Tetralogy of Fallot

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Outline

- Historical Note
- Embryology
- Morphology
- Clinical Features and Diagnosis
- Medical Management
- Indications for and Timing of Surgery
- Surgical Management
- Special Topics in Surgical Management
- TOF with Absent Pulmonary Valve
- TOF with Complete AVSD
- Results of Surgery
- Pulmonary Atresia with VSD

Historical Note

- First description by French physician Etienne Fallot in 1888
- First surgical procedure

  by Alfred Blalock at Johns Hopkins University in 1945
- First successful intracardiac repair (with human cross-circulation)

  by Lillehei at the University of Minnesota at 1954
- First successful repair with a pump oxygenator

  by Kirklin at Mayo Clinic in 1955
- Patch enlargement of RV infundibulum by Warden and Lellehei in 1957
- Transannular patching by Kirklin in 1959
- RV-pulmonary trunk conduit fot TOF/PA by Kirklin in 1965
- Valved extracardiac conduit by Ross in 1966

Embryology

- ***Peripheral PA*** ; from systemic arterial and venous circulations of the primitive foregut
- ***Proximal mediastinal PA*** ; from 6th dorsal arches
- ***Proximal MPA*** ; formed by division of the original conotruncus

Two theories of development of TOF

- Classic theory ; **Unequal spiral septation of conotruncus**
- Van Praagh's theory ; **Underdevelopment of subpulmonary conus**

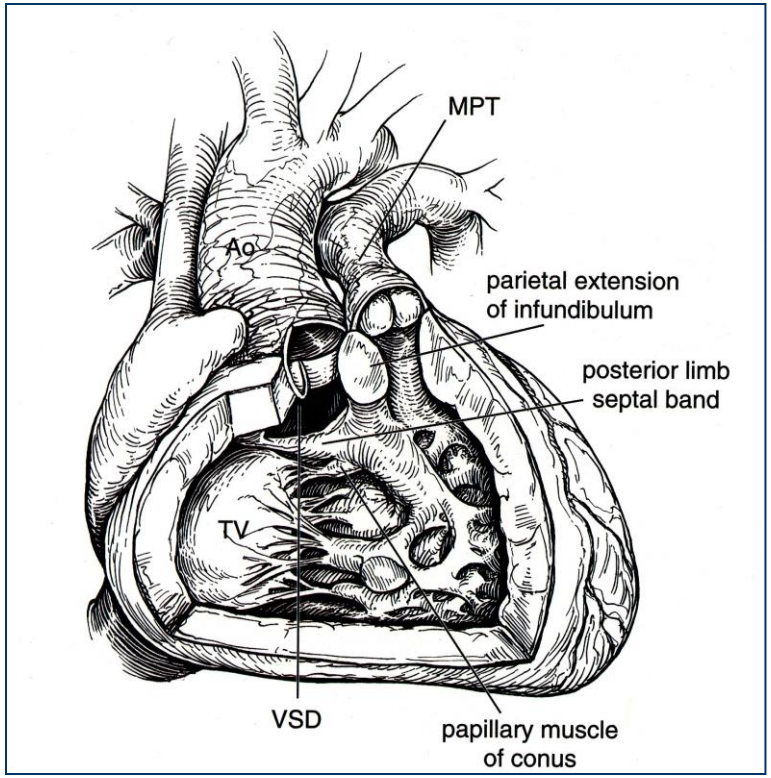
Morphology

One basic morphological abnormality;

Anterior and leftward displacement of the infundibular (conal) septum

- **Overriding of aorta**
- **RVOT obstruction**
- **Large VSD**

TOF

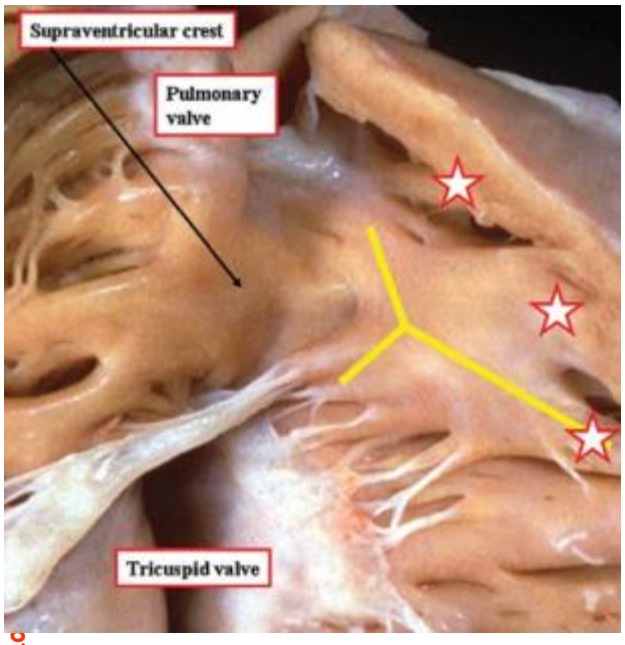


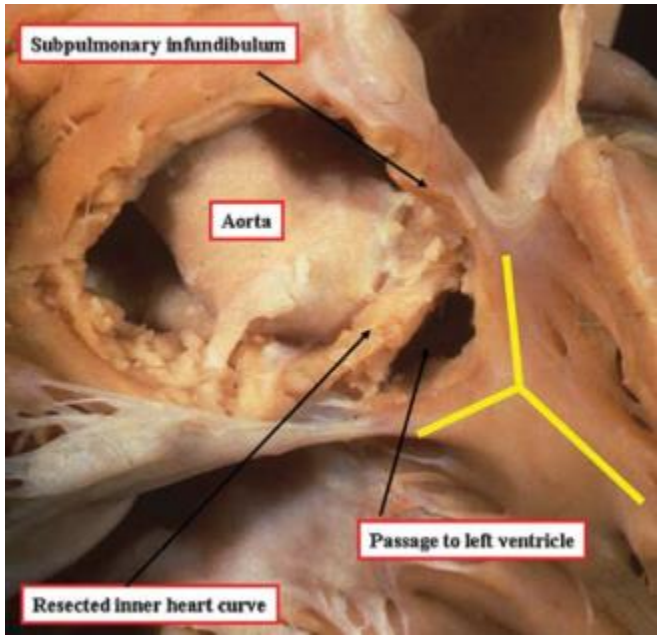
Morphology

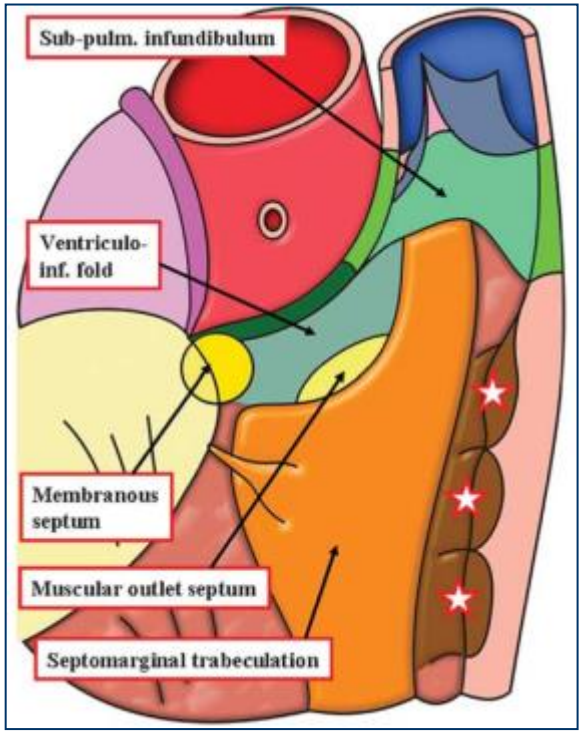
Normal Heart

"Crista supraventricularis (Supraventricular crest)"

defined as the muscular area separating the attachments of the tricuspid and pulmonary valves in the roof of the right ventricle







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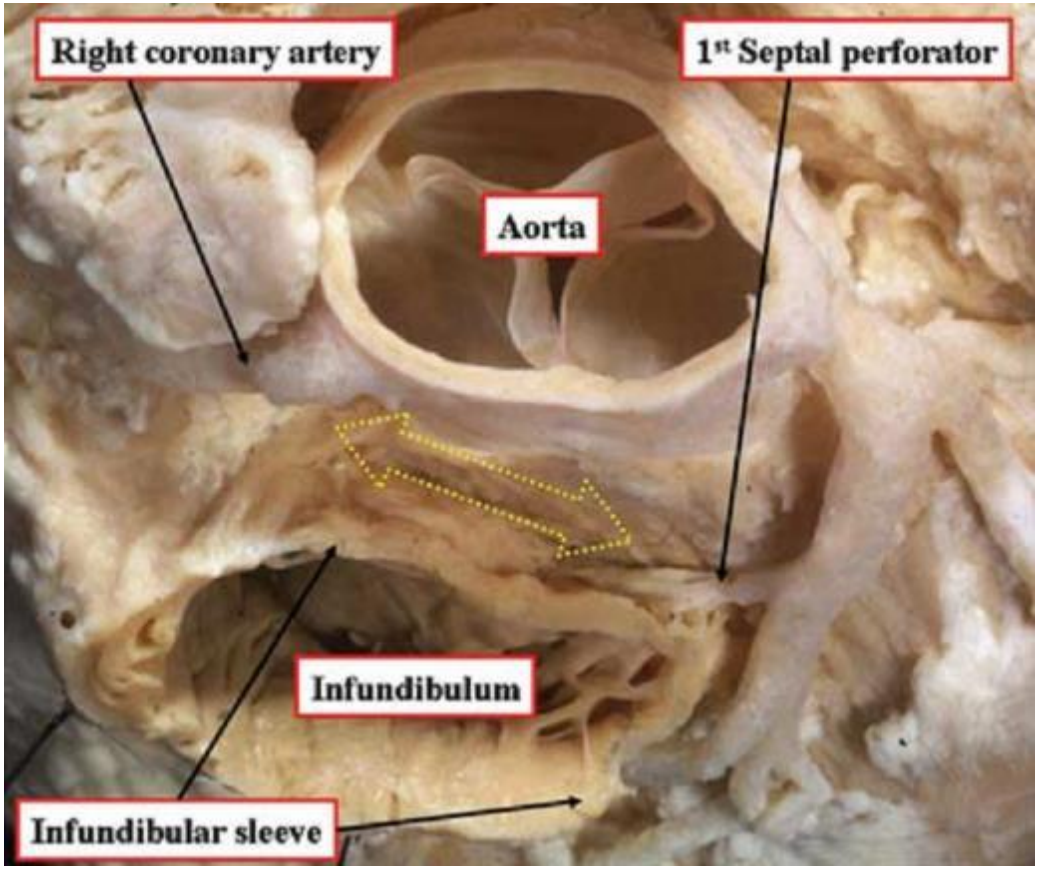

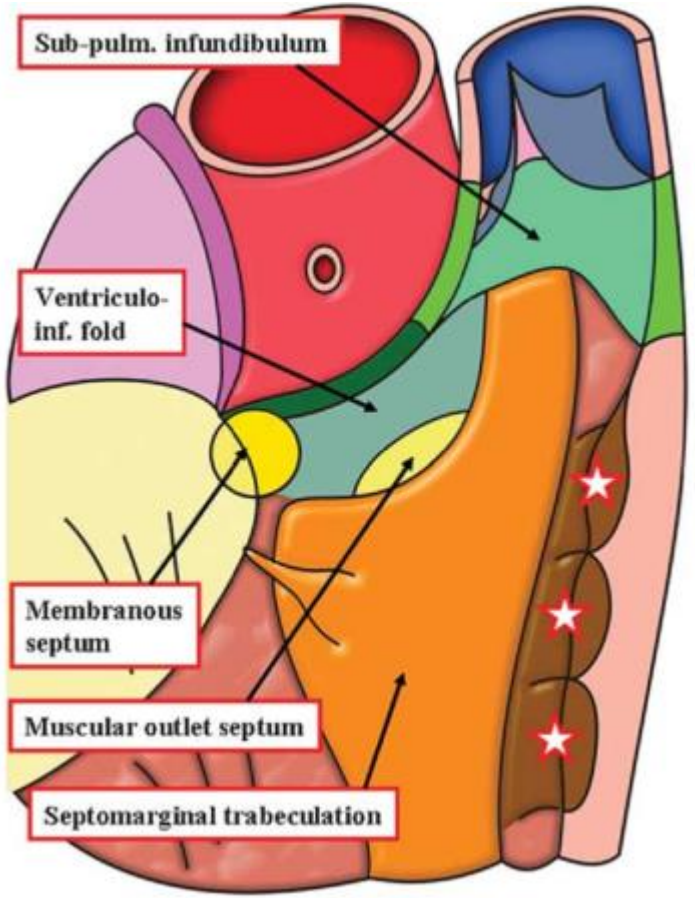

Morphology

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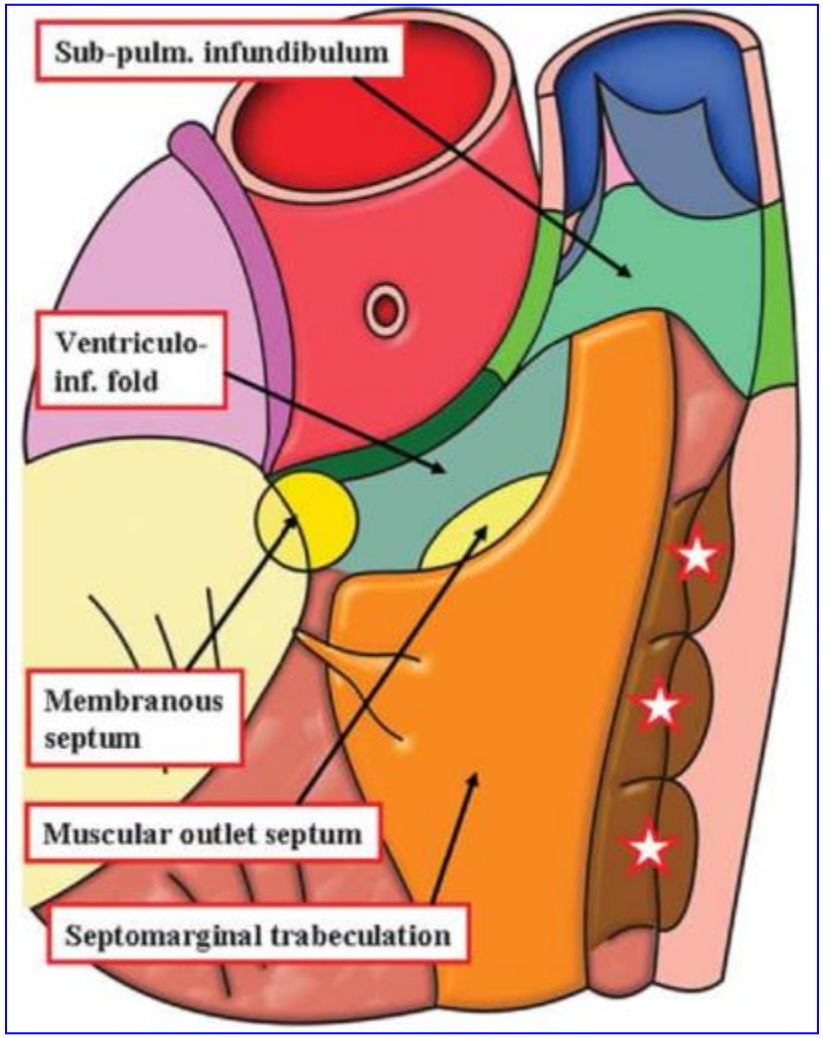

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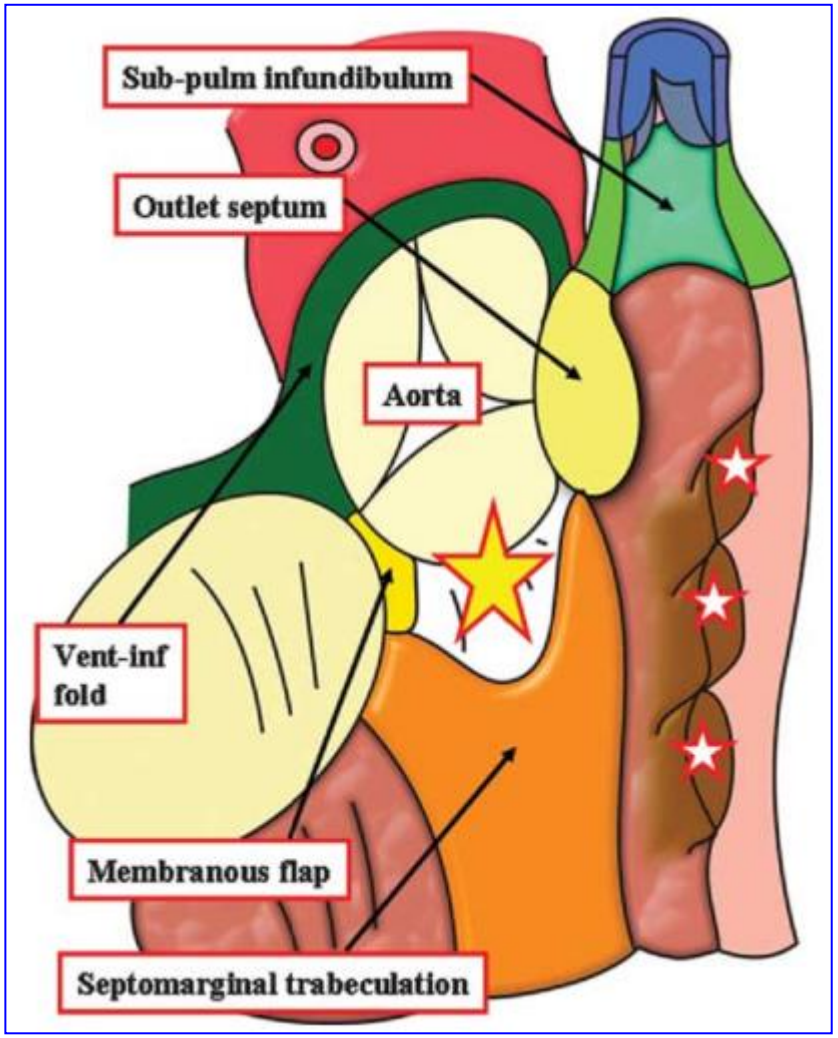

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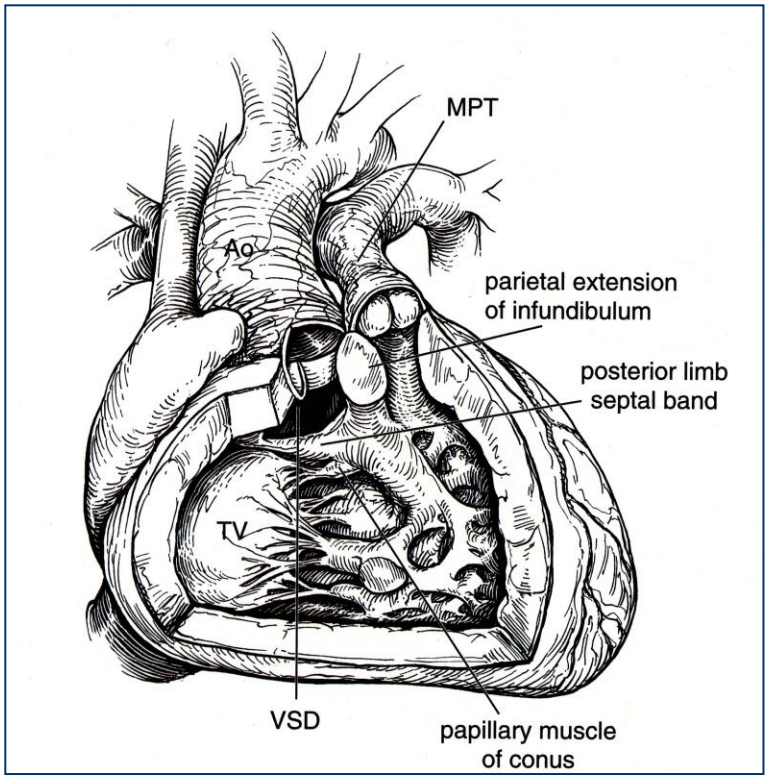
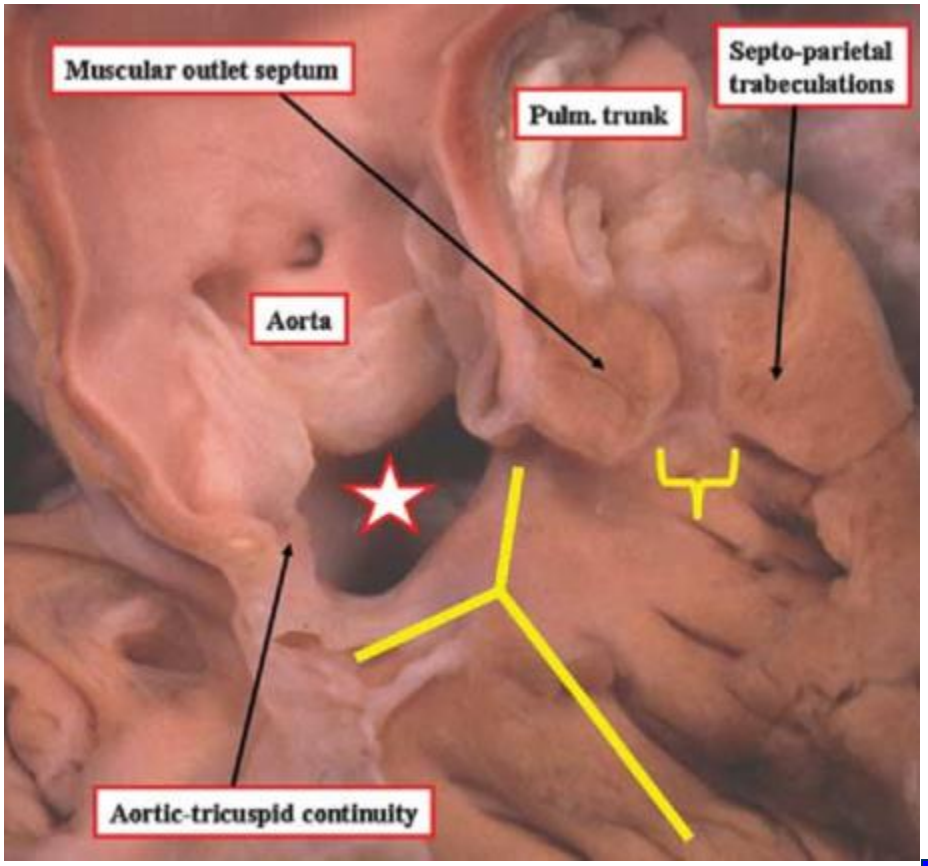


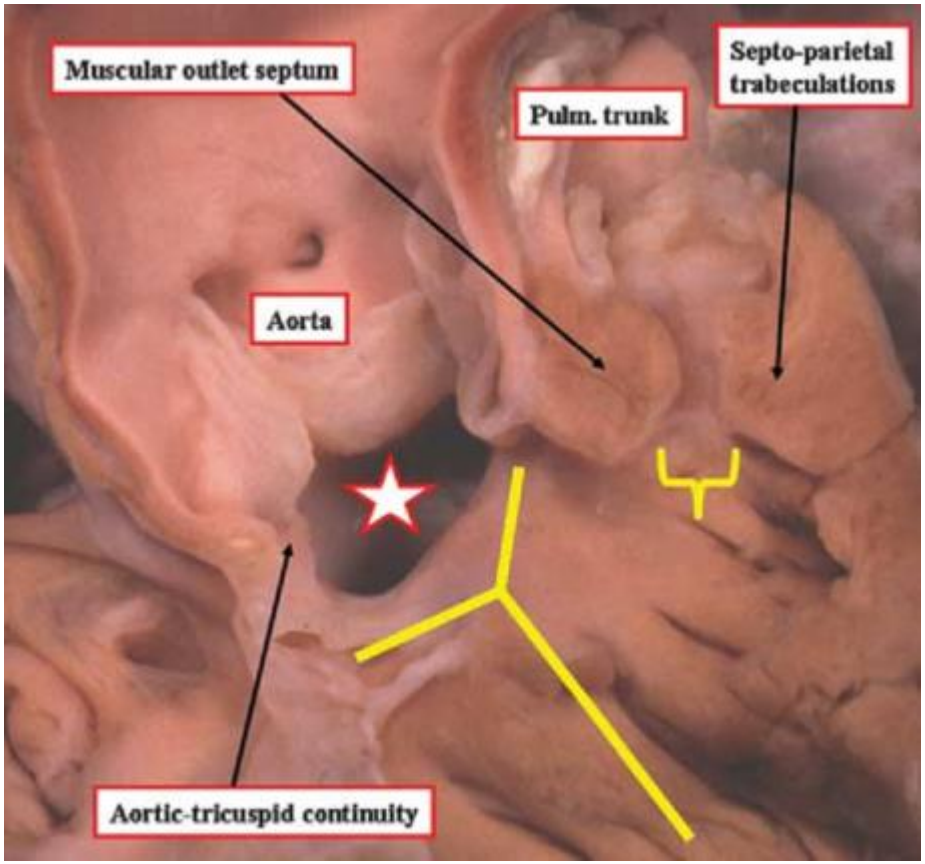


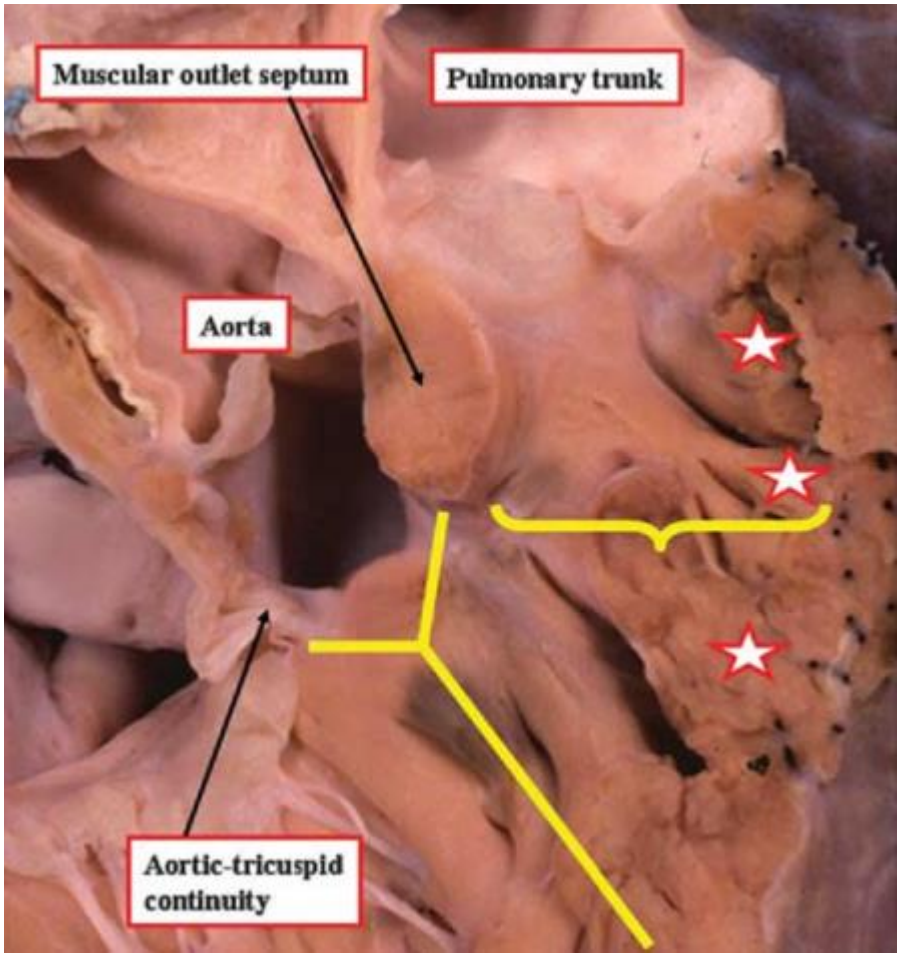
Tetralogy of Fallot

Morphology

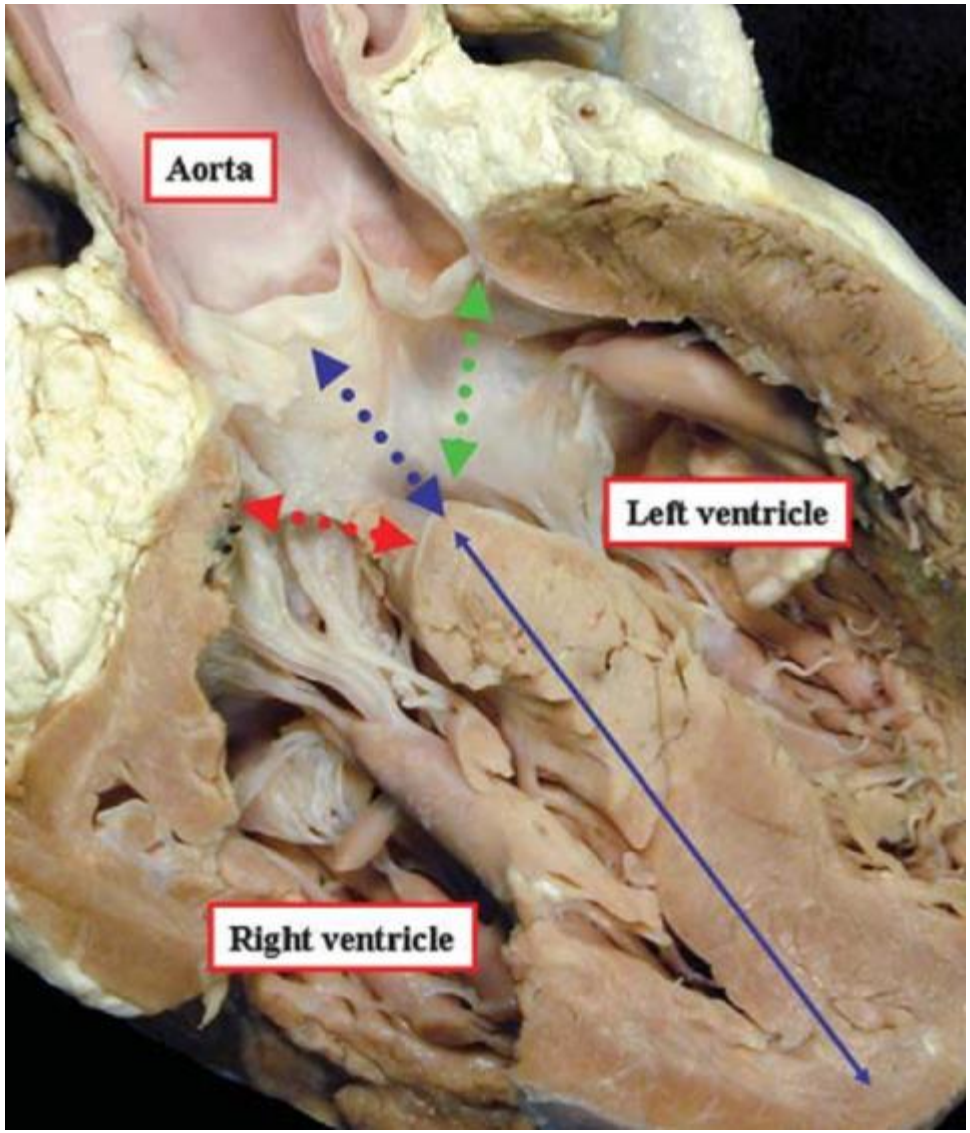
Anteriorly malaligned VSD (Eisenmenger defect)

TOF



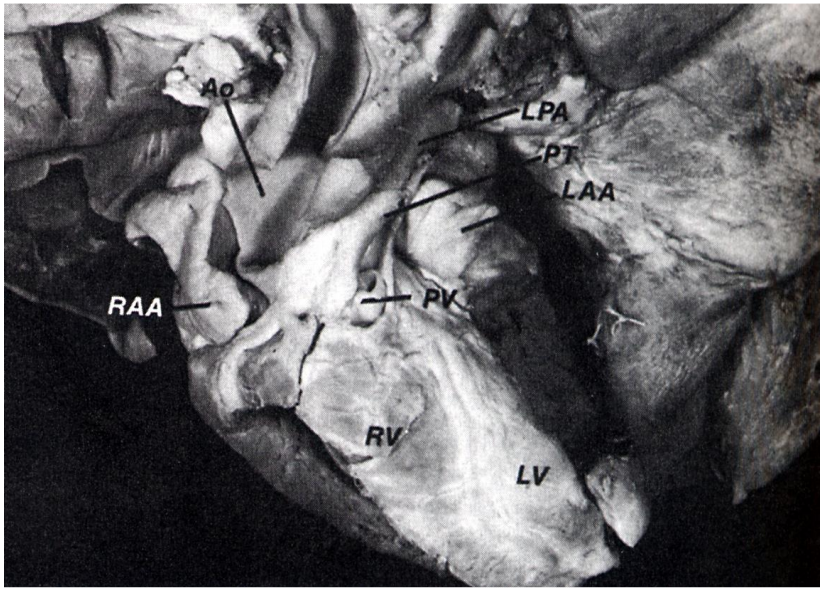


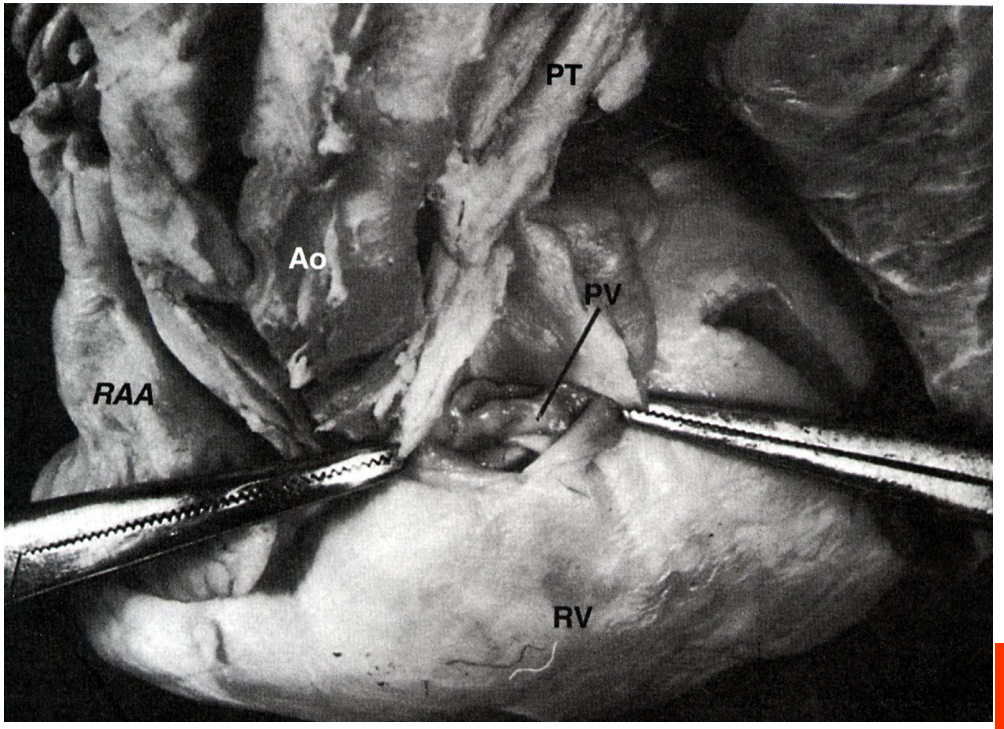
Overriding of the aortic valve



Pulmonary valve and annulus

- Stenotic in 75%
- Leaflets are usually thickened, tethered to the pulmonary artery
- Bicuspid in 75% of cases
- The narrowest part of the outflow tract in only small percentage





MPA and branch PAs

- Usually somewhat diffusely small, often short
- Narrowest portion of MPA is often at sinotubular junction
- Branch PA stenosis in 10%, bilateral in an half of these
- Reduced intraacinar arteries, alveolar size, and alveolar numbers

VSD

- Large anteriorly malaligned
- 25% ; VIF extends to the posterior limb of TSM
  muscle bar beneath tricuspid valve (muscular outlet)
- Additional VSDs in 3-15%

Conduction system

- *SA node, AV node* ; normal in location
- *Bundle of His* ; passes close to the crest of interventricular septum or slightly to the left of the inferior margin of the VSD

Coronary arteries

- 5%, LAD from RCA, dual LAD (one of these from RCA)
- very occasionally, RCA from single LCA, LCA from single RCA
- crossing over RVOT, rarely in the myocardium (not visible)
- can be detected preoperatively by echocardiography

Other anatomic features

- **Pulmonary atresia: 7%**

- **Absence of pulmonary valve leaflet; 5%**

- **25%, right aortic arch**
- **PFO, common, 10%, ASD**

Incidence, Associated Anomaly & Natural History

- 3 / 10000 live birth
- 7-10% of CHD
- M>F
- Genetic defect in 28%

  Chromosome 22q11.2 defect

  Trisomy 21

  VACTERL (Vertebral anomaly, Imperforate anus, Cardiac anomaly, TEF, Esophageal atresia, Renal anomaly, Limb defect)
- 25% die before 1 year of age, 40% before 3 years, 70% before 10 years, 95% before 40 years

Clinical Features and Diagnosis

Presentation

- Severity of RVOTO = Degree of R to L shunt = Degree of cyanosis = Presentation age
- Mild RVOTO → predominant L to R shunt → CHF (pink TOF)
- Most commonly, cyanosis is mild at birth and gradually progresses with age
  (increasing hypertrophy of the RV infundibulum)
- Mechanism of "cyanotic spell" initiation ;
  Reduction of cardiac afterload or preload and tachycardia
  (dehydration, viral infection, vasodilatation by medication, ect)

Laboratory studies

- Routine blood sampling for chromosome 22 (22q11.2) deletion
  10% of TOF/PS have such a chromosomal deletion
  higher incidence in TOF/PA, a right aortic arch and aberrant left subclavian artery

Echocardiography

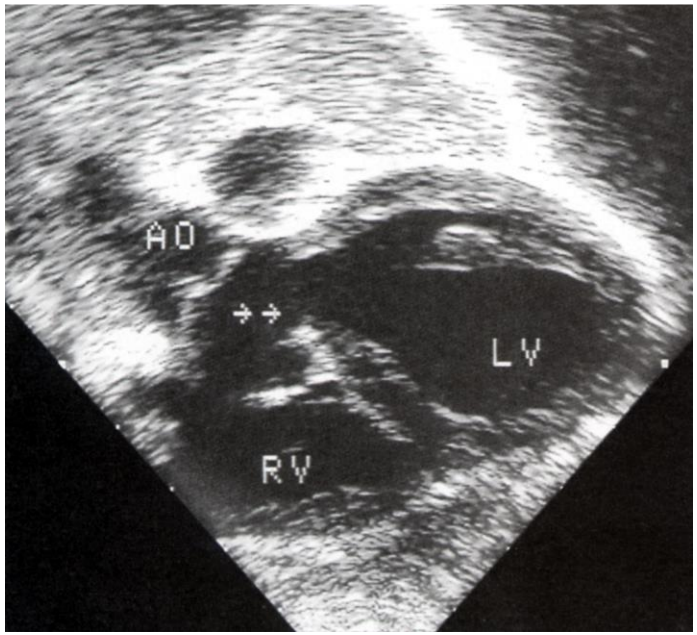
- Usually definitive diagnostic modality
- Detailed elucidation of the important anatomic features including ;
  - number and location of VSDs
  - nature of RVOT
  - anatomy proximal branch PA
  - coronary artery pattern (esp. CAs crossing RVOT)
- Fetal echocardiography ;
  The ultimate impact on survival has not been clearly established

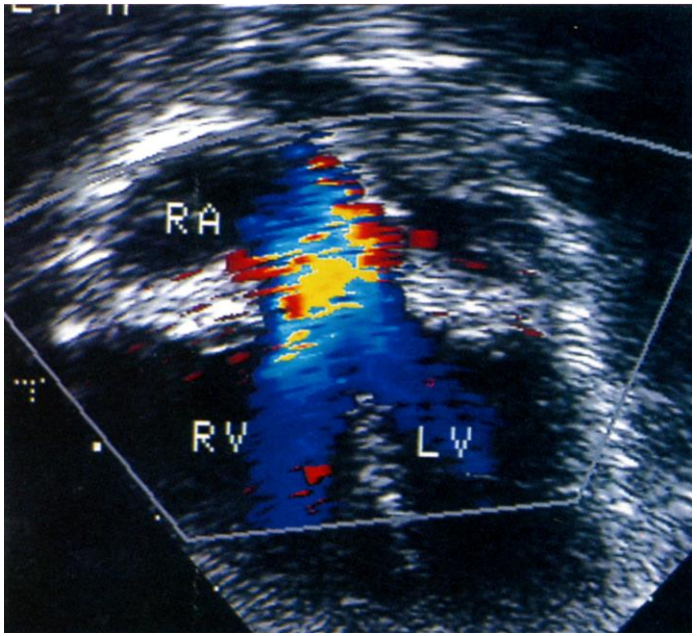
Cardiac catheterization

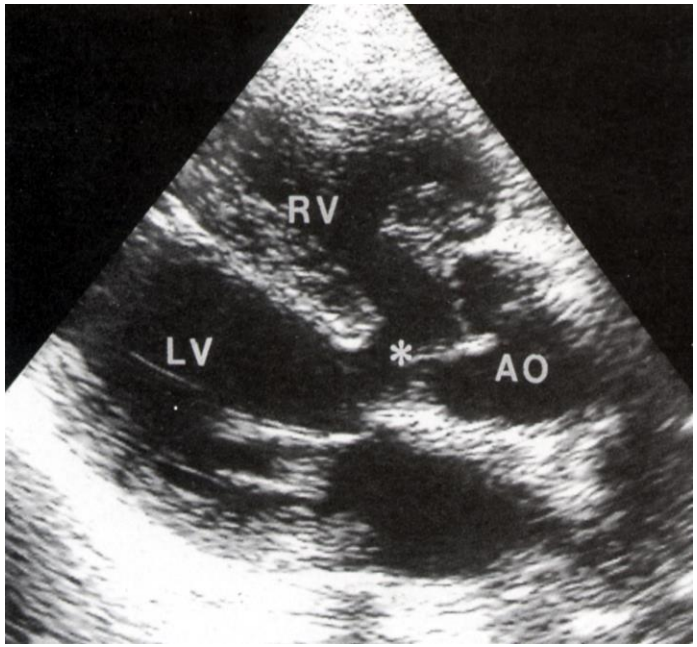
- Rarely indicated in most TOF/PS
  Indication;
  - Concern about important aortopulmonary collateral vessels
  - Multiple VSDs are suspected
- Inducing spasm of RVOT →Urgent operation

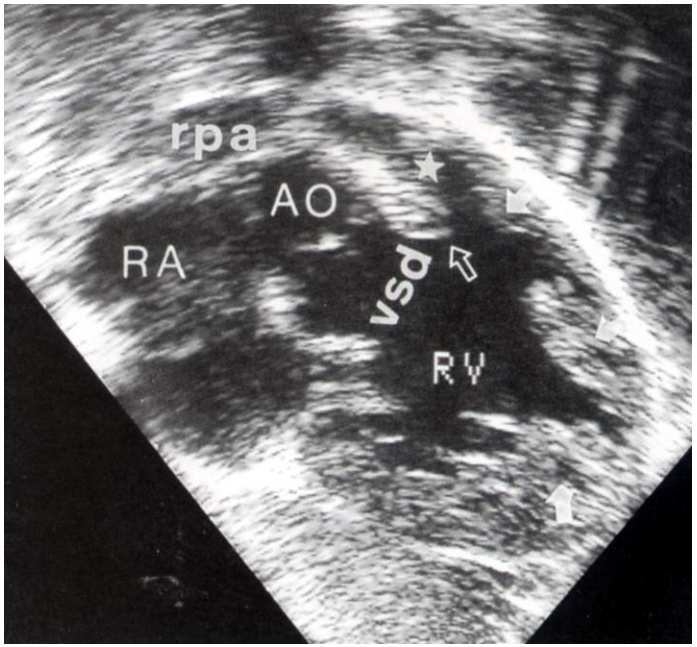
Clinical Features and Diagnosis

Echocardiography

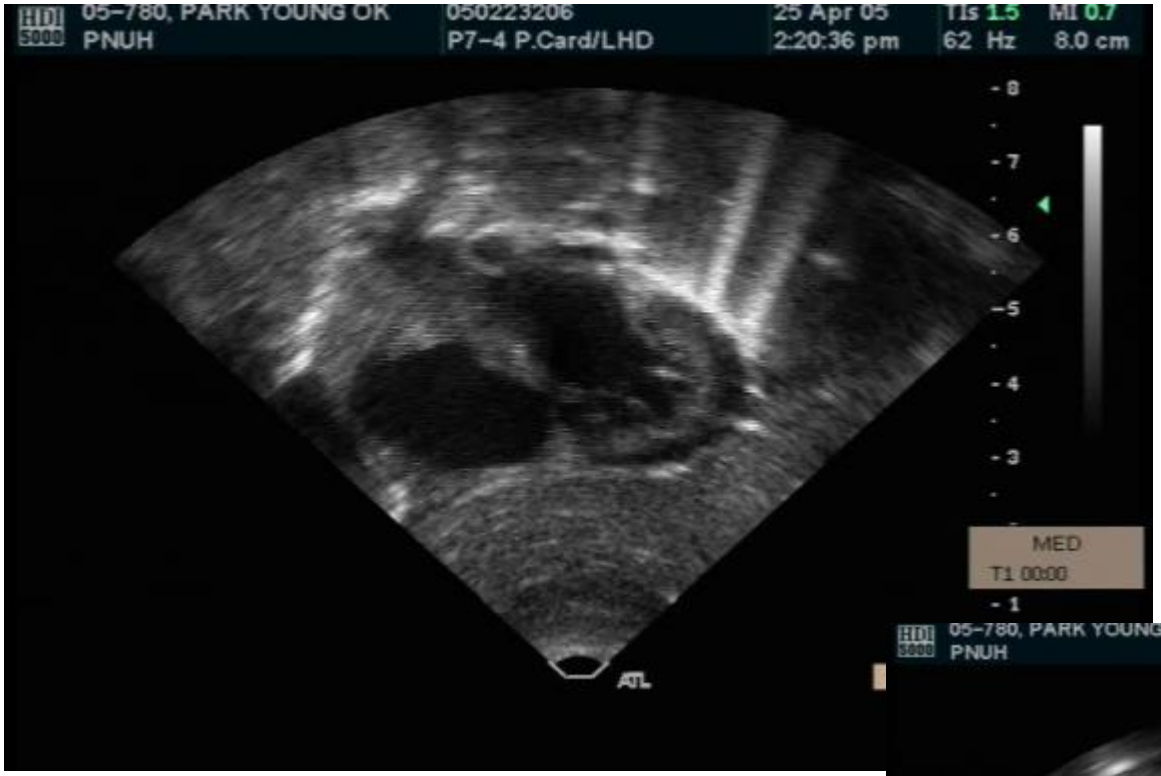


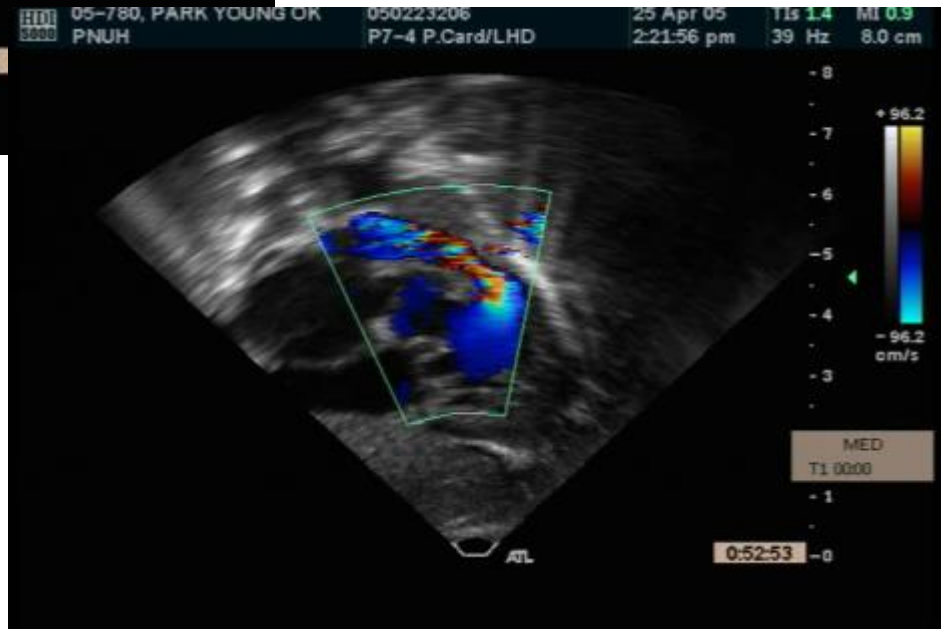






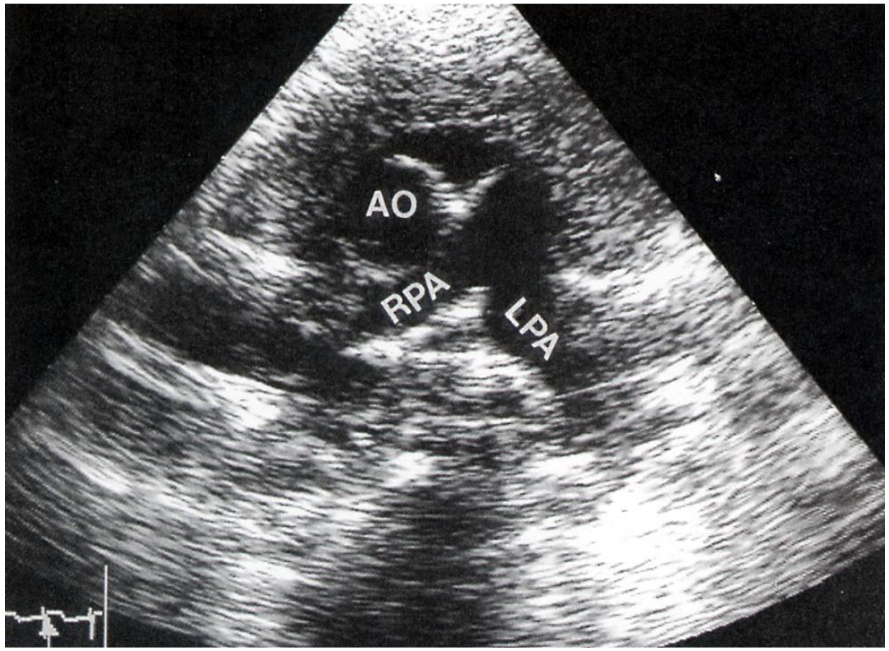
Clinical Features and Diagnosis

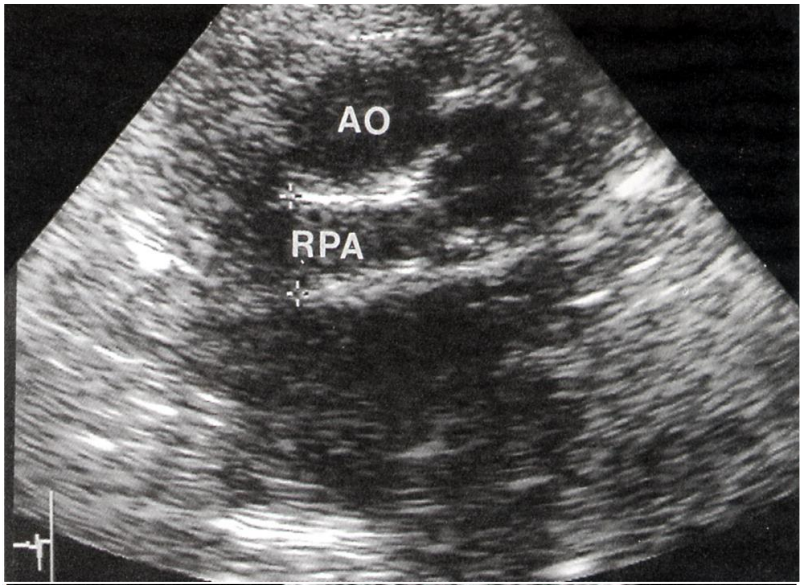


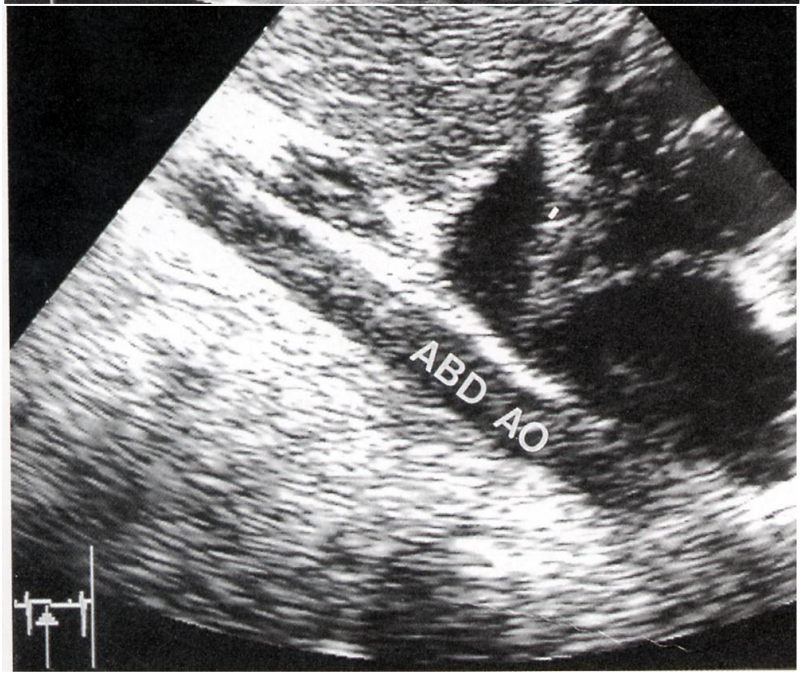


Tetralogy of Fallot

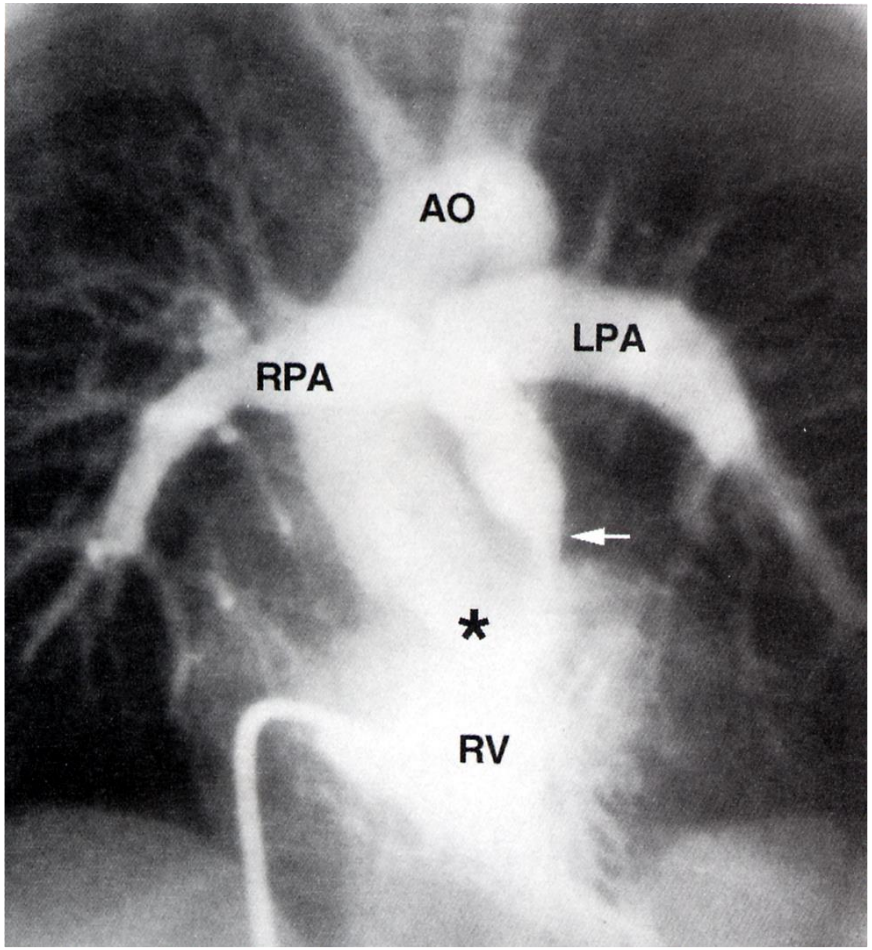
Echocardiography

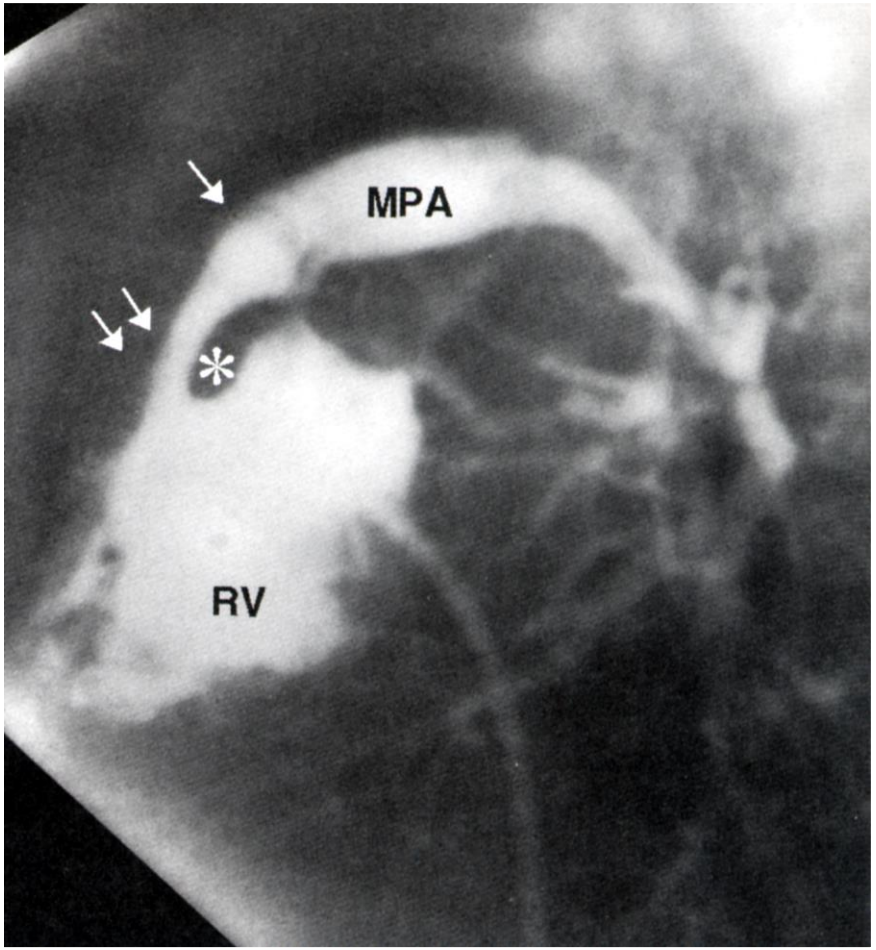






Cardiac catheterization





Medical Management

Outpatient management

- **Aimed at relieving hypoxemia and preventing hypoxic spell (maintaining these infants in a well-hydrated state)**
- **Using a beta-blocking agent such as propranolol to palliate the child is not reliable and should rarely be used**
- **Every effort should be made to protect affected infants from viral infections**
- **Management of hypoxic spell;**
  - **Elimination of predisposing conditions**
    - **dehydration, anemia, increased cathecholamine levels, decreased SVR**
  - **Oxygen**
  - **Sedation**
  - **Barbiturate**
  - **Transfusion of blood or crystalloid**
  - **α-agonist to increase SVR**

Balloon pulmonary valvuloplasty

- **Reduce the need for a transannular patch ?**
- **Disadvantage; excessive pulmonary blood flow, PR, catheter related Cx**

Indications for and Timing of Surgery

- **Symptoms**
  - **1) prostaglandin dependent neonate**
  - **2) worsening cyanosis (O_2 saturation less than 75% to 80%)**
  - **3) cyanotic spell**

- **Elective repair**
  - **4-6 weeks of age (Children's hospital Boston)**

Ductal tissue will declare itself as a possible stenosis at the origin of the LPA)
The incidence of transannular patching between primary repair and two stage approach was very similar

Contraindications to early primary repair

1) anomalous coronary artery
2) multiple muscular VSDs
3) discontinuous central pulmonary arteries

Advantages of early primary repair

- **Normal growth and development of organs (esp. Brain)**
- **Minimization of abnormal lung development**
- **Less need for extensive right ventricular muscle excision**
- **Minimization of lifetime decrease of RV compliance**
- **Better late LV function**
- **Decreased incidence of late dysrhythmias**
- **No PA distortion or scarring around PA and PVOD caused by a shunt**
- **Additional important economic and psychological advantages**

The modified Blalock shunt usually results in scarring and distortion at the level of the distal anastomosis which is an important consideration in the child who requires a highly compliant pulmonary arterial tree to ameliorate the deleterious effects of the pulmonary regurgitation which accompanies a transannular patch.

Timing of Surgery

- **A trend toward earlier intervention with elective repair within the first 3-6 months of life**

Controversies in "Early Primary Repair"

Pro

1) Low operative mortality of early primary repair
2) Avoidance of harmful effect of shunt operation and late repair

Con

1) Higher mortality and morbidity with early primary repair
2) Neonatal myocardium may be less capable of handling of RV volume load (from VSD closure, pulmonary and possible tricuspid regurgitation)
3) Well-executed shunting for TOF in early infancy carries a very low risk of pulmonary artery distortion or early or interval mortality
4) Very low mortality

Symptomatic neonates or young infants with TOF

Shunt (Staged approach) ?

Primary repair ?

Potential disadvantage of staged approach

Long-lasting pressure overload of RV and persistent cyanosis

→ cardiomyocytic degeneration and interstitial fibrosis

→ myocardial dysfunction and ventricular arrhythmia

Potential disadvantage of early primary repair

Frequent need of transannular patch

Adverse effects of early bypass surgery on the neonatal brain

Often complicated and lengthy postoperative recovery in small infants

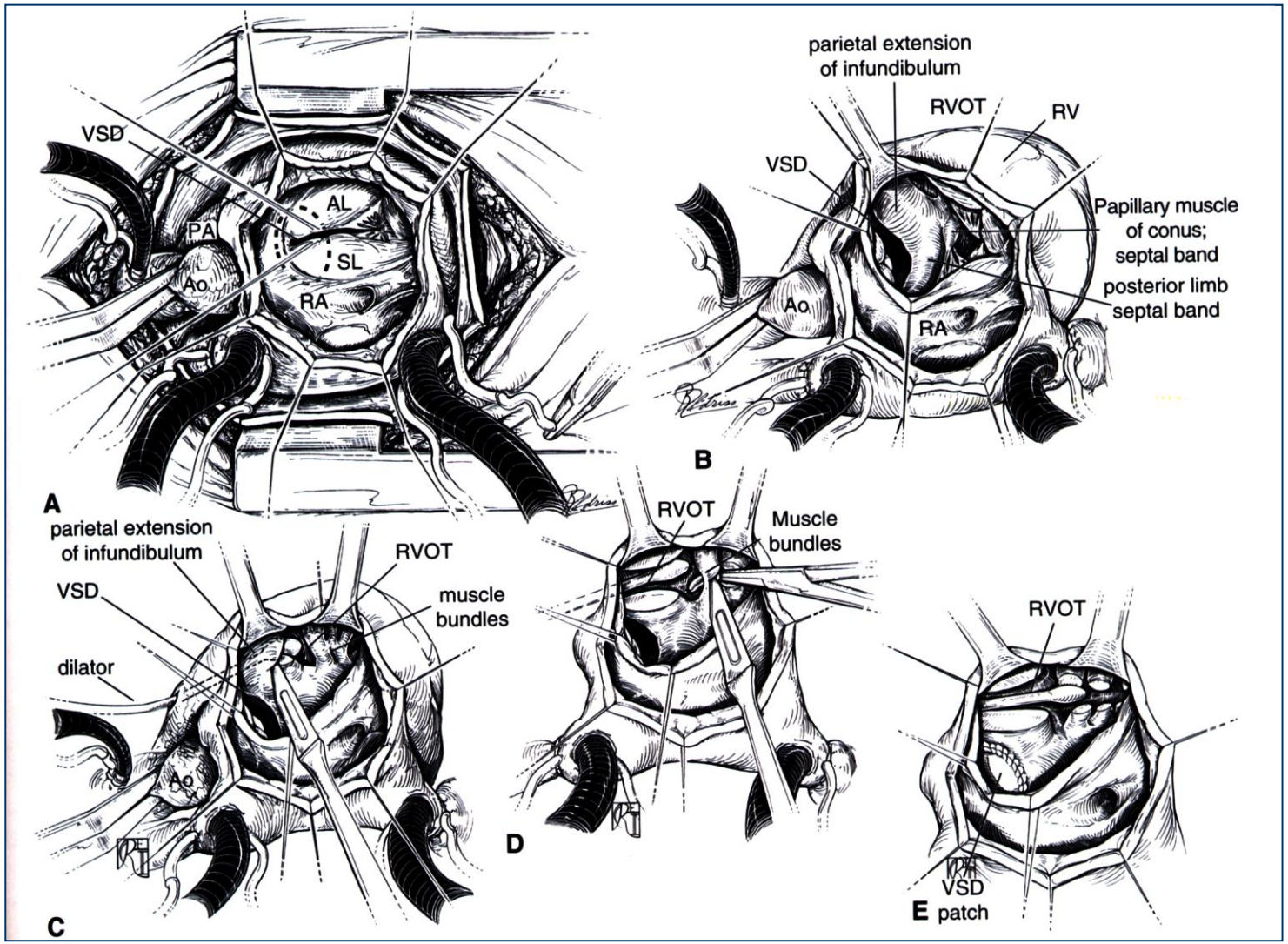
Surgical Management

Goal of surgical therapy

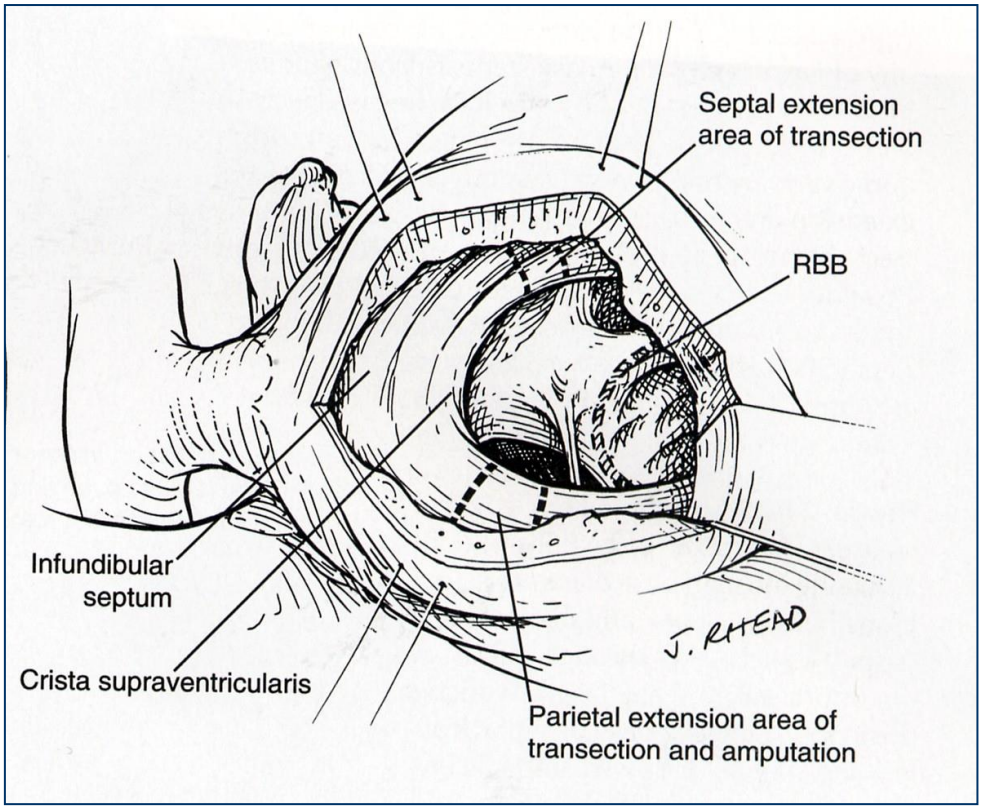
- **Close intracardiac shunts**
- **Provide relatively unobstructed pulmonary blood flow**
- **Maintain normal function of the right ventricle**
- **Maintain normal function of the P - and T - valves**
- **Maintain NSR with minimal morbidity and mortality**

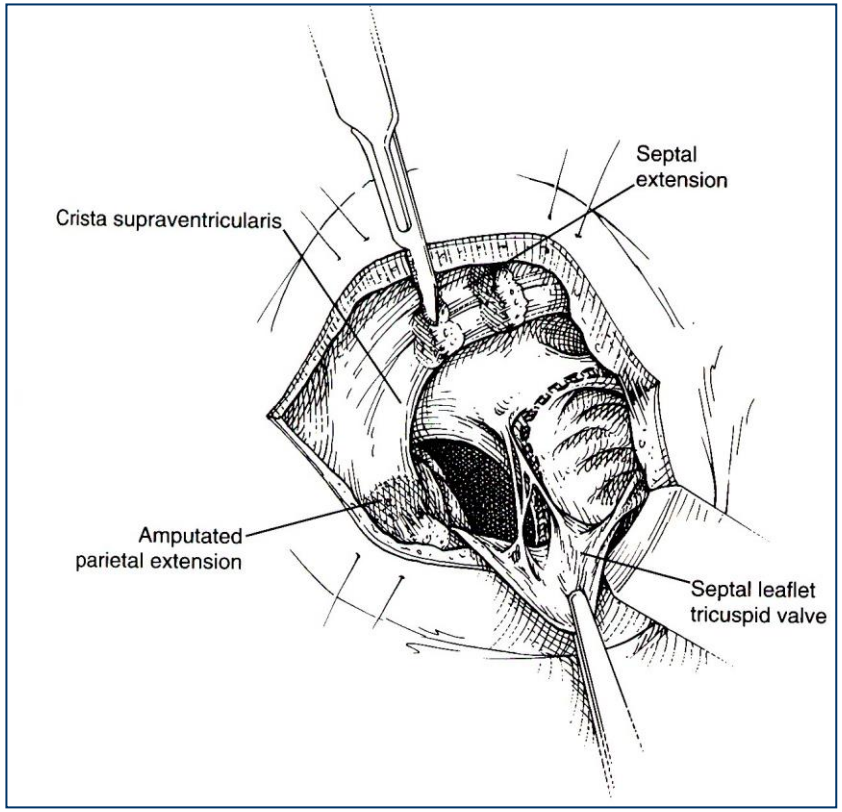
Surgical Management

Infundibular muscle resection through tricuspid valve



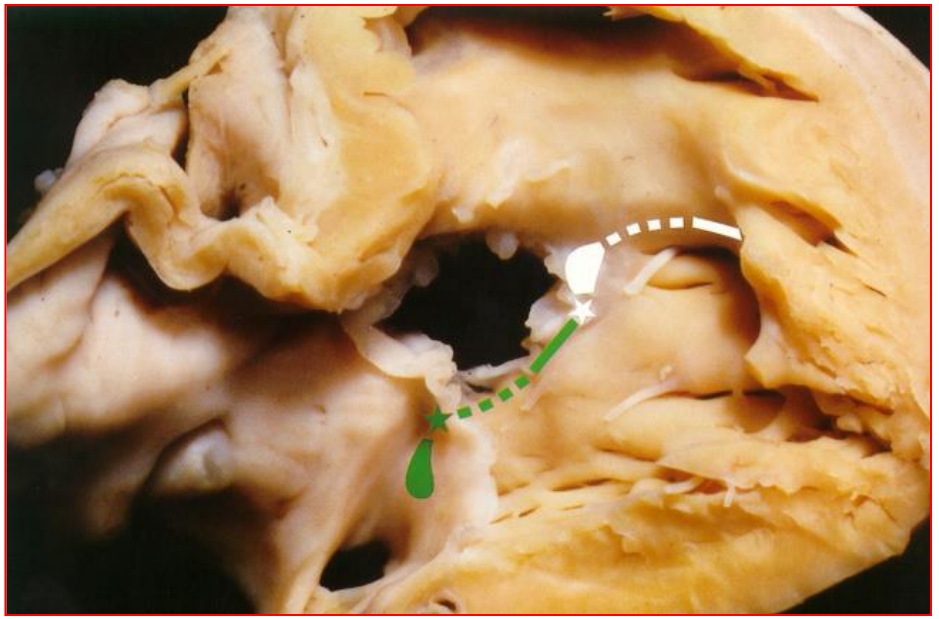
Infundibular muscle resection through ventriculotomy

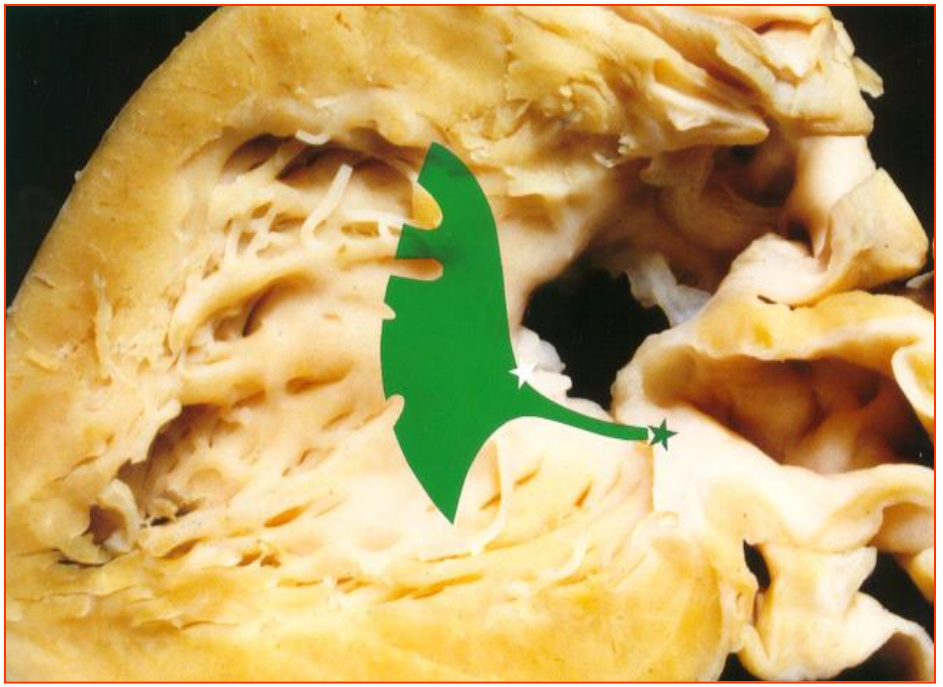




VSD closure

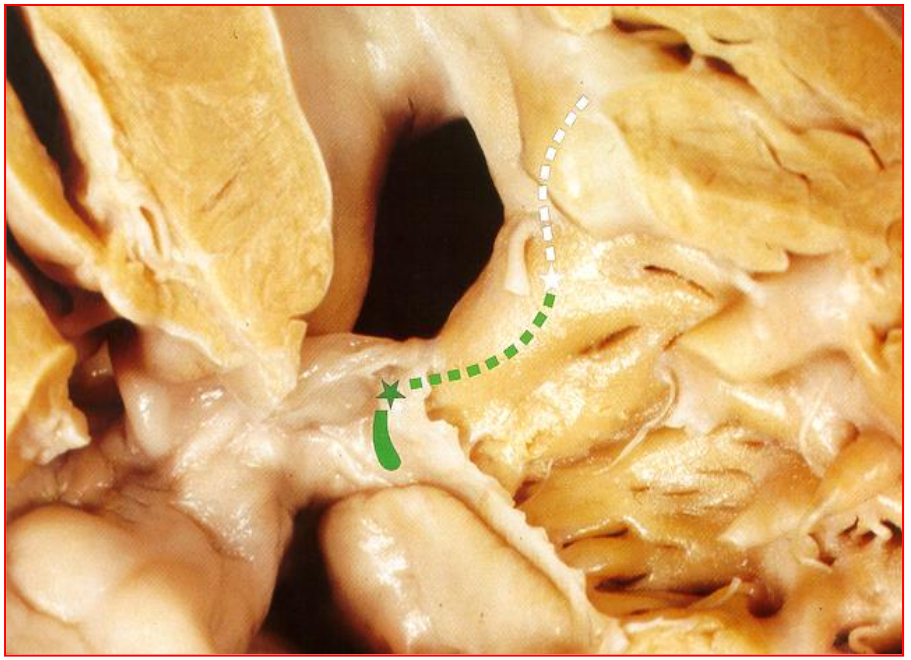
Conduction system
PM VSD

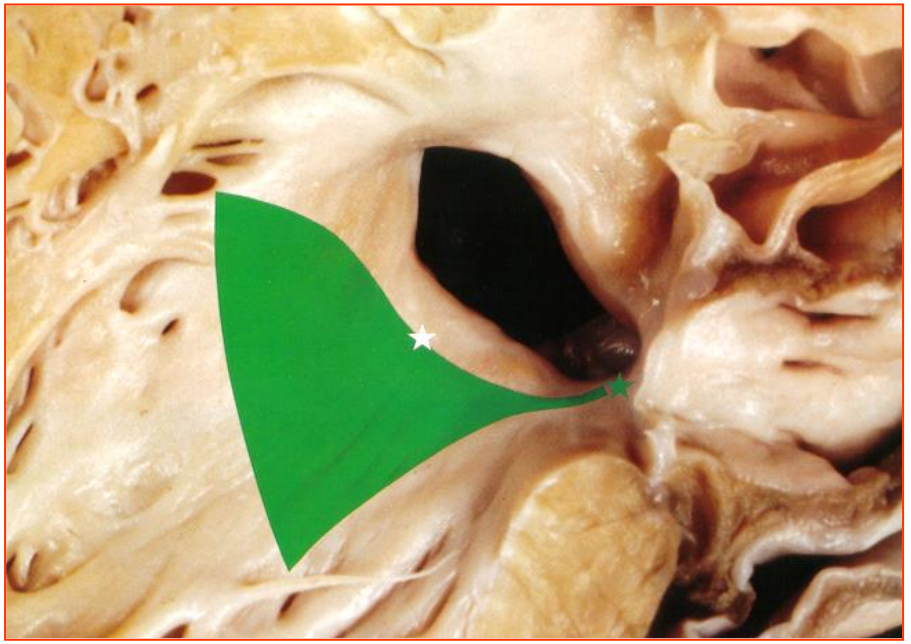




VSD closure

Conduction system
Perimembranous
infundibular VSD



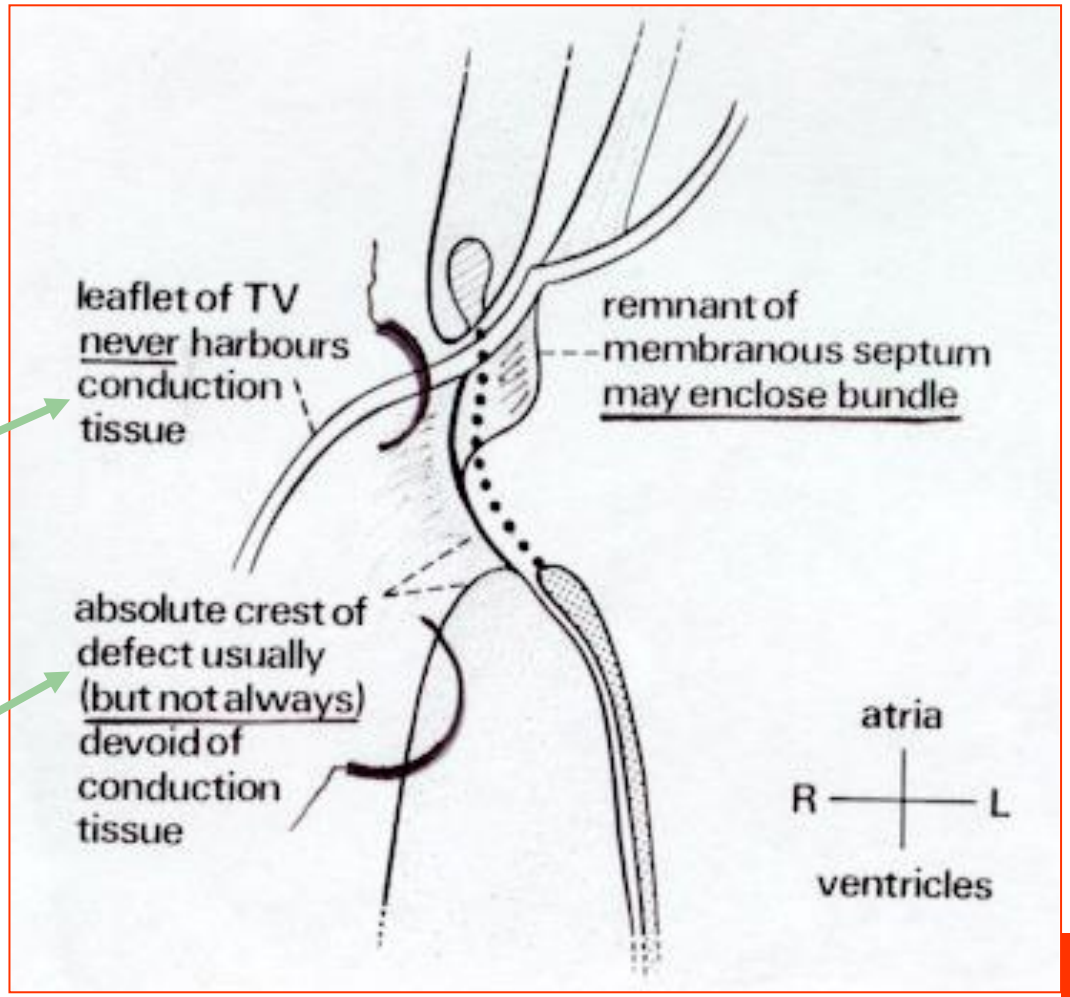


Surgical Techniques to Avoid AV conduction tissue injury in VSD repair

Perimembranous or not ?
(muscle bar beneath TV or not ?)

Placing sutures at the base of TV to protect penetrating bundle

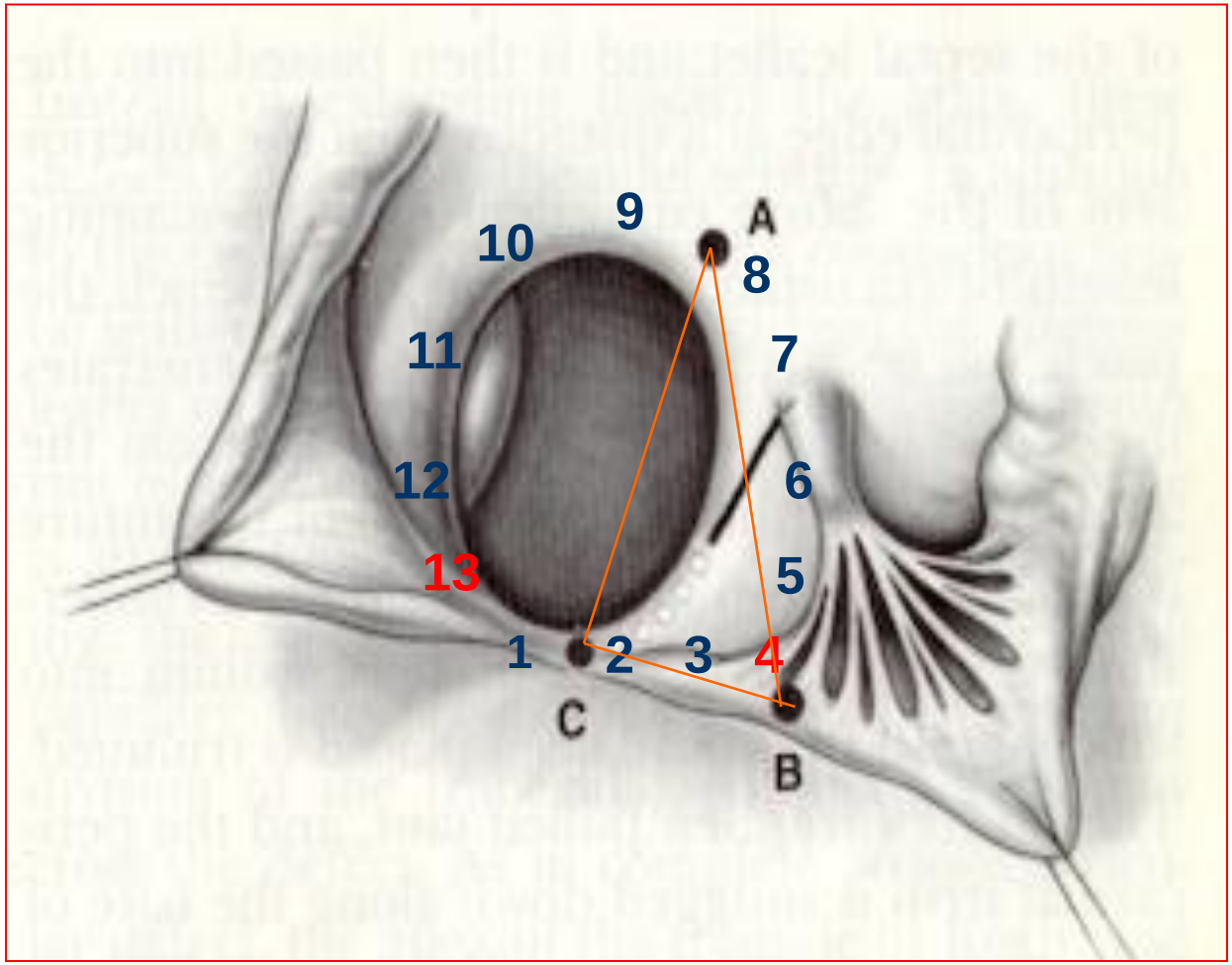
Placing sutures at the right-sided septal wall to protect branching bundle



Surgical Management

VSD closure

PM VSD

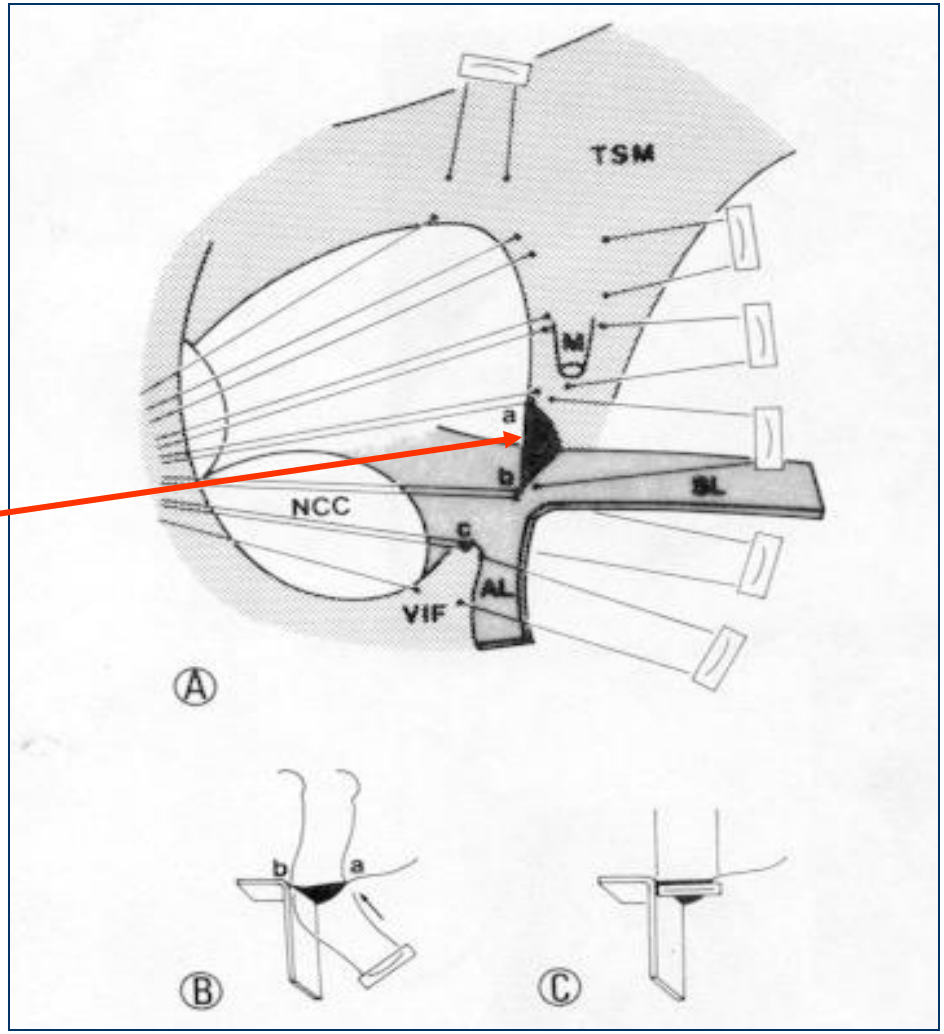


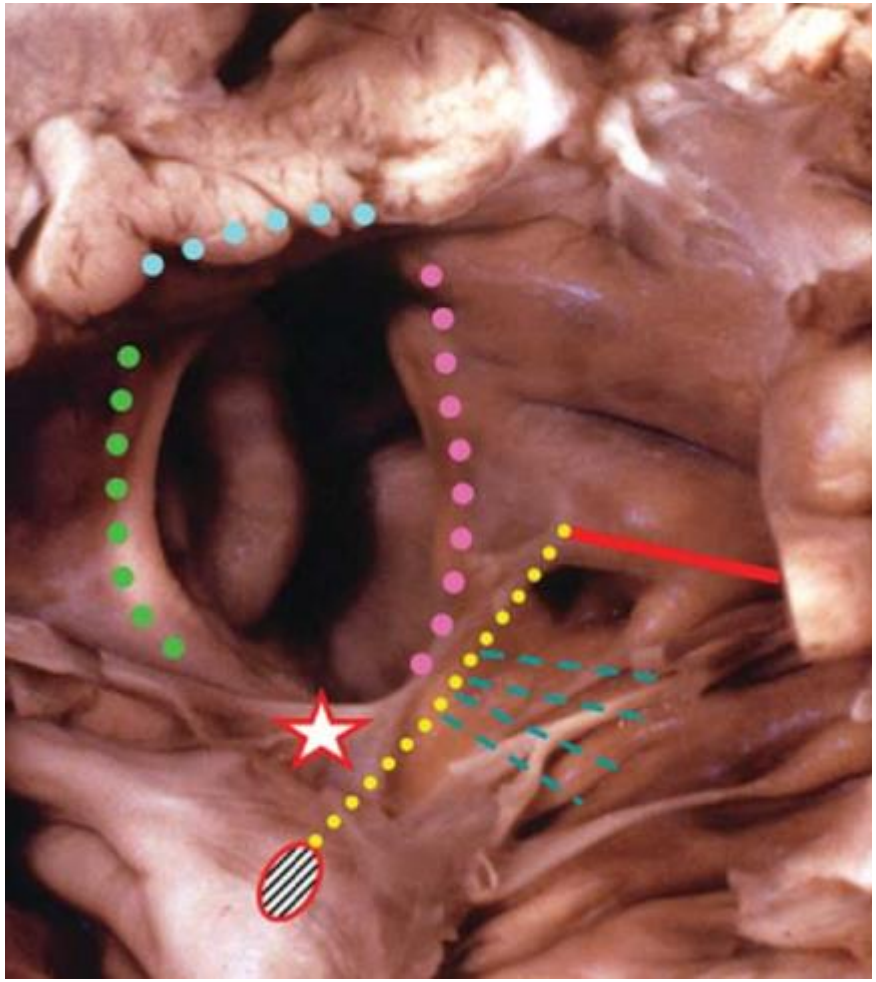
Surgical Management

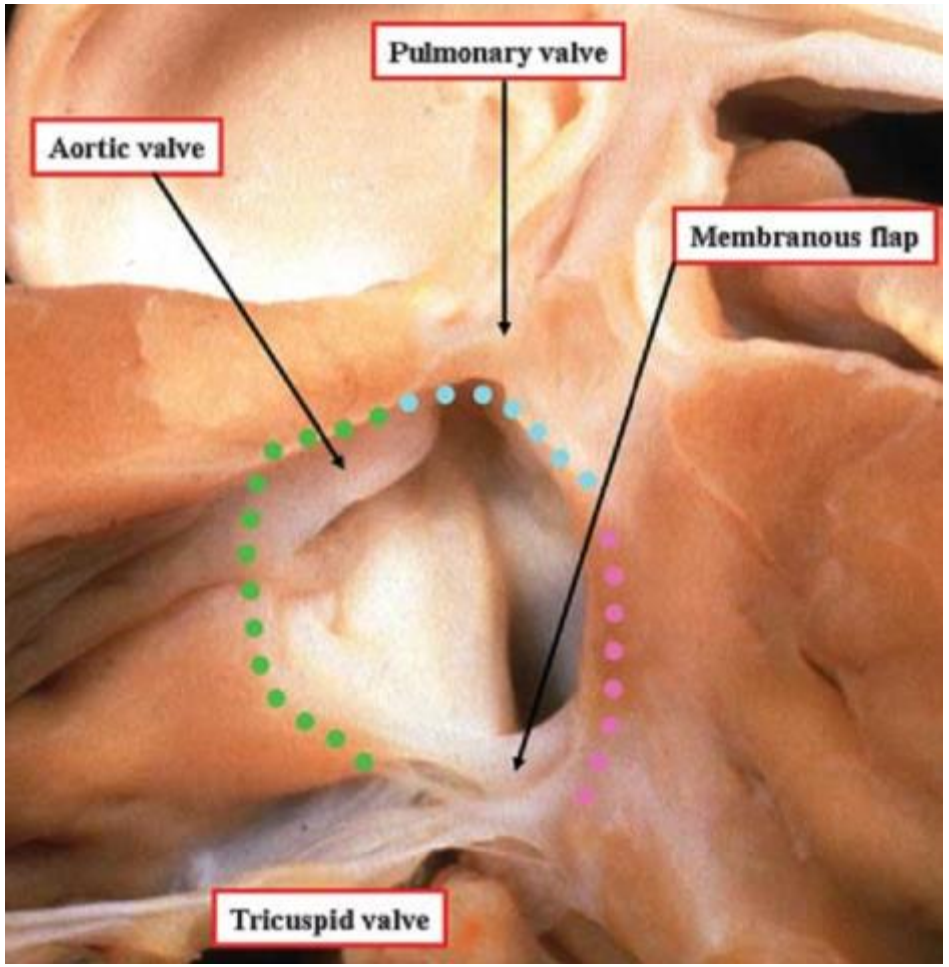
VSD closure

Conduction tissue is composed of specialized muscle cells

Membranous flap







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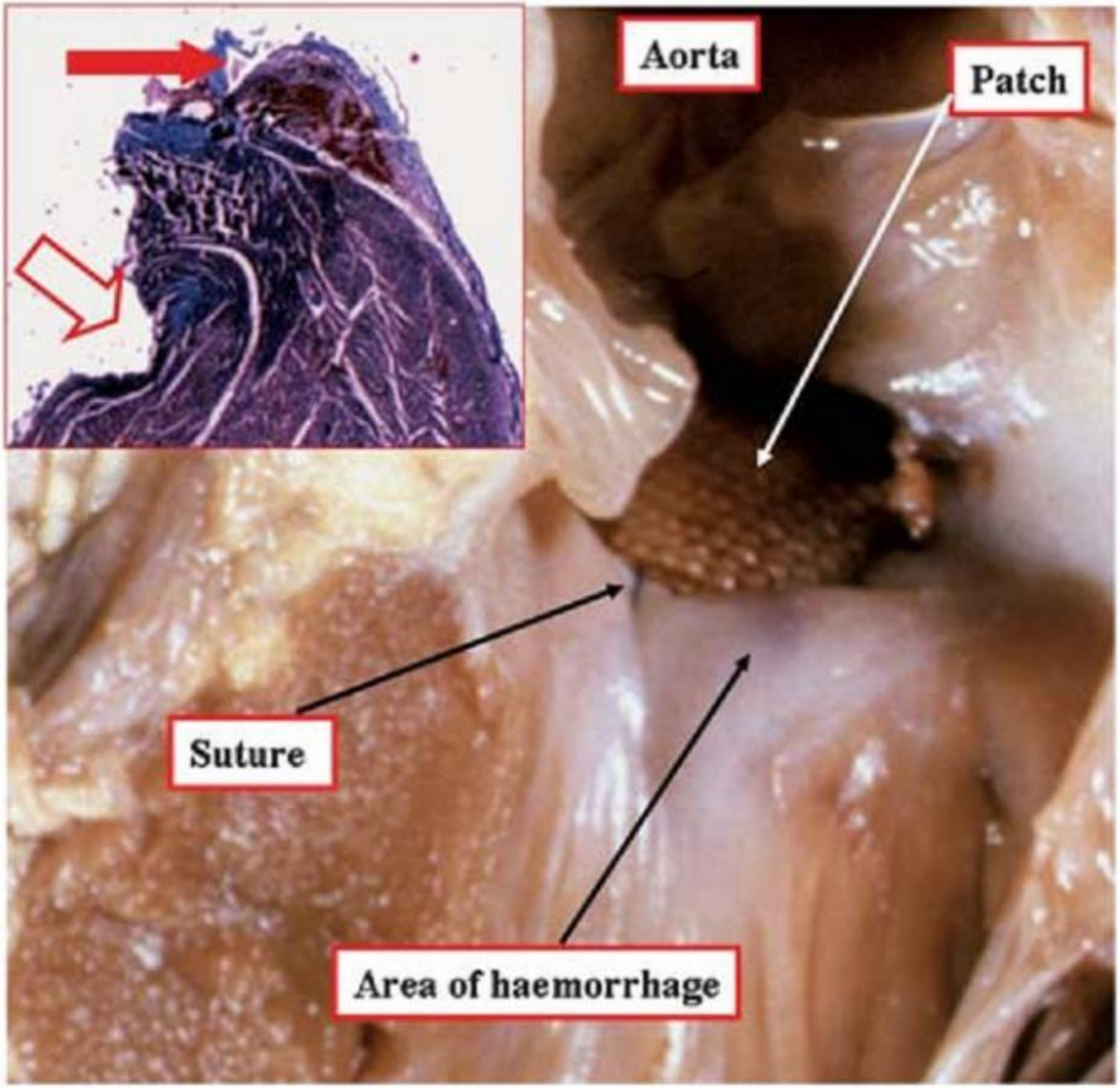
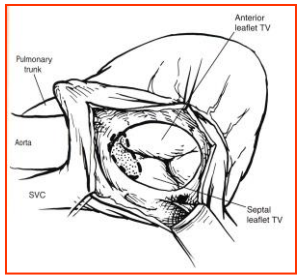
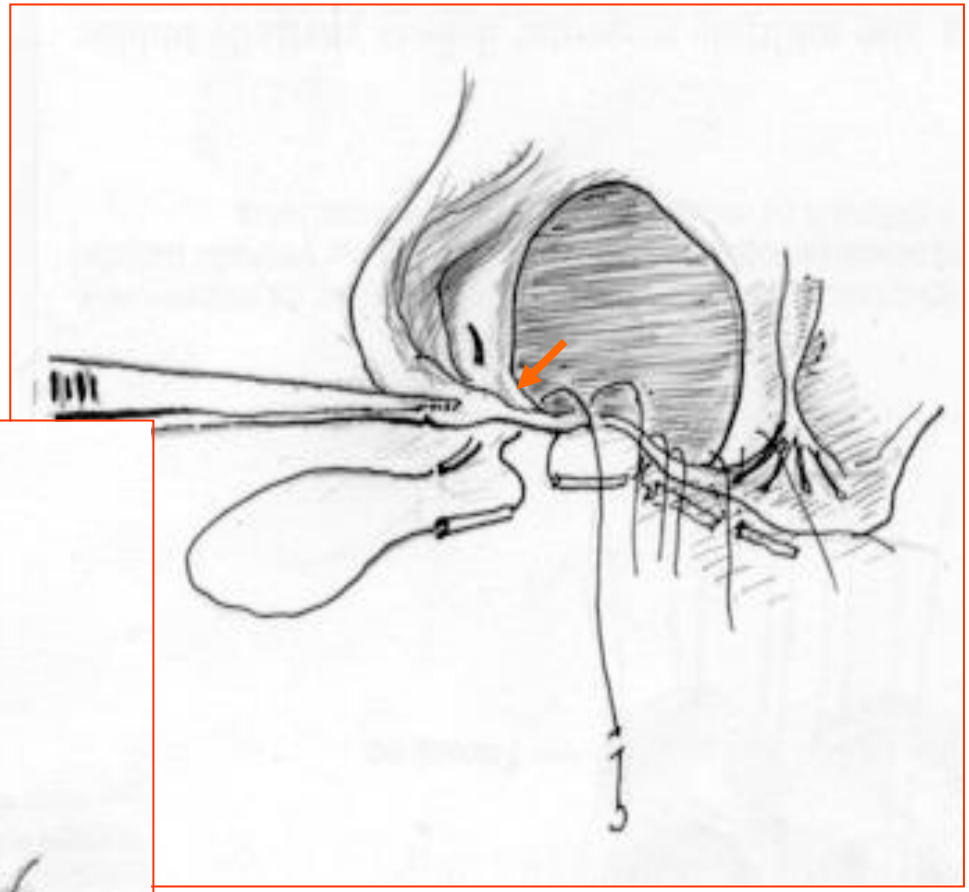
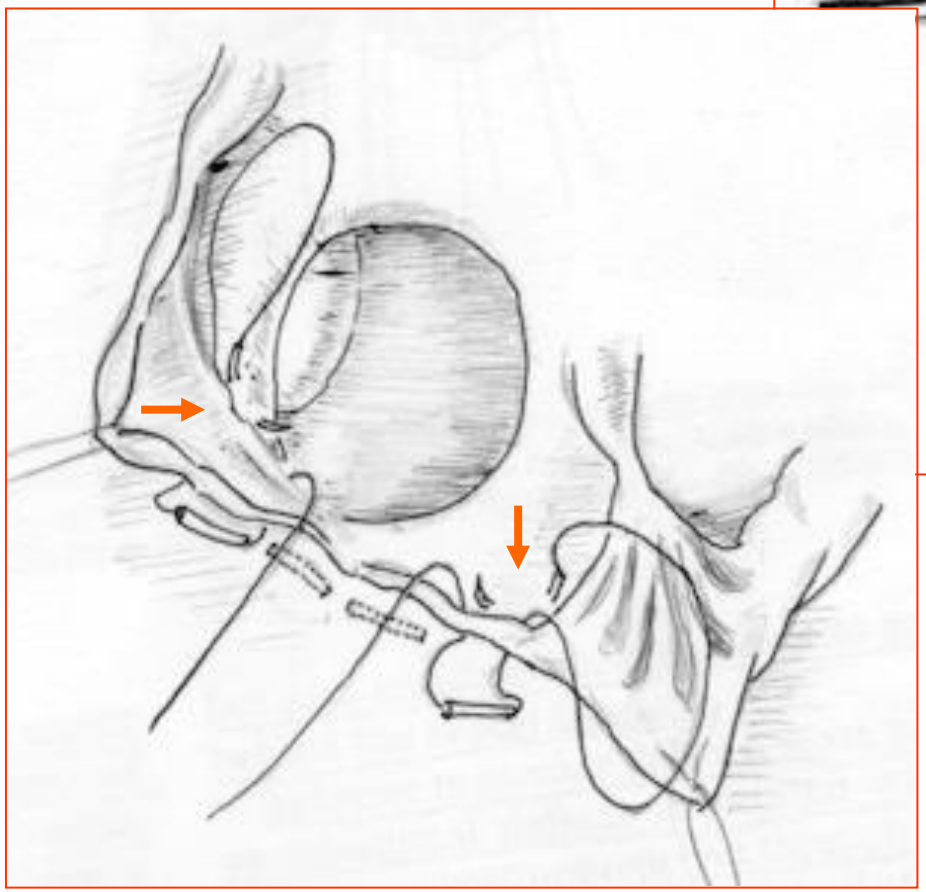

Aorta
Patch
Suture
Area of haemorrhage

Surgical Management

VSD closure



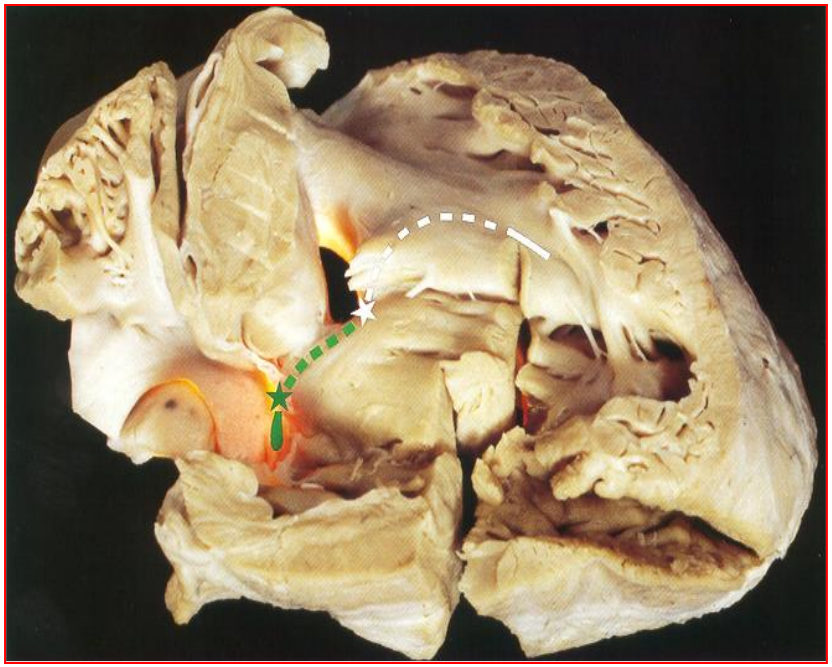


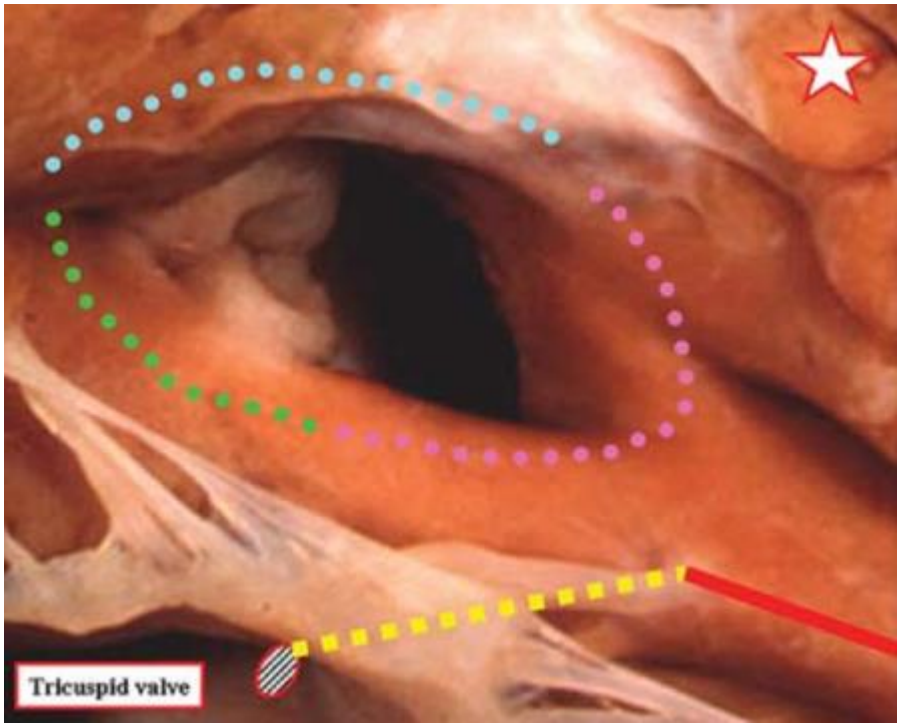


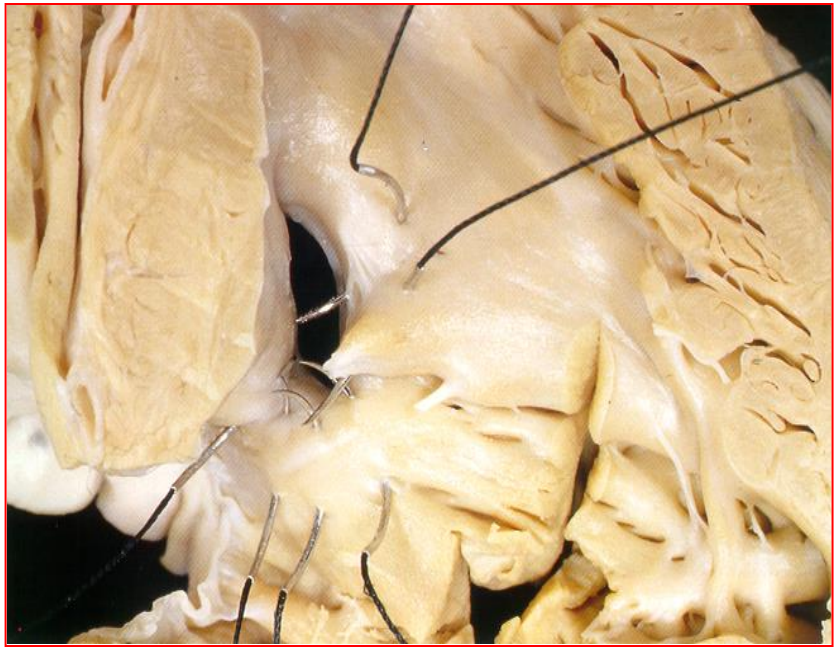
Surgical Management

VSD closure

Conduction system
Muscle bar beneath T - valve
Muscular outlet VSD

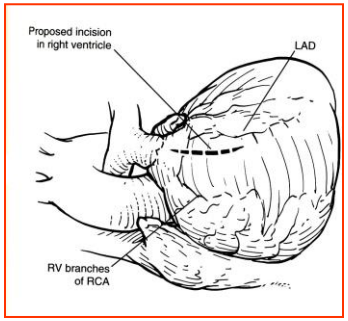


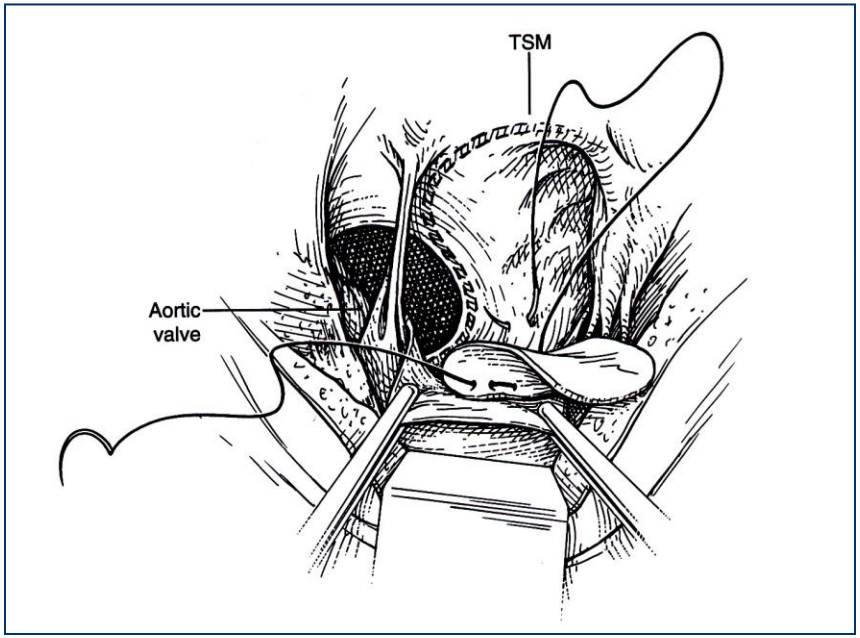


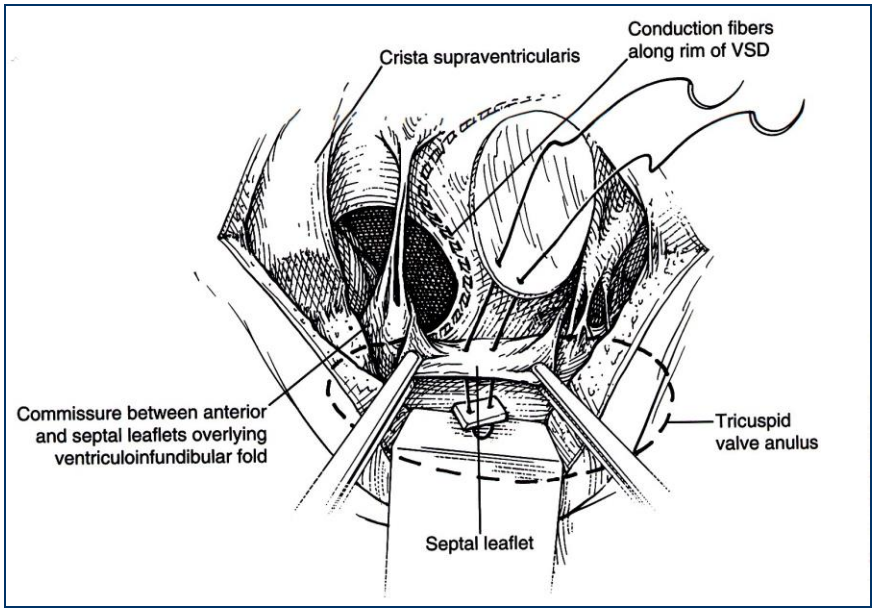


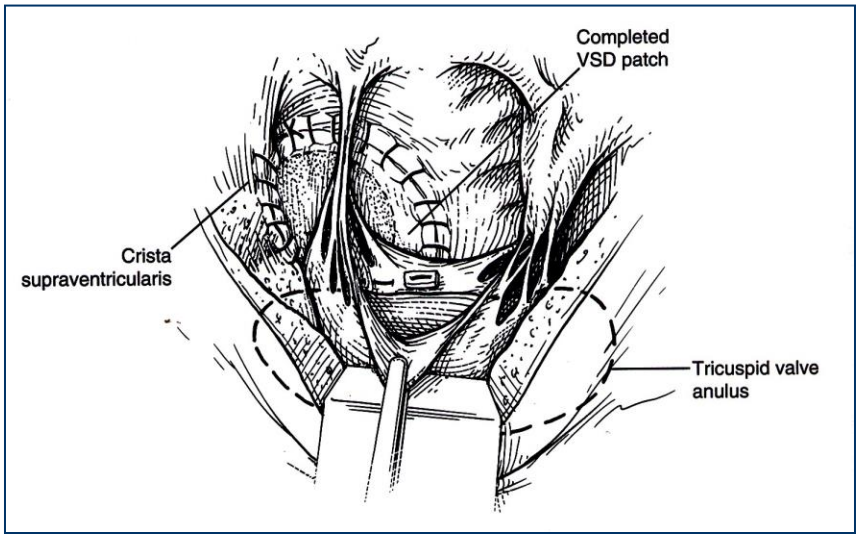
Surgical Management

VSD closure through RV

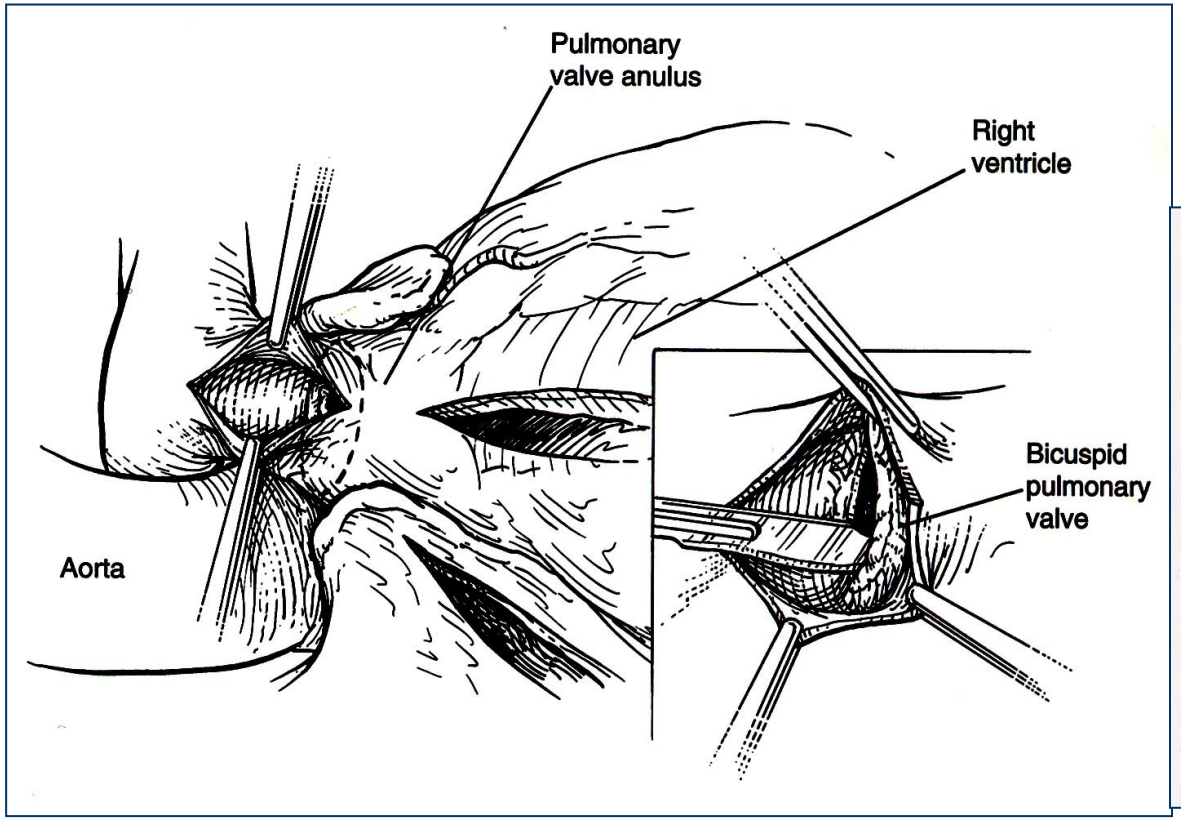


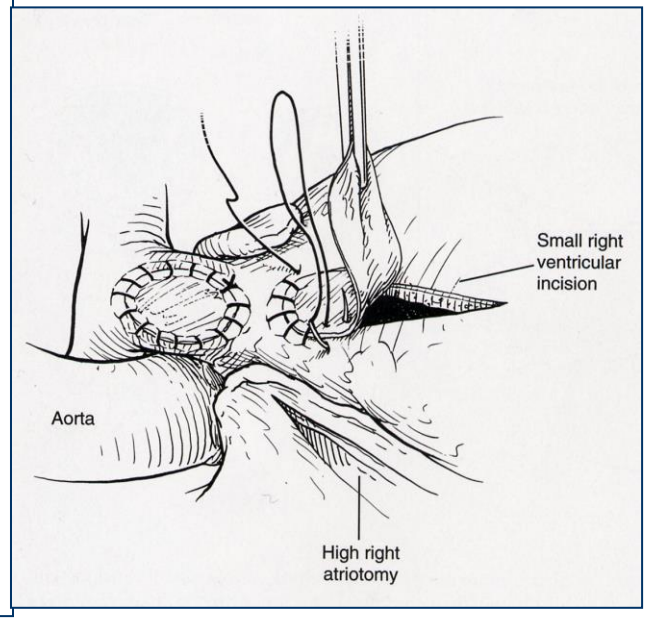




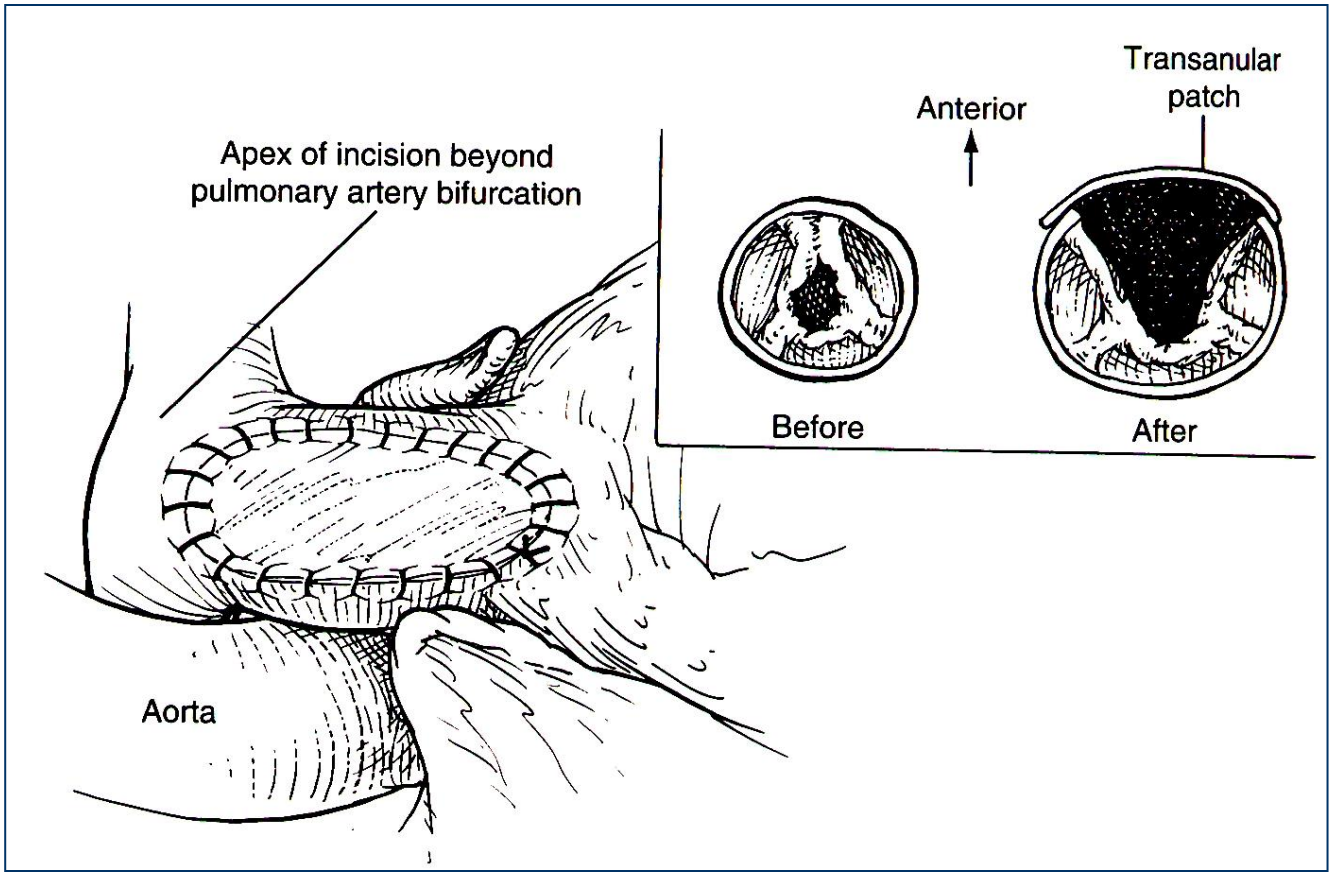


Pulmonary valvotomy





Transannular patch



Transannular patch

Criteria for preservation of pulmonary valve ?

z-valve of pulmonary annulus > -3
diameter of pulmonary annulus (mm) > 0.8mm/kg of body weight

Intraperative relaxed heart, PV annulus > (1-2mm or 0mm ?) + mean PV annulus

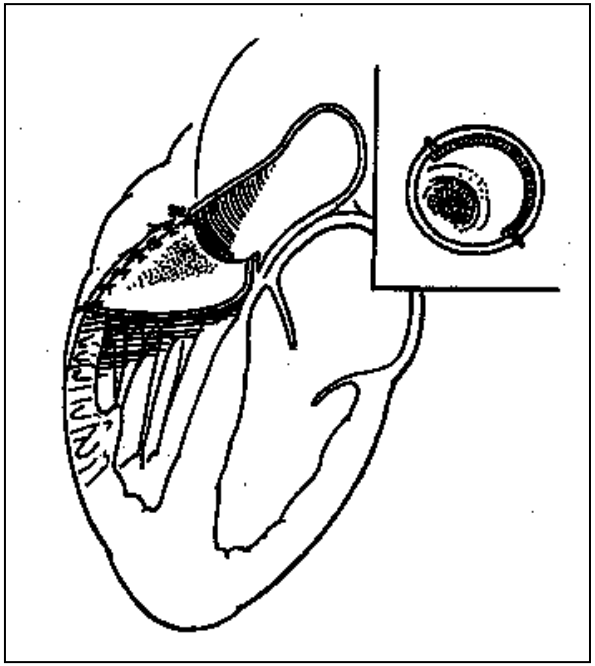
Postrepair RV/LV > 0.7

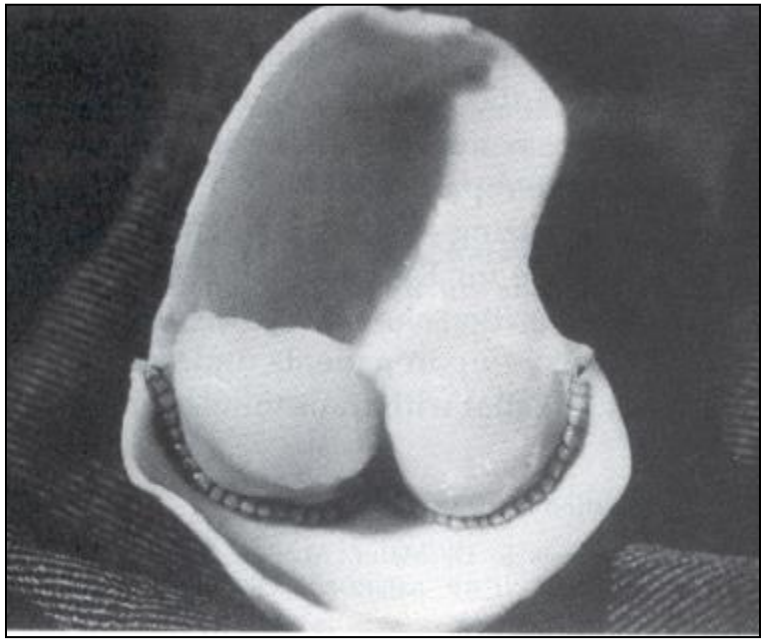
If TAP has not been placed, TAP should be considered.
If TAP has been place, other causes must be considered.

- branch PA stenosis
- hypoplasia of peripheral PAs
- residual VSD
- residual infundibular obstruction

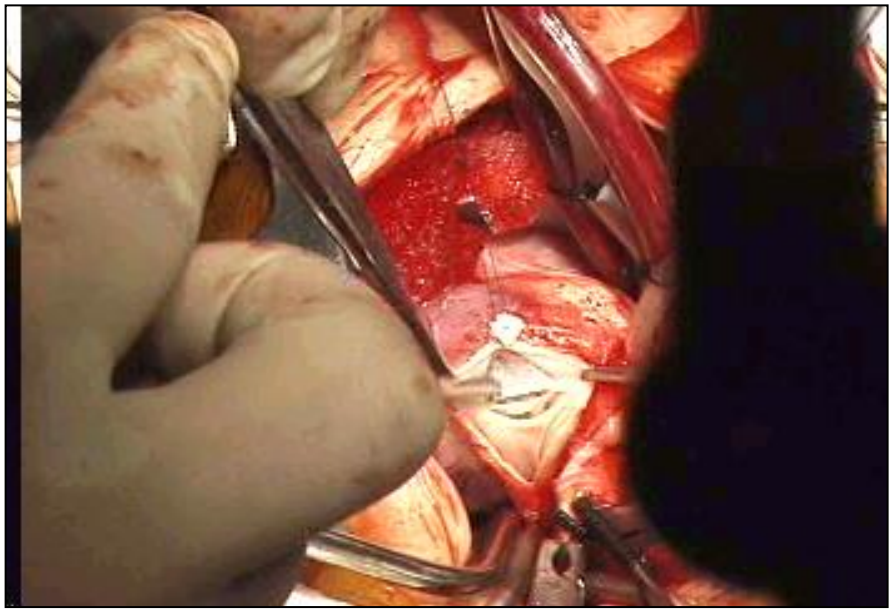
- Often elevation of RV pressure results from dynamic RVOTO.
- Ultra-short acting β-blocker (esmolol) can help in intraoperative differentiation.

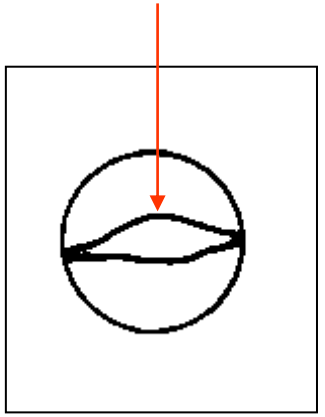
Monocusp Ventricular Outflow Patch



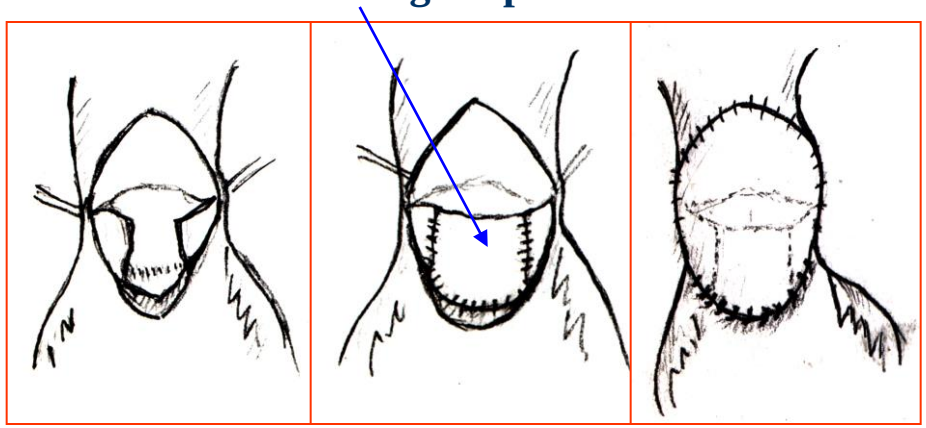


PV Annular Enlargement with Valve Reconstruction

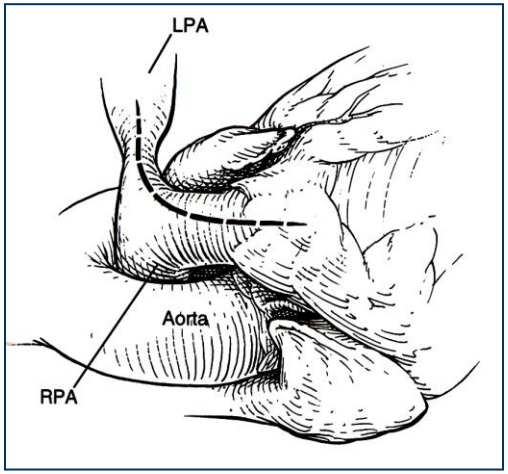


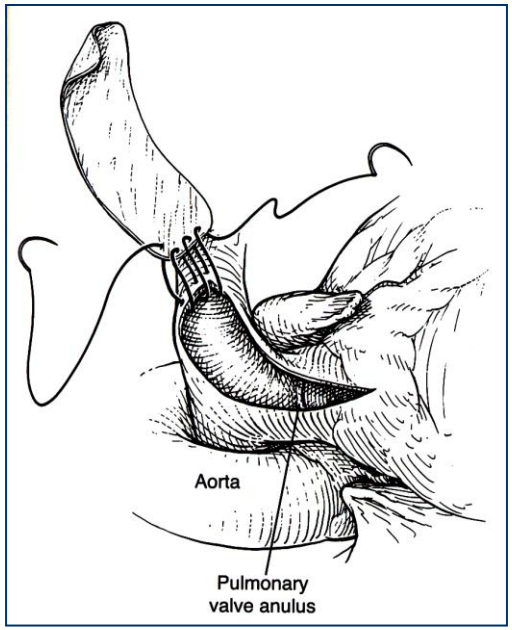


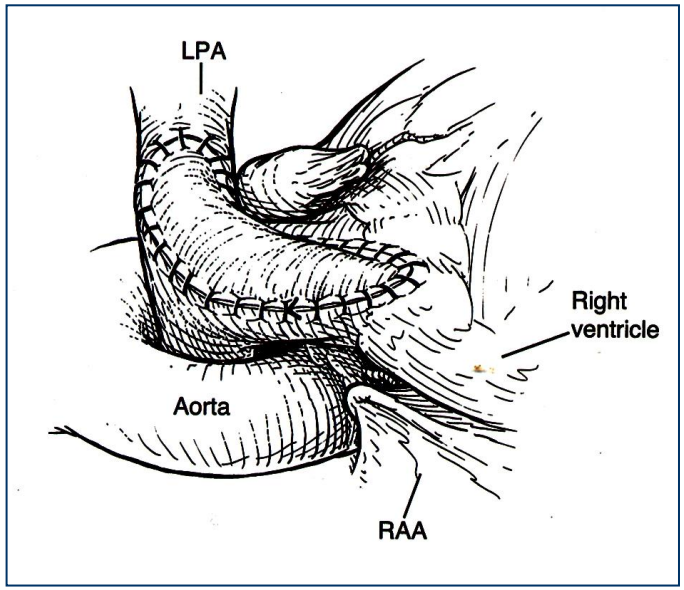
Gore-Tex membrane or autologous pericardium



Pulmonary artery angioplasty





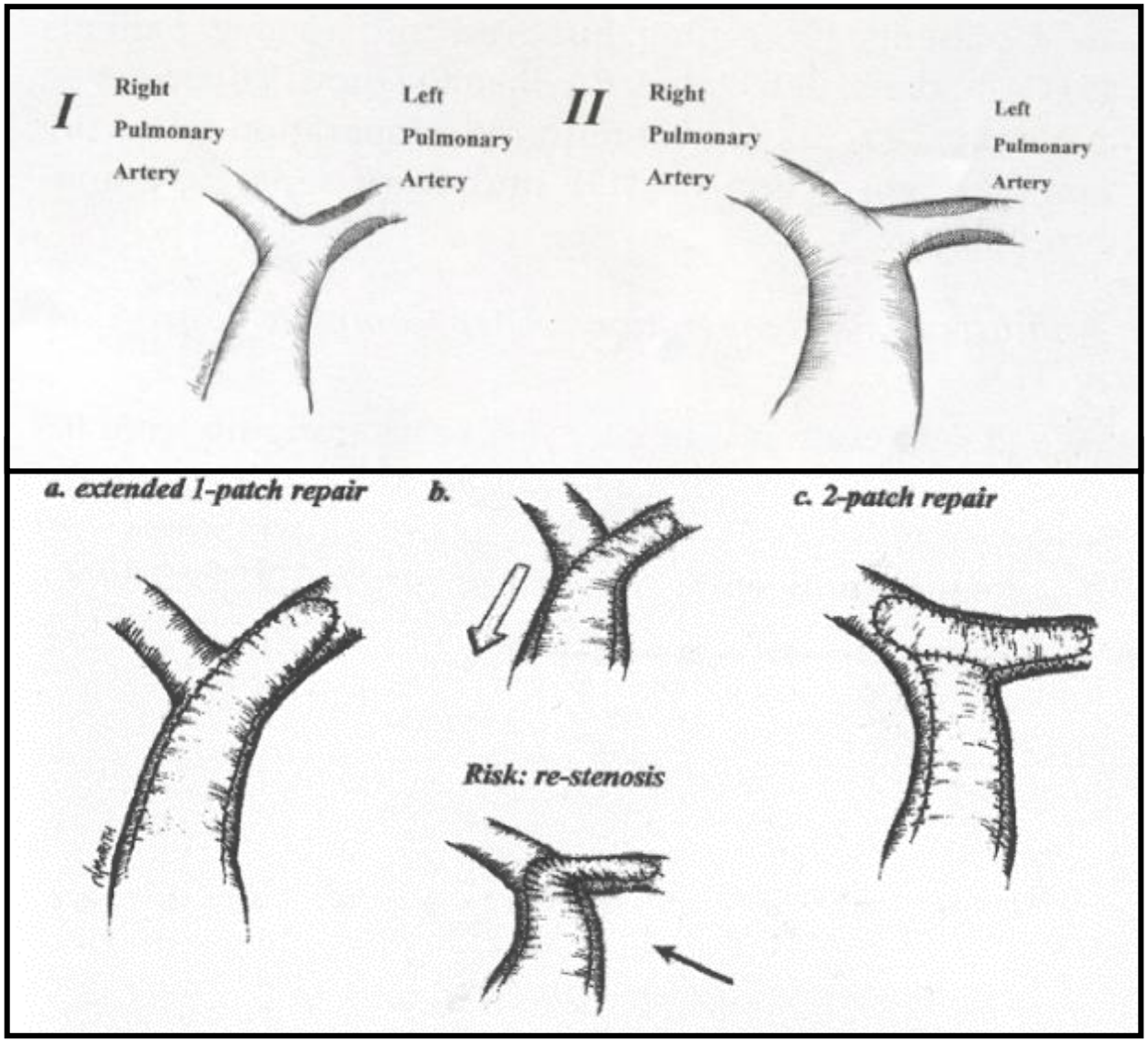


❑ Causes of postoperative LPA stenosis

- **Inadequate enlargement**
- **Aneurysmal dilatation of RVOT patch**
- **Kinking**

Surgical Management

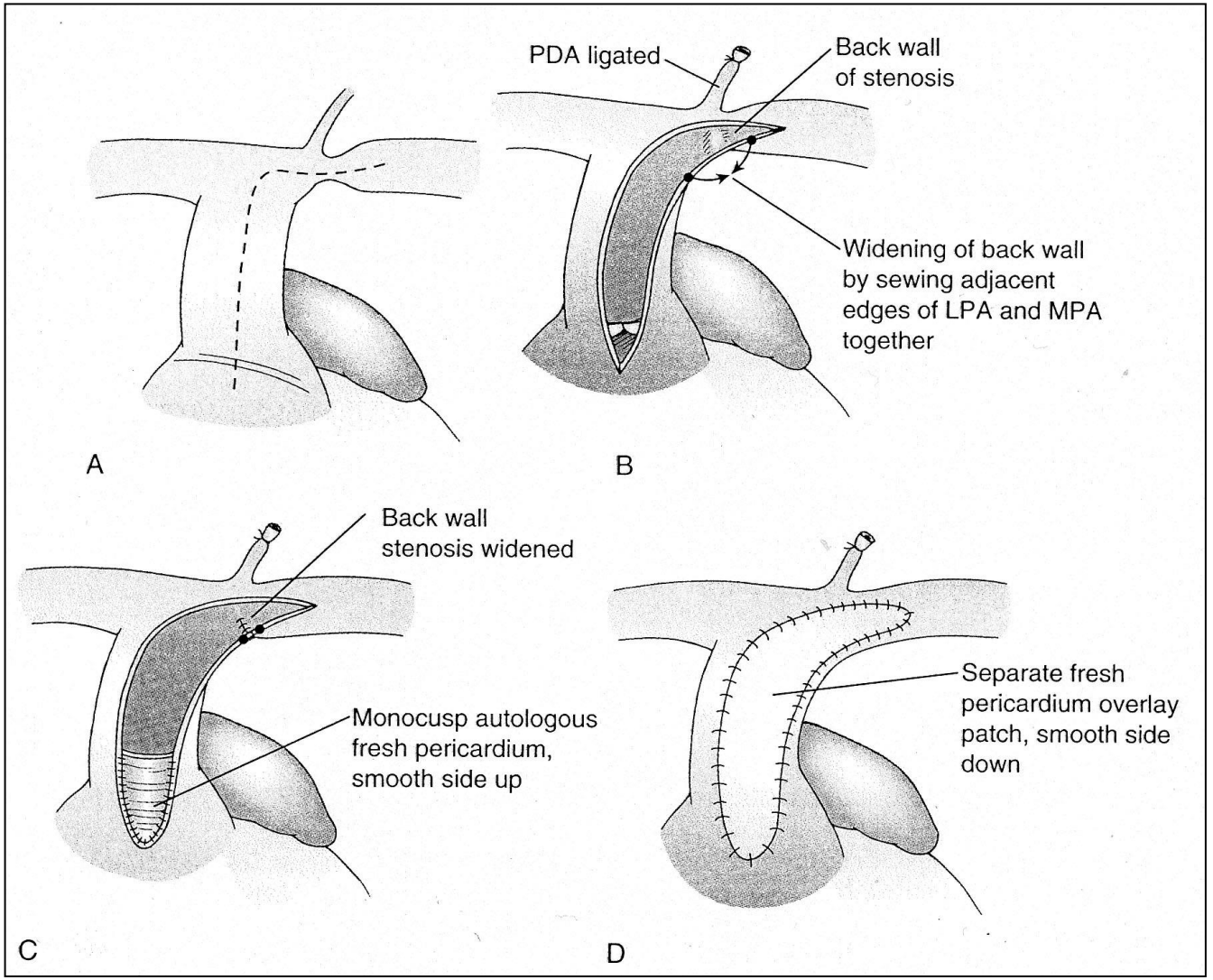
Pulmonary artery angioplasty



Faidutti et al. Ann Thorac Surg 2002;73:96

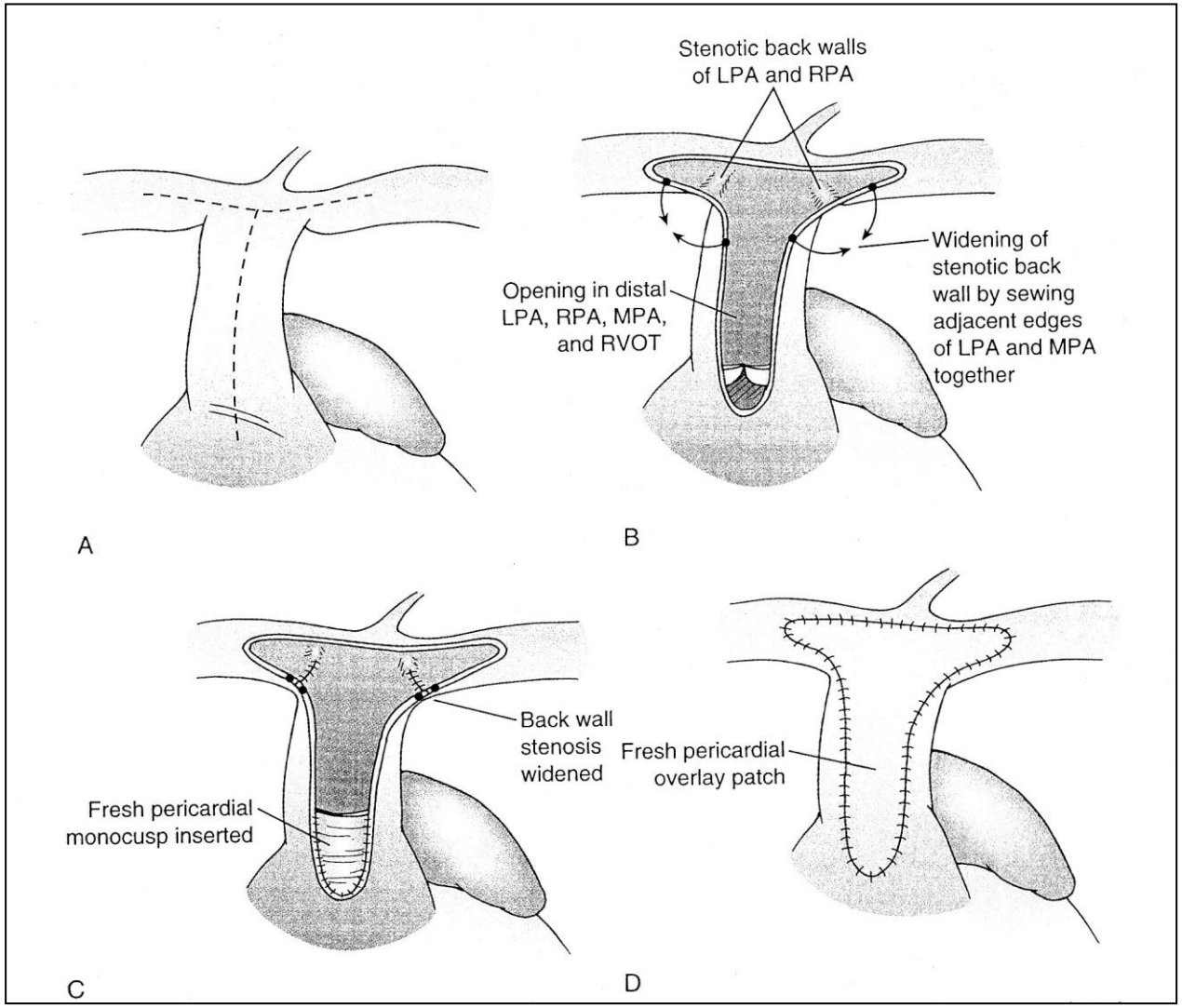
Surgical Management

Pulmonary artery angioplasty



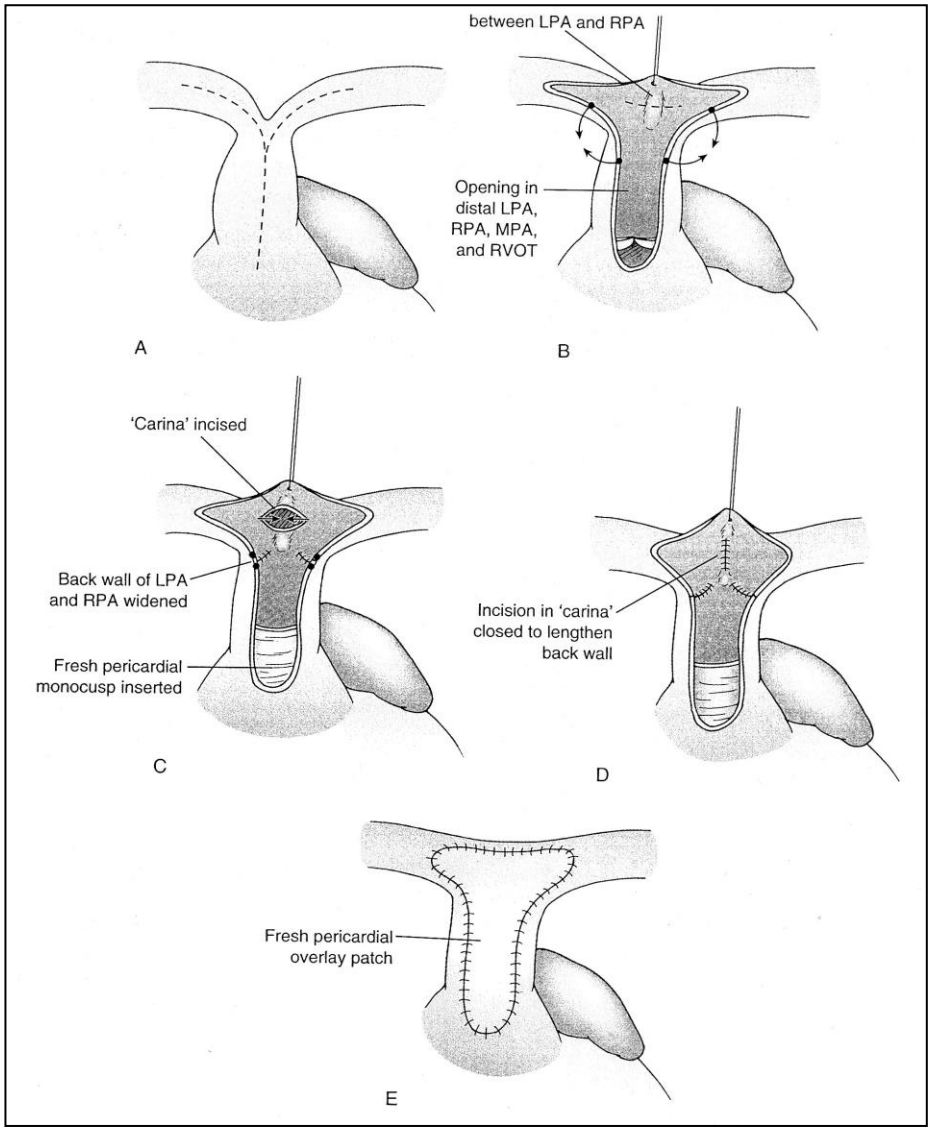
Surgical Management

Pulmonary artery angioplasty



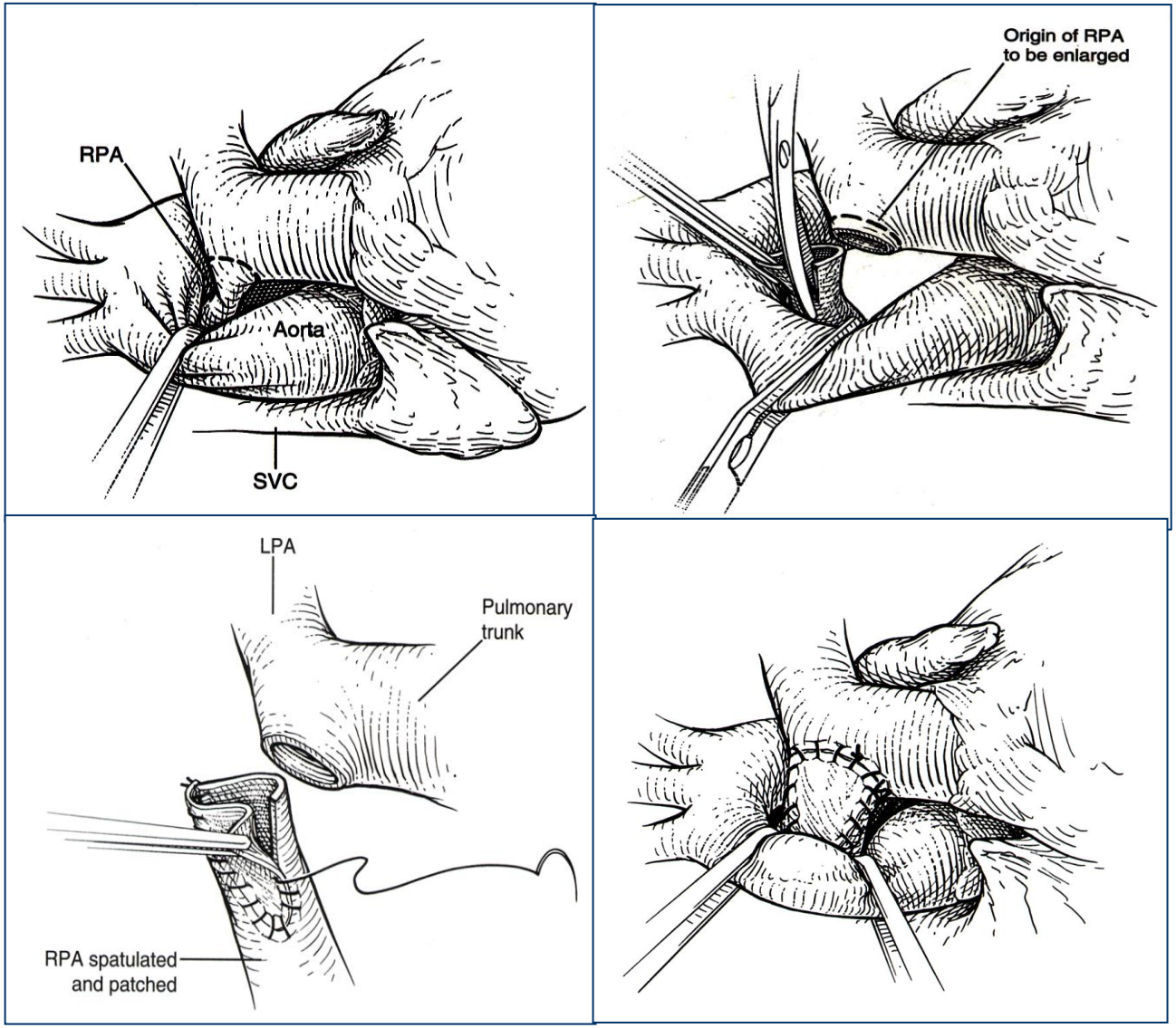
Surgical Management

Pulmonary artery angioplasty

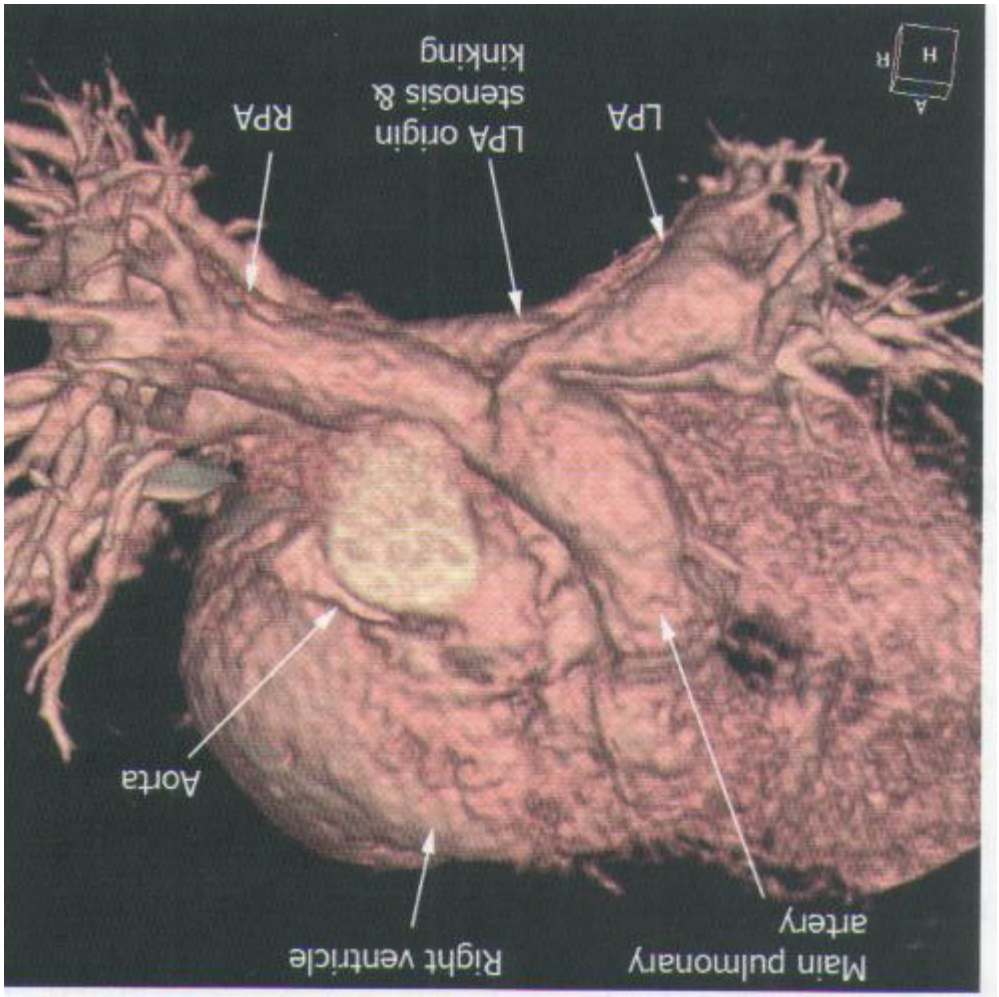


Surgical Management

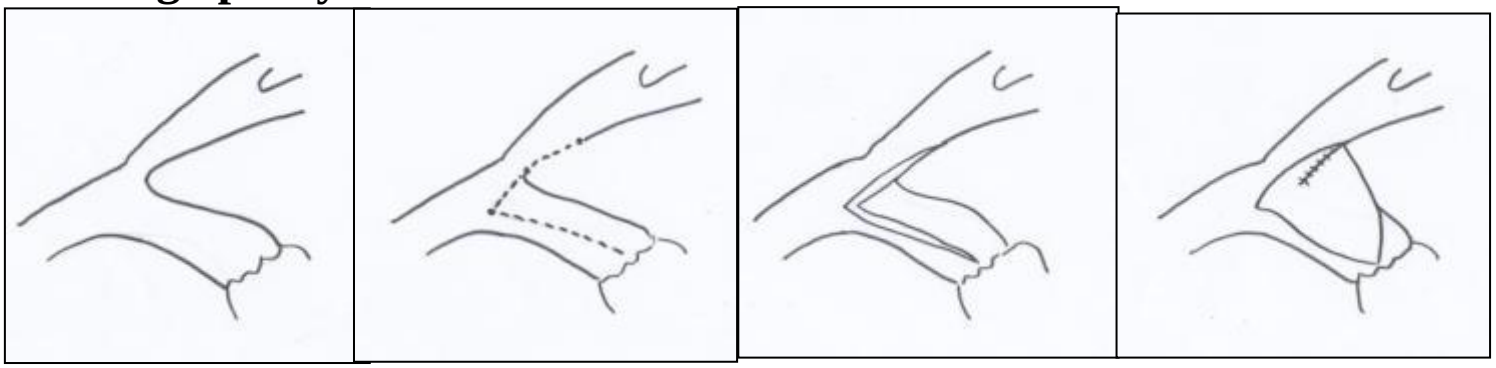
Pulmonary artery angioplasty

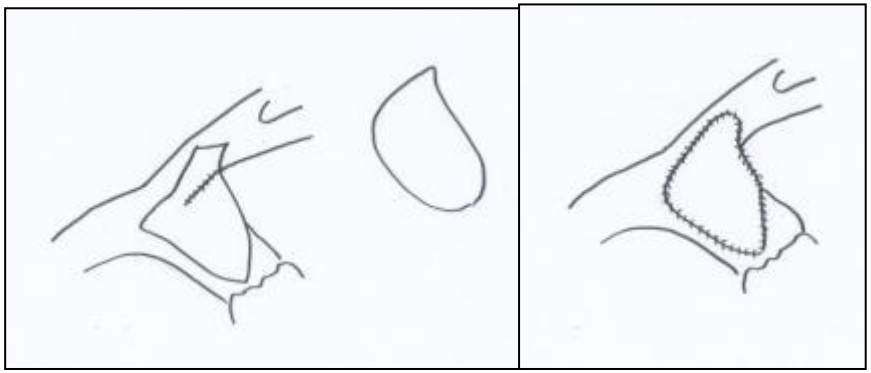


Acute angle of LPA with or without stenosis



LPA angioplasty





LPA & RPA angioplasty

Technical consideration

- **Preservation of moderator band**
  - Central pillar of the RV → preserve or partially divide
- **Avoidance of coronary injury by sutures**
  - Pledgets lying on the endocardial side
- **Management of foramen ovale**
  - High RA pressure is poorly tolerated in neonate or young infant
- **Residual VSD is poorly tolerated**
  - Thin walled and distensible peripheral PAs
  - PR and TR exacerbate RV volume load
  - RV diastolic dysfunction

Special Topics in Surgical Management

1) Hypoplastic pulmonary arteries

McGoon ratio < 1.2
Nakata index < 70

- Uncommon in the patients with TOF/PS
- Hypoplasia is most likely to be a result of underperfusion of the PAs
- Prompt enlargement can be expected when pressure and flow are restored
- In the patient with pulmonary atresia, hypoplasia is more likely due to fixed peripheral arterial stenosis or arbolization anomaly

❑ If after correction of TOF with hypoplastic PAs, intraoperative RV pressure equals or exceed LV pressure, a large perforation should be placed in the VSD patch

2) Use of monocusp valve

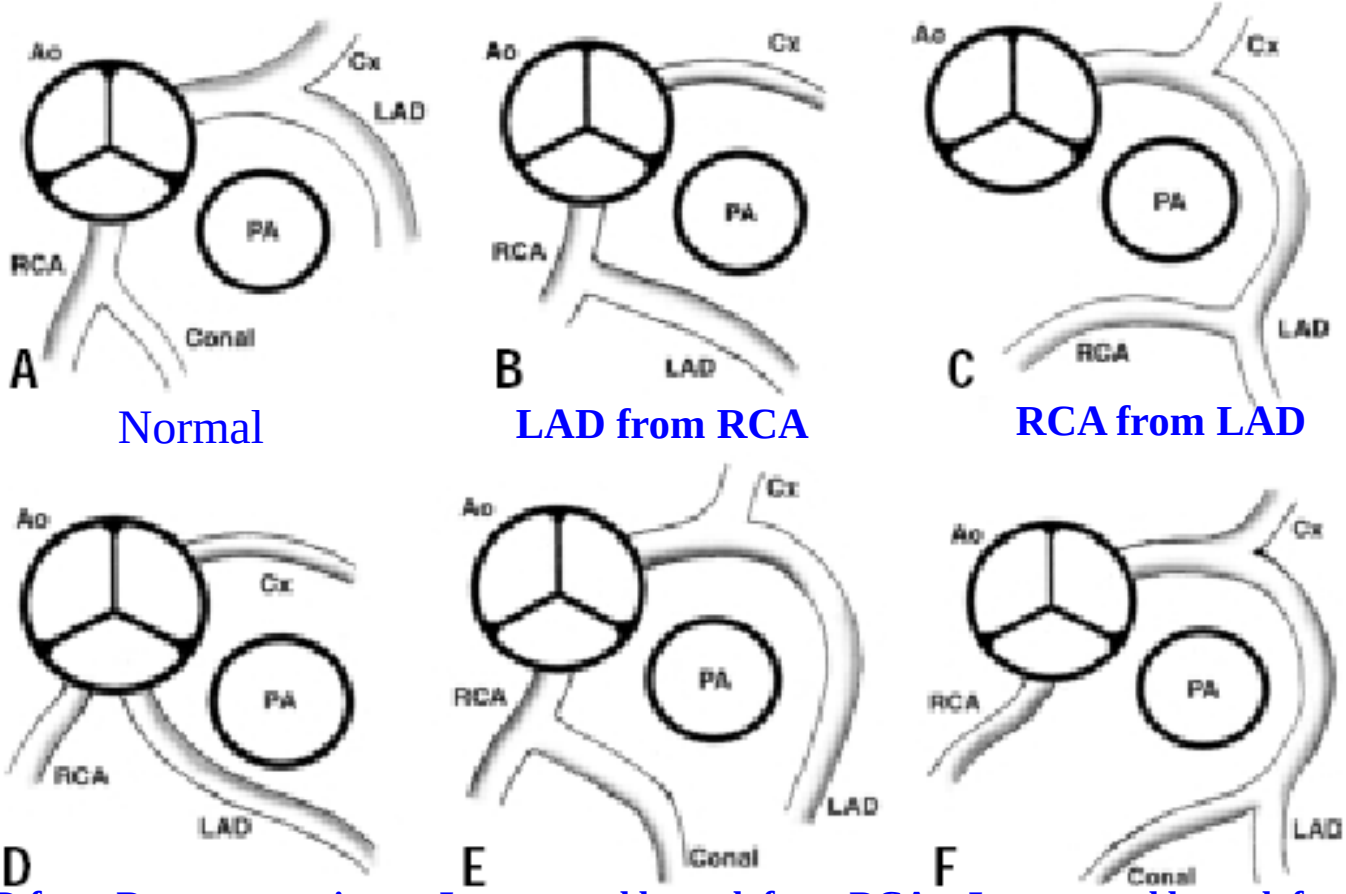
Functions transiently at best

By Bigras et al. (JTCS 1966;112:33) no significant differences in the degree of early postoperative pulmonary regurgitation or in clinical outcomes

By Gundry at al. (JTCS 1994;107-908) 16 of 19 patients had competent monocusp valves immediately after operation, but only one of 7 patients had a competent valve by 24 months postoperatively

❑ **If extensive reconstruction for the branch pulmonary arteries is required or if there is distal disease of the pulmonary vasculature, inclusion of a monocusp in the repair may improve hemodynamics in the immediate postoperative state.**

3) Anomalous coronary artery crossing RVOT



Normal

LAD from RCA

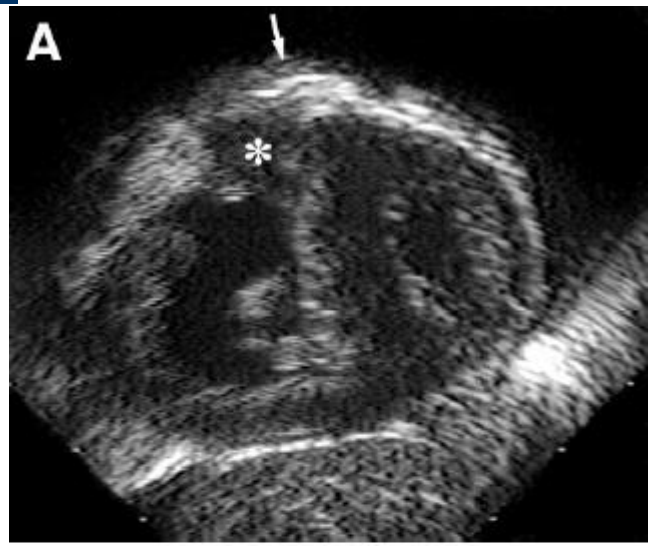
RCA from LAD

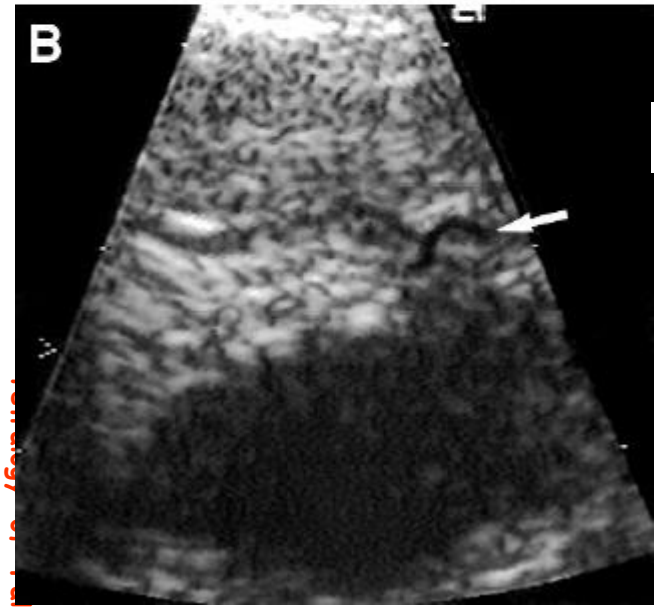
LAD from Rt coronary sinus

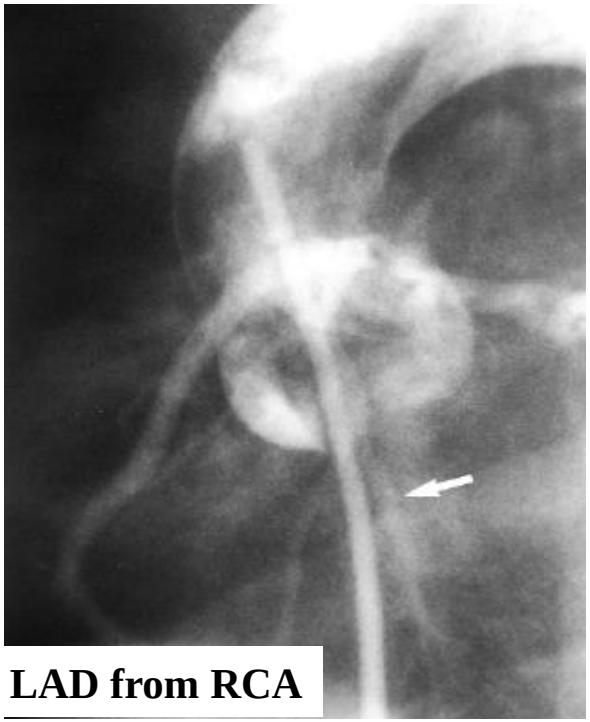
Large conal branch from RCA

Large conal branch from LAD

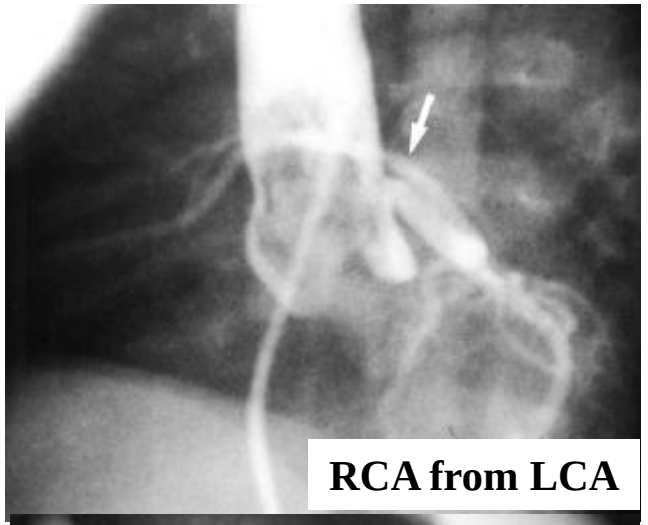
3) Anomalous coronary artery crossing RVOT



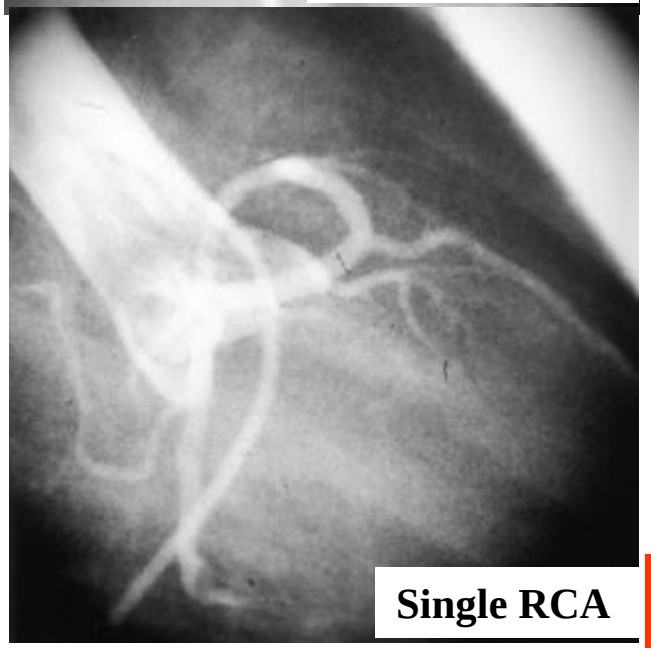




LAD from RCA



RCA from LCA



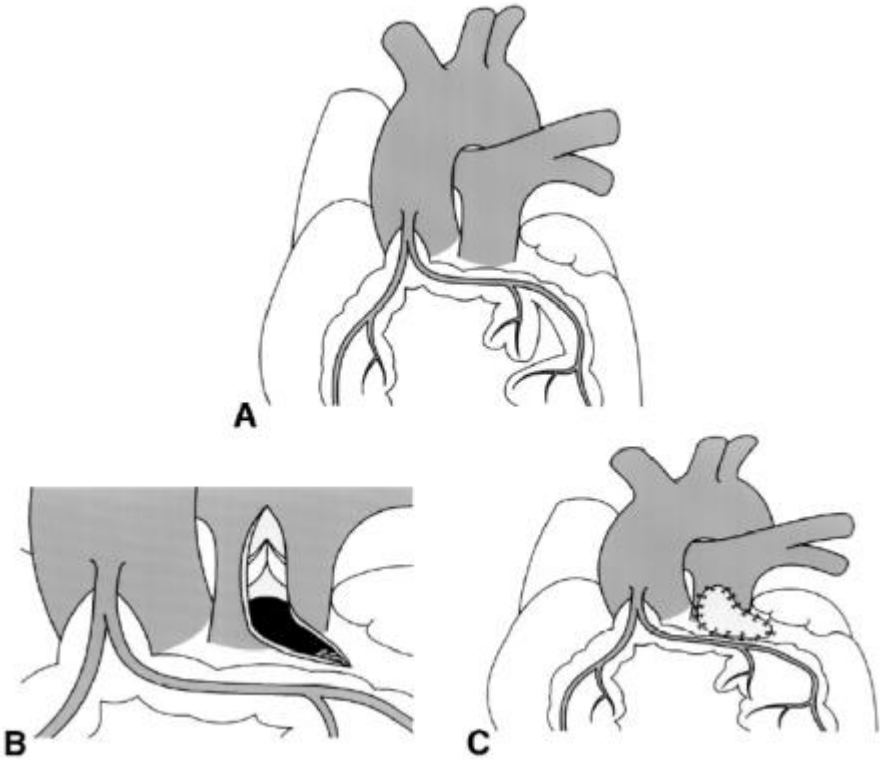
Single RCA

3) Anomalous coronary artery crossing RVOT

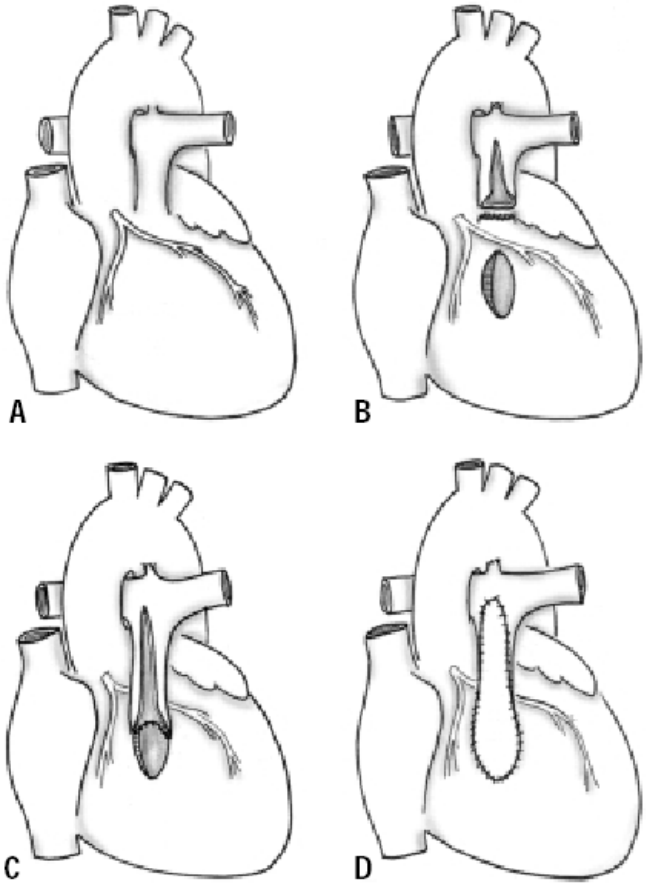
Surgical managements

- Transatrial and transpulmonary approach
- Incision through MPA and PV annulus and to left of anomalous LAD
- RVOT patching under the anomalous coronary artery
- Conduit reconstruction
- Translocation of the pulmonary artery to a distal ventriculotomy
- Proximally based pulmonary artery flap - double outlet technique

3) Anomalous coronary artery crossing RVOT

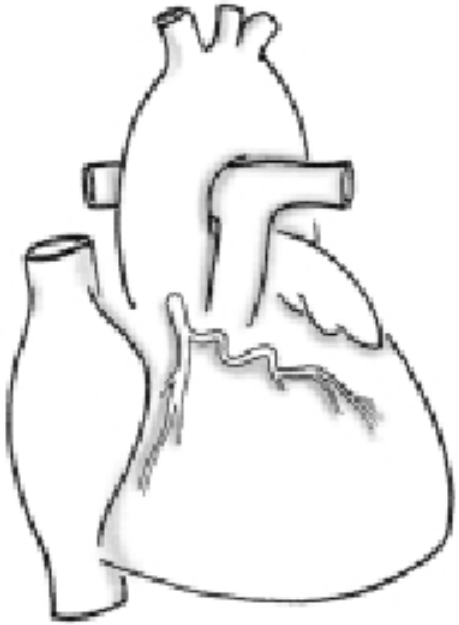


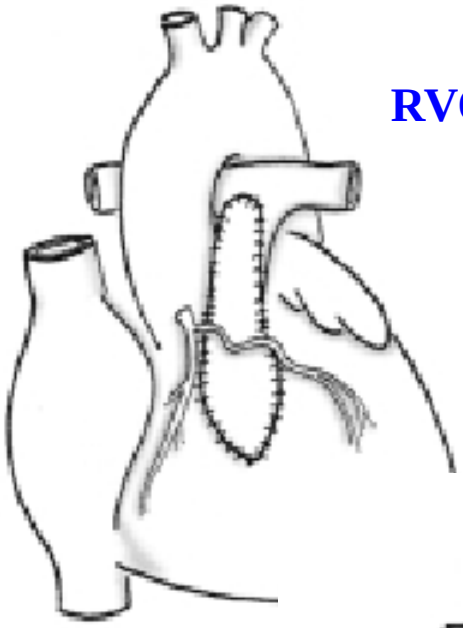
Incision to left of anomalous LAD



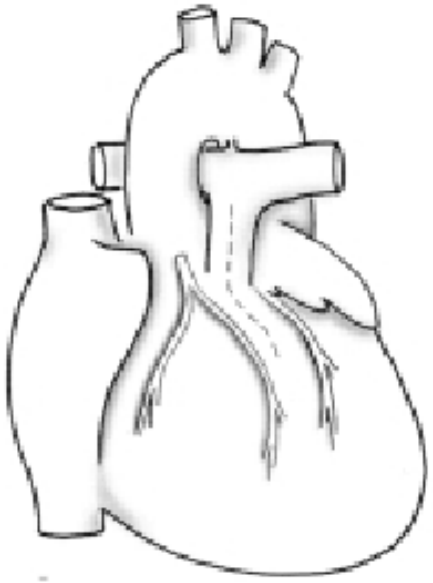
Translocation of MPA to a ventriculotomy

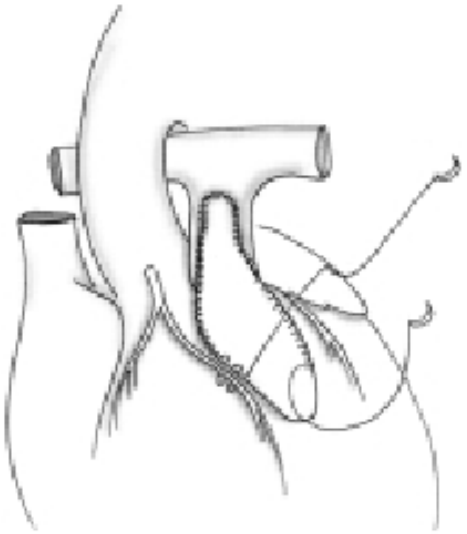
3) Anomalous coronary artery crossing RVOT



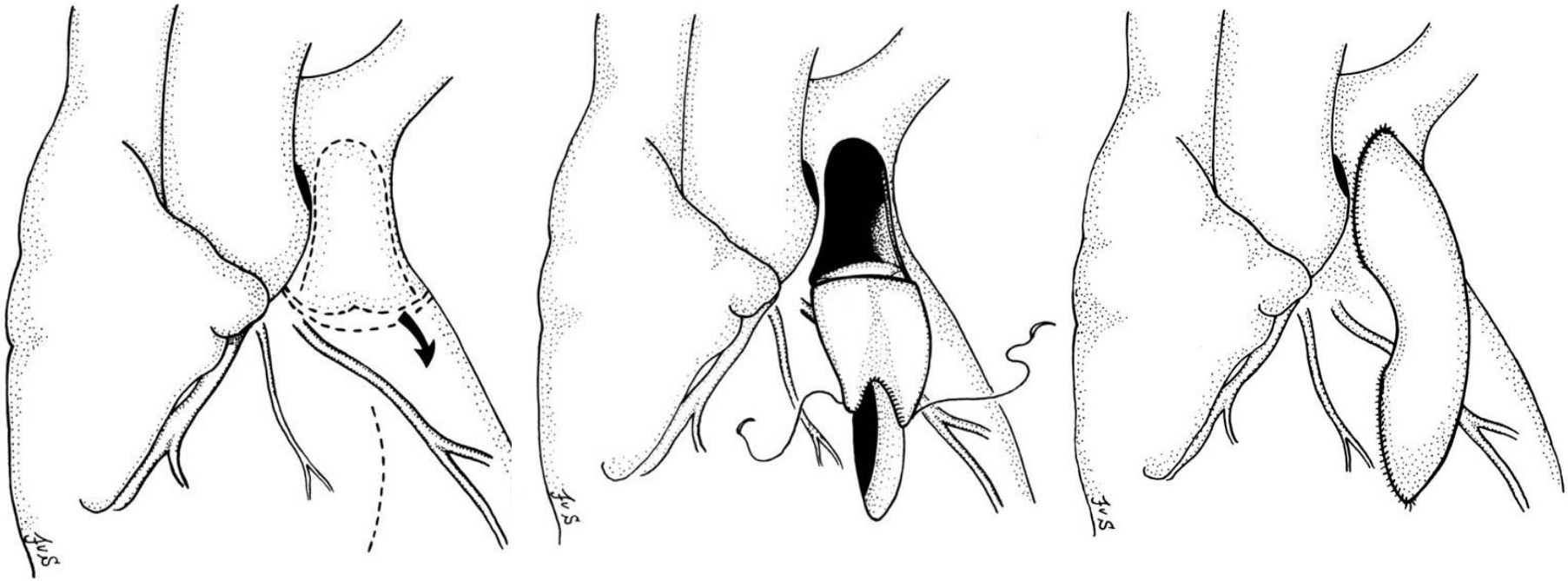


RVOT patching under the anomalous CA





3) Anomalous coronary artery crossing RVOT



Pulmonary artery flap - double outlet technique

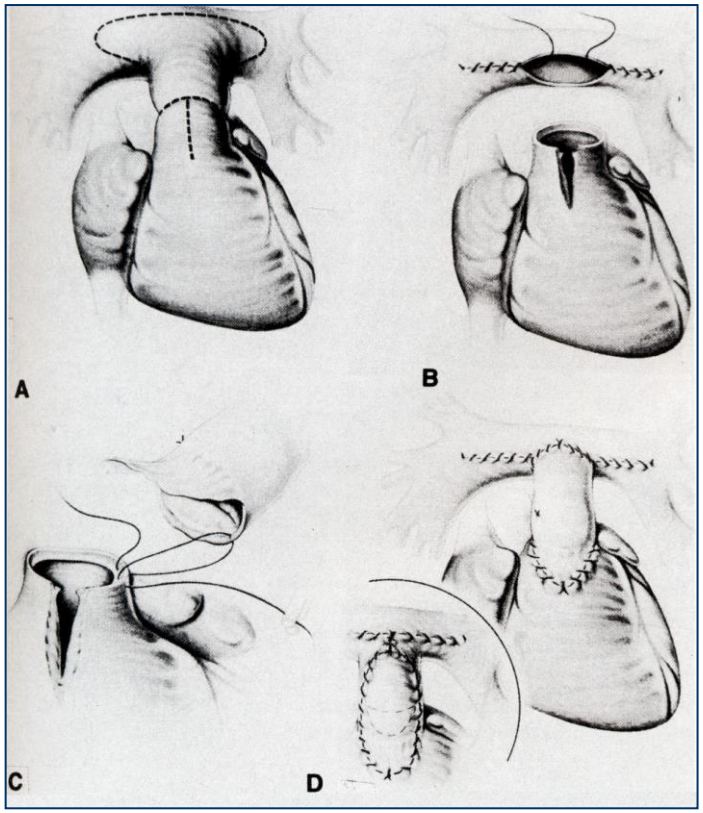
TOF with Absent Pulmonary Valve

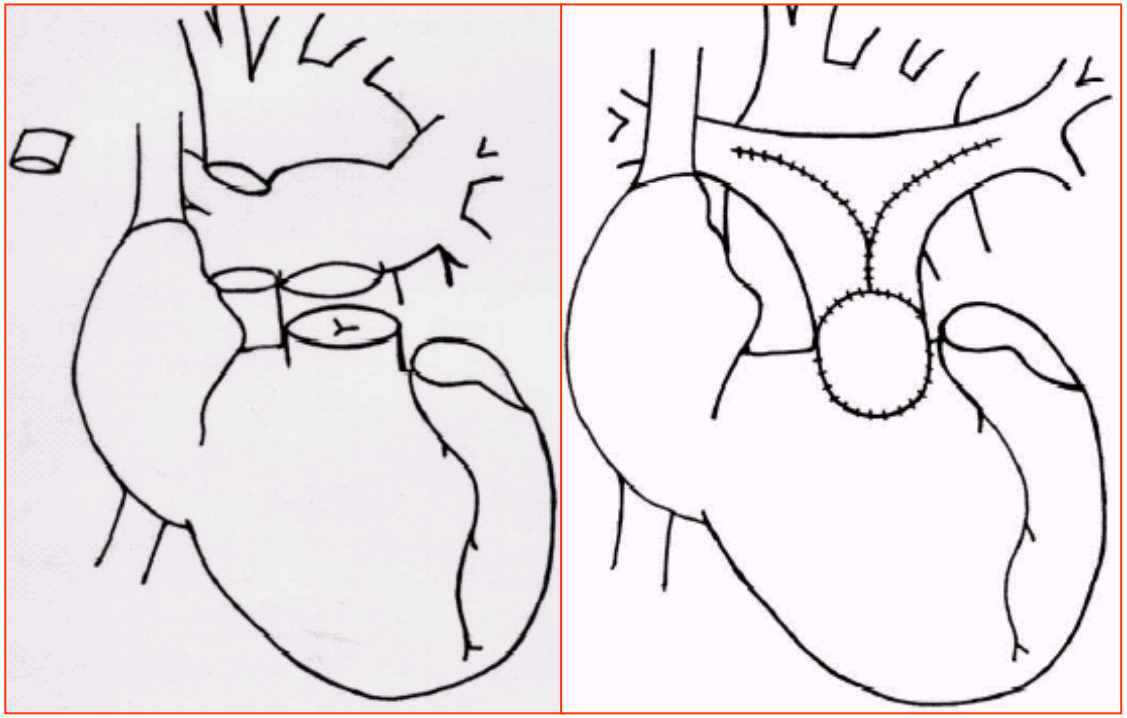
- 5%

- PR through fetal development or abnormality in the vessel wall
  → Aneurysmal dilatation of PAs → Airway compression
- Absence of decompressing PDA → Severe pulmonary hypertension
  → Destroyed PV and Dilated PAs

- Prompt operation in the patient with severe respiratory symptom
- Without severe respiratory symptom, elective repair at 6 mon of age
- Early operative mortality; 21.4%
- 1 year-survival; 77%, 10-year survival; 71% (Ann Thorac Surg 1999;67:1391)

Operative Technique

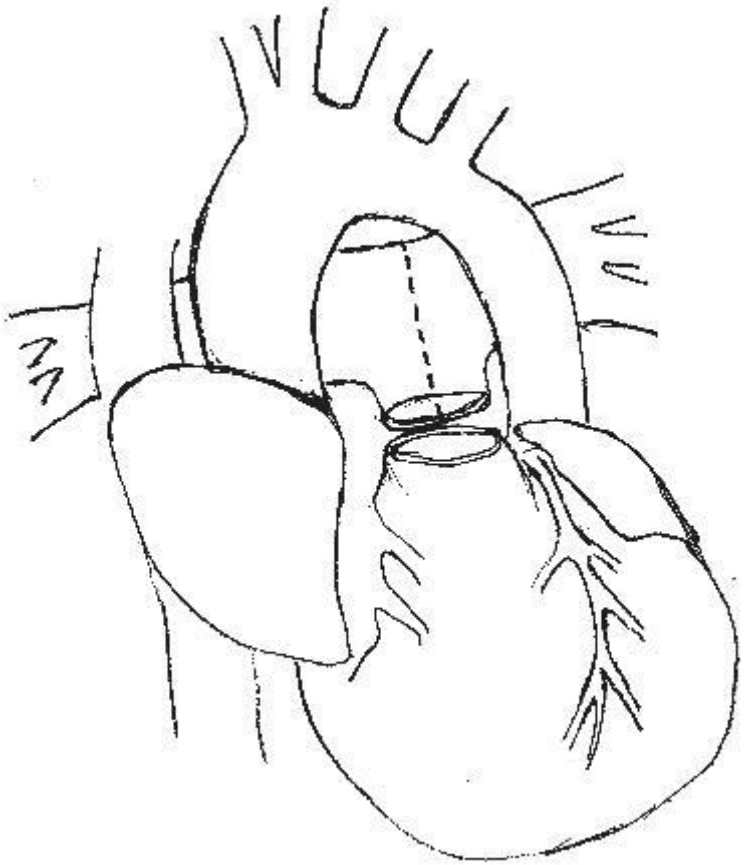


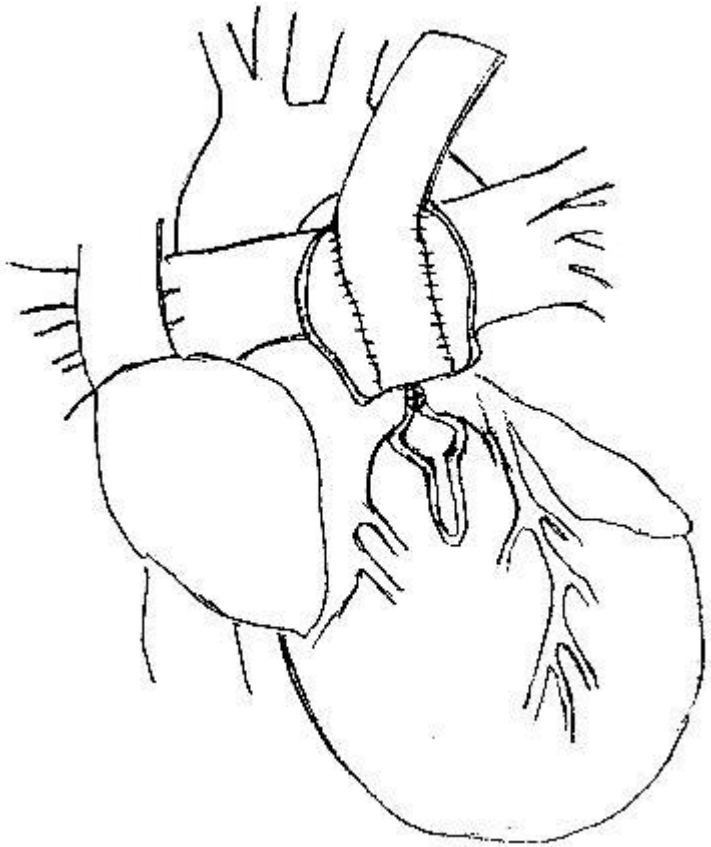


Hraska V, et al. Eur J Cardiothorac Surg 2002;21:711

TOF with Absent Pulmonary Valve

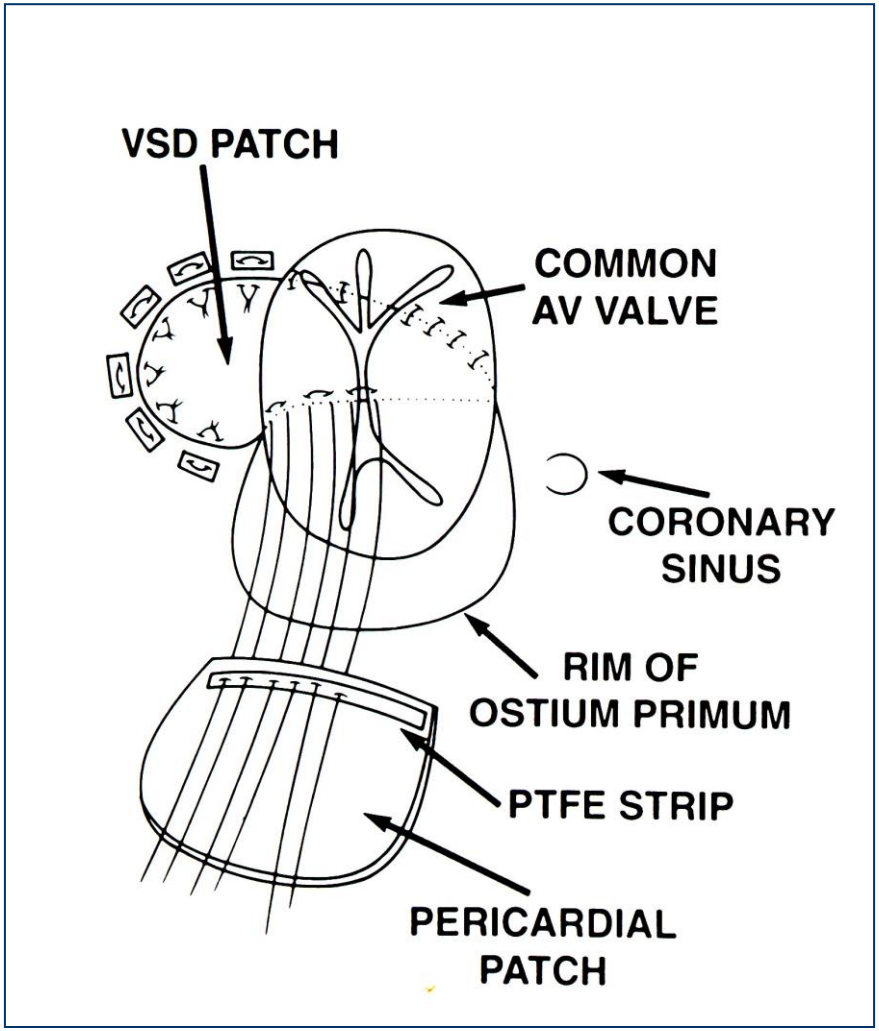
Operative Technique





TOF with Complete AVSD

- 2%
- More common in Down syndrome
- Always Rastelli type C
- RA or RA and RV combined approach for VSD closure
- Transannular patch or pulmonary valve insertion
- Timing of surgery for symptomatic early infants;
  Early primary ? or 6 month of age?



Results of Surgery

Early mortality

Currently 0.5-3%

Complete Repair of Tetralogy of Fallot at the University of Michigan, January 1993 to October 1999, N = 217

Survival	Total	Alive	Dead	95% Confidence Interval
TETRALOGY OF FALLOT (ALL)				
<1 mo	28	27 (96.4)	1 (3.6)	0-10.5
1 mo–1 y	140	135 (96.4)	4 (3.6)	0.5-6.7
>1 y	49	47 (95.9)	1 (4.1)	0.9-7.3
Total	217	209 (96.3)	8 (3.7)	1.2-6.2
TETRALOGY OF FALLOT WITH PULMONARY STENOSIS				
<1 mo	14	13 (92.9)	1 (7.1)	0-20.5
1 mo–1 y	127	123 (96.9)	4 (3.1)	0.1-6.1
>1 y	34	32 (94.1)	2 (5.9)	0-13.8
Total	175	168 (96)	7 (4)	1.1-6.9
TETRALOGY OF FALLOT WITH PULMONARY ATRESIA				
<1 mo	14	14 (100)	0 (0)	N/A
1 mo–1 y	4	3 (75)	1 (25)	17.2-67.2
>1 y	7	7 (100)	0 (0)	N/A
Total	25	24 (96)	1 (4)	3.6-11.6
TETRALOGY OF FALLOT WITH COMPLETE ATRIOVENTRICULAR SEPTAL DEFECT				
<1 mo	0	0	0	N/A
1 mo–1 y	9	9 (100)	0 (0)	N/A
>1 y	8	8 (100)	0 (0)	N/A
Total	17	17 (100)	0 (0)	N/A

Summary excludes repairs performed with or after unifocalization procedures for multiple aortopulmonary collateral arteries.
Values in parentheses are percentages.
N/A, Not applicable.

Results of early primary repair

Table 21-1. Summary of One-stage Early Repair of TOF

Authors	Time period of operation	No. of patient	Operation	Age at operation	Early death	Follow-up duration	Late death	Reoperation
Reddy (1995)[125]	1992-1995	30 Neonate (10) TOF/PA (5) Sx (33%)	TAP (60%)	53d (4-90d)	1 (3.3%)	19mo	No	No
Stellin (1995)[33]	1978-1994	51 TVA (A) TA-TPA (B)	TAP (65%) TVA (43%)	4.2mo (21d-6mo)	1 (2%)	7.3yr (A)	No	5 (10%) RVOTO (2) Subaortic stenosis (1) PPM (1) Other (1)
Pigula (1999)[126]	1988-1996	99 TOF/PS (73) TOF/PA (26) 신생아 (57) Sx (90%)	*TAP (84%) TVA (96%)	27d (2-87d)	*2 (2.7%)	4yr (1-8.8yr)	*1 (1.4%)	*12 (16%)
Parry (2000)[68]	1992-1999	42 Sx (0)	TAP (24%)	62d (4-87d)	No	38mo (12-64mo)	No	1 RVOTO
Tamesberger (2008)[59]	1995-2006	90 Neonate (25)	TAP (66%) TVA (93%)	49d (3-116d)	No	56mo (4-146mo)	2 (2%)	7 (8%) RVOTO (4) PR (1) VSD (1) PA aneurysm (1)
Kantorova (2008)[60]	1996-2005	61 Neonate (12) Sx (44%)	TAP (72%)	3.3mo (0.02-6mo)	1 (1.6%)	4.5yr	1 (1.7%)	6 RVOTO (6)

* Patients with TOF-PA excluded

* NC: nonconfluent, PPM: permanent pacemaker insertion, Sx: symptomatic, TAA: transatrial approach to VSD, TAP: transannular patching, TA-TPA: transatrial-transpulmonary approach to VSD, TVA: transventricular approach to VSD

Results of primary repair in neonate (I)

Table 21-2. Summary of total repair in TOF neonate

Authors	Time period of operation	No. of patient	Operation	Age at operation	Early/Late death	Major postoperative complication	Follow-up duration	Reoperation
Di Donato (1991)[39]	1973-1988	27 TOF/PS 14 TOF/PA 13 Sx (100%)	TAP (25) RV-PA conduit (2)	3d	5 (19%)/ 2 (9%)	3 (14%) Respiratory failure Low cardiac output syndrome JET	5yr	6 (27%) RVOTO (6)
Hirsch (2000)[28]	1988-1999	61 TOF/PS 31 TOF/PA 24 TOF/NC 6 Sx (100%)	TAA (62%) TVA (38%) *TAP (29/31) *RV-PA conduit (2/31)	16d	1 (1.6%) 4 (6.7%)/	N.A.	62mo	22 (39%) RVOTO (19) Branch PA (11) PR (4) VSD (1)
Kolcz (2005)[127]	1998-2004	46 TOF/PA 5 Sx (30%)	TVA (100%) TAP (88%)	5d (1-29d)	3 (4.5%)/ No	N.A.	35mo	3 (6.8%) RVOTO
Tamesberger (2008)[59]	1995-2006	25 Sx (100%)	TAP (84%)	15d (3-28d)	No	2 (8%)	N.A.	3 (12%) RVOTO (2) PR (1)
Kanter (2010)[65]	2002-2008	20 Sx (100%)	TAA (100%) TAP (100%)	9.7d	No/ 2 (10%)	N.A.	47mo	4 (22%) RVOT (3) PPM (1)

Results of primary repair in neonate (II)

Table 21-2. Summary of total repair in TOF neonate

Di Donato (1991)[39]	1973-1988	27 TOF/PS 14 TOF/PA 13 Sx (100%)	TAP (25) RV-PA conduit (2)	3d	5 (19%)/ 2 (9%)	3 (14%) Respiratory failure Low cardiac output syndrome JET	5yr	6 (27%) RVOTO (6)
Hirsch (2000) [28]	1988-1999	61 TOF/PS 31 TOF/PA 24 TOF/NC 6 Sx (100%)	TAA (62%) TVA (38%) *TAP (29/31) *RV-PA conduit (2/31)	16d	1 (1.6%)/ 4 (6.7%)	N.A.	62mo	22 (39%) RVOTO (19) Branch PA (11) PR (4) VSD (1)
Kolcz (2005) [127]	1998-2004	46 TOF/PA 5 Sx (30%)	TVA (100%) TAP (88%)	5d (1-29d)	3 (4.5%)/ No	N.A.	35mo	3 (6.8%) RVOTO
Tamesberger (2008)[59]	1995-2006	25 Sx (100%)	TAP (84%)	15d (3-28d)	No	2 (8%)	N.A.	3 (12%) RVOTO (2) PR (1)
Kanter (2010) [65]	2002-2008	20 Sx (100%)	TAA (100%) TAP (100%)	9.7d	No/ 2 (10%)	N.A.	47mo	4 (22%) RVOT (3) PPM (1)

* Patient with TOF-PA excluded

* JET; junctional ectopic tachycardia, N.A.; not available, NC; nonconfluent, PPM; permanent pacemaker insertion, Sx; symptomatic, TAA; transatrial approach to VSD, TAP; transannular patching, TVA; transventricular approach to VSD

Results of Surgery

- **Main causes of reoperation**
  - RVOT obstruction
  - Pulmonary valve regurgitation
  - Residual VSD
  - Permanent pacemaker implantation

- **Main causes of cardiac death in long-term survivors**
  - Sudden death
  - Heart failure

Long-term Follow-up

1) RV performance and functional status

- "Excellent long-term survival"
- By Alexiou et al, 20-year survival ; 98%, 99% in NYHA class I
- By Knott-Craig et al, 20-year survival; 98%,
  freedom from reintervention on RVOT after 20 years; 86%
- Performance during exercise testing is well maintained in most reports

University of Munich, Germany

1958-1977, 490 patients excluded operative or 1-year death

Mean age at repair; 12.2 ± 8.6 years

10-, 20-, 30, 36-year survival rate; 97%, 94%, 89%, and 85%

Mortality increased 25 years postoperatively from 0.24%/year to 0.94%/year

- Main causes of cardiac death in long-term survivors
  - Sudden death (arrhythmia)
  - Heart failure

2) Pulmonary valve regurgitation - PVR

- **2% of patients at 10-year follow-up and 12% after 20 years.**
- **PVR secondary to pulmonary insufficiency is rarely necessary in the absence of a transannular patch**
- **Risk factors;** ***TAP (large RVOT patch), Peripheral PA stenosis Residual VSD, Duration after TOF repair Early repair or late repair ?***

Degree of PR depends on

Size of pulmonary valve coaptation defect

Pulmonary afterload

Diastolic physiology

2) Pulmonary valve regurgitation - PVR

Indication for PVR

- not well established
  - **a) symptomatic** (decreased exercise tolerance, decreased functional class)
  - **b) progressive RV enlargement and RV dysfunction**
  - **c) ventricular and supraventricular arrhythmia**

Emory University School of Medicine

RV/LV volume >2 in MRI

Increasing TR in the presence of severe PR

University Children Hospital, Zurich (Eur Heart J 2005;26:2721)

RV end-diastolic volume > 150ml/m^2

Normal RV function and size by MRI

End-diastolic volume ≤ 108ml/m^2

End-systolic volume ≤ 47ml/m^2

RV ejection fraction ≥ 50%

2) Pulmonary valve regurgitation - PVR

Clinical effect of PVR

- **RV end-diastolic volume >170 ml/m^2 or RV end-systolic volume >85 ml/m^2**
  → no "normalized" RV volume after surgery (Am J Cardiol 2005;95:779)
- **Only 44% had normalization of RV size and symptomatic relief** (ATS 2002;73:1794)
- **Late PVR failed to normalize RV volume**

- **RV end-diastolic volume ≤ 160 ml/m^2 or RV end-systolic volume ≤ 82 ml/m^2**
  → "normalized" RV volume after surgery (Circulation 2007;116:545)
- **PVR is associated a substantial decrease in arrhythmias and stabilization of the QRS duration**

2) Pulmonary valve regurgitation - PVR

Choice of valve at PVR

- **Bioprosthesis (most preferred), Mechanical valve, Homograft**

- **Consideration of mechanical valve**

  When anticoagulation is required

  (mechanical valve at aortic or mitral position, atrial fibrillation)

  Rapid degeneration of previous tissue valve

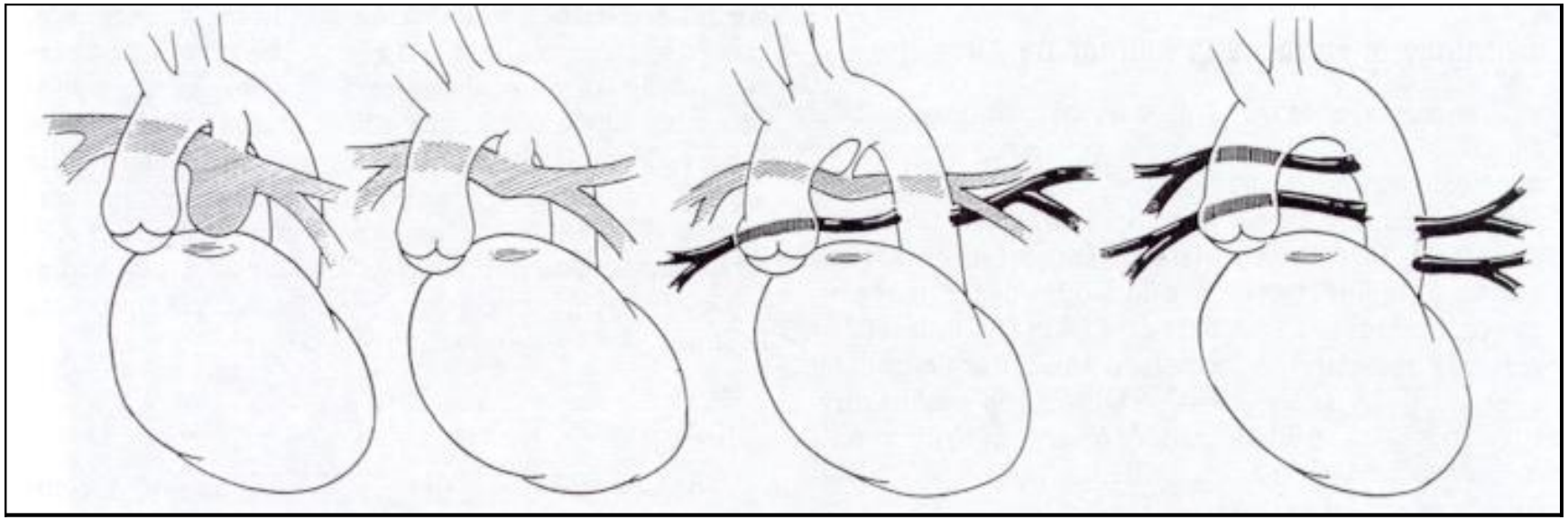
  Previous multiple reoperation

3) Arrhythmias

- **In patients followed for more than 20 years after TOF;**
  - *2-4% for atrial fibrillation / flutter*
  - *3-4% for sustained ventricular tachycardia*
  - *2-4% for sudden cardiac death*
- **Holter monitoring (24-hour) detects a higher percentage of arrythmia;**
  - *19% for sustained ventricular tachycardia*
  - *23% for atrial fibrillation / flutter*
- **Risk factors;**
  - *Elevated RV volume and pressure*
  - *Decreased right and left ventricular ejection fractions*
  - *Pulmonary regurgitations*
  - *RVOT aneurysm and significant stenosis*
- **QRS duration greater than 180msec**
  predicting the occurrence of life threatening ventricular ectopy
- **Significant ventricular ectopy - ventricular approach; 39%** (Dietl CA et al circulation 1994;90:II7)
  transatrial approach; 2.8%
- **Severe PR - ventricular approach; 25.9%, transatrial approach 12.5%**

Tetralogy of Fallot with Pulmonary Atresia

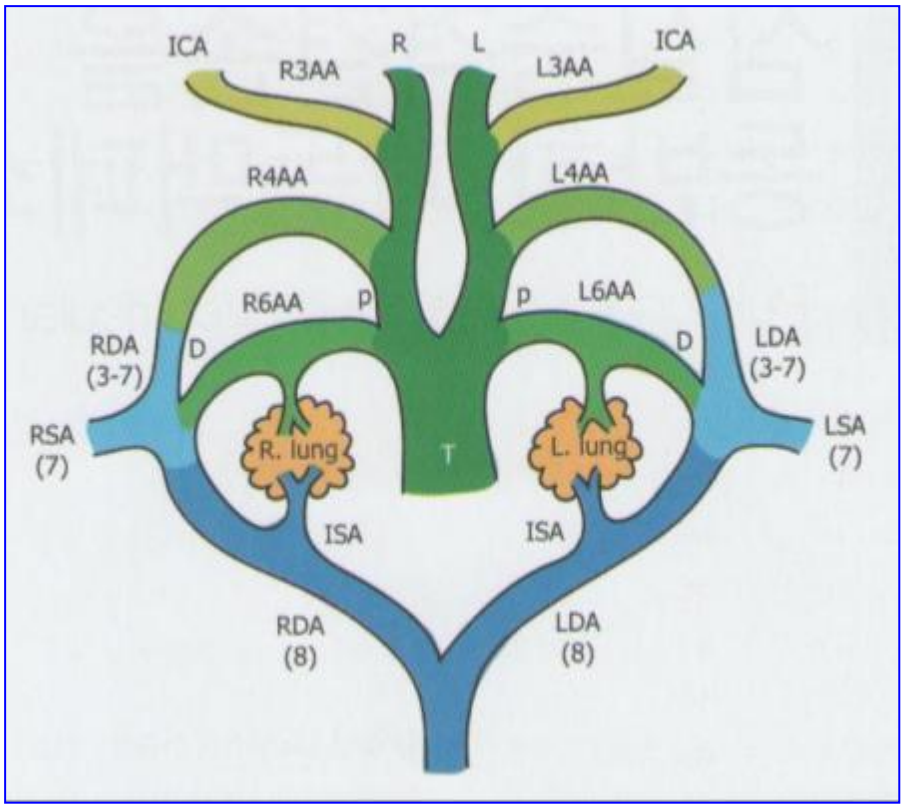
Spectrum



Tetralogy of Fallot with Pulmonary Atresia

Embryology

- ***Peripheral PA*** ; from systemic arterial and venous circulations of the primitive foregut
- ***Proximal mediastinal PA*** ; from 6th dorsal arches
- ***Proximal MPA*** ; formed by division of the original conotruncus



Anatomy

Central pulmonary artery

- **Confluent, nonconfluent, or absent**
- **If present, variable size, from just over a millimeter to normal size**
- **"gull wing" or "seagull" - small confluent pulmonary arteries**
- **Nonconfluent central pulmonary arteries, from PDA or MAPCA**

Aortopulmonary collateral arteries

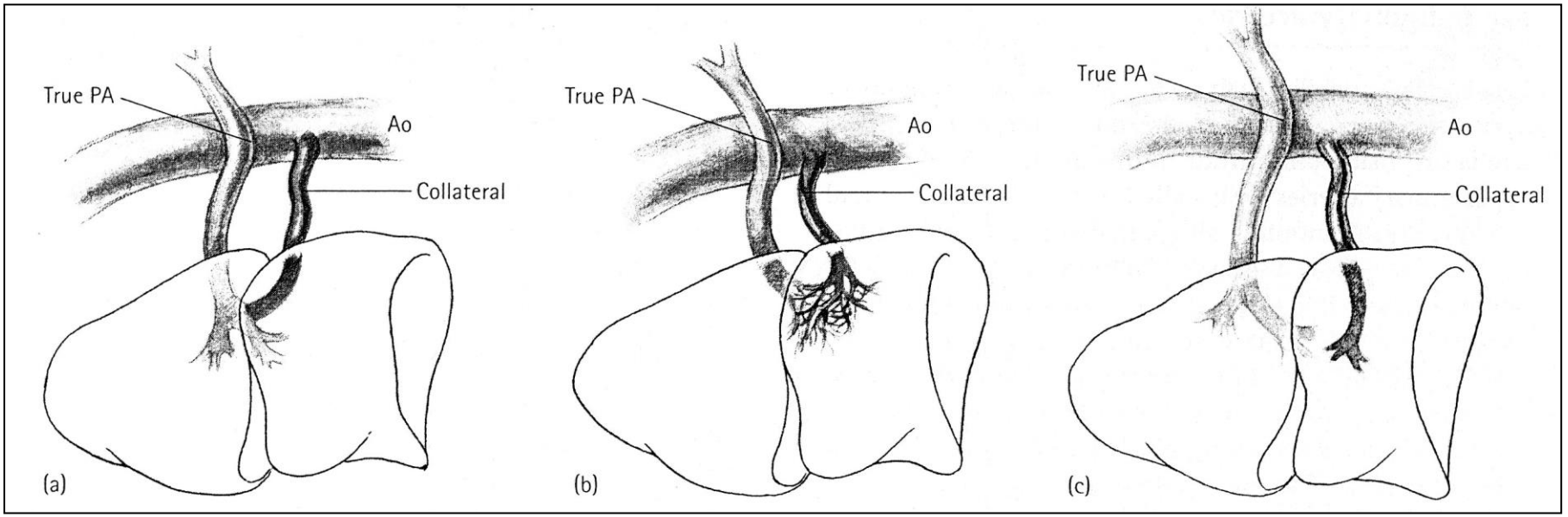
- **MAPCAs; from embryonic aortic arch (resemble midsize muscular artery)**
- **Intrapulmonary artery; from foregut (classified as musculoelastic artery)**
- **Communicating MAPCA or Noncommunicating MAPCA**

- Highly variable pulmonary arterial morphology, but **some predictable pattern**

1) The PDA connected to a central PA; peripheral PA distribution is normal
(no systemic collateral arteries in that hemithorax,
ductus arteriosus does not coexist with MAPCAs in the same lung)

2) The right upper lobe and the left lower lobe segments often are supplied by single, noncommunicating MAPCAs
(from subclavian artery and descending aorta)

Anatomy


True PA
Ao
Collateral
(a)
True PA
Ao
Collateral
(b)
True PA
Ao
Collateral
(c)

Natural History

- **Variable history dependent on the anatomy of the pulmonary arteries**
  - *ductus-dependent circulation*
  - *excessive pulmonary circulation with heart failure*
  - *local pulmonary hypertension*
  - *acquired occlusion or stenosis of MAPCAs can cause worsening cyanosis*

- **Survey for 218 patients having intervention or not** (Bull K et al. J Am Coll Cardiol 1995;491)
- *survival at 1 year ; 60%*
  of those alive at 1 year ; 65% survived 10 years
  from age 10 to 35 ; only 16% survived

- **65% presented in infancy presented with**
  - *50% being cyanotic*
  - *25% having heart failure*
  - *25% having "well-balanced" circulation*

- **Subsequent death was most commonly associated /c cardiac operation**

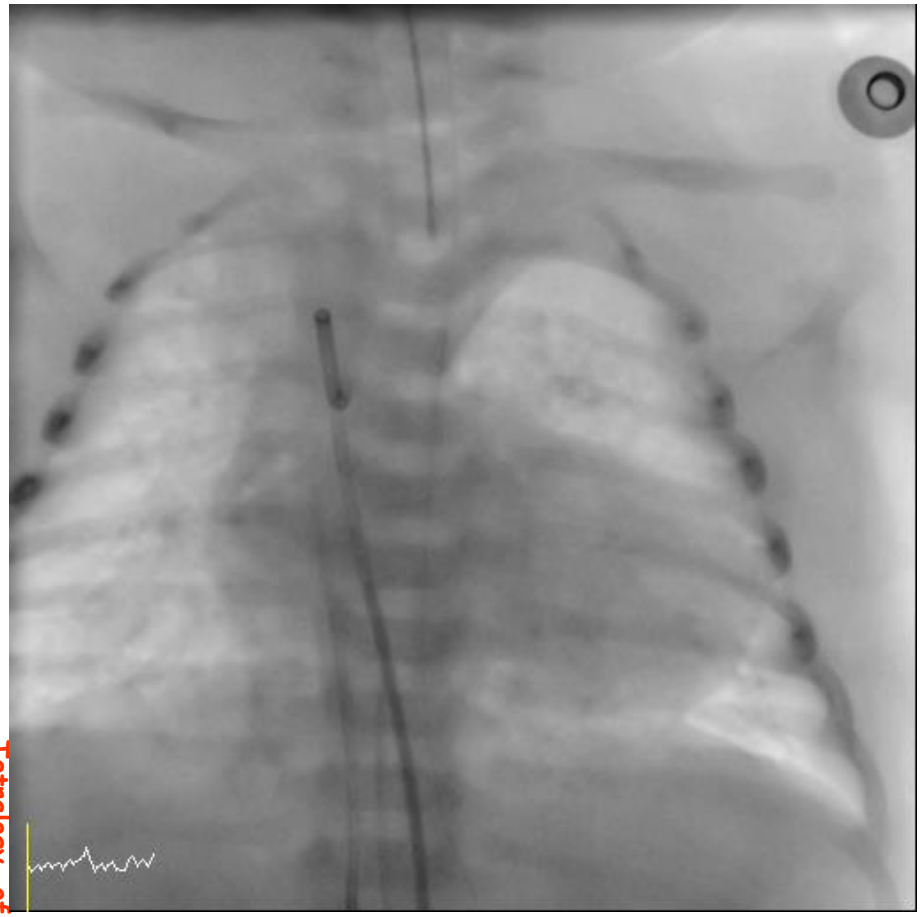
Associated Anomalies

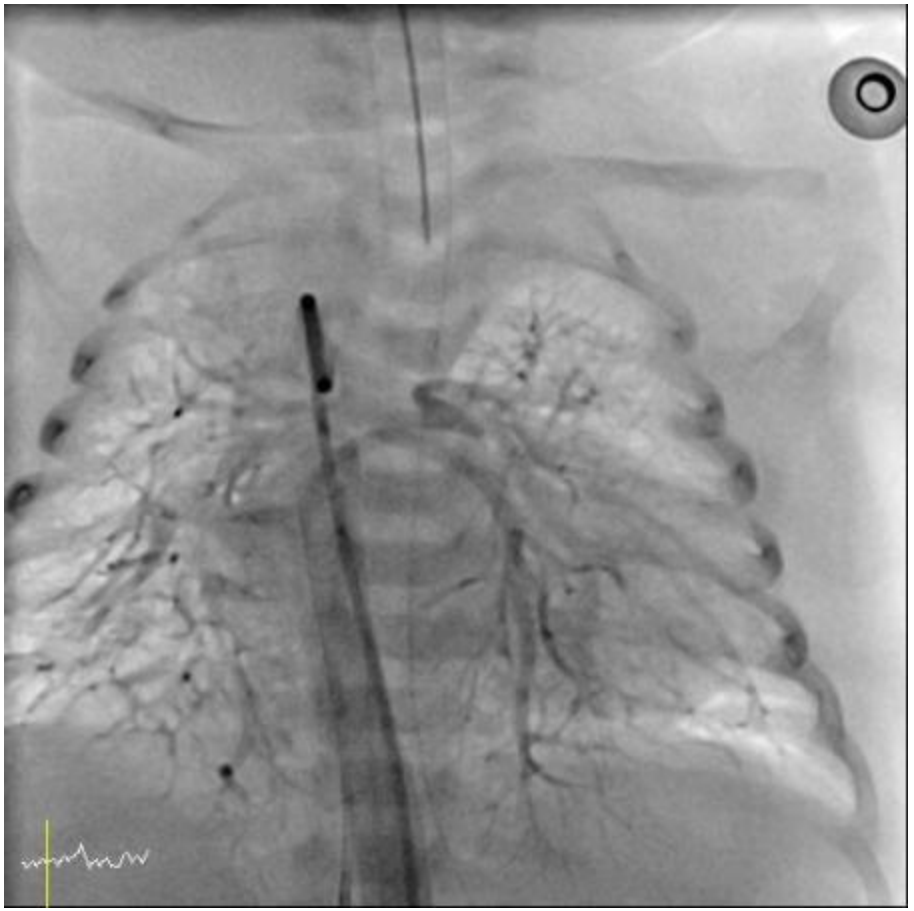
- Velo-cardio-facial syndrome in about 10%
- DiGeorge syndrome may also be associated
- Deletion of chromosome 22q11 in 30%
  (higher prevalence in those having MAPCAs)
- Right aortic arch in 20%

Diagnostic Studies

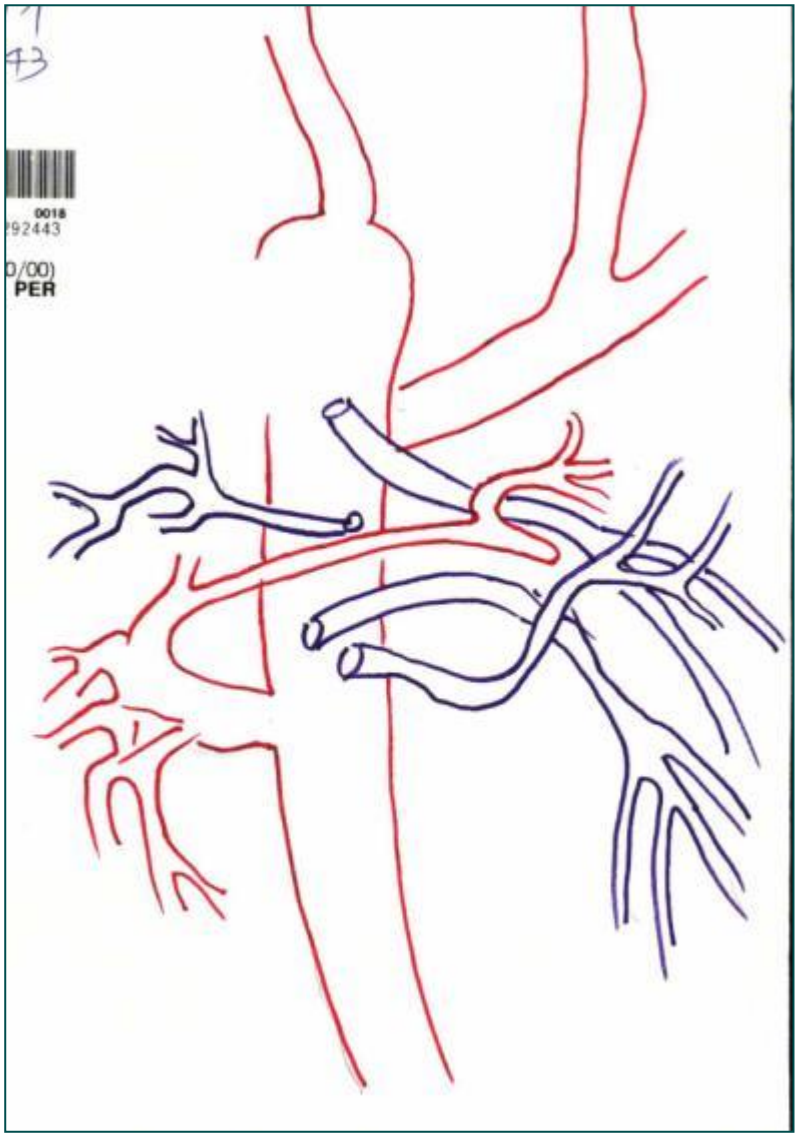
- Cardiac catheterization
  central diagnostic tool for MAPCA and native PAs evaluations
- Direct hemodynamic measurements of individual collaterals
  distal pressure > mean pressure of 20-25mmHg
  → PVOD has already developed or will develop within the lung segments
- It is frequently necessary to use retrograde venous wedge angiography to identify tiny true central pulmonary artery
- MRI ; particularly helpful in understanding complex collateral and PA anatomy and aid in the planning of unifocalization procedures

Before operation

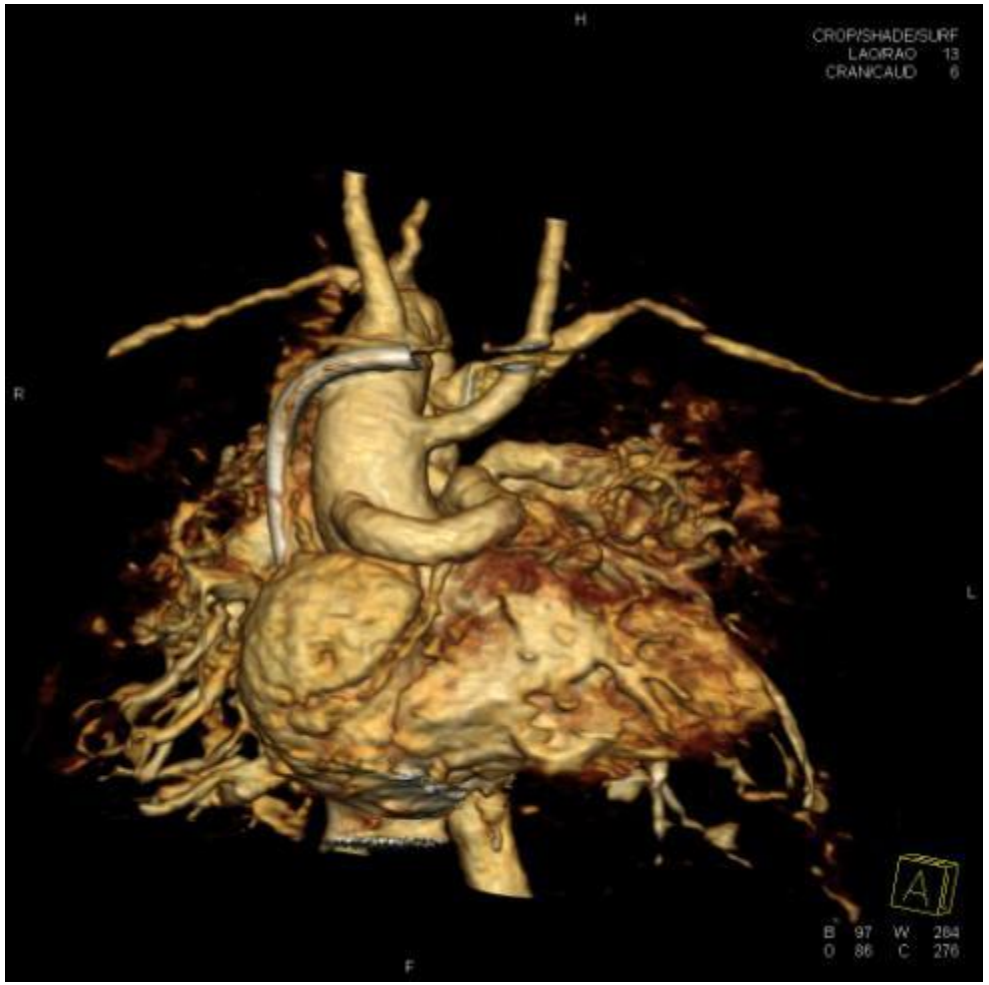




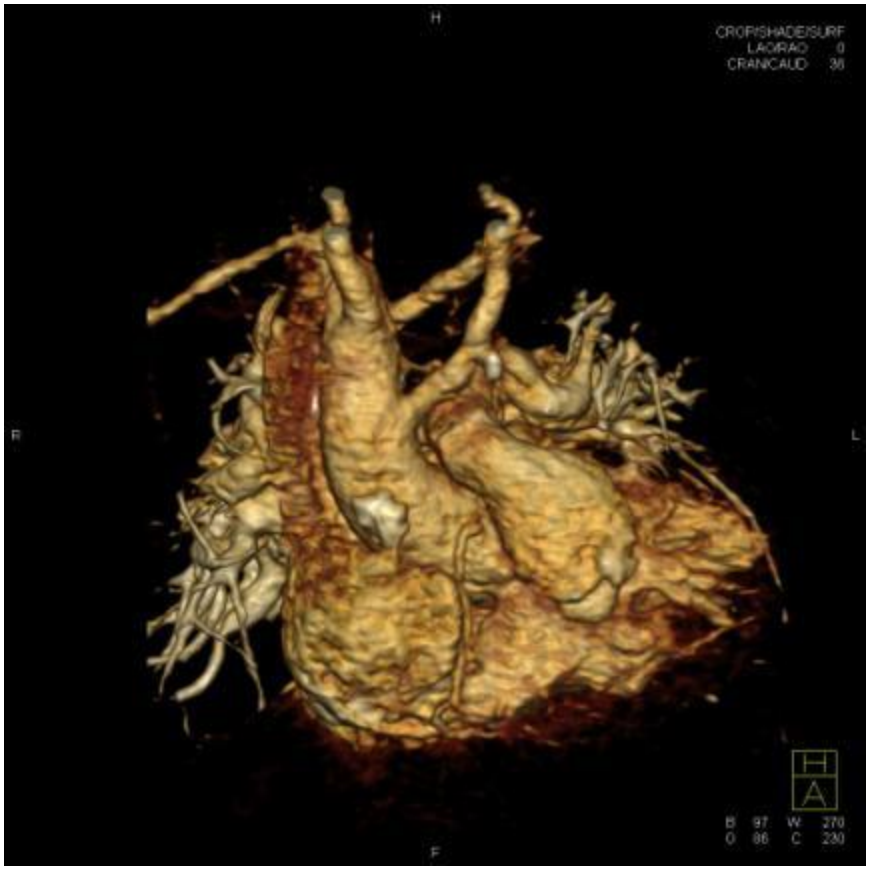
Before operation



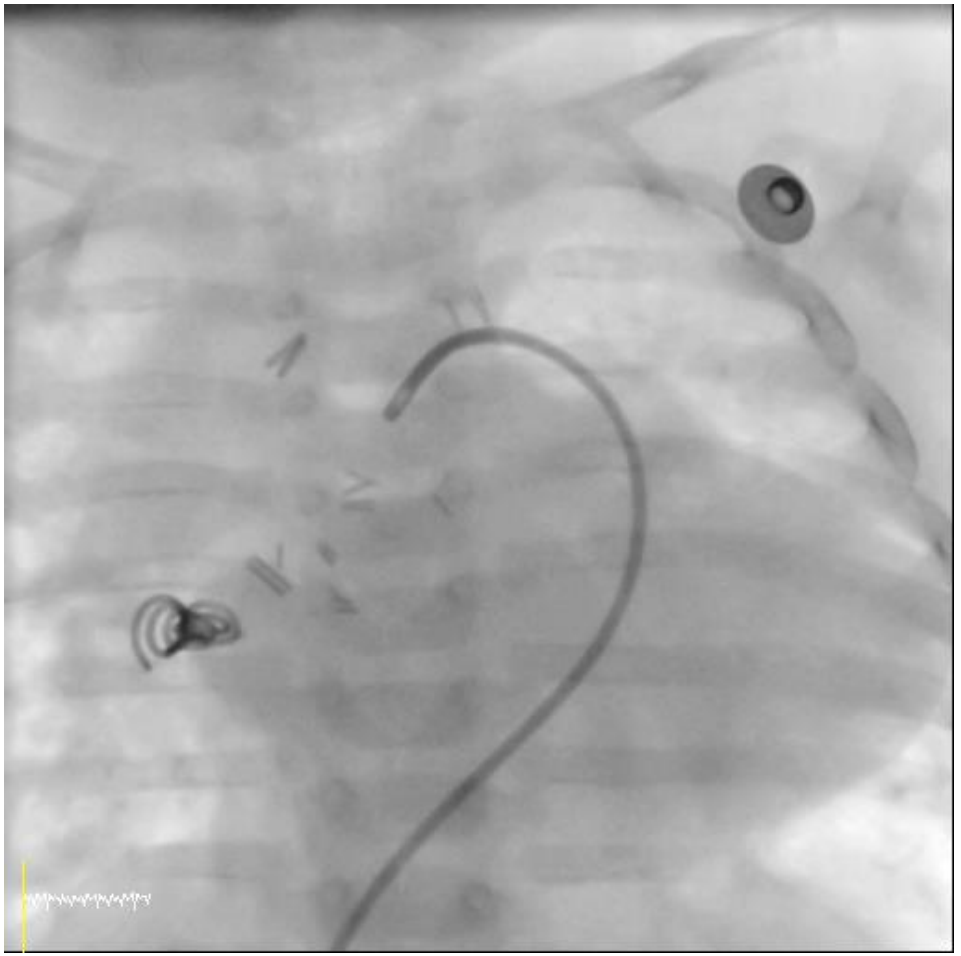
After central shunt



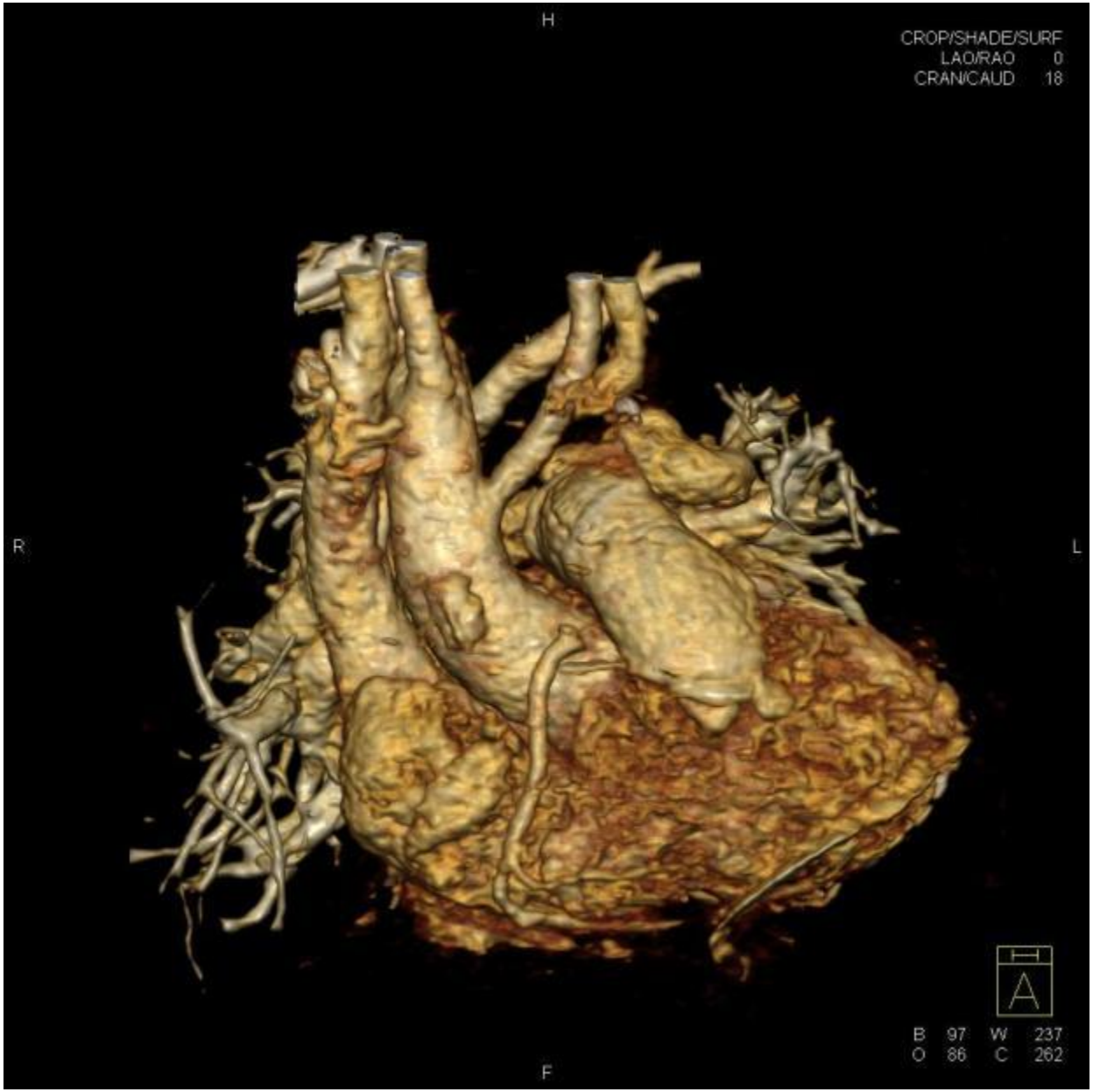
After LPA unifocalization and RV-PA conduit



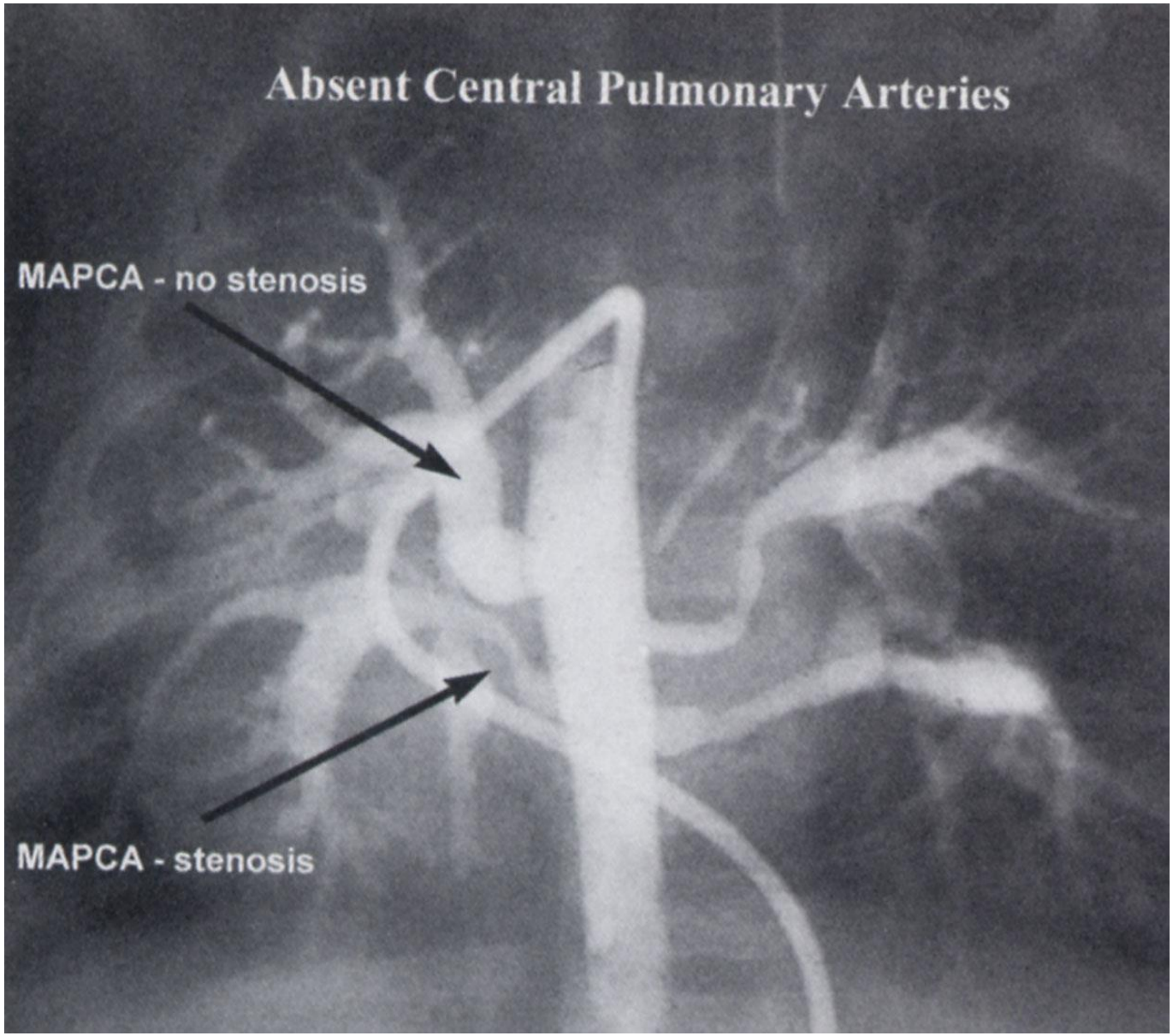
After LPA unifocalization and RV-PA conduit



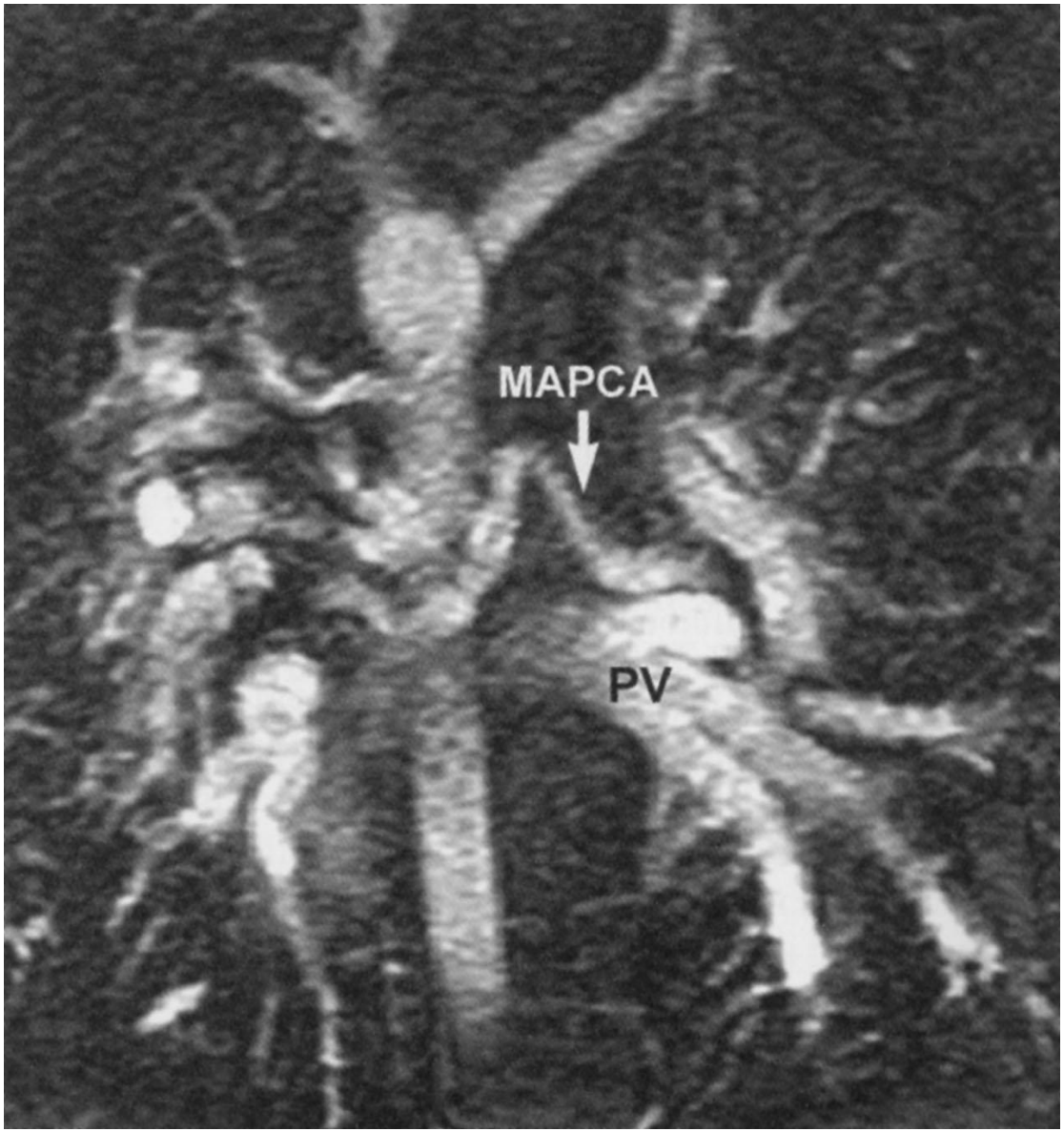
After VSD closure and conduit change with PA angioplasty



Tetralogy of Fallot with Pulmonary Atresia

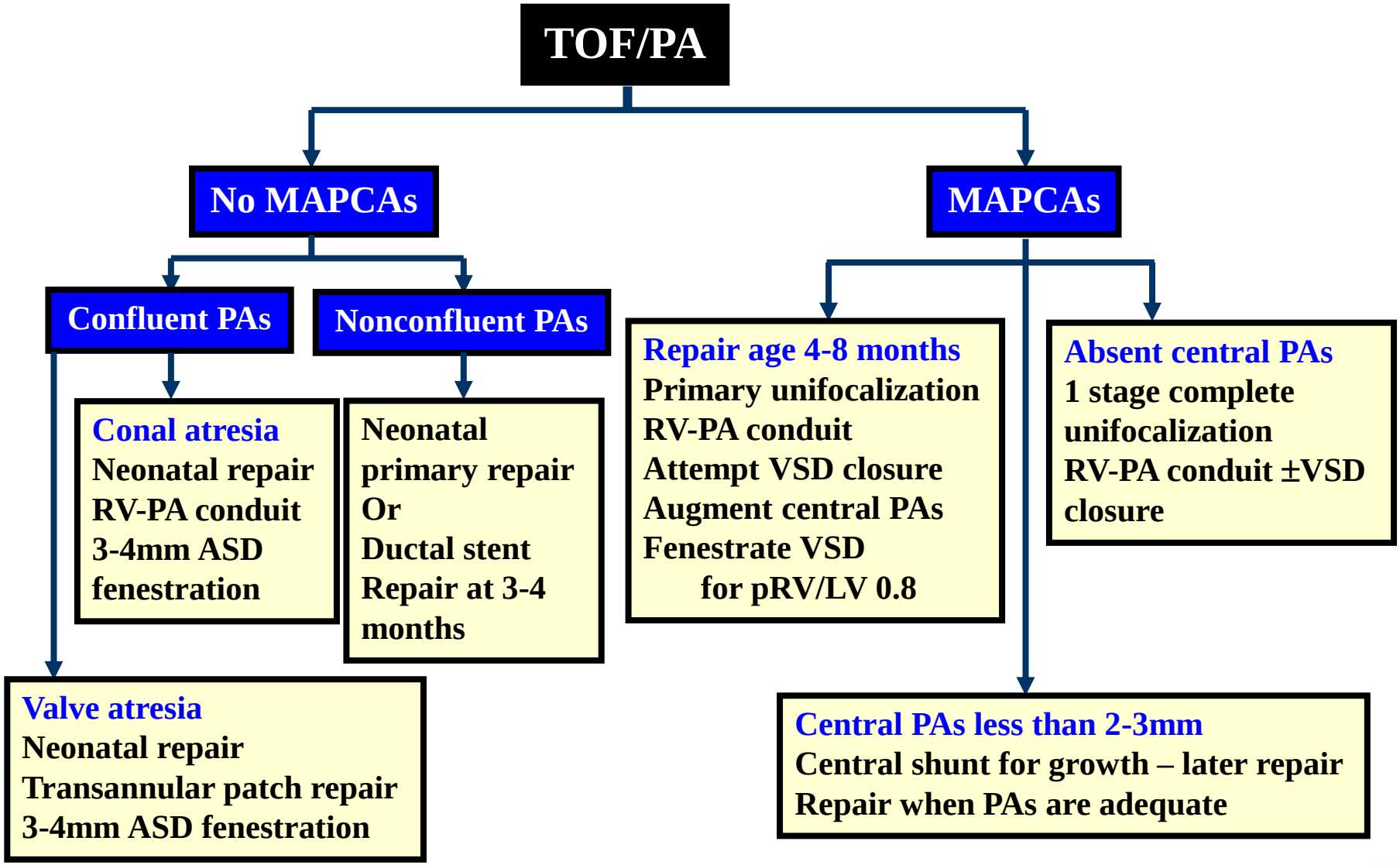


Tetralogy of Fallot with Pulmonary Atresia



Repair Strategies

Decision making algorithm for TOF/PA at HSC in Toronto



Repair Strategies

TOF/PA/absent MAPCAs/present PDA

- **Primary repair within the first 2 weeks of life using RV-PA conduit or transannular patch (Hospital for Sick Children in Toronto)**
  Foramen ovale is narrowed to 3-4mm
  Use of delayed sternal closure and peritoneal drain
- **Second alternative; initial shunt or RV-PA conduit**
- **then, complete repair around 6-10 months of age**

TOF/PA/MAPCAs

- **Single stage complete unfocalization with RV to PA conduit and VSD closure at age of 4-8 months**
  High RV pressure > 80% of RV/LV → VSD fenestration
- **Staged repair consists of**
  1) inducing growth of the central PAs with central shunt or RV-PA conduit
  2) staged thoracotomy-based unfocalization of MAPCAs
  3) RV-PA conduit with VSD closure

Repair Strategies

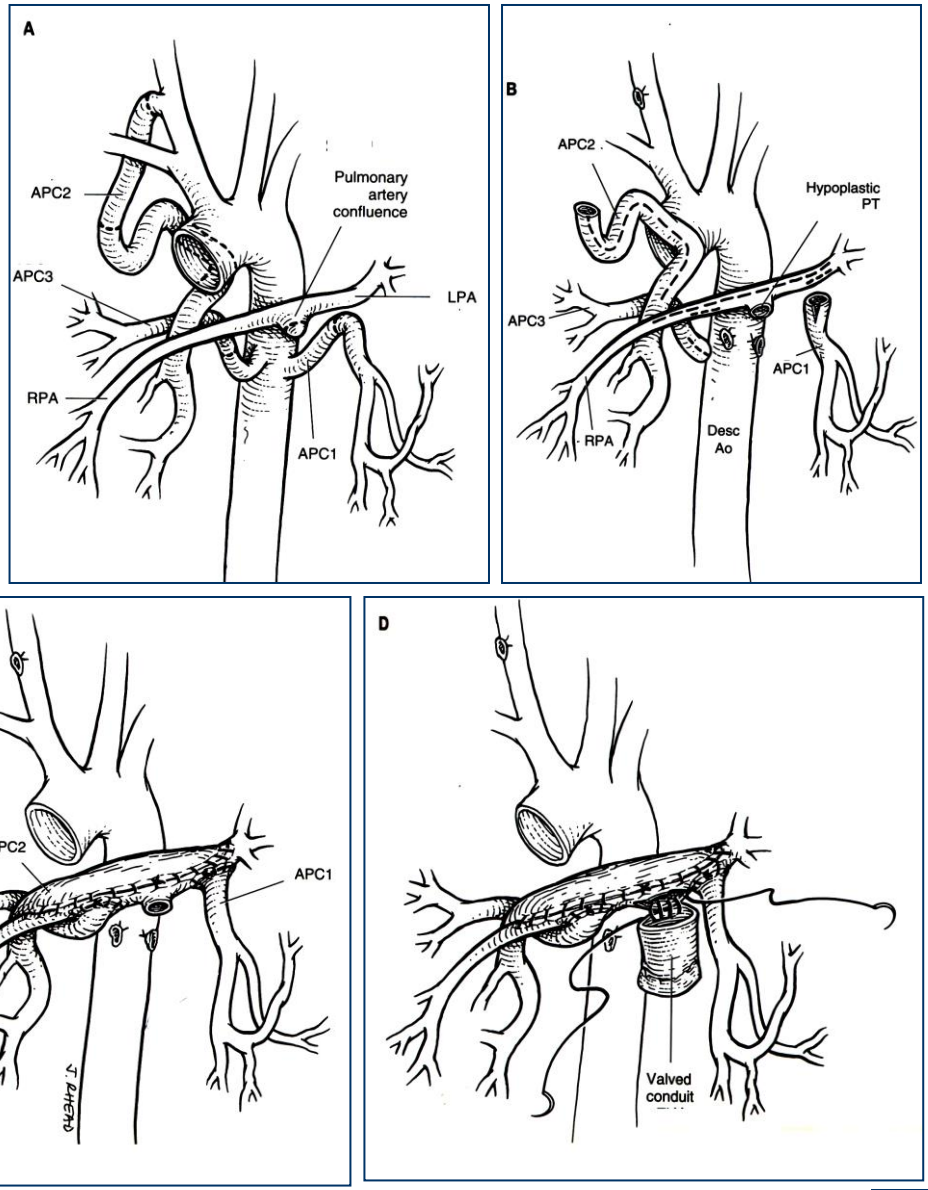
Rationale for early single stage unifocalization for TOF/PA/MAPCAs

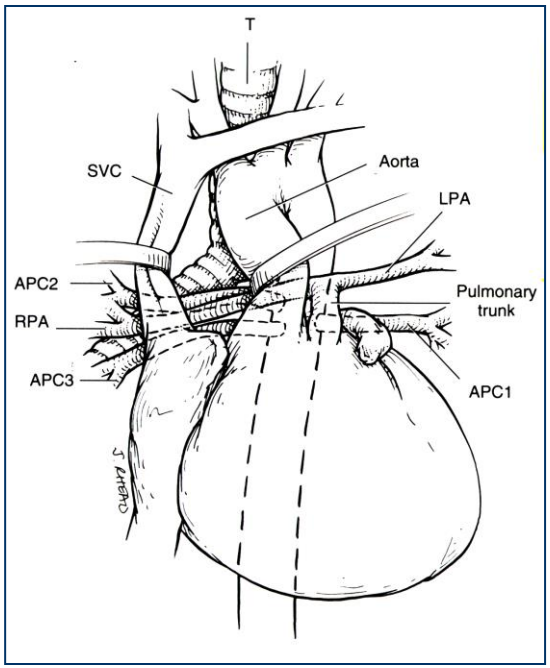
- **One can incorporate all segments of blood supply to the lung before stenosis develop in the MAPCAs and before potential changes of PH occur**
- **Preferred age for single staged repairs; 4 to 8 months of age (HSC)**
  - **Improved tolerance of long operations as compared to young infancy**
  - **Prior to the development of risks of PVOD**
  - **Prior to the development of MAPCA stenosis**

Rationale for staged unifocalization for TOF/PA/MAPCAs

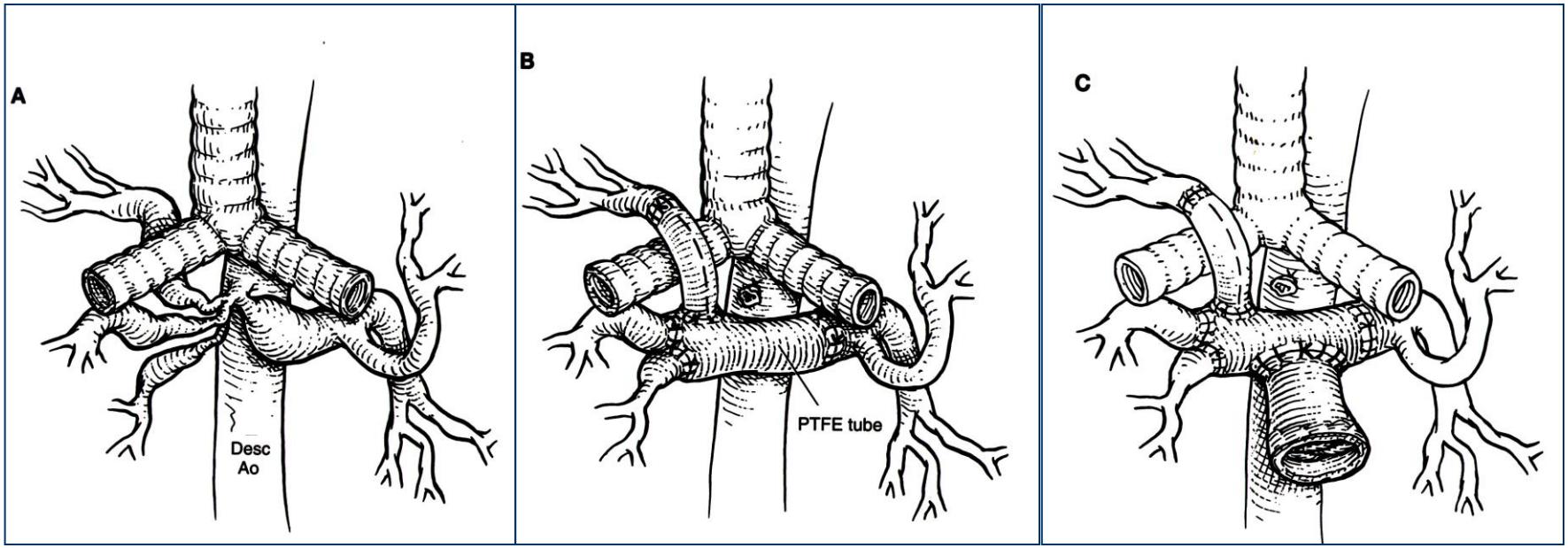
- **Small central PAs needs to be "rehabilitated" to normal size with shunt or RV to PA conduit**
- **To gain exposure to the distal MAPCA, hilar and intraparenchymal dissection is facilitated thu unilateral or staged bilateral thoracotomies**
- **Identification and mobilization of MAPCAs is much easier through posterolateral thoracotomy than a sternotomy approach**
- **Single stage unifocalization is a long and tedious procedure (very stressful to a child)**

Technique for single stage complete unifocalization



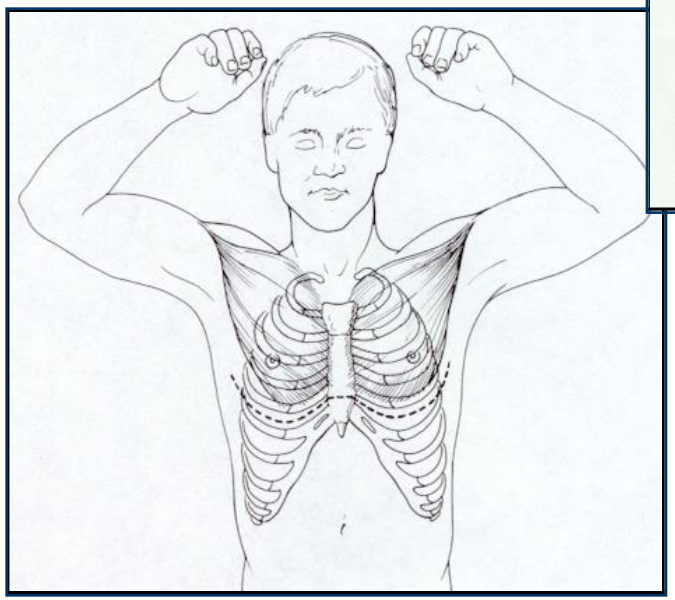


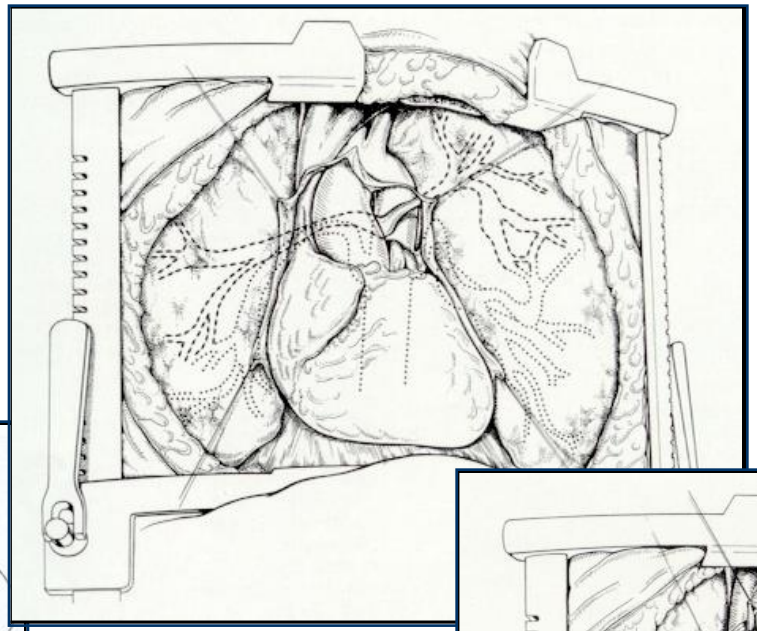
Technique for single stage complete unifocalization

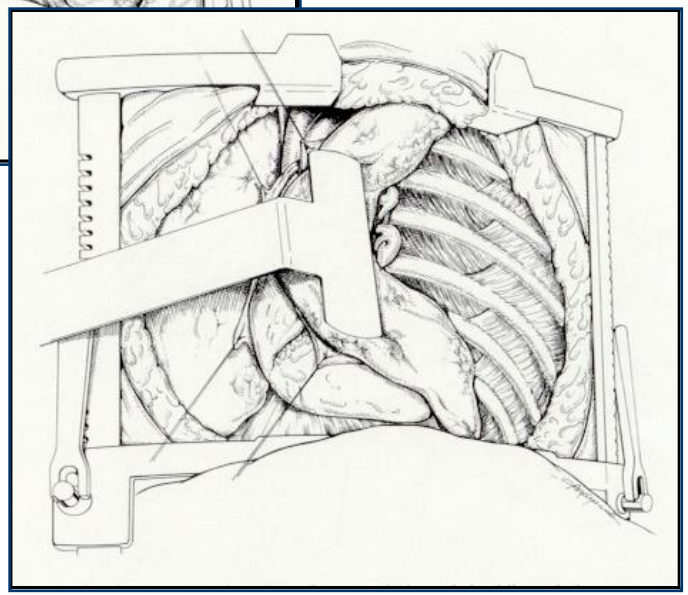


Technique for single stage complete unifocalization

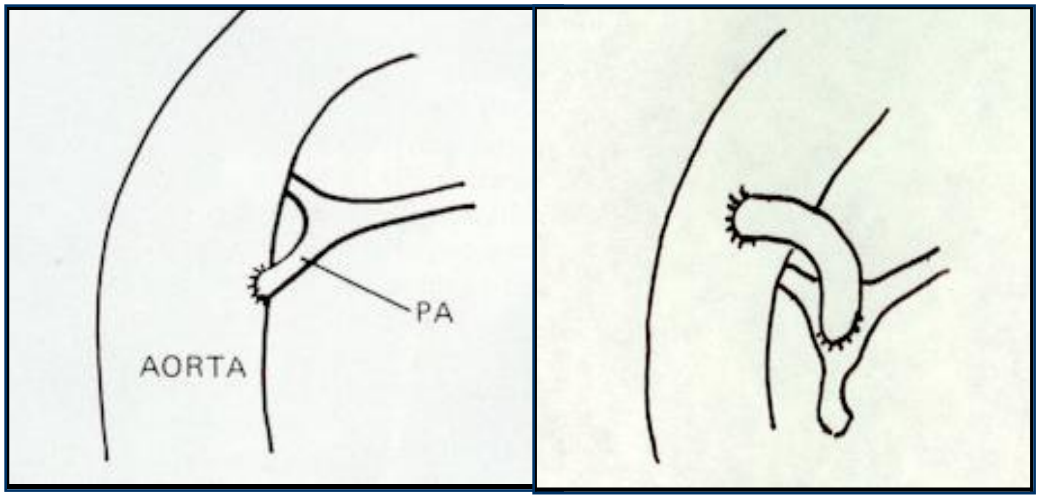
Clamshell Incision (bilateral thoracosternotomy)



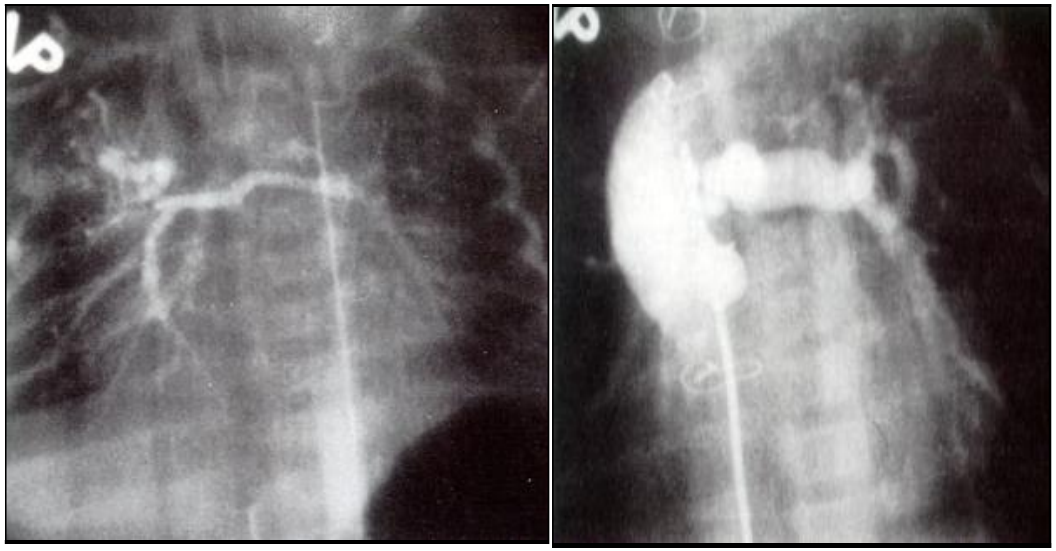




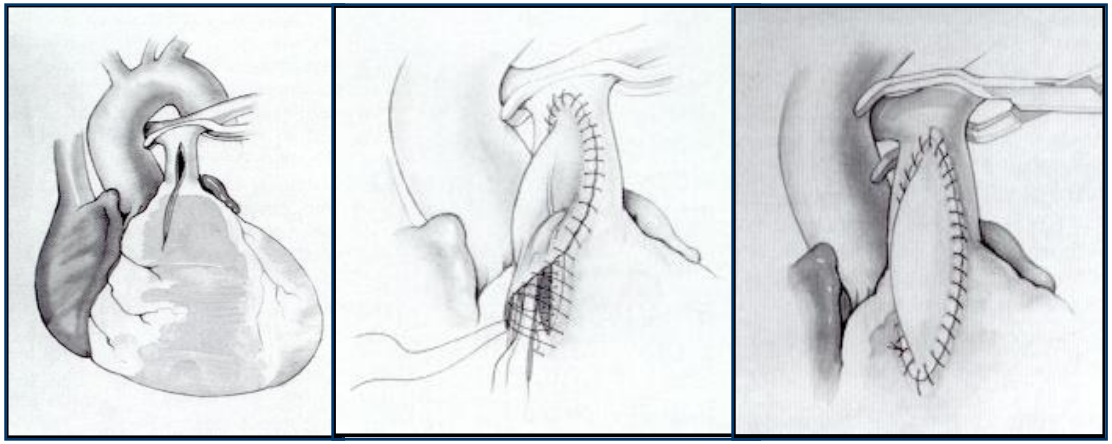
Technique for staged unifocalization



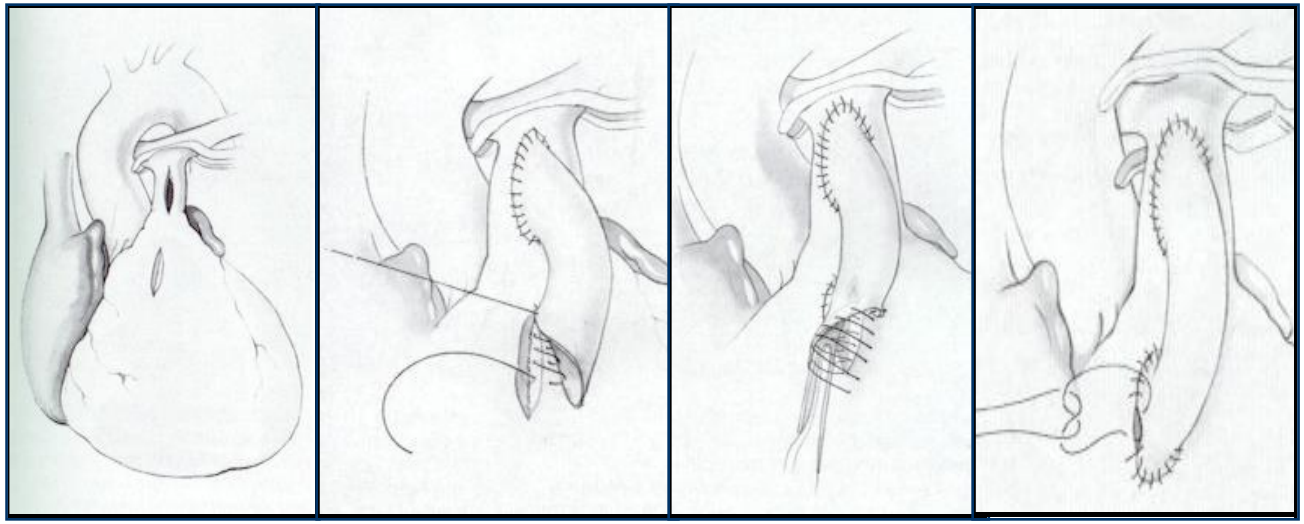
Central shunt



Technique for staged unifocalization



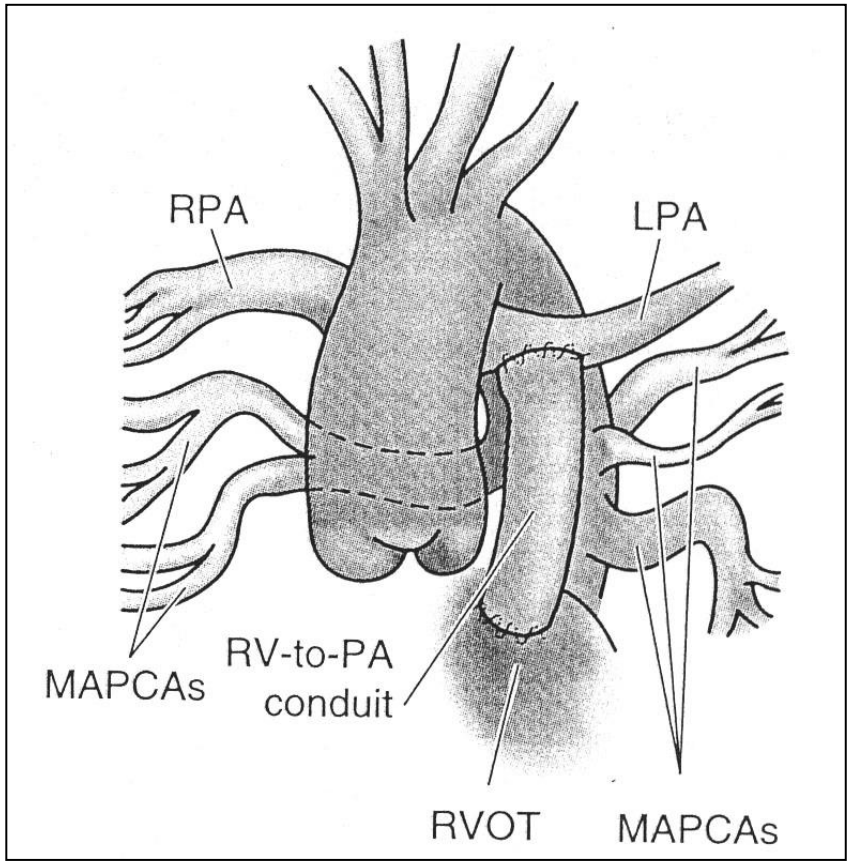
RVOT patch

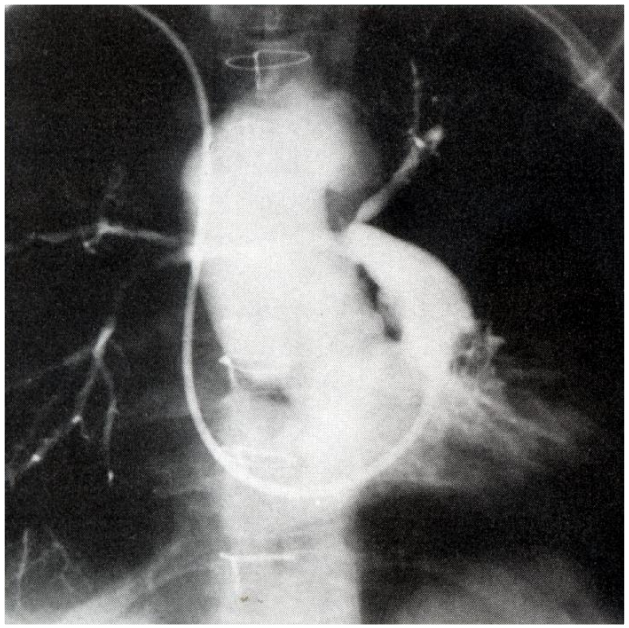


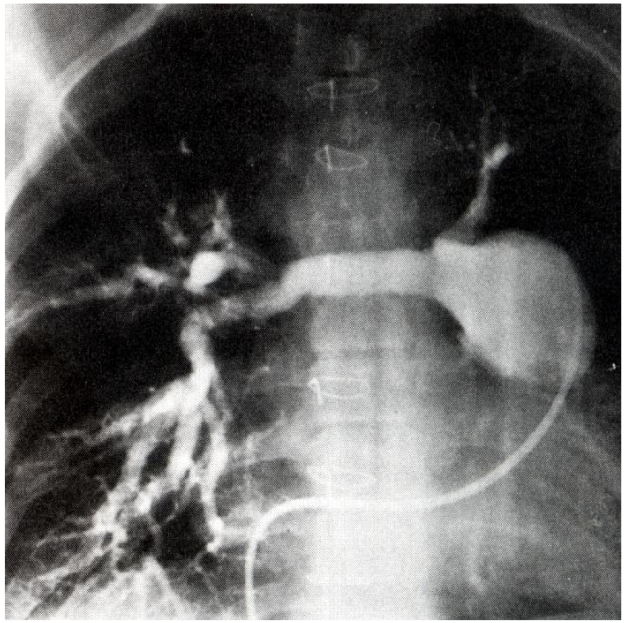
RV-PA conduit

Technique for staged unifocalization

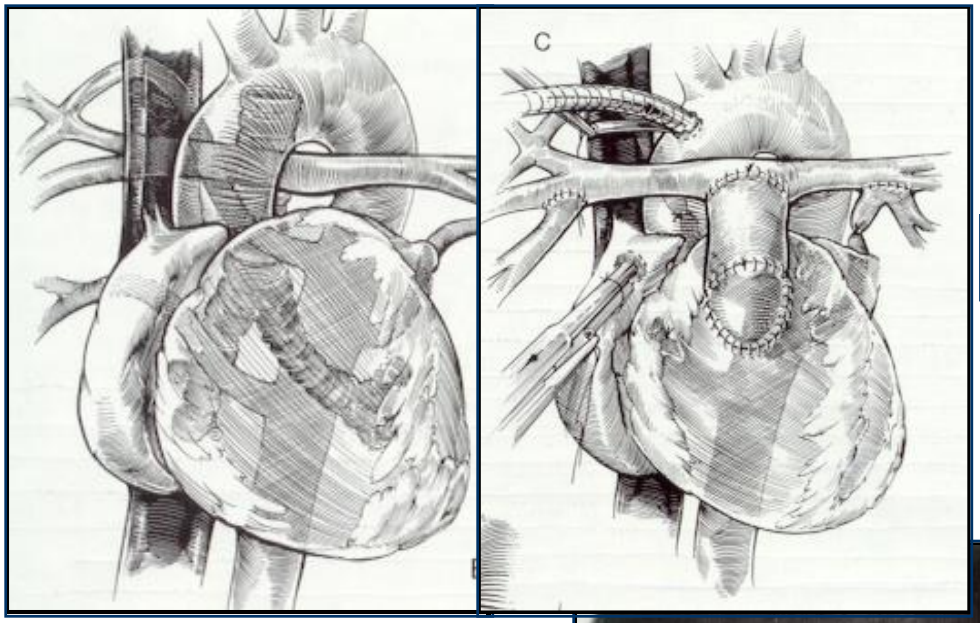
RV-PA conduit



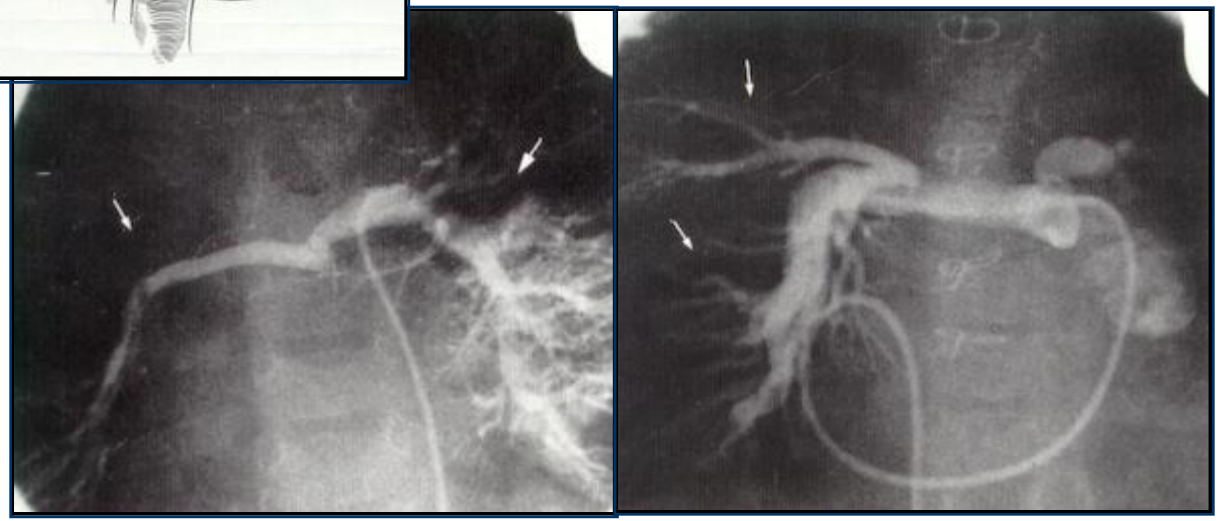




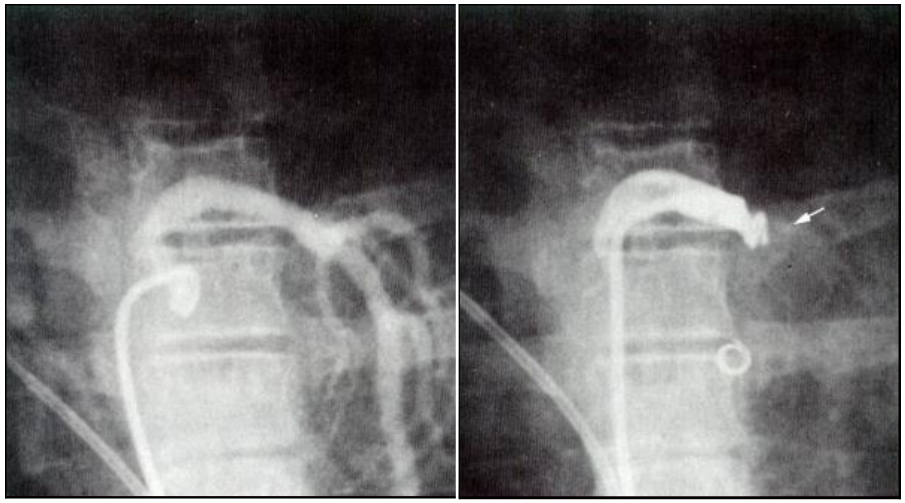
Technique for staged unifocalization



RV-PA conduit + Unifocalization

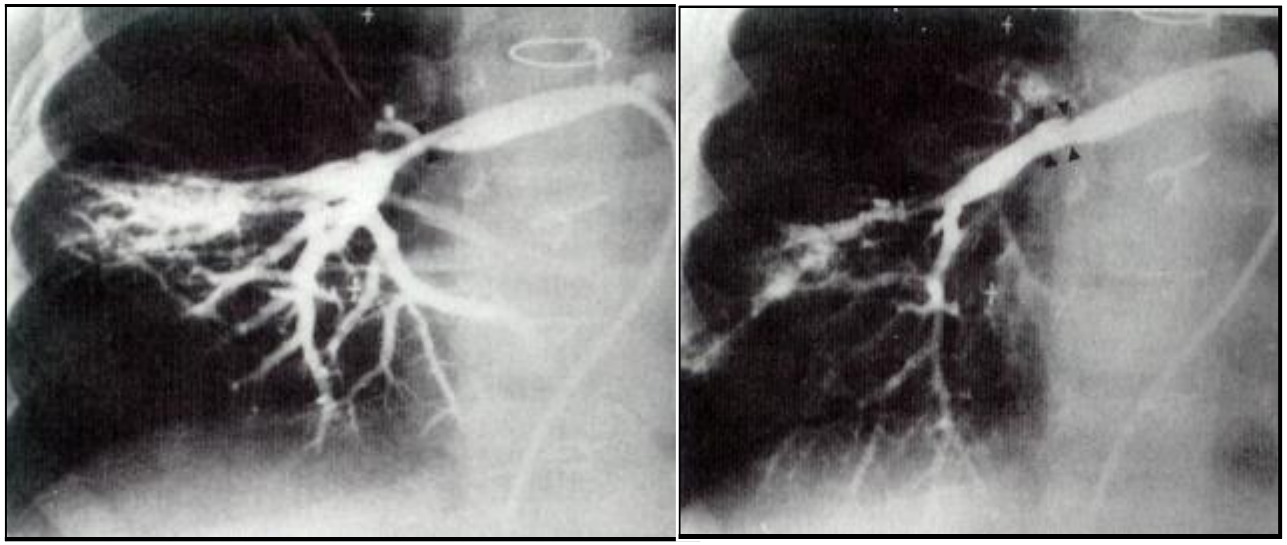


Technique for staged unifocalization



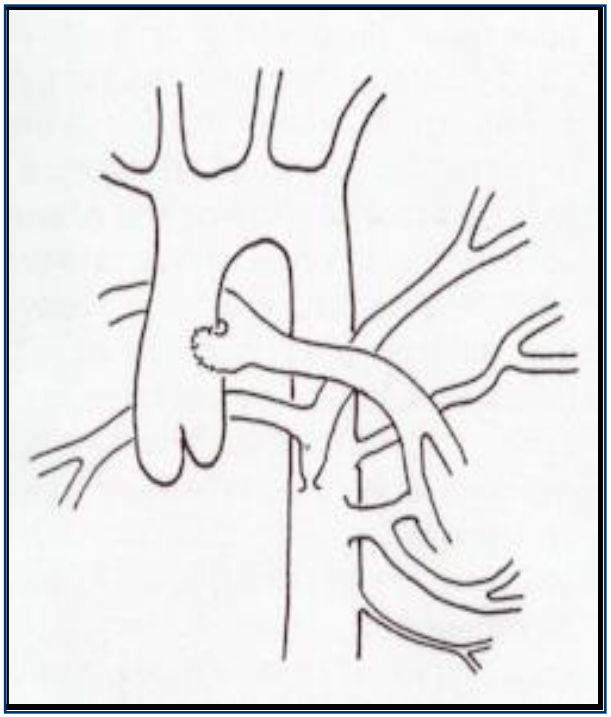
Coil embolization

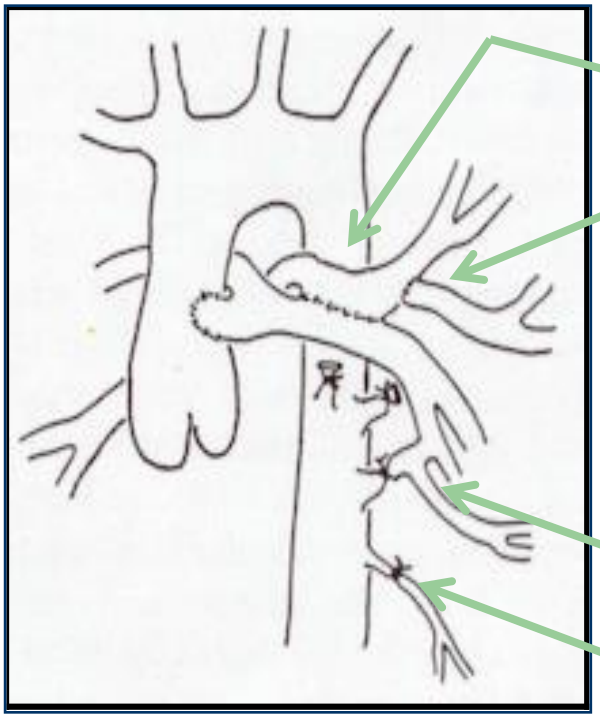
Balloon angioplasty



Technique for staged unifocalization

through bilateral thoracotomy





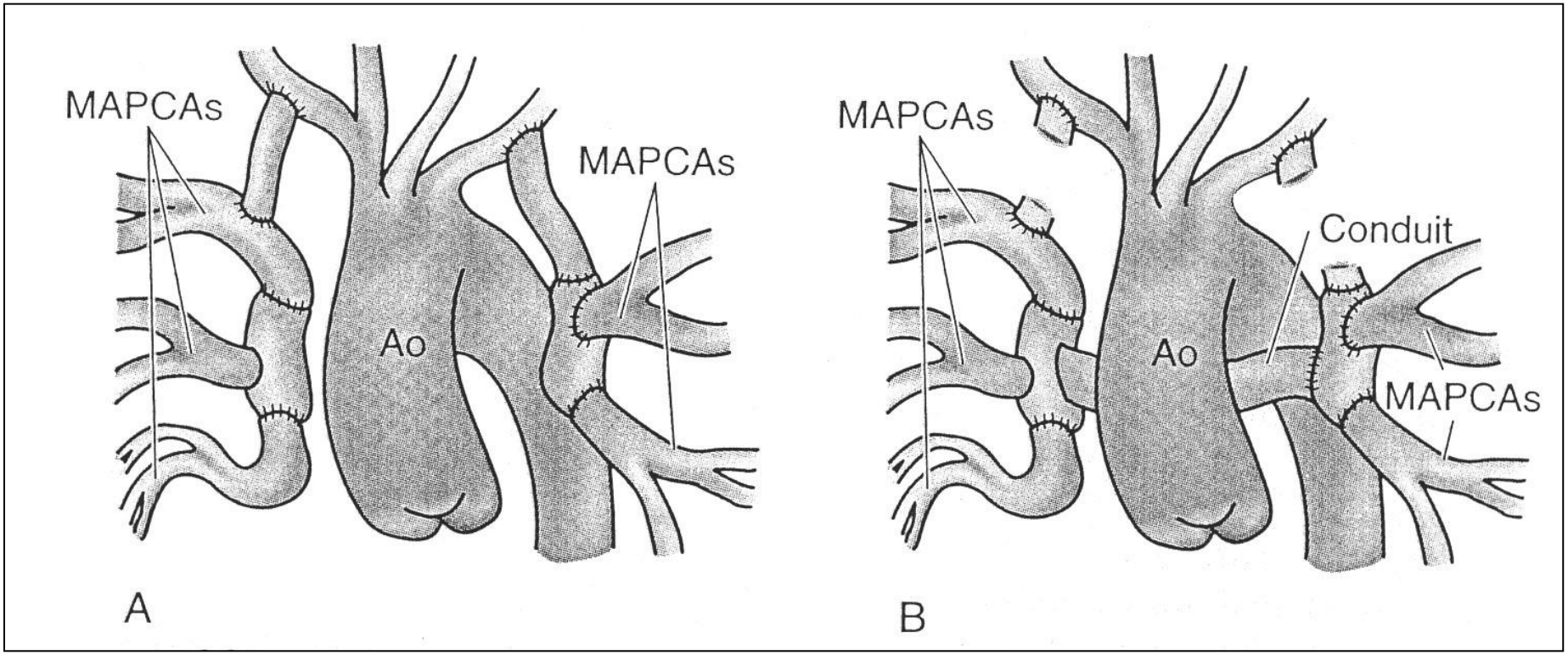
transplantation

graft interposition

ligation

Technique for staged unifocalization

through bilateral thoracotomy



Anatomic predictors of successful VSD closure

- Calculated pRV/LV ratio by the Birmingham formula < 0.7
- No sizeable MAPCAs remained
- More than 15 of lung segments connected to native PA
- PA index for all MAPCAs and central PAs (Total neopulmonary artery index) $\geq 200 mm^2/m^2$

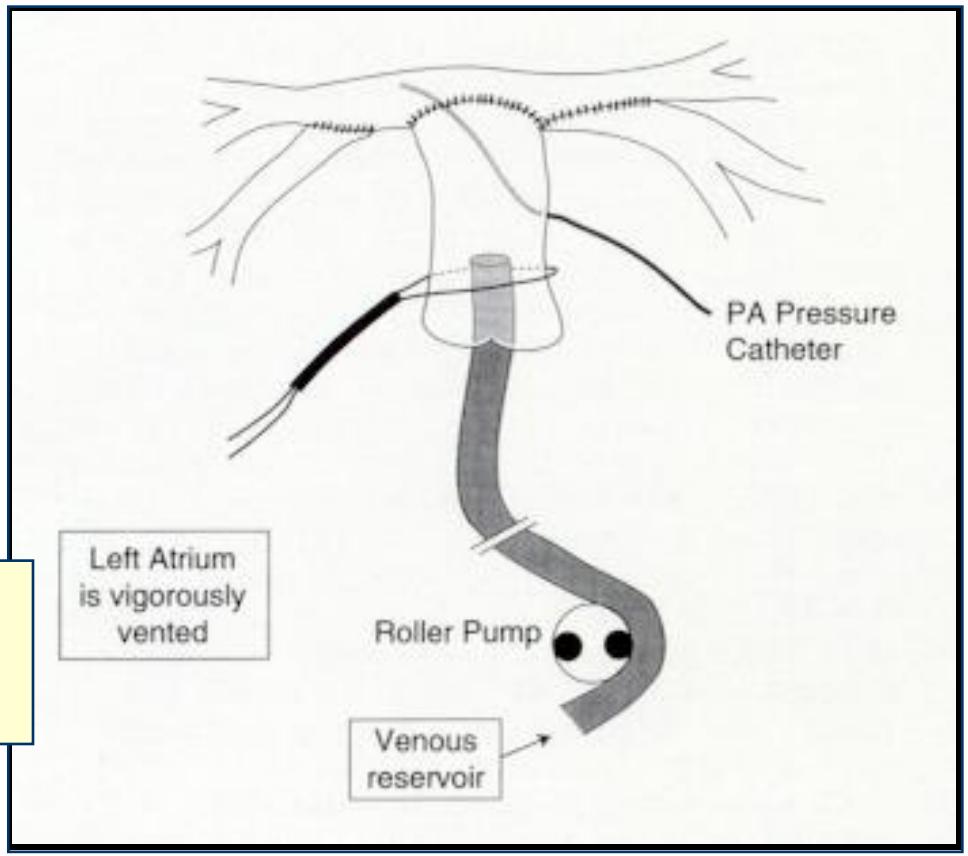
Functional predictors of successful VSD closure

- Net left-to-right shunt
- SpO_2 is typically in the high 80s or low 90s
- At a cardiac index of 2.5L/min/m^2 and PA presure of less than 30mmHg after unifocalization

VSD closure criteria by Hanley FL.

Intraoperative pulmonary flow study

≤ 25 mmHg at 2.5 L/min/m²



Results of Surgery for TOF/PA/MAPCAs

Table 22-1. Summary of surgical outcomes in patients with pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries

Authors (year)	Time period of operation	No. of patients	Strategy of unifocalization	Age at first operation	Early death	Follow-up duration	Late death	Outcomes at last follow-up
Reddy (2000)[71]	1992-1998	85	Single-stage	5m (10d – 37y)	9 (10.6%)	22m (1 – 69m)	7	84% 1YSR 74% 4YSR 93% total repair
Carotti (2003)[53]	1994-2002	37	Integrated approach	39m (22d – 13y)		43m (1 – 85m)		81% 7YSR 85% total repair
Gupta (2003)[65]	1983-2000	104	Staged	7d (3d – 22y)	11.5%	10.2y	5%	
Duncan (2003)[49]	1993-2001	46	Staged	7.2m (17d – 23y)	0	44m (1 – 79m)	1 (2.2%)	61% total repair
d'Udekem (2005)[69]	1975-1995	82	Staged	1.4y (7d – 34y)	4% + 8%	14.2y (3m – 25y)	9	51% 12YSR (total repair) 65% total repair
Ishibashi (2007)[50]	1982-2004	113	Staged	6.3y (1.1m – 34y)		8.8y (0.8 – 23.3y)		80.9% 5YSR 73.8% 10YSR 80.5% total repair
Davies (2009)[54]	1989-2008	216	Staged/ Single	2y	6%	2.3y	6%	89% 3YSR 73% total repair
Honjo (2009)[67]	2003-2008	20	Single-stage	7.7m (2-197m)	0	31m (8-66m)	5%	94% 1YSR 95% total repair
Malhotra (2009)[38]	1992-2007	462	Single-stage	7.7m (10d – 39y)	5.9%	NR	NR	86% 5YSR 90% total repair

* d: days, m: months, y: years, YSR: year survival rate

Result of staged repair of TOF/PA/MAPCAs

(from The Children's Hospital at Cleveland Clinic Foundation)

- 1993 – 2001, 46 patients
- 28 (61%) underwent complete repair with VSD closure
  median number of operations ; 3 (1 - 6)
- Mean PA pr/ mean systemic pr = 0.36 (0.19 – 0.58)
- 2 (7.1%) required VSD fenestration
- 18 (39%) underwent 1 or more staging operations
  (good candidates for complete repair)
- No hospital mortality
- 1 late death

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