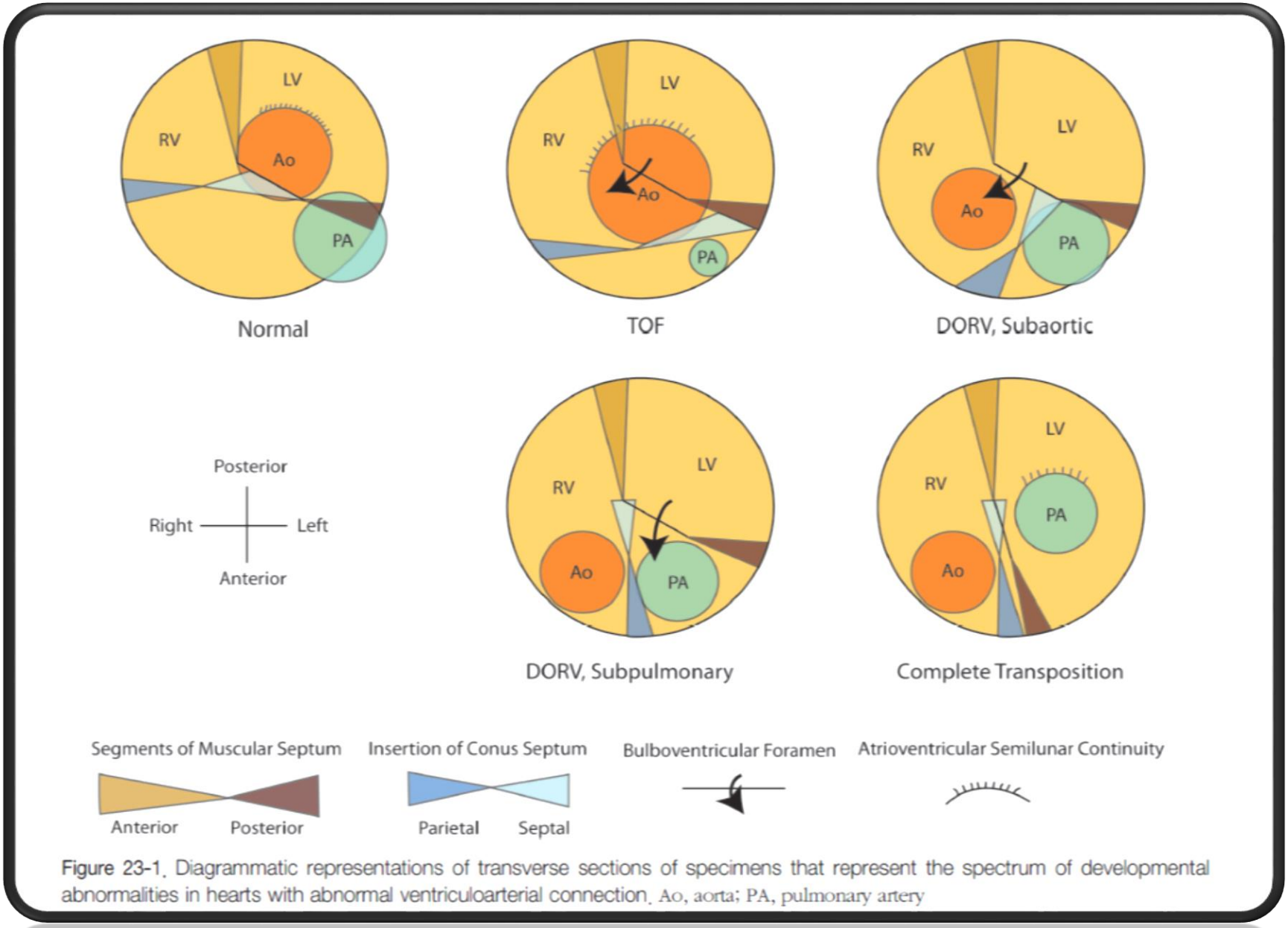


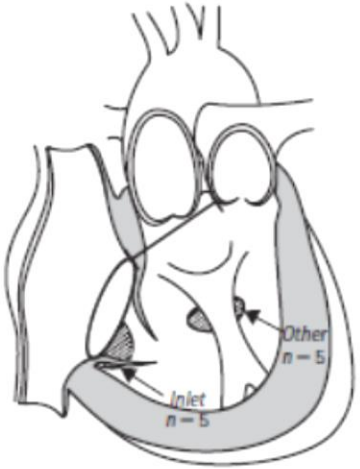
Figure 23-1. Diagrammatic representations of transverse sections of specimens that represent the spectrum of developmental abnormalities in hearts with abnormal ventriculoarterial connection. Ao, aorta; PA, pulmonary artery

Subaortic VSD

$n = 73$	SubAo VSD n (%)	SubP VSD n (%)
	31(42)	27(37)
Pulmonary stenosis	15(48)	5(19)
Aortic stenosis	4(13)	4(15)
Ao arch obstruction	1(3)	14(52)
TPD < Ao annulus	2(6)	14(52)

TPD = Tricuspid to pulmonary valve distance

Subpulmonary VSD



Noncommitted VSD

$n = 73$	Noncommitted n (%)	Doubly committed n (%)
	10(14)	5(7)
Pulmonary stenosis	5(50)	2(40)
Aortic stenosis	3(30)	3(60)
Ao arch obstruction	2(20)	2(40)
TPD < Ao annulus	4(40)	0

TPD = Tricuspid to pulmonary valve distance

Doubly committed VSD

Figure 23.9 *Important anatomic features of VSD groups in a review of 73 patients who underwent biventricular repair of DORV between 1981 and 1991 at Children's Hospital Boston. (From Aoki* et al. *Results of biventricular repair for double outlet right ventricle.* J Thorac Cardiovasc Surg, *1994; 107:340, with permission from Elsevier.)*

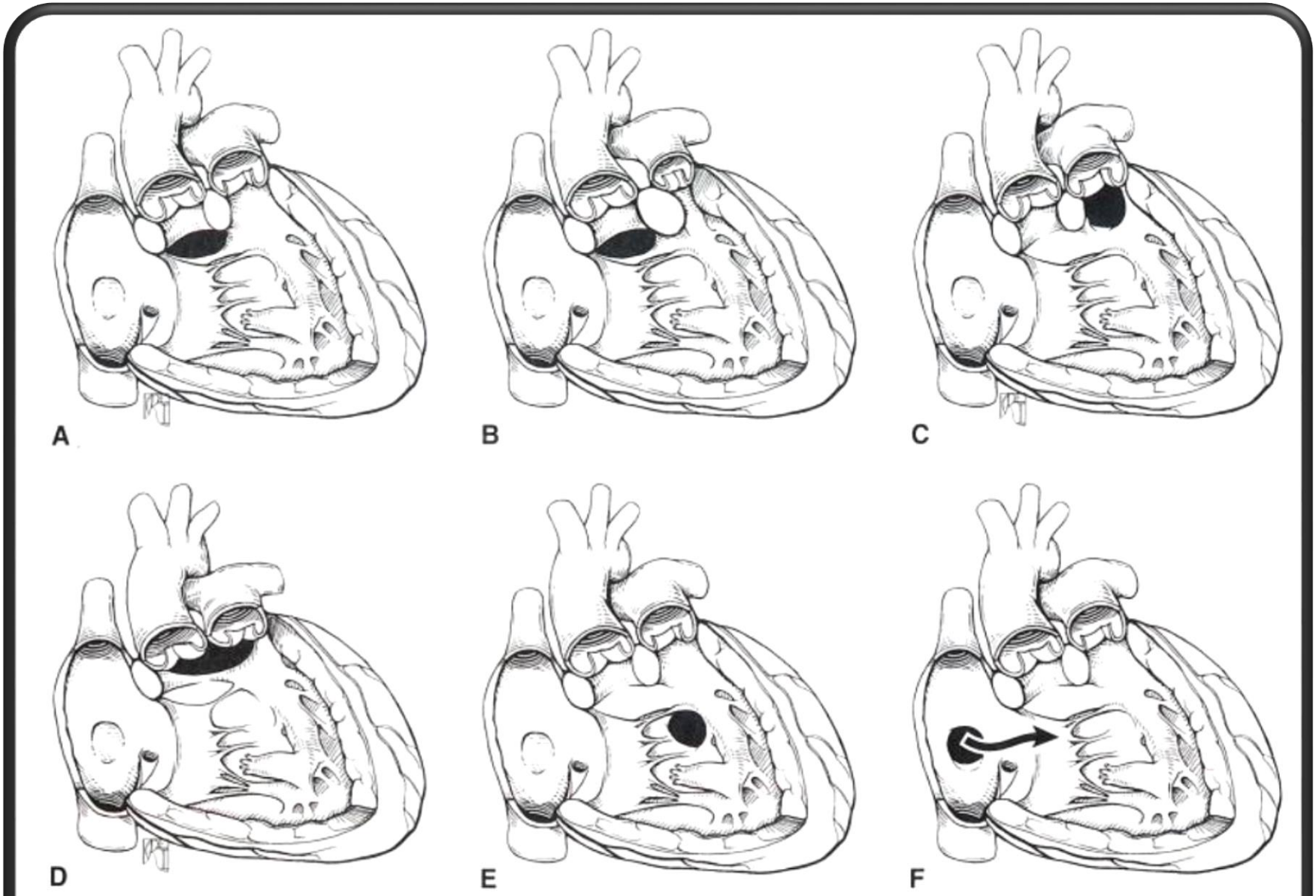


Fig. 24-3 The relationship of the VSD to the great arteries in DORV. **A,** Subaortic VSD without pulmonary stenosis. **B,** Subaortic VSD with pulmonary stenosis. **C,** Subpulmonary VSD (Taussig-Bing malformation). **D,** Doubly-committed VSD. **E,** Noncommitted (remote) VSD. **F,** Intact interventricular septum.

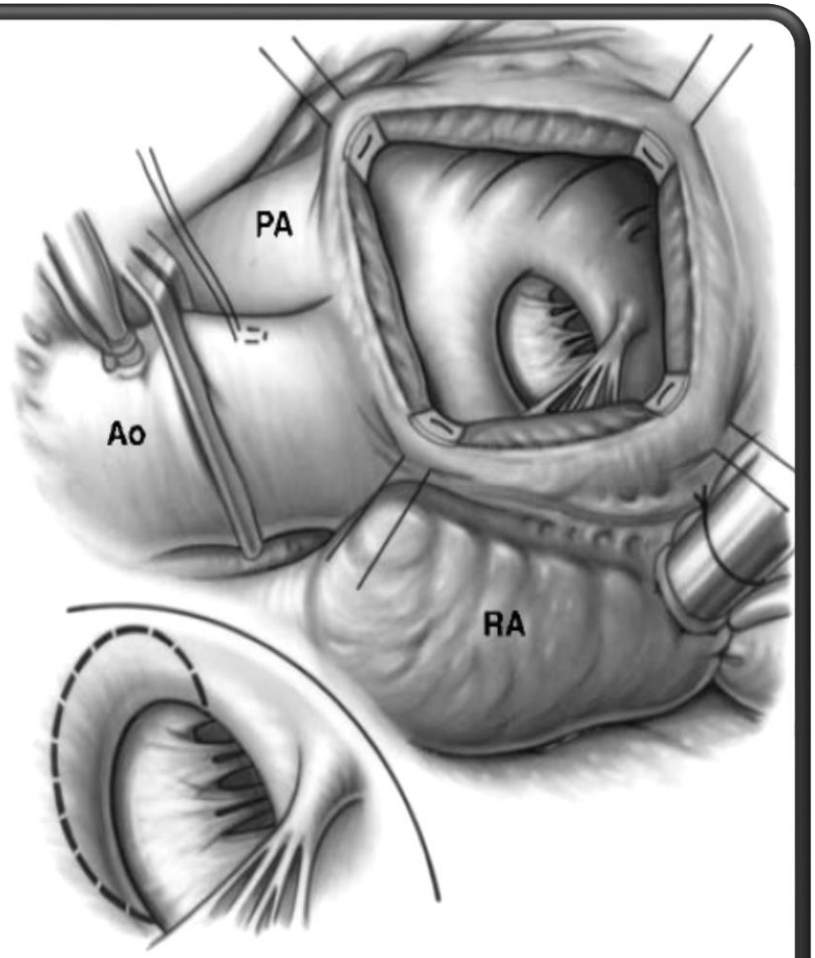


Figure 32.2

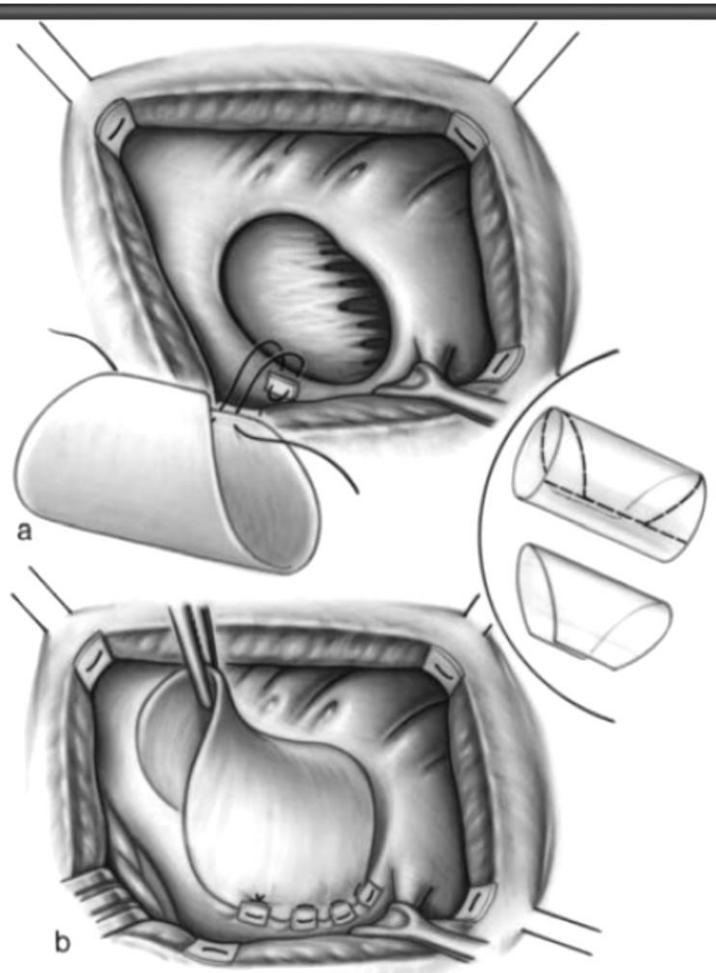


Figure 32.3

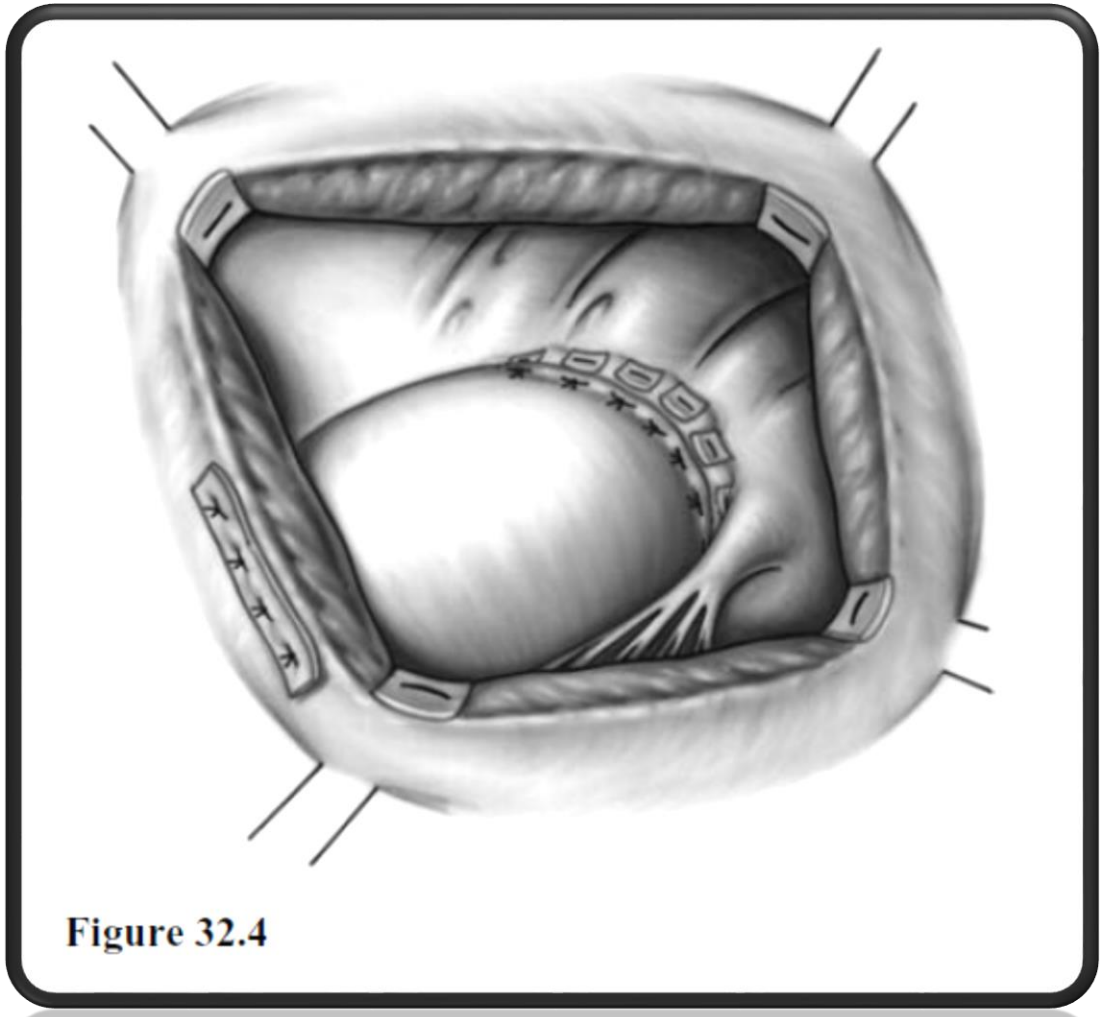


Figure 32.4

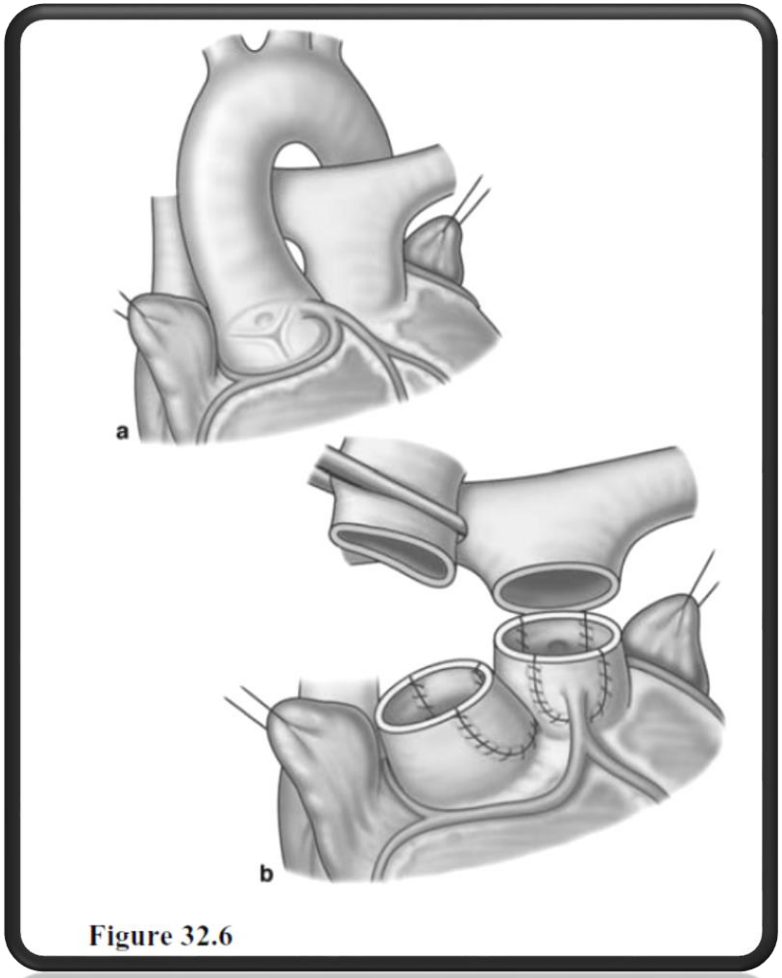


Figure 32.6

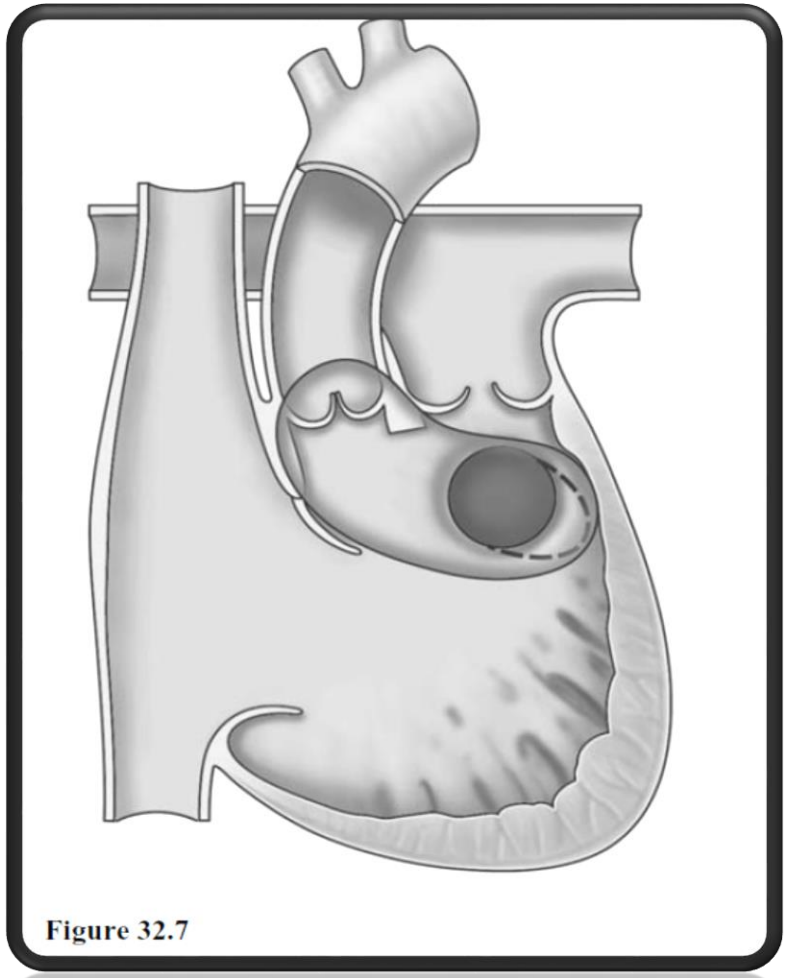


Figure 32.7

TV-PV distance

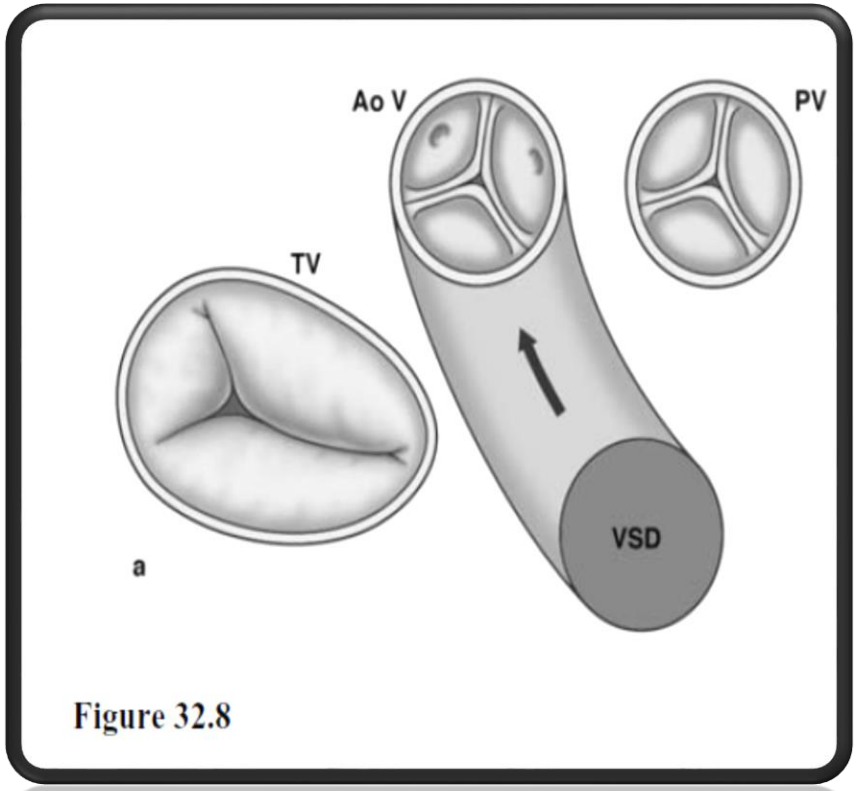
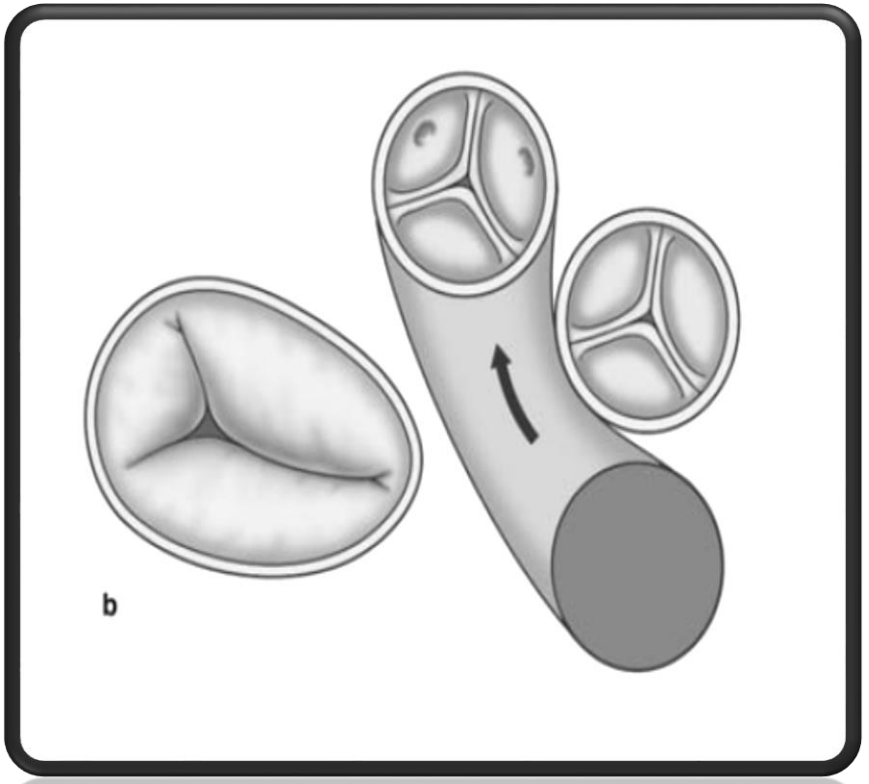


Figure 32.8



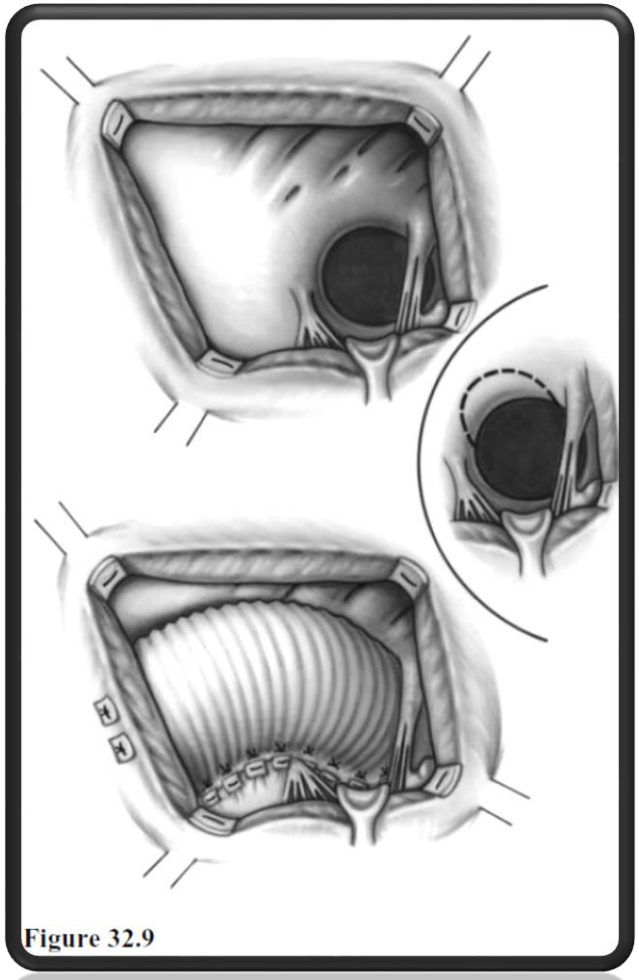


Figure 32.9

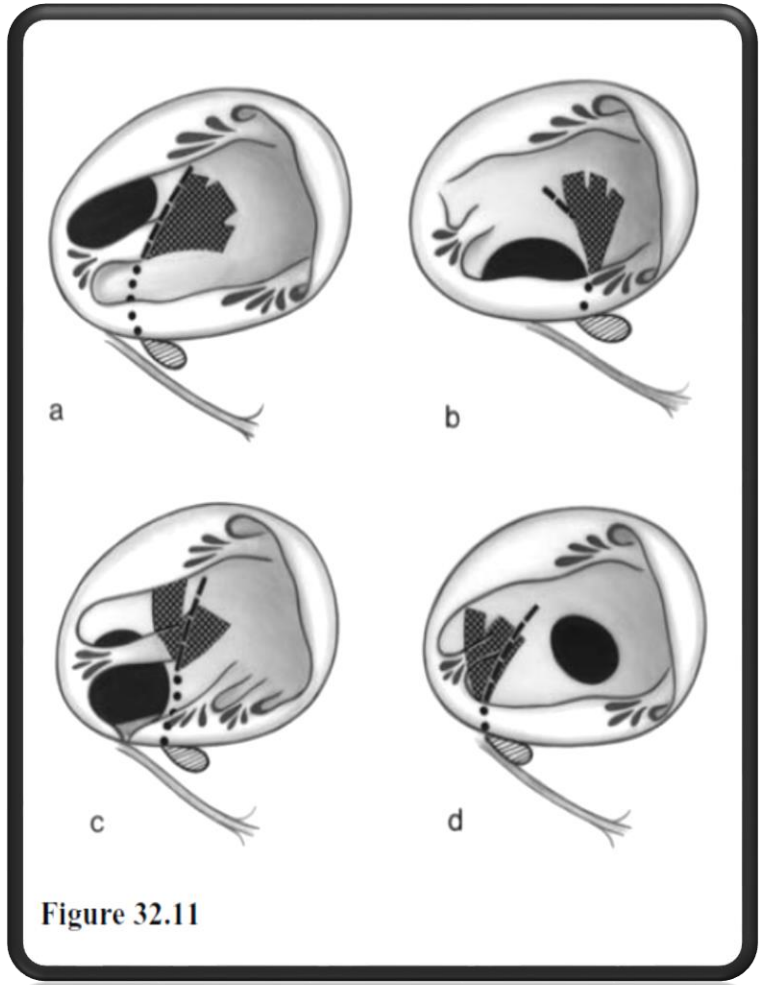


Figure 32.11

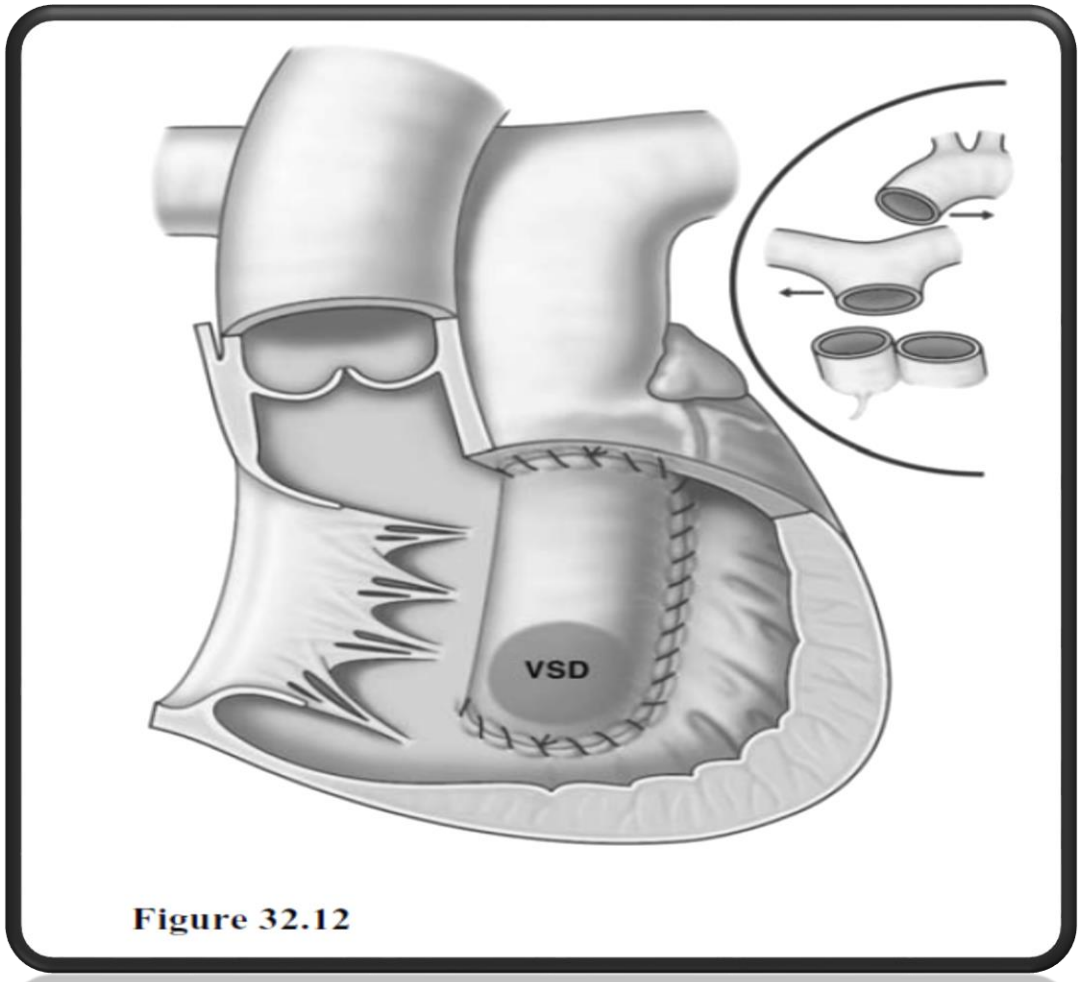


Figure 32.12

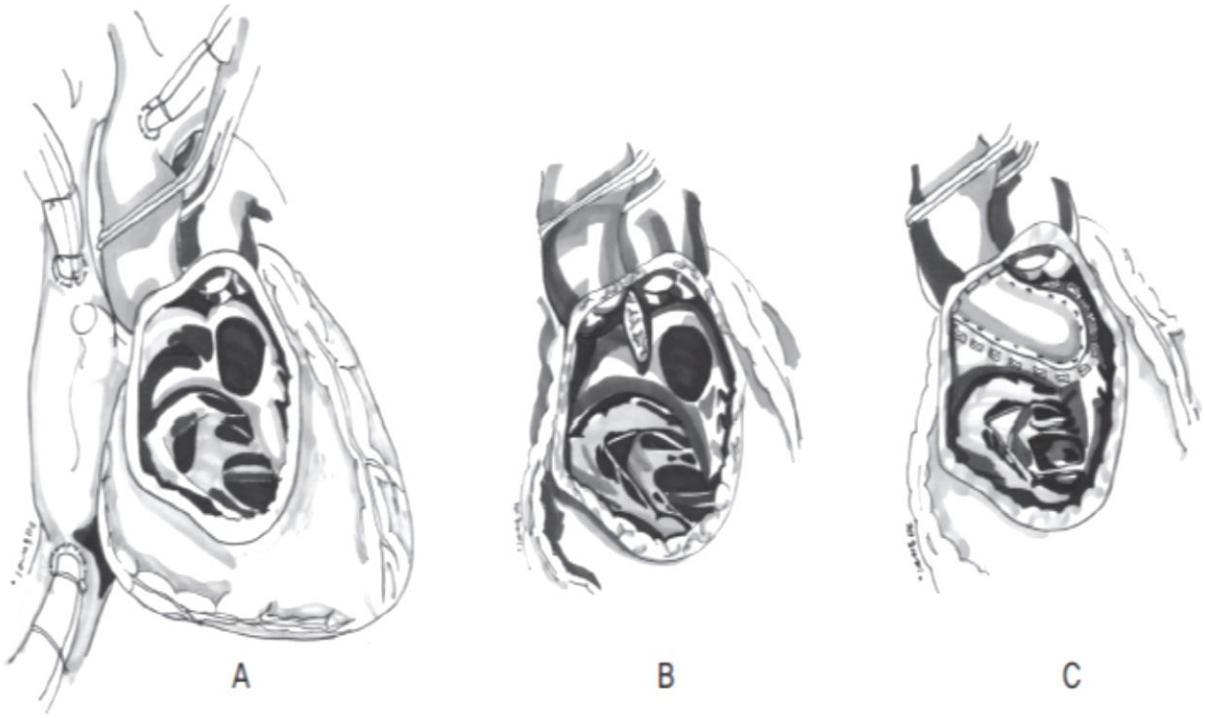


Figure 23-3. Kawashima Intraventricular repair: Artist's cut-away view of the Taussig-Bing heart with side-by-side great arteries. (A) The subpulmonic ventricular septal defect (VSD) and subaortic conus are represented in relationship to the great arteries and semilunar valves. (B) The excised subaortic conus is shown in preparation for (C) the left ventricular-to-aorta Intraventricular tunnel, shown here contructed with a Dacron patch and interrupted pledged sutures.

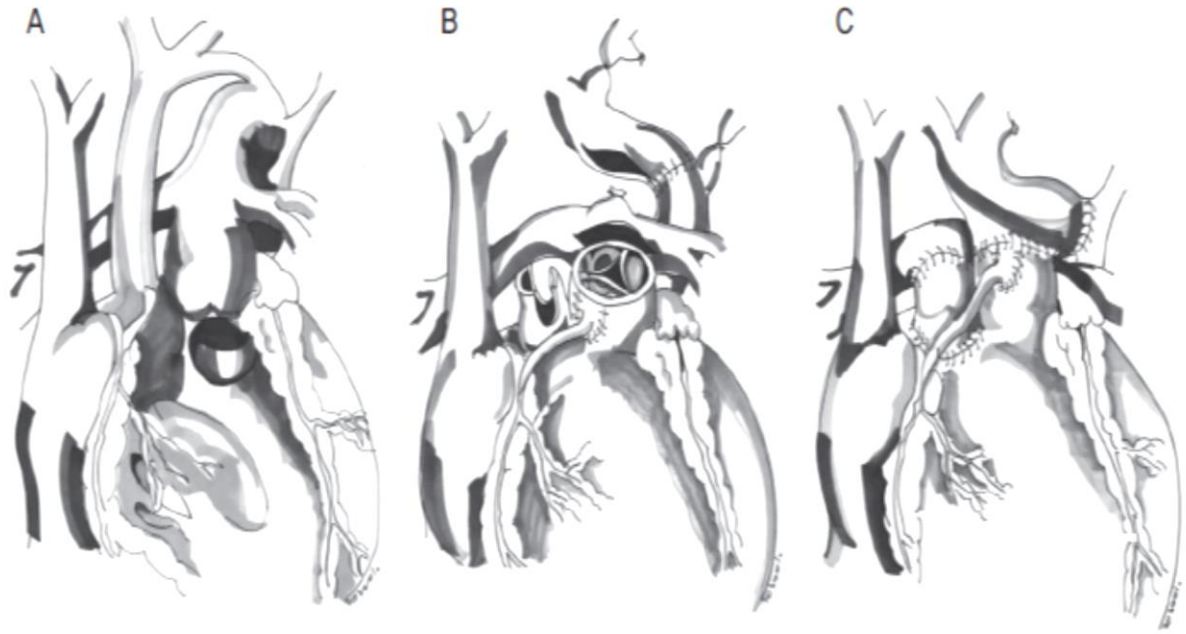


Figure 23-4. Arterial switch, ventricular septal defect closure, and aortic relocation: one-stage neonatal repair of (A) the Taussig-Bing heart with subaortic obstruction, small ascending aorta, and coarctation of the aorta. (B) After great artery transection and coronary artery transfer are accomplished, the coarctation is repaired under circulatory arrest by coarctectomy with end-to-end anastomosis between the "proximal" ascending and descending aorta. A longitudinal incision is made in the lateral ascending aorta, which is now oriented in a transverse position for anastomosis to the neoaorta. (C) Completed repair showing the neopulmonary artery reconstruction with a valved homograft after subvalvular resection.

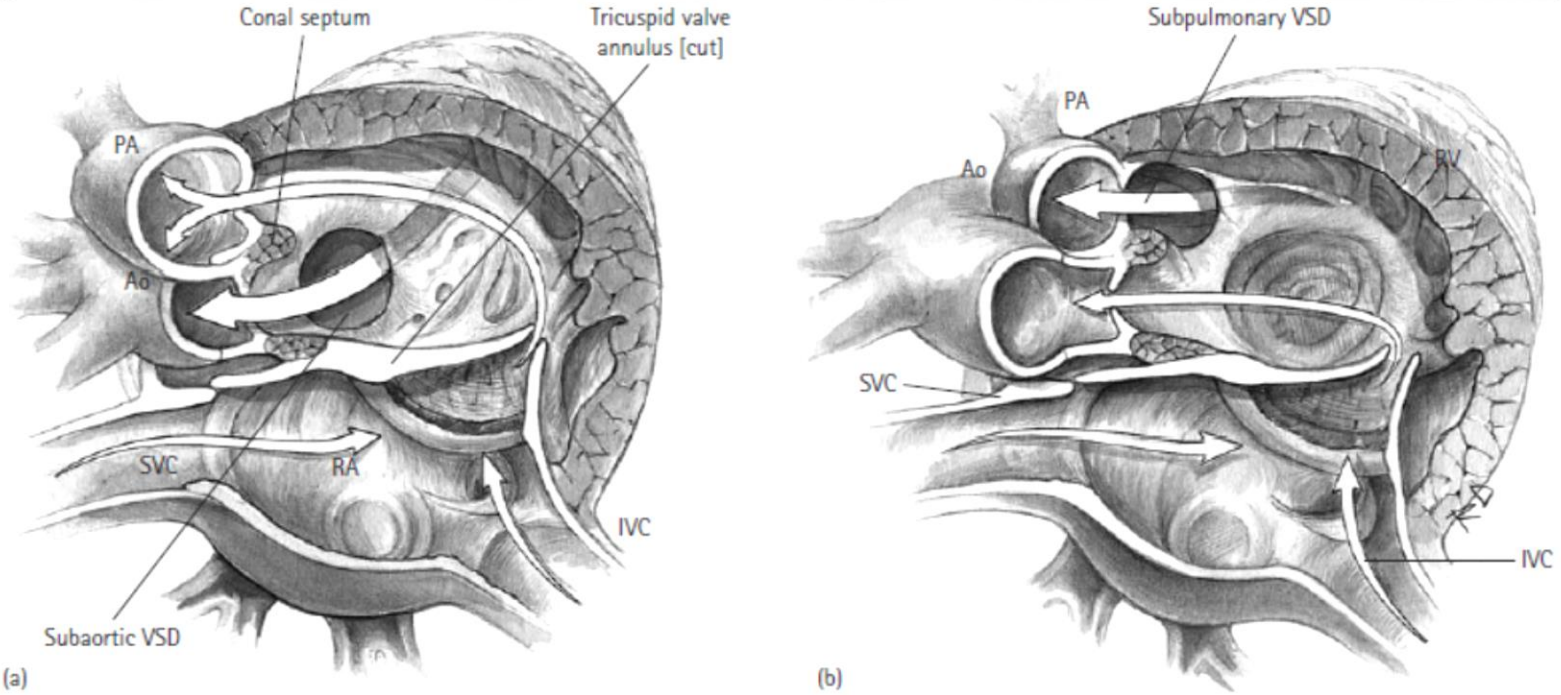


Figure 23.2 *(a) Double outlet right ventricle with subaortic VSD. Physiologically this anomaly is similar to tetralogy of Fallot. Left ventricular blood is predominantly directed to the aorta after passing through a subaortic VSD. (b) Double outlet right ventricle with subpulmonary VSD. Physiologically this anomaly is similar to transposition of the great arteries. Left ventricular blood passes through the subpulmonary VSD into the pulmonary arteries so that oxygen saturation is higher in the pulmonary artery than in the aorta.*

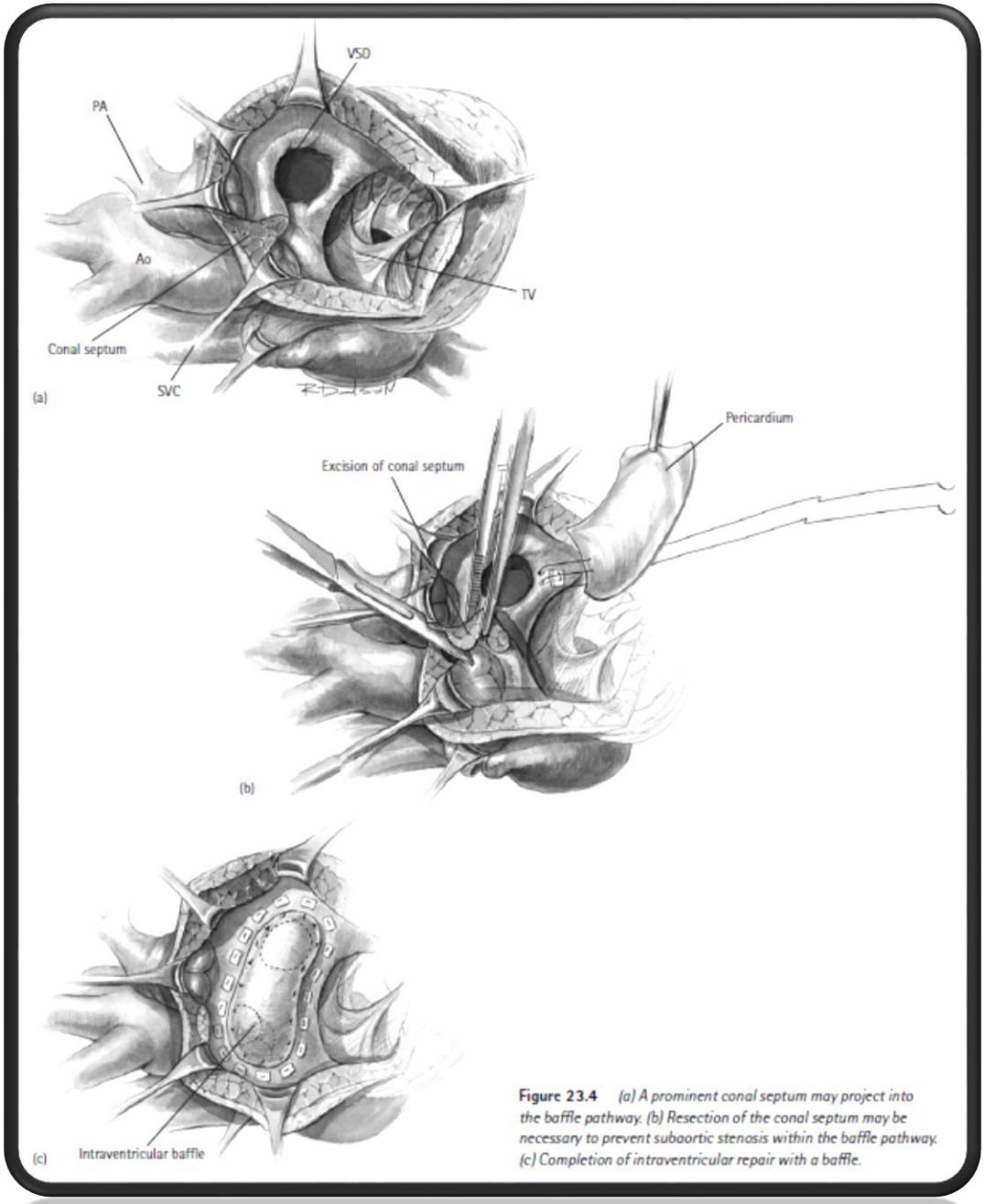


Figure 23.4 *(a) A prominent conal septum may project into the baffle pathway. (b) Resection of the conal septum may be necessary to prevent subaortic stenosis within the baffle pathway. (c) Completion of intraventricular repair with a baffle.*

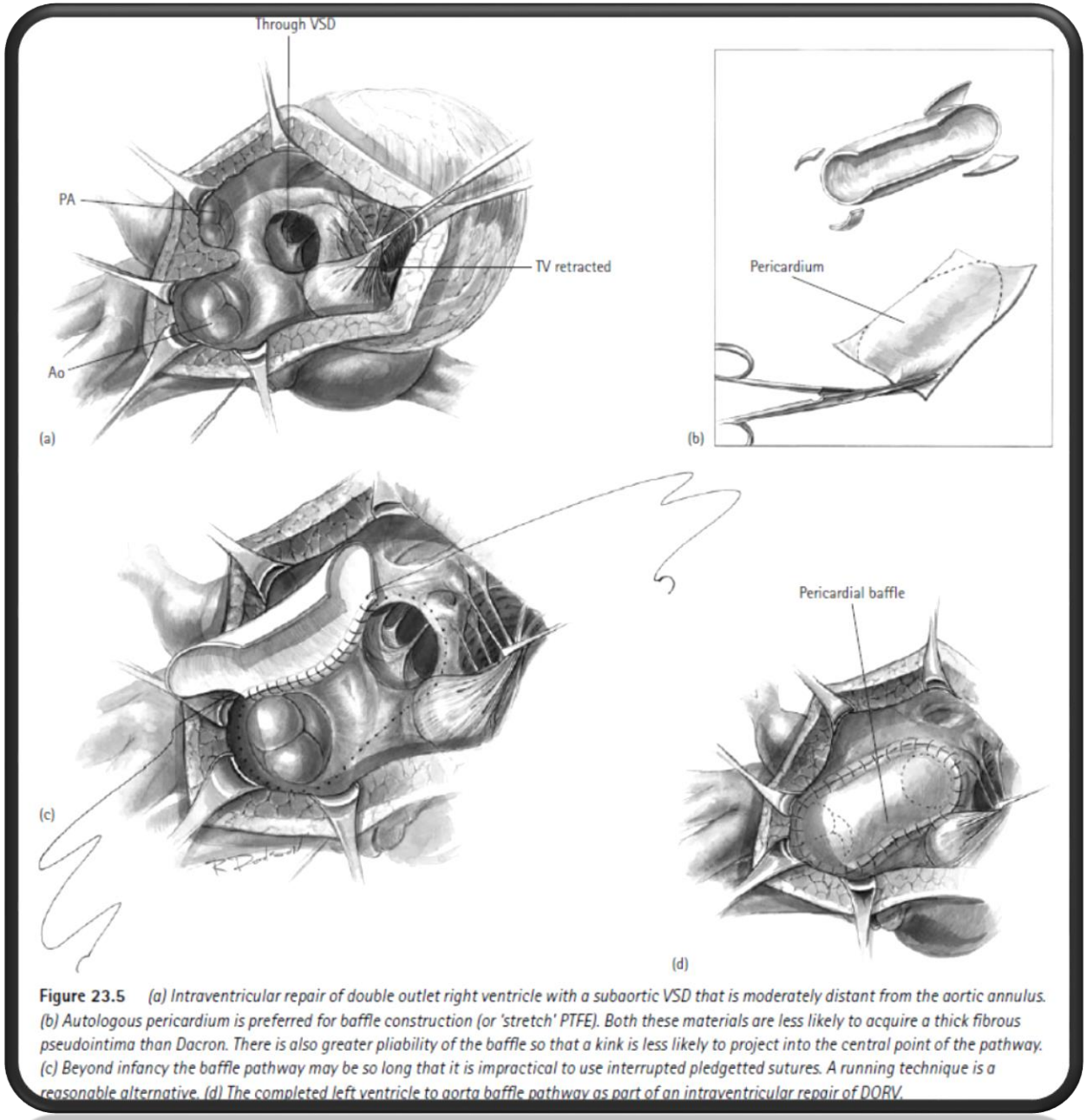


Figure 23.5 *(a) Intraventricular repair of double outlet right ventricle with a subaortic VSD that is moderately distant from the aortic annulus. (b) Autologous pericardium is preferred for baffle construction (or 'stretch' PTFE). Both these materials are less likely to acquire a thick fibrous pseudointima than Dacron. There is also greater pliability of the baffle so that a kink is less likely to project into the central point of the pathway. (c) Beyond infancy the baffle pathway may be so long that it is impractical to use interrupted pledgetted sutures. A running technique is a reasonable alternative. (d) The completed left ventricle to aorta baffle pathway as part of an intraventricular repair of DORV.*

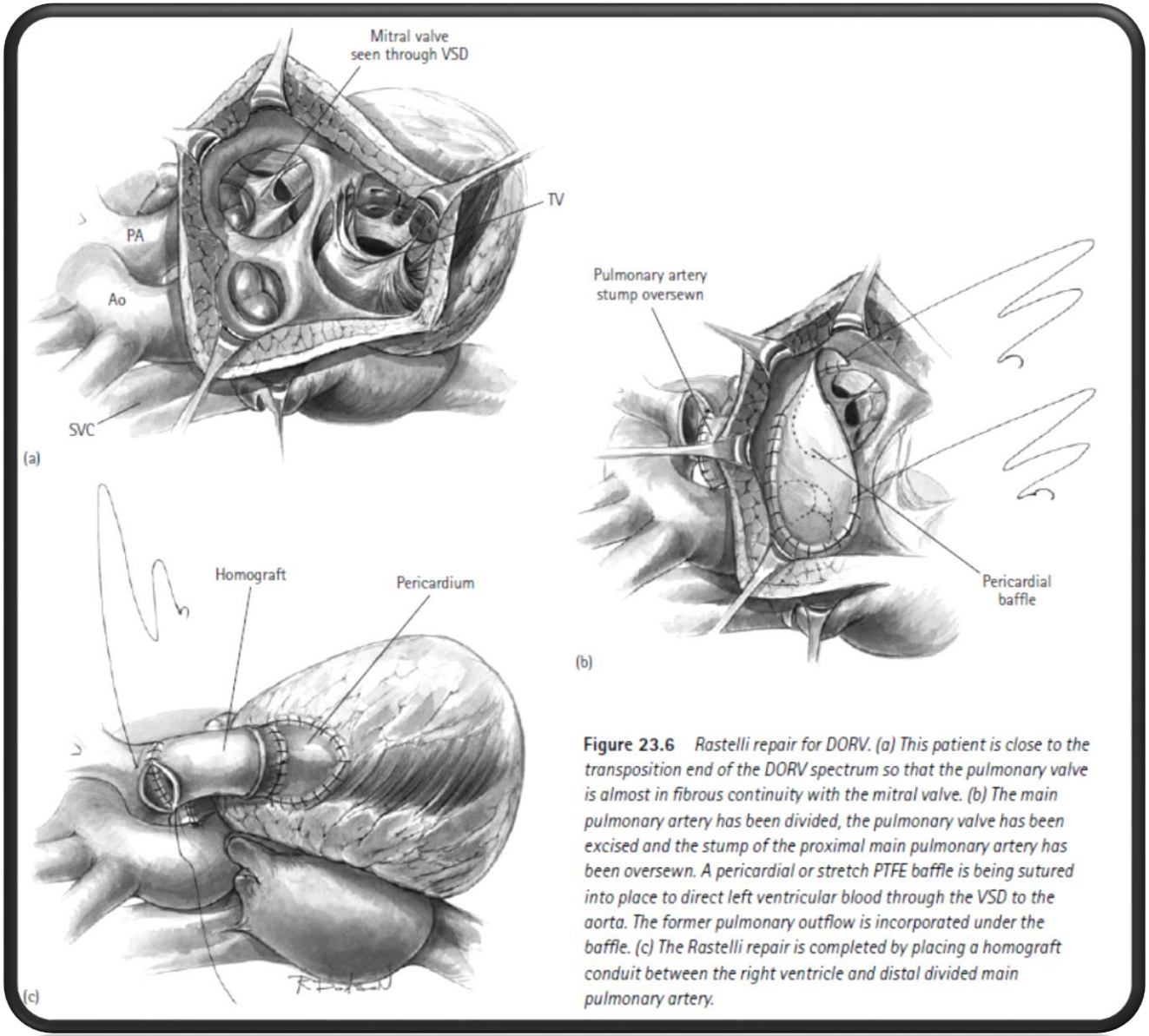


Figure 23.6 *Rastelli repair for DORV. (a) This patient is close to the transposition end of the DORV spectrum so that the pulmonary valve is almost in fibrous continuity with the mitral valve. (b) The main pulmonary artery has been divided, the pulmonary valve has been excised and the stump of the proximal main pulmonary artery has been oversewn. A pericardial or stretch PTFE baffle is being sutured into place to direct left ventricular blood through the VSD to the aorta. The former pulmonary outflow is incorporated under the baffle. (c) The Rastelli repair is completed by placing a homograft conduit between the right ventricle and distal divided main pulmonary artery.*

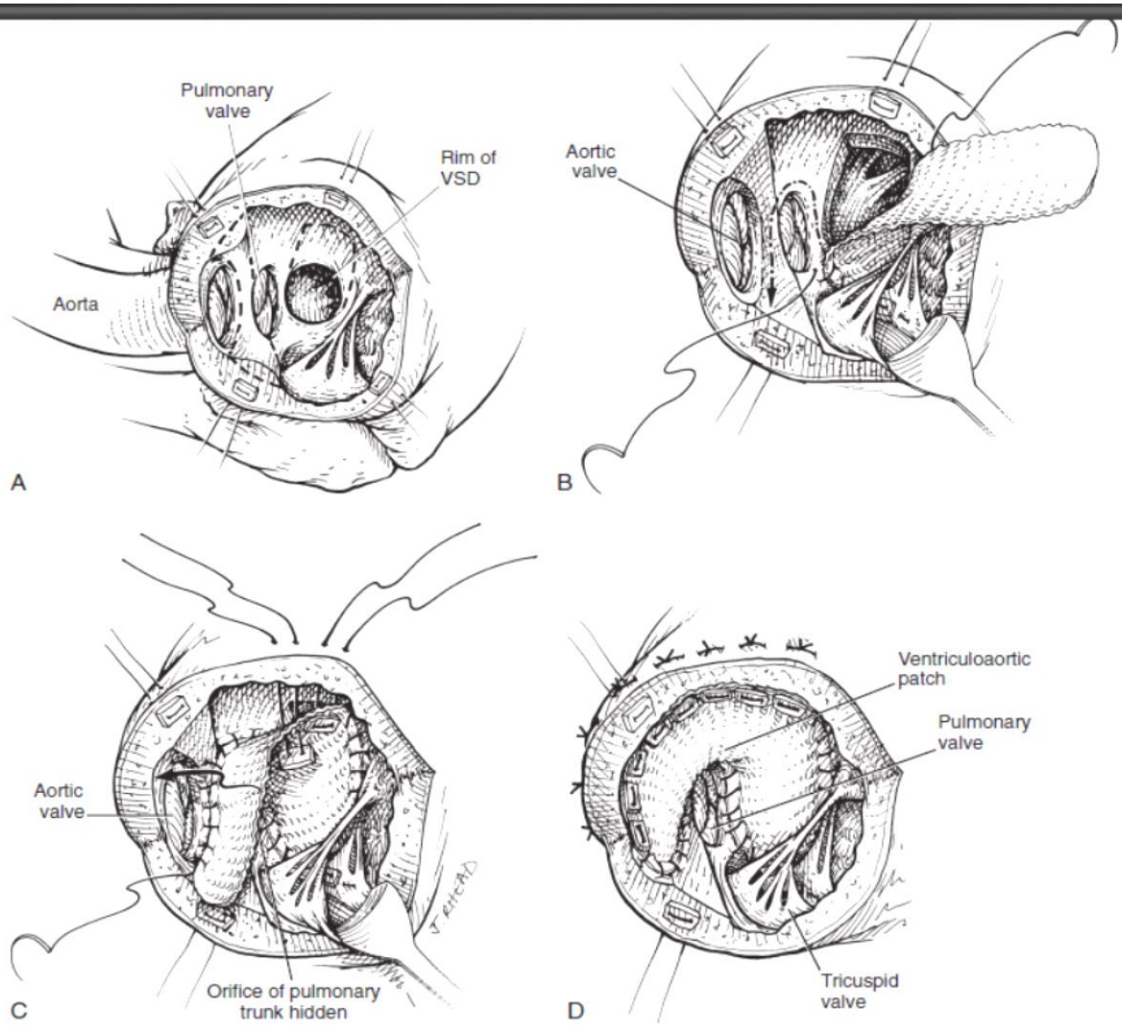
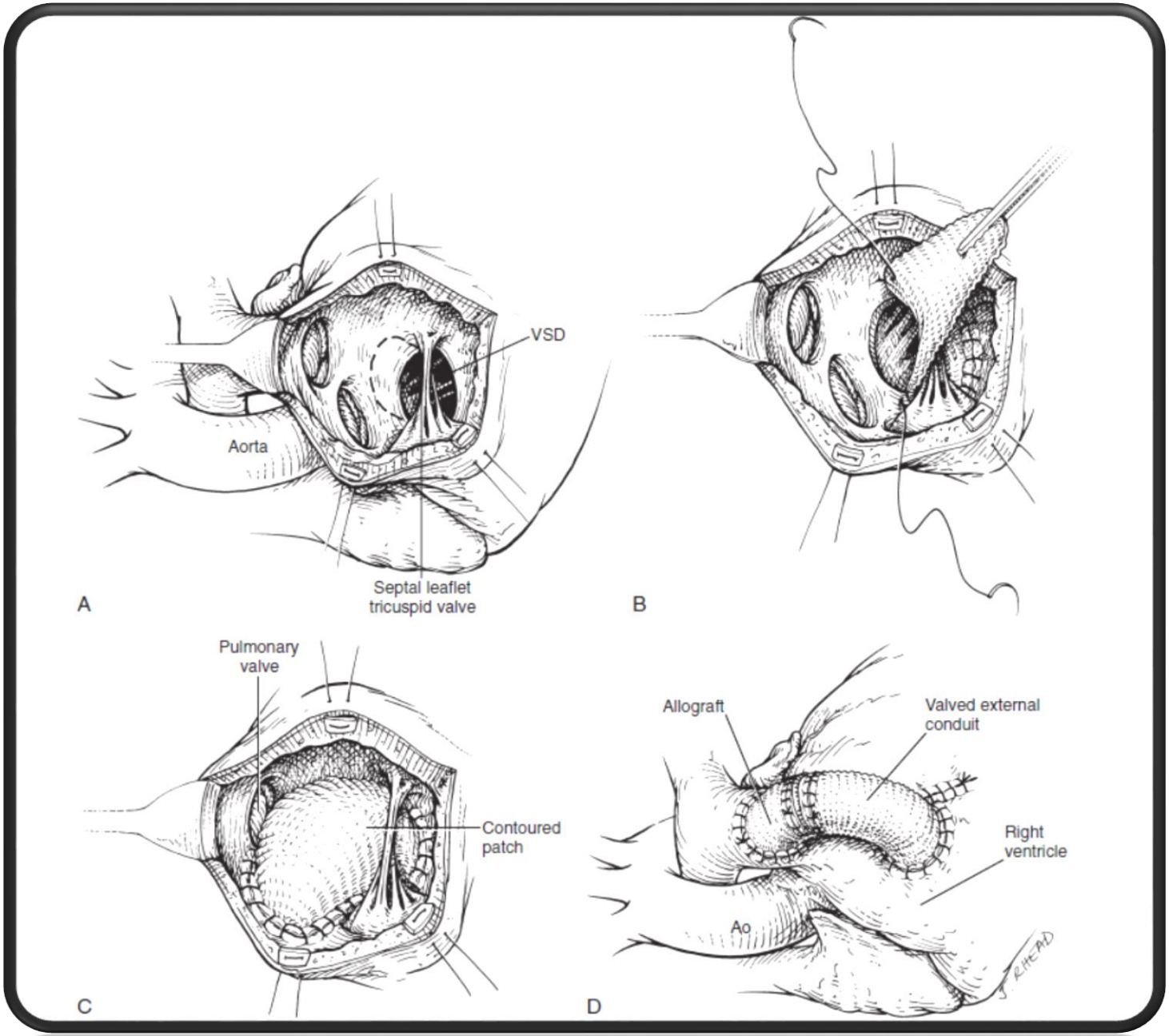


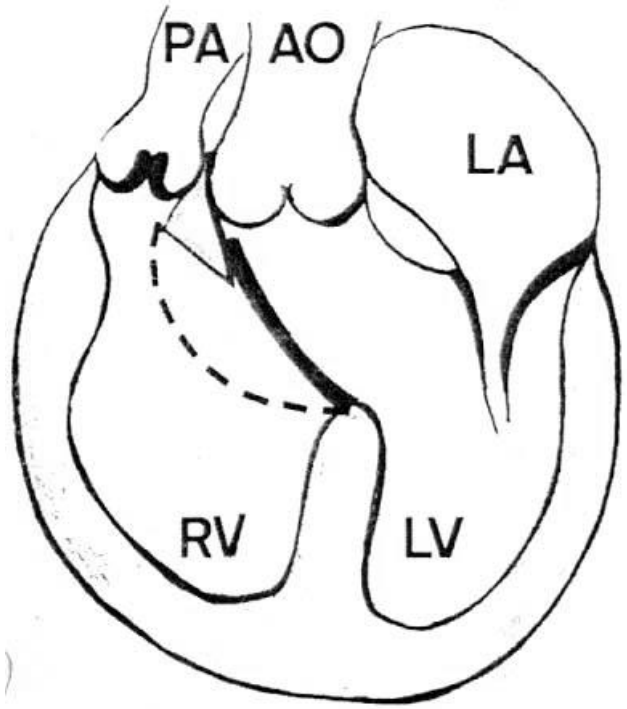
Figure 53-15 Repair of double outlet right ventricle with subpulmonary ventricular septal defect *(VSD)* (Taussig-Bing heart) when there is only a short distance between tricuspid and pulmonary valves (Patrick-McGoon method[M9,P1,P3]). Aorta is usually anterior to pulmonary trunk. **A,** Through a transverse right ventriculotomy, the VSD is enlarged and much of the inlet septum excised *(dashed line)*. **B,** A contoured polyester (or polytetrafluoroethylene) patch is cut from a tube graft whose diameter is about 20% larger than that of the aorta. Initial suturing is performed in the usual manner, preventing damage to the bundle of His. A continuous suture is used, often supplemented by interrupted stitches. **C,** With the leftward arm of the continuous suture, suture line is continued leftward along the posterior and then leftward margin of the ventriculopulmonary trunk junction, and finally along the anterior margin of this junction *(dashed line)*. With the other arm of the suture, the patch is sutured inferior to the VSD, to the right side of the septum, and then anterior to the defect. **D,** Insertion of spiraled patch has been completed. Left ventricular blood now passes beneath the patch to the aorta, while right ventricular blood passes to the pulmonary trunk behind the tunnel.


VSD
Aorta
Septal leaflet
tricuspid valve
A
B
Pulmonary
valve
Contoured
patch
C
Allograft
Valved external
conduit
Right
ventricle
Ao
D

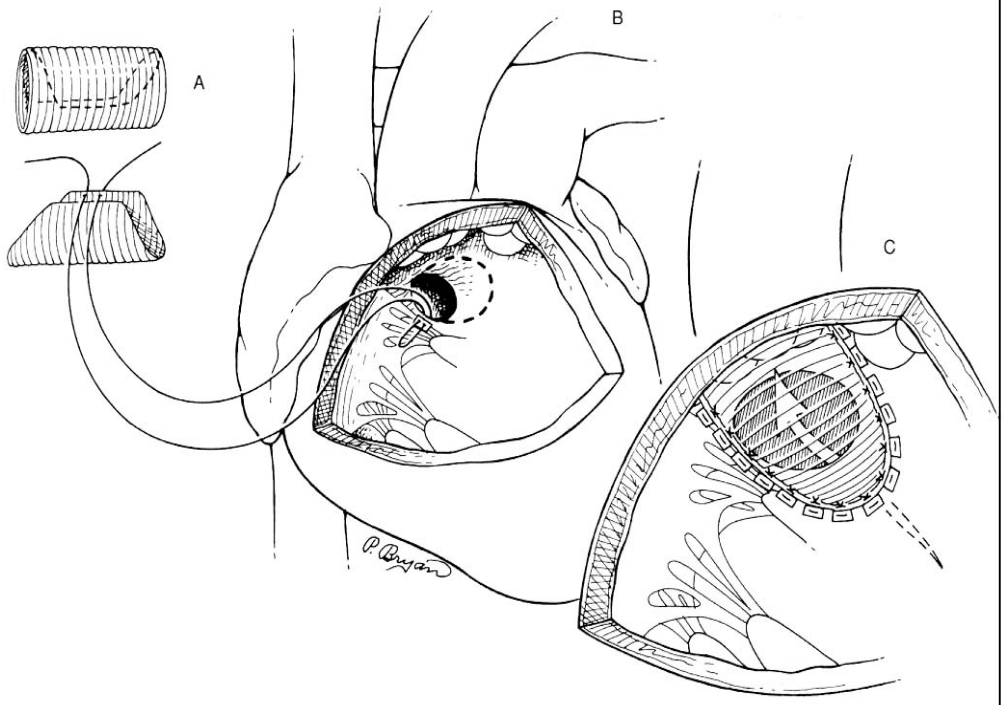
Surgery and timing for repair

VSD type	About 3 months	Intraventricular repair (IVR)
TOF type	3 - 6 month	IVR + RVOT rec.
TGA type	Early and late infancy	•ASO + VSD tunnel •Senning/Mustard + VSD tunnel •DKS+VSD tunnel+RV-PA •REV •Kawashima •Nikaidoh
Remote VSD	Infancy? After palliation?	•Biventricular repair •Univentricular repair

Intraventricular repair



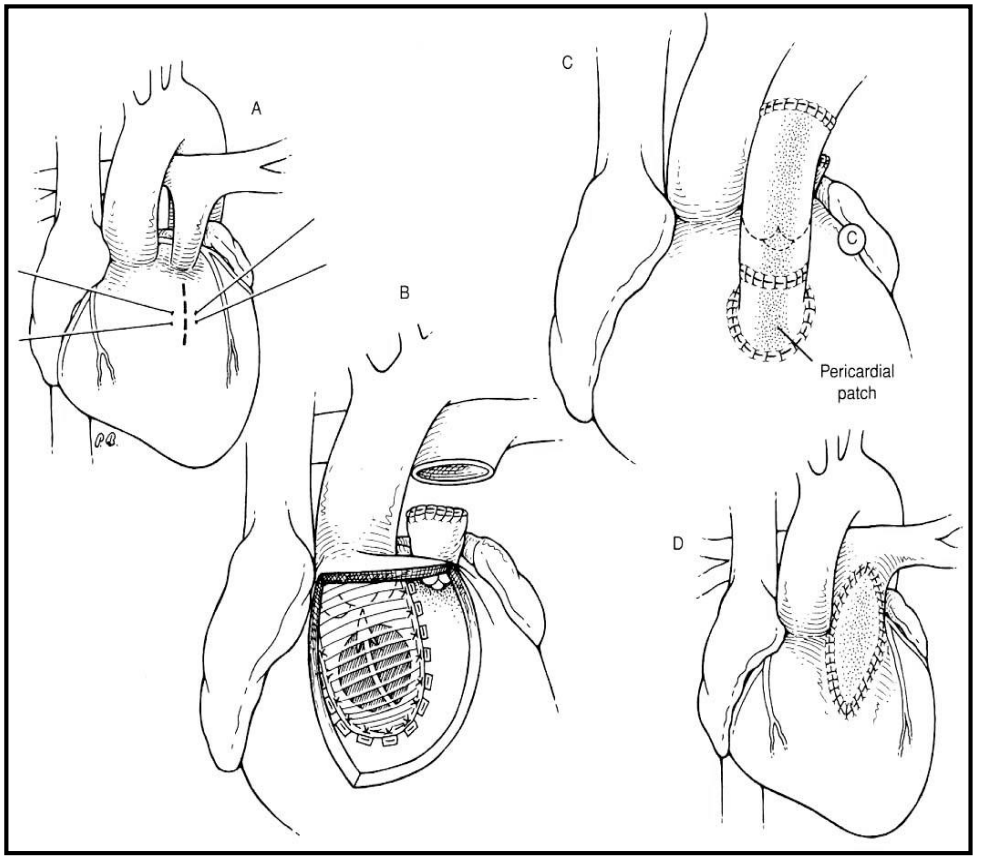
Conal resection



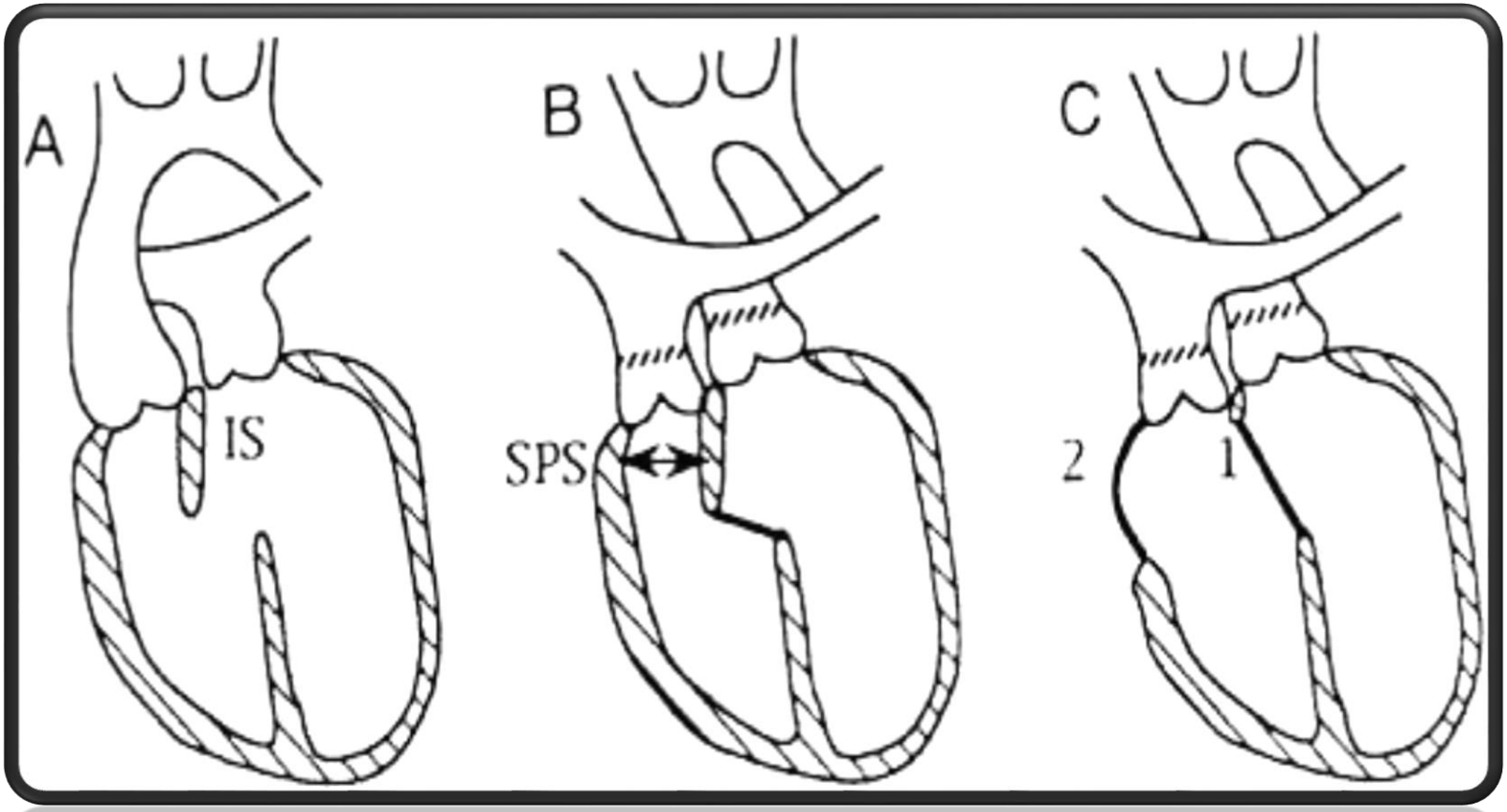
VSD extension

DORV with subaortic VSD, PS

- Valved conduit
  - Major coronary arteries crossing RV
  - High PVR or distal pulmonary obstruction
- Transannular patch



DORV, subpulmonic VSD (Taussig-Bing Anomaly)



DORV, subpulmonic VSD (Taussig-Bing Anomaly)

Taussig-Bing 기형에서의 대혈관치환술은 완전대혈관전위증과 비교하여 몇 가지 차이점이 있다.

1) 관상동맥 전이 방법의 차이이다. 완전대혈관전위증의 경우 통상적인 관상동맥 배열이 흔한 반면, Taussig-Bing 기형의 경우 다 양하고 복잡한 형태의 관상동맥 배열 (single coronary artery, itramural coronary artery) 이 관찰된다

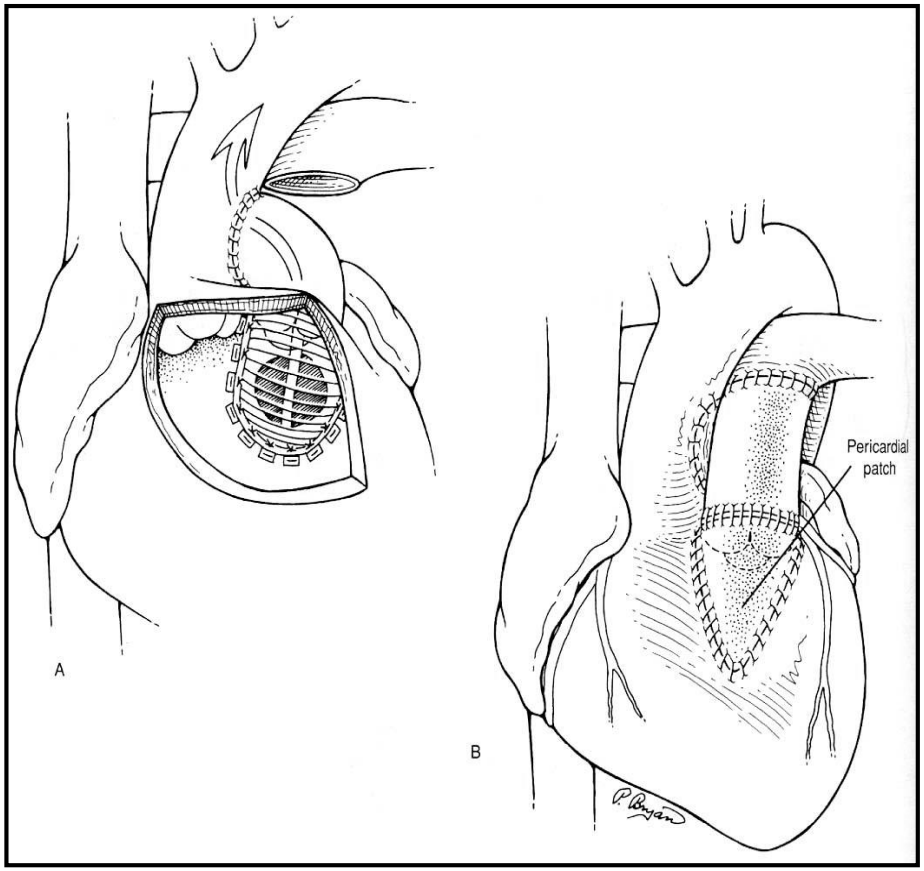
2) Taussig-Bing 기형에서 대혈관의 배열이 다양한 데, 측측 배열의 대혈관에서 Lecompte 수기(maneuver)의 시행 여부이다

3) Taussig-Bing 기형에서 대동맥과 폐동맥의 심한 크기 차이를 보인다 상행대동맥이 폐동맥에 비해 심하게 작으며, 특히 대동맥궁협착이 있는 경우 차이는 더 심해진다

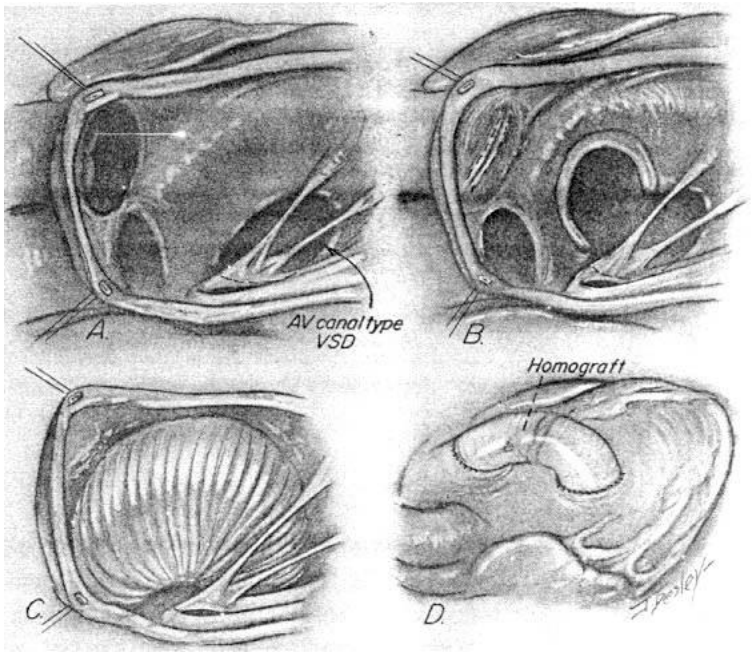
4) 대동맥하 협착, 대동맥 축착의 동반이 많다

Damus-Kaye-Stansel procedure, tunnel closure of VSD and RV-PA conduit

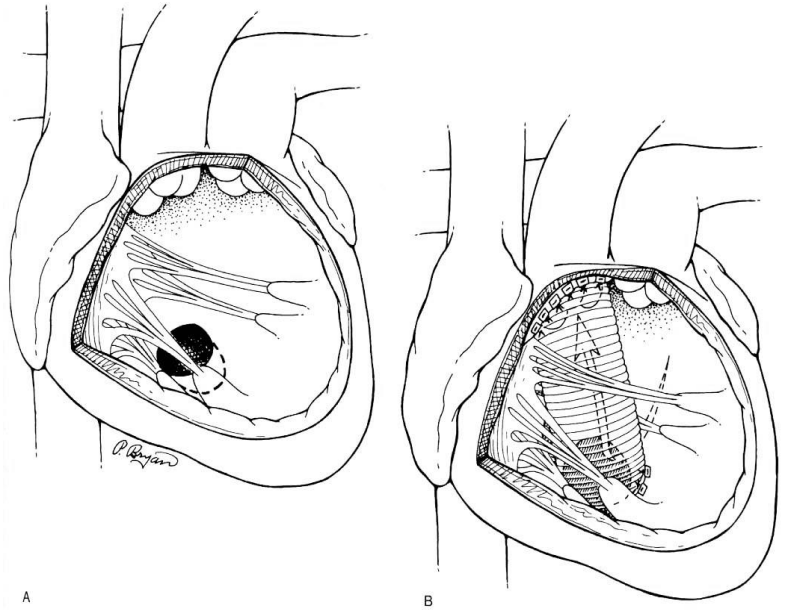
- Significant subaortic stenosis
- To risky to perform arterial switch (intramural coronary artery, single coronary artery)
- Aortic valve insufficiency - aortic valve should be closed



DORV with noncommitted VSD



Intraventricular tunneling



Intraventricular conduit

DORV with noncommitted VSD

- Uni-ventricular repair

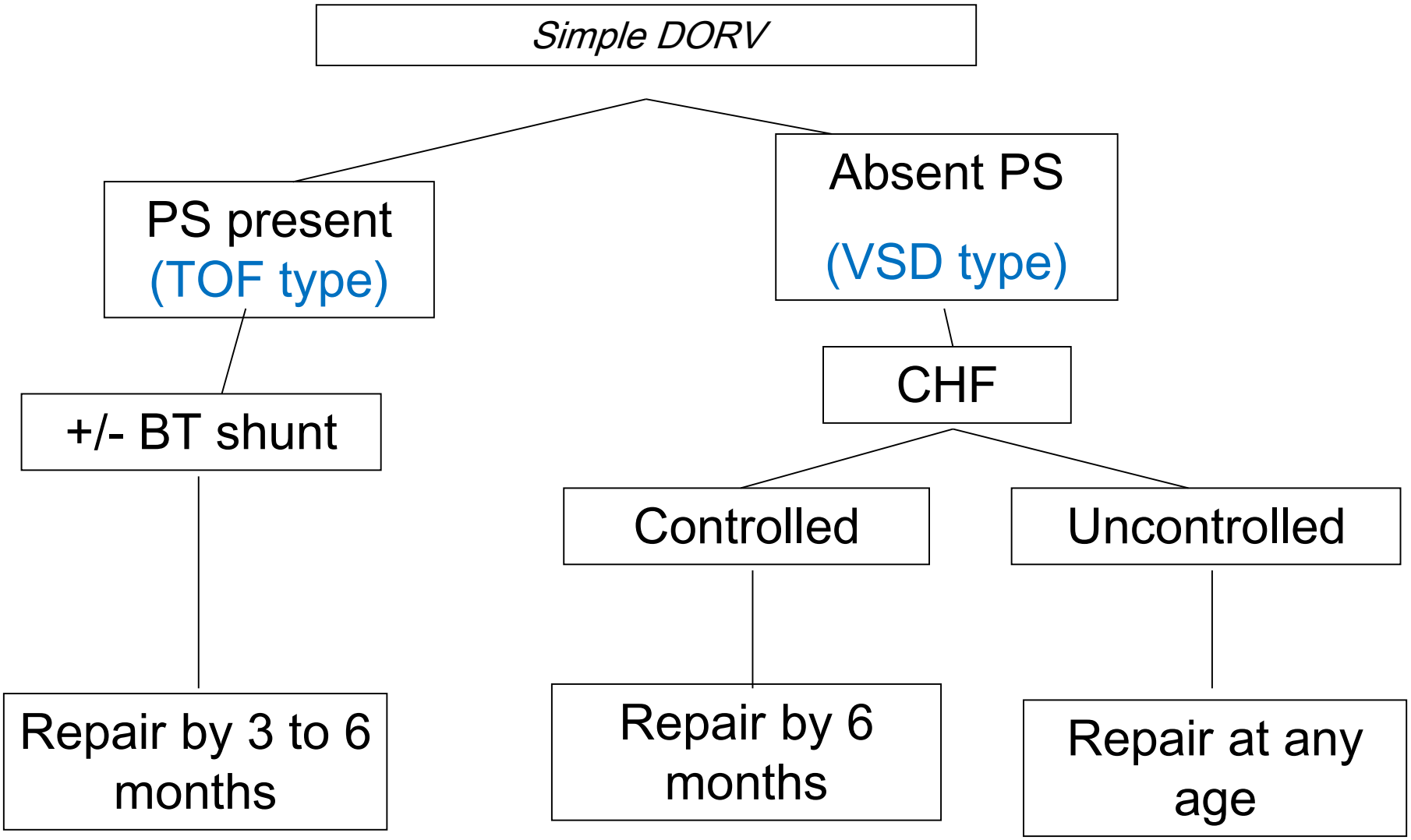
  Anomalies of the atrioventricular valves

  Multiple muscular VSDs

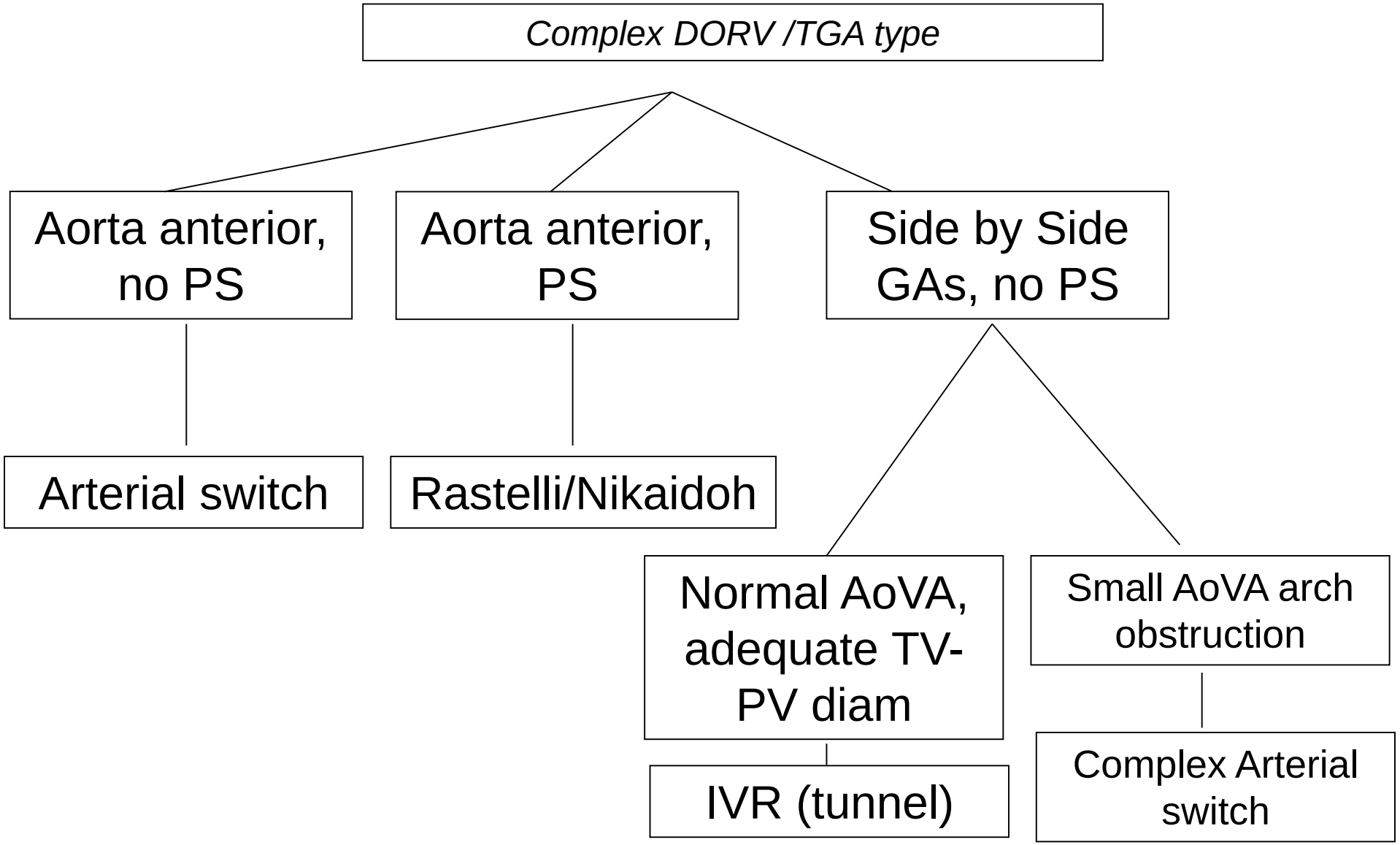
  Inability to reliably channel the remote VSD to aorta

  Hypoplasia of the right or left ventricle

DORV decision tree



DORV decision tree



Complication

- Heart block
- Residual left-to-right shunt
- LVOT obstruction
  - Inadequate enlargement of VSD
  - Poor configuration of intraventricular tunnel patch
- RVOT obstruction
  - Obstructing muscle bundle or patch
- Myocardial dysfunction
- Coronary ischemia

Table 23-1. Summary of surgical treatment of remote DORV

Authors (year)	Time period of operations	No of patients	Previous palliations	Techniques of repair	Age at repair	Early death	Follow-up	Late death	Reoperation
Belli (1999)[41]	1987-1997	23	9 PAB (7) COA repair (5) BTS (2)	IVR 21 ASO 2	20no (50d–10yr)	2 (9%)	58mo	No	8 (35%) Subaortic stenosis (6) TV repair + RVOTR(1) AVR (1)
Barbero-Marcial (1999)[43]	1987-1999	18	7 BTS (4) PAB (3)	IVR (Multiple patches)	4.7yr (2mo-13yr)	2 (11%)	2.7yr (1mo-12yr)	3 (16.6%)	MV repair (1)
Lacour-Gayet (2002)[77]	1998-2001	10	7 PAB	ASO	16mo (3wk-4.5yr)	1	20mo (4mo-4yr)	No	No
Artrip (2006)[21]	2000-2005	10	8 BTS (3) PAB (5) COA repair (2)	IVR 7 ASO 3	11mo (9d-4yr)	1	20mo	1	NA
Hu (2010)[79]	2006-2009	6	NA	Double-root translocation	*4yr (1-16yr)	No	*22mo (2-36mo)	없음	No

(ASO; arterial switch operation after LV–PA tunneling, AVR; aortic valve replacement, BTS; Blalock–Taussig shunt, COA; coarctation of the aorta, IVR; intraventricular rerouting, MV; mitral valve, PAB; pulmonary artery banding, TV; tricuspid valve)
NA; not available
* 4 patients with Taussig–Bing anomlay included

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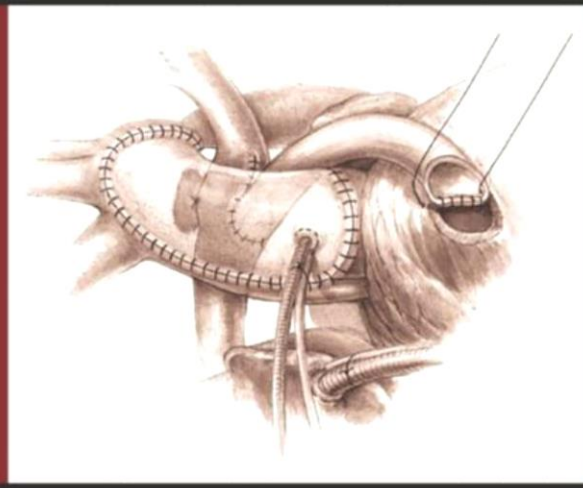

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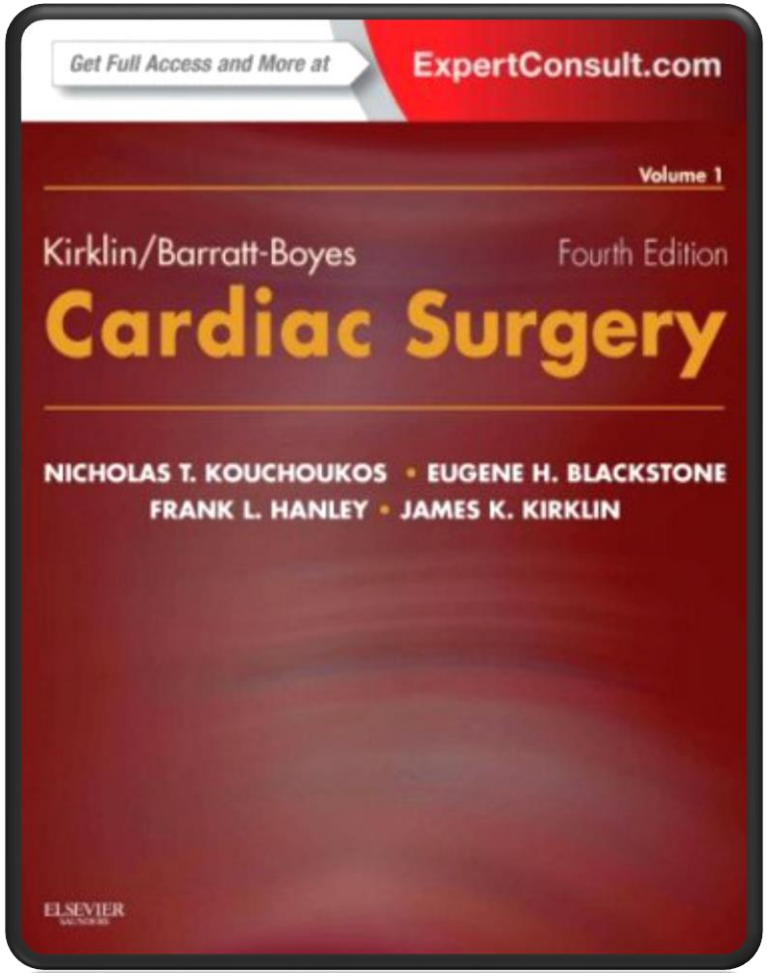
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