Oklahoma Heart Institute

Adult congenital heart surgery

Ajit K. Tharakan, MD

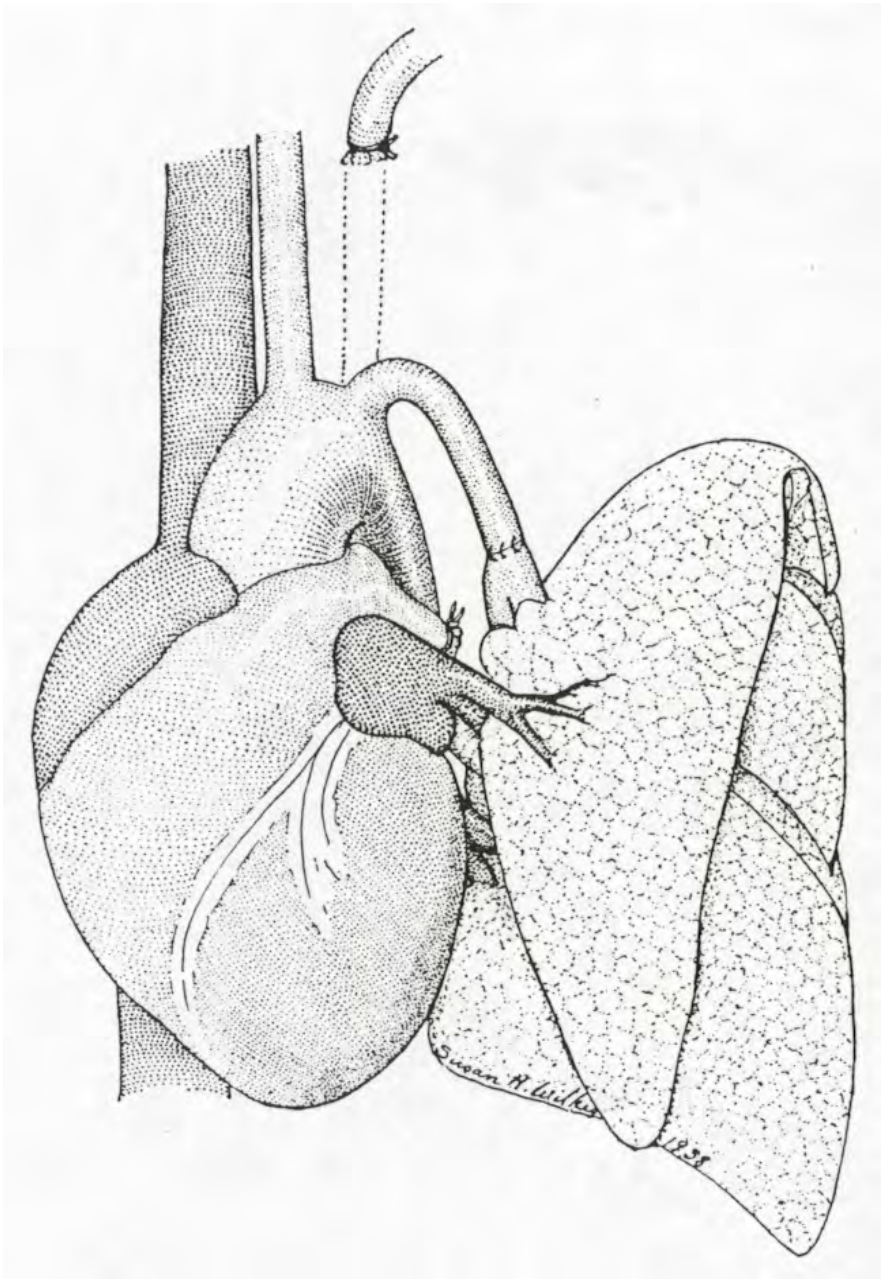
Cardiothoracic Surgery

No conflict of interest

- Evolution of surgery for CHD
- Bethesda classification- ACHD
- Common pathology seen in adults
- Surgical cases

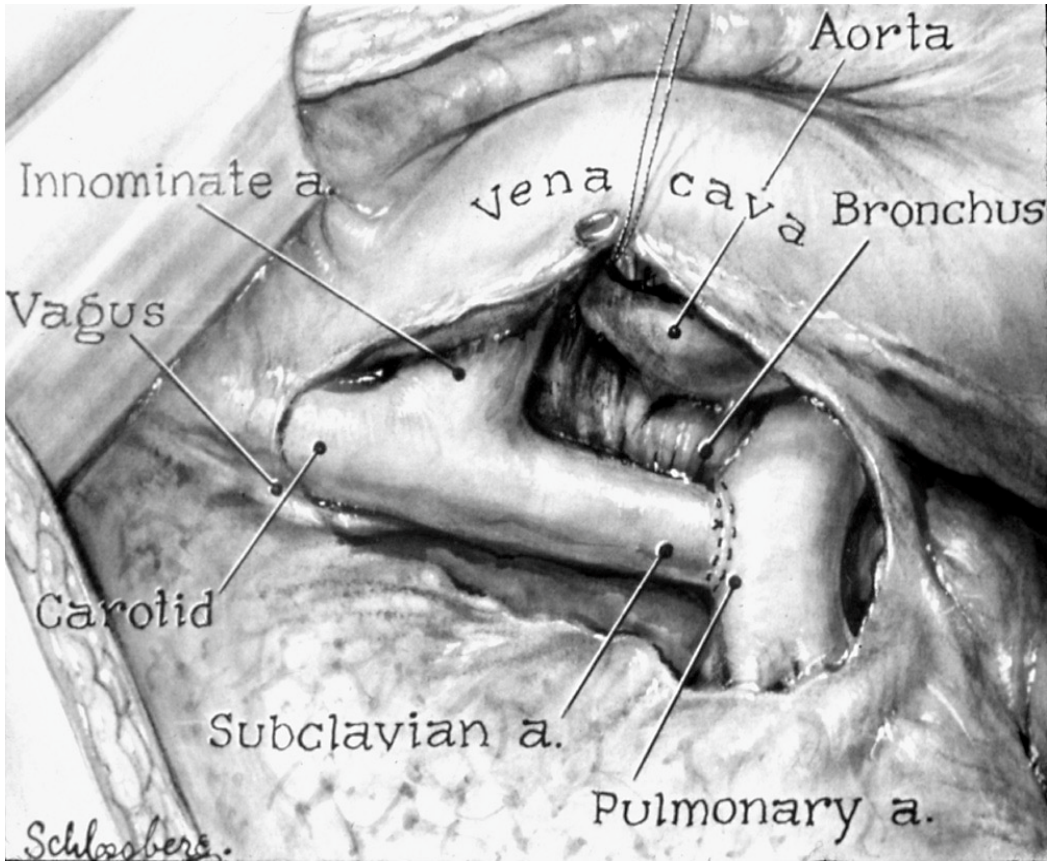
The treatment of congenital heart disease is largely hygienic. Fresh air, avoidance of cold and of all conditions liable to induce bronchial irritation and a carefully regulated diet are the most essential elements… Bloodletting, under these circumstances, might be freely employed.

Sir William Osler's Collected Papers on the Cardiovascular System, 1889



Levy SE, Blalock A.
Experimental Observations on the Effects of Connecting by Suture the Left Main Pulmonary Artery to the Systemic Circulation. **The Journal of Thoracic Surgery 1939;8:525.**

Blalock Taussig Shunt



Eileen Saxon, the frail, 15-month-old child who made surgical history as the first patient to undergo the Blue Baby operation.

TABLE 3 Clinical Experience by J. H. Gibbon, Jr., with Total Cardiopulmonary Bypass Using his Screen Oxygenator

Patient	*Age*	*Date of Surgery*	*Diagnosis Preoperatively*	*Postoperative Results*
1	15 mo	February, 1952	ASD	Had PDA (only); died in OR
2	18 yr	May 6, 1953	ASD	Lived (long term)
3	5.5 yr	July 1953	ASD	Died in OR
4	5.5 yr	July 1953	ASD	Had also VSD, PDA; died in OR
5	Data NA	Data NA	Data NA	Died
6	Data NA	Data NA	Data NA	Died

ASD, atrial septal defect; PDA, patent ductus arteriosus; OR, operating room; VSD, ventricular septal defect; NA, not available. From Gibbon JH Jr. Application of a mechanical heart and lung apparatus to cardiac surgery. *Minn Med* 1954;37:171–185; Time News Magazine. *Historic operation*. 1953:70.

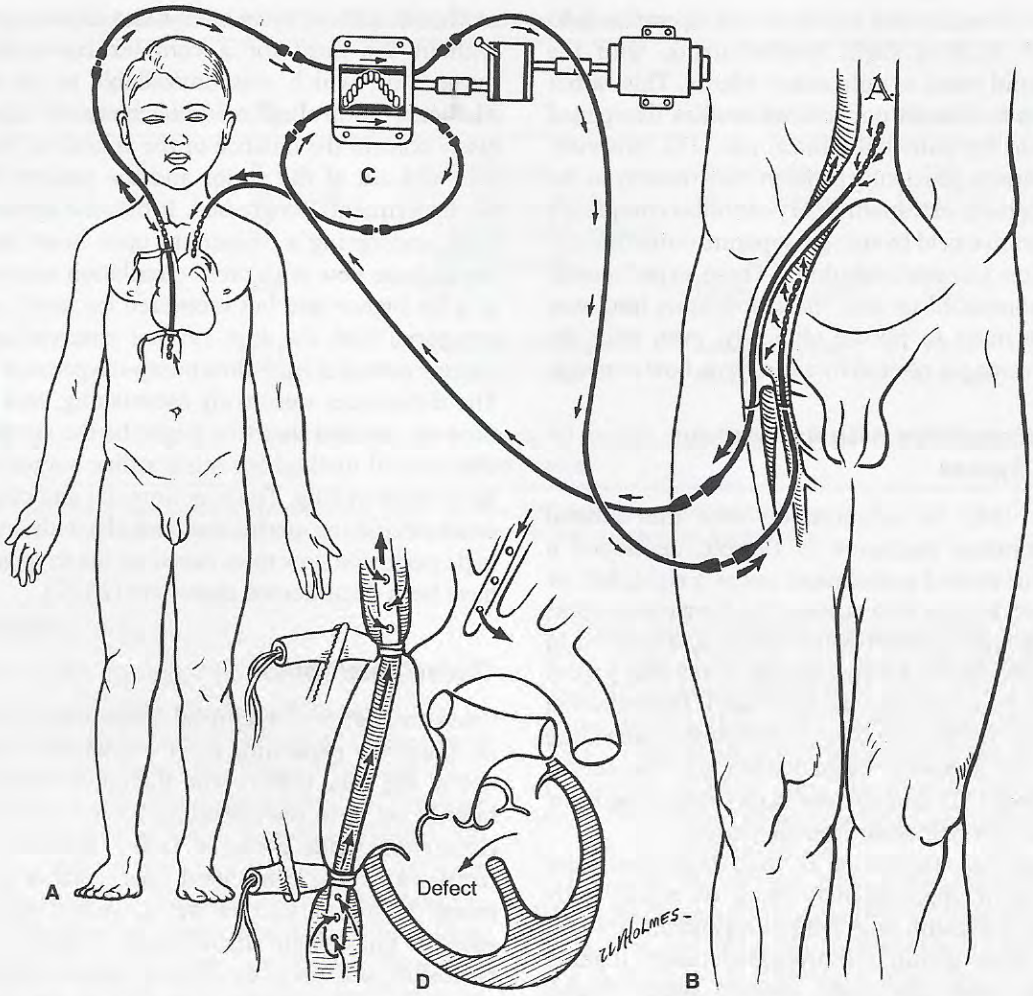
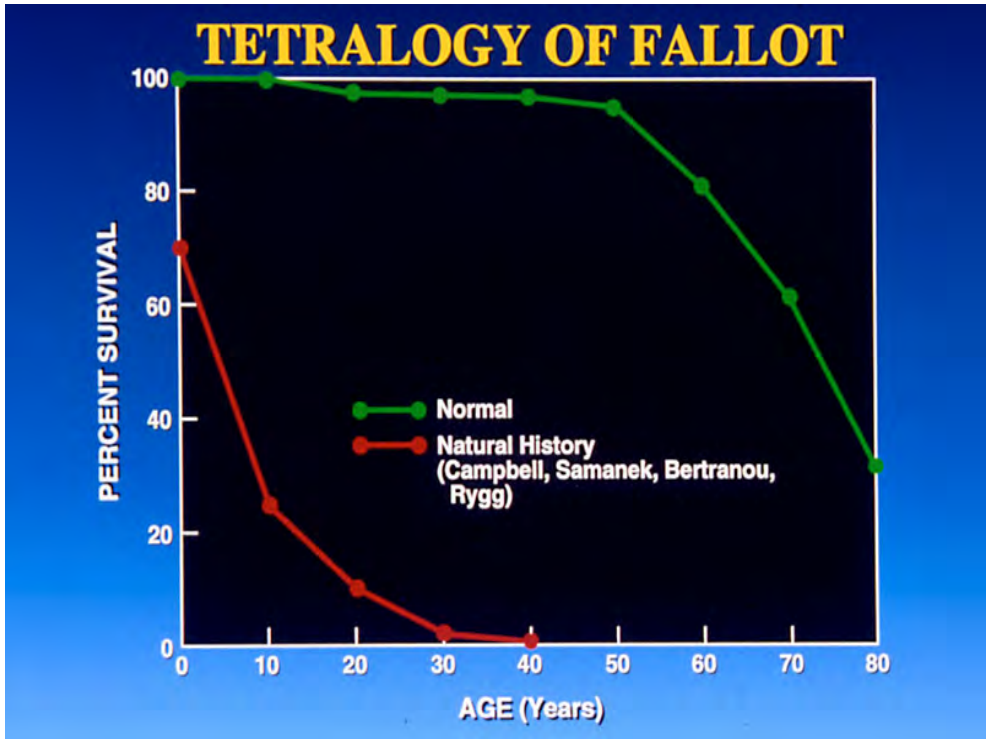


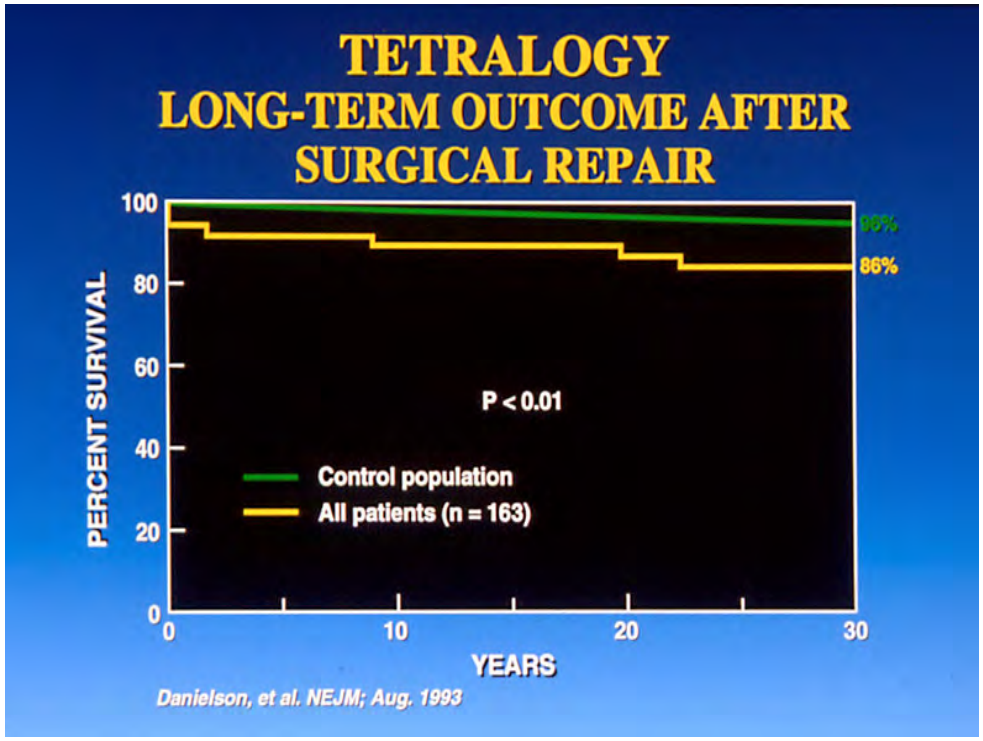
FIGURE 2. Direct-vision intracardiac surgery using extracorporeal circulation by means of controlled cross-circulation. **A:** The patient, showing sites of arterial and venous cannulations. **B:** The donor, showing sites of arterial and venous (superficial femoral and great saphenous) cannulations. **C:** The single Sigmamotor pump controlling precisely the reciprocal exchange of blood between the patient and donor. **D:** Close-up of the patient's heart, showing the vena caval catheter positioned to draw venous blood from both the superior and inferior venae cavae during the cardiac bypass interval. The arterial blood from the donor was circulated to the patient's body through the catheter that was inserted into the left subclavian artery.

TABLE 6 Results of Direct-Vision Intracardiac Surgeries with Cardiopulmonary Bypass by Cross-Circulation in 45 Patients from March 26, 1954, to July 9, 1955[a]

Abnormality	*Corrective Operations*	*Patients*	*Mortality*	
			Hospital	*Late (After 30 yr)*
VSD	Suture closure	27	8	2
PDA (with severe pulmonary hypertension)	Exploratory ventriculotomy, division of ductus	1	0	0
Tetralogy of Fallot	Closure of VSD, correction of infundibular/valvularpulmonary stenosis	10	5	3
Atrioventricularis communis	Closure of ostium primum, VSD; repair of valvular deformities	5	3	1
Isolated infundibular pulmonary stenosis	Resection of infundibulum	1	0	0
Pulmonary stenosis, ASD, anomalous pulmonary venous return	Pulmonary valvotomy, ventricular and atrial cardiotomies, transposition of anomalous pulmonary veins, closure of septal defects	1	1	0
Total		45	17	6

[a]Cross-circulation was used exclusively from its inception through February 1955. Beginning March 1, 1955, other bypass methods (bubble oxygenator, dog-lung oxygenator, arterial reservoir) were used for lower-risk patients. Cross-circulation was reserved for high-risk patients. By July 1955, the bubble oxygenator had become the sole method. VSD, ventricular septal defect; PDA, patent ductus arteriosus; ASD, atrial (secundum) septal defect.


TETRALOGY OF FALLOT
PERCENT SURVIVAL
100
80
60
40
20
0
0
10
20
30
40
50
60
70
80
AGE (Years)
Normal
Natural History
(Campbell, Samanek, Bertranou, Rygg)


TETRALOGY
LONG-TERM OUTCOME AFTER SURGICAL REPAIR
PERCENT SURVIVAL
100
80
60
40
20
0
0
10
20
30
YEARS
P < 0.01
Control population
All patients (n = 163)
90%
86%
Danielson, et al. NEJM; Aug. 1993

Congenital Heart Disease

- Most common birth defect ~ 8 per 1000
- 90% survive to adulthood
- Atleast 800,000 adults in the US
  - 47% simple defects
  - 38% complex defects
  - 15% very complex defects
- Only a fraction have appropriate cardiology f/u
- Most commonly present in adulthood with arrhythmias and CHF

Adult congenital heart disease

(Bethesda Conference classification)

- Simple defects
  - Isolated secundum ASD's, VSD's
  - Isolated valve disease (pulmonary / aortic stenosis)
  - Ligated/ occluded PDA
  - Repaired ASD/ VSD
- Recommend evaluation "atleast once" in a adult congenital heart center.

Adult congenital heart disease

(Bethesda Conference classification)

- Moderate defects
  - PAPVC
  - AV canal defects
  - Coarctation of aorta
  - Ebstein's anomaly
  - Primum ASD, sinus venosus ASD
  - Subvalvular or supravalvular aortic stenosis
  - Moderate or severe pulmonary stenosis/ regurgitation
  - Tetralogy of Fallot
  - Complicated VSD

Adult congenital heart disease

(Bethesda Conference classification)

- Complex disease
  - Cyanotic lesions
  - Double outlet ventricle
  - Eisenmenger's syndrome
  - Fontan Procedure
  - Single ventricle
  - Pulmonary vascular obstructive disease
  - TGA
  - Truncus

General Management

- Preoperative evaluation
  - Natural history
  - Sequelae of previous interventions
  - Development of acquired diseases
  - Development of CAD
  - TEE/ TTE
  - Cardiac catheterisation

Procedures requiring reoperation

- Redo median sternotomy
  - CT scan to assess adherance of vascular structures to the back of the sternum
  - Femoral cannulation
  - Need for DHCA
  - Use of goretex or pericardial patch
- Bleeding
  - Platelet dysfunction, hepatic congestion
  - Use of autologous blood, antifibrinolytics

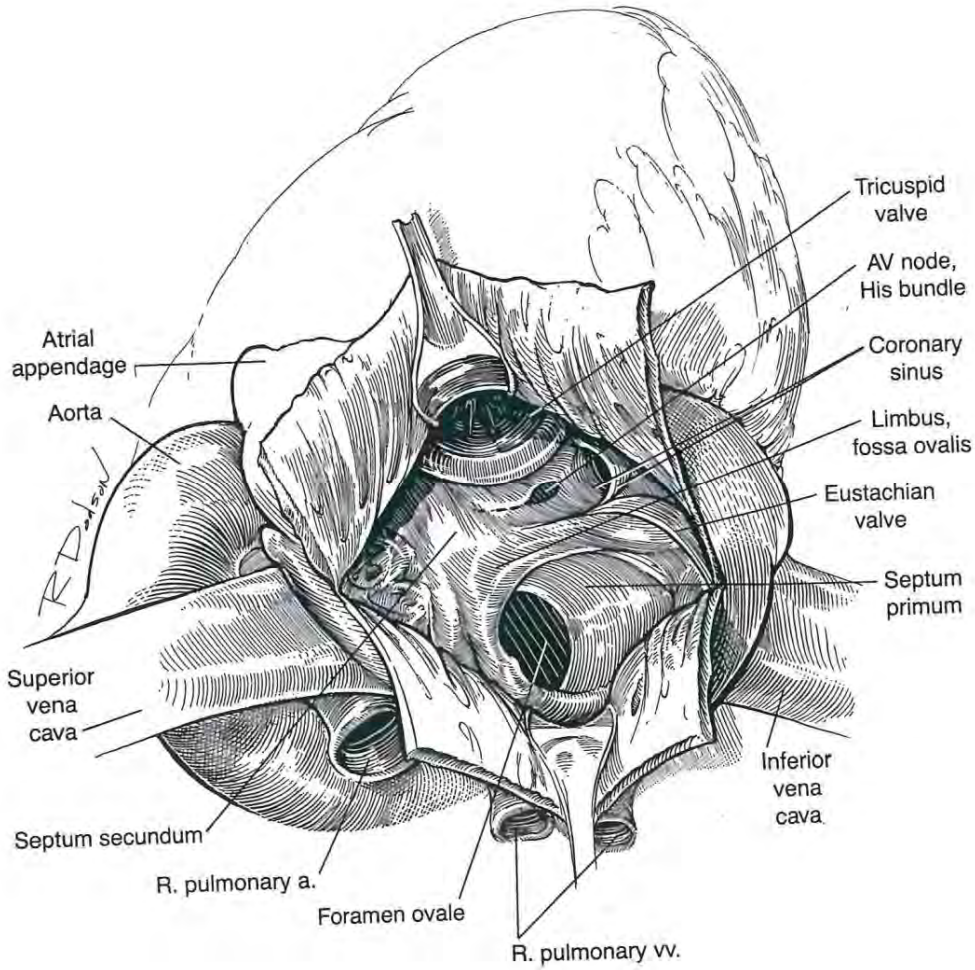
Myocardial Protection

- Right and left ventricular hypertrophy, dysfunction and dilatation
- Noncoronary/ bronchial collateral flow
- Need for LV vent, deep hypothermia and frequent antegrade and retrograde cardioplegia

Post operative care

- More significant post operative myocardial dysfunction
- Need for direct LA line

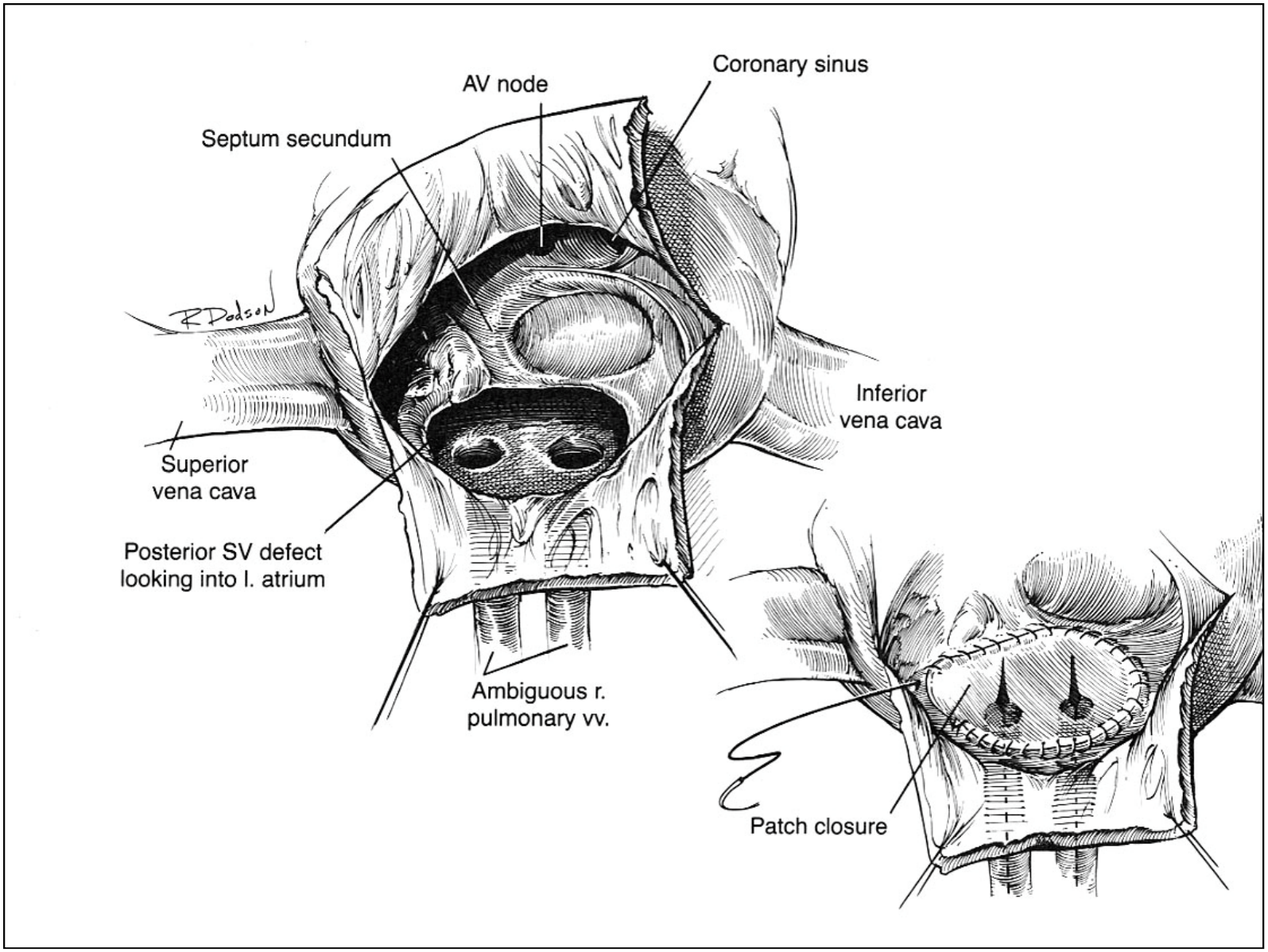
ASD


Tricuspid valve
AV node, His bundle
Coronary sinus
Limbus, fossa ovalis
Eustachian valve
Septum primum
Inferior vena cava
Atrial appendage
Aorta
Superior vena cava
Septum secundum
R. pulmonary a.
Foramen ovale
R. pulmonary vv.

Drainage of at least one pulmonary vein drains to the right atrium, SVC or IVC

Operative Procedure

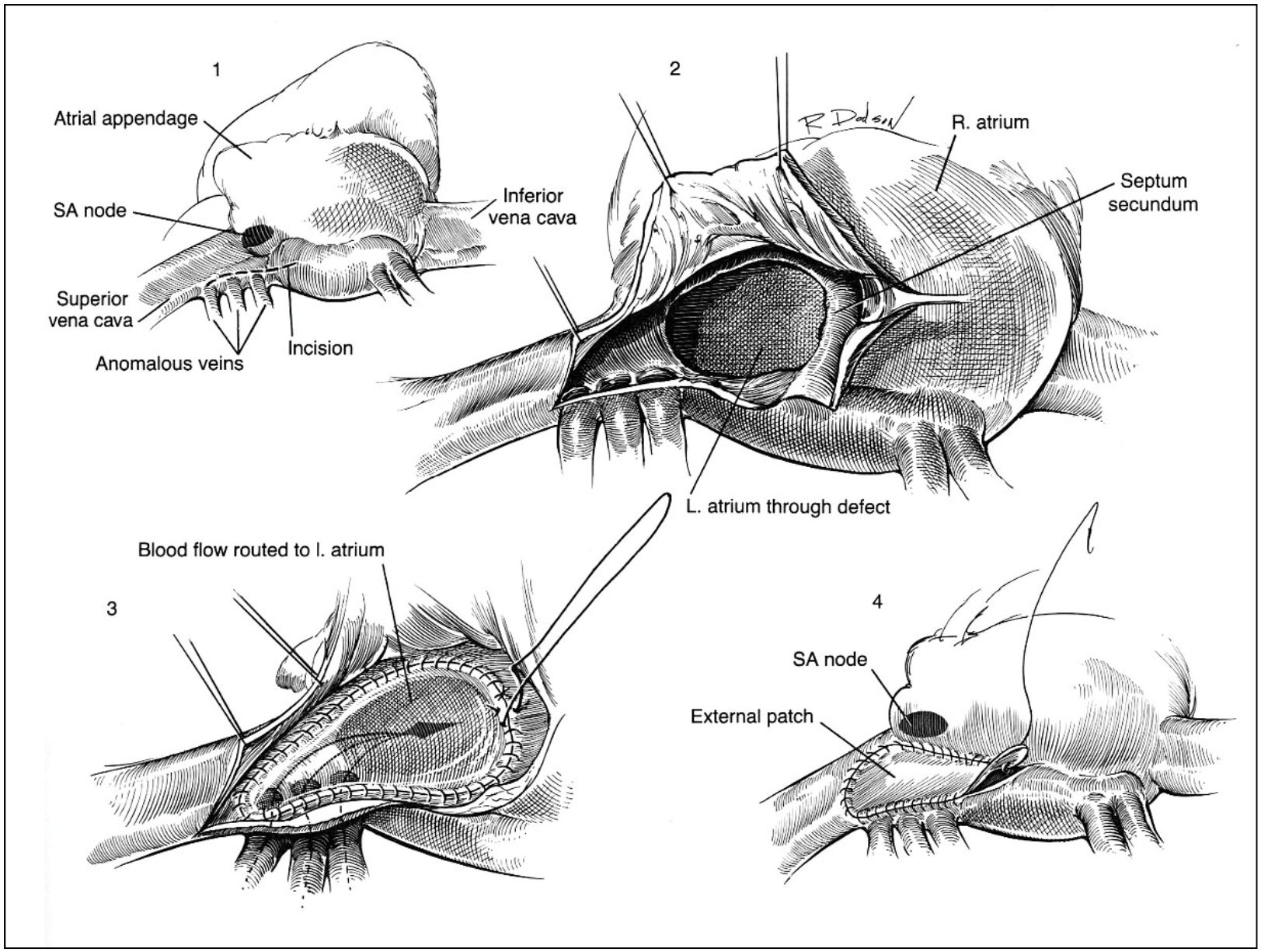
Kirklin, Ellis and Wood - 1965


Coronary sinus
AV node
Septum secundum
R Dodson
Inferior
vena cava
Superior
vena cava
Posterior SV defect
looking into l. atrium
Ambiguous r.
pulmonary vv.
Patch closure

The Problem

Baffle diversion of the PAPVR and ASD closure can obstruct the SVC

Schuster, Gross and Colodny - 1962


1
Atrial appendage
SA node
Superior vena cava
Anomalous veins
Incision
Inferior vena cava
2
R. atrium
Septum secundum
L. atrium through defect
3
Blood flow routed to l. atrium
4
SA node
External patch

Atriocavoplasty
Late Sinus Node Dysfunction

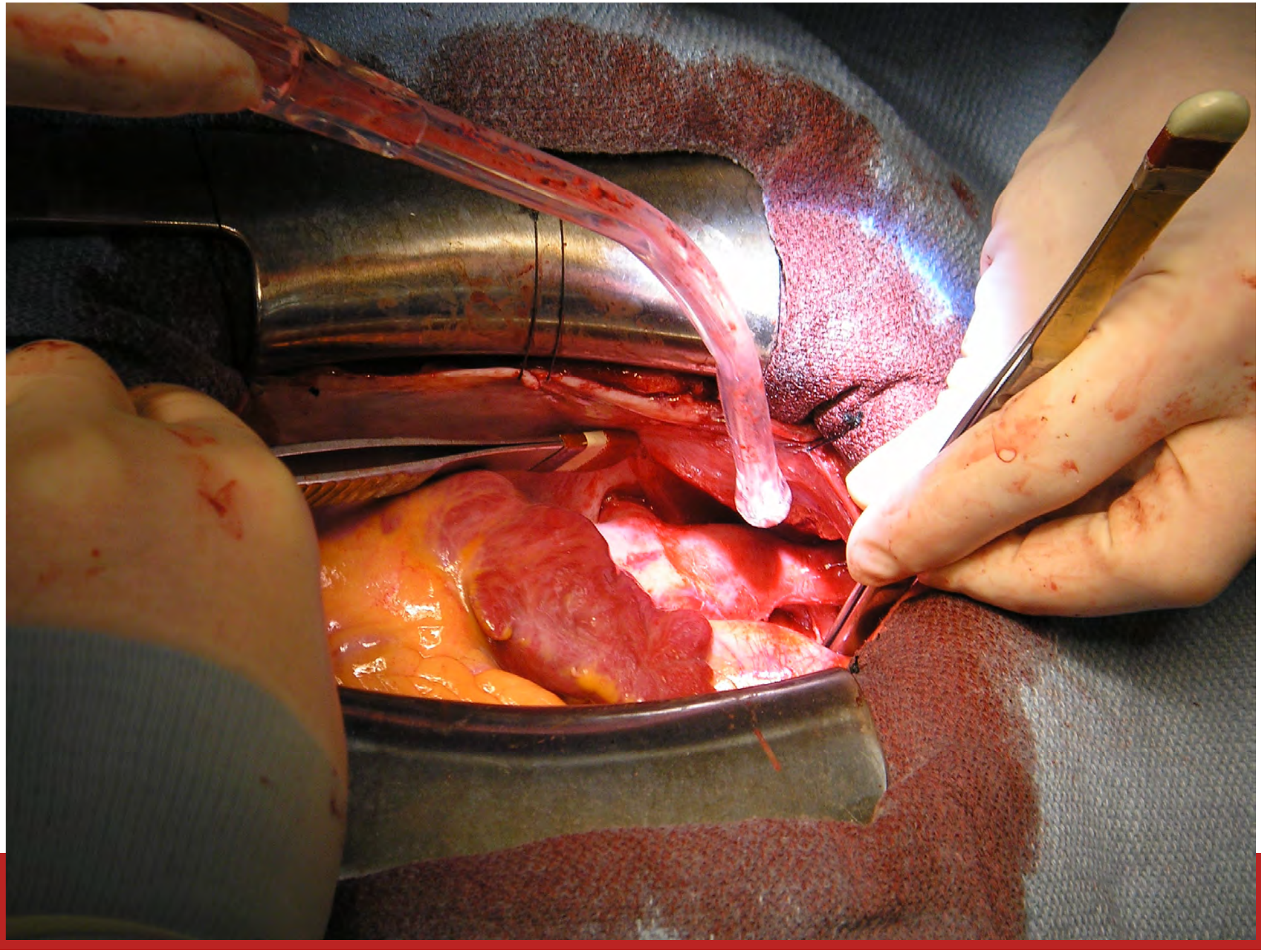
- Agrawal - 1997 3/27 (11%)
- Pathi et al - 1995 0/6 (0%)
- DeLeon et al - 1993 3/23 (13%)
- Cooley et al - 1991 2/6 (33%)

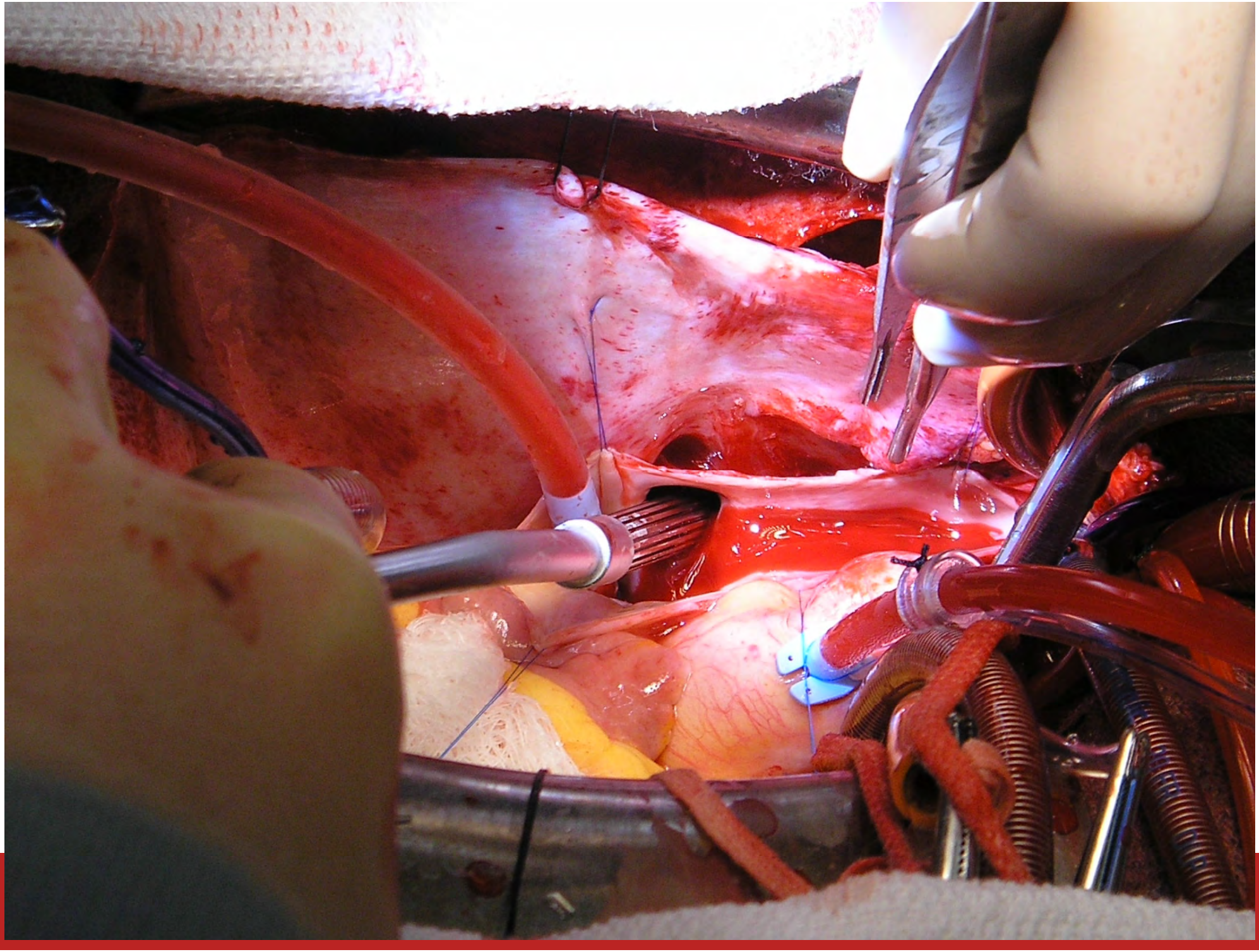
History & Physical

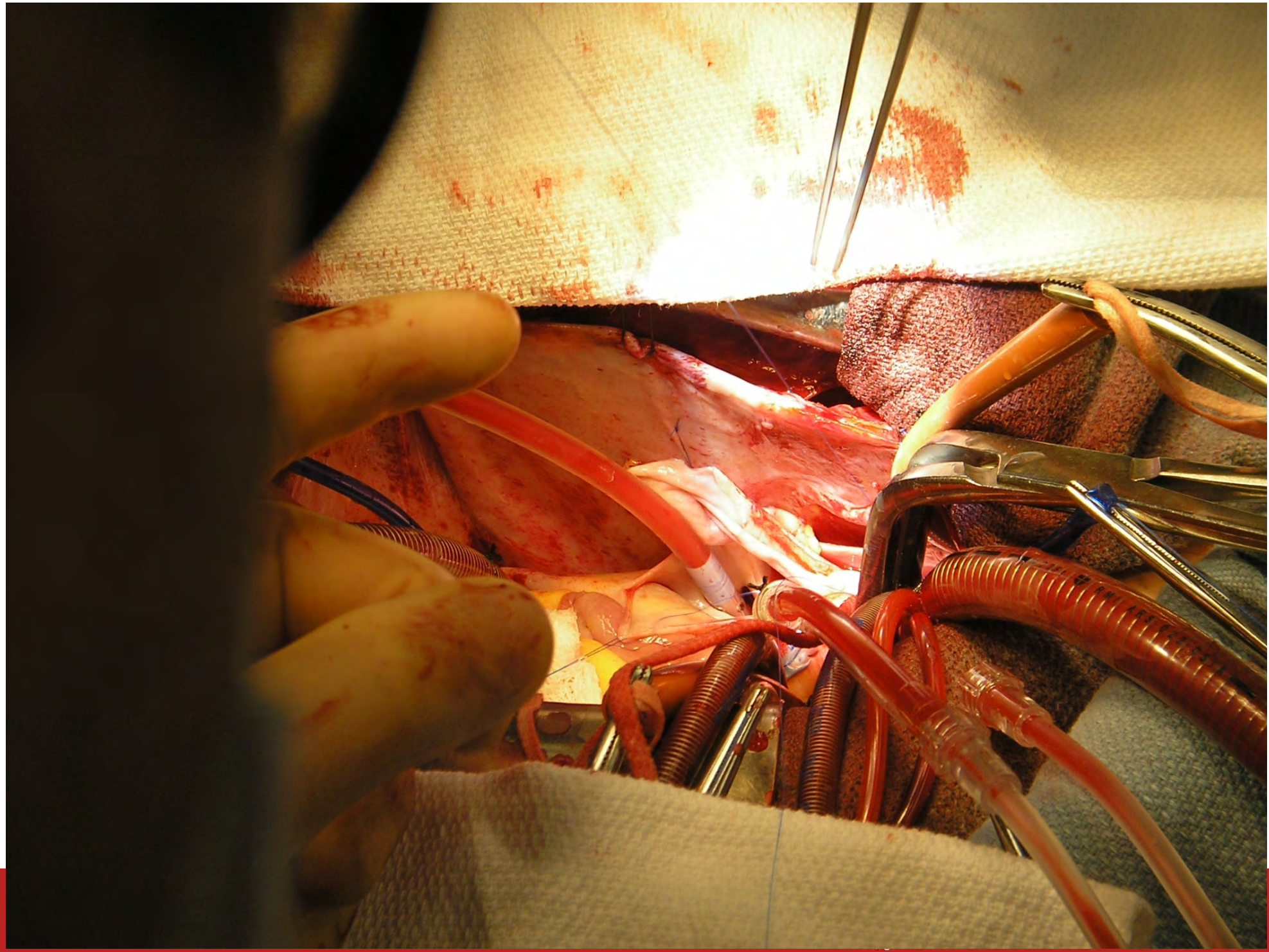
- 66 yr old female
- Murmur since childhood- asymptomatic
- Recent dry cough
- Abnormal CXR

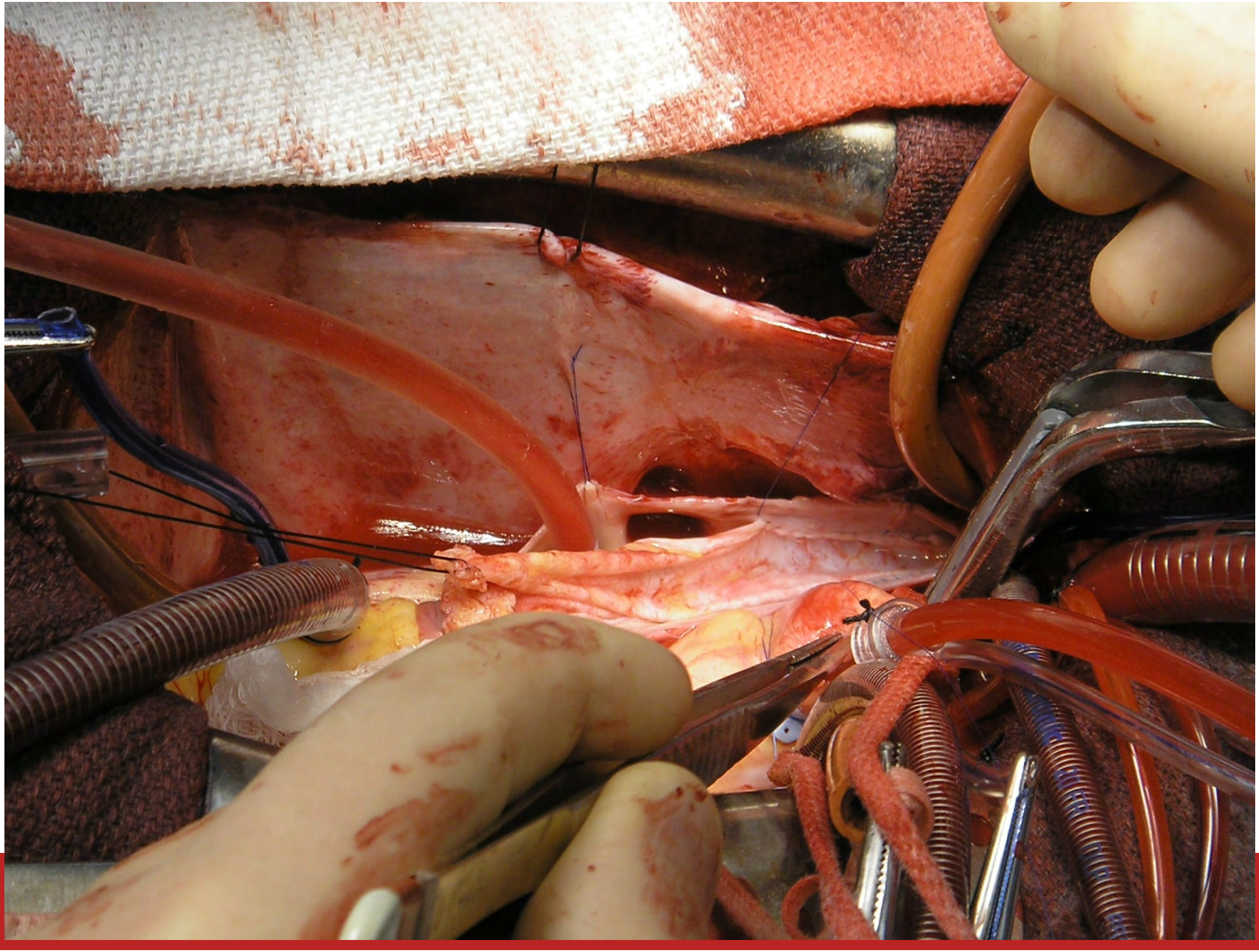
TEE

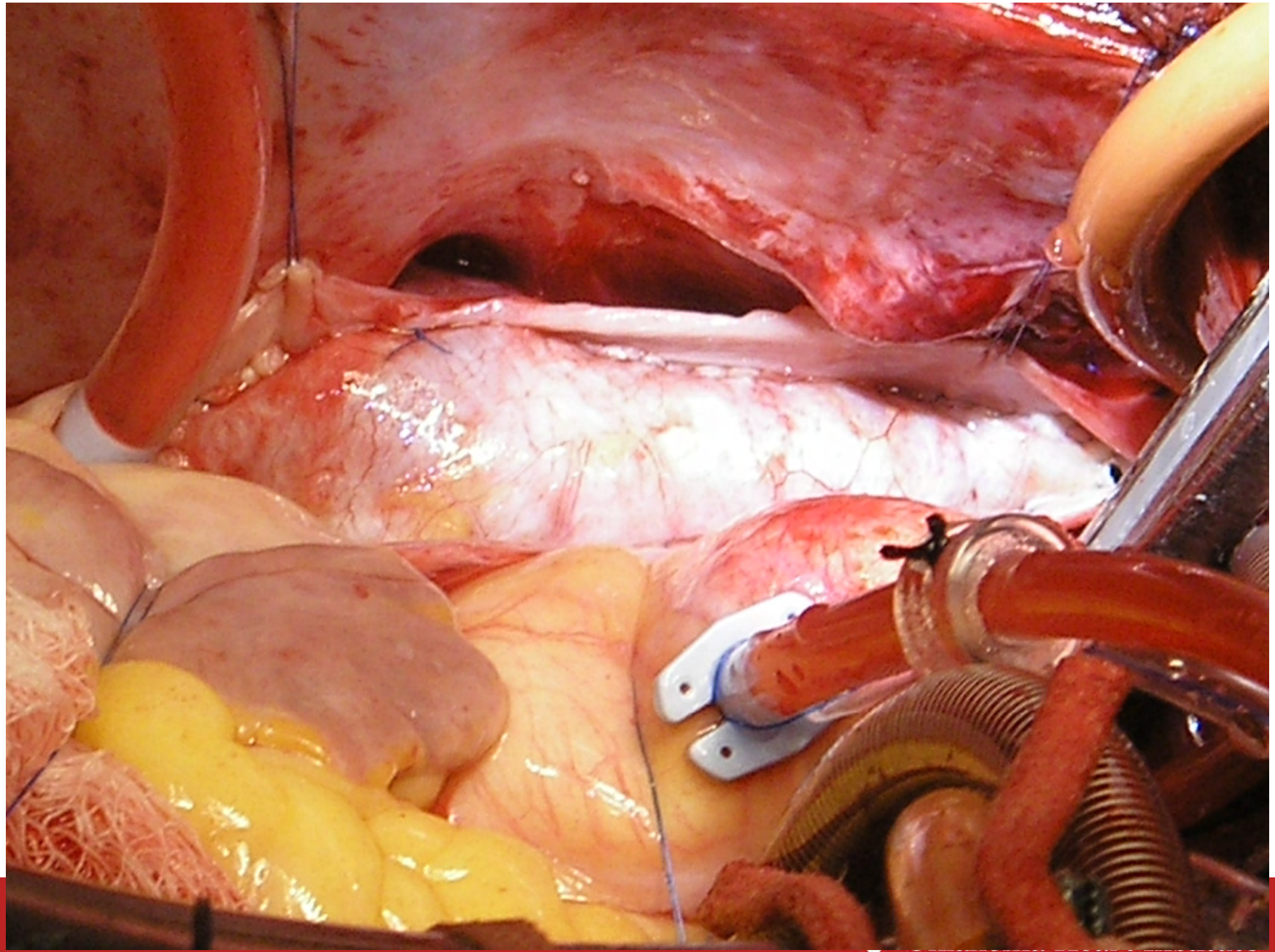
- PAPVC of the RUPV to the SVC/RA junction.
- LU, LL, RL pulmonary veins - normal
- RUPV vein dilated
- SV ASD measuring 1.5 cm
- Bidirectional shunting

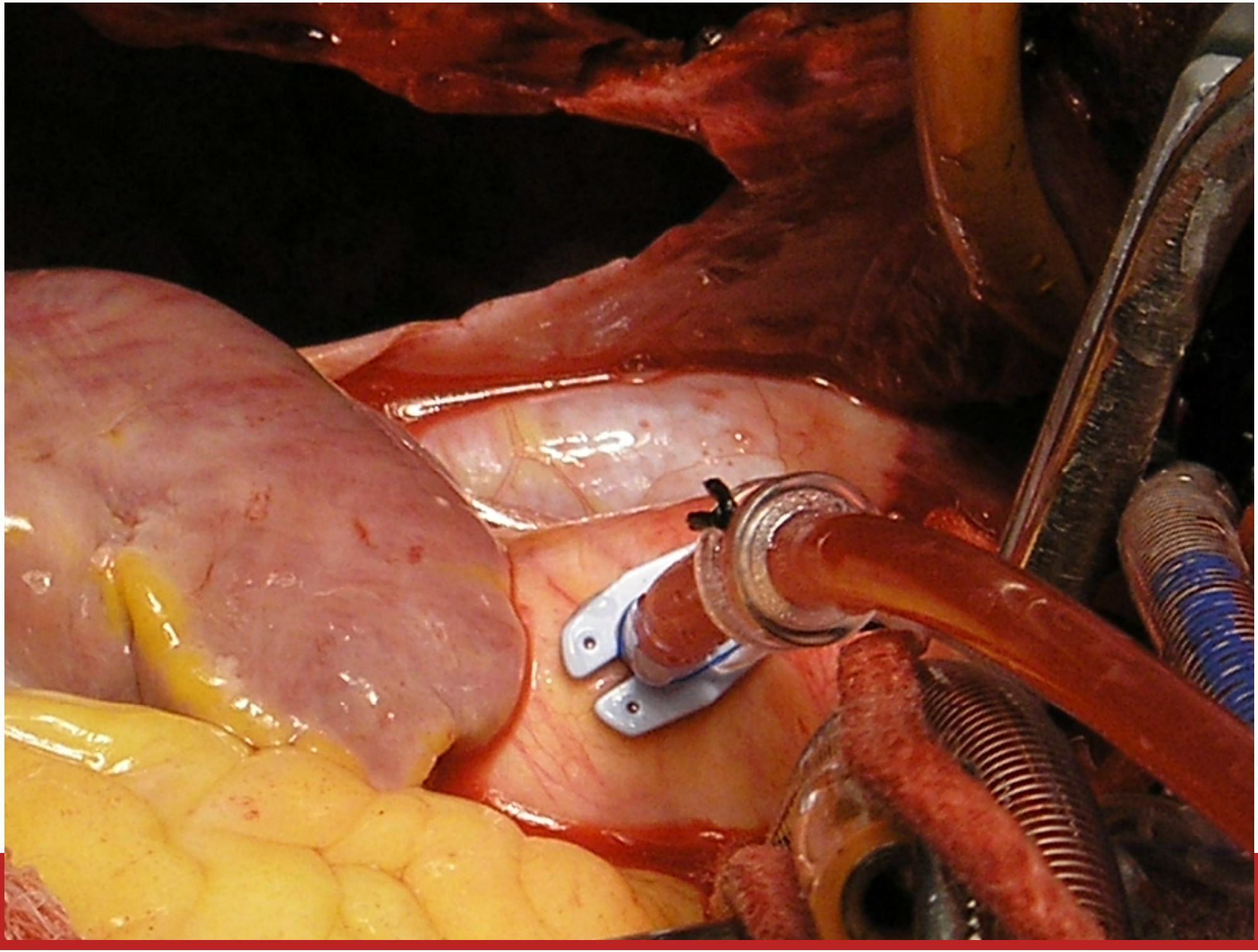






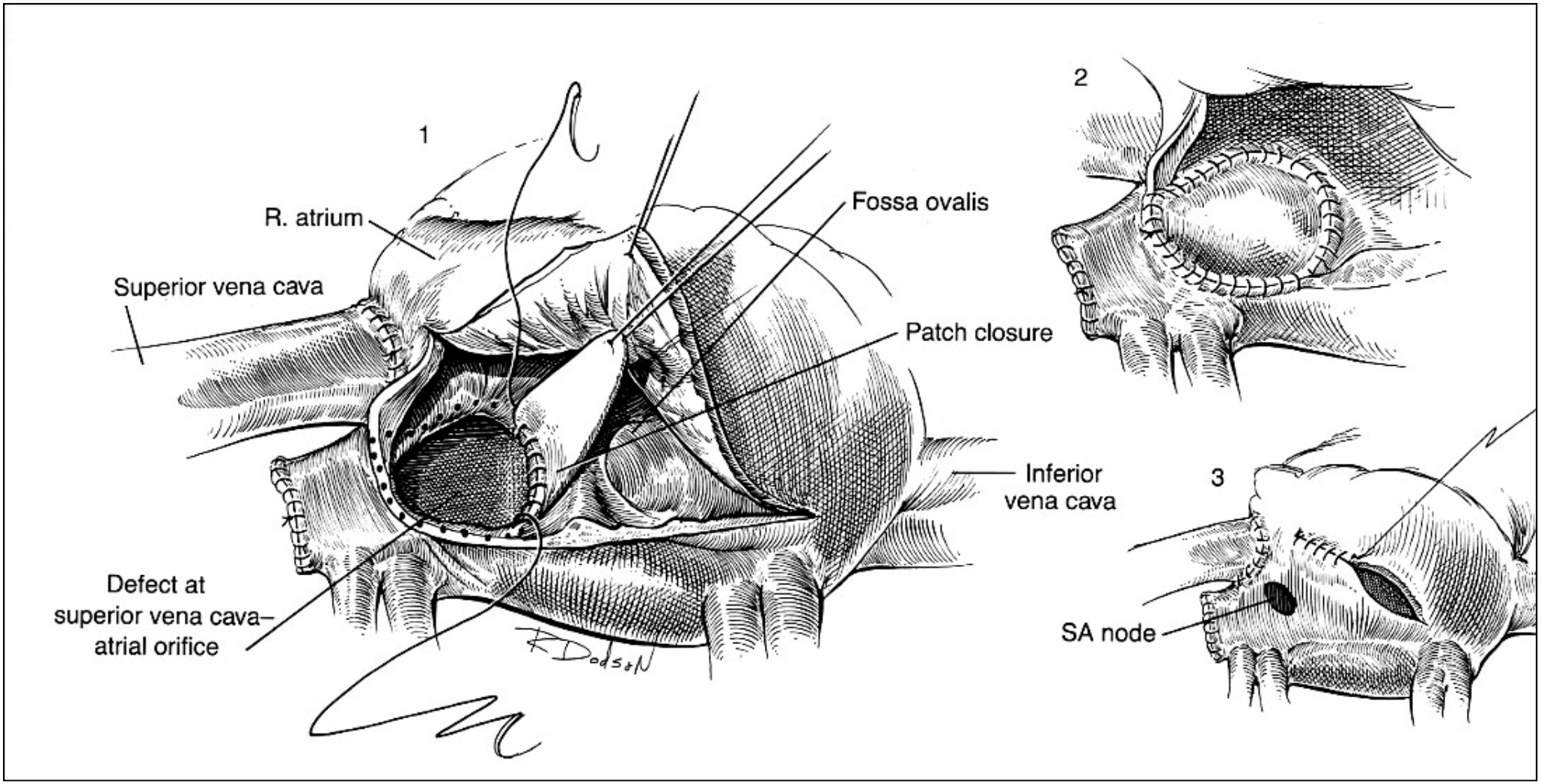






Warden, Gustafson, Tarnay and Neal - 1984

The Warden Procedure



Warden Procedure
Late Sinus Node Dysfunction

- Warden et al - 1984 3/13 (23%)
- Freedom et al - 1984 0/6 (0%)
- Gaynor et al - 1995 0/11 (0%)

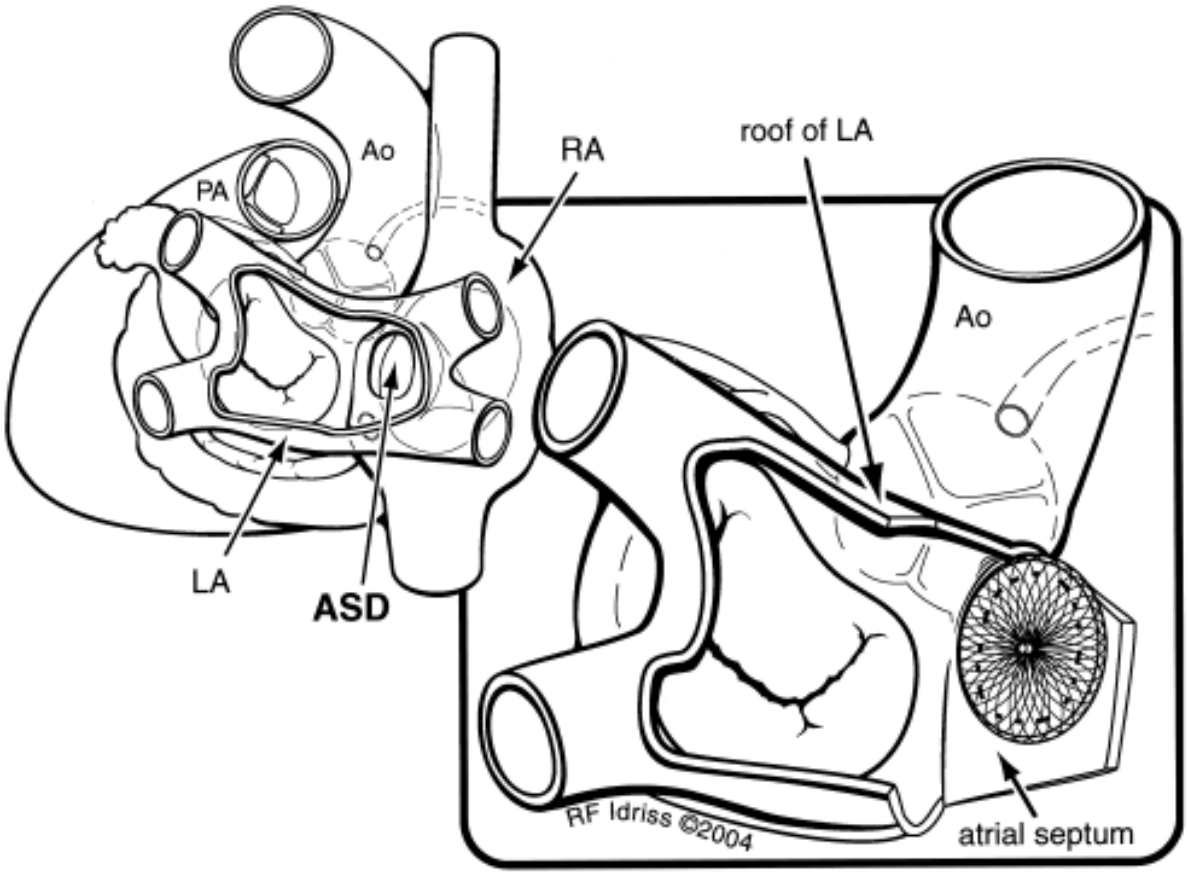
Catheterization and Cardiovascular Interventions 63:496–502 (2004)

Erosion of Amplatzer Septal Occluder Device After Closure of Secundum Atrial Septal Defects: Review of Registry of Complications and Recommendations to Minimize Future Risk

Zahid Amin,[1*†] MD, Ziyad M. Hijazi,[2†] MD, John L. Bass,[3†] MD, John P. Cheatham,[4†] MD, William E. Hellenbrand,[5] MD, and Charles S. Kleinman,[5†] MD

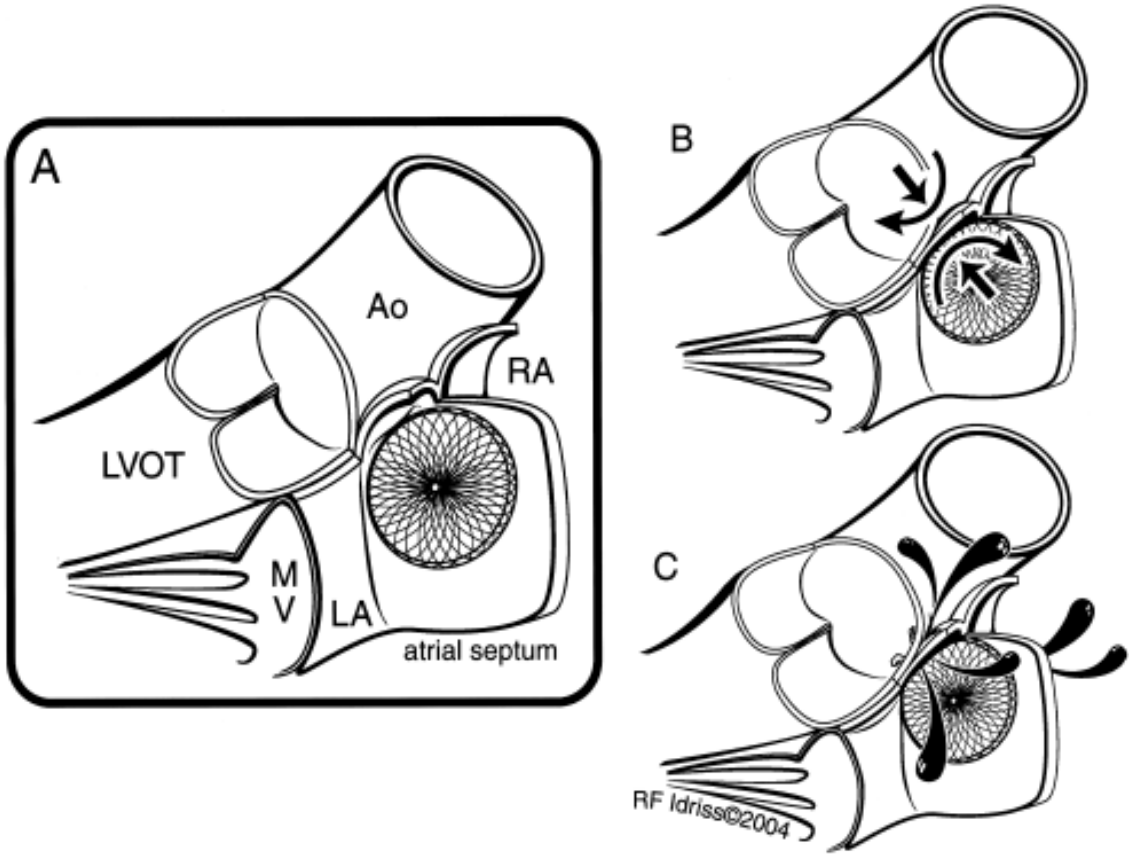
The objectives of this study were to identify possible risk factors that may lead to erosion of the Amplatzer septal occluder (ASO) and recommend ways to minimize future risk. There have been rare occurrences of adverse events with development of pericardial effusion after ASO placement. Identification of high-risk cases, early recognition, and prompt intervention may minimize the future risks of adverse events. In all patients who developed hemodynamic compromise after ASO placement, echocardiograms (pre-, intra-, and postprocedure), atrial septal defect (ASD) size (nonstretched, stretched), size of the device used, cineangiograms, and operative records were reviewed by a panel selected by AGA Medical Corporation. The findings were compared to the premarket approval data obtained from FDA-approved clinical trials that were conducted in the United States, before the device was approved. A total of 28 cases (14 in United States) of adverse events were reported to AGA Medical. All erosions occurred at the dome of the atria, near the aortic root. Deficient aortic rim was seen in 89% and the defect described as high ASD, suggesting deficient superior rim. The device to unstretched ASD ratio was significantly larger in the adverse event group when compared to the FDA trial group. The incidence of device erosion in the United States was 0.1%. The risk of device erosion with ASO is low and complications can be decreased by identifying high-risk patients and following them closely. Patients with deficient aortic rim and/or superior rim may be at higher risk for device erosion. Oversized ASO may increase the risk of erosion. The defect should not be overstretched during balloon sizing. Patients with small pericardial effusion at 24 hr should have closer follow-up. *Catheter Cardiovasc Interv 2004;63:496–502.*

Key words: atrial septal defect; Amplatzer septal occluder; catheterization; complications



- Defect in close proximity to anterior and superior rim

(Z. Amin et al. Erosion of ASOD after closure of secundum ASD's. Catheter Cardiovasc Interv 63 2004)

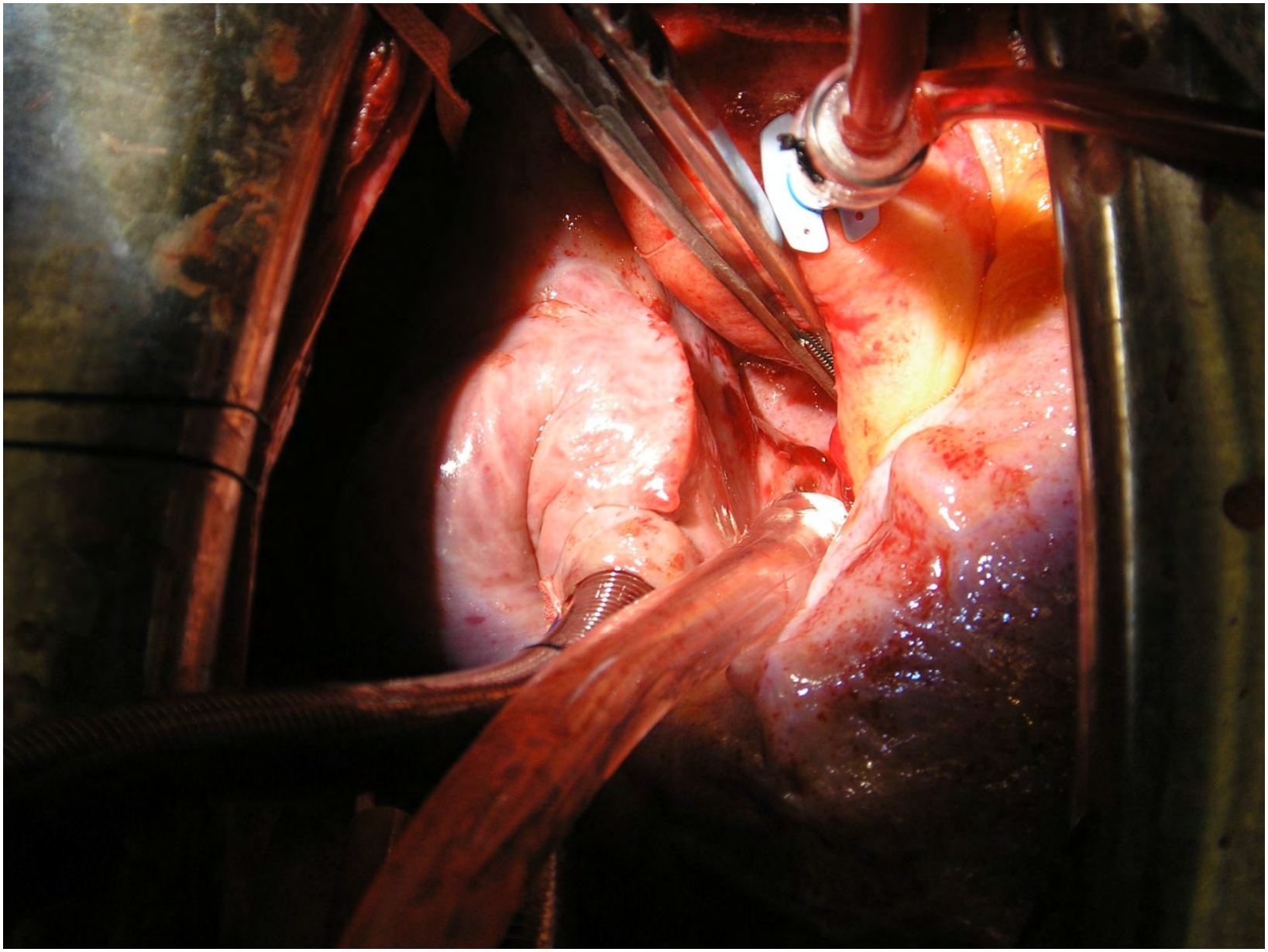


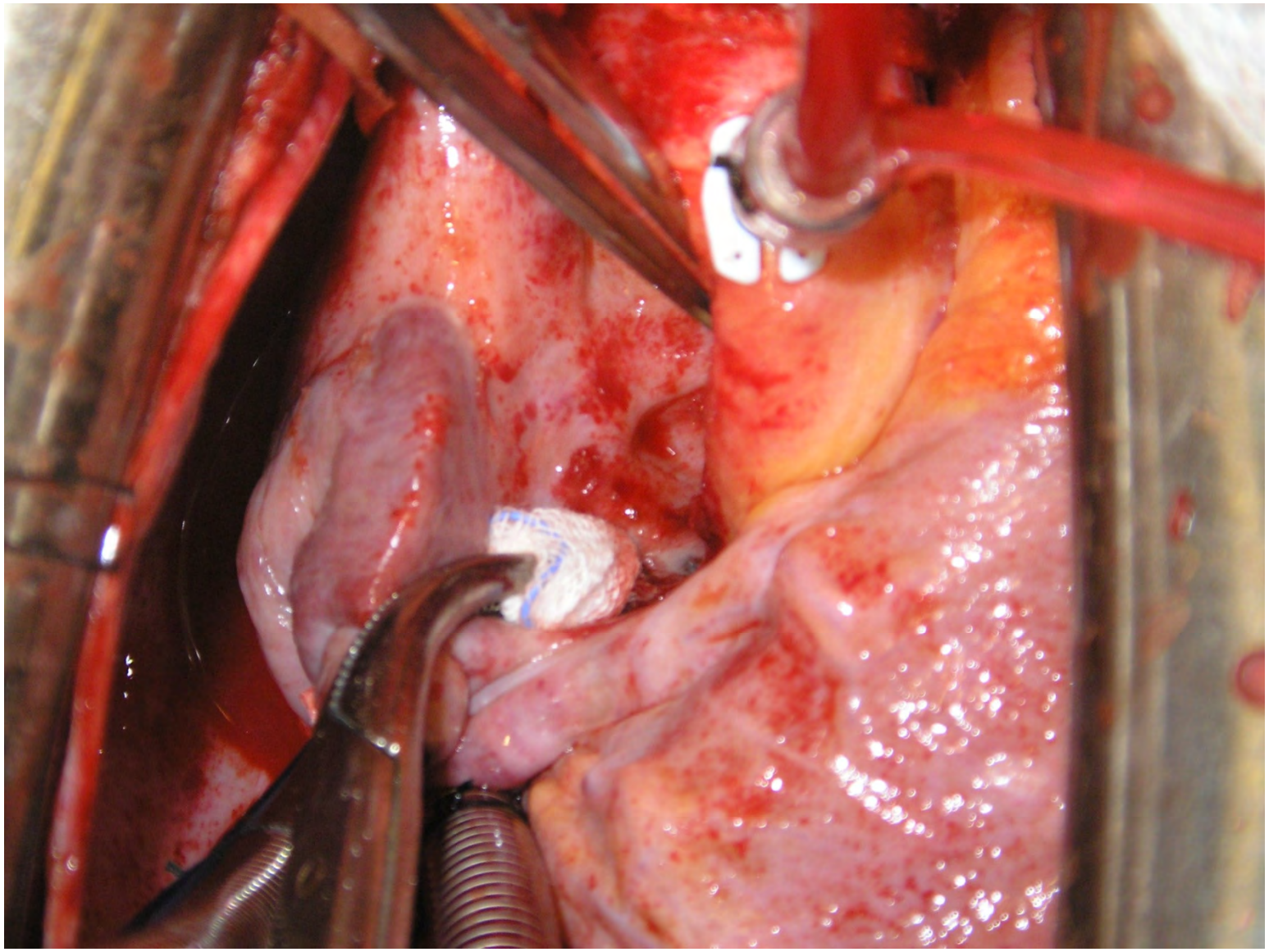
- ASO deployed in the ASD *(Note close proximity of free wall of L&R atrium to the edge of device)*
- With each cardiac cycle, the device results in bruising of atrial wall and/ or aorta

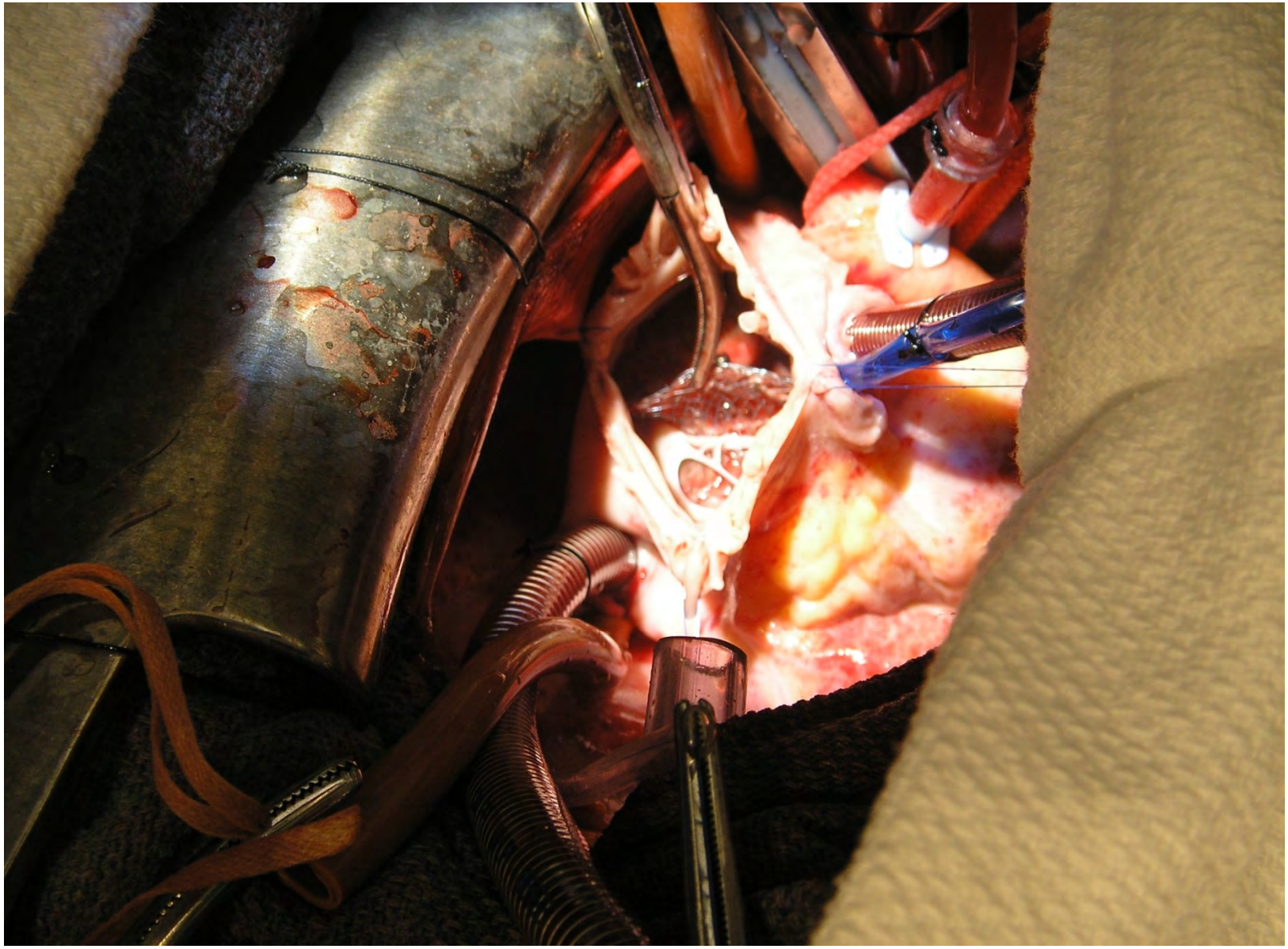
(Z. Amin et al. Erosion of ASOD after closure of secundum ASD's. Catheter Cardiovasc Interv 63 2004)

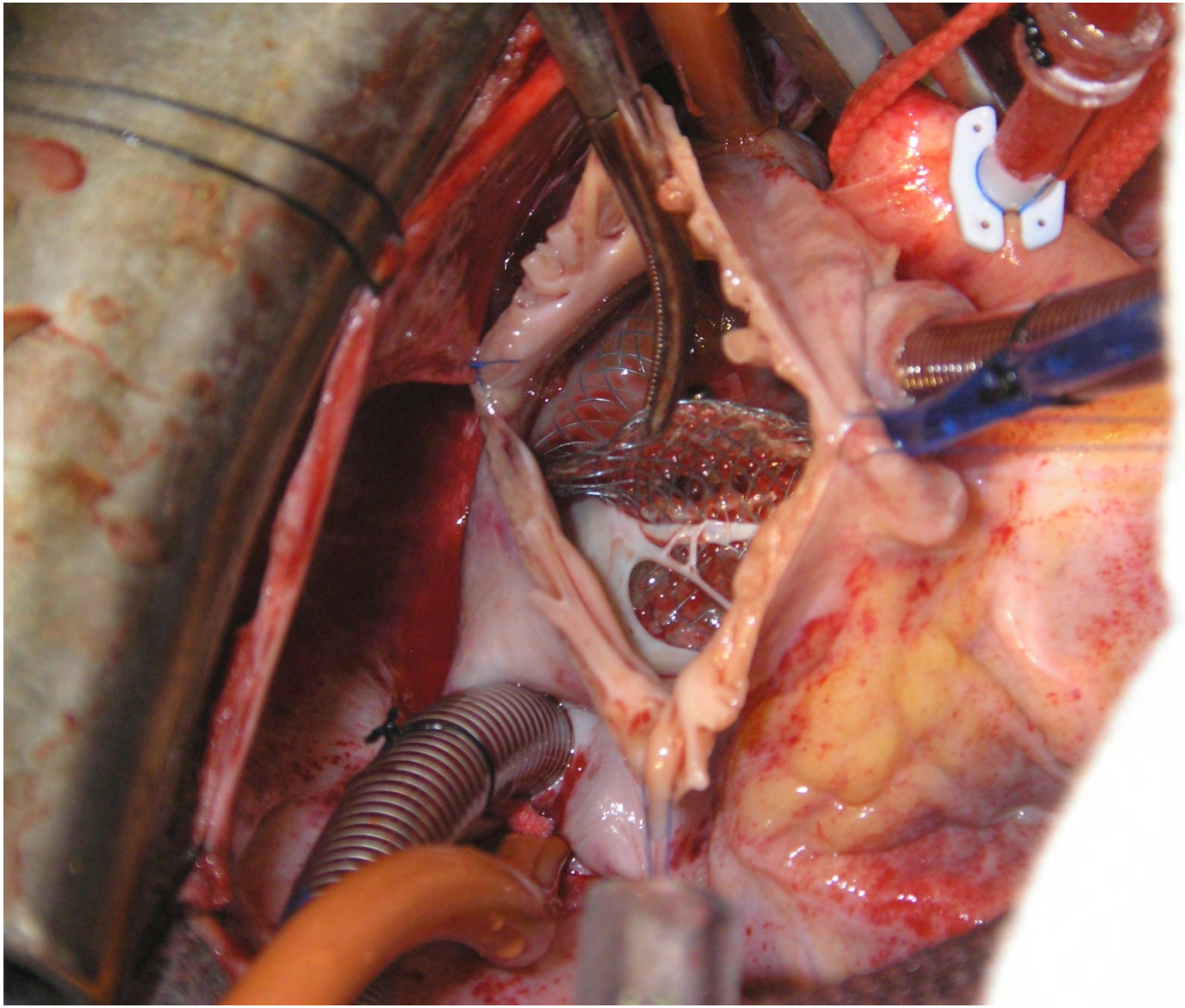
- Risk factors:
  - Deficient rims (esp anterior and superior)
  - Oversizing of the device
  - Device to unstretched ASD ratio was higher in adverse event group compared to the FDA trial group
- All erosions occurred at the dome of the atria, near aortic root
- Incidence in US: 0.1%

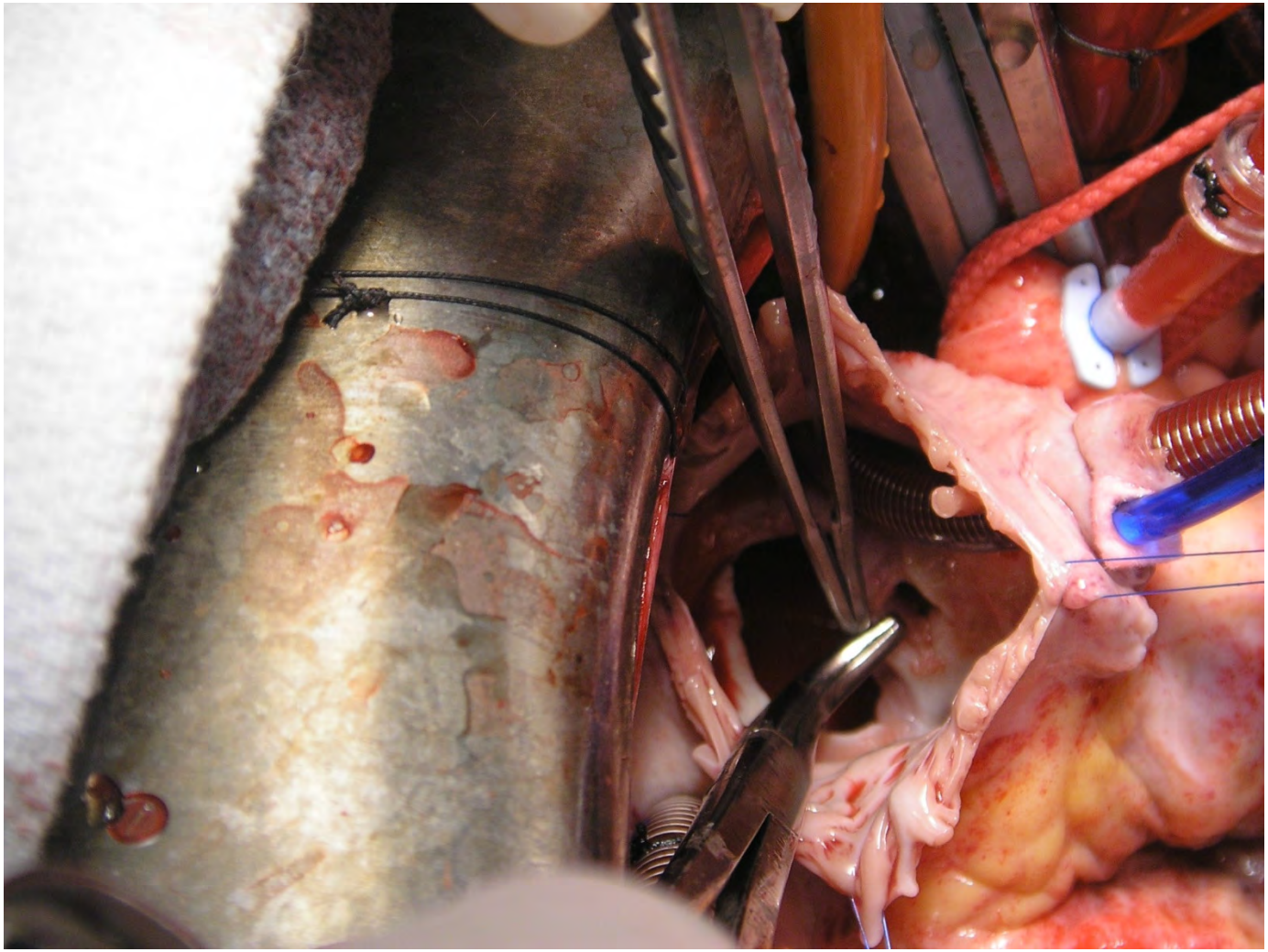
(Z. Amin et al. Erosion of ASOD after closure of secundum ASD's. Catheter Cardiovasc Interv 63 2004)

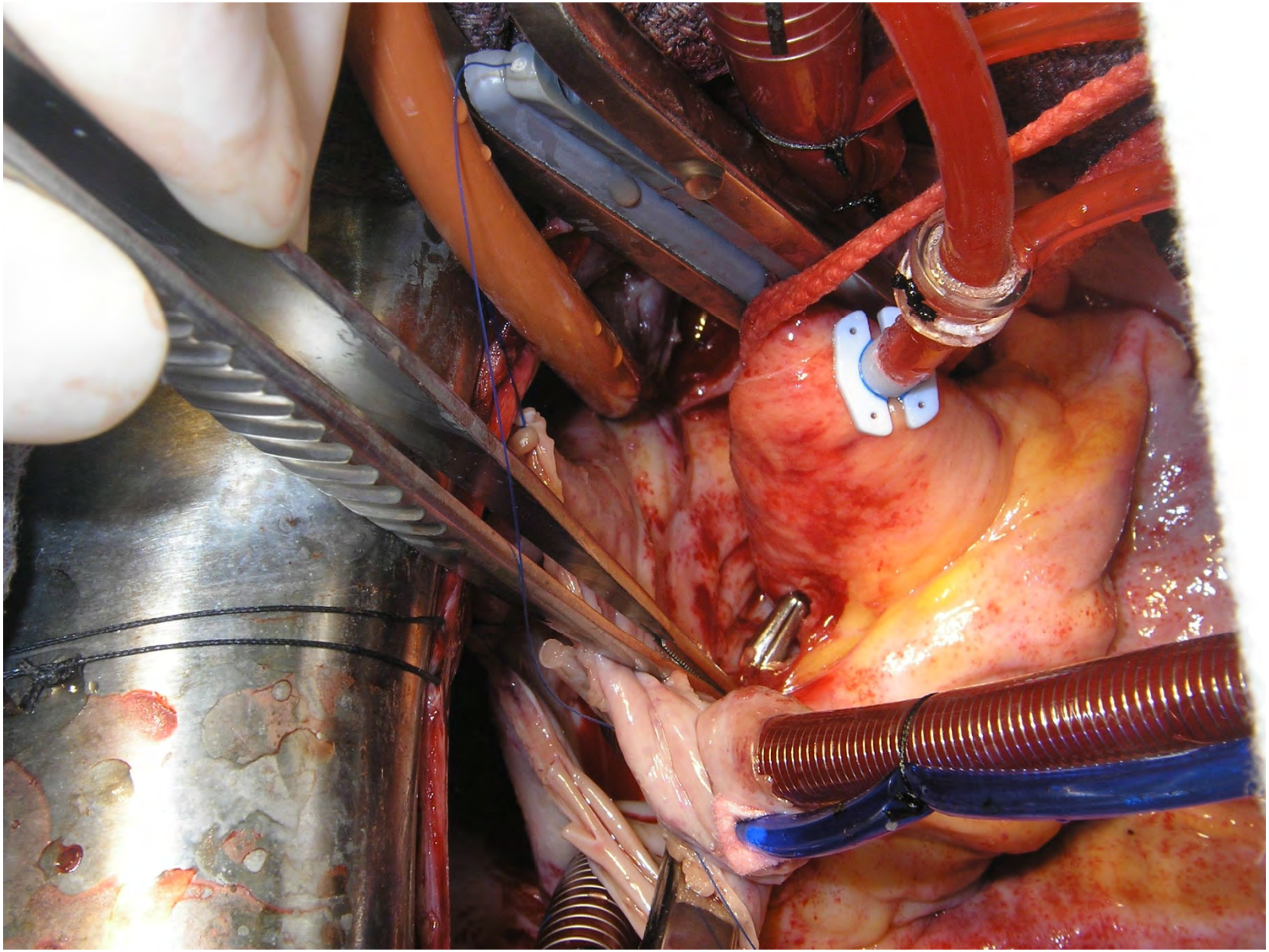


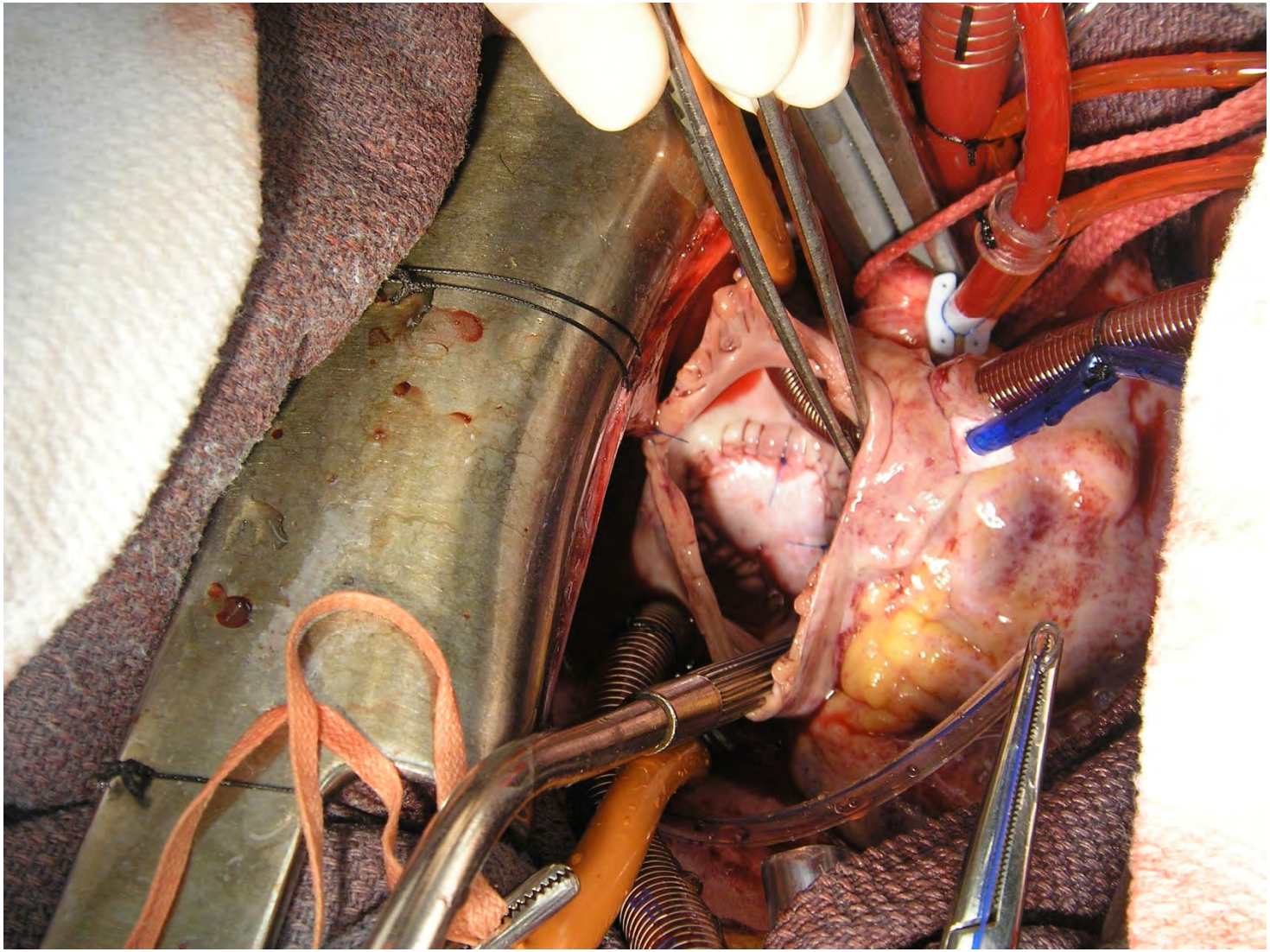










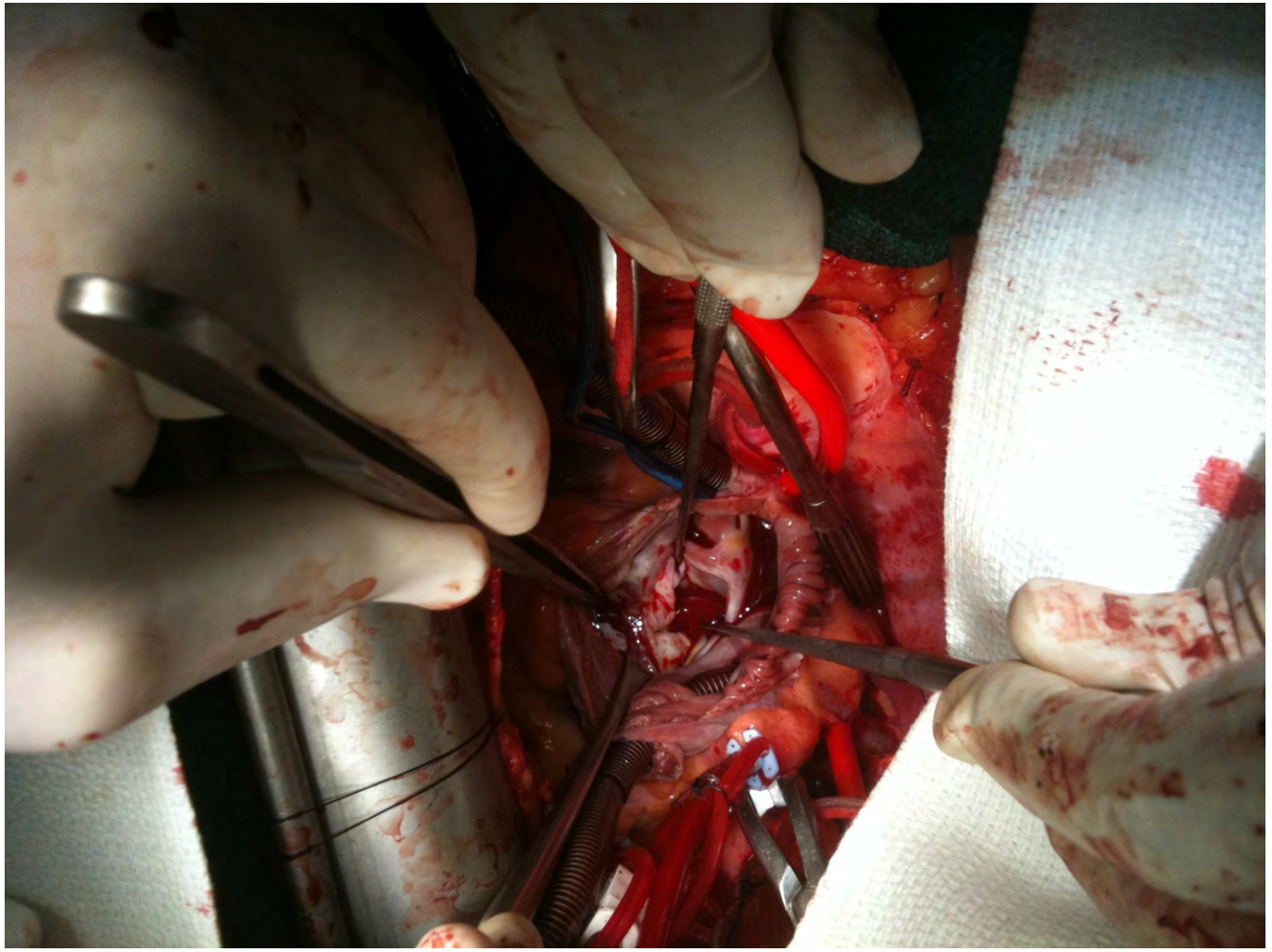


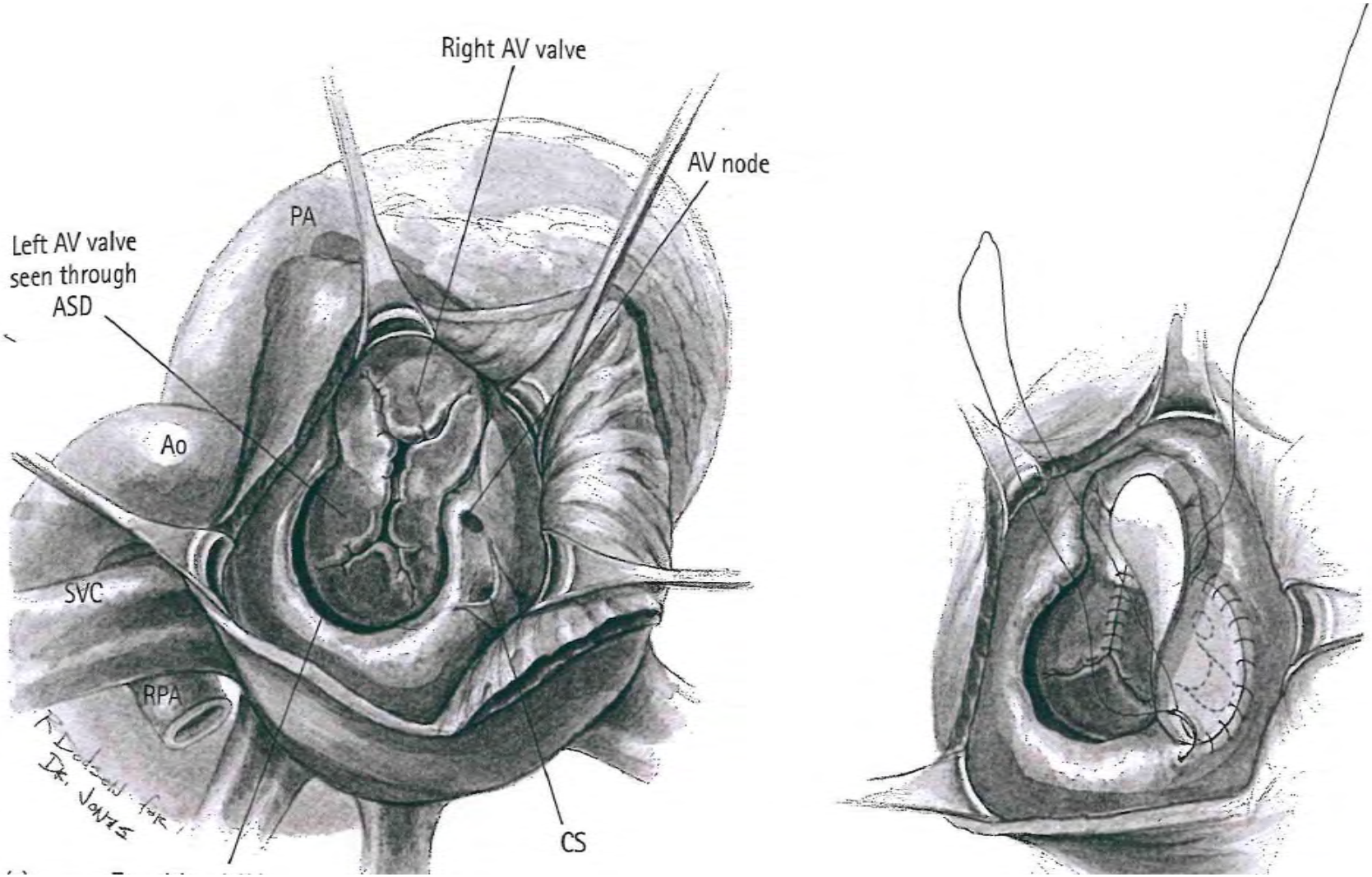

VISCOT
MEDICAL LLC
EAST HANOVE
METRIC 1 2 3 4 5 6 7

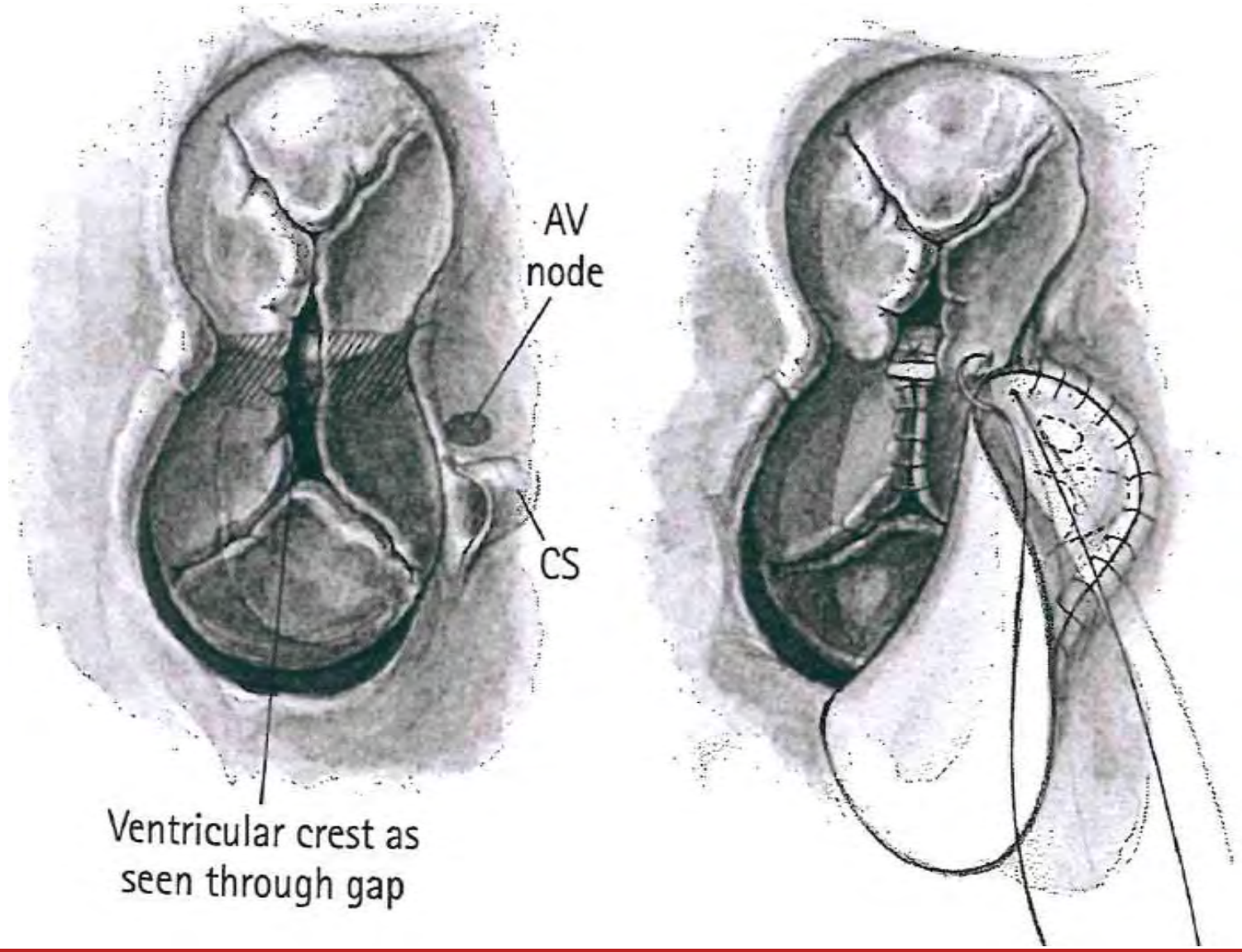
Surgery

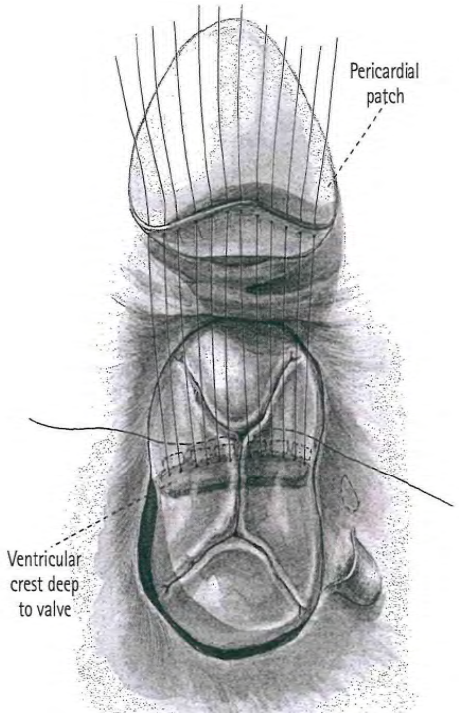
- RA was opened and the device was removed
- Erosion through the roof of Rt atrium and through the adventitia and media of the aortic root
- 3 cm pericardial patch was sewn to close the defect with a continuous 4-0 prolene
- RA defect closed from inside with 2 layers of 4-0 prolene
- Aortic rent closed in 2 layers
- Cross clamp time 31 mins + 11 mins

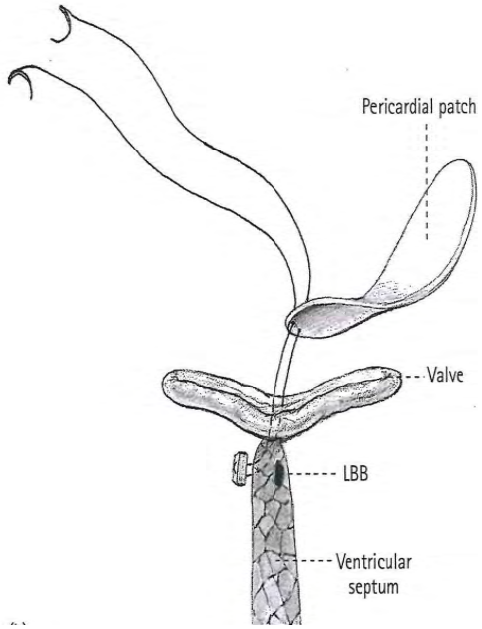
Ostium Primum defects & late reoperations after CAVC

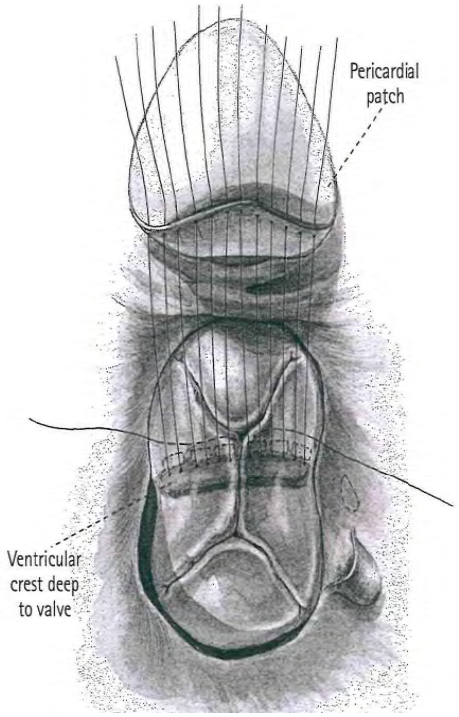


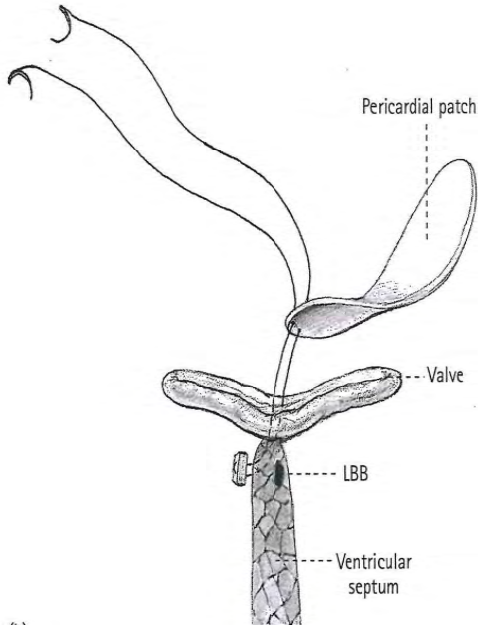

Right AV valve
AV node
PA
Left AV valve
seen through
ASD
Ao
SVC
RPA
CS

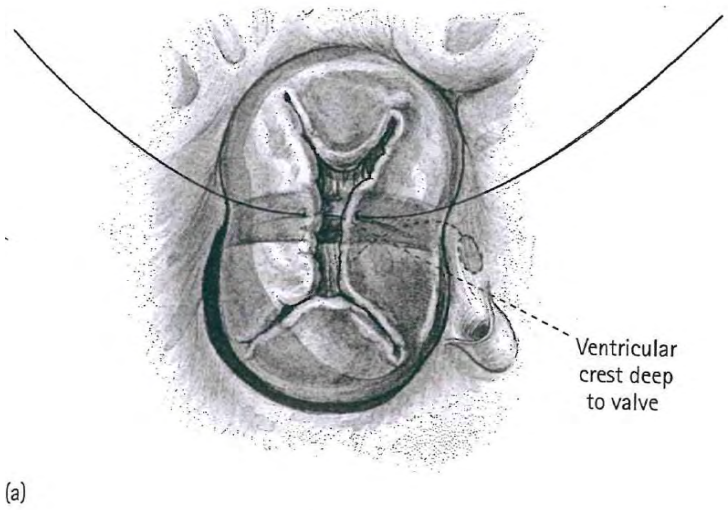

AV
node
CS
Ventricular crest as
seen through gap


Pericardial patch
Ventricular crest deep to valve

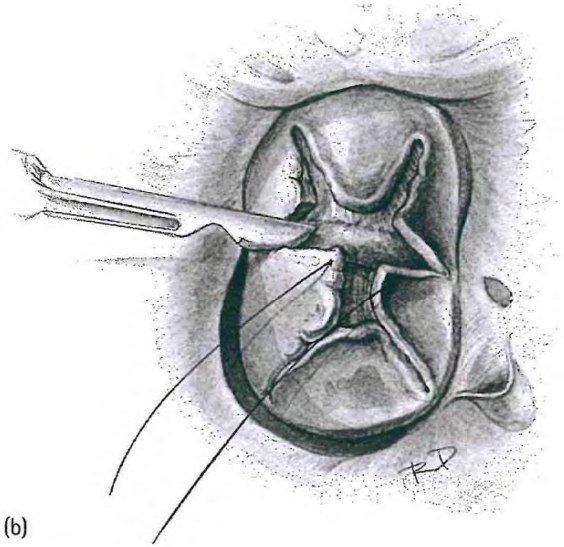

Pericardial patch
Valve
LBB
Ventricular septum


Pericardial patch
Ventricular crest deep to valve

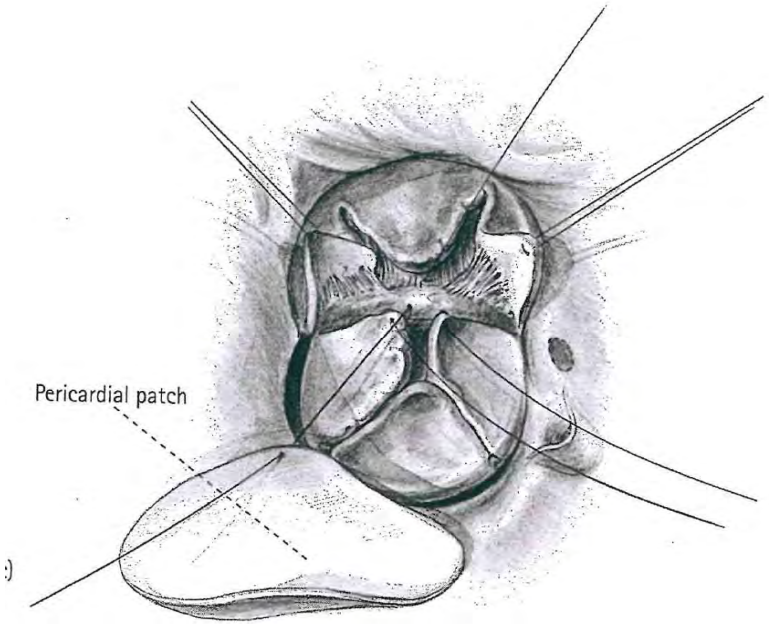

Pericardial patch
Valve
LBB
Ventricular septum


Ventricular
crest deep
to valve

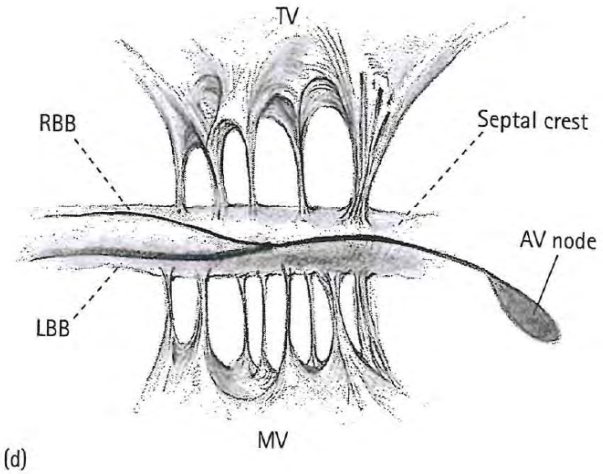
(a)



(b)


Pericardial patch

(c)


TV
RBB
Septal crest
AV node
LBB
MV

(d)

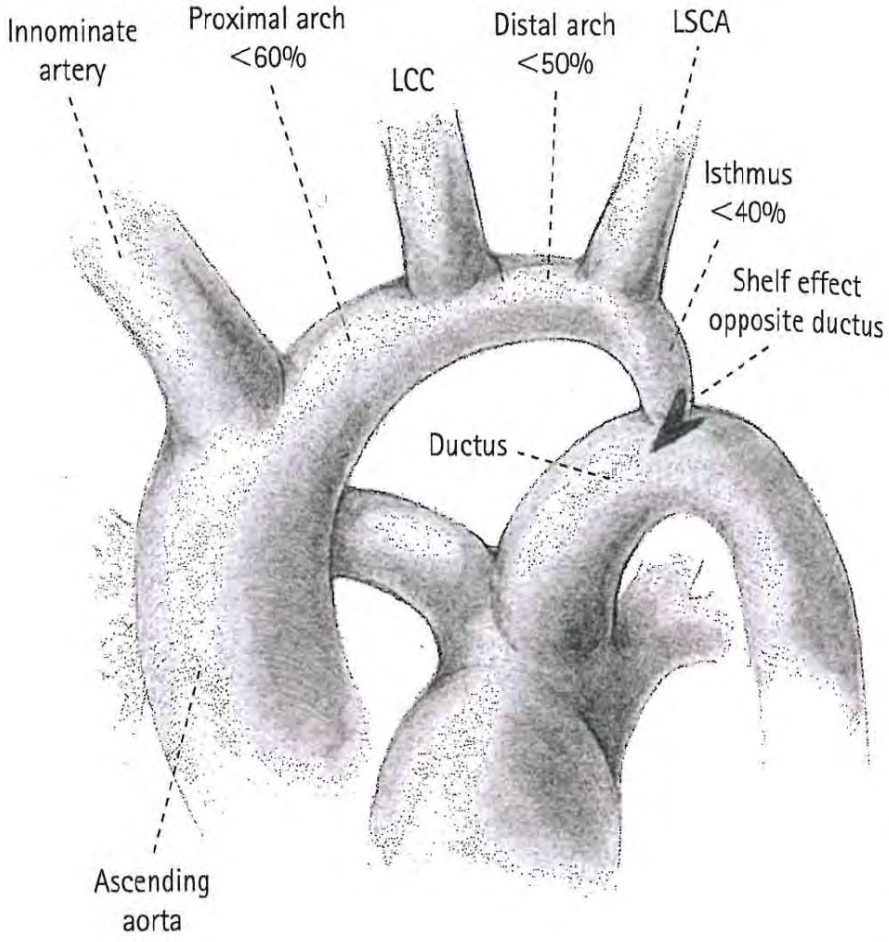
Late reoperations after repair of AV canal

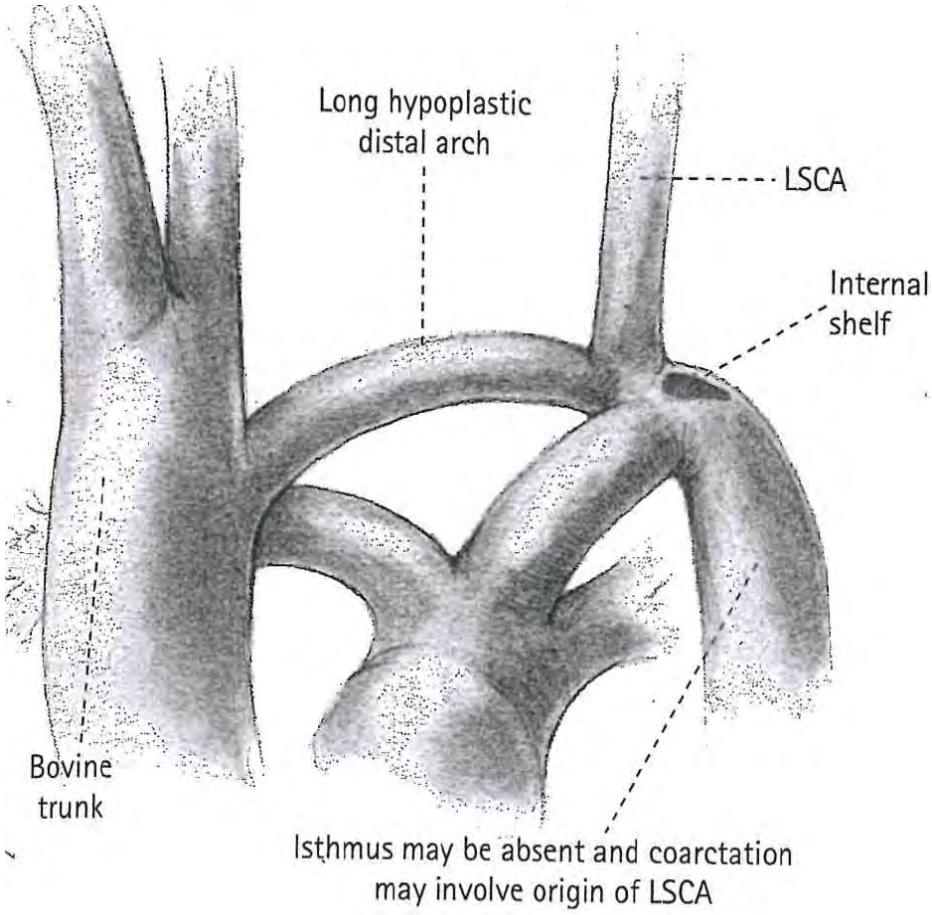
- Late AV valve regurgitation
  - 10-30% incidence of AV valve malfunction
  - Most common cause- residual or recurrent cleft in the anterior leaflet
  - Repair/ replacement

Late reoperations after repair of AV canal

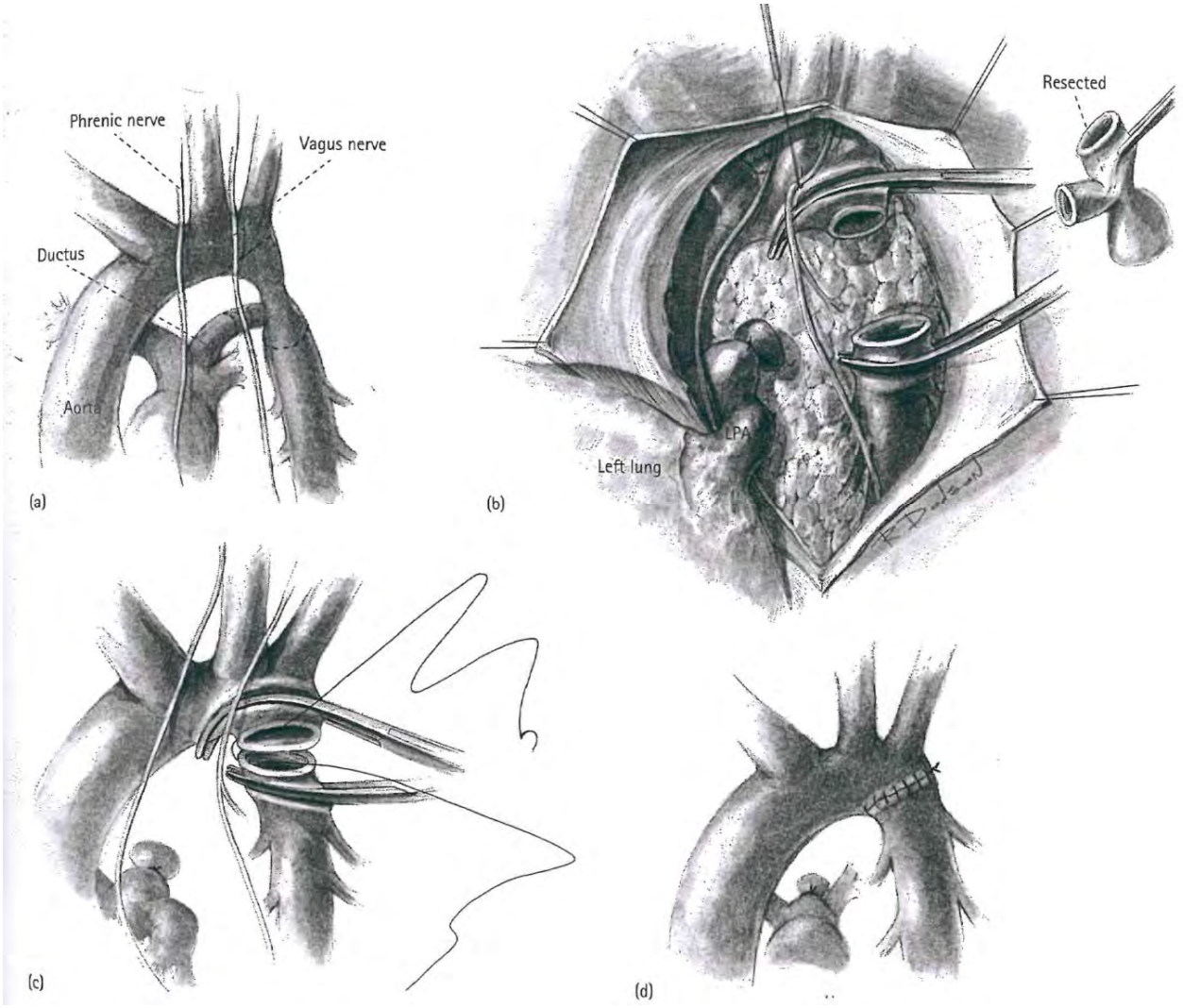
- LVOTO after AV canal repair
  - left superior leaflet chordae that are attached to the outlet septum,
  - outgrowths of accessory valve tissue in the left ventricular outflow tract.
  - subaortic membrane
  - abnormal papillary muscle position.

Coarctation of Aorta

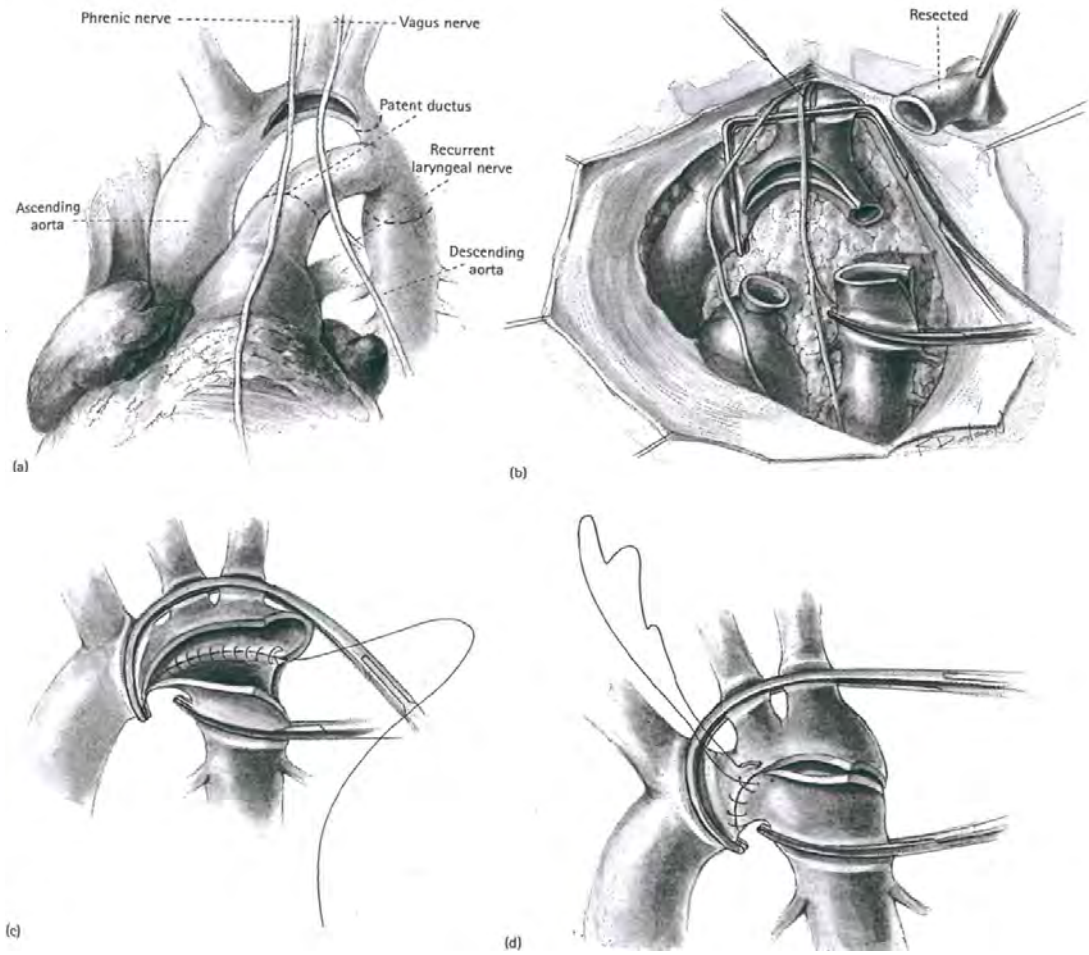




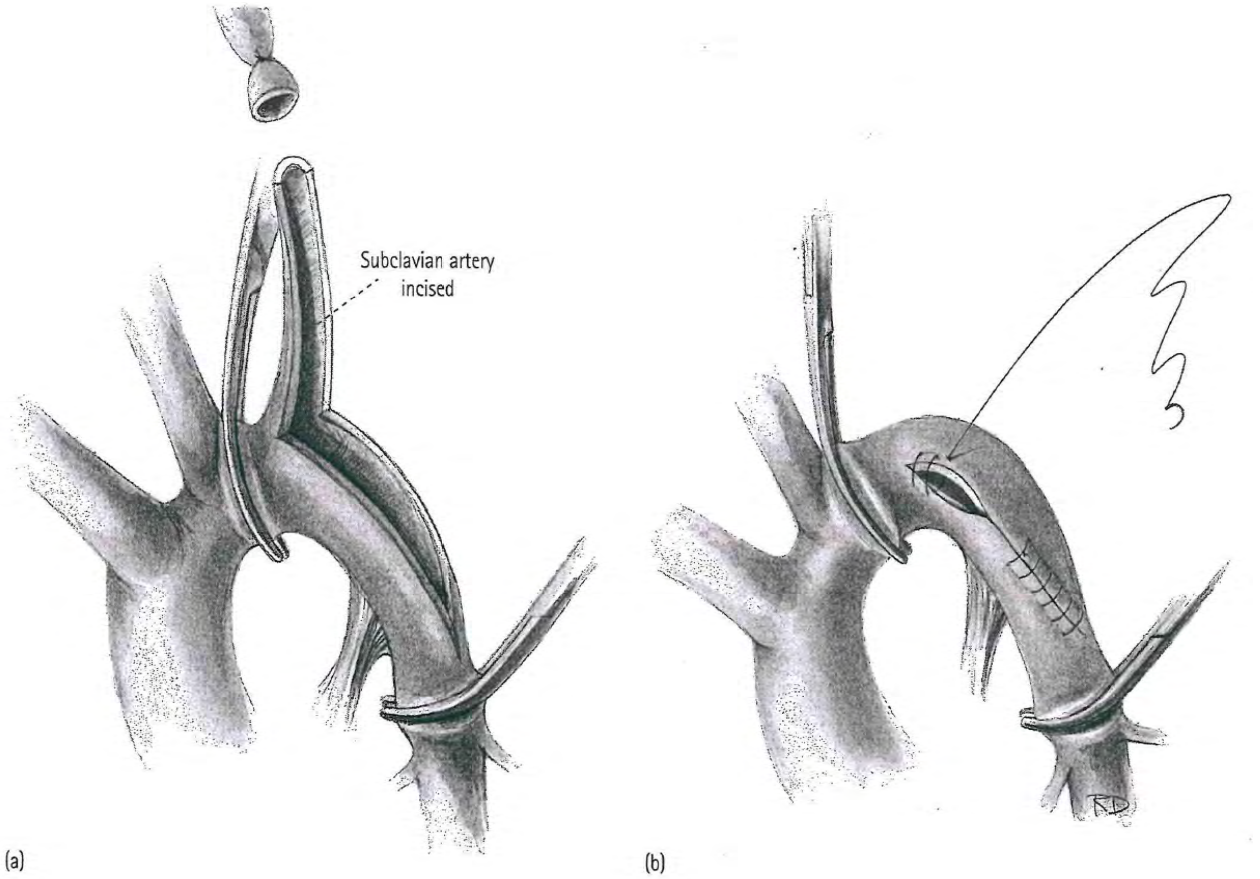
Resection & E-E Anastomosis



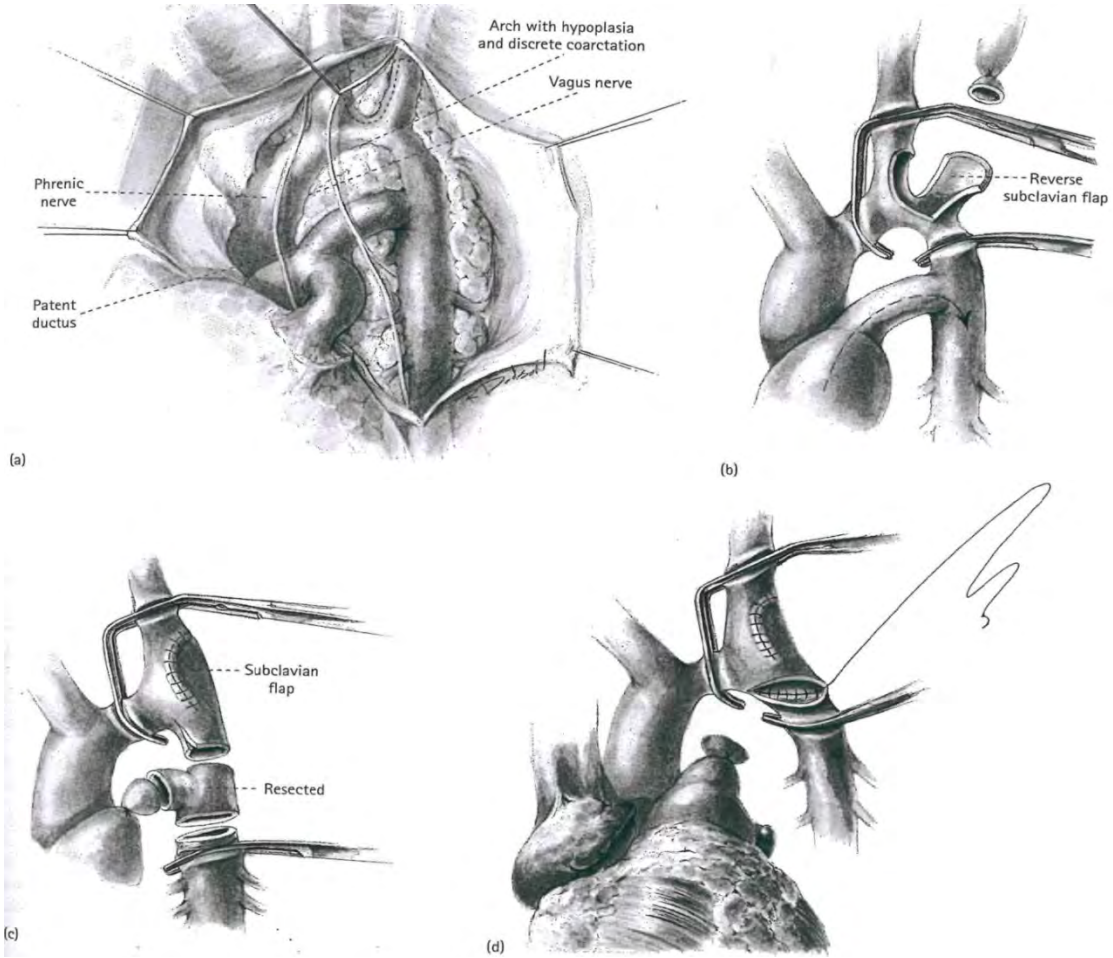
Extended Resection & E-E Anastomosis



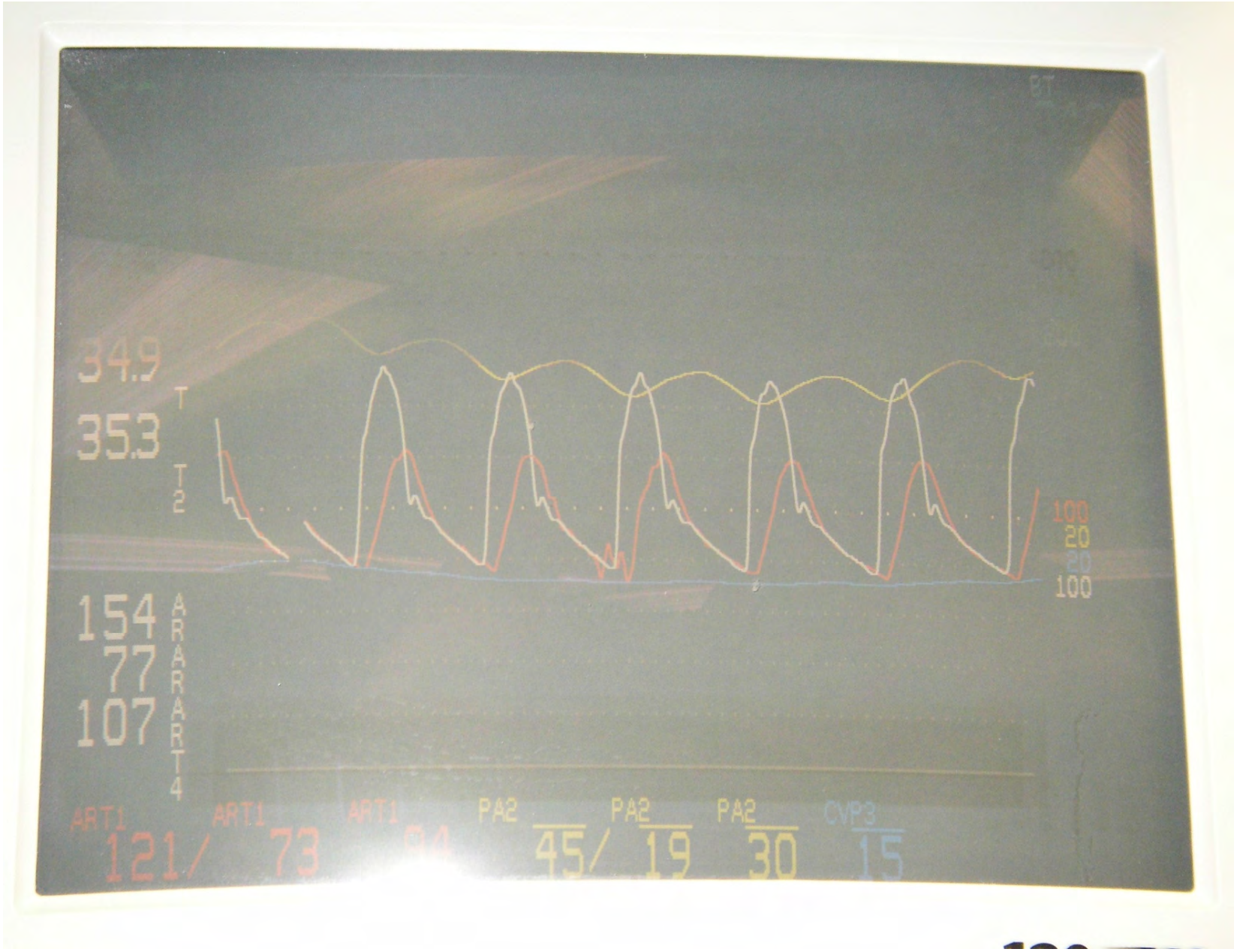
Subclavian Patch aortoplasty

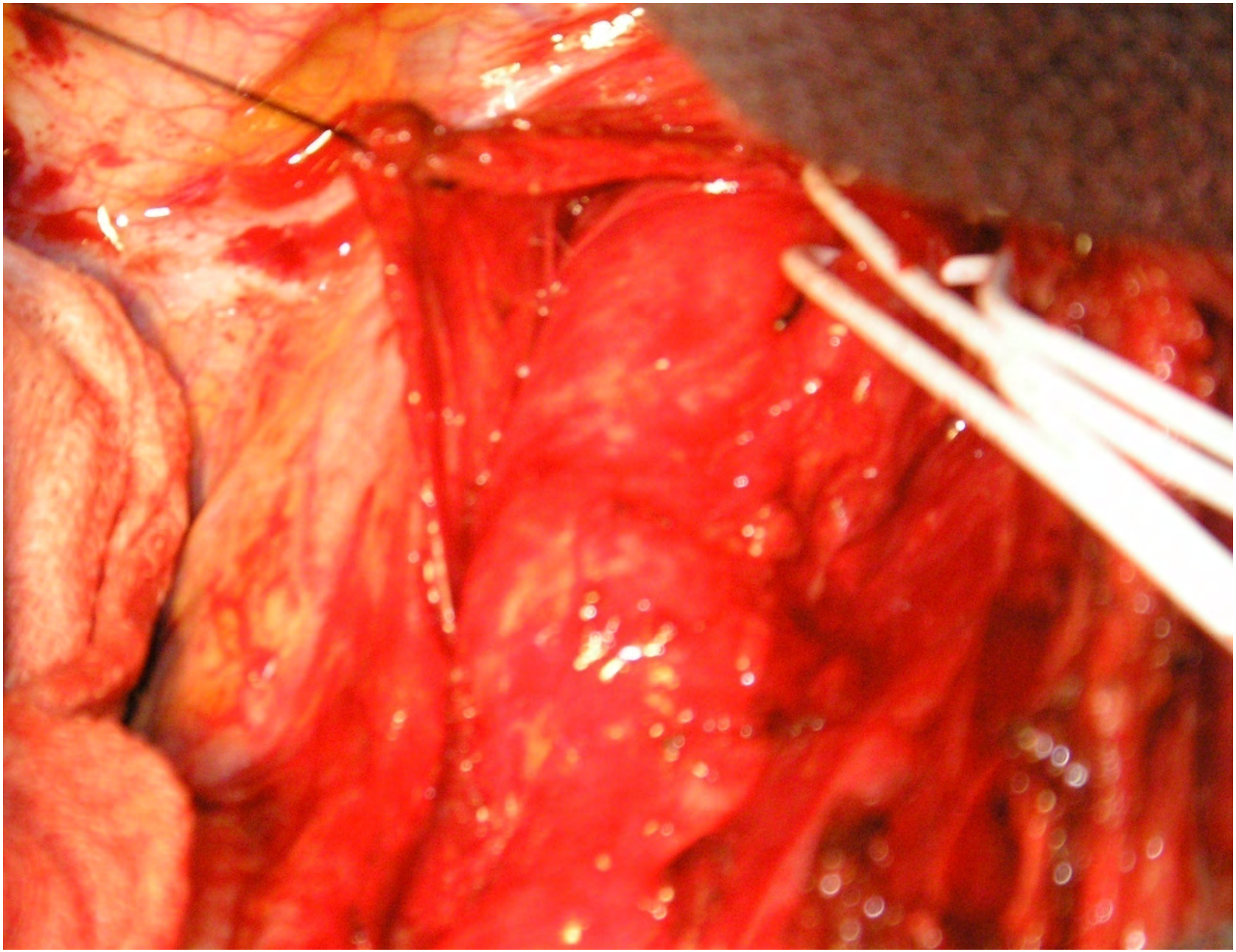


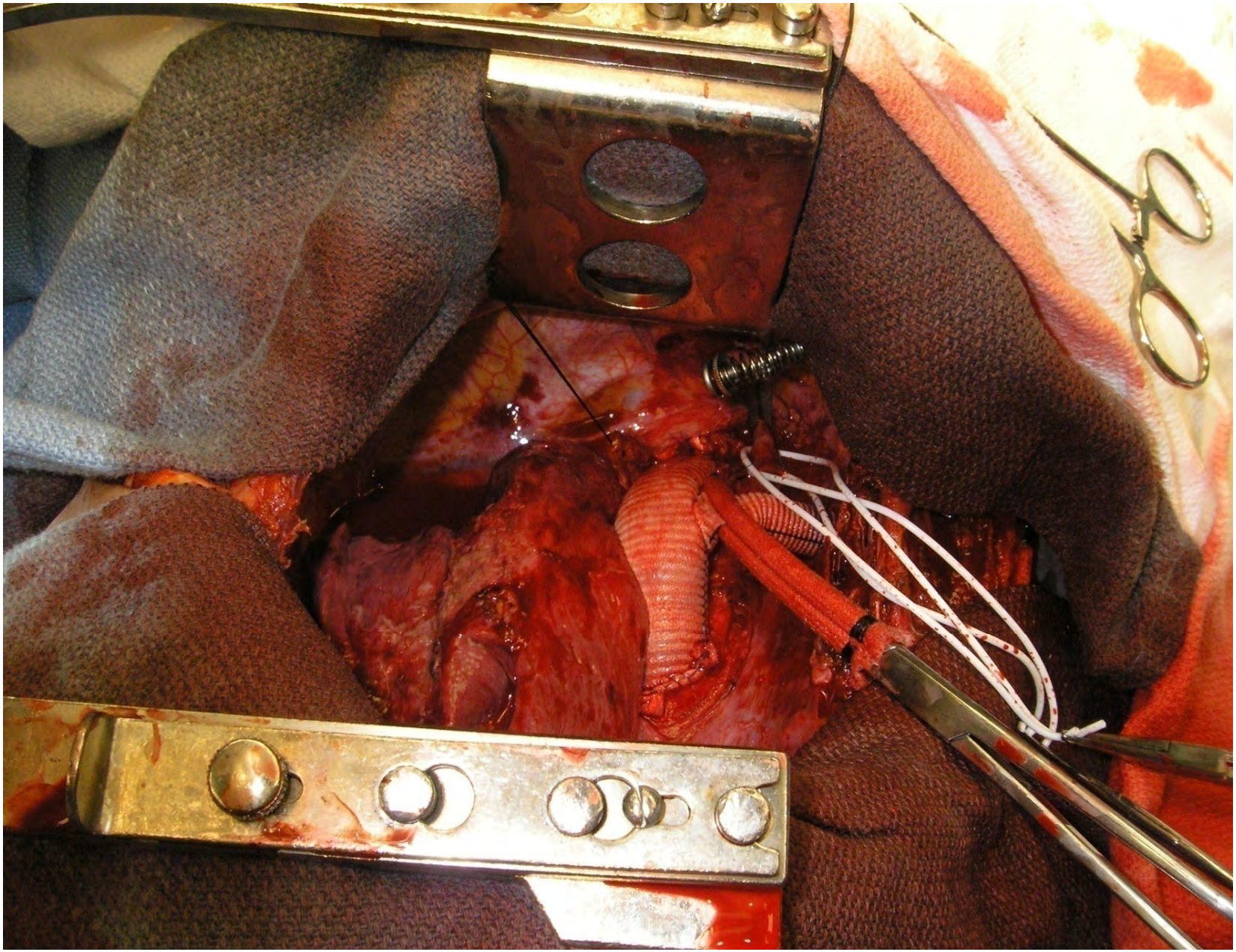
Reversed Subclavian Patch Plasty

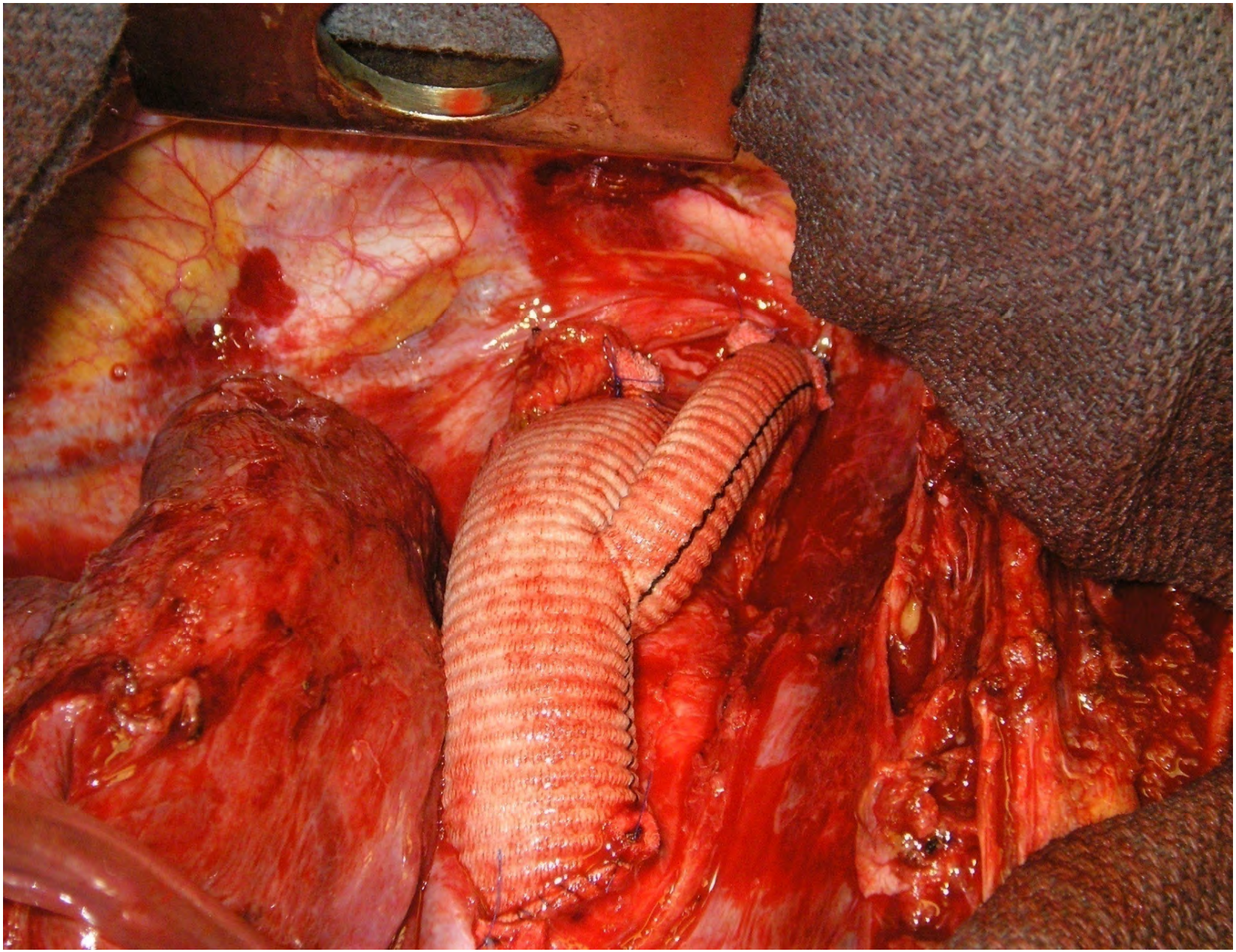


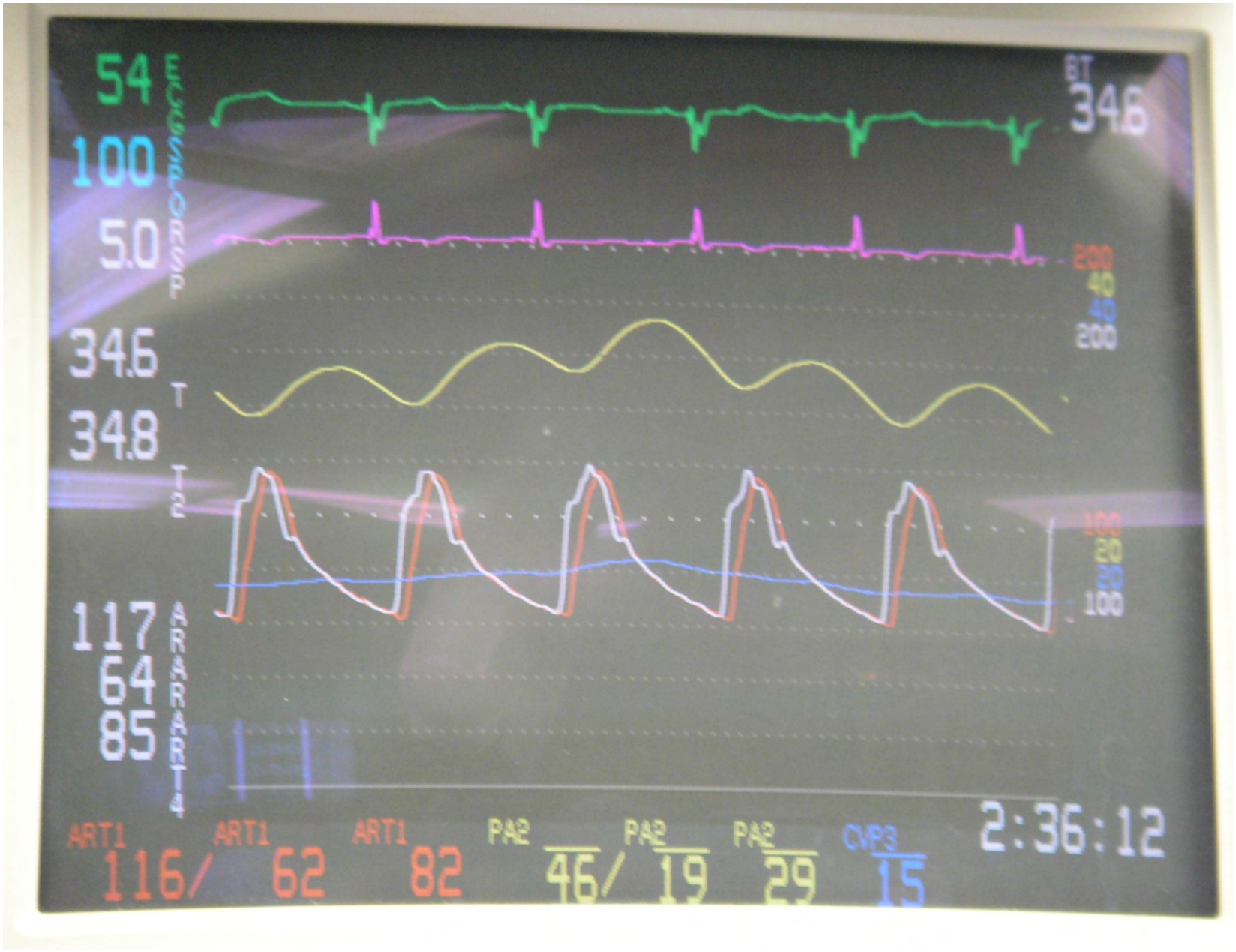
- 45 yr old gentleman
- Presented with chest pain and shortness of breath
- CT angiogram showed a coarctation of aorta distal to the left subclavian artery


34.9
T
35.3
T2
154
77
107
ART4
100
20
20
100
ART1
121/ 73
ART1
ART1
PA2
45/ 19
PA2
30
PA2
CVP3
15





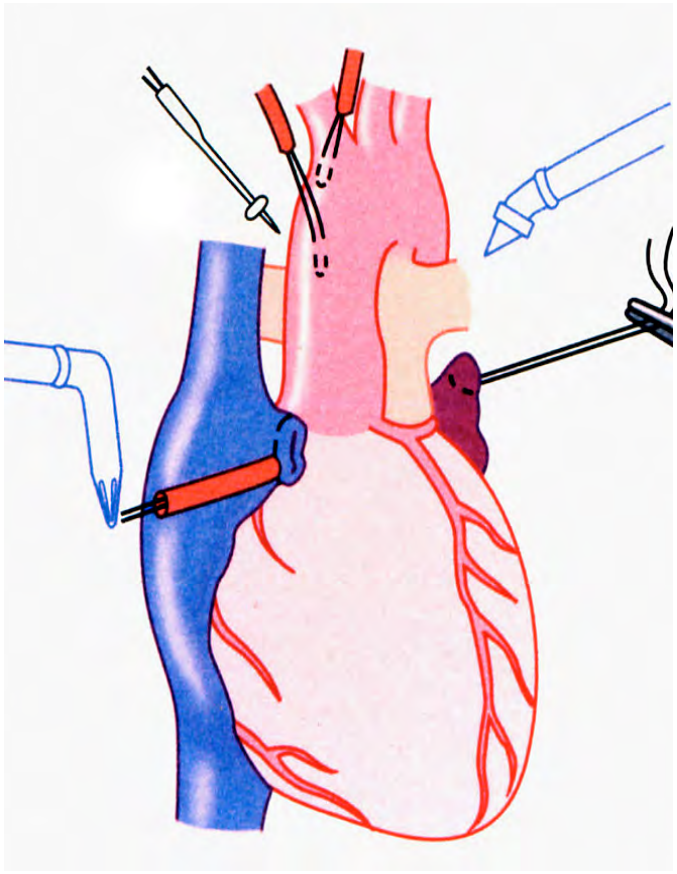


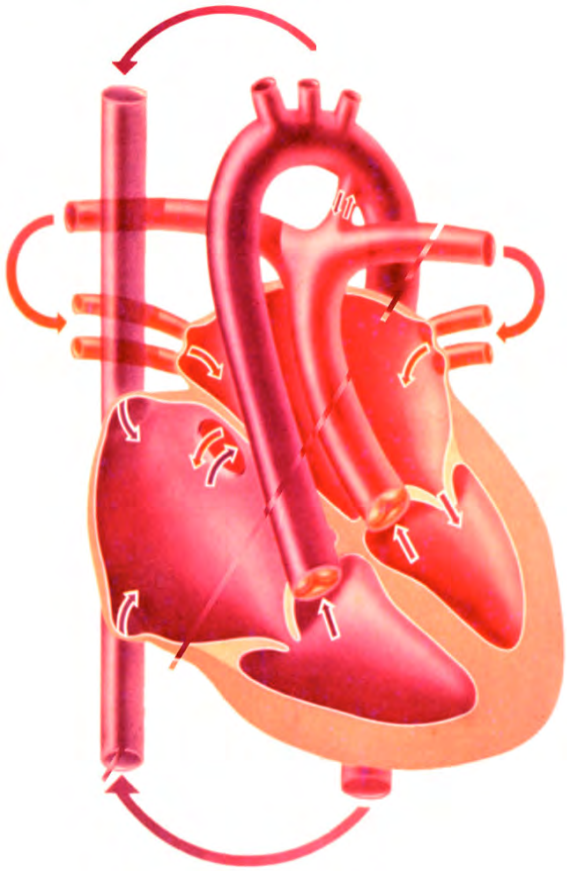

54
100
5.0
34.6
34.8
117
64
85
ART1
116/ 62
ART1
82
ART1
PA2
46/ 19
PA2
29
PA2
CVP3
15
2:36:12

Outcomes

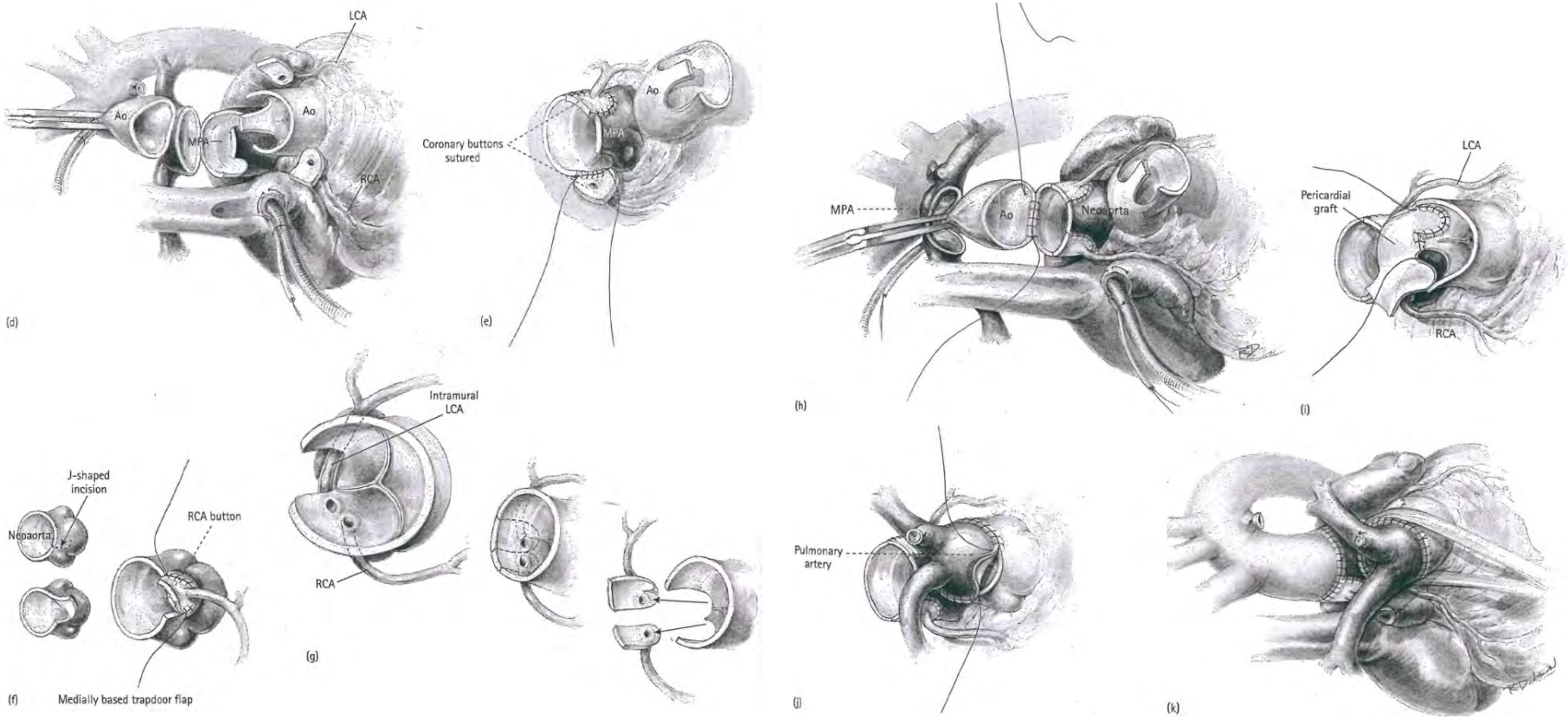
- Excellent
- Recoarctation
  - Defined as a residual gradient > 20-30 at rest
- Catheter based techniques used for primary and recurrent coarctation

Tranposition of Great Arteries





DTGA
Arterial Switch Operation



Atrial Switch Operations

- Mustard- Pericardial baffle used to redirect blood
- Senning- Atrial septum used to redirect blood
- Right becomes systemic ventricle
- 70-80% survival at 20 yrs

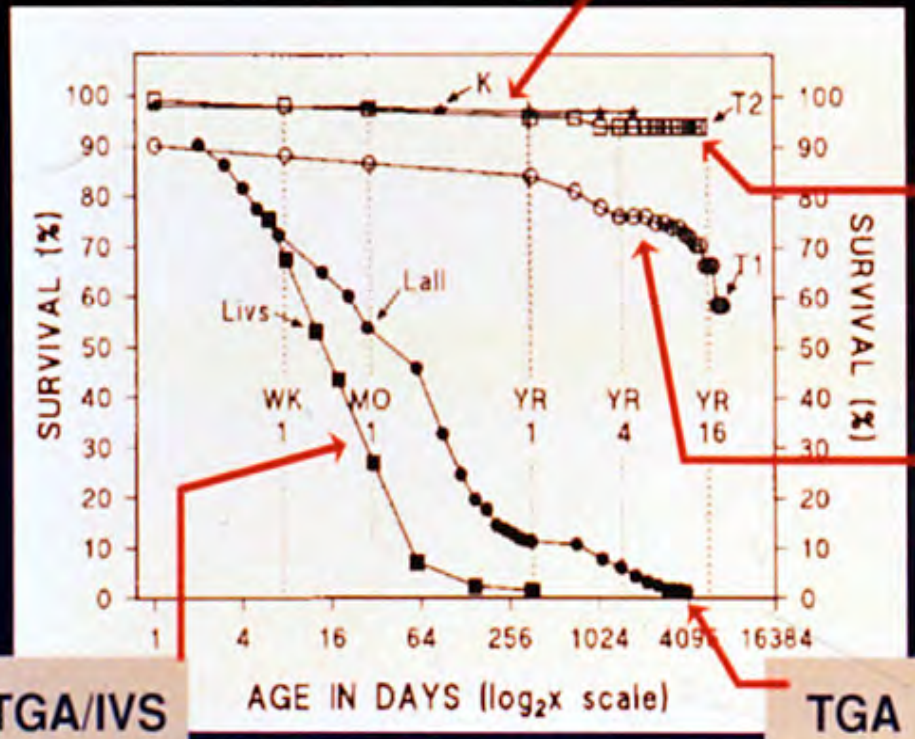
Atrial Switch Operation

- 10 % symptomatic at rest, 60% with exercise
- Atrial arrhythmias common
- Baffle obstruction leads to PV or Systemic venous congestion and eventually systemic ventricular failure
- Sudden death is most common cause of mortality after an atrial switch operation

Reoperation

- Left ventricular retraining with PA banding and arterial switch
- Repair of baffle obstruction with pericardial patch
- May need tricuspid valve replacement

Transposition: Outcomes



TGA '85-'89 "ASO"

TGA/IVS '74-'84
Recent "Mustard Era"

TGA/IVS '63-'73
"Mustard Era"

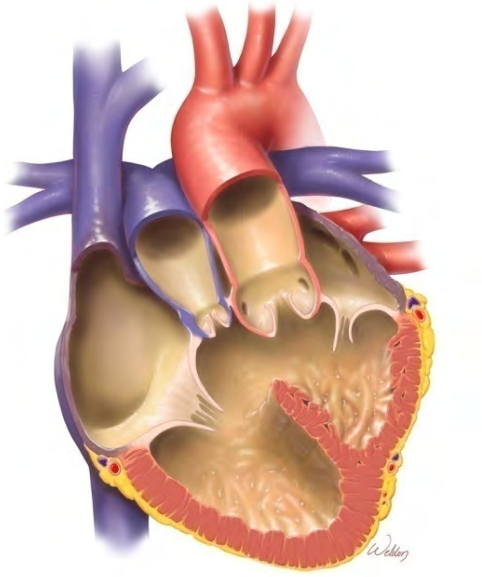
TGA/IVS
'57-'64

TGA
'57-'64

Paul & Wernovsky, Moss & Adams, 1995

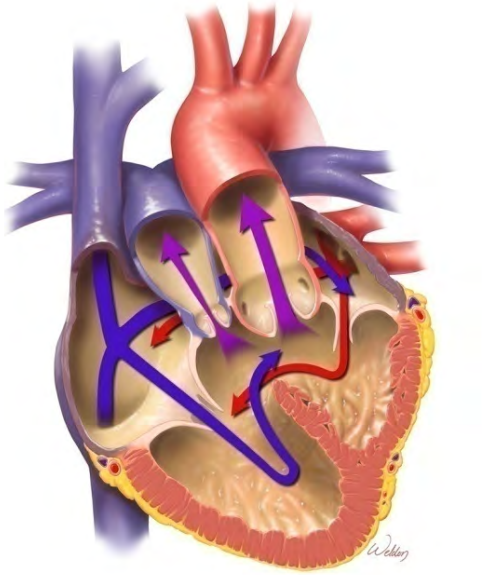
CCTGA

- 17 year old, 68 kg male
- CC-TGA, VSD, PS
- Heart failure
  - Decreased exercise tolerance
  - 3rd degree heart block
- Cardiac catheterization

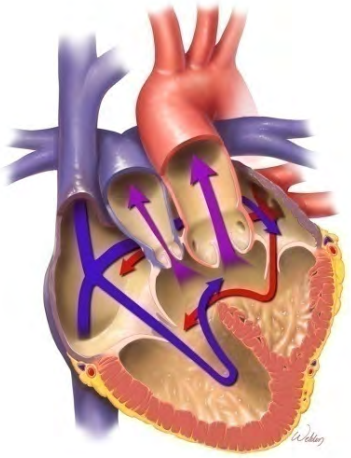


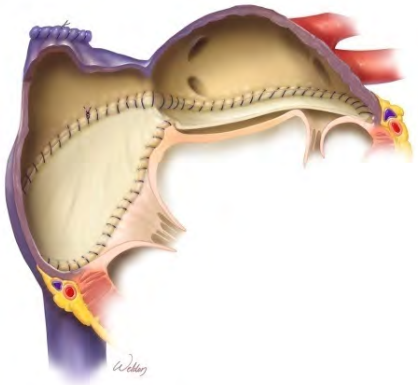
CCTGA

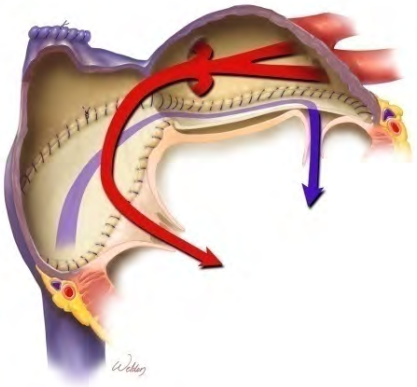
- RA →MV→LV→PA
- LA→TV→RV→Ao
- VSD (75%),
- PS (75%),
- Ebstein like anomaly (> 75%)

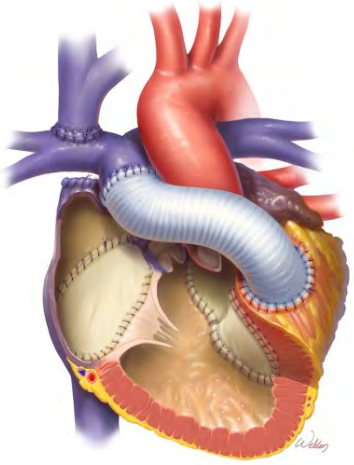


The Double Switch

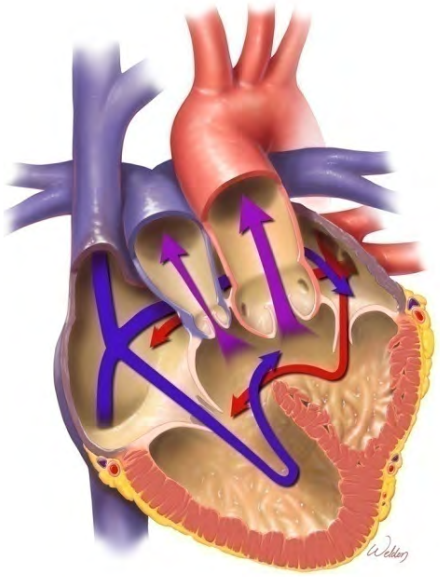


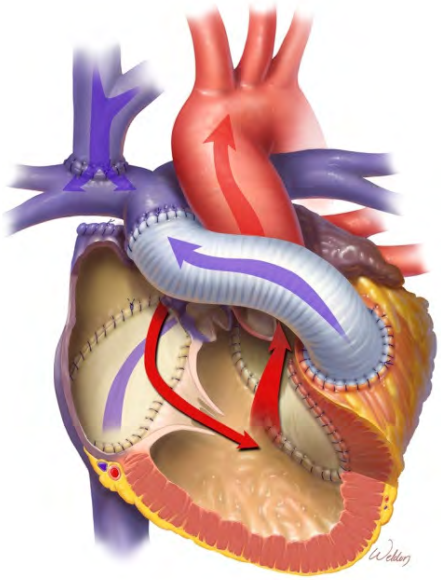






The Double Switch

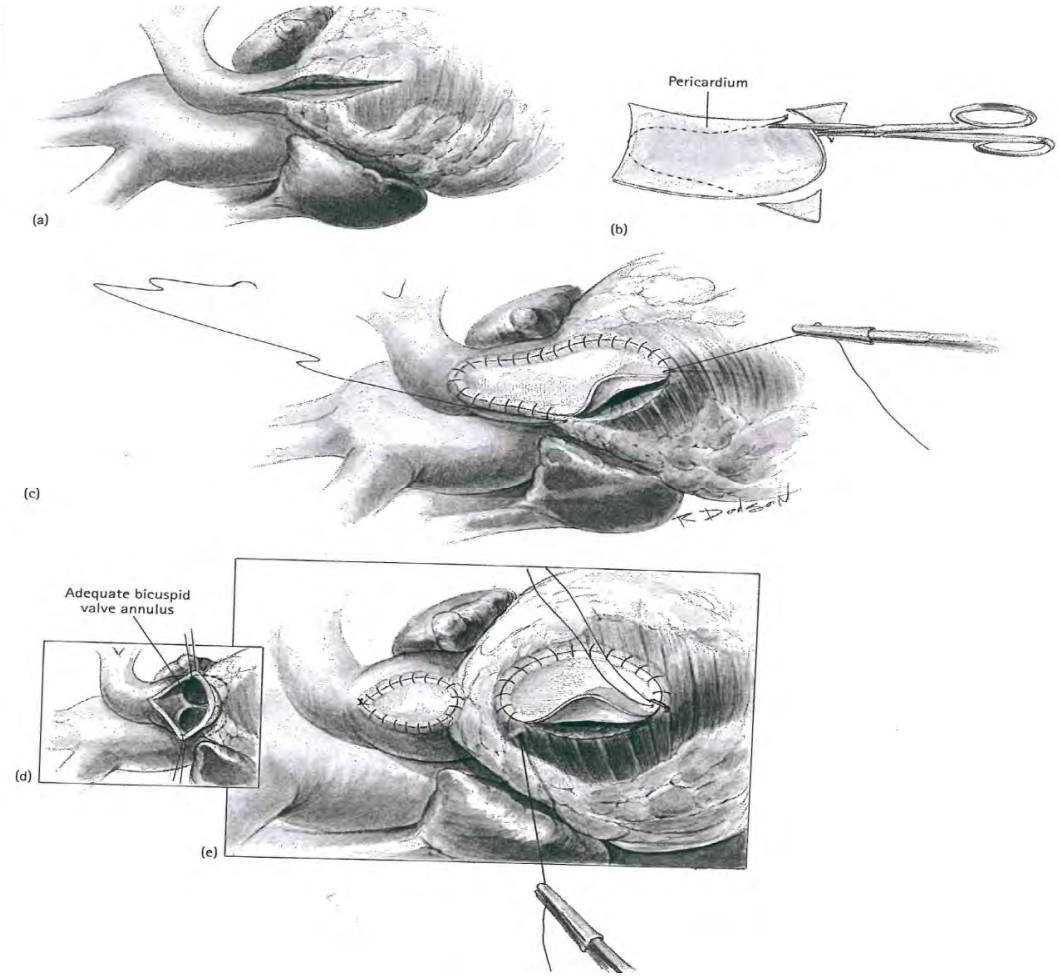




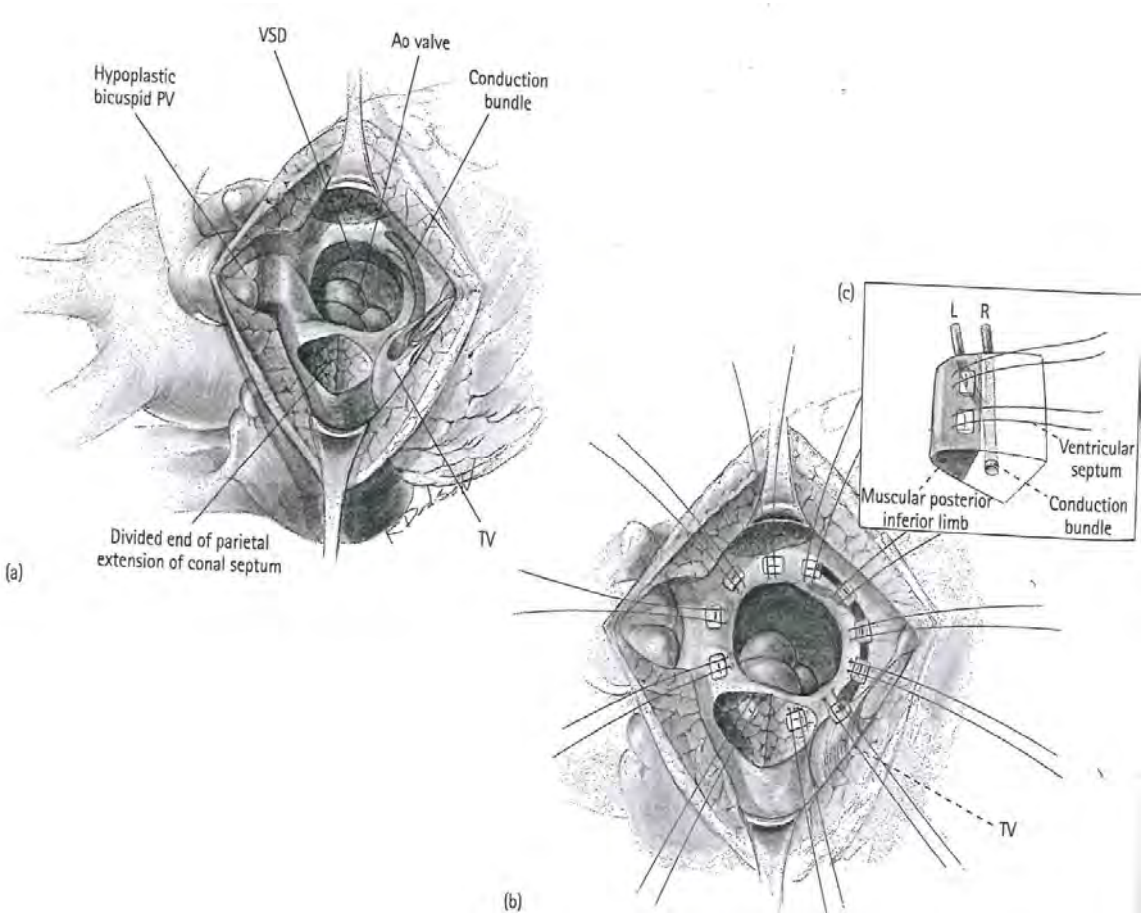
CCTGA

- Systemic (tricuspid) AV valve regurgitation → Valve replacement
- AV Block → Pacemaker
- End stage heart failure → Heart transplantation

Tetralogy of Fallot



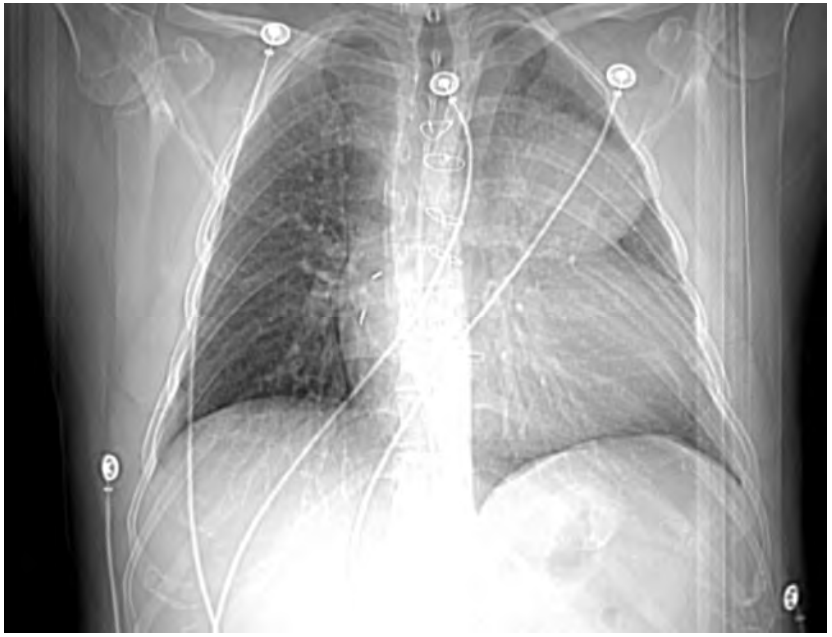
Complete repair



Indications for late reoperations

- Residual VSDs
- Residual or recurrent obstruction to pulmonary blood flow,
- Severe pulmonary insufficiency with progressive right ventricular dilatation and dysfunction

Post Rastelli repair- Pulmonary artery pseudoaneurysm




P

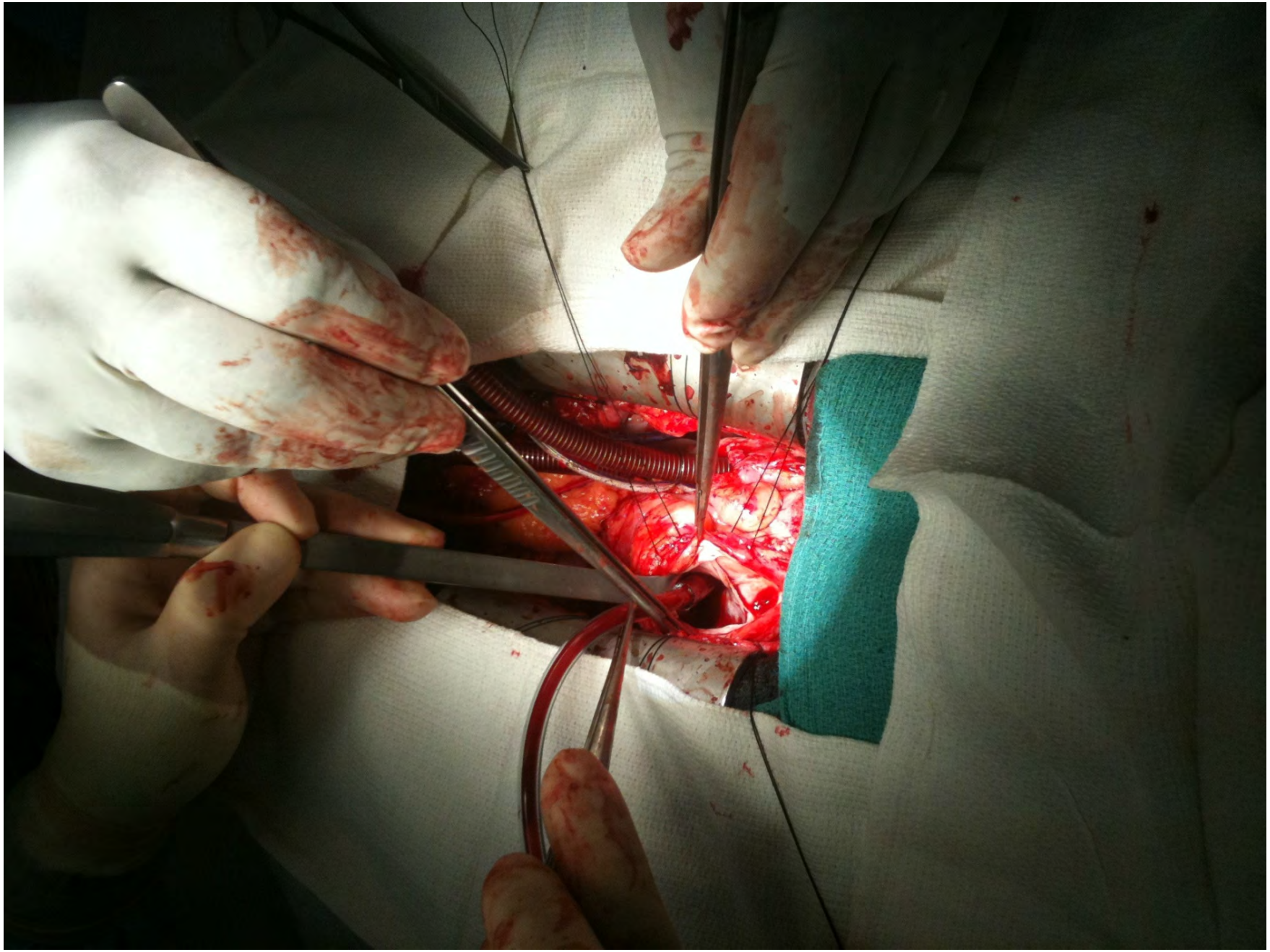

AM
P

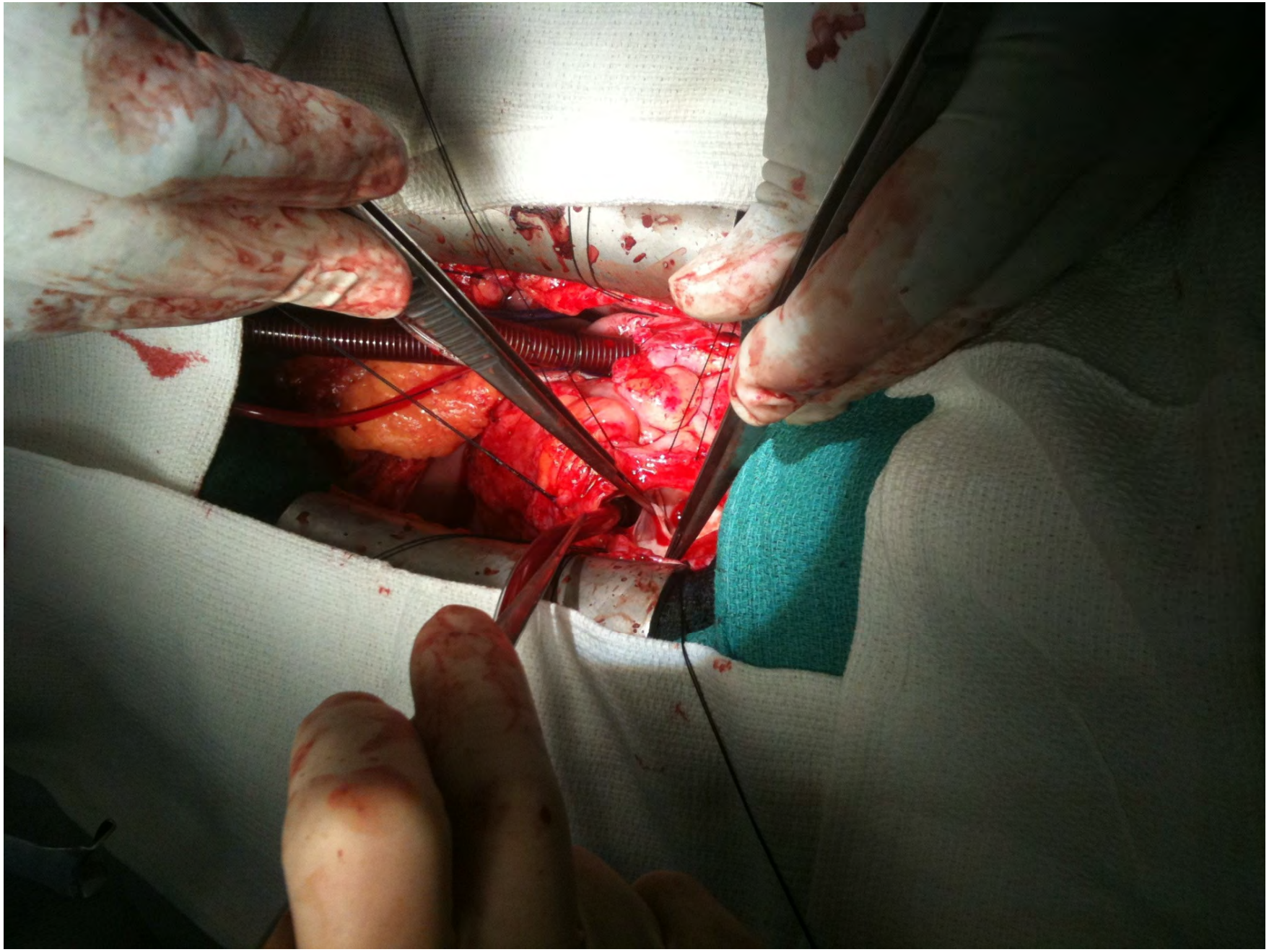

AM
P


86.3 mm
AM
P


F


F





Single Ventricle Palliation
FONTAN

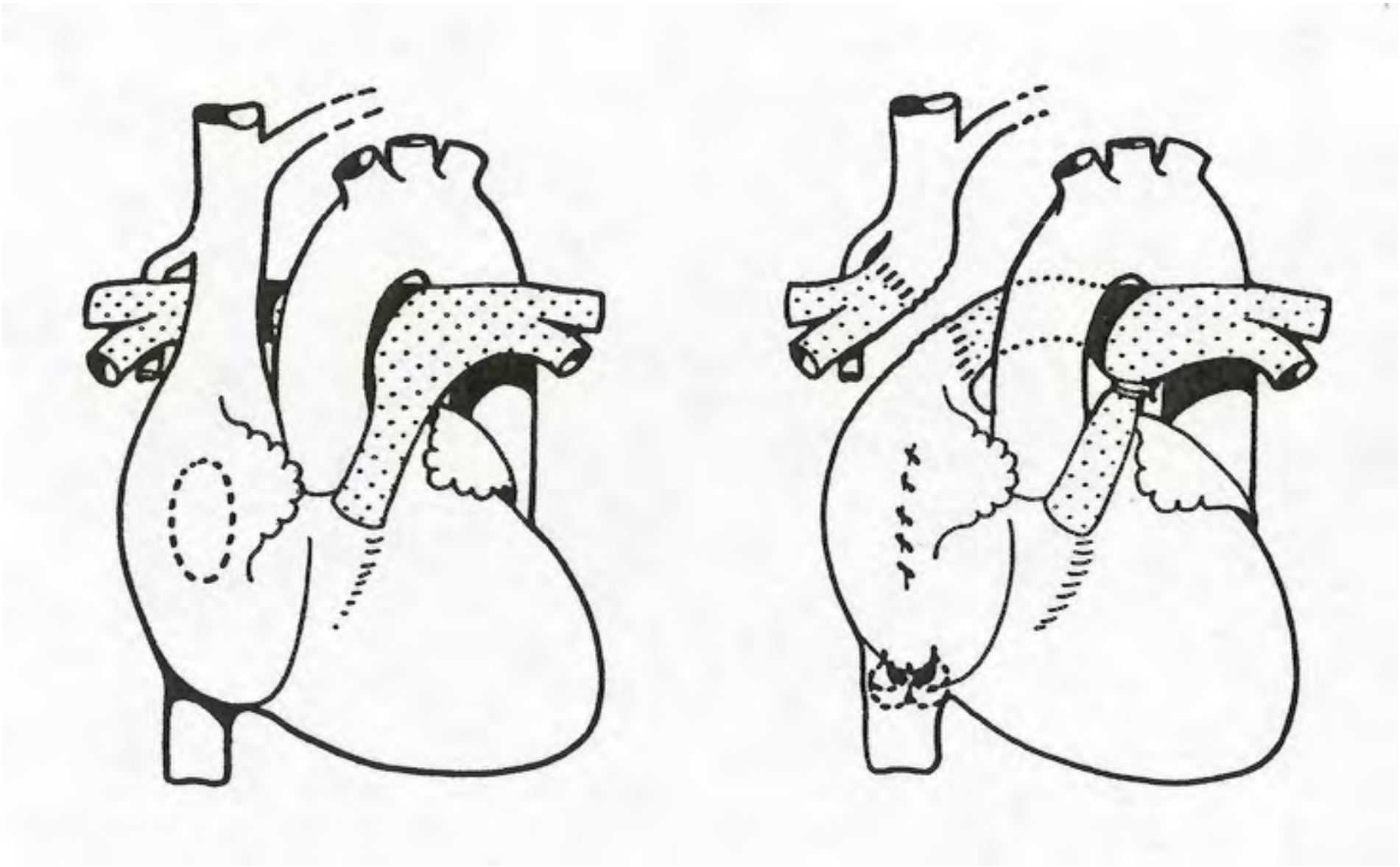
Elevated systemic venous pressure is the adequate driving force for pulmonary blood flow and a pumping right ventricle is not necessary

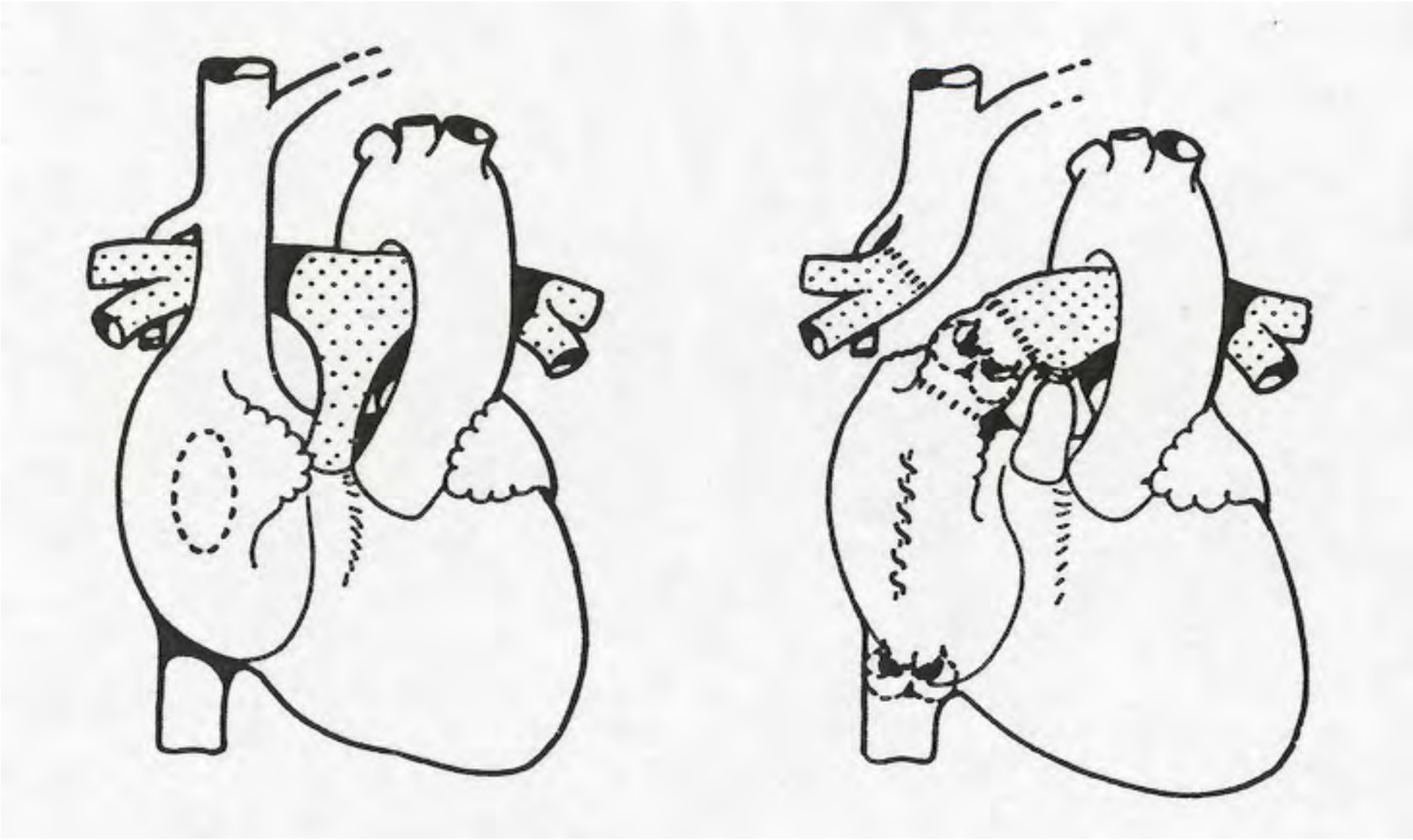
SURGICAL REPAIR OF TRICUSPID ATRESIA

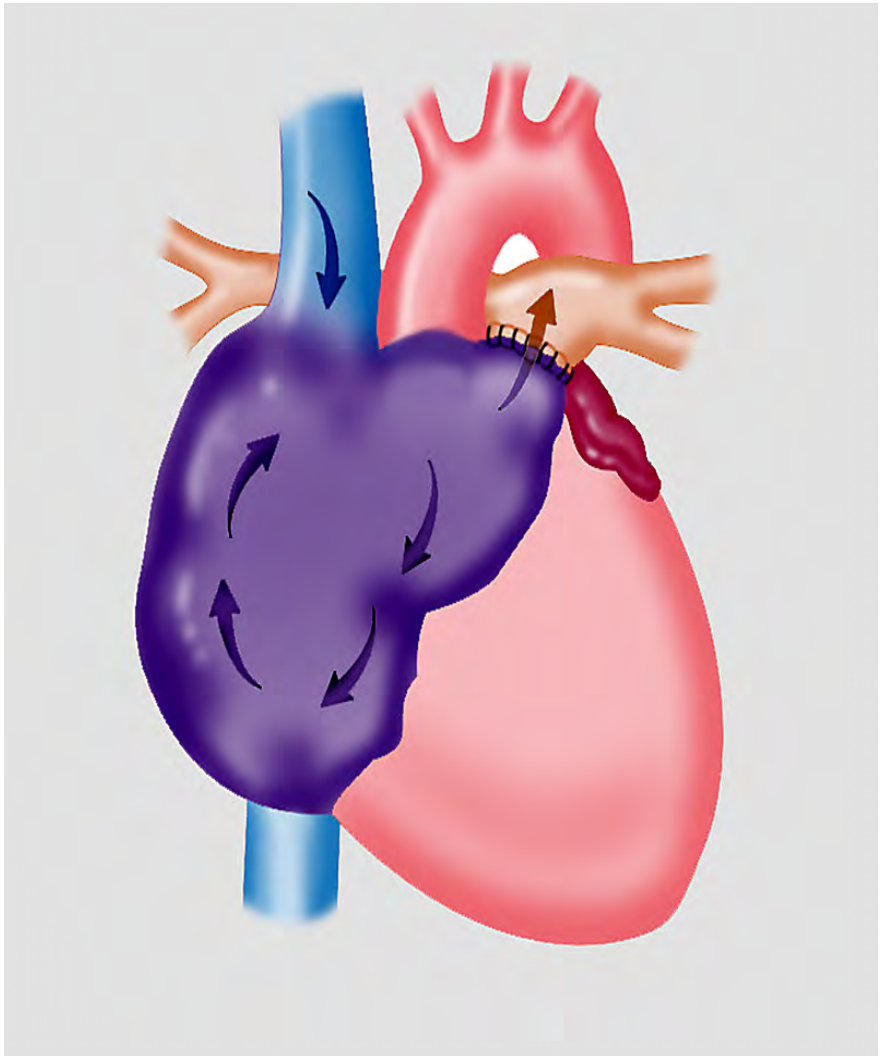
F. Fontan and E. Baudet

University of Bordeaux
Bordeaux, France

Thorax 1971;26:240-8



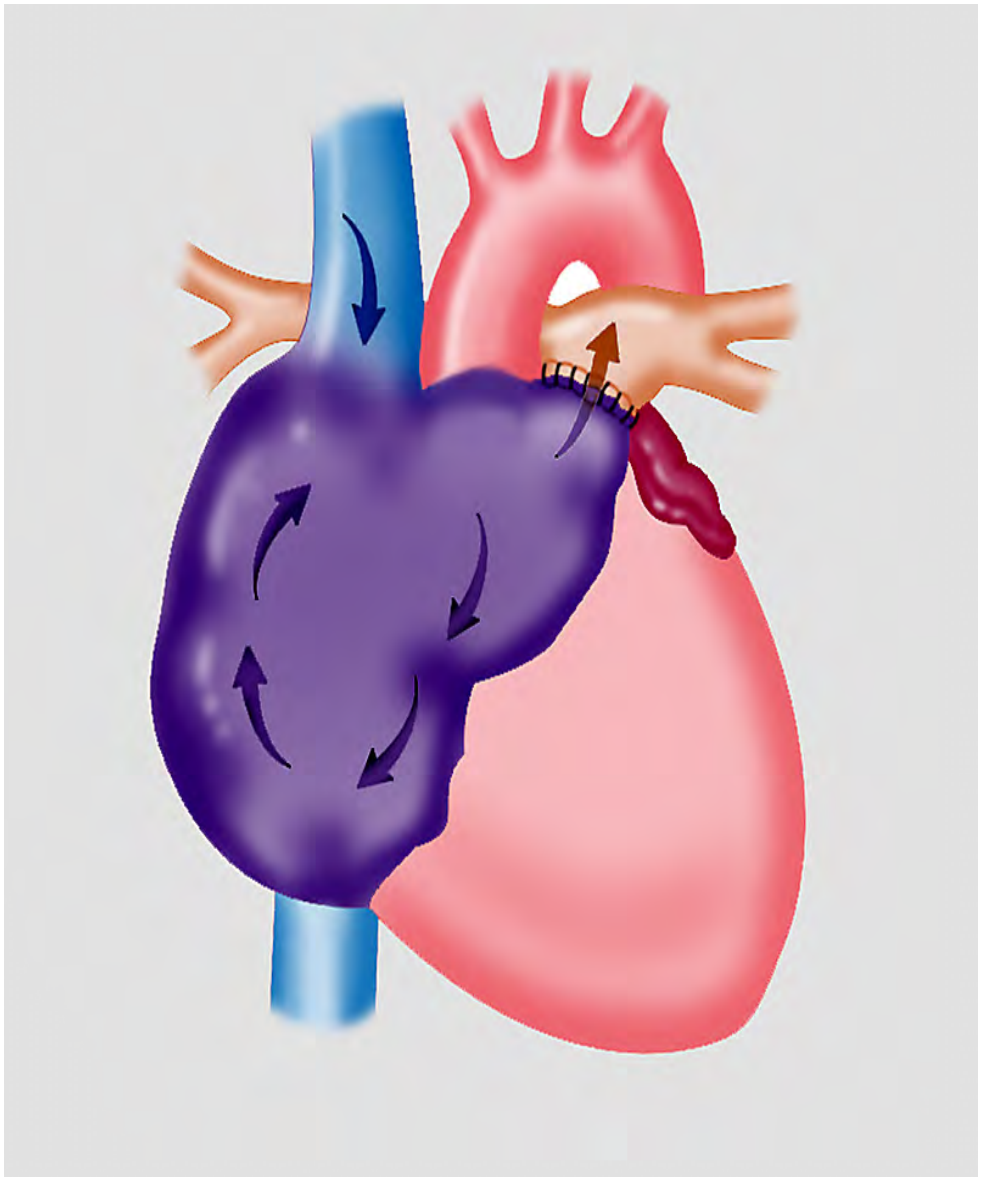




Turbulent Flow

Parietal Stasis

Thrombus Formation and Fontan circuit obstruction

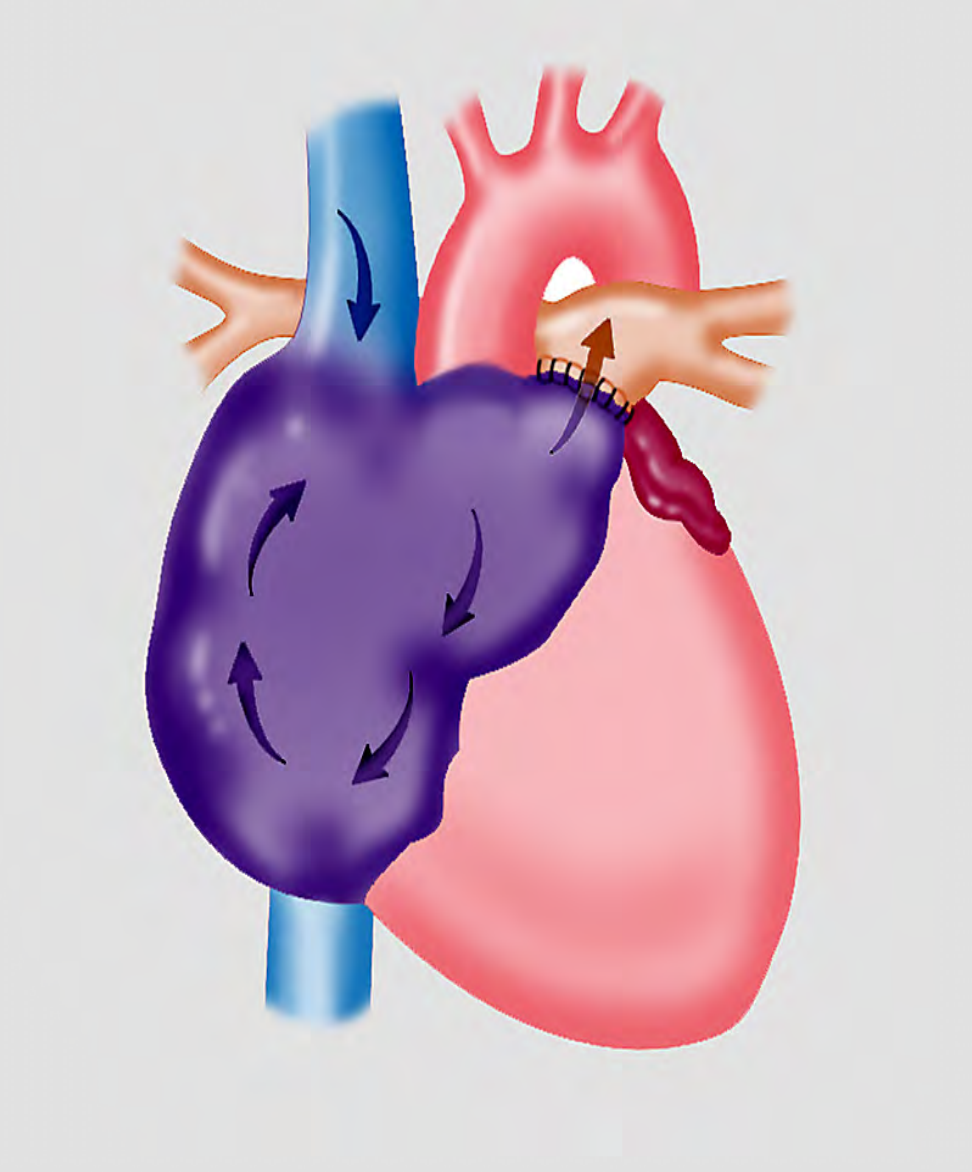


Elevated CVP

CS HTN

PV compression

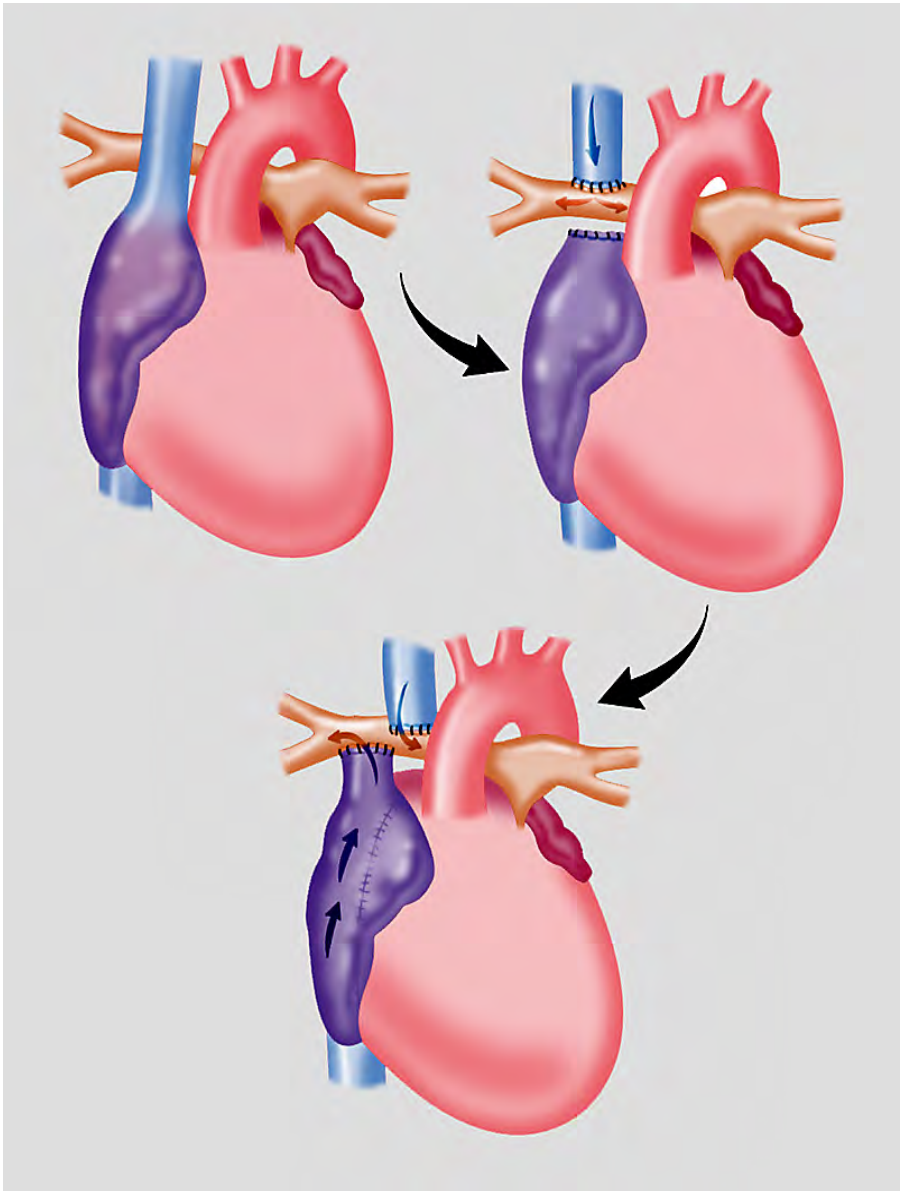
Atriopulmonary Connection



RA Dilation

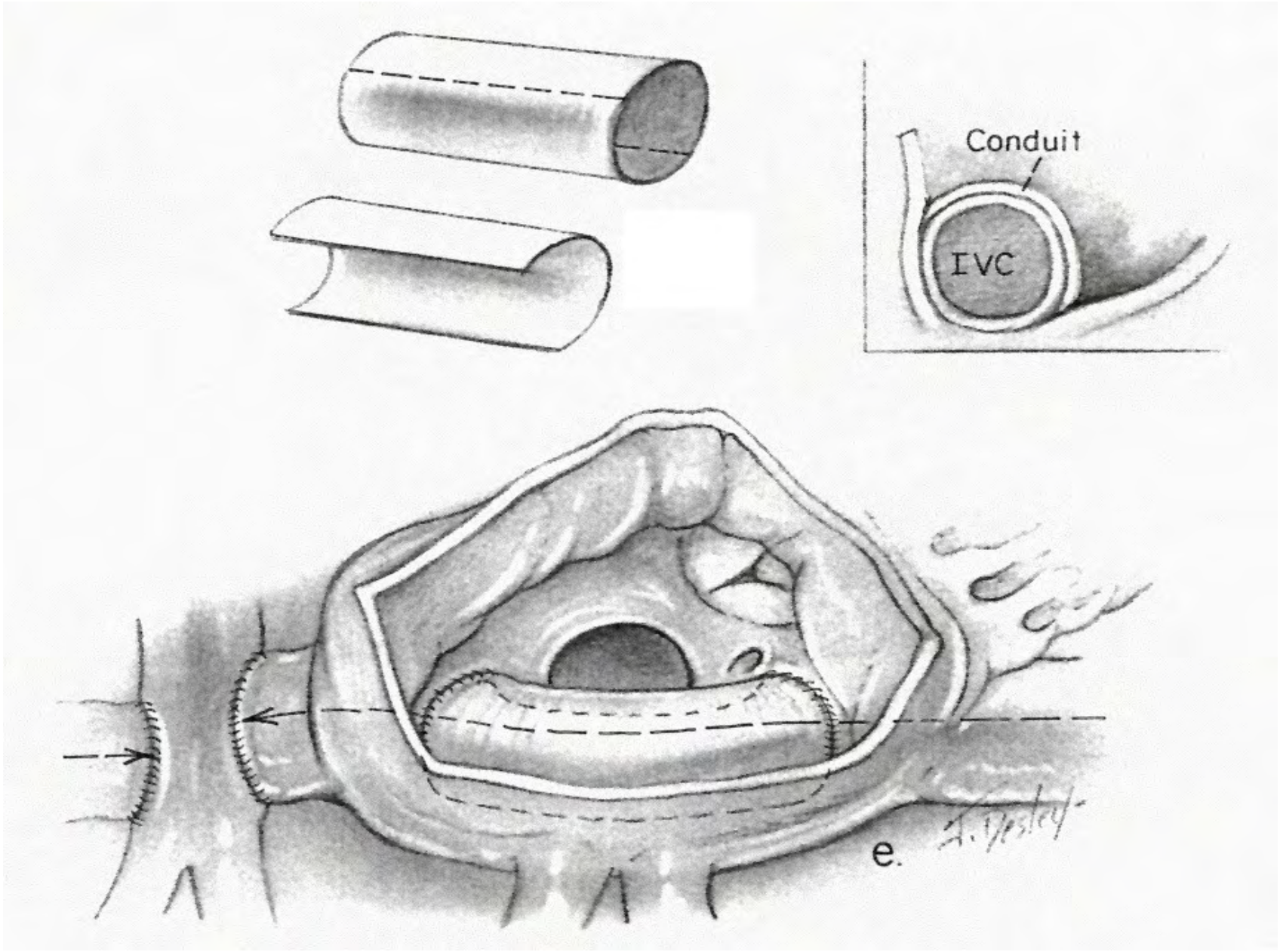
SV Arrhythmia

AV Synchrony and further compromise of ventricular function



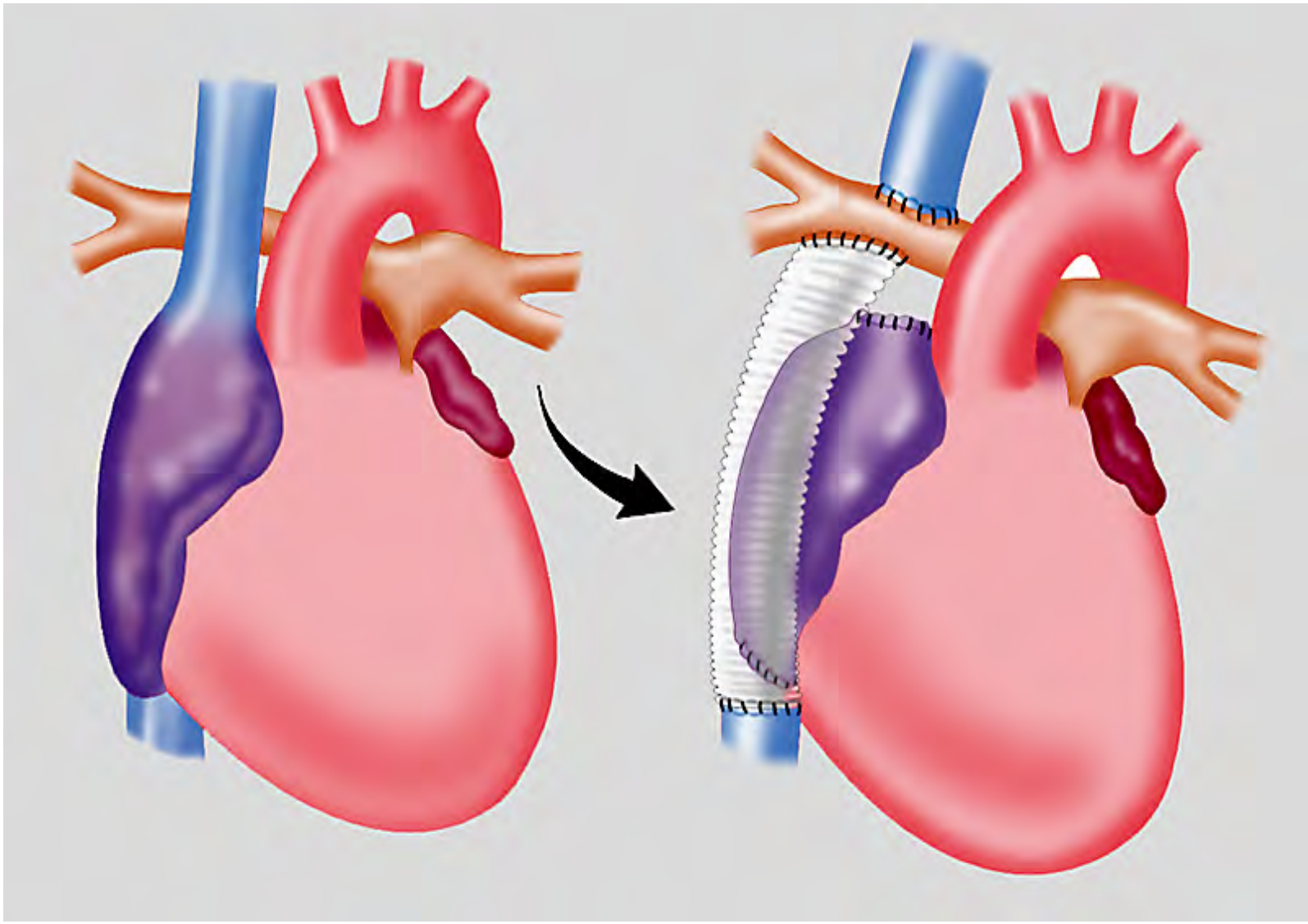
Lateral Tunnel TCPC;
Benefits

- Gold Standard for Fontan Completion
- Growth Potential
- Source of endothelium


Conduit
IVC
e.

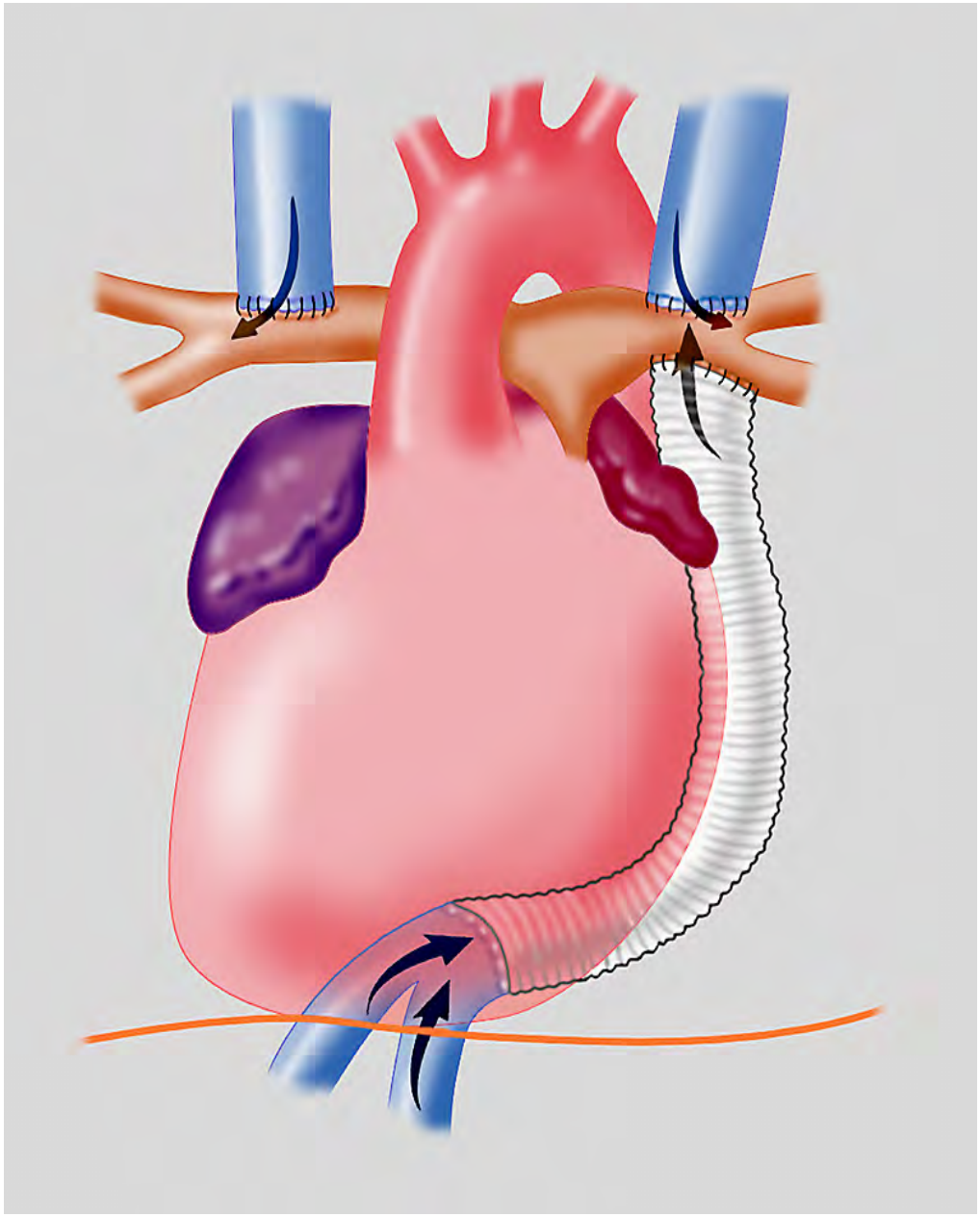
Lateral Tunnel TCPC;
The Downside

- Atriotomy (implies CPB, X-clamp)
  - Post CPB cardiovascular function
- Heavy suture load to the RA
- SA node proximity



Extracardiac Conduit TCPC; Benefits

- No atriotomy (no cross-clamp)
  - Warm, beating heart
  - Decreasing need for fenestration
- No atrial suture lines
- SA node



Extracardiac Conduit TCPC; The Downside

- Growth potential
  - Age of operation
- Conduit issues
  - Obstruction
  - Calcification,Degeneration

REVISION OF PREVIOUS FONTAN CONNECTIONS TO TOTAL EXTRACARDIAC CAVOPULMONARY ANASTOMOSIS: A MULTICENTER EXPERIENCE

Carlo F. Marcelletti, MD[a]
Frank L. Hanley, MD[b]
Constantine Mavroudis, MD[c]
Doff B. McElhinney, MD[b]
Raul F. Abella, MD[a]
Stefano M. Marianeschi, MD[a]
Francesco Seddio, MD[a]
V. Mohan Reddy, MD[b]
Ed Petrossian, MD[b]
Teresa de la Torre, MD[a]
Luisa Colagrande, MD[a]
Carl L. Backer, MD[c]
Adriano Cipriani, MD[a]
Fiore S. Iorio, MD[a]
Francis Fontan, MD[d]

J Thorac Cardiovasc Surg 2000;119:340-6

Patients

- 31 patients (19.9±8.8 year old)
- AP (21), AV (1), LT (1)
  - 20 had preop tachyarrhythmia
  - 3 had 1st or 2nd AV block
  - 5 had PM (complete AV block)

Technique

- Conversion to ECC TPCP (no fen!)
- 68% aggressive reduction atrioplasty
- 6 mapping/cryoablation/PM
- 2 right atrial modified maze

Results

- 2 operative deaths
- 1 late death ,1 transplant (RHYTHM)
- All survivors has improved functional status (n=20 NYHA I, n=7 NYHA II)
  - 4 DCEPM, 6 persistence of preop arrhythmia, 1 new A-fib (11/29)
  - All those having rhythm control surgery were controlled, only 1 required medication

Total cavopulmonary conversion and maze procedure for patients with failure of the Fontan operation

Constantine Mavroudis, MD
Carl L. Backer, MD
Barbara J. Deal, MD
Christopher Johnsrude, MD
Janette Strasburger, MD

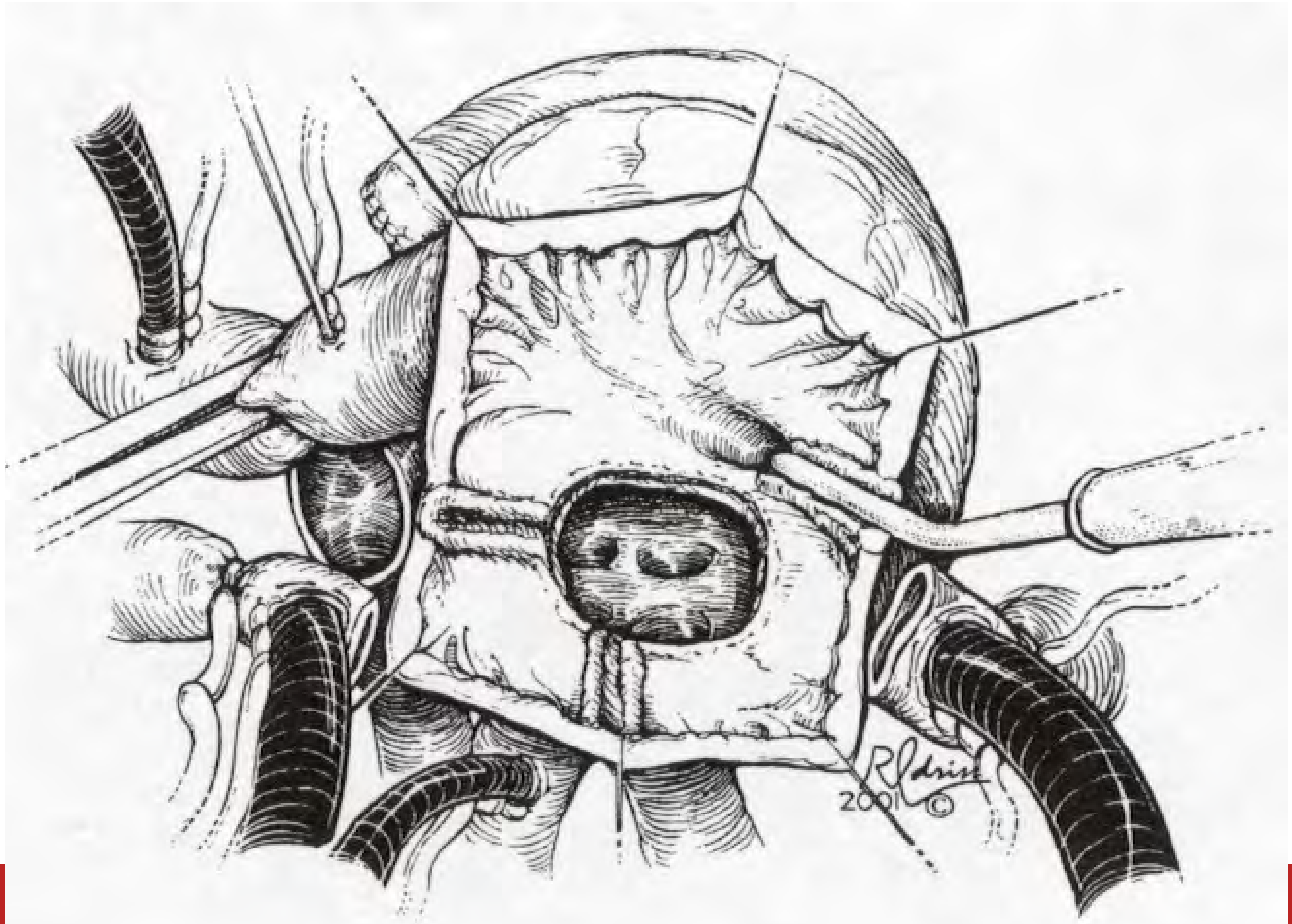
J Thorac Cardiovasc Surg 2001;122:863-71

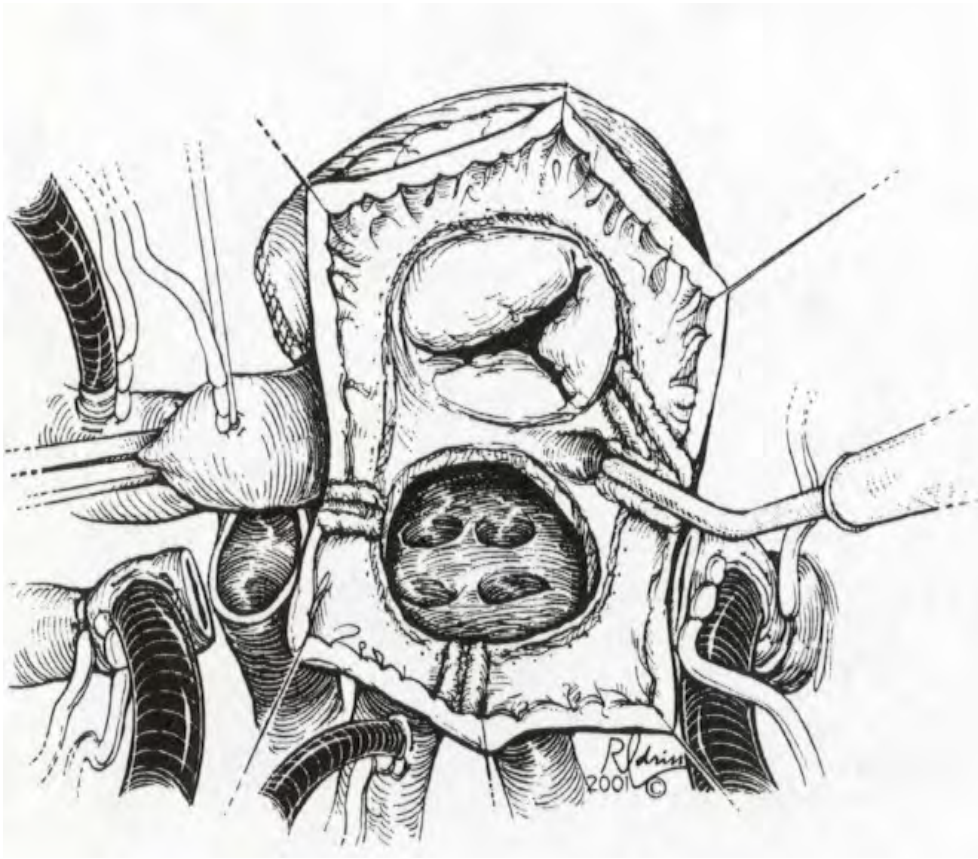
Patients

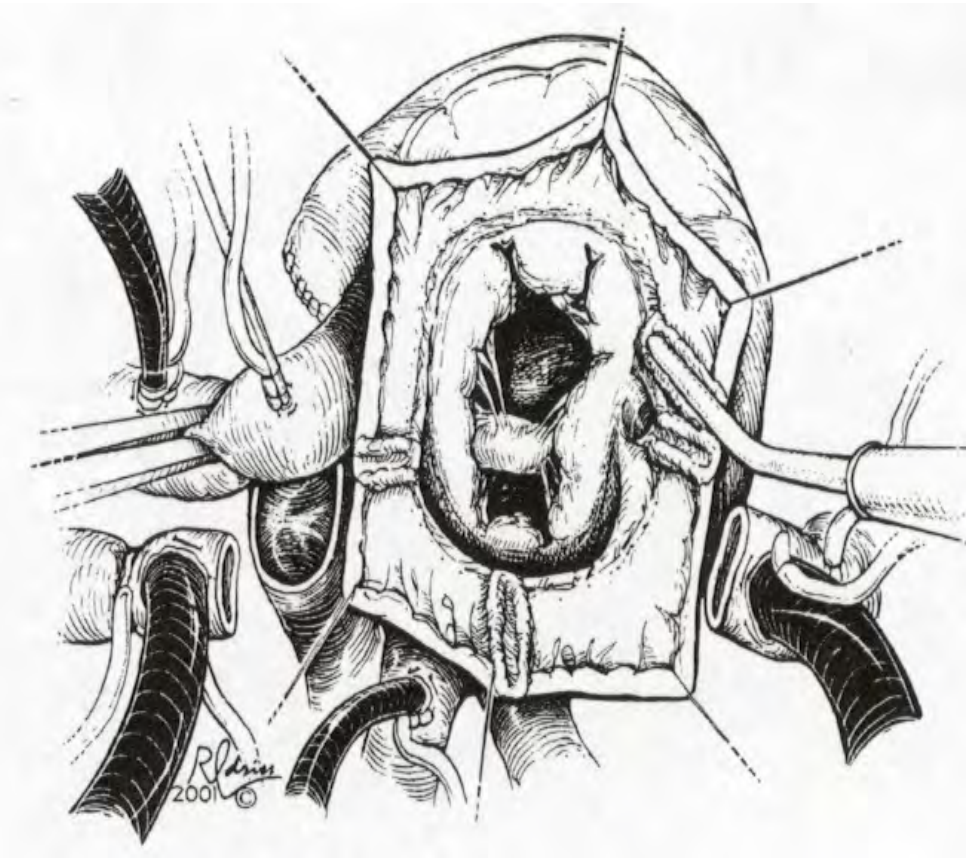
- 40 patients (18.7±9.0 years old)
- AP (31), AV (6), LT (2), IAG (1)
  - all but 2 had rhythm disturbance

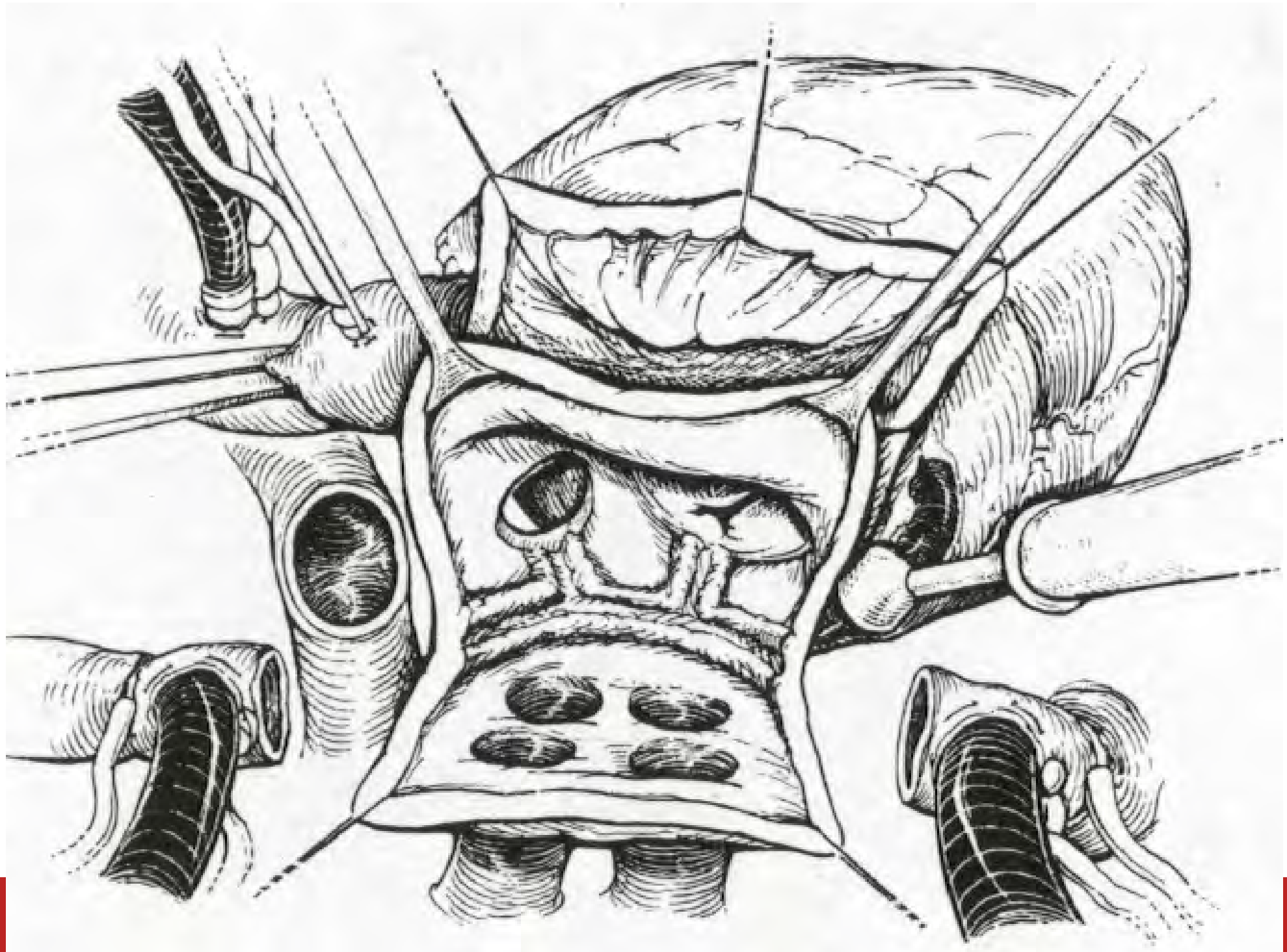
Technique

- 38 TCPC (32 ECC, 6LT), 1 RA-PA anastamotic revision, 1 “one and a half” ventricular repair
- 40 had arrhythmia procedures
- 38 pacemakers (33 atrial, 5 DC)


2001 ©







Results

- No surgical deaths, 1 late death (MI)
- 5 patients with tachyarrhythmia (12.5%), 4 (10%) require medication
- 0 patients undergoing modified maze, 0 patients undergoing maze-Cox III have rhythm disturbance

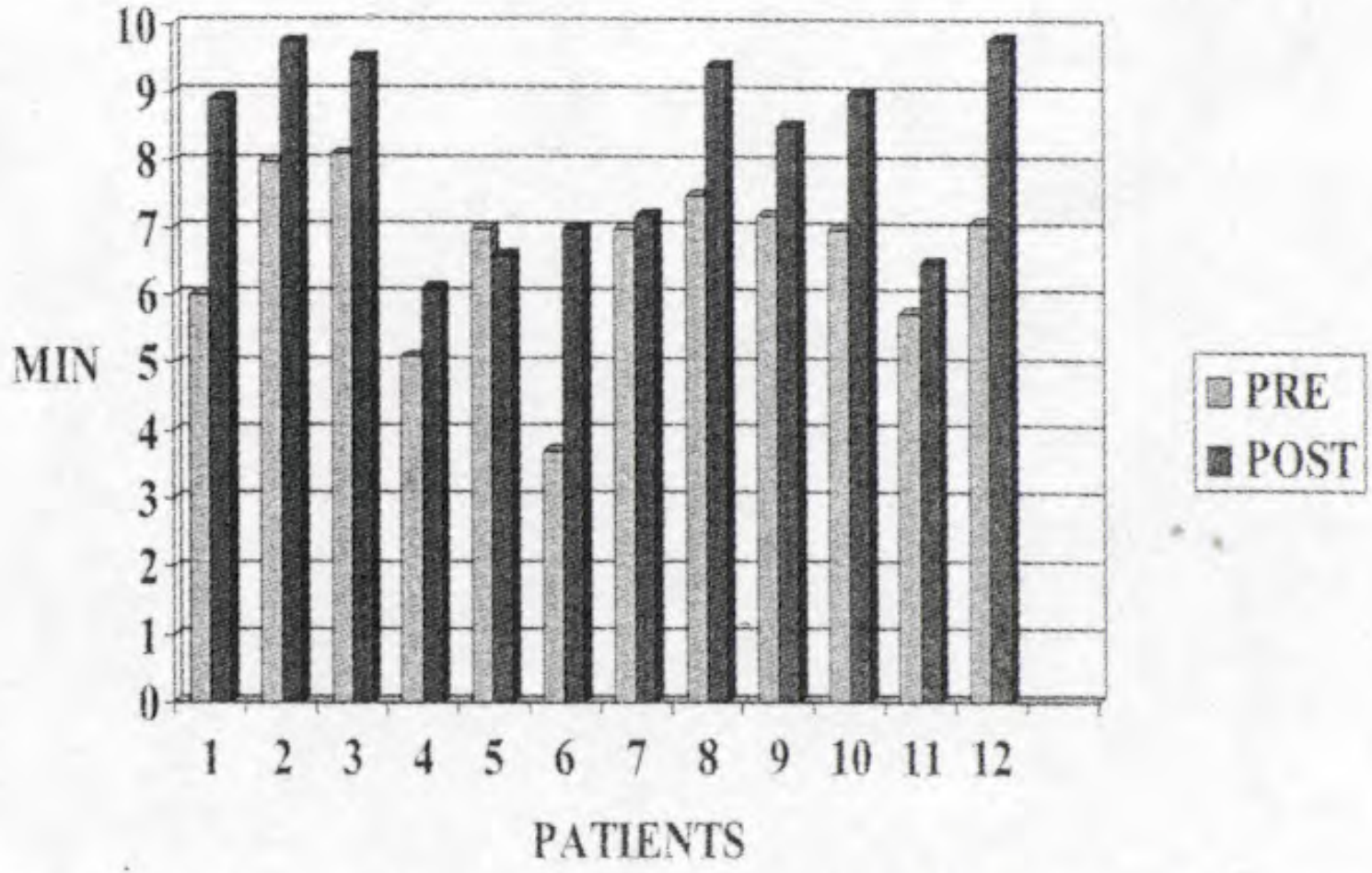
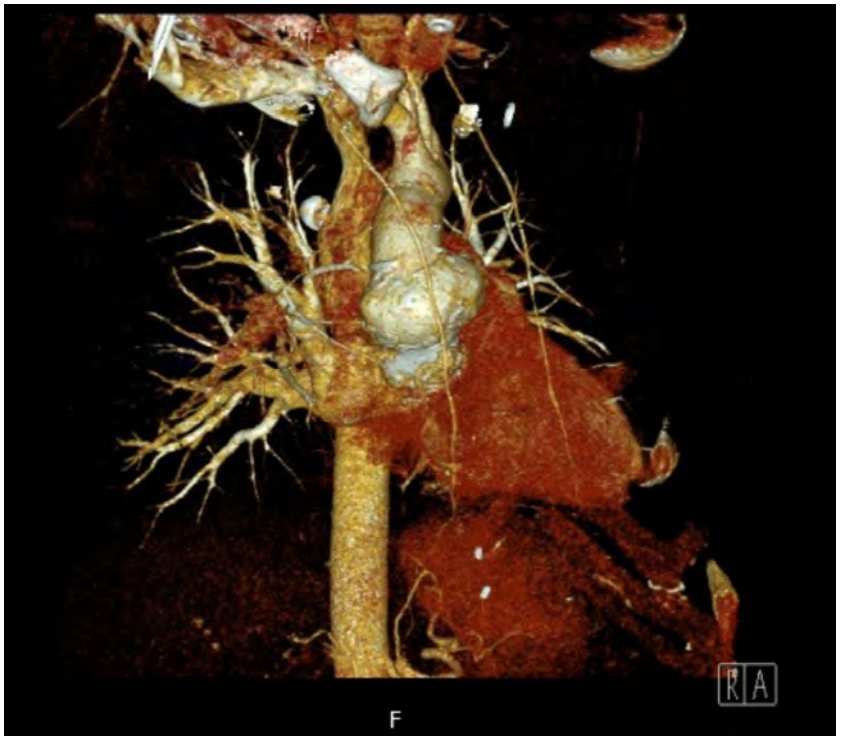


Figure 6. Bar graph showing the results of exercise tolerance (Bruce protocol) before and after Fontan conversion and arrhythmia surgery ($P < .05$; Student *t* test).

Degenerations of Homografts


PM
P

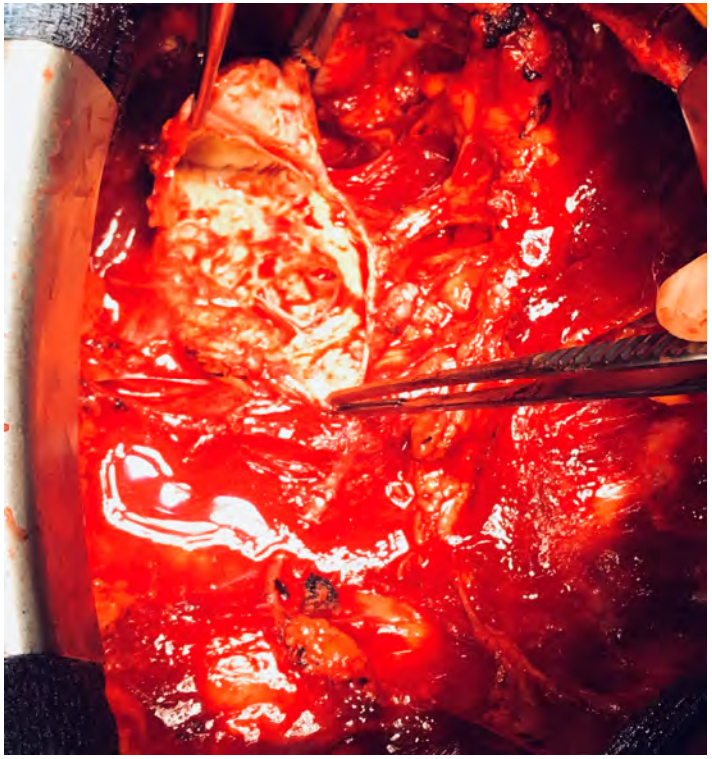

PM
F
L

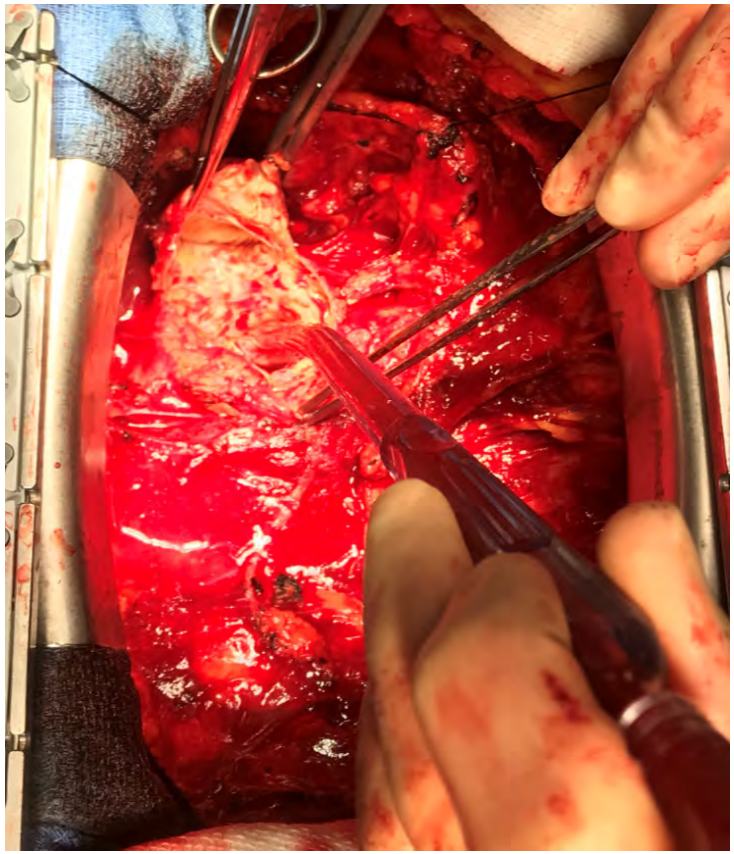

RA
F

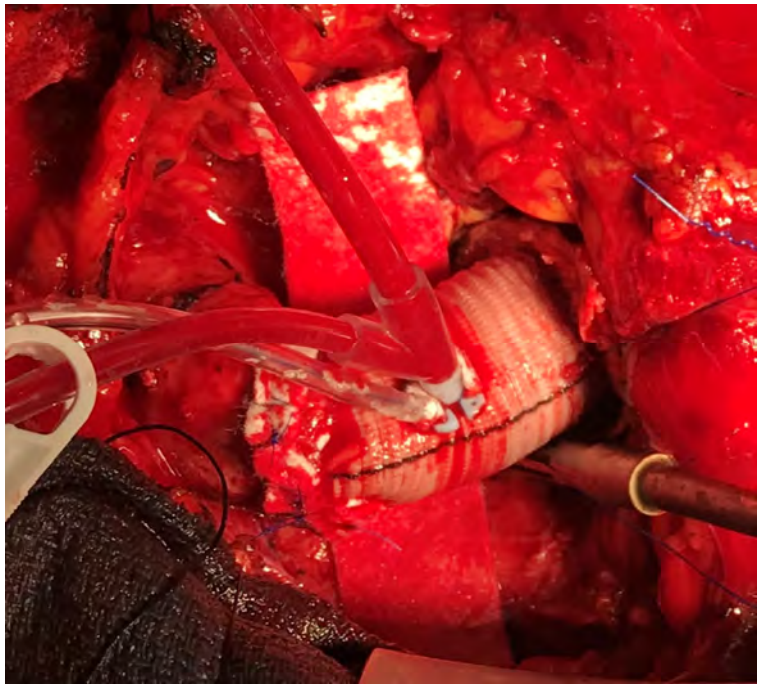

A
F


55.4 mm
P

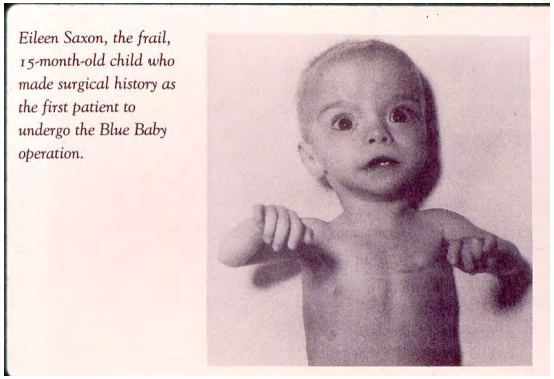

P







All patients do need follow-up monitoring because there are long-term residua and sequelae associated with repair of nearly all the defects



"If I have seen farther, it is by standing on the shoulders of giants"

Isaac Newton to Robert Hooke - 1676

Oklahoma Heart Institute

Thank you