

2016년 흉부외과 전공의 연수강좌

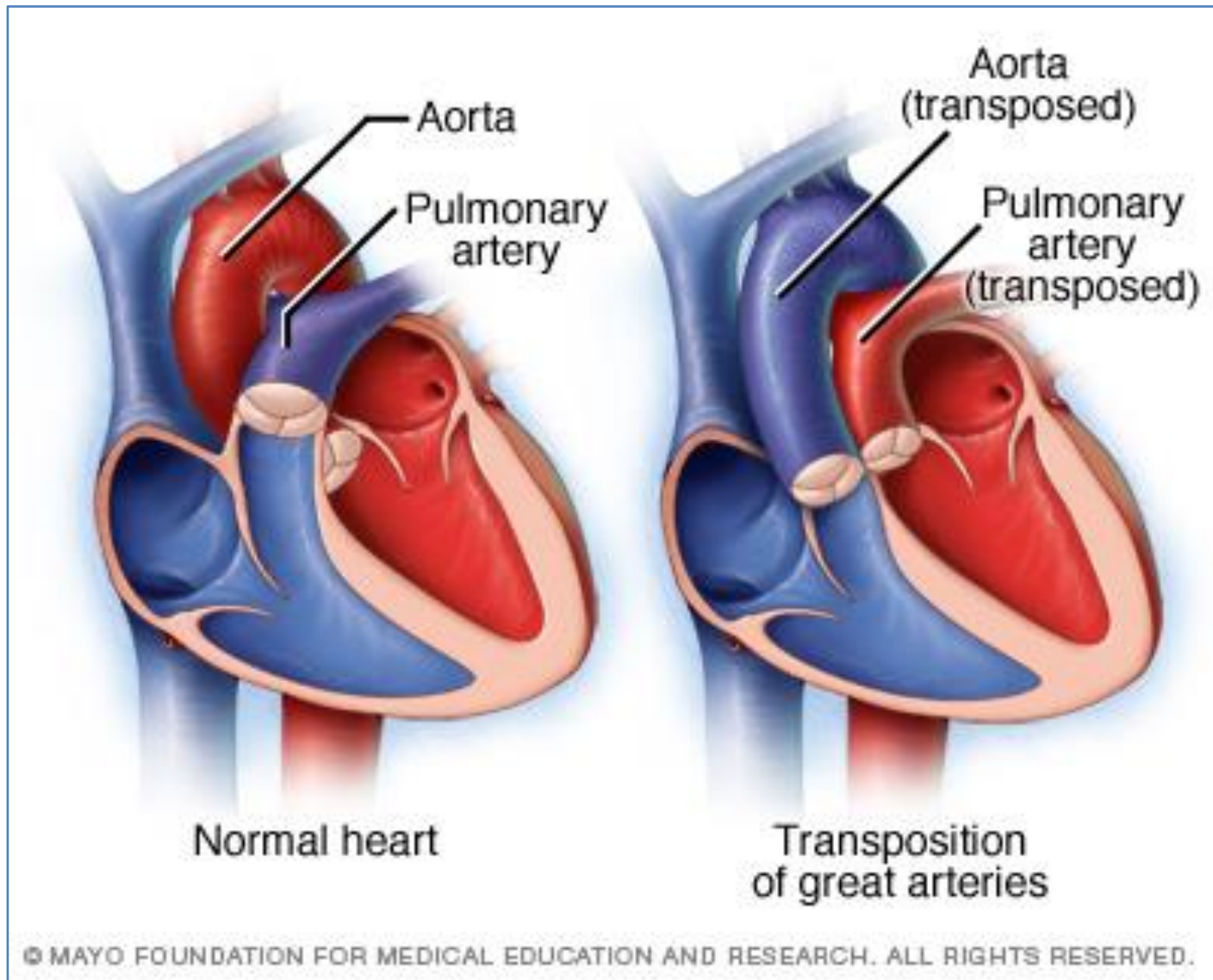
Transposition of Great Arteries

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http://www.mayoclinic.org/~media/kcms/gbs/patient%20consumer/images/2013/08/26/11/07/ds00733%20mcdc7_transpositionthu_jpg.png

학습목표

- TGA의 수술시기를 알 수 있다
- Coronary artery pattern을 기술할 수 있다
- Atrial switch operation을 이해한다
- TGA,VSD,PS의 수술방법을 구분할 수 있다
- Arterial switch operation후의 long-term complication을 이해할 수 있다

Definition

- Concordant atrio-ventricular connections
- Discordant ventriculo-arterial connections

History

- Baillie가 1797년 처음 형태학적으로 기술하였고, 1814년 Farre가 전위(transposition) 라는 표현을 사용하였다.
1971년 Van Praagh가 명확히 정의하였다
- Senning 과 Mustard 가 1959년과 1963년에 각각 심방전 위술을 처음으로 성공시켰다. 1975년 Jatene 등에 의해 처음으로 성공적인 동맥전환술(arterial switch operation, ASO)이 시행되었다

Anatomy of TGA

- Intact ventricular septum ("Simple" TGA) = 50%
- VSD (perimembranous) = 25%
- VSD and pulmonary stenosis = 25%
- Patent foramen ovale in nearly all
- Coarctation rare

Incidence

- 모든 선천성 심기형의 5-7% 빈도로 발생
- 청색증 심장기형 중 활로 4징증에 이어 두 번째로 흔하다.
- 남자에서 약 3:1로 발생빈도가 높다

Classification

- TGA with IVS – 신생아기 수술 필요
- TGA with VSD – 3개월 이내 수술 필요
- TGA with VSD, PS(LVOTO) – Pailliative 수술 후 수술시기 결정

Physiology

- Separate parallel circulation
- Degree of cyanosis depends on mixing
- VSD patients less cyanotic
- LV thickness/function diminished > 1 month

Clinical Presentation

- Cyanosis: simple > VSD
- Earlier presentation: simple > VSD and
PS > VSD
- Soft systolic murmur

Diagnosis

- CXR
 - Normal at birth
- Echo
 - Posterior branching great vessel
 - Intracardiac anatomy defined
 - Coronary ostia defined
- Catheterization
 - Septostomy or coronary anatomy

Medical management

- Prostaglandin E1 infusion to maintain PDA
- Balloon atrial septostomy (Rashkind, 1966)
- Correct acid-base abnormalities
- Increase pulmonary blood flow / mixing

Pathophysiology

- Two parallel circulations
- Mixing between the parallel circulations: PDA, ASD, VSD
- Rapid progression of pulmonary vascular disease
- LVOTO
- LV pressure
- 관상동맥형태

Single / Intramural

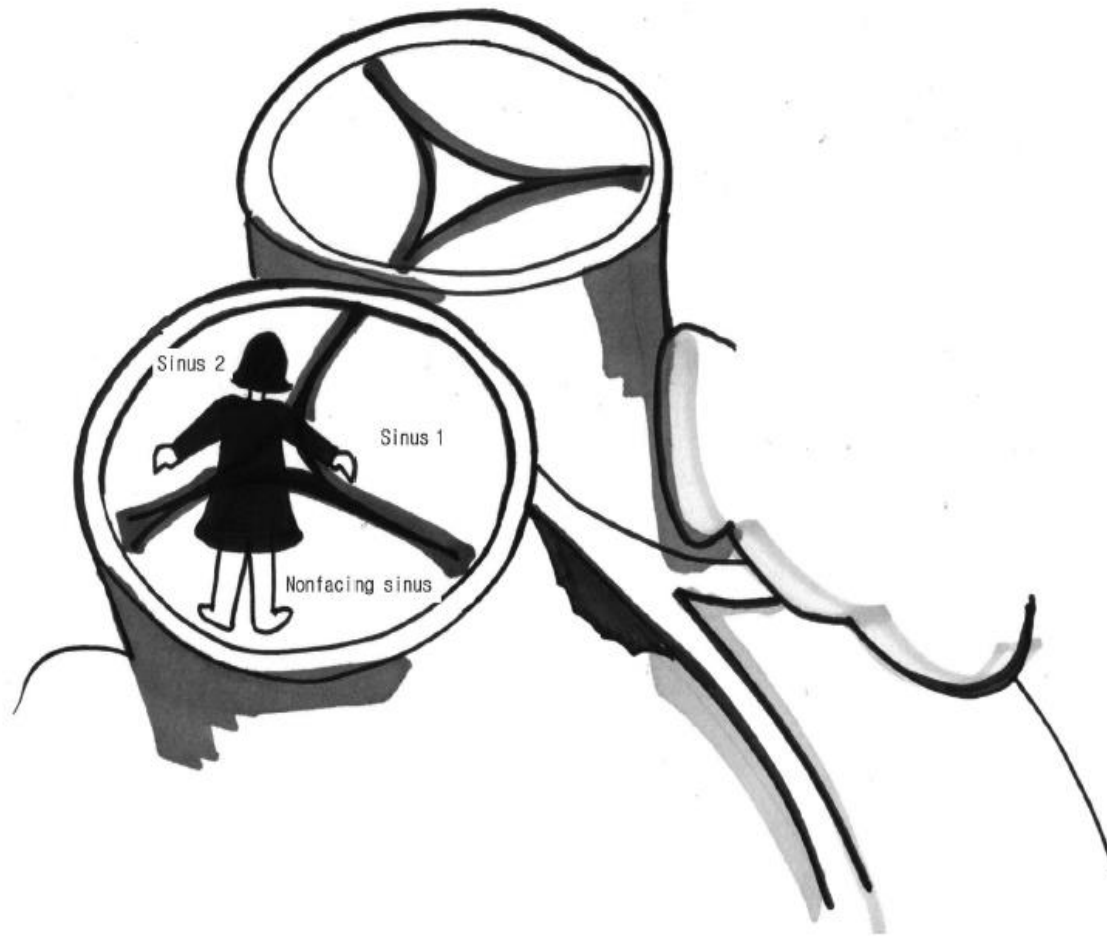


Figure 24-1. Lenden convention, Sinus 1 is the sinus adjacent to the pulmonary artery on the right-hand side of the observer, Sinus 2 is the sinus adjacent to the pulmonary artery on the left-hand side of the observer

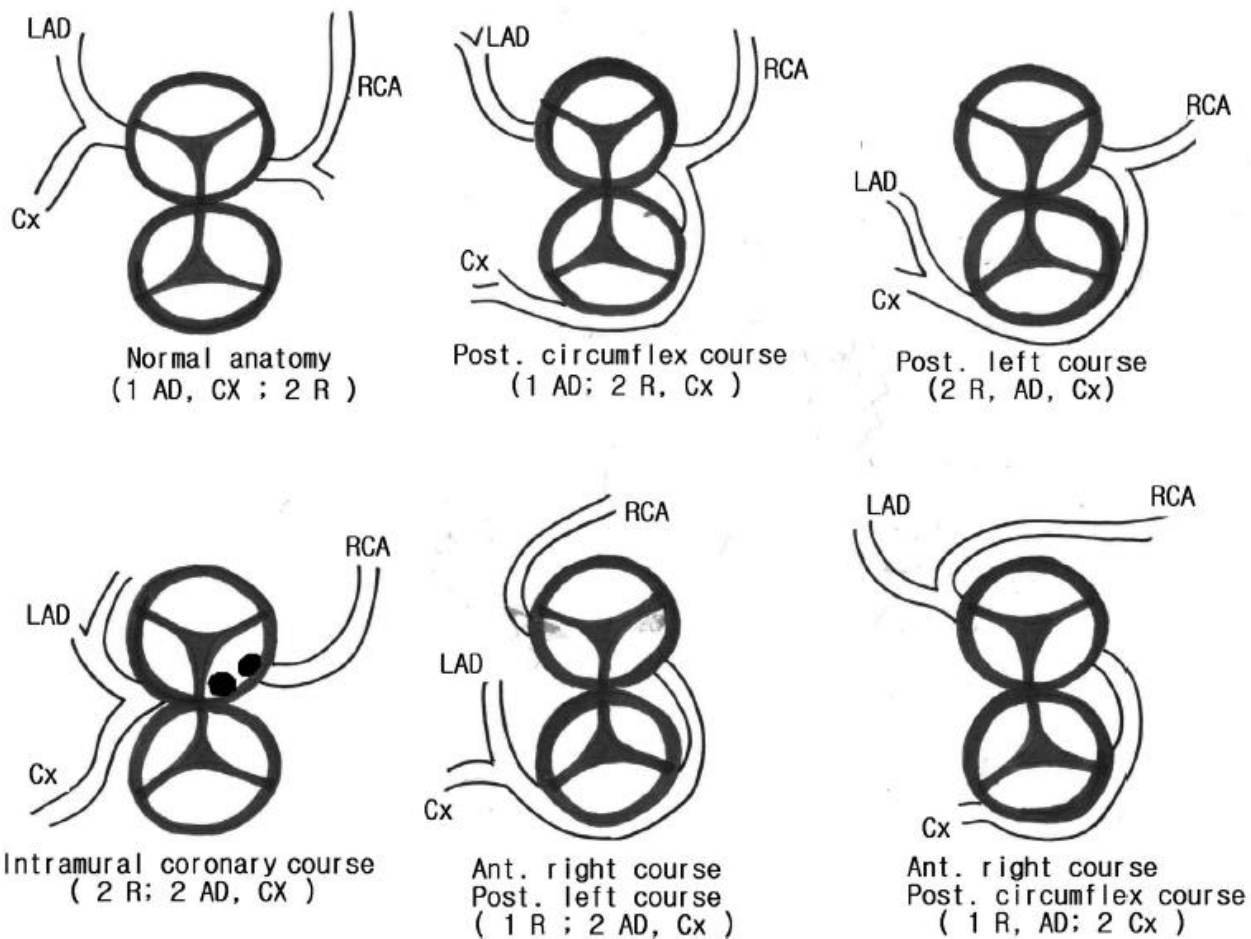
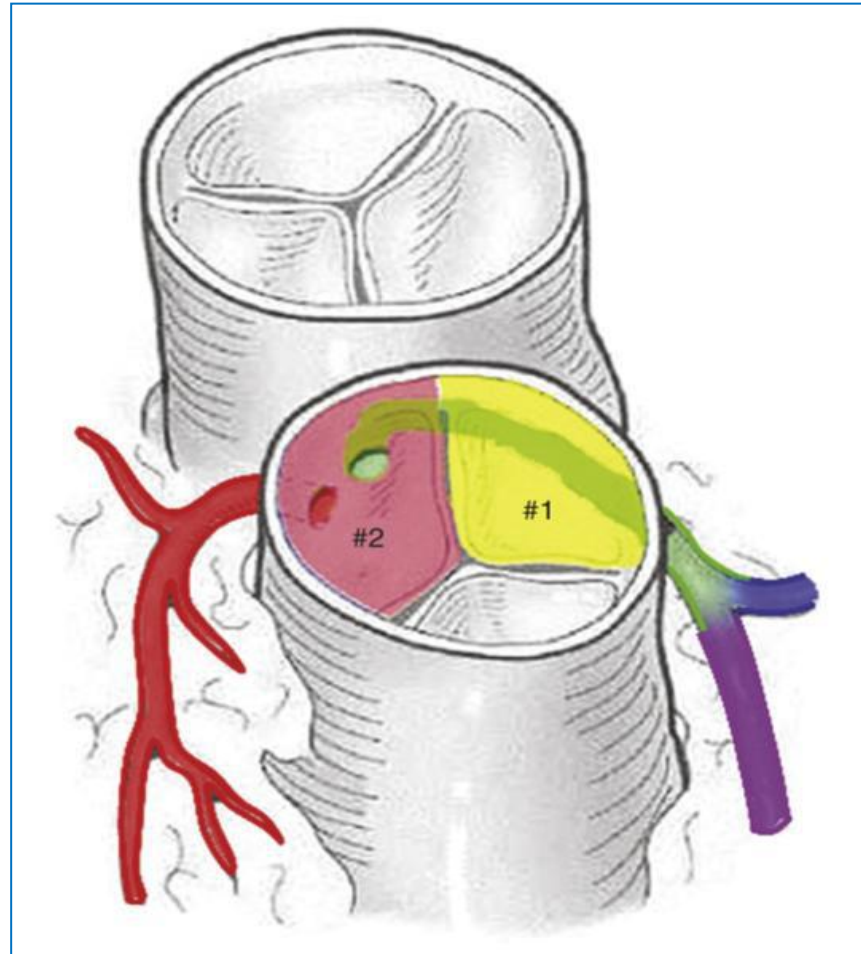


Figure 24-2. The six commonest types of coronary artery anatomy in d-TGA. AD, left anterior descending artery; Cx, circumflex coronary artery; R, right coronary artery

Coronary Anomalies

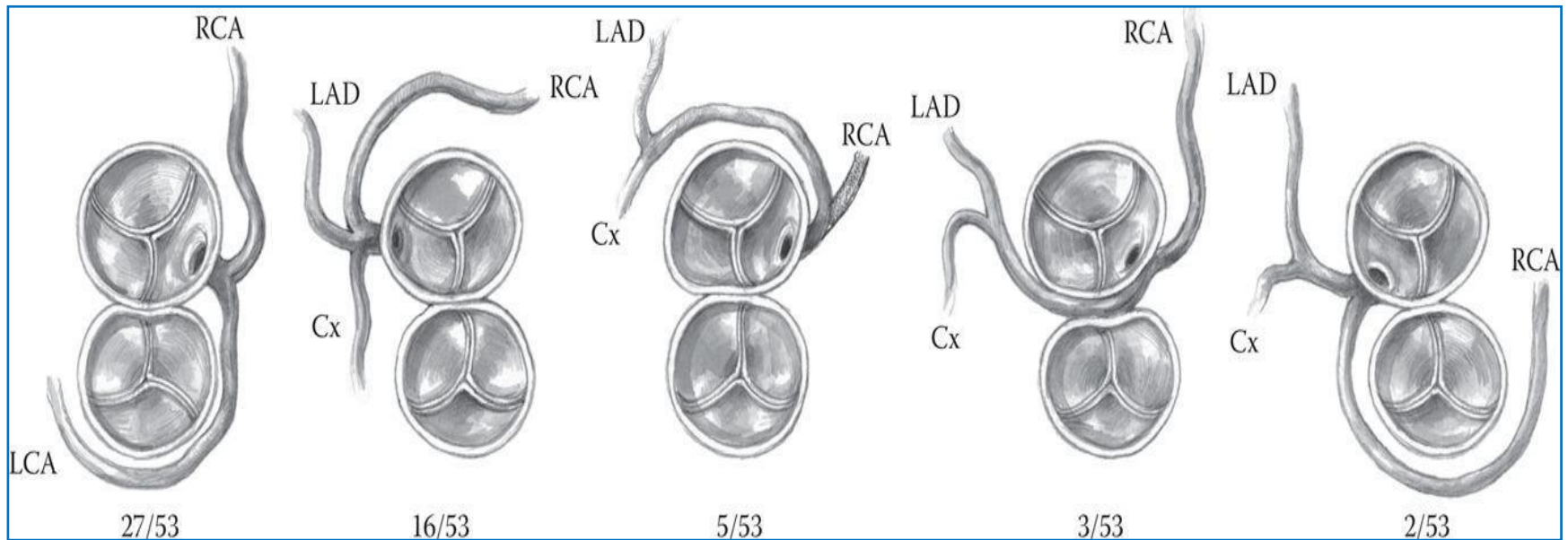
- Intramural coronary artery
- Single coronary ostium
- Coronary ostial atresia, stenosis

Intramural Coronary Artery



Anderson RH. Paediatric cardiology. 3rd ed. 2010

Single Coronary Artery

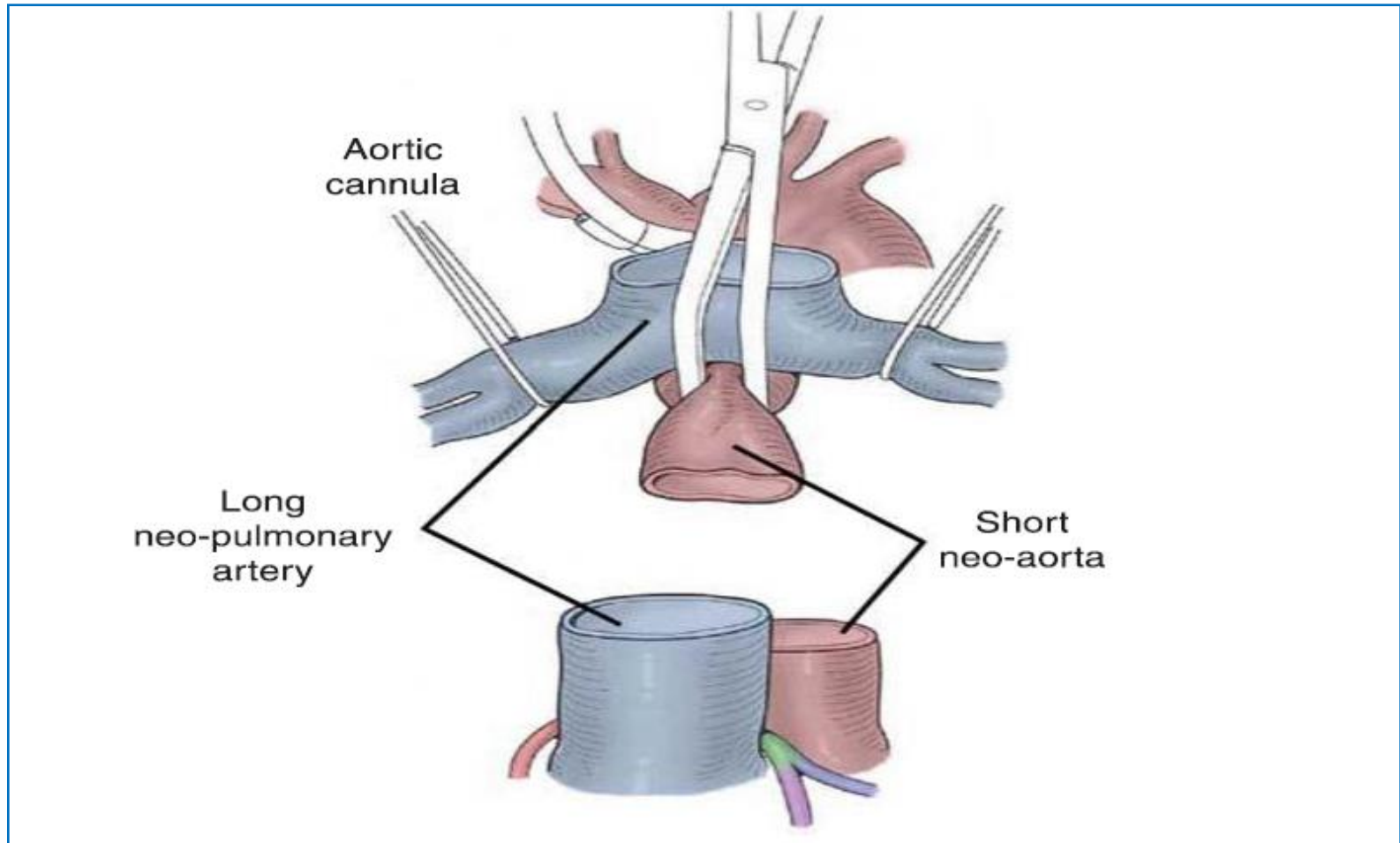


Jonas RA. Comprehensive surgical management of congenital heart disease.
2nd ed. 2014

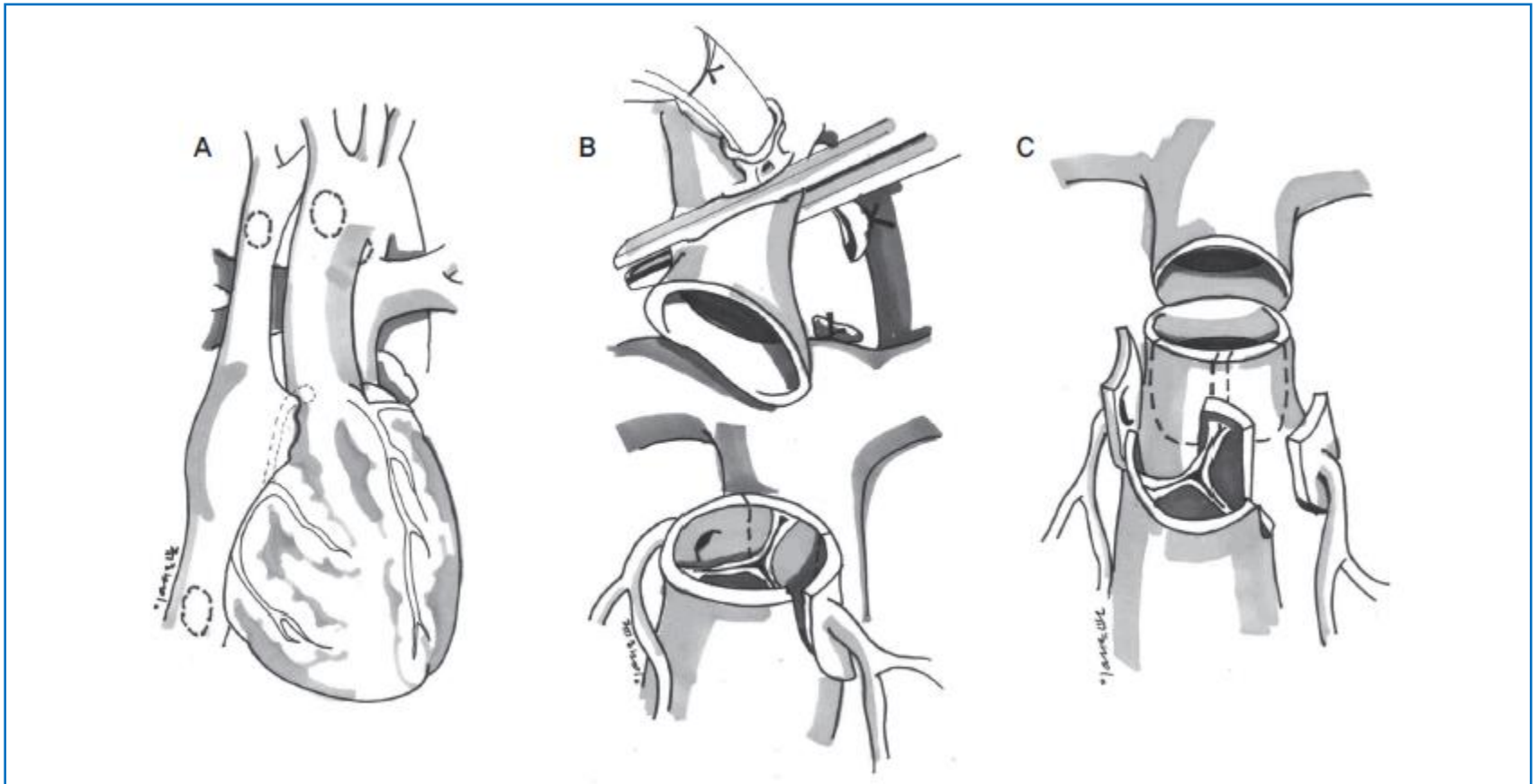
Surgical Management

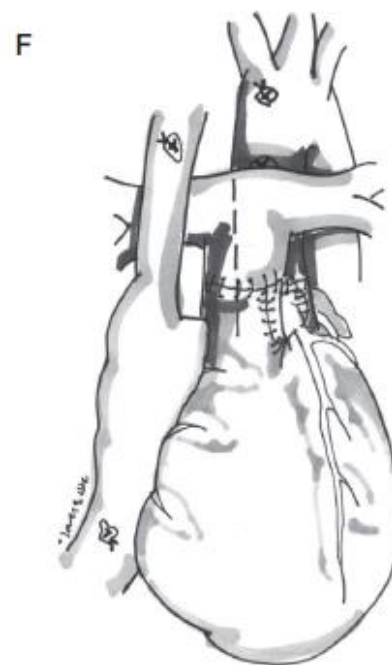
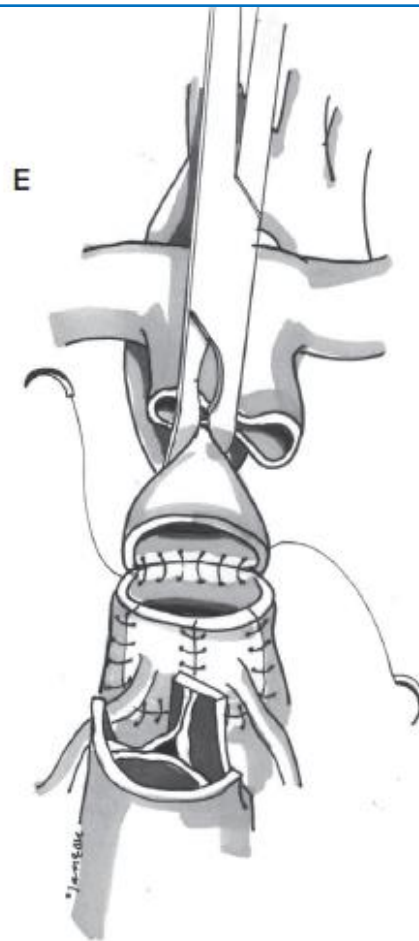
- Atrial switch procedure (Senning, Mustard)
- Arterial switch procedure
- Rastelli / REV / Nikaidoh procedure
- Arterial switch after LV retraining

Lecompte Maneuver



Arterial Switch Operation

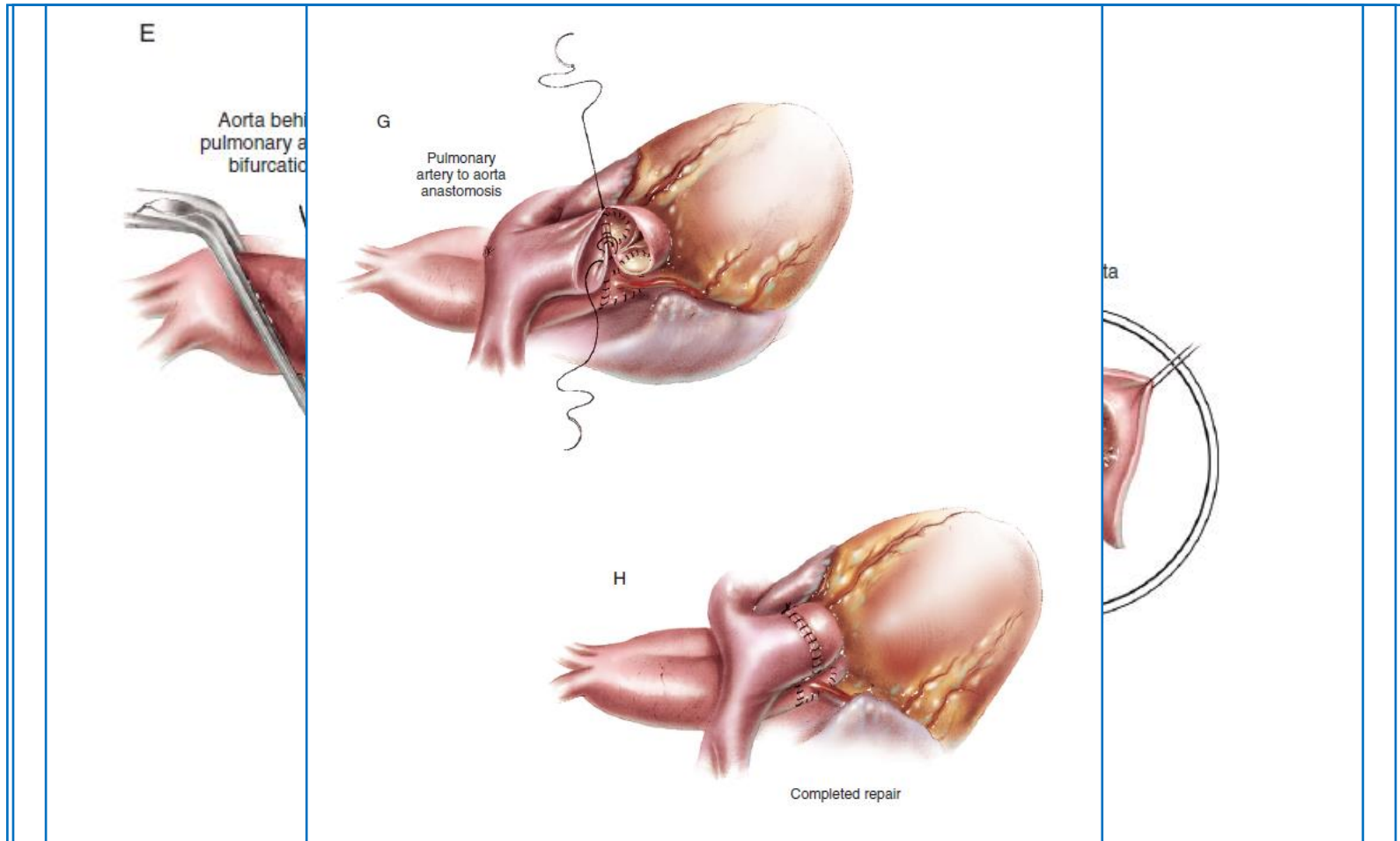




Steps in Arterial Switch Procedure

- Excision coronary arteries
- Transfer coronary arteries to neo-aorta
- LeCompte maneuver
- Neo-aorta anastomosis
- Reconstruct neo-pulmonary root -
pericardium

Arterial Switch Operation



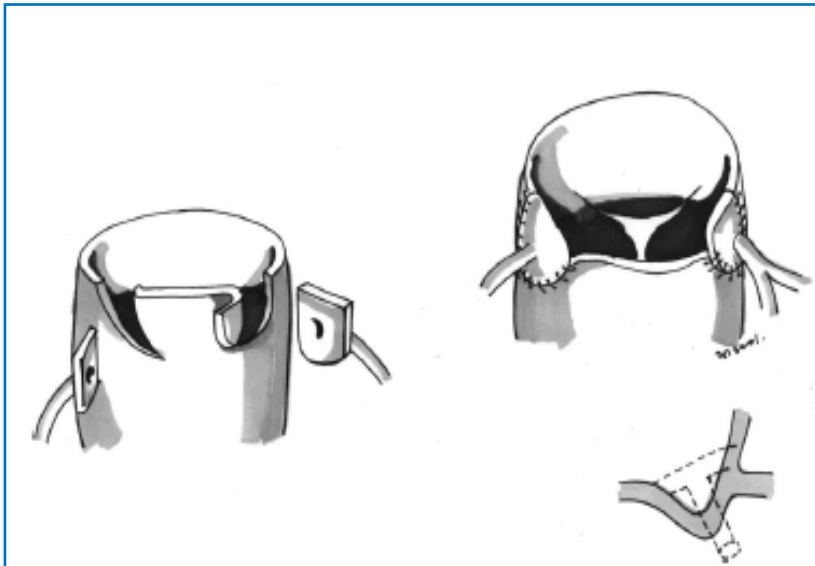


Figure 24-4, Trapdoor formation, Medially based flap incisions are used for coronary reimplantation,

Open technique

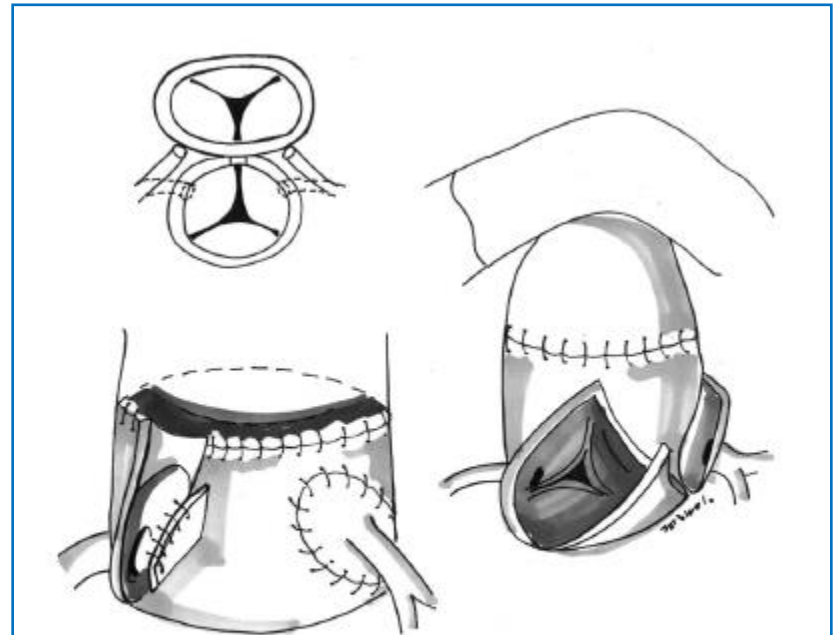


Figure 24-5, Coronary artery transfer after neo-aorta anastomosis, Marking sutures on the point of superior margin of each commissure, Reimplantation of coronary artery transfer along the oblique incision is done after neo-aorta reconstruction,

Close technique

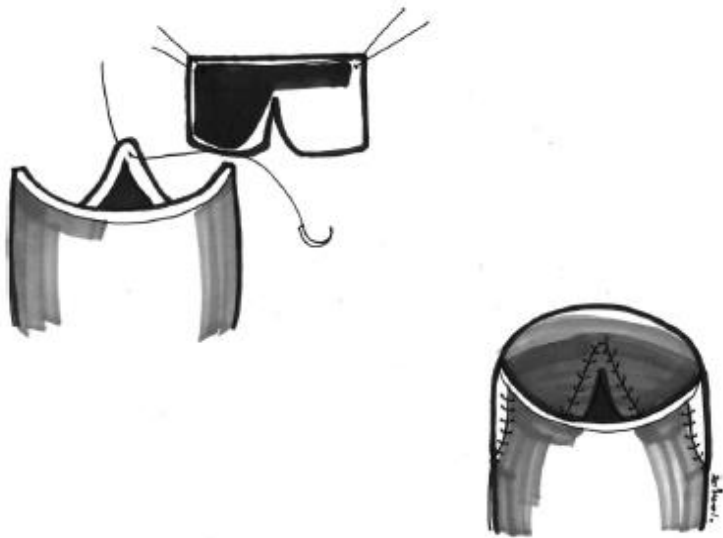


Figure 24-6. The defect in the aorta from which the coronary arteries have been excised is augmented with a pantaloon-shaped patch of pericardium.

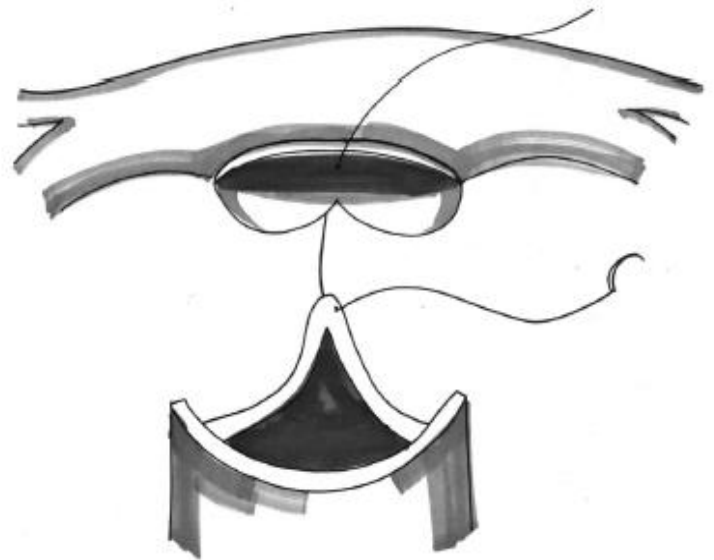


Figure 24-7. Direct pulmonary artery reconstruction to avoid pericardial or prosthetic patches and to hold the potential for unlimited tissue growth.

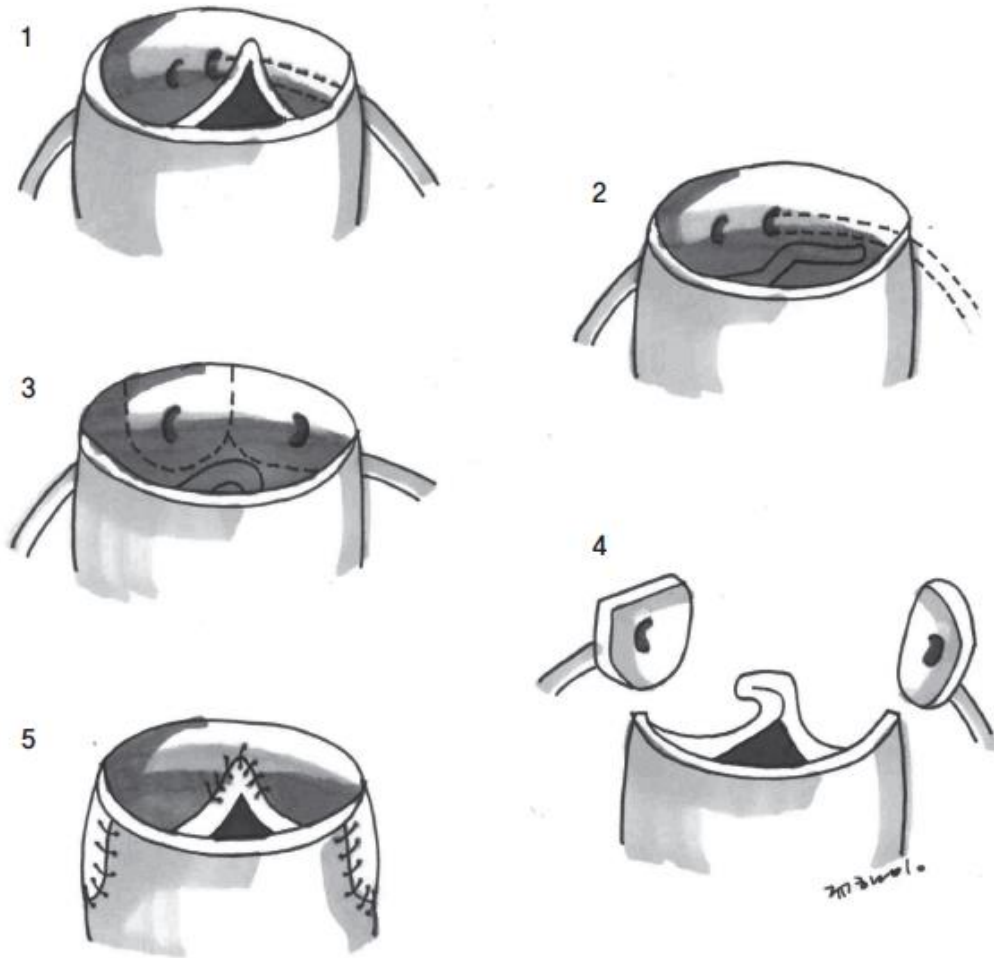
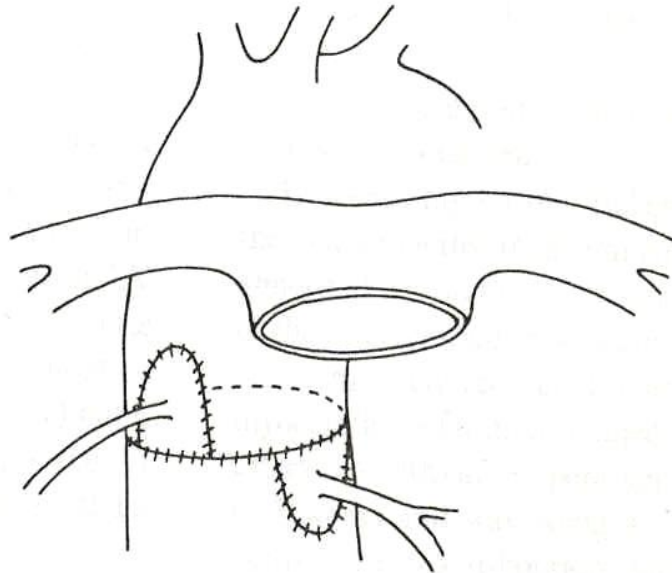
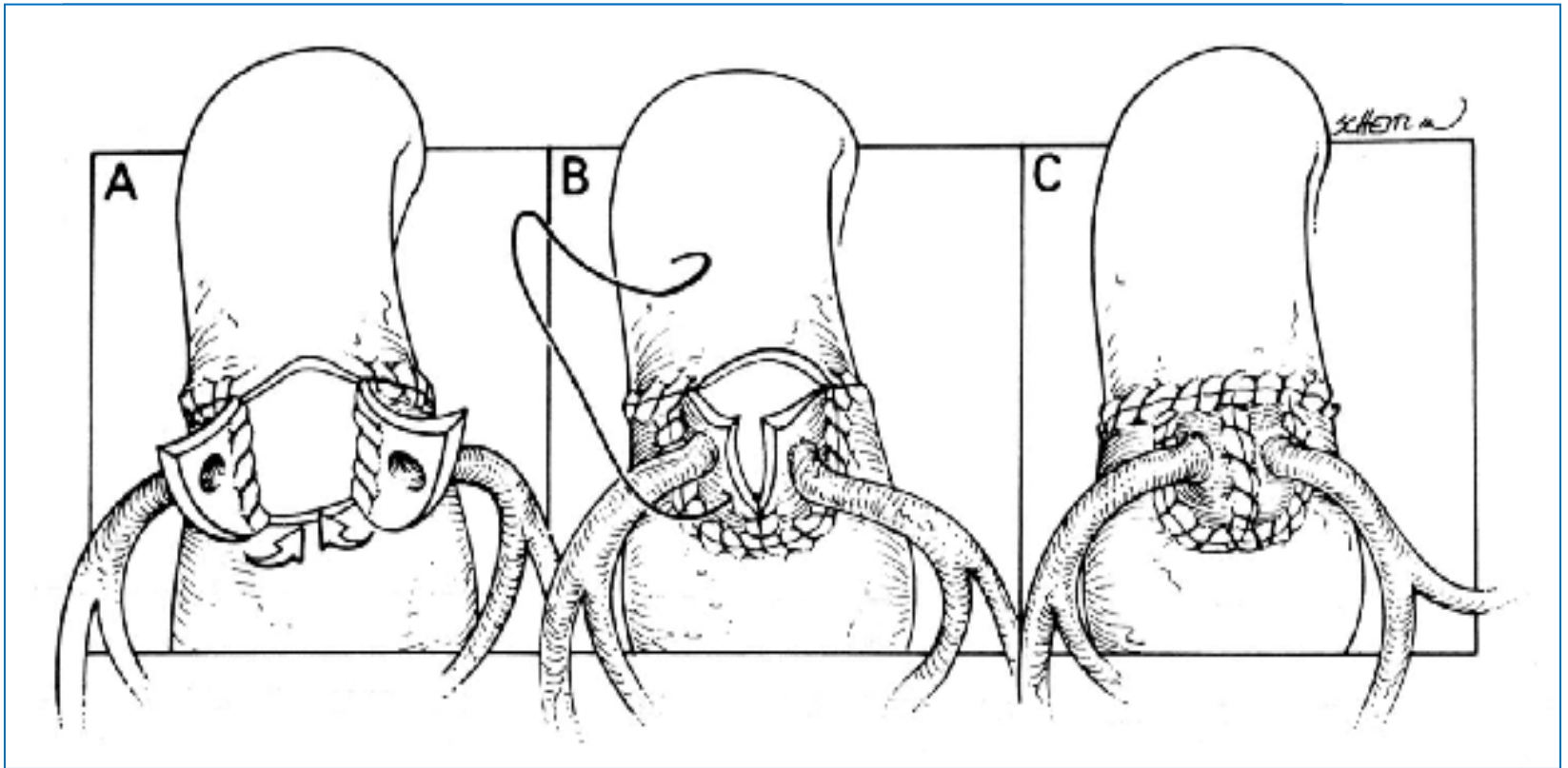


Figure 24-8. If juxtacommissural origins of either or both coronary arteries from the facing sinuses are present, excision of a portion of the native aortic valve may be necessary to allow mobilization.

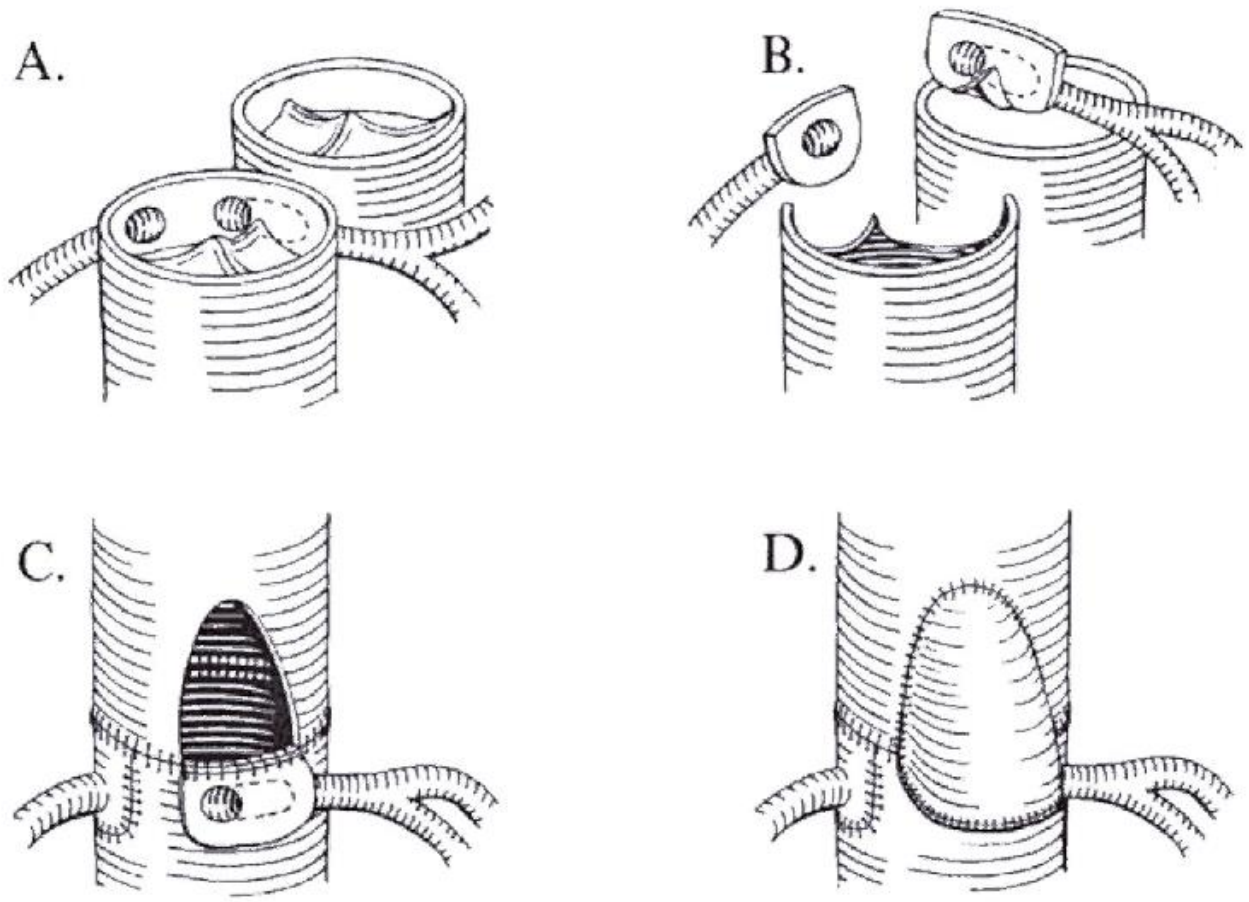
- Preserve a harmonious curve of the coronary trunks.
- In the middle of the appropriate sinus



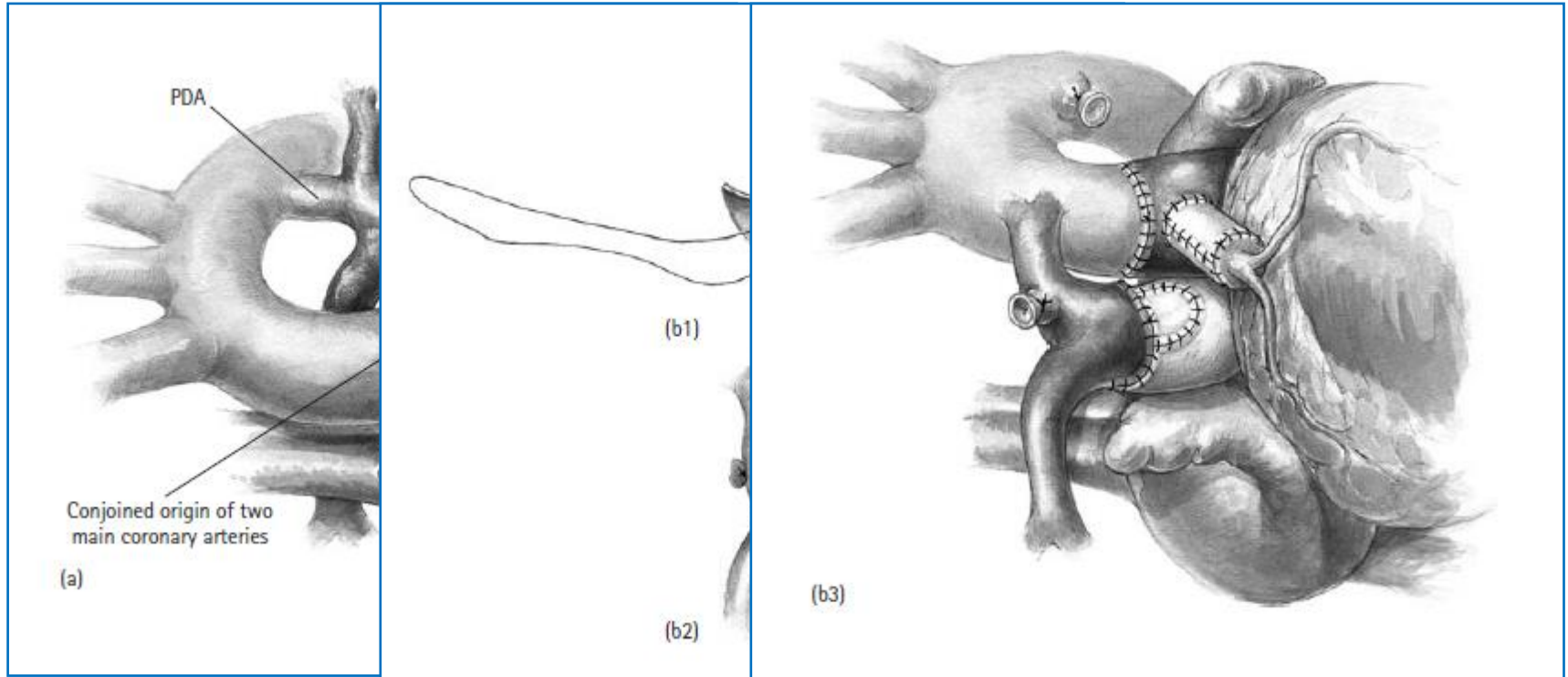
J Thorac Cardiovasc Surg 1988;96:354-63



Eur J Cardio-thorac Surg 1994;8:74-8



Single Coronary Artery



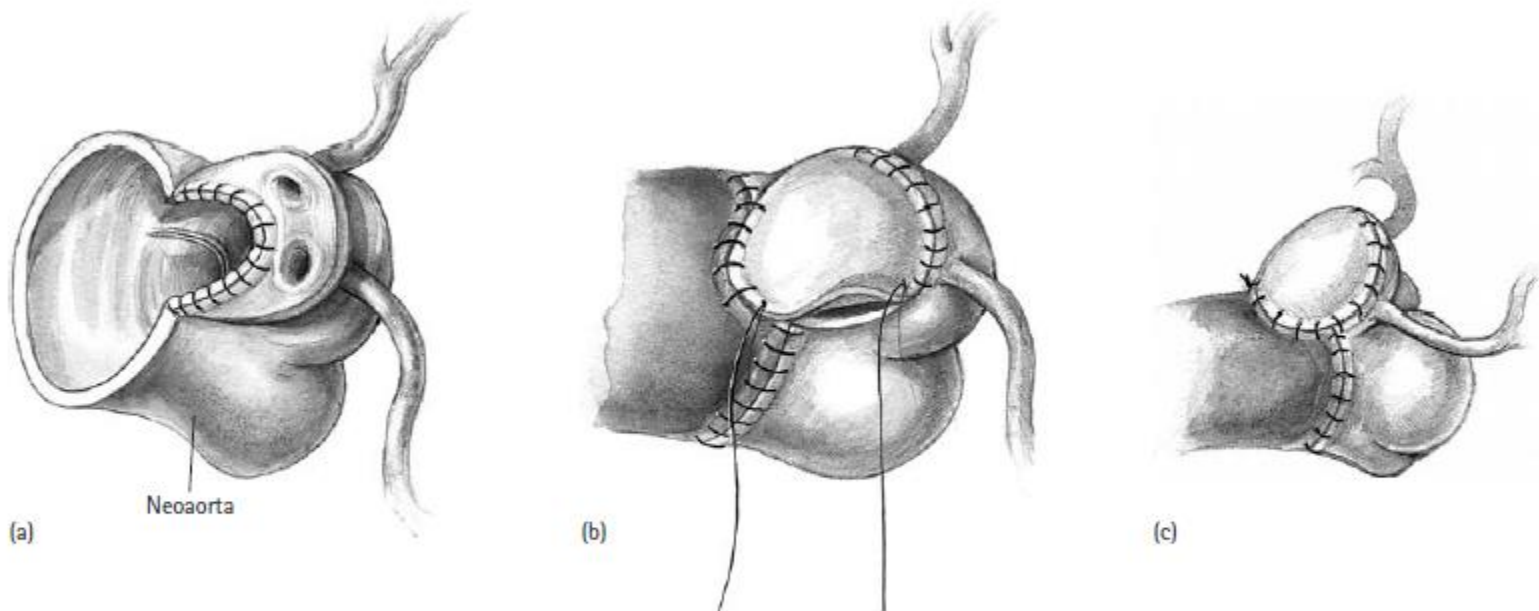


Figure 15.6 *In the extremely rare instance of a single coronary artery running between the pulmonary artery and aorta the button is rotated through 90° and roofed with pericardium.*

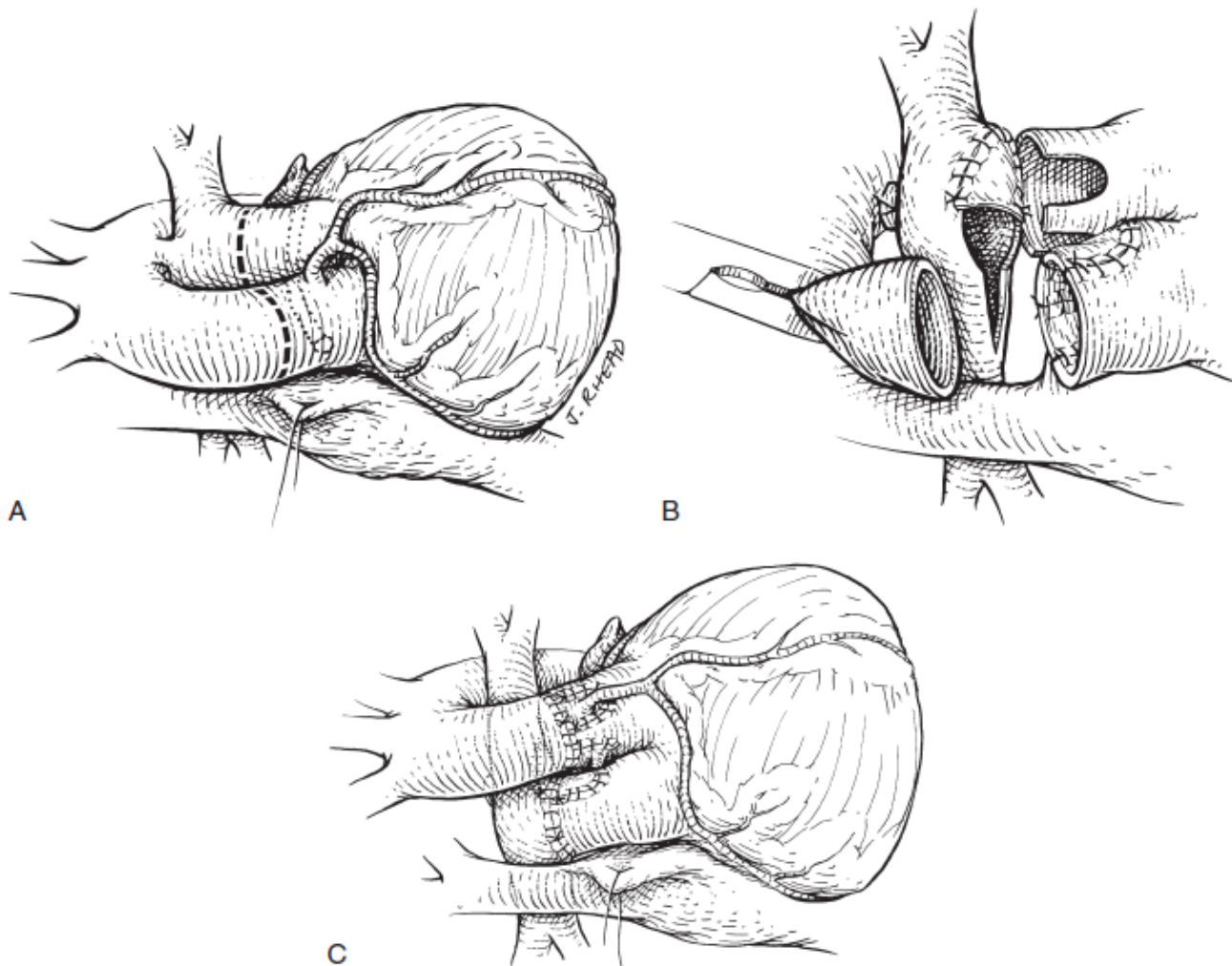


Figure 52-31 Arterial switch operation for transposition of great arteries with side-by-side great arteries and aorta to right, with coronary pattern of 1LR-2Cx. This coronary pattern is common with this great artery orientation. **A**, Dashed lines show proposed transection sites

Postoperative care

- Respiratory and supportive care

- Adequate ventilation
- mild respiratory alkalosis (pH 7.50 – 7.60, pCO₂ 28 – 35 mmHg)
- Sedation and pain relief -> ↓ oxygen consumption, metabolic expenditure, prevention of pulmonary hypertensive crisis

Postoperative care

- LV is poorly compliant after ASO
- Rapid infusion of fluid should be avoided.
- Boluses of fluid are generally given by infusion in quantities of 5 –10 ml/kg.
- High LAP or CVP pressure should be avoided

Postoperative care

- • Increased afterload might result in low cardiac output and ventricular failure.
-
- • Afterload reduction should be maintained at least 48 – 72 hours during the period of LV adaptation to the SVR
-
- • Blood pressure : 50 – 70 mmHg

Postoperative care

- Retraining the left ventricle after arterial switch operation: emerging uses for the left ventricular assist device in pediatric cardiac surgery

[J Cardiothorac Vasc Anesth. 2000 Aug;14:454-6](#)

- ECMO

Results of Arterial Switch

- Operative mortality = 2-5%
- Higher mortality
 - single coronary/intramural coronary
- Supravalvular PS = 10-15%
- Sinus rhythm > 95%
- 90% survival at 5 years

Risk Factors for ASO

- Presence of an intramural coronary artery
- Low birth weight
- Older age of patients with simple TGA
- longer periods of circulatory arrest
- Multiple VSD's
- Augmentation of the aortic arch

Senning Procedure

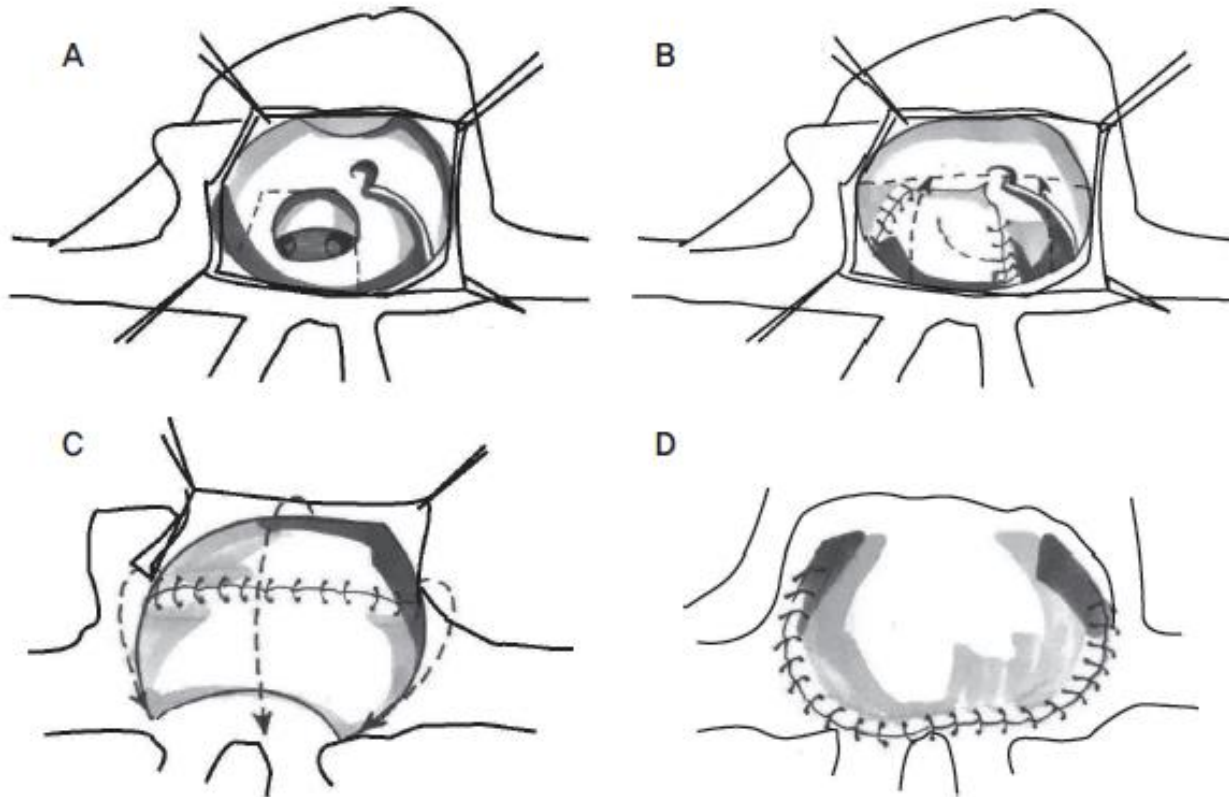
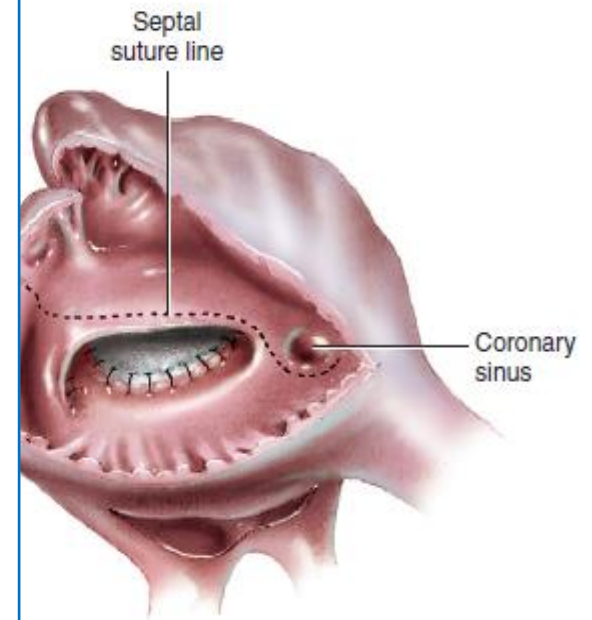
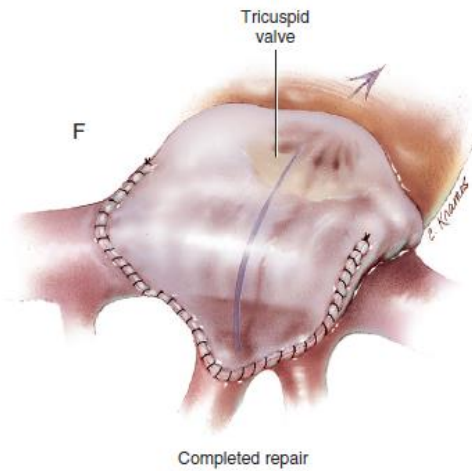
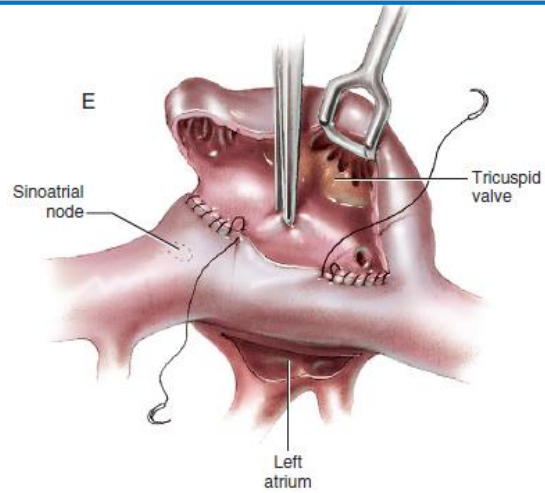


Figure 24-9. Senning procedure. A, View from the right atrium showing incision lines (dashed lines) to create a flap of the posterior atrial septum. B, The posterior flap of atrial septum is augmented with a piece of pericardium and sutured inferiorly over the origins of the pulmonary veins in the left atrium. C, After completion of the venous baffle, the anterior wall of the right atrium is sutured to an opening made in the left atrium posterior to the interatrial septum. D, Completed suture line showing repair with autologous tissue.

Senning Procedure



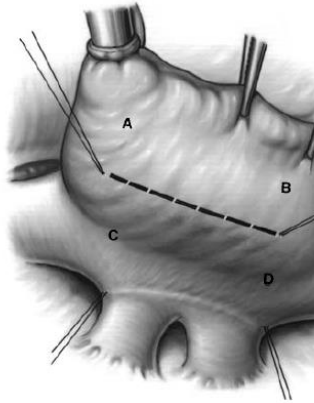


Figure 34.1

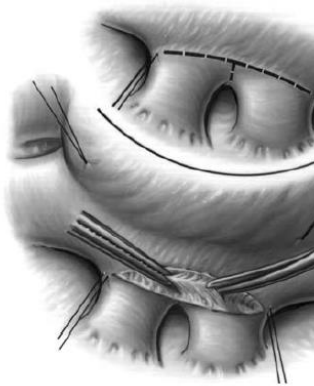


Figure 34.2



Figure 34.3

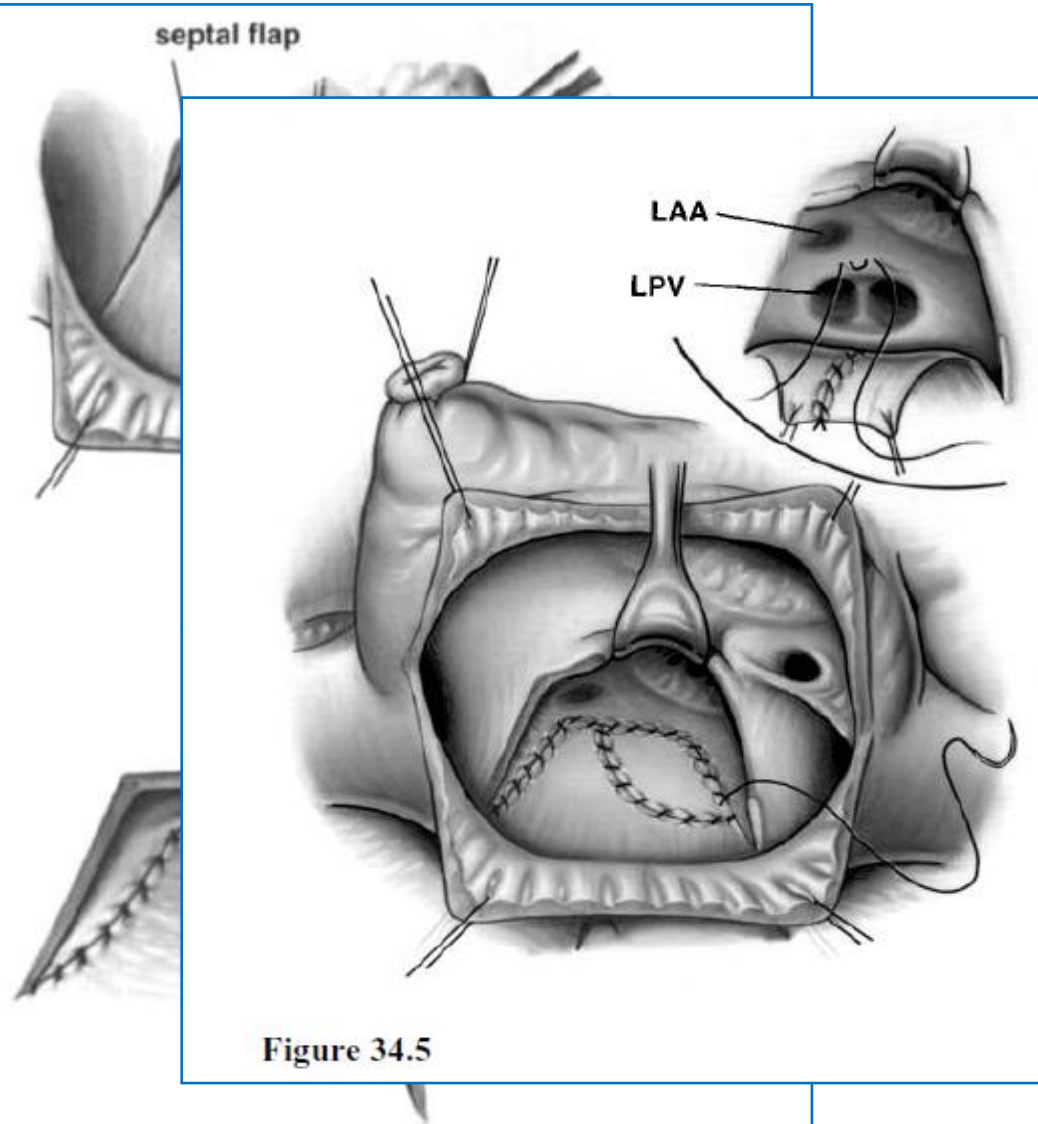


Figure 34.5

Figure 34.4

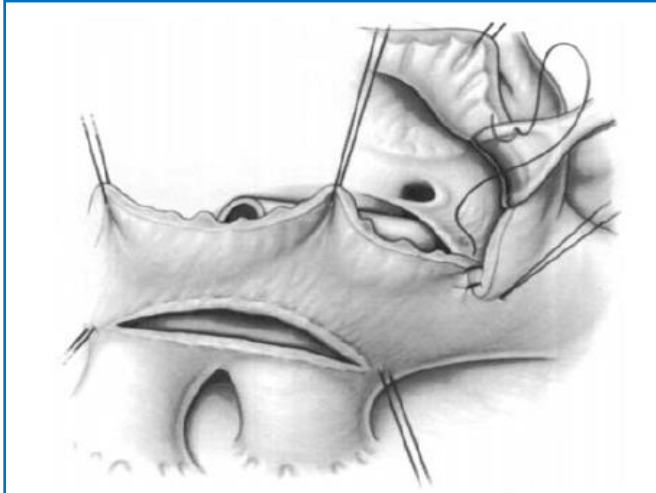


Figure 34.6

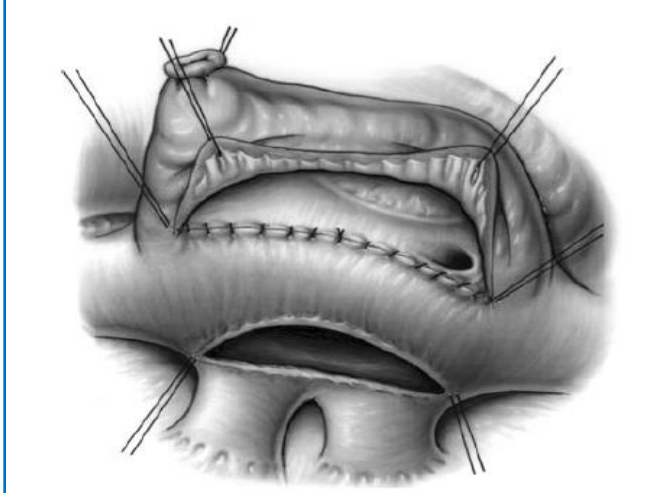


Figure 34.7

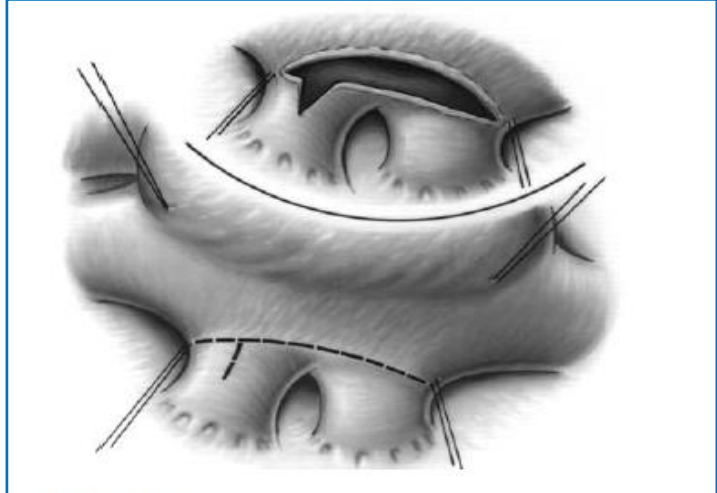


Figure 34.8

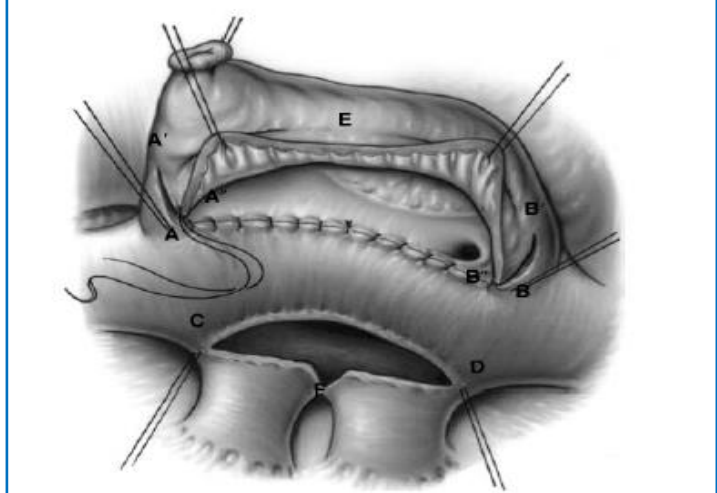


Figure 34.9

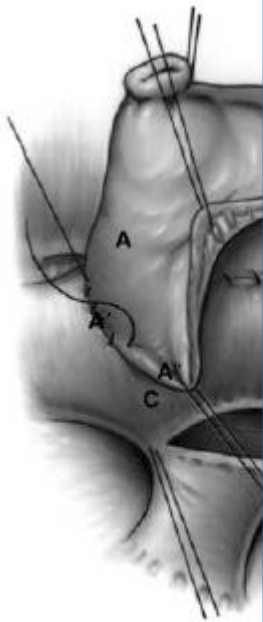


Figure 34.10

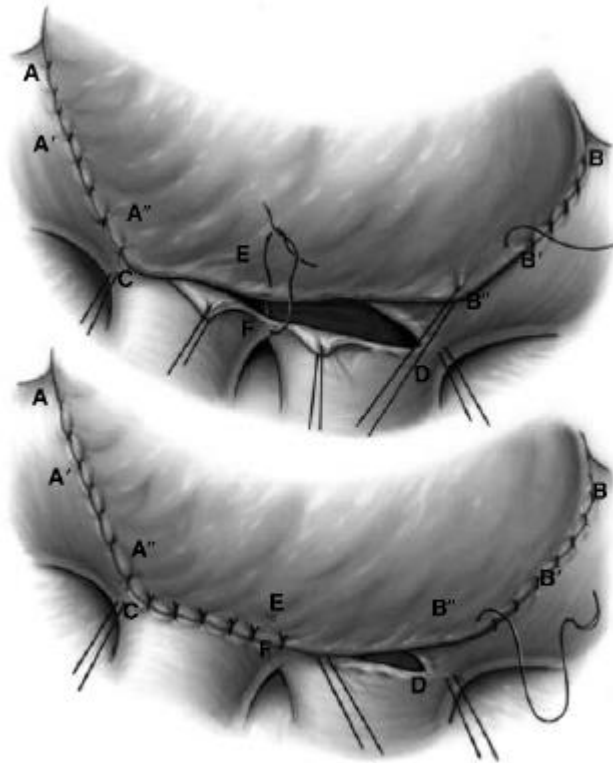


Figure 34.11

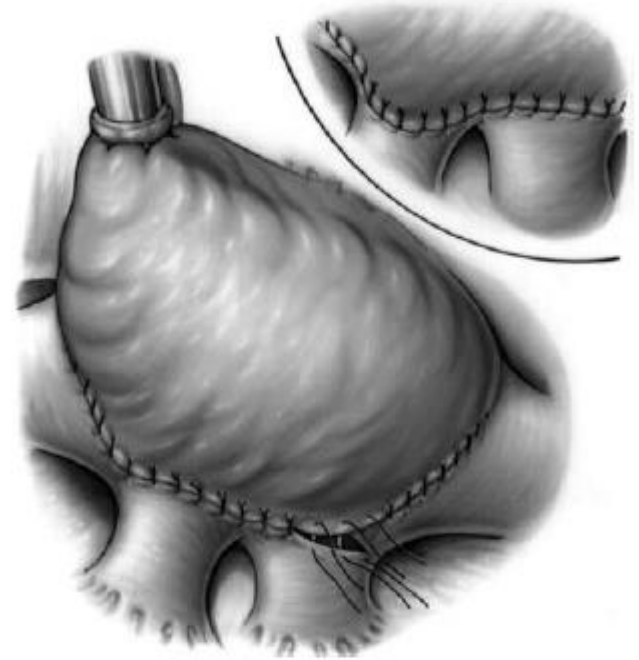
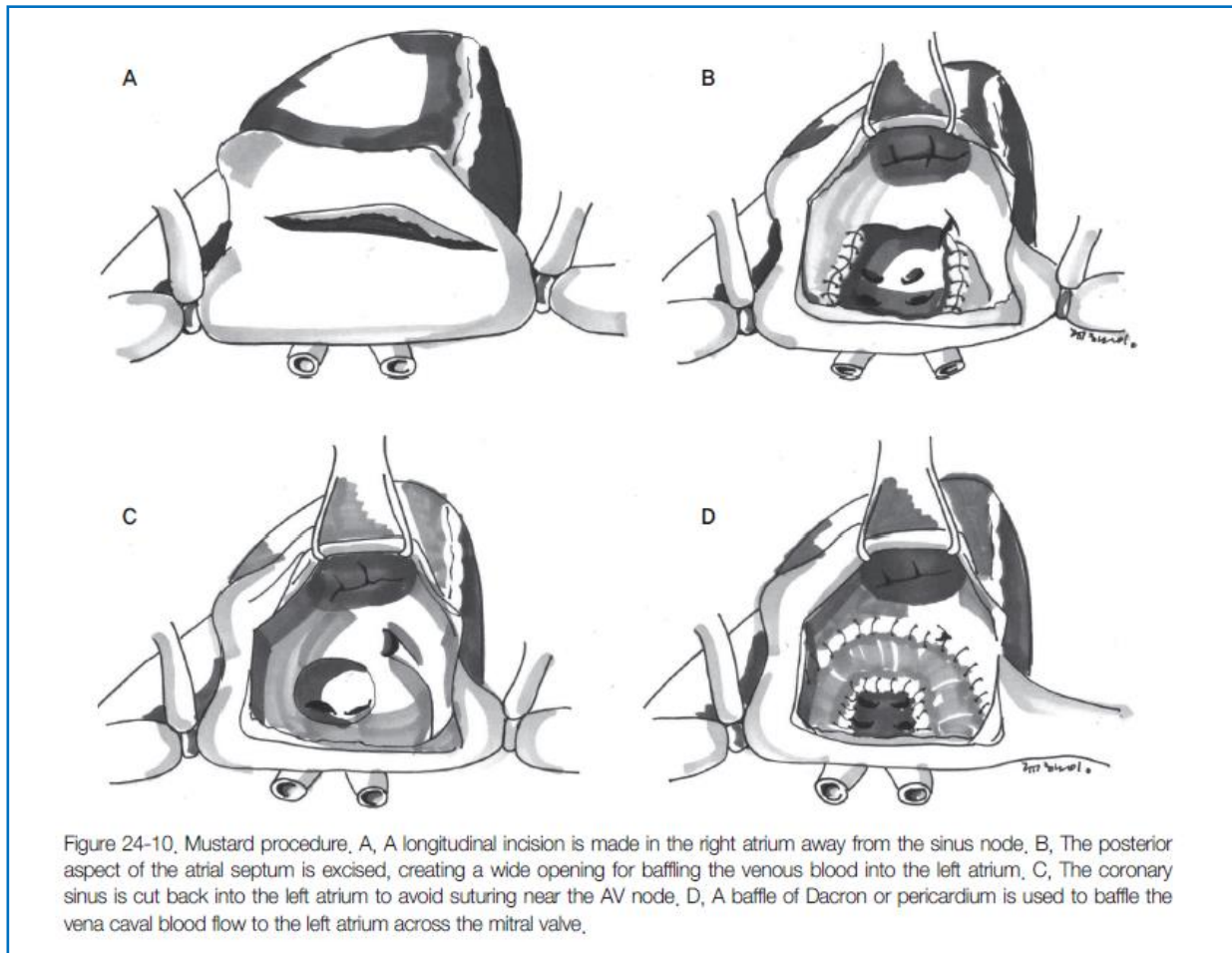


Figure 34.13

Mustard Procedure



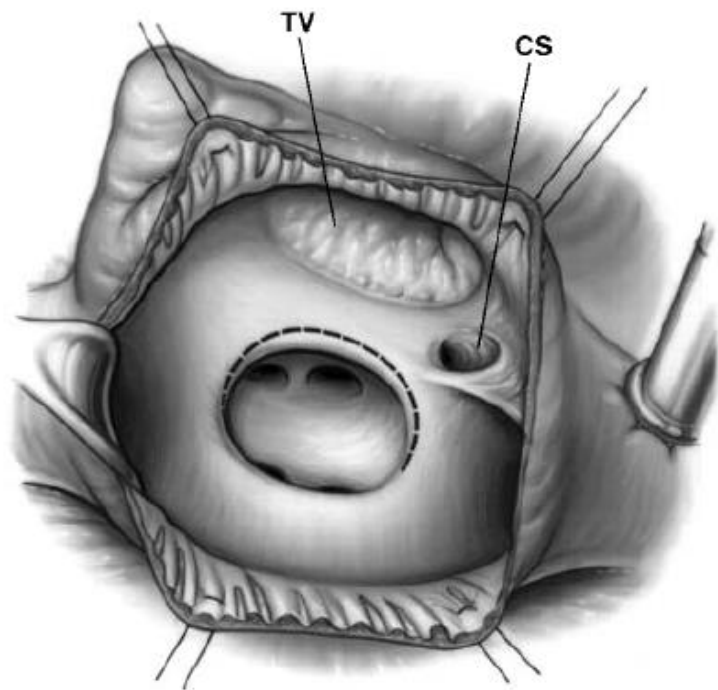


Figure 33.5

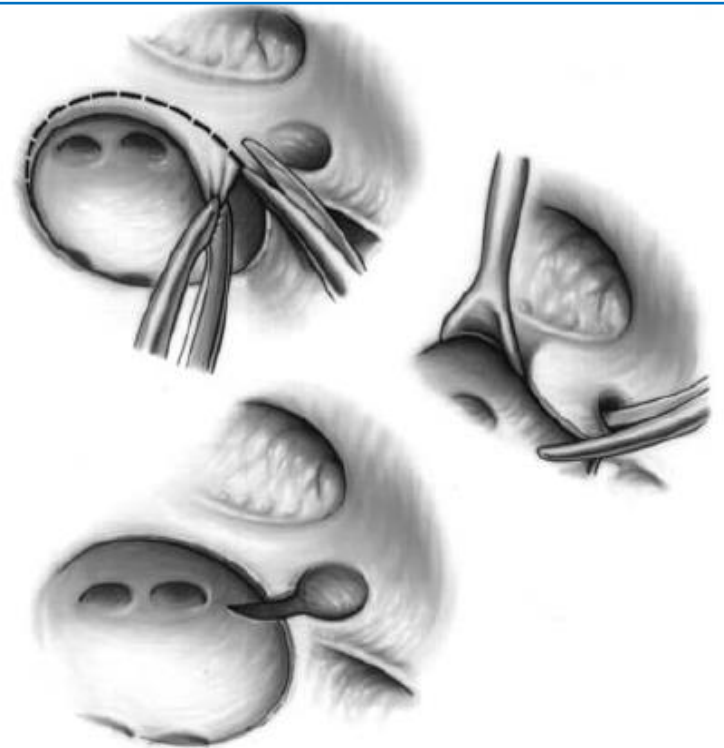


Figure 33.6

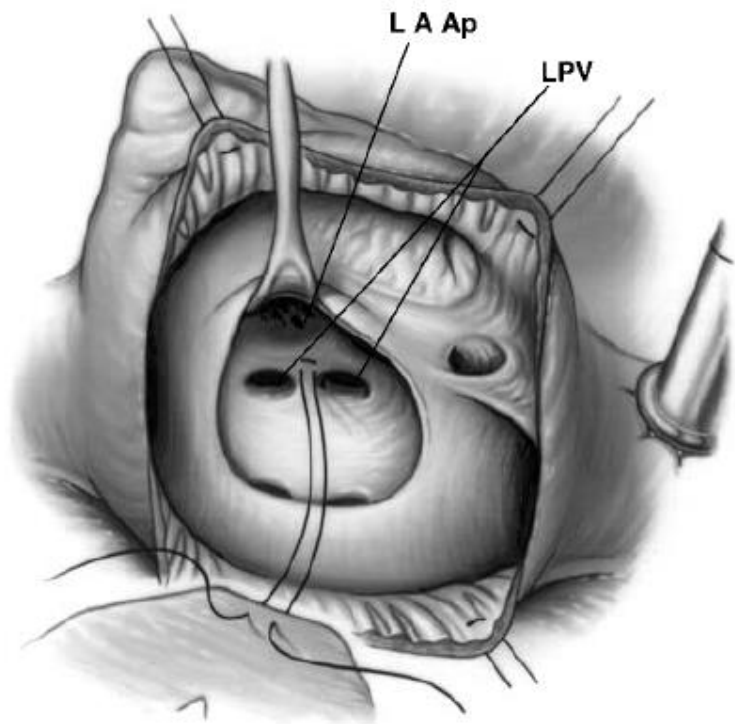


Figure 33.7

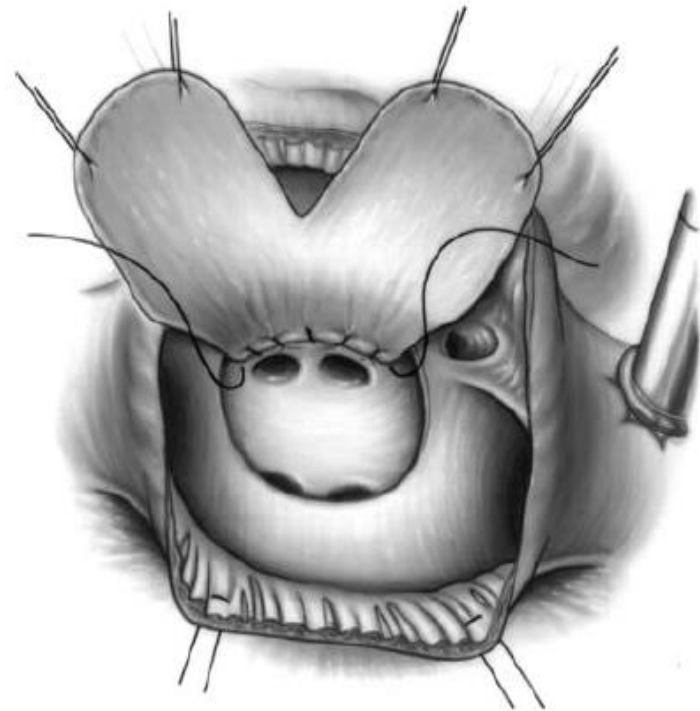
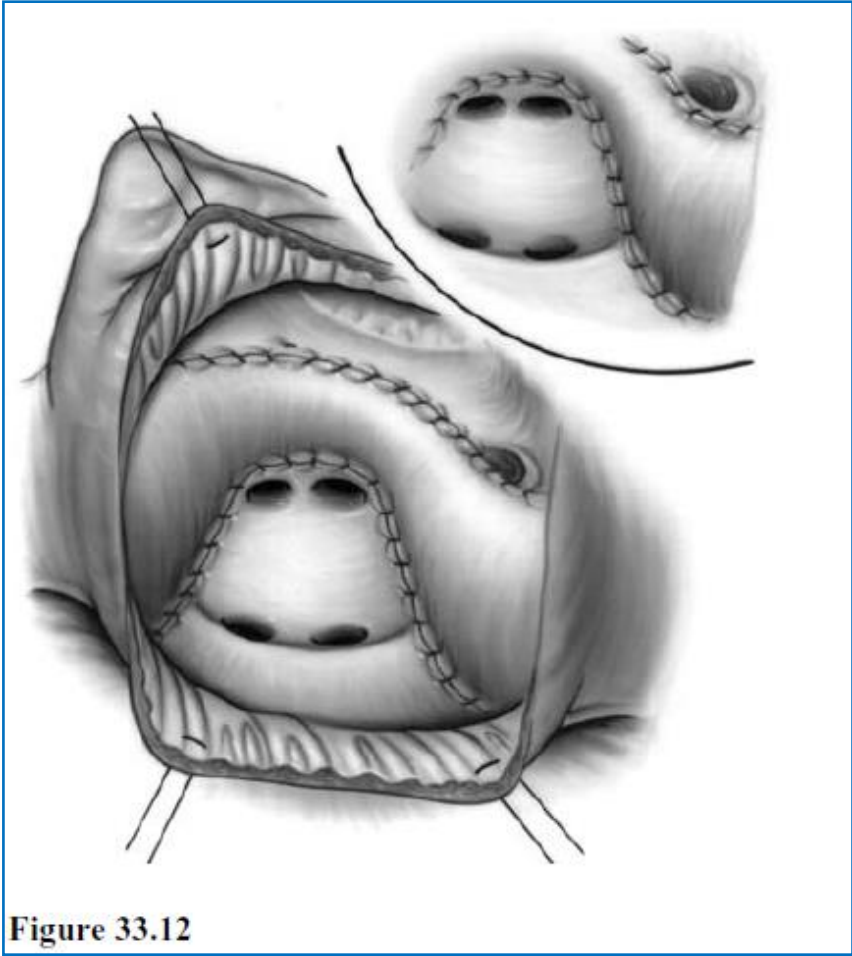
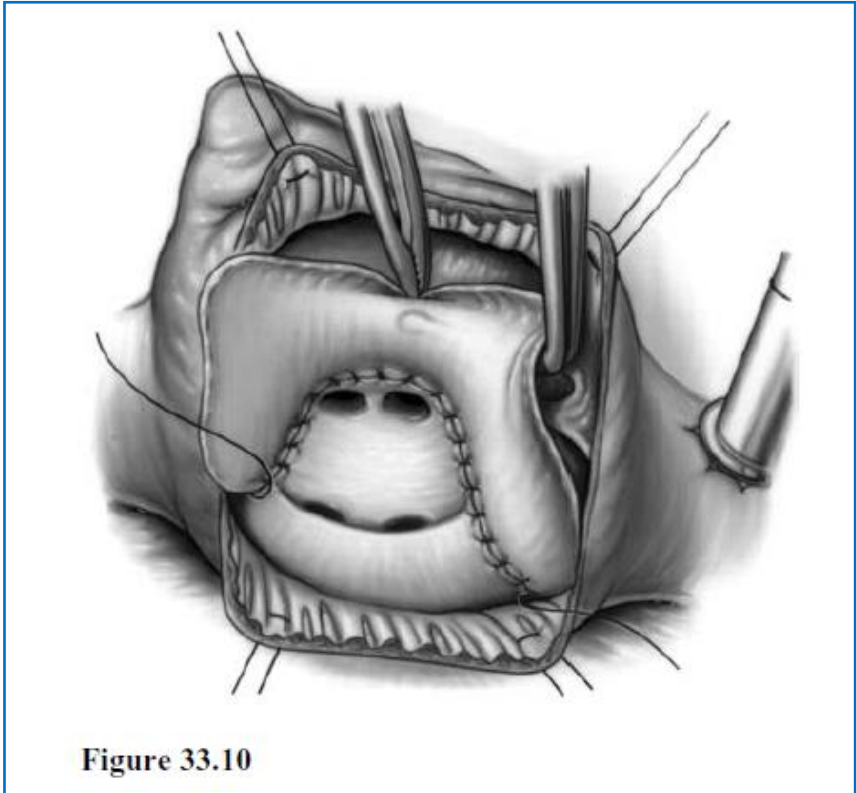


Figure 33.8



Disadvantages of Atrial Switch

- SVC obstruction - Mustard > Senning
- Supraventricular arrhythmias
- Baffle leaks
- Tricuspid insufficiency
- Late RV failure

TGA, VSD and PS

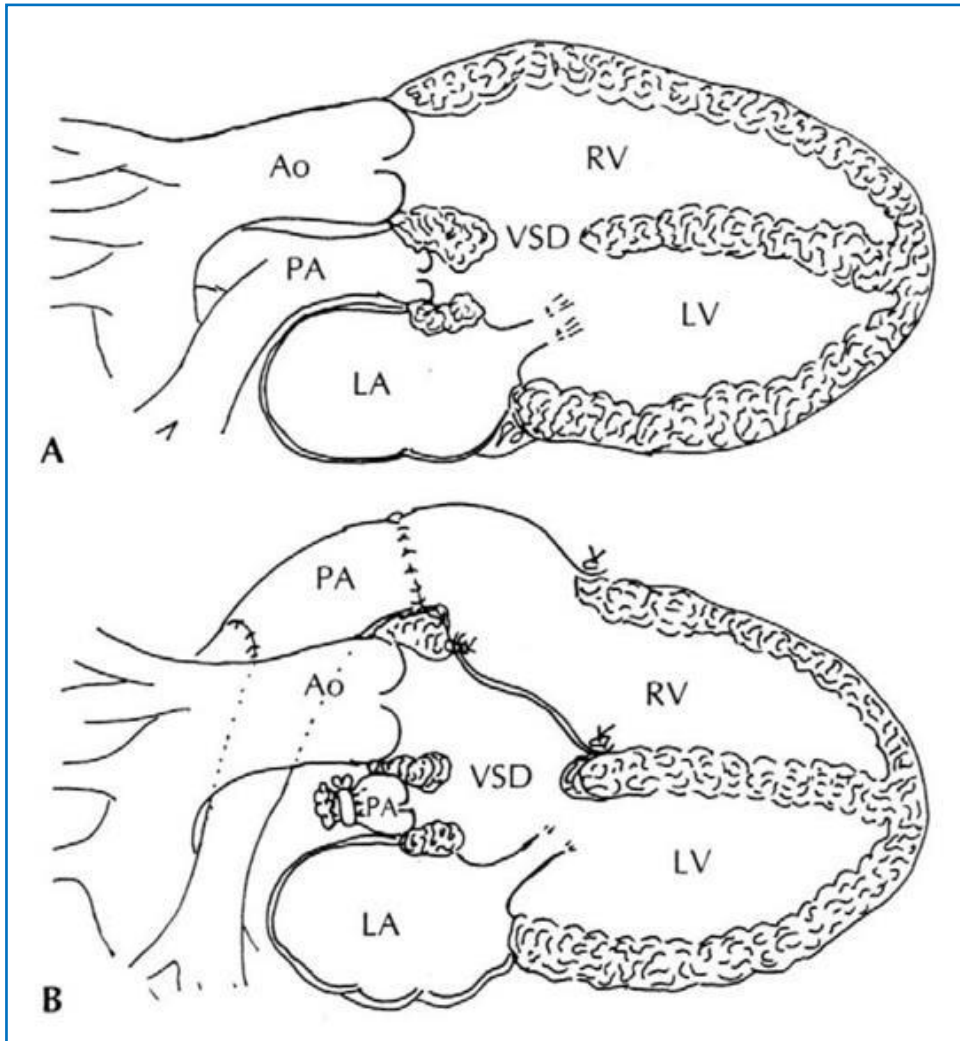
- Shunt when symptomatic as infant
- Rastelli repair - 1-2 years of age
- Operative mortality = 5-10%
- Late reoperation for conduit replacement
- Atrial Septectomy

TGA, VSD and PS

- Goals of surgery
 - 1.Unobstructed LV outflow tract / RV outflow tract
 - 2.Competent aortic and pulmonary valve
- Surgical options
 - 1.Arterial switch \pm LVOTO relief
 - 2.Rastelli procedure
 - 3.REV procedure
 - 4.Nikaidoh procedure or its variants
 - 5.Pulmonary root translocation

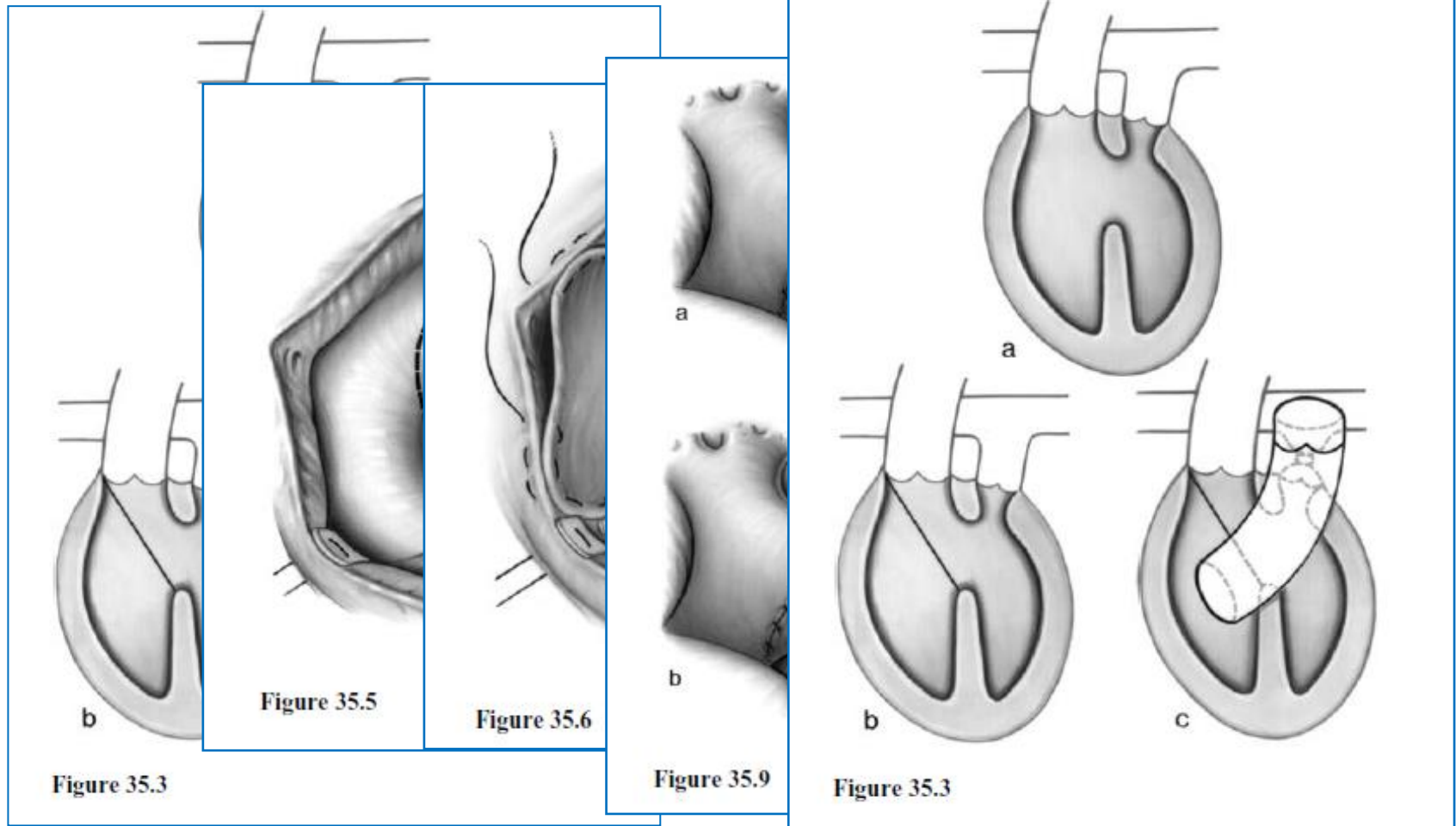
Rastelli Procedure

- LV to aorta baffling \pm VSD enlargement
- RV to pulmonary artery (valved) conduit
- Poor long-term survival reported
- LVOTO, RVOTO, arrhythmia, sudden death

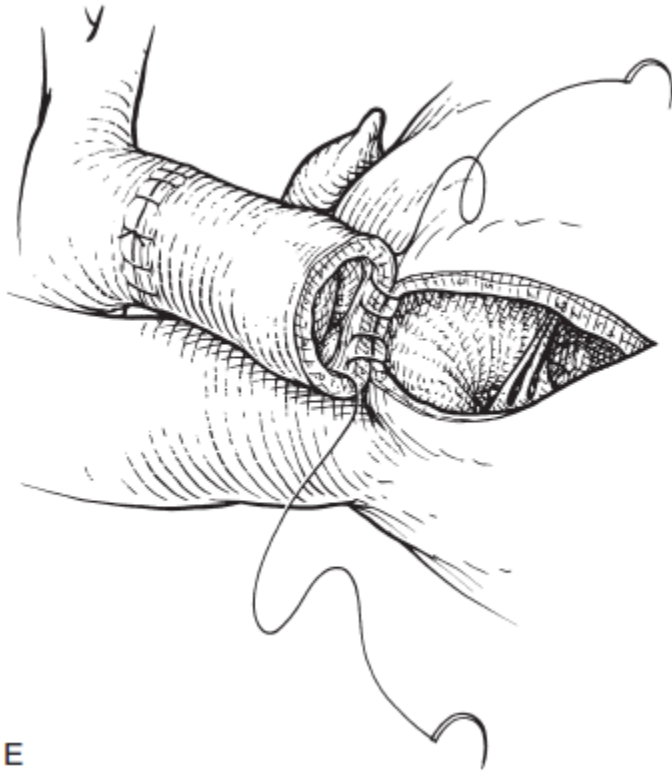


Ann Thorac Surg 2005;79:2089-93

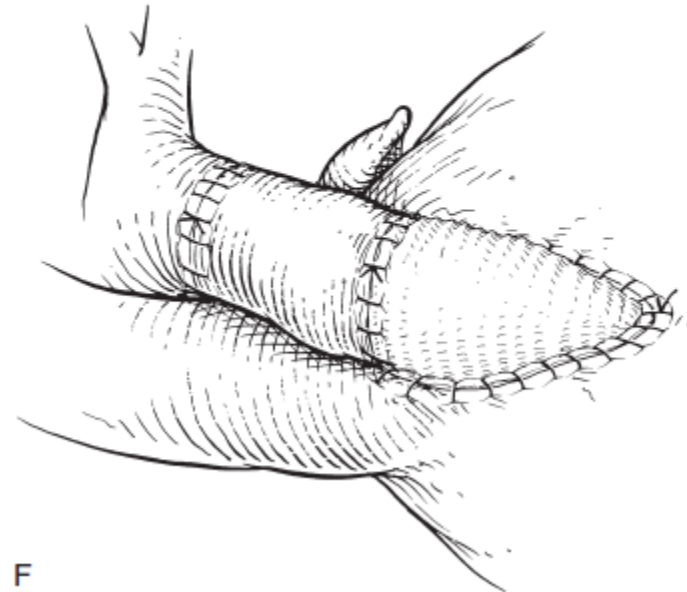
Rastelli operation



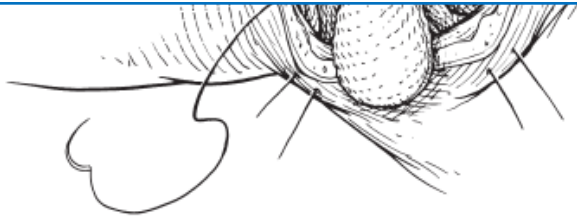
Pulmonary trunk



E



F



C

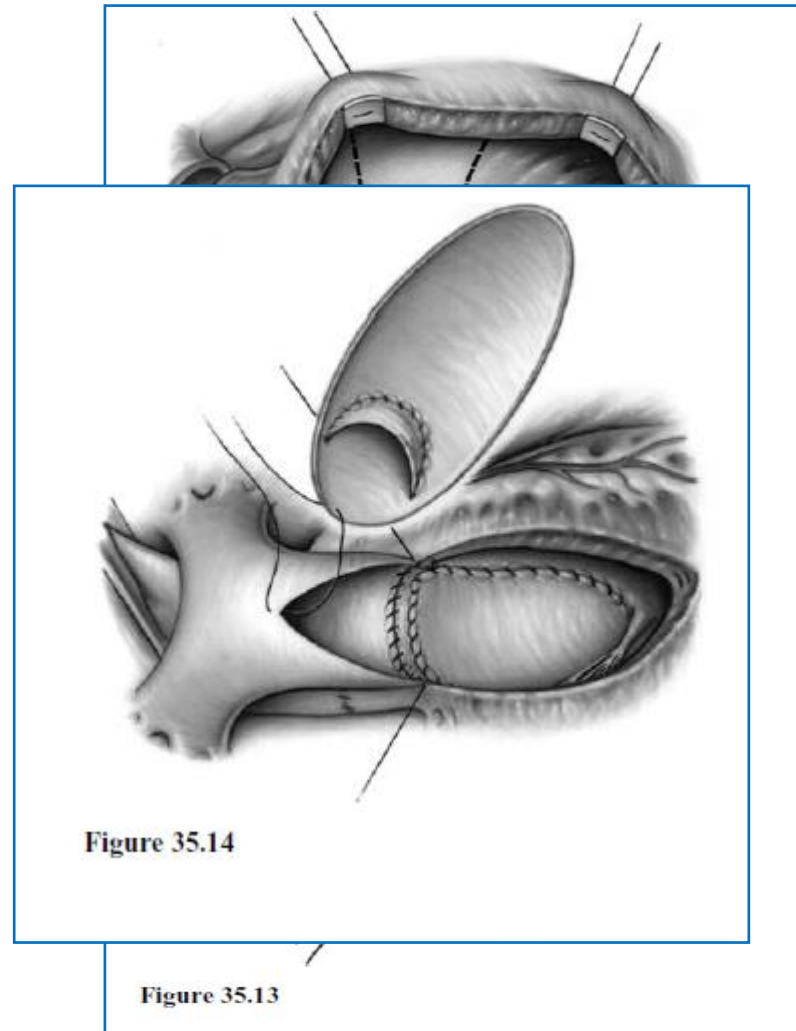
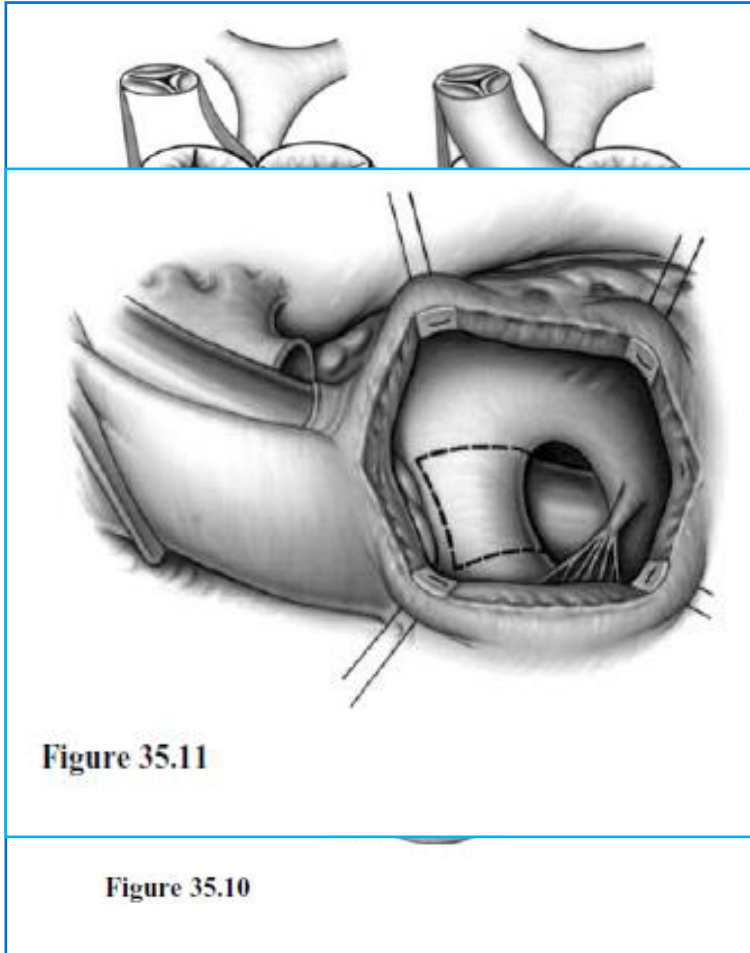


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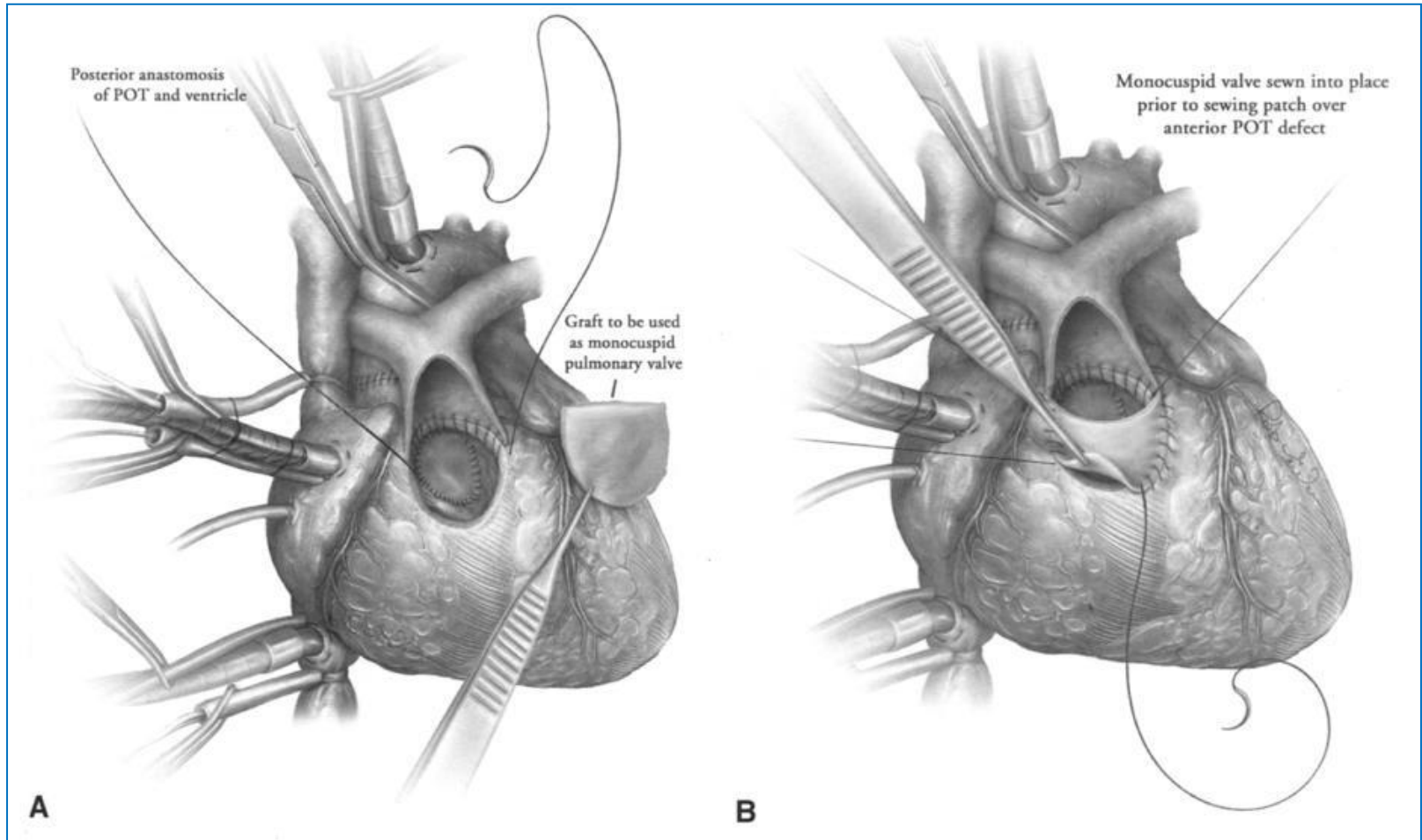
REV Procedure

- Aggressive VSD enlargement to prevent LVOTO
- Avoidance of the use a RV to PA conduit

REV Procedure

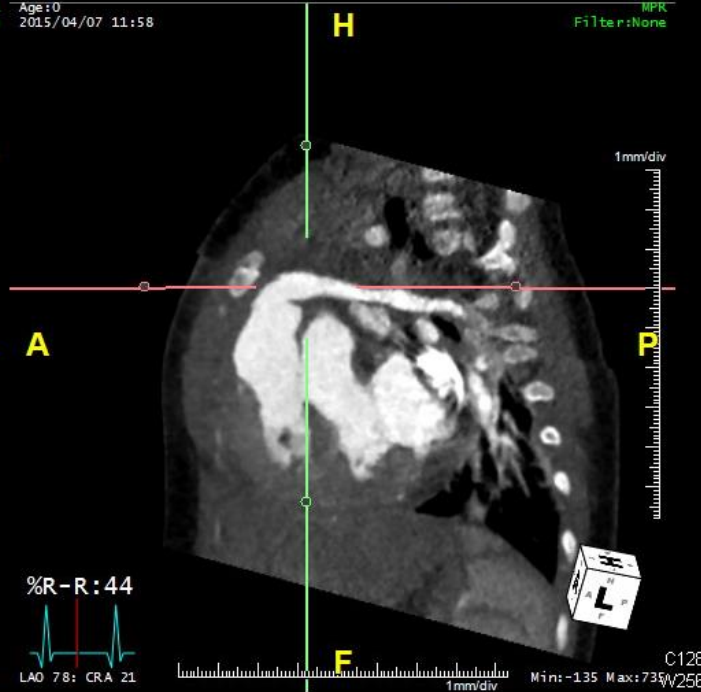
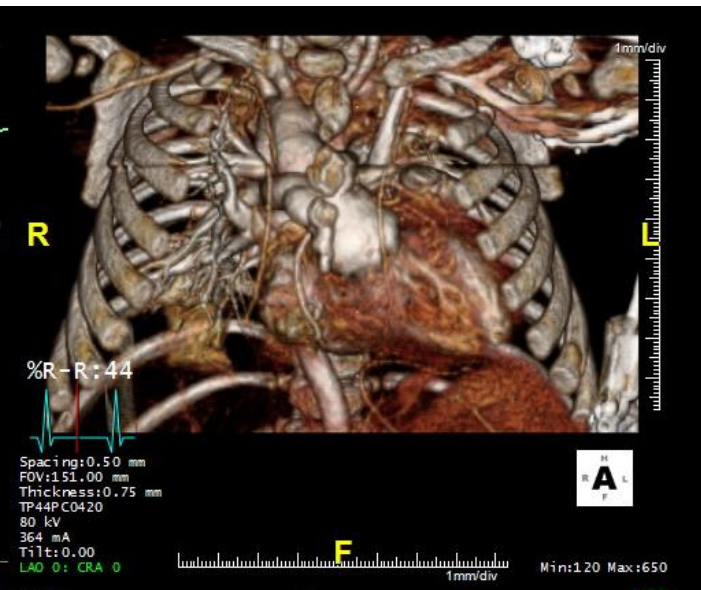
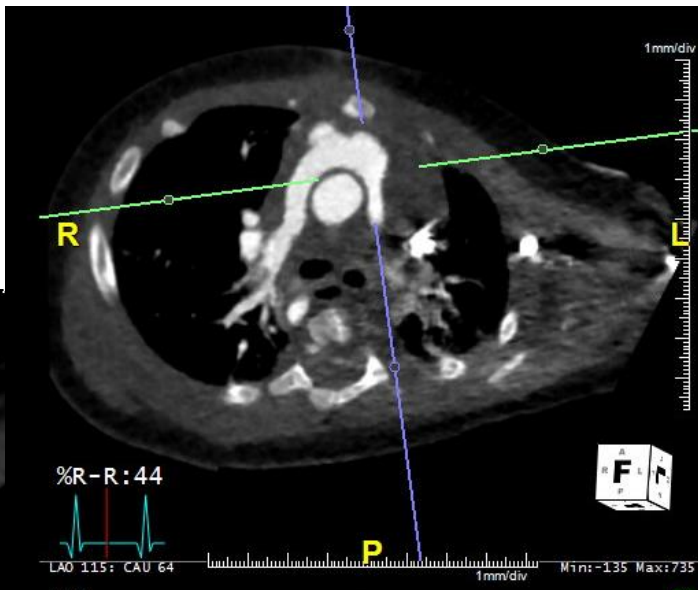


REV Procedure (RVOT Reconstruction)



Nikaidoh Procedure

- Aortic translocation
- Half-turned truncal switch
- Double root translocation
- Ross-Switch-Konno

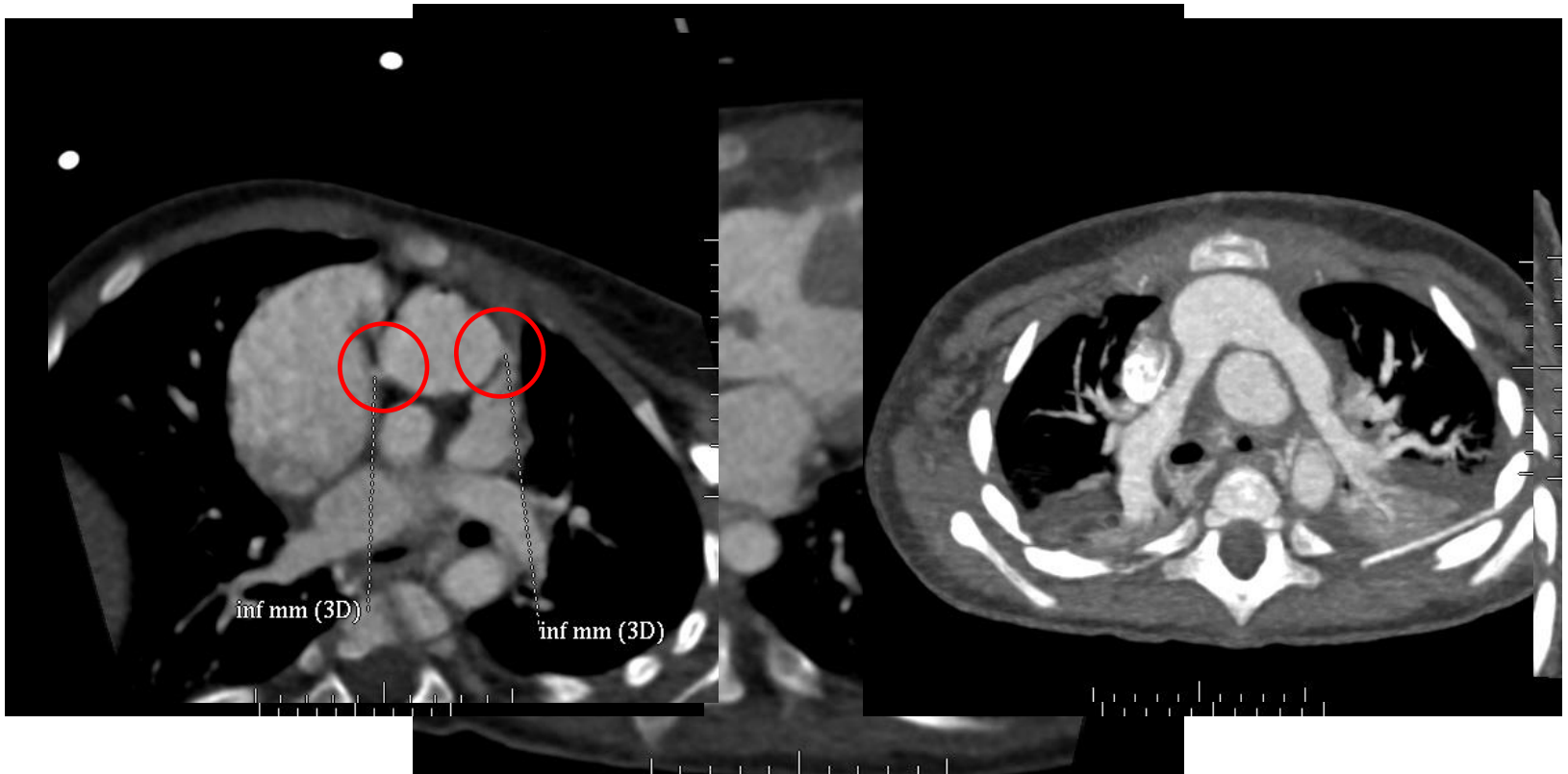


Patient

- 20Mo / 9.8Kg / M
- TGA with
 - VSD (PM)
 - PS (valvar and subpulmonary by post. Deviated conal septum)
- 9 days: BAS
- 22 days: RMBT

Surgical Options

Rastelli or REV
Nikaido, Nikaido Modification
Nikaido op.



Aortic root translocation

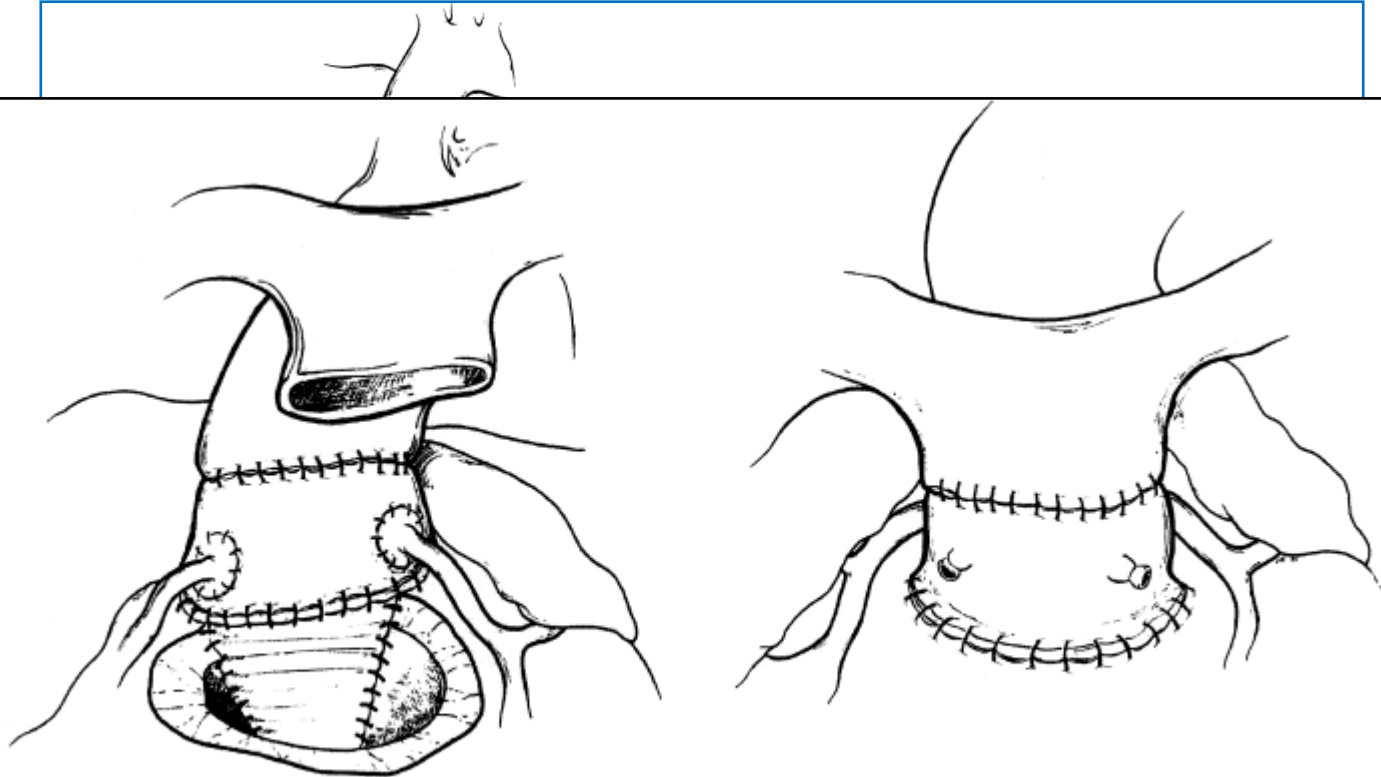
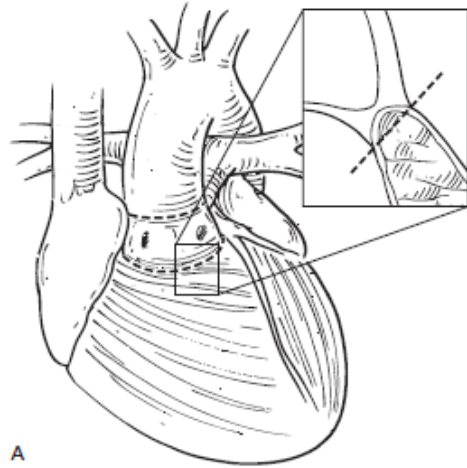
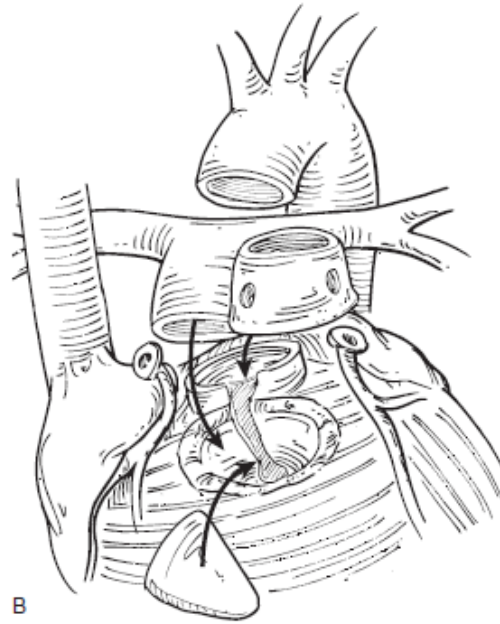


Figure 125-11

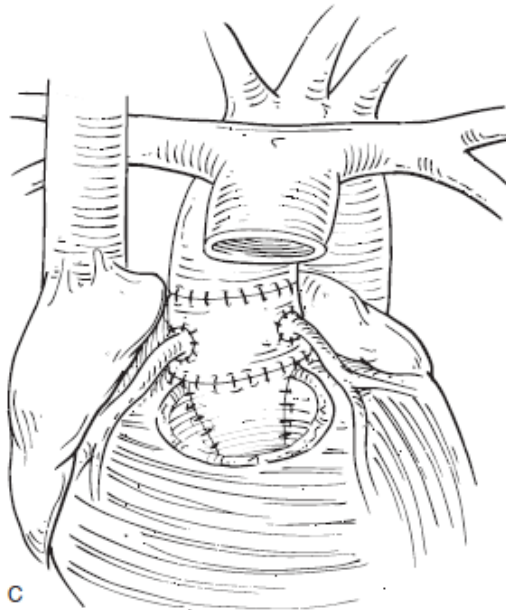
Aortic root autograft. Right ventricle to pulmonary conduit, with the pulmonary arteries brought anterior to the aorta.



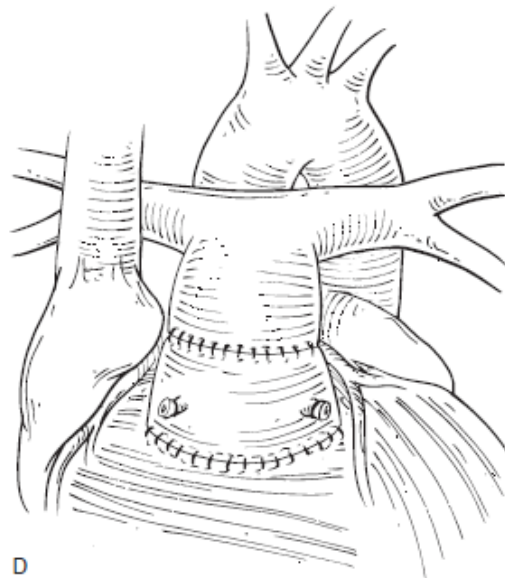
A



B

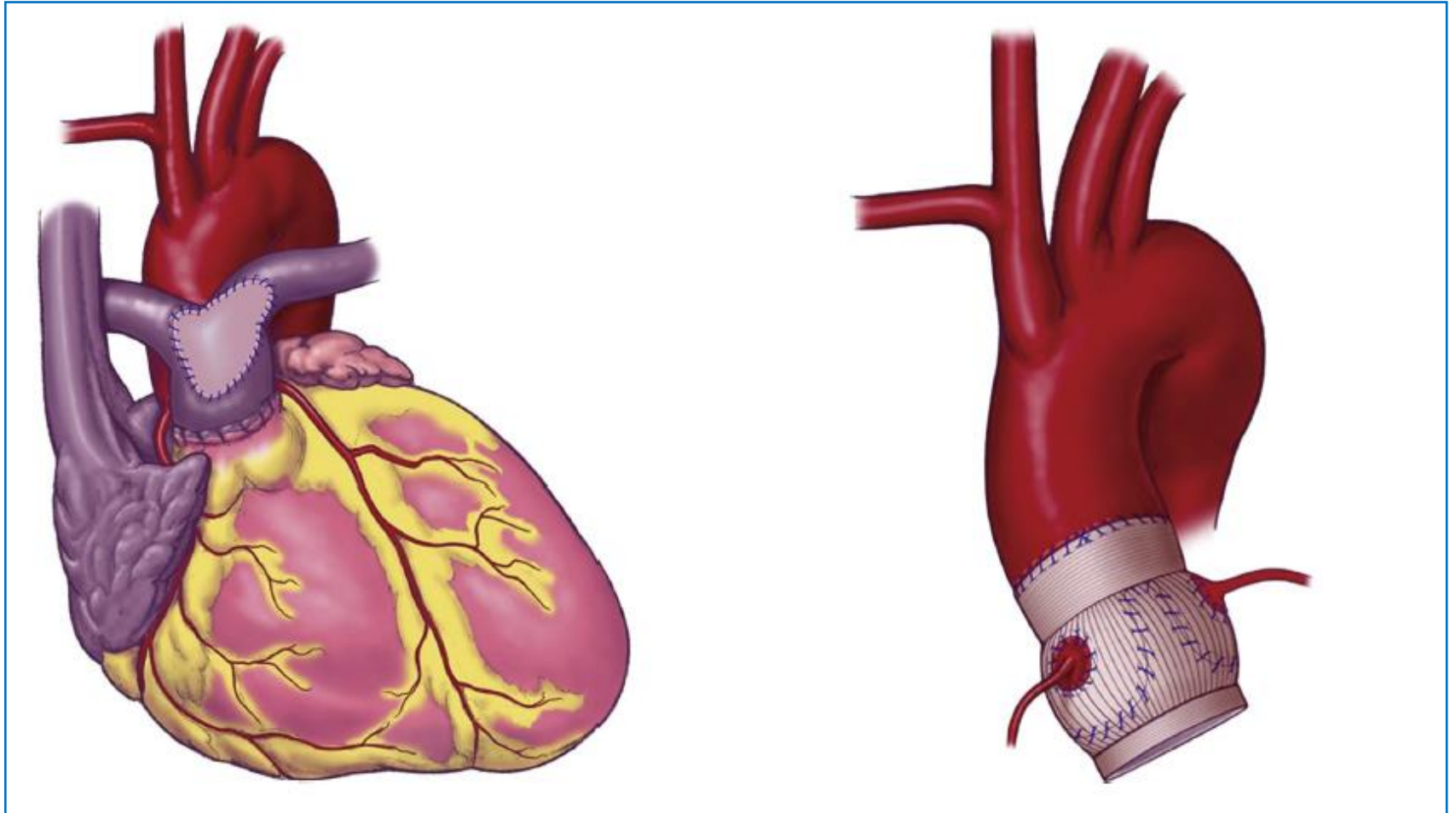


C



D

Reoperation After ASO



Reoperation After ASO

- Supravalvular pulmonary stenosis

- **The most common cause for re-operation (7-28%)**

- Possible causes

- Pericardial patch constriction

- Anastomotic site tension

- Suture purse-string effect

- Inadequate dissection of pulmonary artery

Reoperation After ASO

- Supravalvular pulmonary stenosis

- Double pericardial patch -- should be avoided(Paillole, 1988)
- A single large pantaloons pericardial patch
- More extensive dissection and mobilization of pulmonary a. into the hila (Yamaguchi, 1990)
- Balloon angioplasty for postoperative pulmonary artery stenosis(Nakanishi, 1993) -- 50% success rate

Reoperation After ASO

- **Factors** implicated for neo-aortic insufficiency
- - Previous pulmonary artery banding
 - Thickening and deformity of the pulmonary valves and sinuses
- - Aortic root dilation
 - Distortion of the commissure
 - Deformity of the neo-aortic root
 - Discrepancy in size between the PA and aorta
 - Regurgitation preoperatively

Reoperation After ASO

- Supravalvular pulmonary stenosis
- Neoaortic insufficiency
- **Coronary stenosis**

- 2 – 11%
- Cause of early death and morbidity
- Late coronary events – < 2%

학습목표

- TGA의 수술시기를 알 수 있다
- Coronary artery pattern을 기술할 수 있다
- Atrial switch operation을 이해한다
- TGA,VSD,PS의 수술방법을 구분할 수 있다
- Arterial switch operation후의 long-term complication을 이해할 수 있다

Summary

- TGA/IVS simple - Arterial switch < 14 days
- TGA/IVS simple with intramural L main - Mustard/Senning
- TGA/VSD - neonatal switch and VSD closure
- TGA/VSD/PS - shunt; Rastelli 1-2 years

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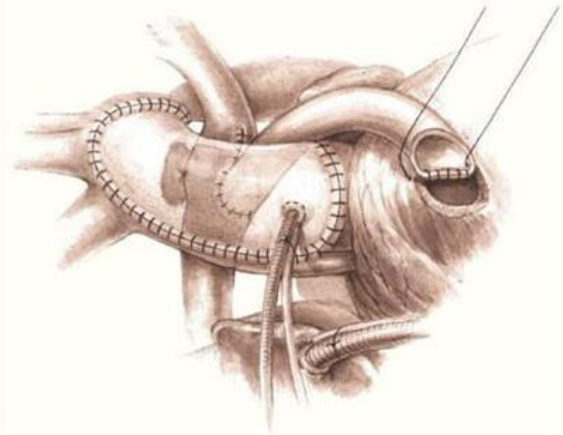
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