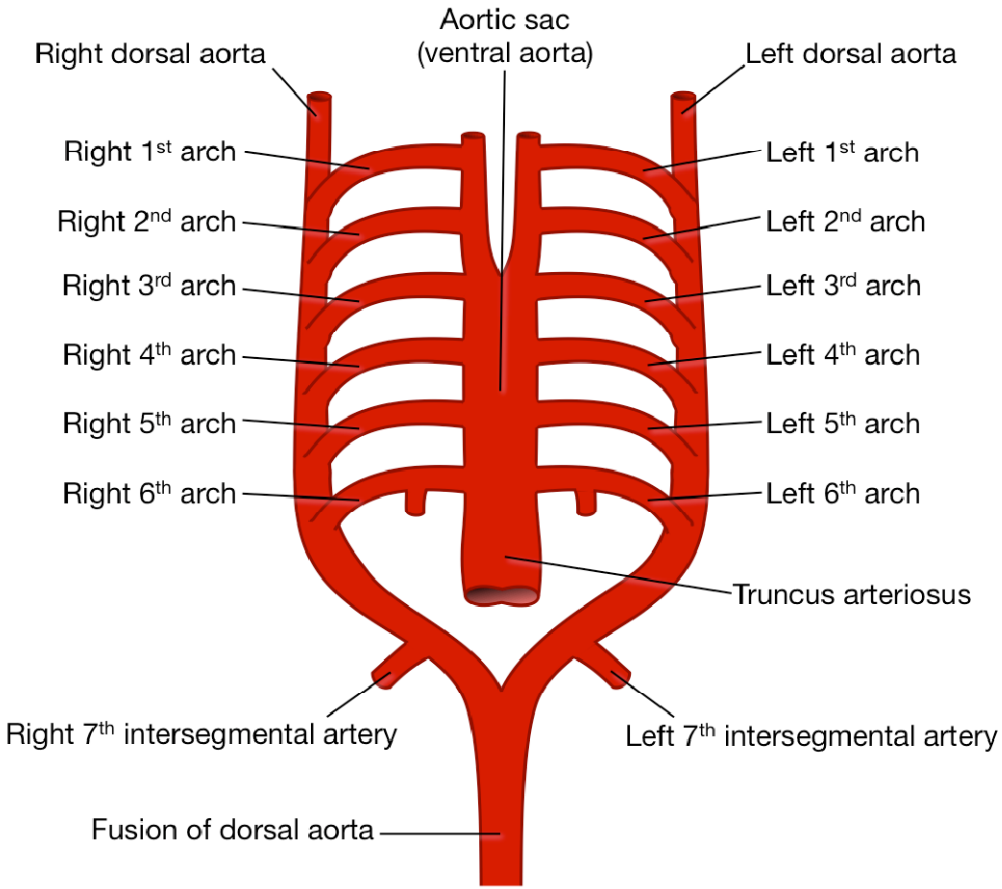
Aortic arch obstruction

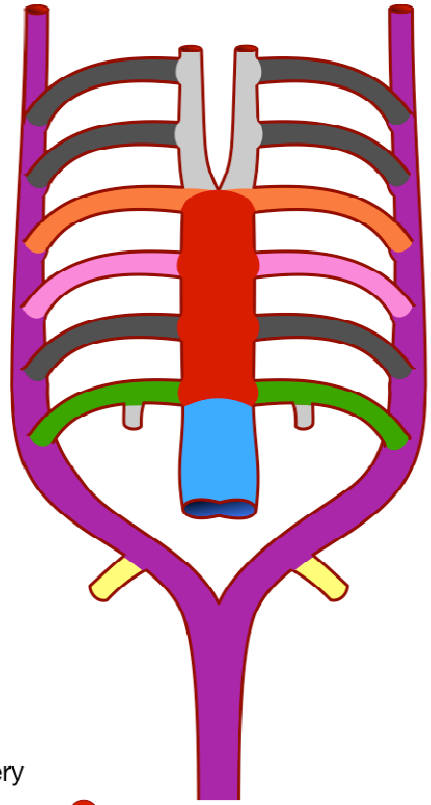
Kwang Ho Choi, M.D.
Department of Thoracic and Cardiovascular Surgery
Pusan National University Yangsan Hospital

Development of the aortic arch and branches



Embryonic pharyngeal arch arteries

Development of the aortic arch and branches



- 3rd Pharyngeal arch artery
- 4th Pharyngeal arch artery
- 6th Pharyngeal arch artery
- 7th Intersegmental artery
- Aortic sac
- Truncus arteriosus
- Dorsal aorta

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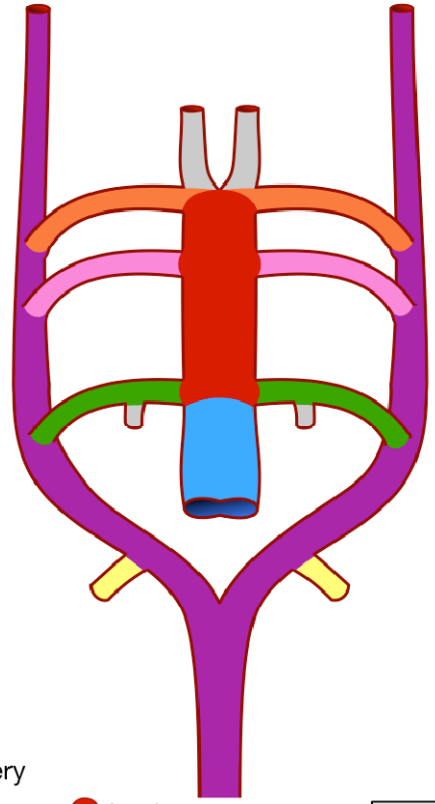
Development of the aortic arch and branches



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6 WEEKS

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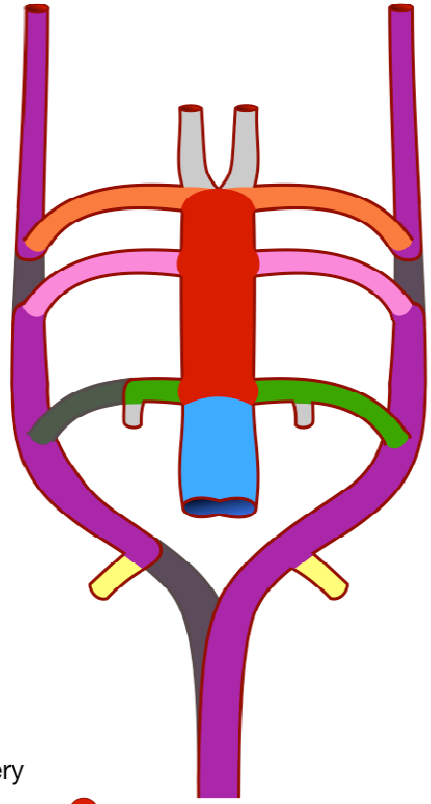

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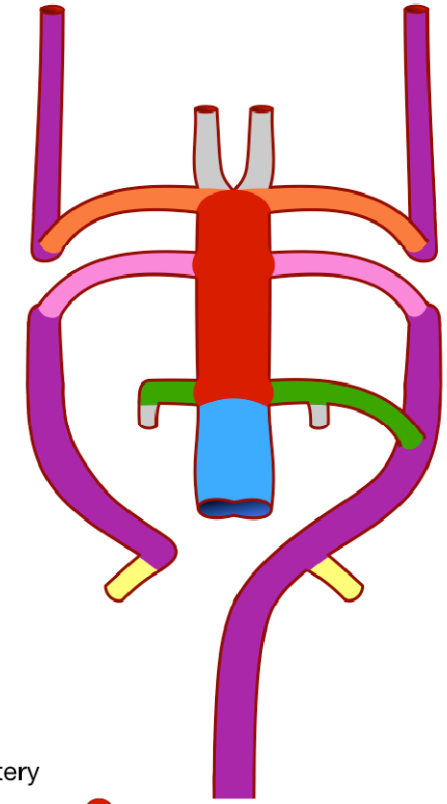

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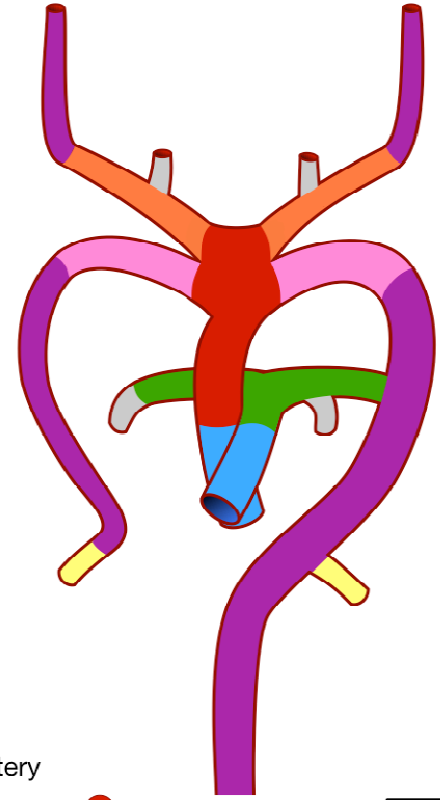

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7 WEEKS

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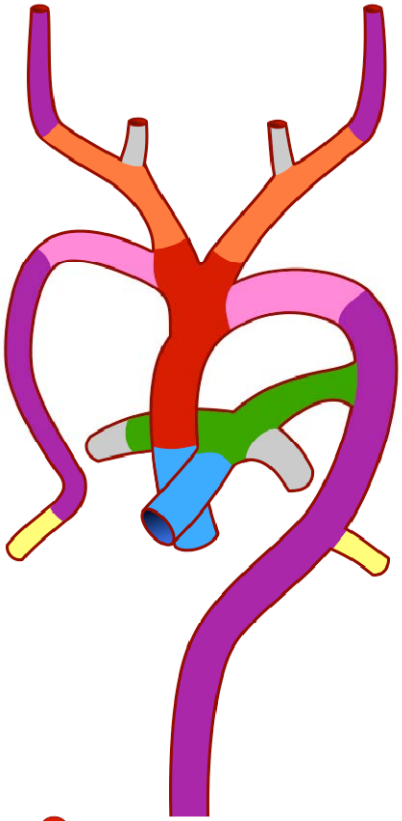

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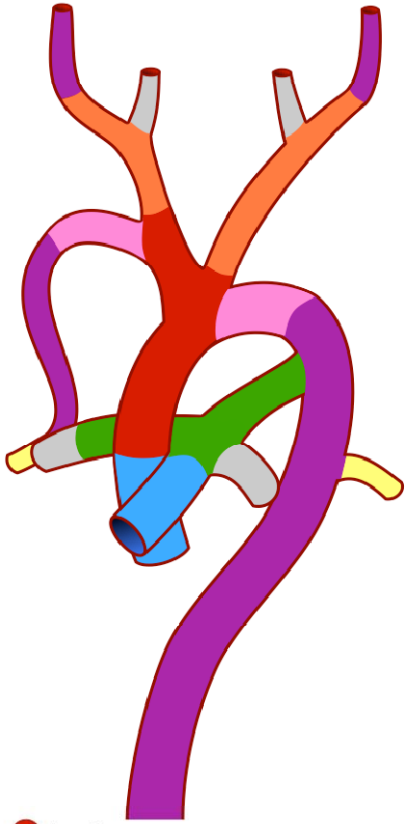

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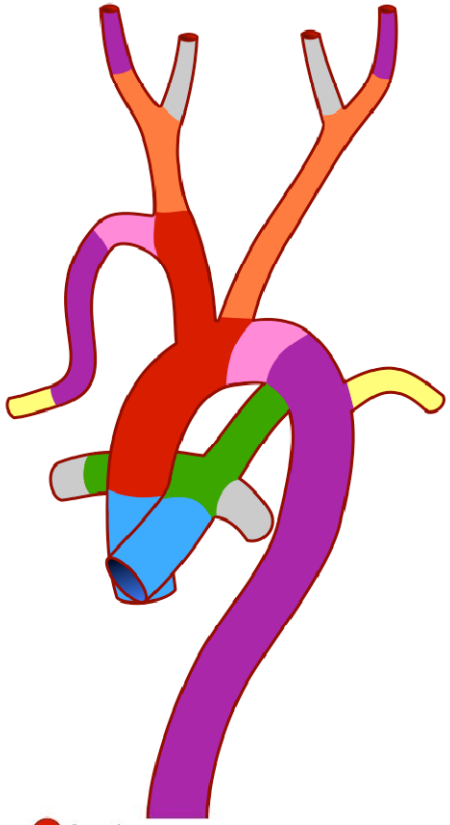

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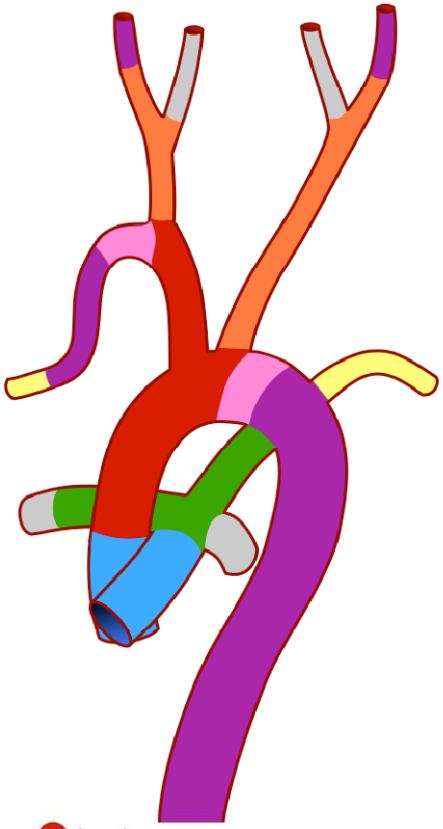

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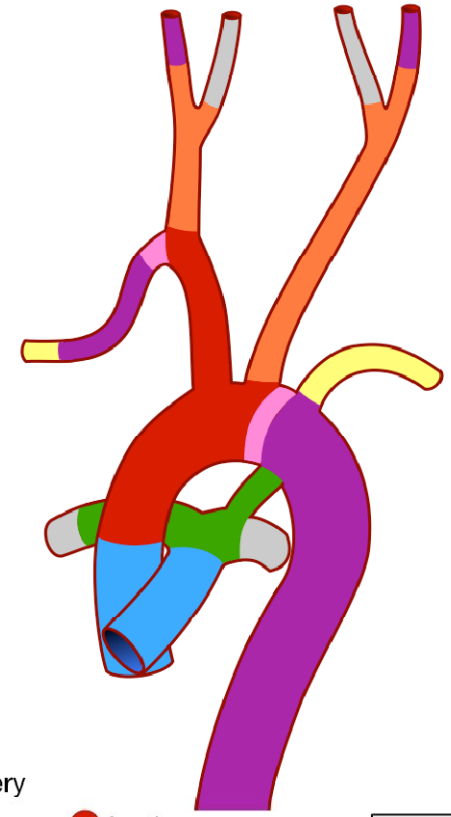

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8 WEEKS

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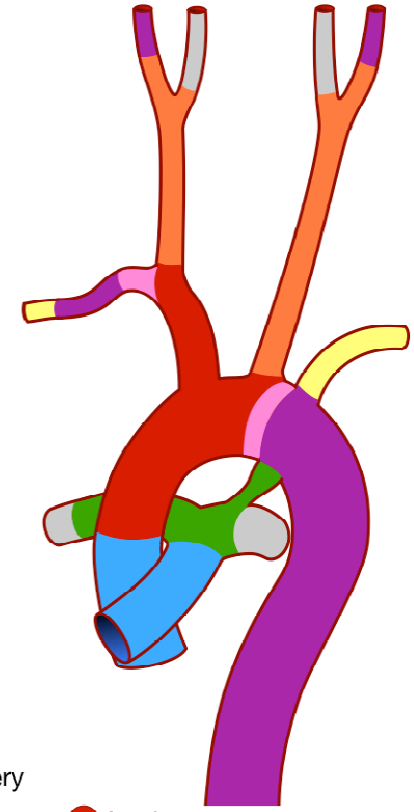

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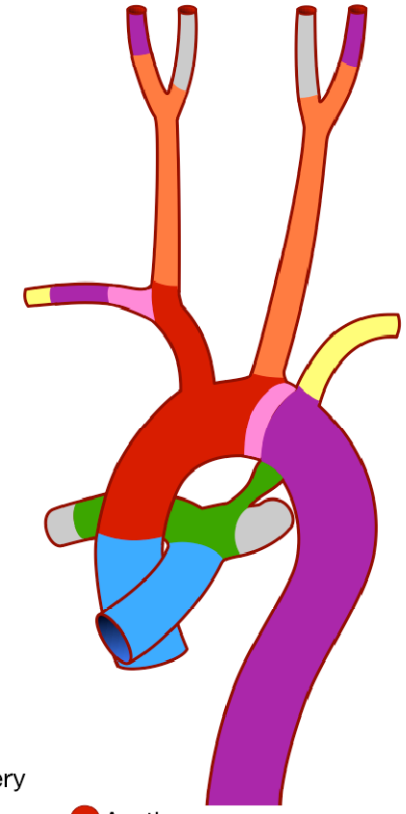

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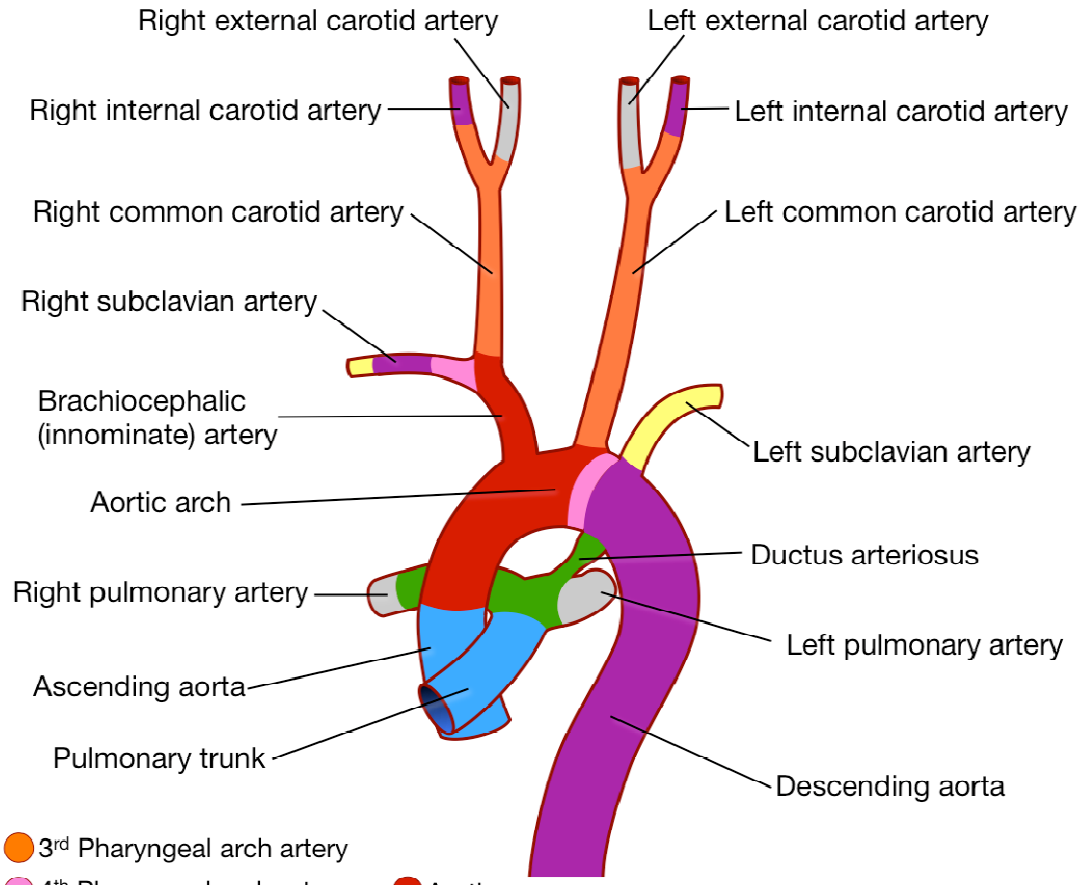

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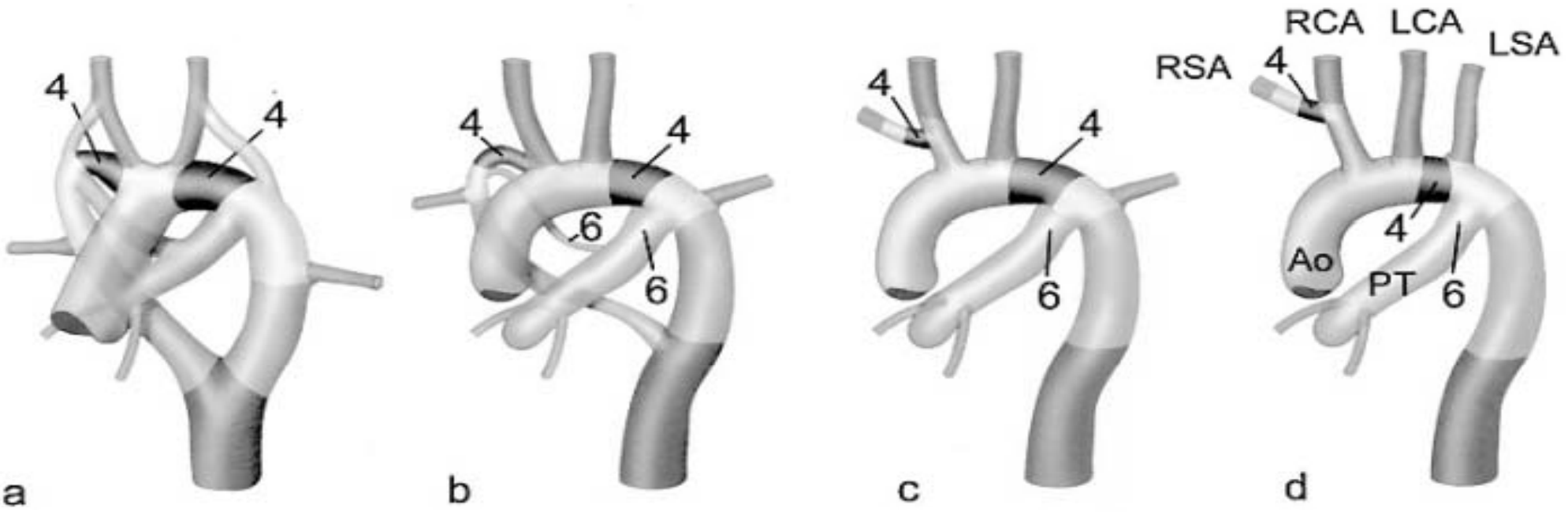
Coarctation of Aorta (CoA)

Coarctation of Aorta

- Definition
  - Congenital narrowing of the descending thoracic aorta
  - Usually occurring just distal to the LSCA
  - Site of insertion of the ductus arteriosus (ligamentum arteriosum)

Coarctation of Aorta

Abnormal development of left 4th and 6th aortic arches

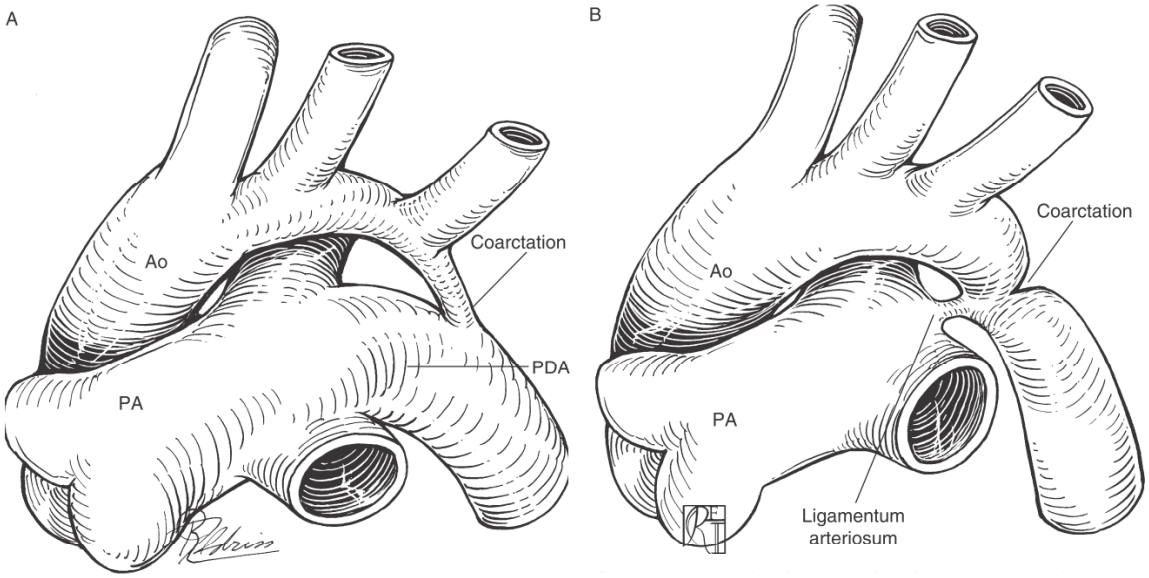


Gittenberger-De Groot, A.C. Bartelings, M.M. Deruiter, M.C. Poelmann, R.E. Basics of Cardiac Development for the Understanding of Congenital Heart Malformations. *Pediatric Research.* 2005; 57 (2): 169-176.

- Prevalence
  - 0.2 ~ 0.6 / 1000 live births (8^{th})
  - 5 ~ 8 % of cases of congenital heart disease
- Associated anomaly
  - Patent ductus arteriosus, bicuspid aortic valve, ventricular septal defect, mitral valve abnormalities
- Clinical presentation
  - From cardiovascular collapse in neonate after ductal closure to asymptomatic hypertension in adult

Embryology and anatomy

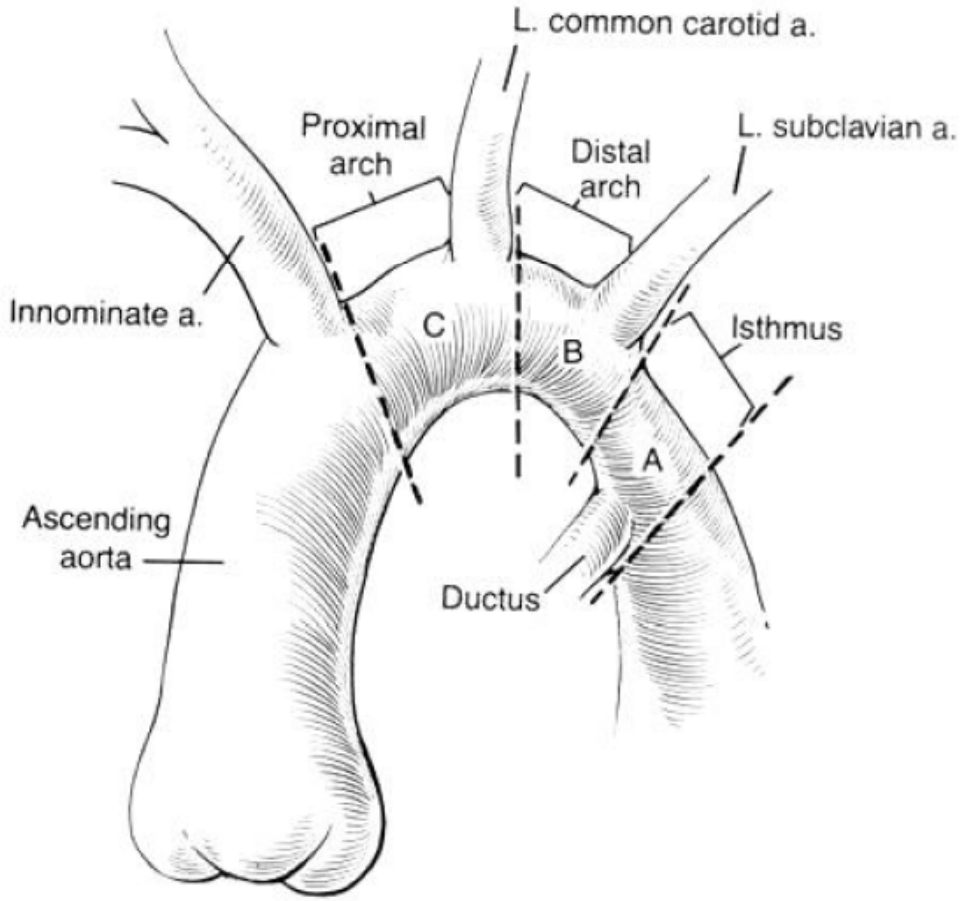
- Morgagni in 1760 : localized constriction of descending aorta
- Bonnet in 1903
  - Infantile vs. adult type
  - Key differences : patent of ductus arteriosus



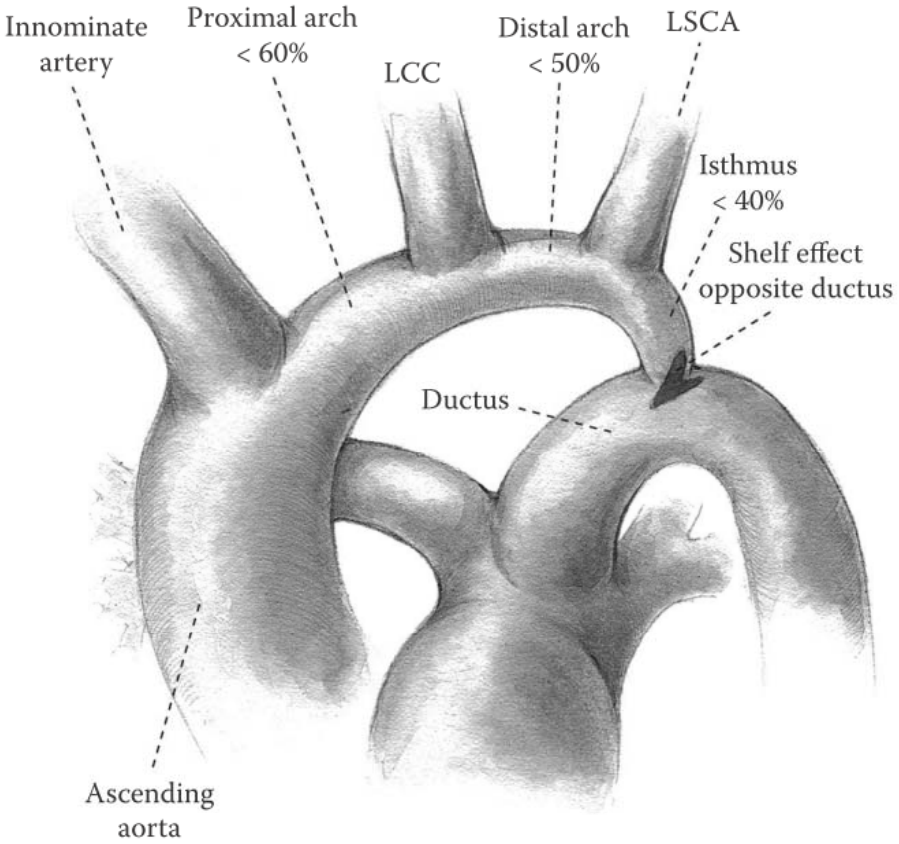
- Critical factors
  - Patent of ductus arteriosus
  - Size of transverse arch
  - Degree of narrowing at the coarctation site

- Three group of classification system
  - I : patients with isolated coarctation
  - II : patients with coarctation and ventricular septal defect
  - III : patients with coarctation and complex intracardiac anomaly

3 components of aortic arch



Aortic arch hypoplasia



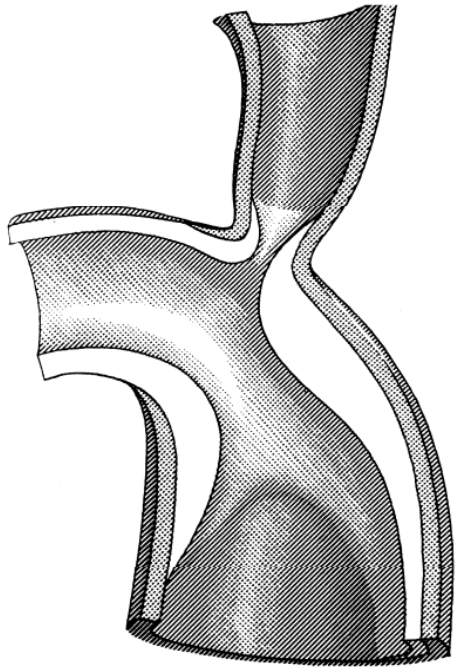
- Arch / Aao diameter < 50%
- Arch < Bwt. (Kg) + 1
- Z-score < -2

Two theories of CoA (I)

- Flow theory
  - Hypothesis that blood flow through the chambers and great arteries determines the size
  - If L-R shunt in fetus, left heart blood flow decreased -> aortic isthmus decreased
  - Lack of fetal blood flow across the aortic isthmus
  - VSD, Bicuspid aortic valve, congenital aortic stenosis, congenital mitral stenosis
  - Corroborating evidence : reduced right heart output (TOF, PS, TA..) lesions are almost never associated CoA

  - no obvious intracardiac defects ??

Two theories of CoA (II)



- Ductal sling theory
  - Skoda (1855) : abnormal extension of contractile ductal tissue into the aorta
  - Microscopic exam : obstructing shelf was composed of cells similar to that found in ductus arteriosus
  - Contraction and fibrosis of “ ductal sling “ at the time of ductal closure → constriction of the aorta and coarctation

Pathophysiology, Clinical presentation

- Critical Neonatal Coarctation
  - Severe juxtaductal constriction of aorta → profound circulatory collapse in first month of life
  - Closure of ductus → restrict blood flow to lower body → absence of femoral pulses
  - Pale, poorly perfused, tachypnea, pulmonary edema
  - Metabolic acidosis → renal failure, hepatic failure, NEC, seizure, death

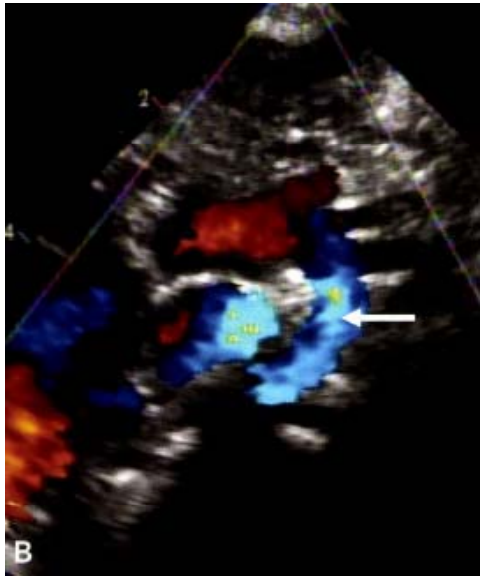
Pathophysiology, Clinical presentation

- Infant Coarctation
  - Slowly closure of ductus → collateral vessel develop
  - Tachypnea, failure to thrive
  - Irritability, sweating, difficulty with feeding

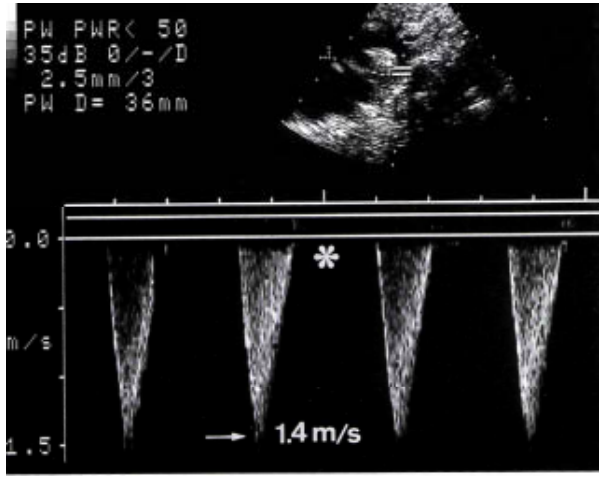
Pathophysiology, Clinical presentation

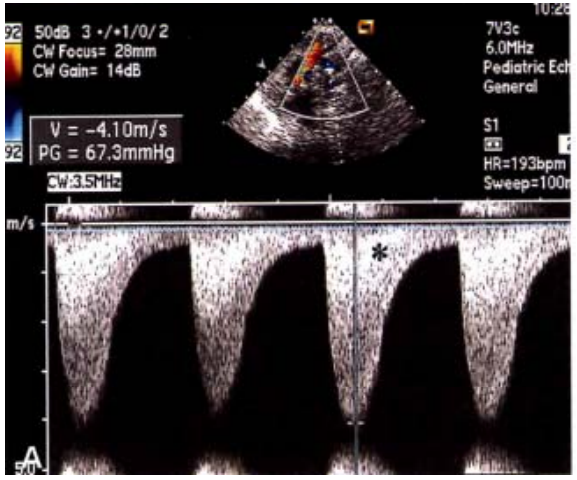
- Older Child and Adult
  - Uncommon for mild or moderate coarctation with undiagnosed
  - History of exercise intolerance, fatigue of lower extremities
  - Excellent collateral vessel development → could be asymptomatic
  - High blood pressure in upper extremities
  - Diminished pulse pressure of femoral pulse
  - Rib notching on chest x-ray

Diagnostic studies



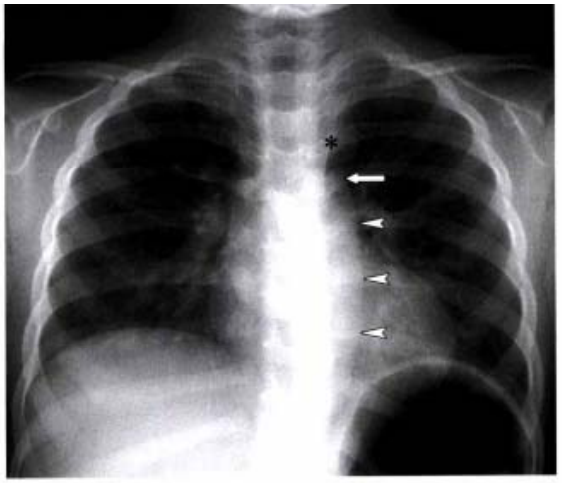
- Echocardiography
  - Two dimensional echocardiography with Doppler
  - Lack of pulsatile flow in the descending aorta
  - Anatomic coarctation site
  - Size of transverse arch
  - Other intracardiac anomaly

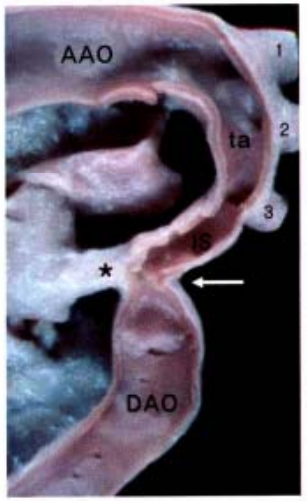




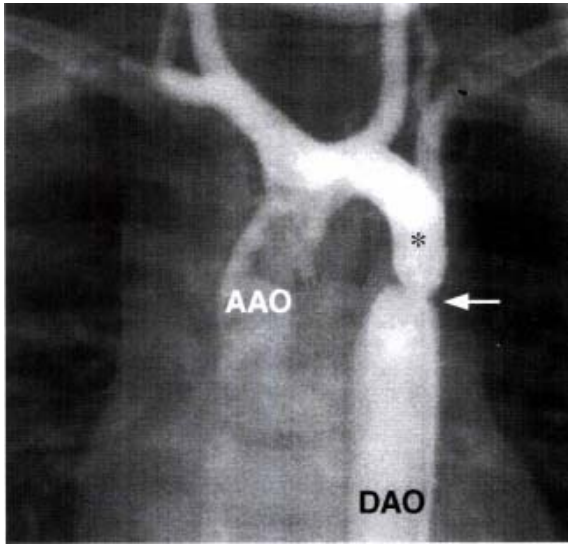
- Chest X-ray
  - Rib notching
  - Classic 3 sign : dilatation of LSCA, narrowing

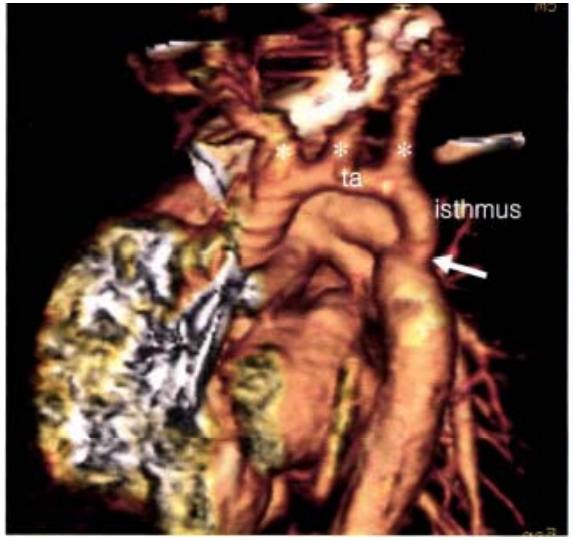
    CoA, post dilatation of descending aorta





- Angiogarphy





- CT scan
- MRI

Medical management

- Critical neonatal coarctation
  - PGE1 infusion
  - Optimize the ratio of pulmonary to systemic resistance
    - Reducing FiO_2 to 21%
    - pCO_2 above 45mmHg
  - Optimize cardiac output : dopamine
  - Enteral feeding : careful watch!!

Surgical indication

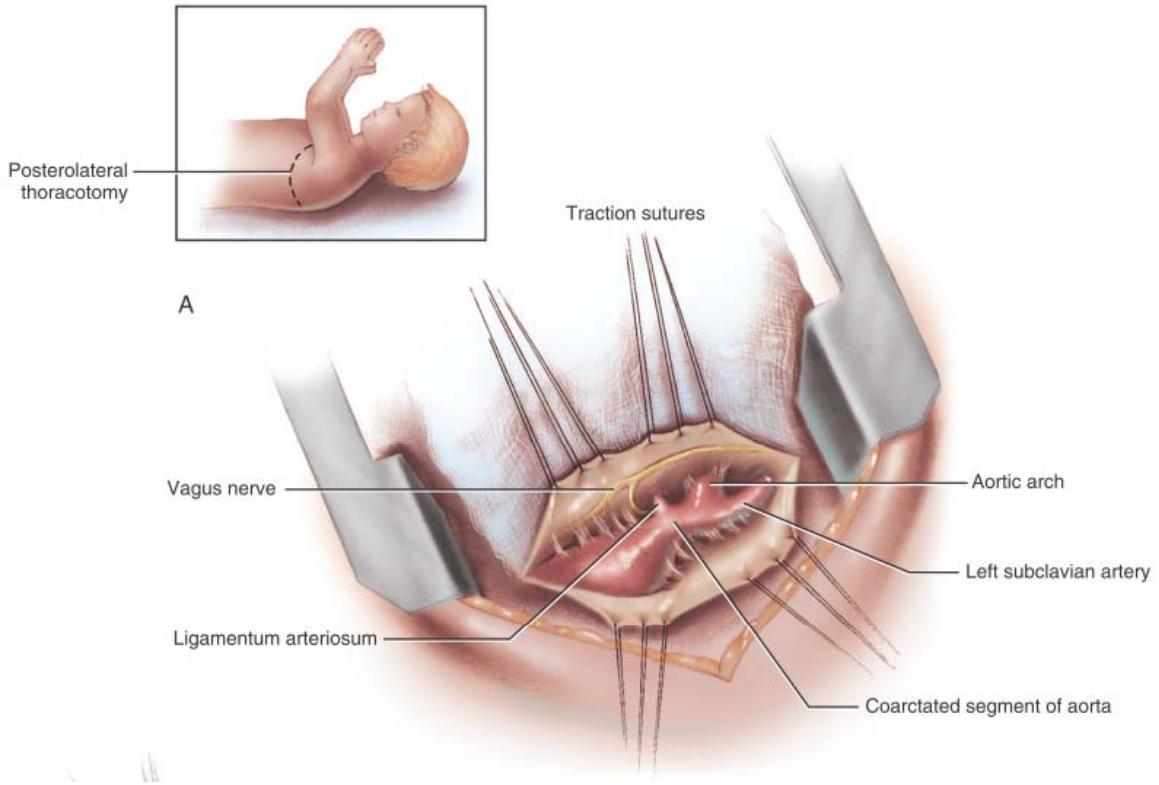
- Unresponsive to medical therapy
- Hypertension > 2SD
- Diameter loss of 50%
- Blood pressure gradient > 20 ~ 30mmHg
- CoA with CHF at any age

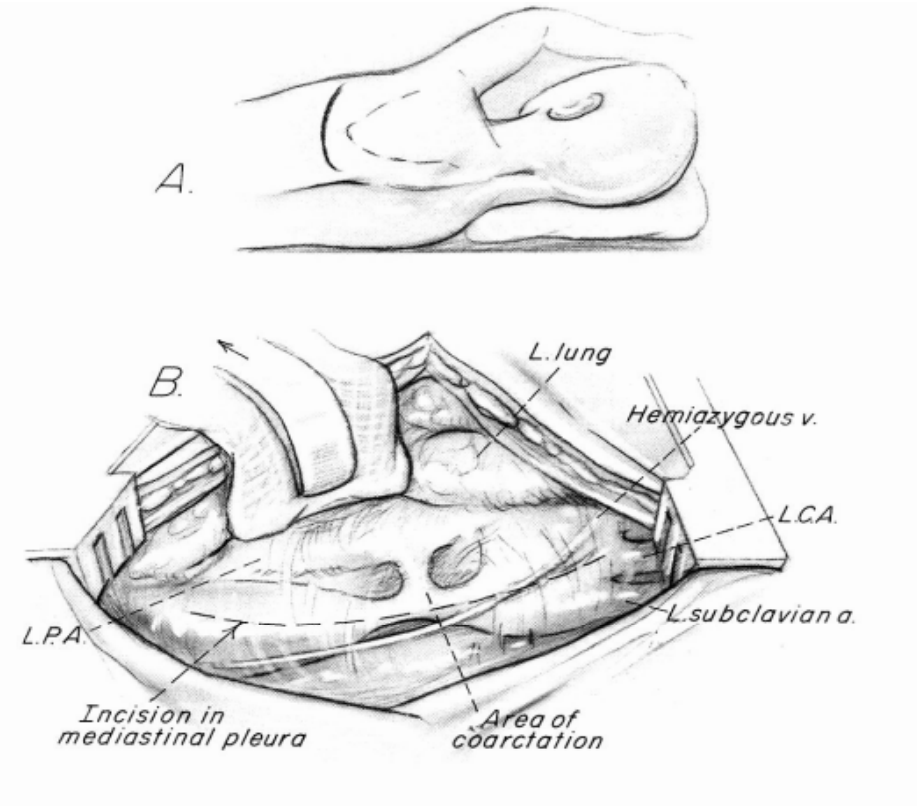
General consideration for Surgical Tx.

- Thoracotomy vs. Sternotomy
  - Arch hypoplasia
  - Associated cardiac anomaly
  - One stage vs. staged

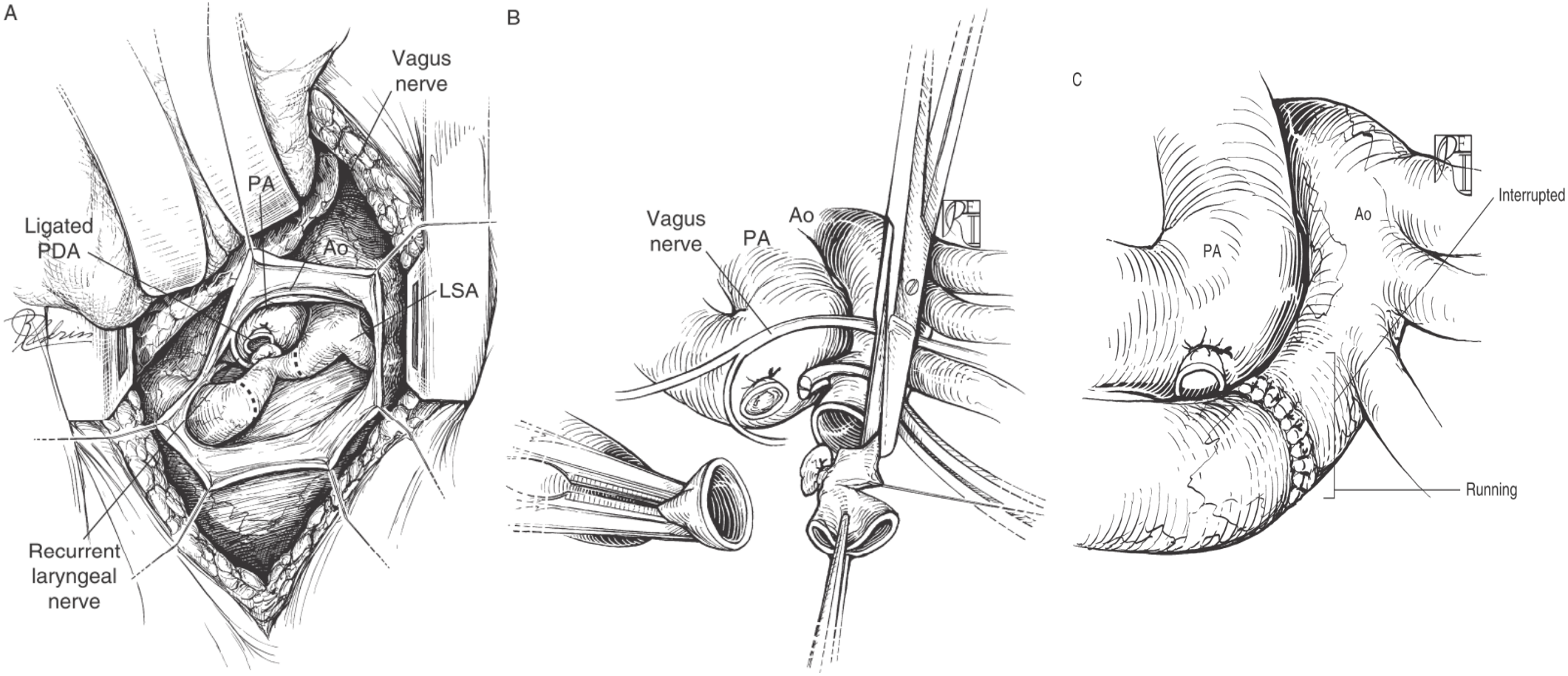
General consideration for Surgical Tx.

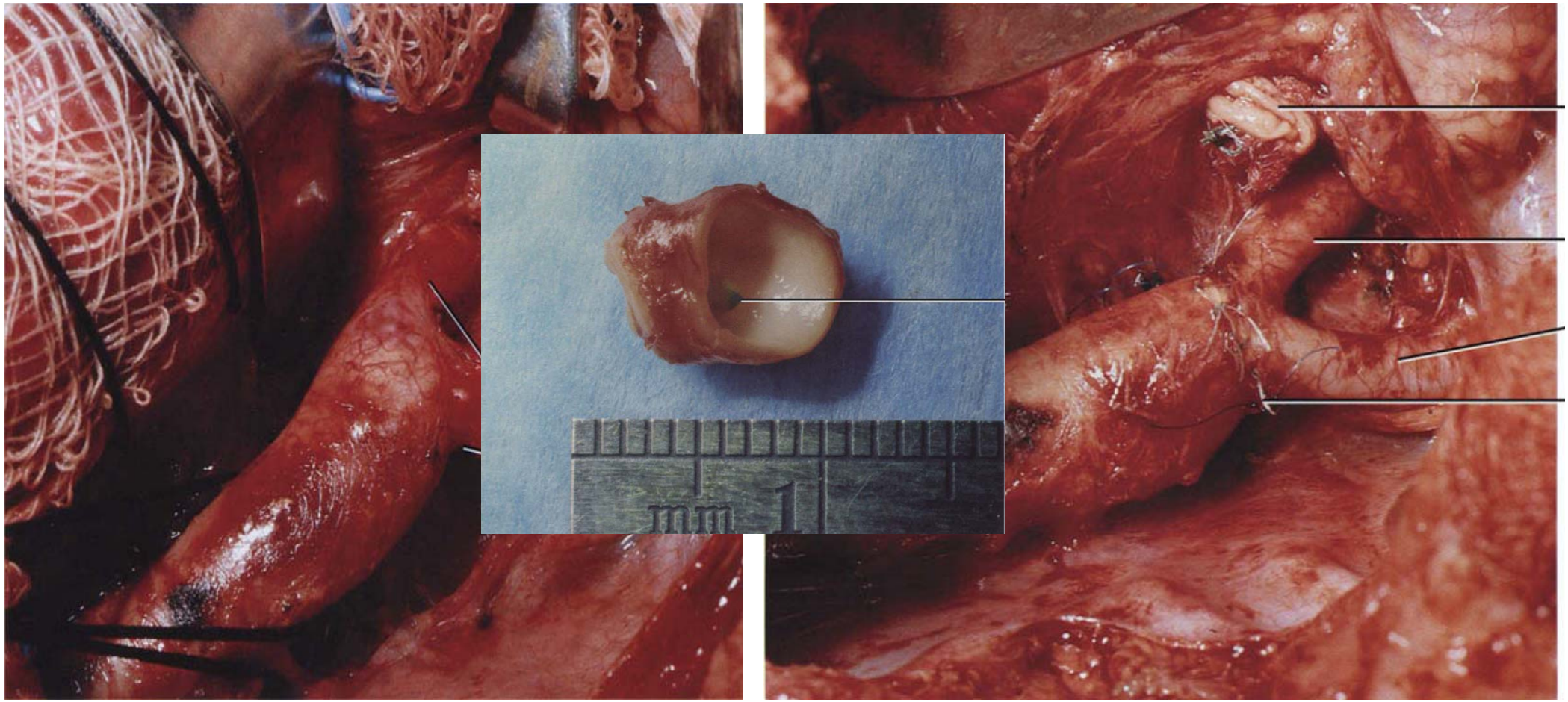
- Left posterolateral thoracotomy incision via 3rd or 4th ICS
- Median sternotomy for patients with intracardiac anomaly
- Arterial pressure monitoring : right radial artery
- NIRS
- Lymphatics channels : preserved or ligated and divided
- Vagus nerve and recurrent laryngeal nerve preservation
- Large collateral intercostal vessels : mobilized and encircled with vessel loops or ligated and divided for fully mobilize


Posterolateral thoracotomy
A
Traction sutures
Vagus nerve
Aortic arch
Left subclavian artery
Ligamentum arteriosum
Coarctated segment of aorta


A.
B.
L. lung
Hemiazygous v.
L.C.A.
L.subclavian a.
L.P.A.
Incision in mediastinal pleura
Area of coarctation

End-to-End anastomosis

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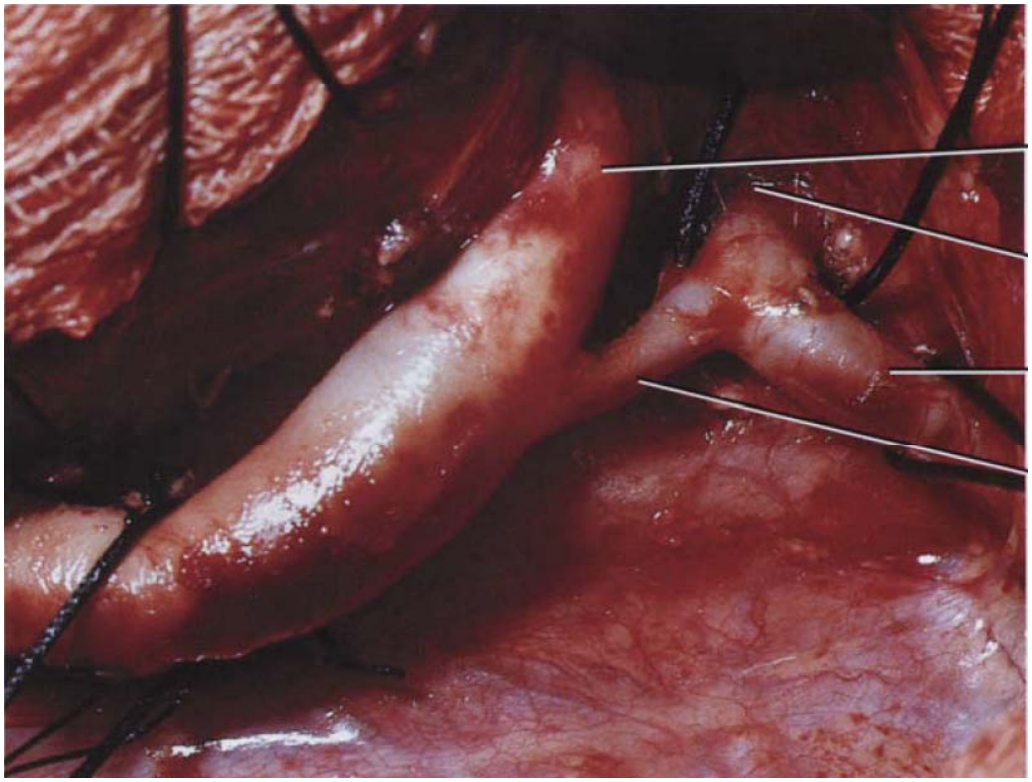
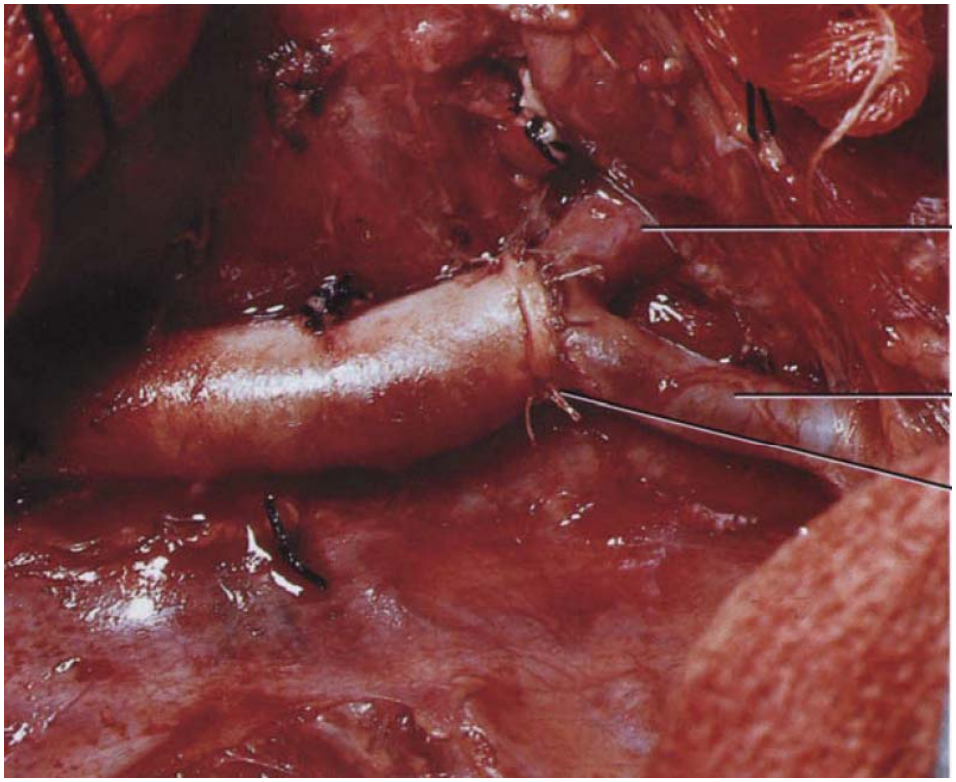
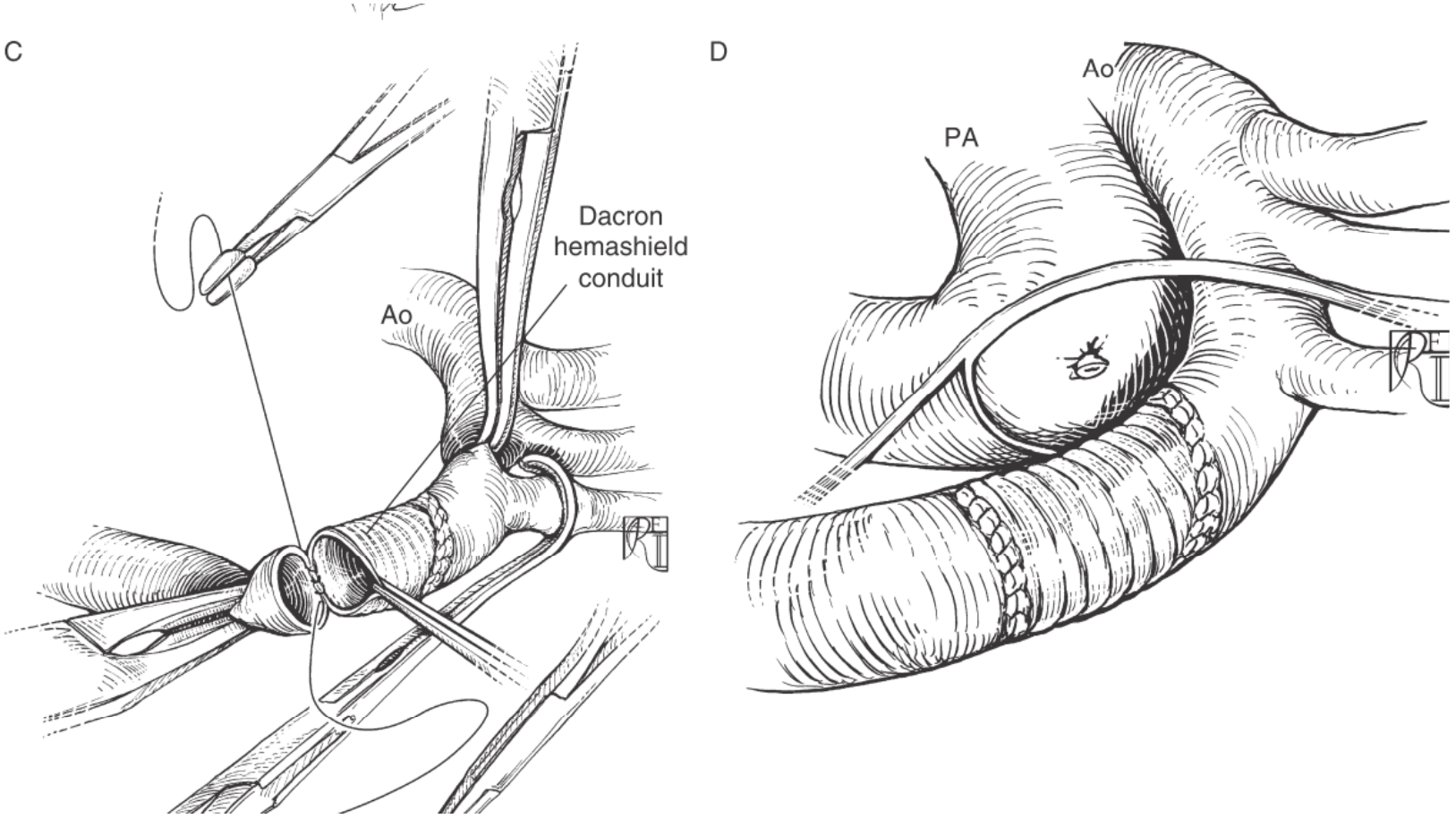
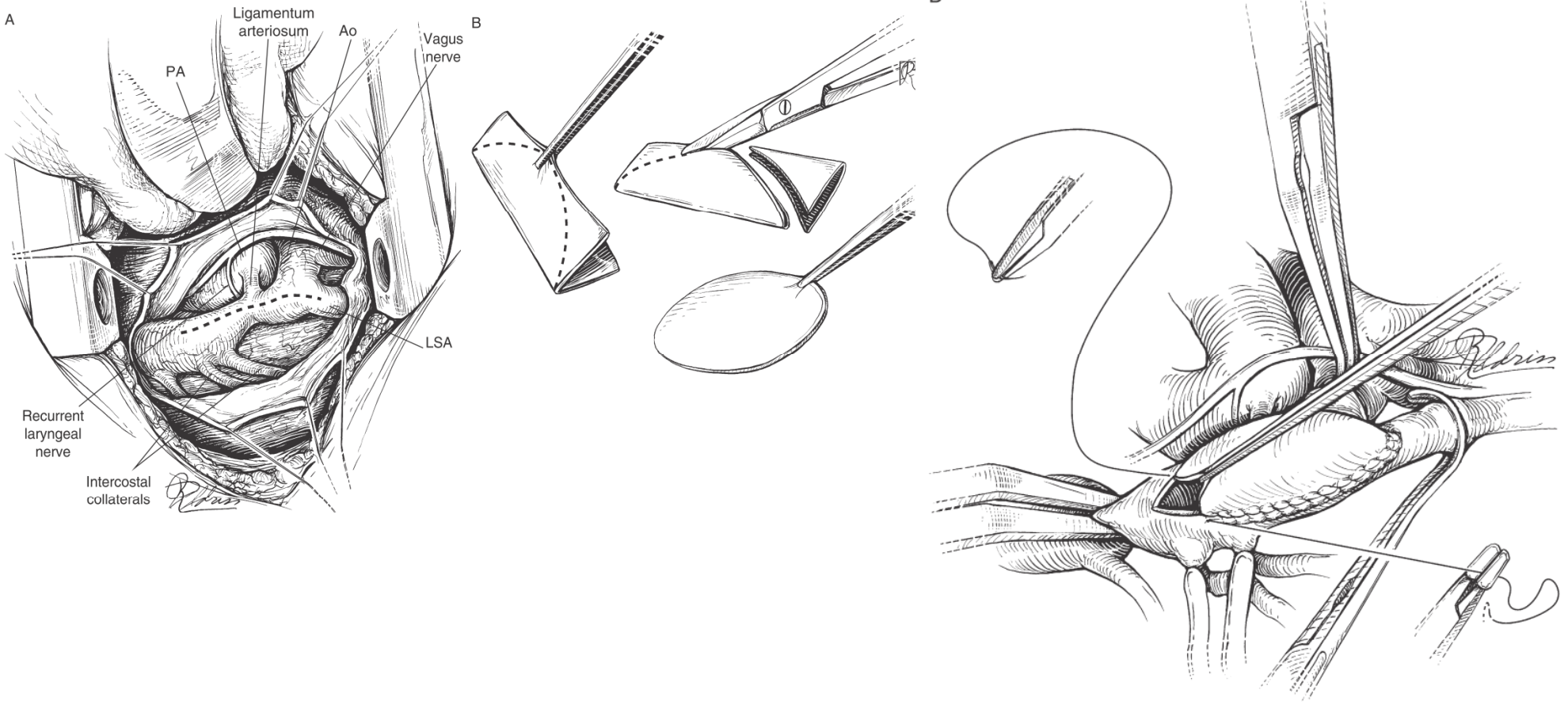


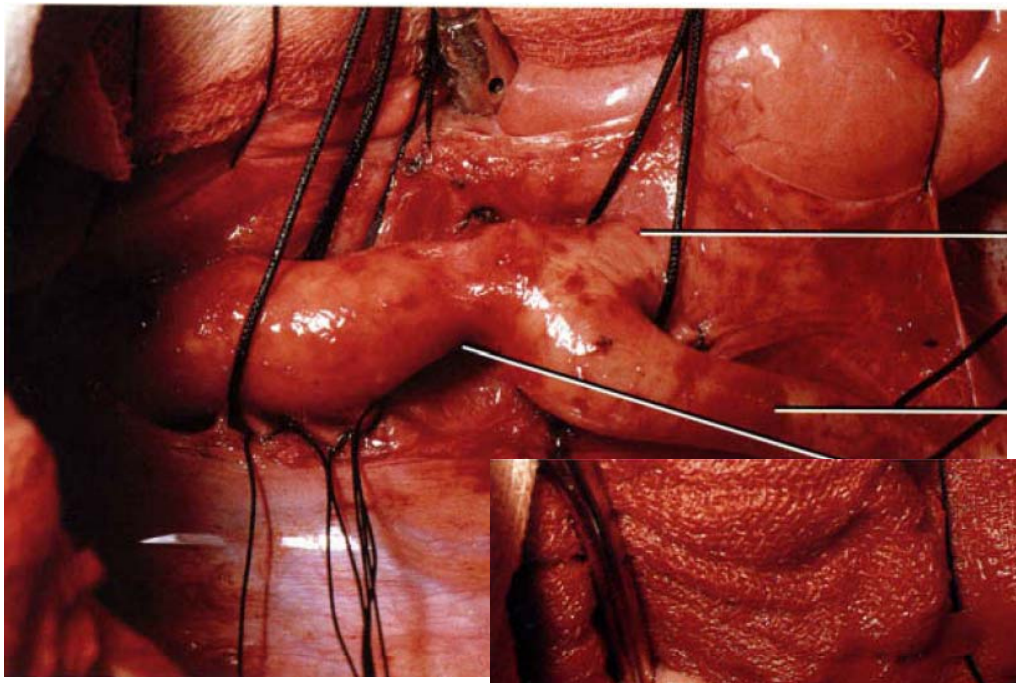


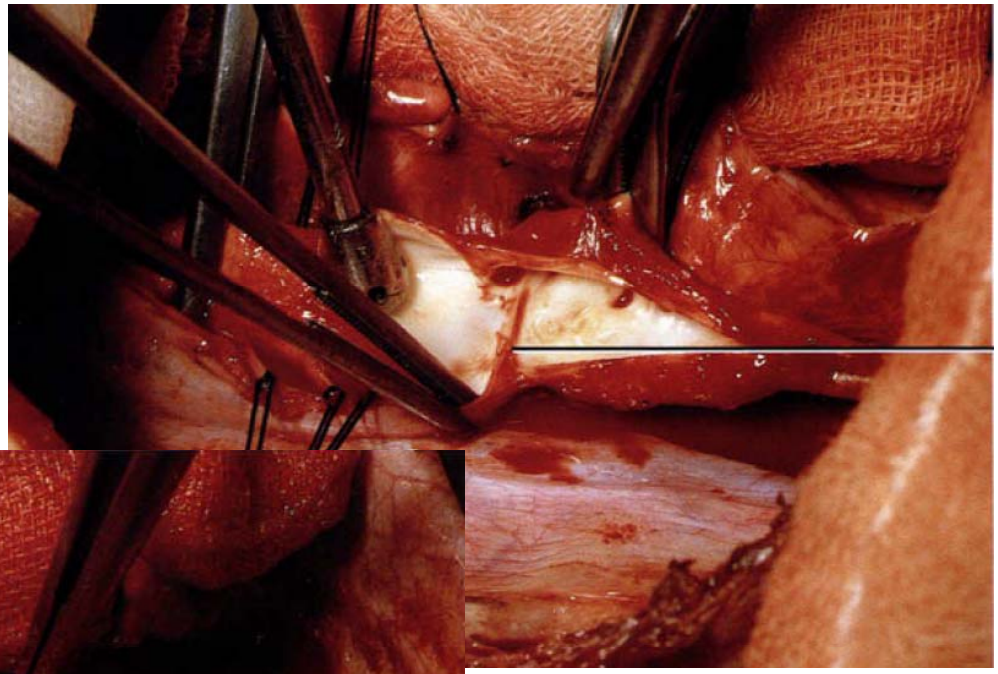
Prosthetic interposition graft

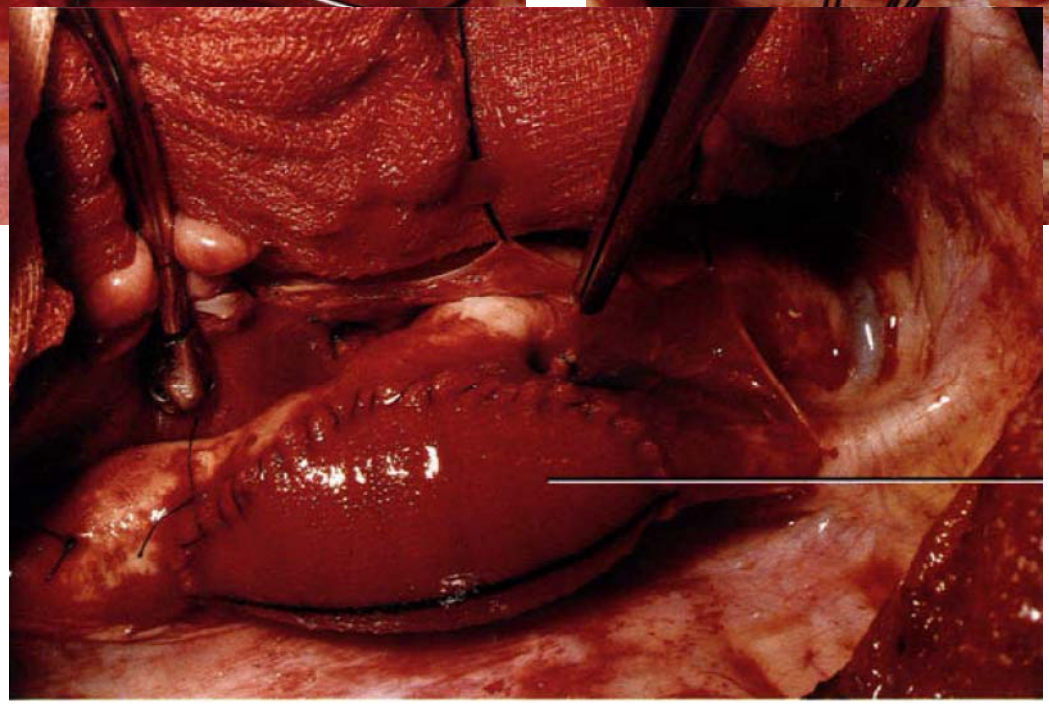


Prosthetic patch aortoplasty

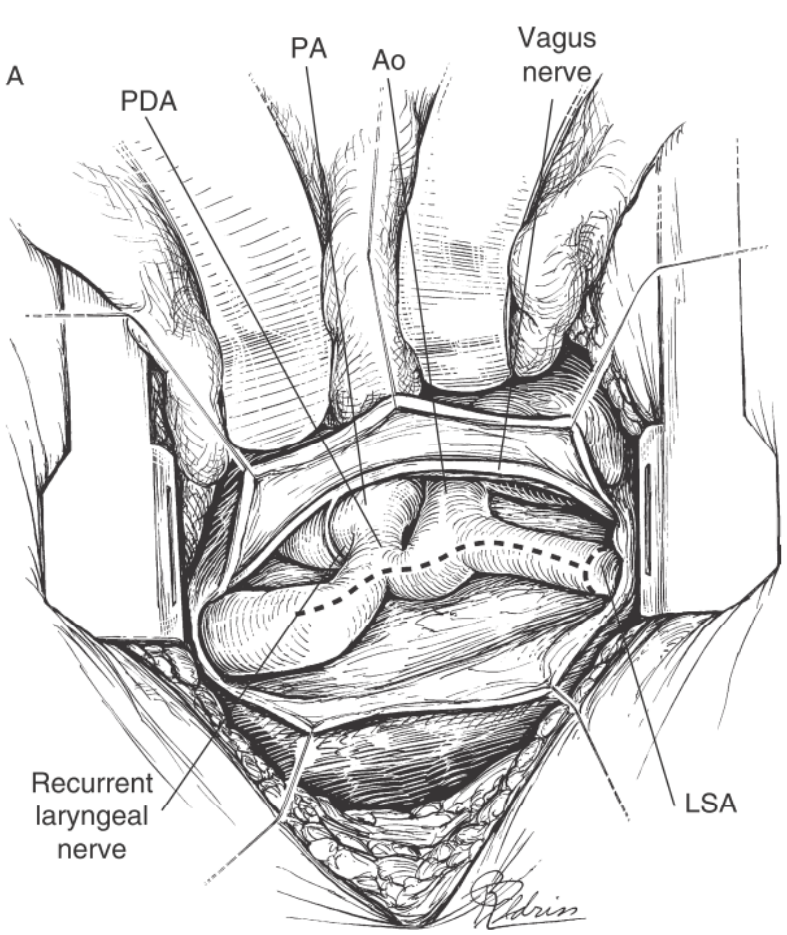


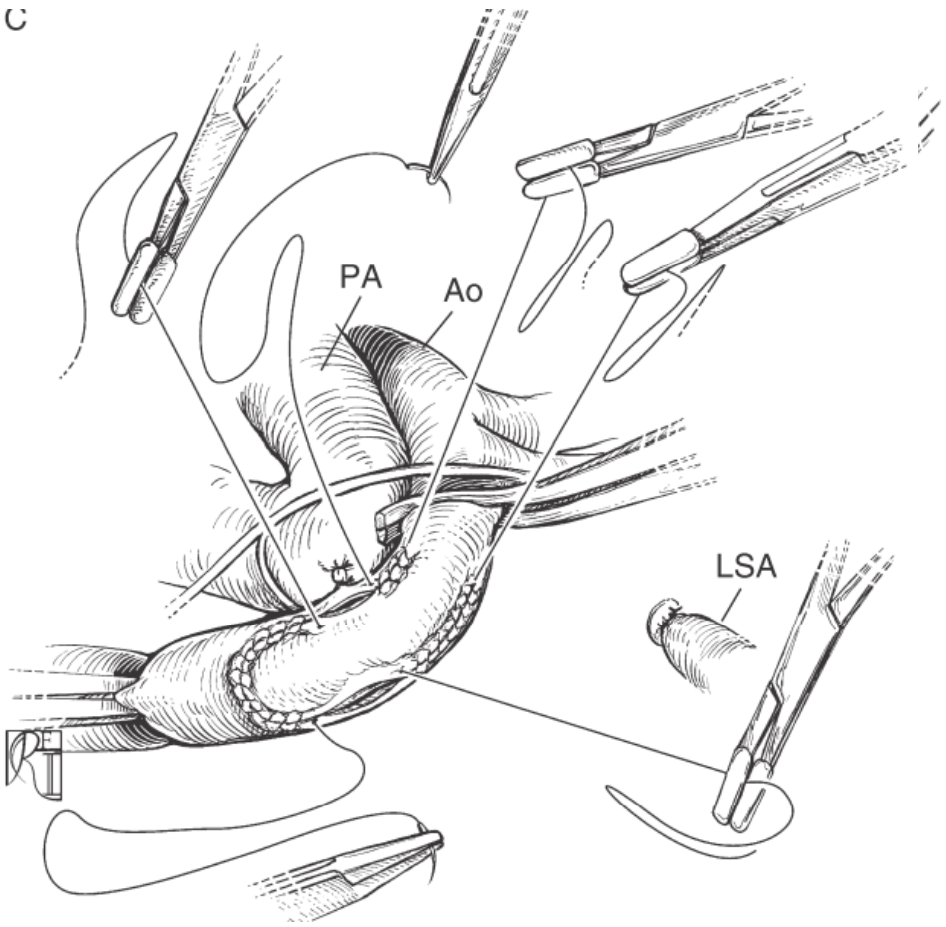


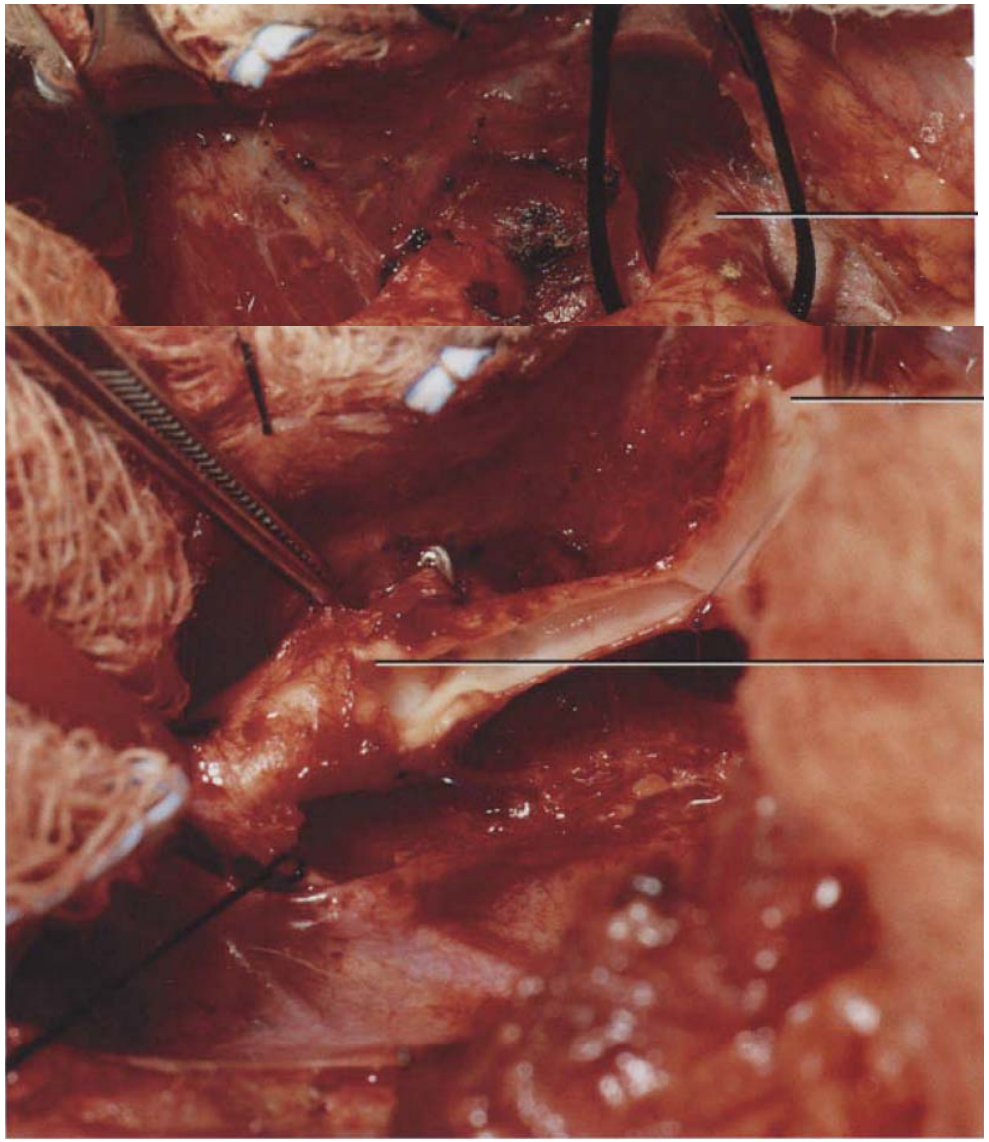


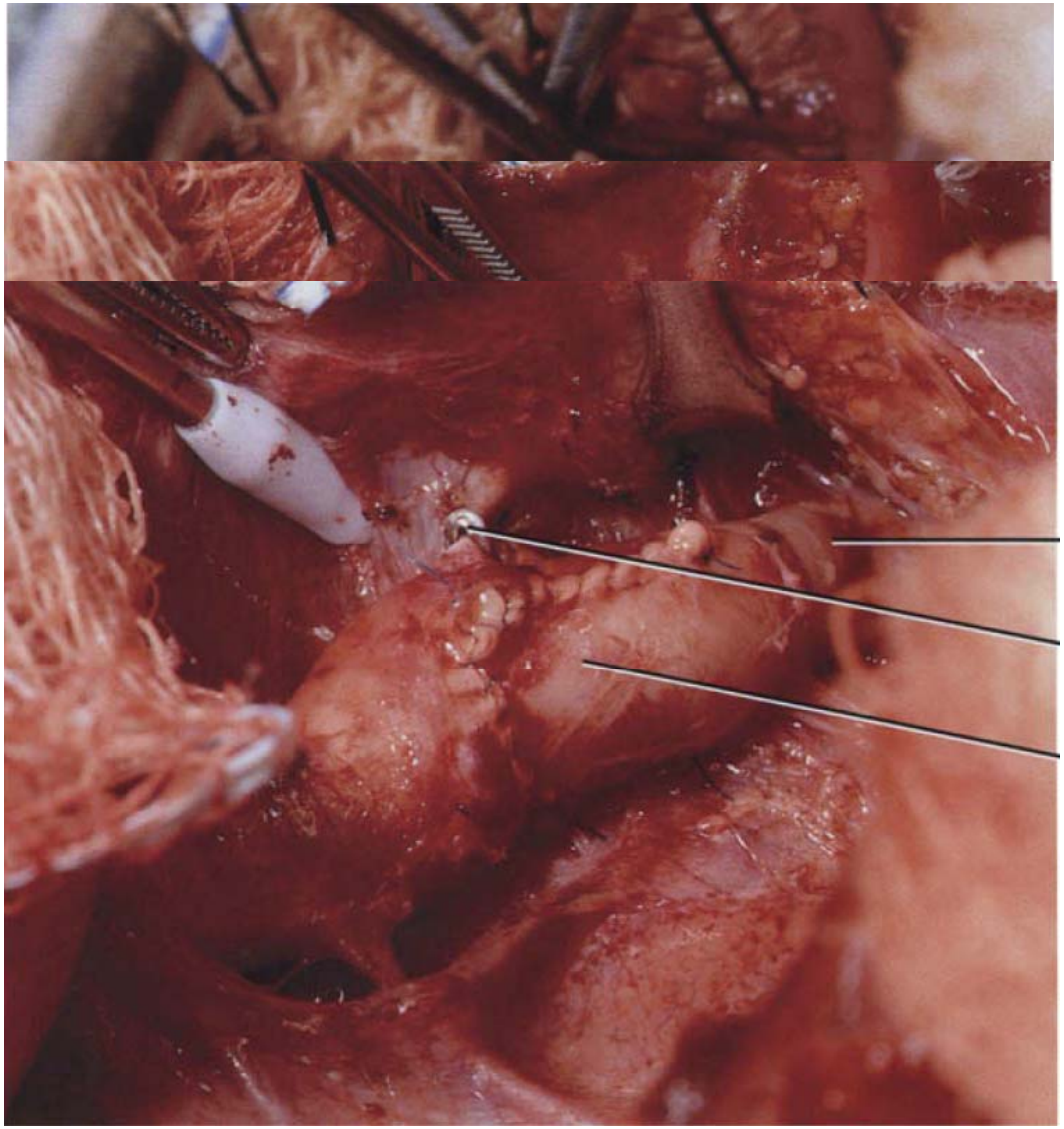


Subclavian flap aortoplasty

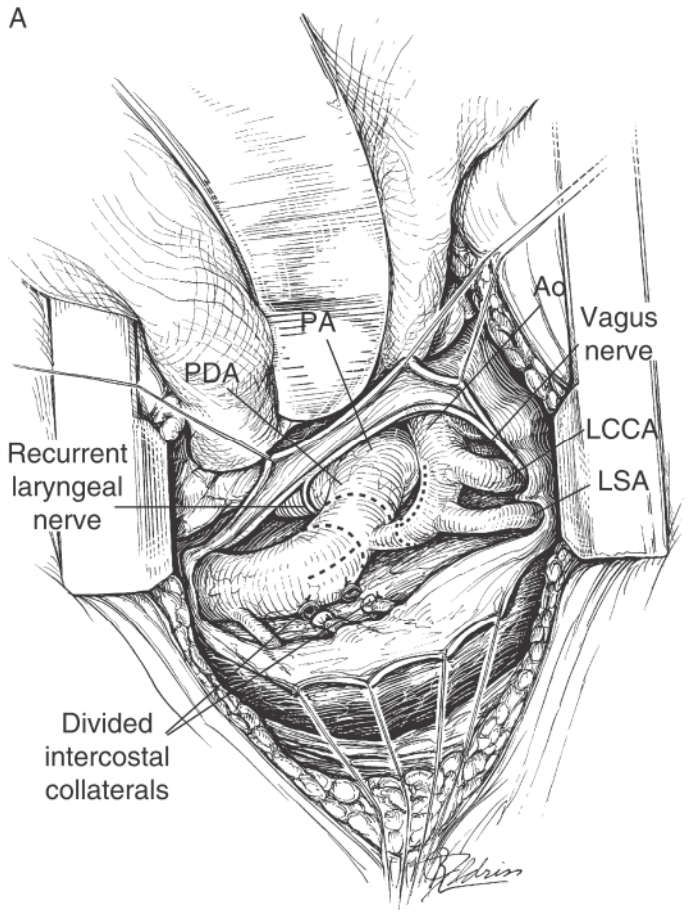


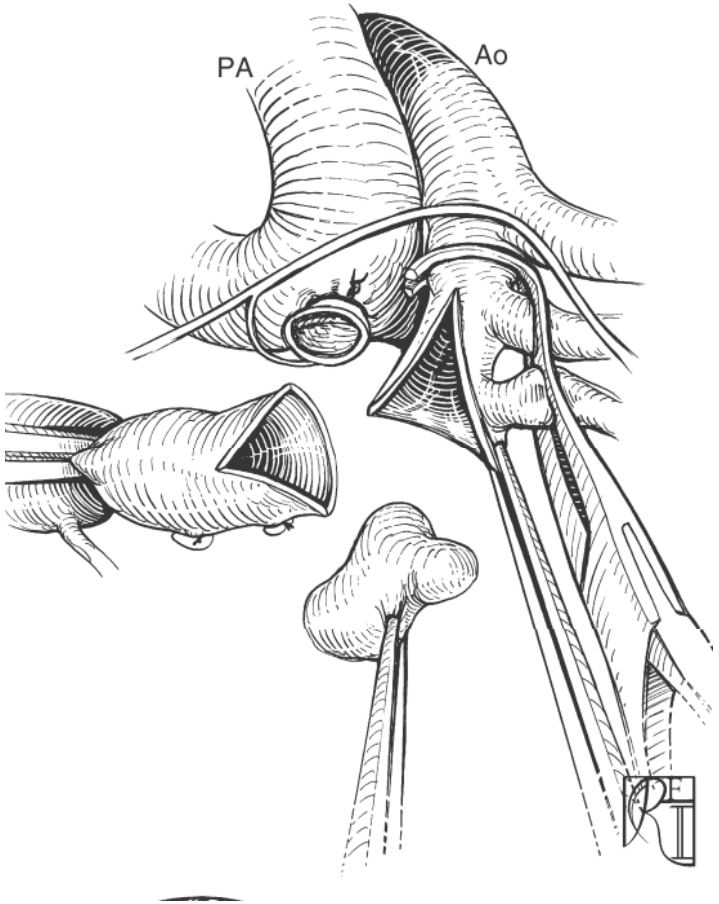


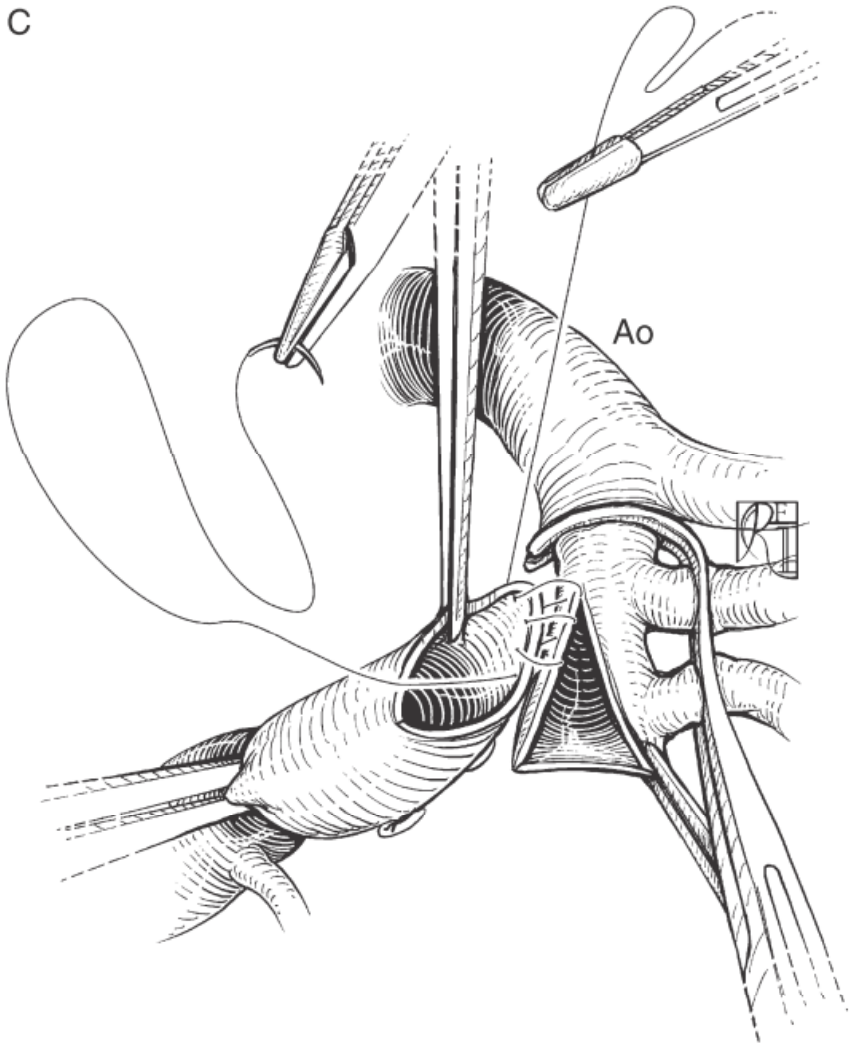


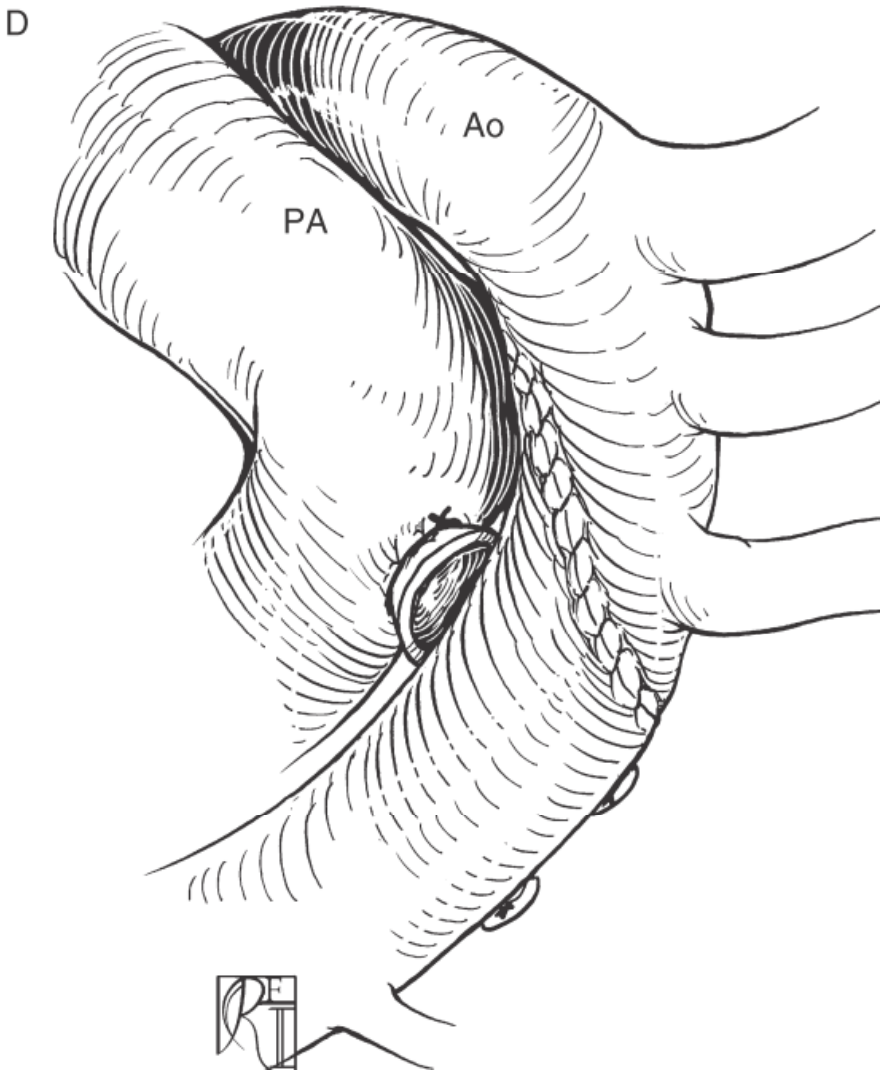


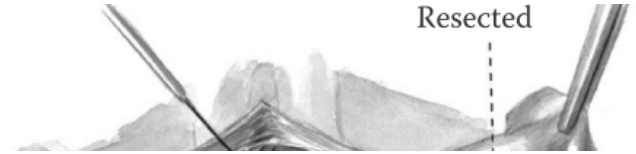
Extended End-to-End anastomosis

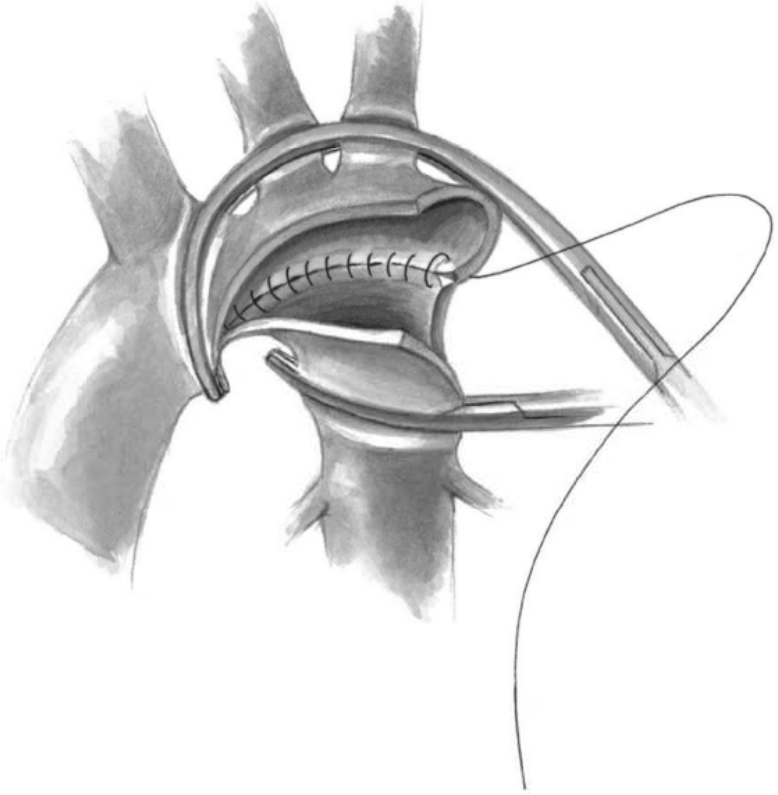




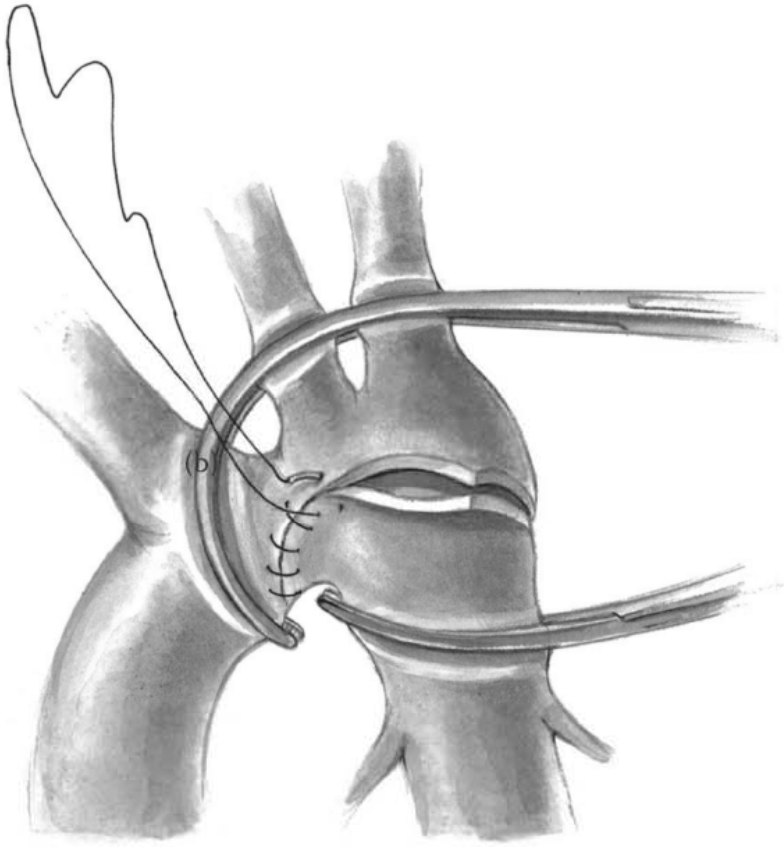

C
Ao


D
Ao
PA



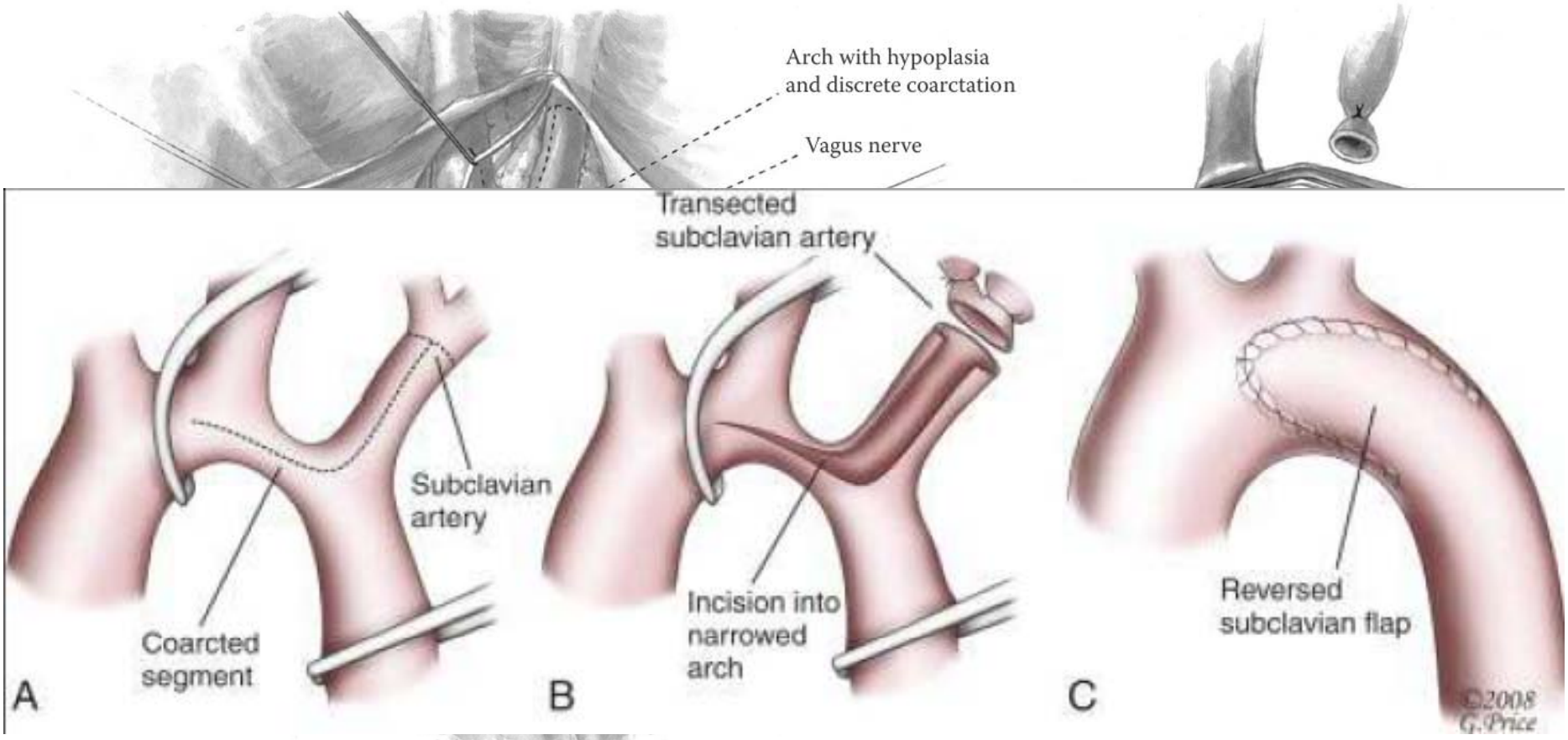


(c)

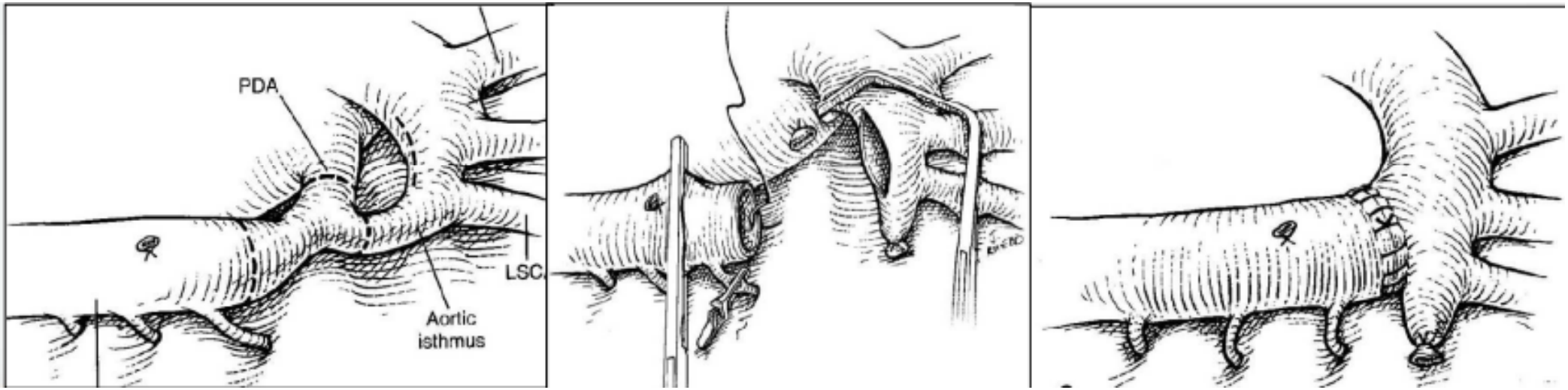


(d)

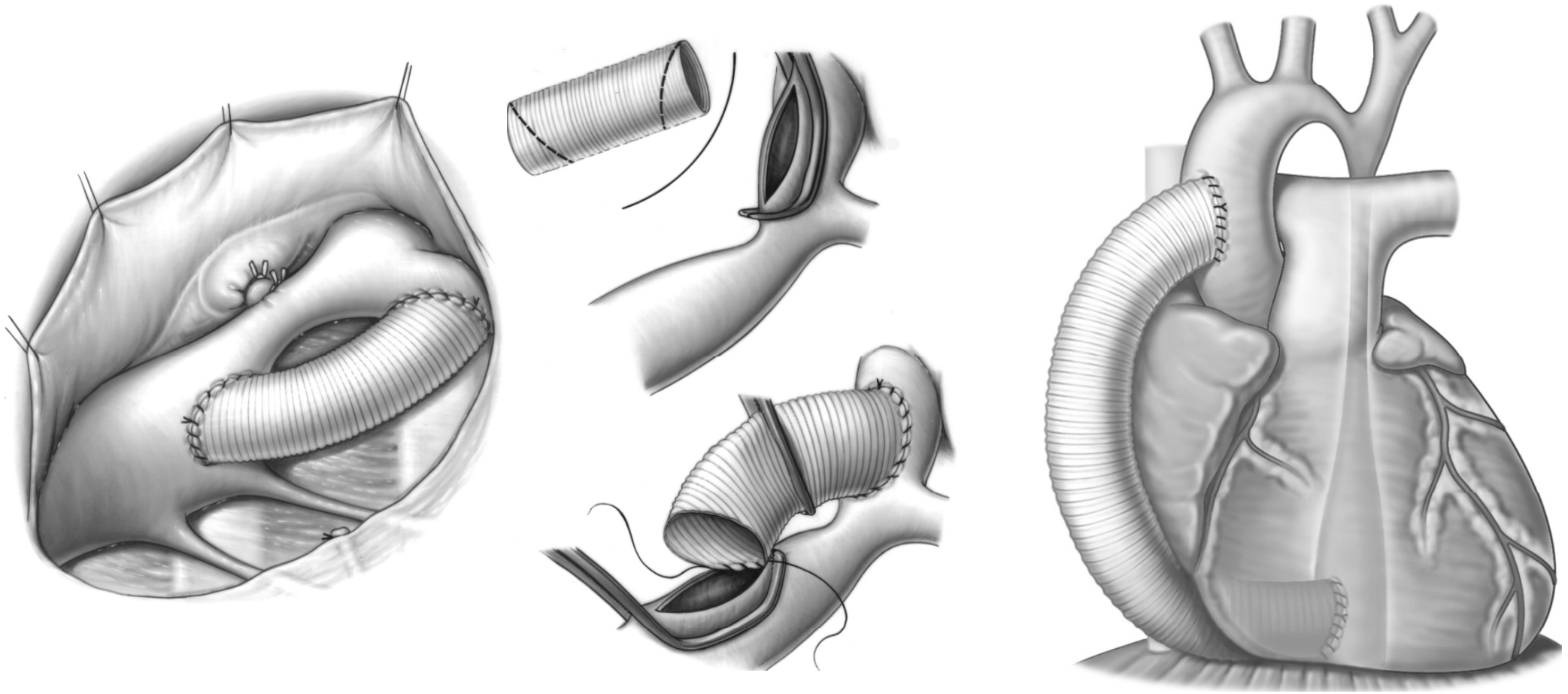
Reverse subclavian flap aortoplasty



End-to-side anastomosis



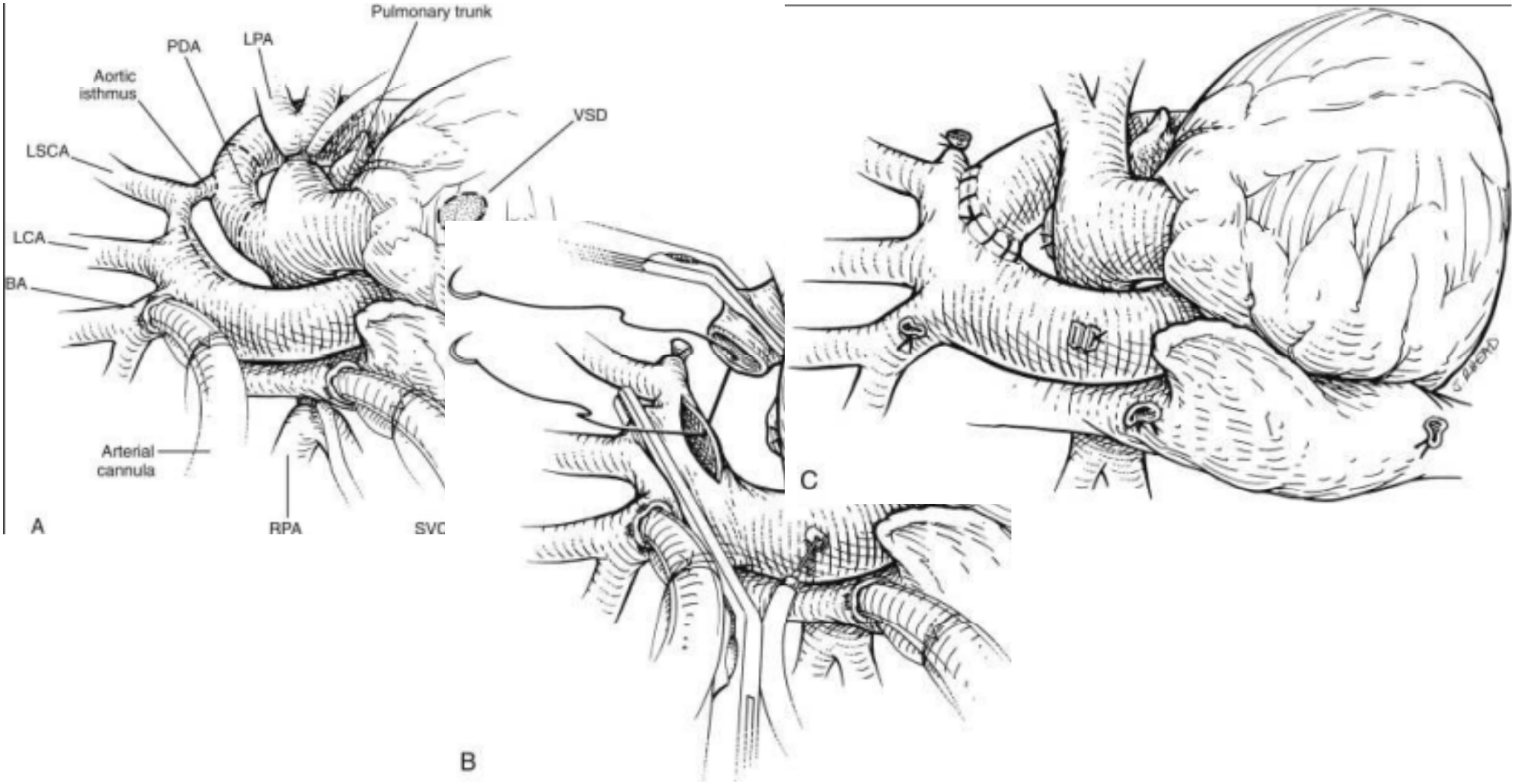
Extra-anatomic bypass graft



Median sternotomy

- CoA with intracardiac defects
  - Staged : CoA repair with PA banding first

    Repair of intra-cardiac defect second
  - Simultaneous repair
    - Cardiopulmonary bypass with hypothermia
    - Total circulatory arrest vs. low-flow perfusion
    - Myocardial perfusion during aortic arch repair


Pulmonary trunk
PDA
LPA
Aortic isthmus
VSD
LSCA
LCA
BA
Arterial cannula
RPA
SVC
A
B
C

DHCA vs. Regional perfusion

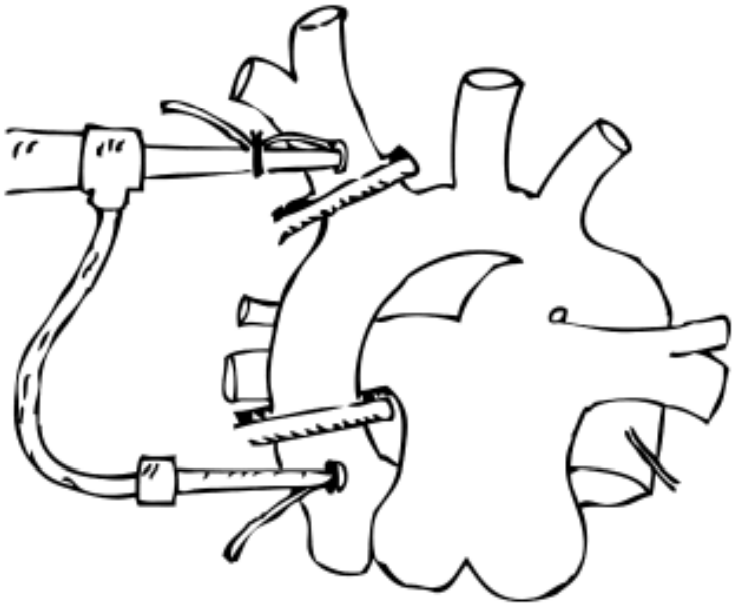
	DHCA	SCP
Advantage	Clear operation field More accurate correction esp. in small babies	Potentially neuroprotective
Disadvantage	Poor neurodevelopmental outcome → Safe duration of circulatory arrest ???	Crowded operative field Technically demanding Lack of randomized trial

Brain protection during arch repair

- Regional cerebral perfusion
  - Via innominate artery → graft, direct
  - Flow rate : 50mL/Kg
  - Right radial artery pressure > 30mmHg
  - Perfusate temperature : 24 ℃ (at rectal temperature 25 ℃)

Table 5. Antegrade Selective Cerebral Perfusion and Selective Cerebral and Myocardial Perfusion: Management Protocols Reported

Author	Year	Pts	Flow[a]	Temp °C	Hema
ASCP					
Asou	1996	14	50	22	
McElhinney	1997	7	30	18	
Pigula	2001	15	20	18	
Tchervenkov	2001	18	18–76	18	
Andropoulos	2003	34	63	17–22	
Hoffman	2004	9	30–70	18–20	25
Dent	2006	22	30	18	28
Amir	2005		20–40	22–25	25
SCMP					
Kostelka	2004	24	30–40	18–28	>
Lim	2002	48	50–100	18	



[a] Flow rates are mL (kg · min).

ASCP = antegrade selective cerebral perfusion; SCMP = selective cerebral and myocardial perfusion.

Complications

Table 14.6 Potential complications of surgery.

Recoarctation
Paradoxical hypertension
Paraplegia
Recurrent laryngeal nerve injury
Left arm ischemia
Hemorrhage
Aneurysm formation
Chylothorax
Horner's syndrome
Phrenic nerve injury
Cerebral-vascular accident

Paradoxical postoperative hypertension

- Two hypertensive responses

  1) Occurs immediately and subsides within 24 hours (systolic)

  - Release of the stretch on the baroreceptors in the carotid artery

  → markedly increased in sympathetic activity

  2) 48~72hr, 1/3 of first phase hypertension (diastolic)

  - Elevated renin and angiotensin
  - Beta blocker and Nitroglycerine, Nitroprusside

Paraplegia

1) ACC time as short as possible

2) Careful technical anastomosis so reapplication of clamps is not necessary

3) Moderate hypothermia

4) High proximal blood pressure

5) No acidosis

6) Adequate distal mean blood pressure (> 45mmHg)

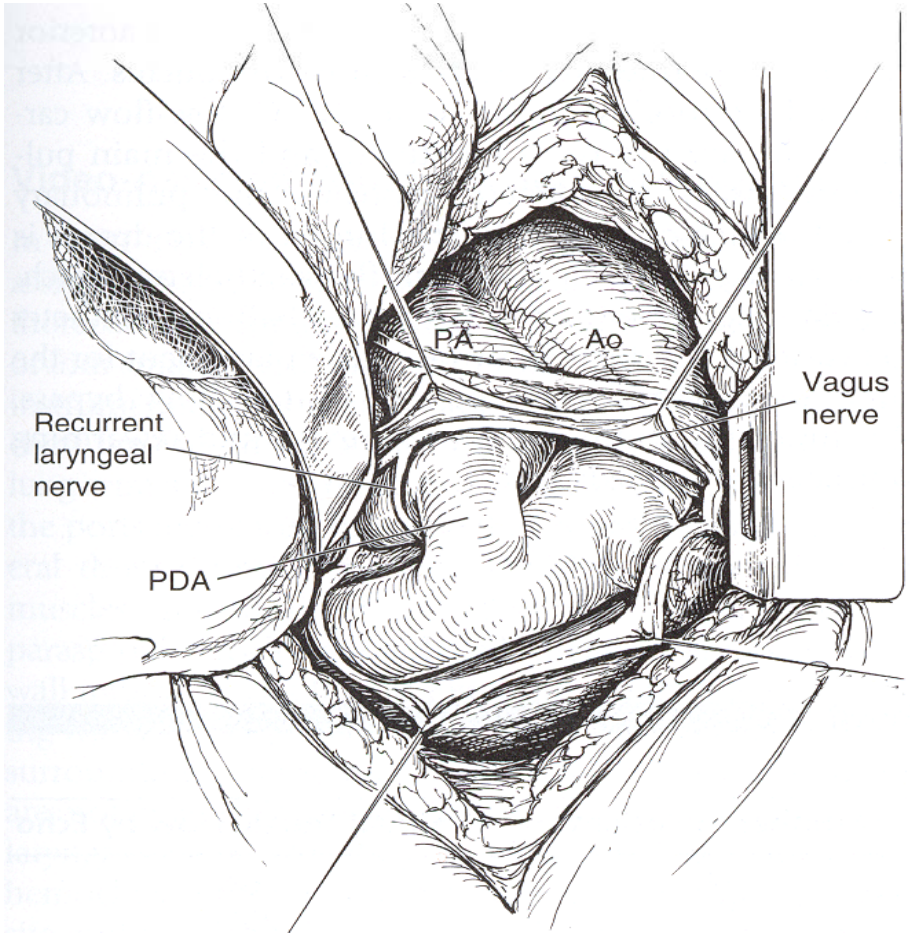
Aneurysm formation

- Prosthetic patch angioplasty

Chylothorax

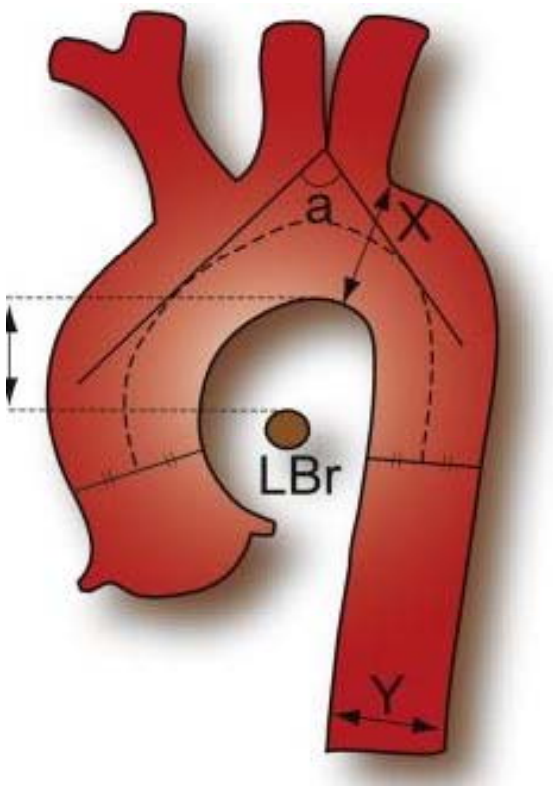
- Near the subclavian artery
- Conservative TX : NPO, Somatostatin, MCT milk

Nerve injury



Left main bronchus compression

- Longer course and smaller caliber compared to right
- Geometric change after coarctoplasty
- Tension on anastomosis site

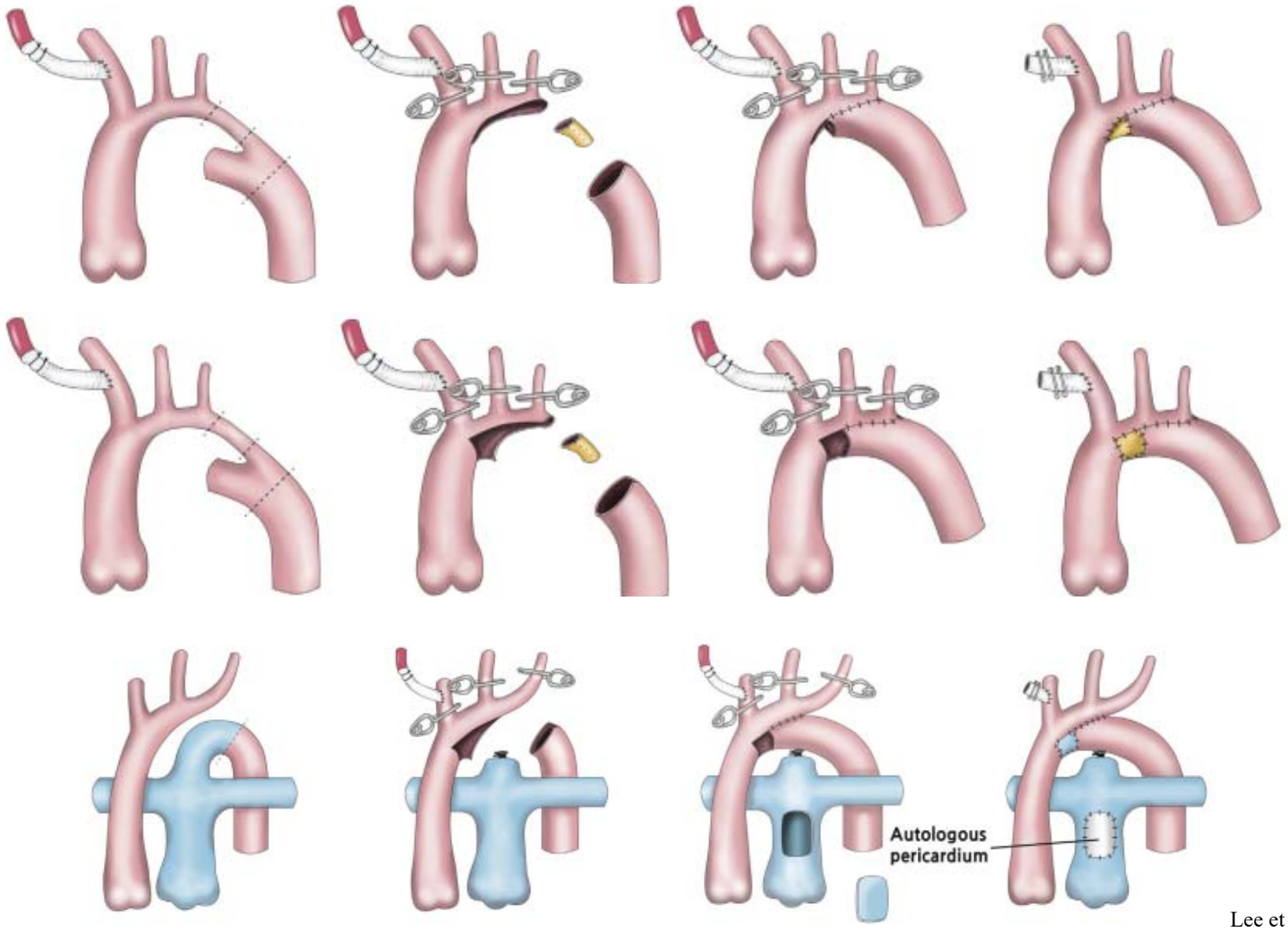


To avoid airway problem

- Extensive dissection arch vessels and descending aorta
- Tension free anastomosis
- Arch repair using autologous MPA patch

Airway problem management

- Aortopexy
- RPA anterior translocation



Lee et al. ATS 2016

Long term follow-up

- Hypertension : age at operation
  - Exclude recurrent coarctation
  - Subclavian flap repair > End-to-end anastomosis (Kenny et al.)
- Aneurysm : synthetic patch aortoplasty
  - Mismatch in compliance between patch and native aortic wall
  - cf.) subclavian flap aortoplasty : remained ductal tissue
  - Balloon aortoplasty

Recoarctation

Table 14.2 Results of resection with end-to-end anastomosis.

Author	Age	Year	Patients	Mortality	Recoarctation
Williams [66]	<1 yr	1980	176	66 (38%)	39 (33%)
Cobanoglu [67]	<3 mo	1985	55	16 (29%)	3 (8%)
Körfer [68]	<4 mo	1985	55	2 (4%)	3 (6%)
Ziemer [69]	<1 mo	1986	24	8 (33%)	4 (25%)
Brouwer [70]	<2 yr	1991	32	2 (6%)	4 (13%)
Kappetein [71]	<3 yr	1994	48	5 (10%)	41 (86%)[a]
Van Heurn [13]	<3 mo	1994	42	5 (10%)	11 (30%)
Quaegebeur [72][b]	<1 mo	1994	139	20 (14%)	6 (4%)
TOTALS			571	124 (21%)	111 (19%)

Table 14.3 Results of patch aortoplasty. (NS, not specified; PTFE, polytetrafluoroethylene,)

Author	Age	Year	Patients	Operative mortality	Recoarctation	Aneurysm	Patch
Yee [84]	<1yr	1984	100	0	10 (12%)	0	PTFE
Clarkson [82]	>15 yr	1985	38	NS	6 (16%)	5 (13%)	Dacron
Hehrlein [80]	2 d–64 yr	1986	317	16 (5%)	4 (1.3%)	18 (6%)	Dacron
Del Nido [85]	3 d–32 yr	1986	63	1(2%)	8 (13%)	3 (5%)	Dacron
Ungerleider [86]	NS	1991	54	0	2 (5%)	0	PTFE
Backer [87]	5.1 yr (mean)	1994	125	4 (3%)	10 (8%)	0	PTFE
Walhout [81]	1.8 yr (mean)	2003	118	4 (3%)	30 (25%)	8 (7%)	PTFE
TOTALS			815	25 (3%)	70 (9%)	34 (4%)	

Table 14.4 Results of subclavian flap aortoplasty. (NS, not specified.)

Author	Age	Year	Patients	Mortality	Recoarctation
Metzdorff [106]	<2 mo	1985	60	11 (18%)	10 (17%)
Ziemer [69]	<1 mo	1986	70	8 (11.4%)	9 (15%)
Ehrhardt [108]	<1 mo	1989	45	14 (31%)	7 (23%)
Milliken [97]	<1 mo	1990	123	11 (9%)	20 (16%)
Van Heurn [13]	<3 mo	1994	15	1 (7%)	6 (42%)
Quaegebeur [72][a]	<1 mo	1994	112	9 (8%)	12 (12%)
Allen [93]	<3 mo	2000	53	0	2 (4%)
Jahangiri [109]	<1 yr	2000	185	6 (3%)	11 (6%)
Pandey [105]	< 4 yr	2006	399	51 (12.8%)	61 (15.3%)
Barreiro [110]	<1 yr	2007	119	5 (4%)	12 (11%)
TOTALS			1181	116 (10%)	150 (13%)

Table 14.5 Results of resection with "extended" end-to-end anastomosis.

Author	Age	Year	Patients	Operative mortality	Recoarctation
Lansman [9]	6 mo	1986	17	1 (6%)	2 (12%)
Vouhé [19]	3 mo	1988	80	21 (26%)	6 (10%)
Zannini [112]	3 mo	1993	21	4 (19%)	4 (23%)
Van Heurn [13]	3 mo	1994	77	5 (6%)	8 (11%)
Kappetein [71]	3 yr	1994	26	4 (15%)	0%
Conte [21]	<1 mo	1995	307	23 (7%)	24 (9%)
Van Son [116]	<1 mo	1997	25	0	1 (4%)
Wood [118]	<1 yr	2004	181	1	4 (2.2%)
Wright [117]	<1 yr	2005	83	2 (2%)	4 (6%)
Thomson [119]	<1 yr	2006	191	9 (5%)	7 (4.2%)
Kaushal [53]	1 yr	2009	201	4 (2%)	8 (4%)
TOTALS			1209	74 (6%)	68 (6%)

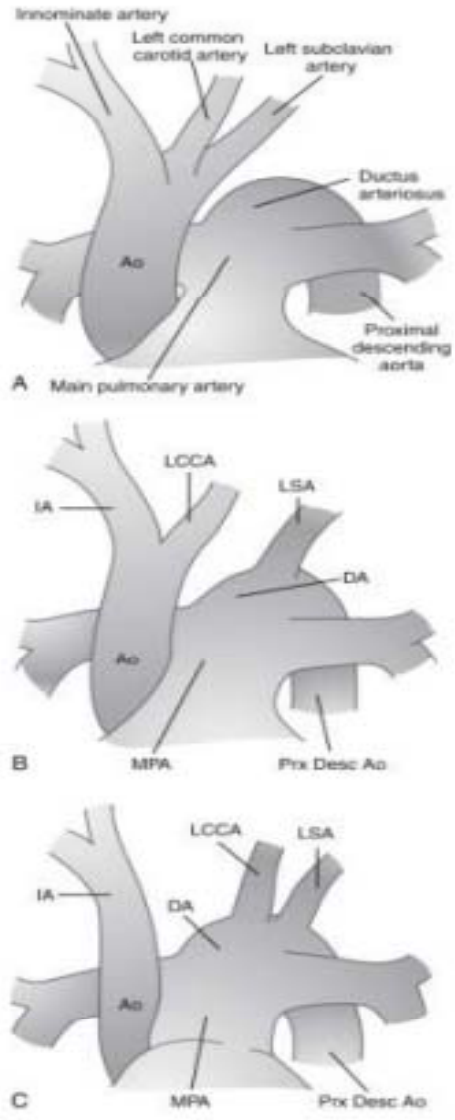
Prevention of recoarctation

- Operative procedure
  - Successfully address transverse arch hypoplasia
  - Resection of all ductal tissue
  - Prevention of residual circumferential scarring at anastomosis site
  - Tension free anastomosis

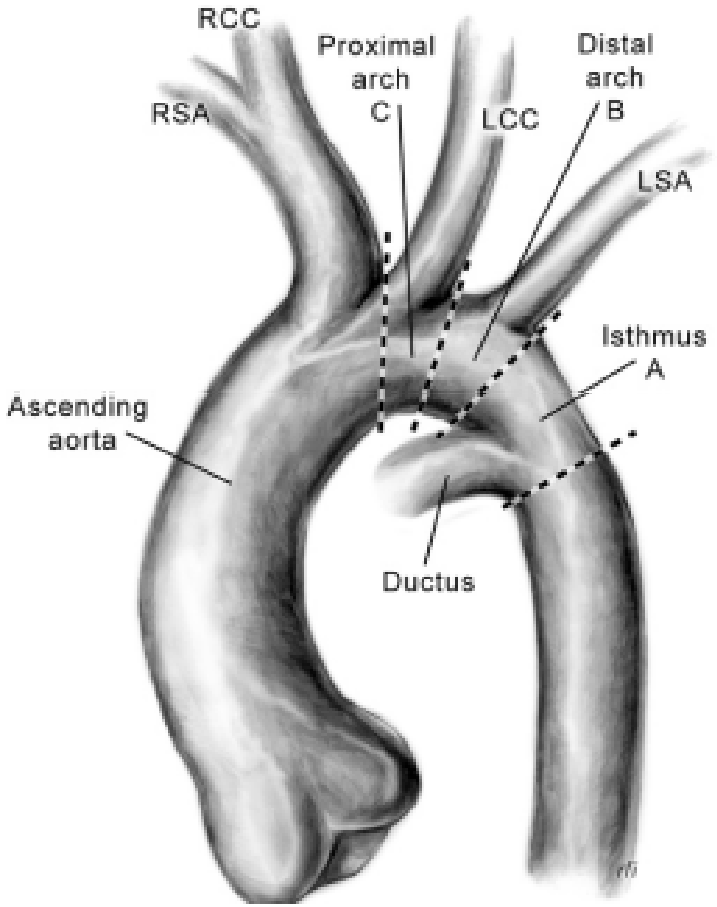
Interrupted Aortic Arch (IAA)

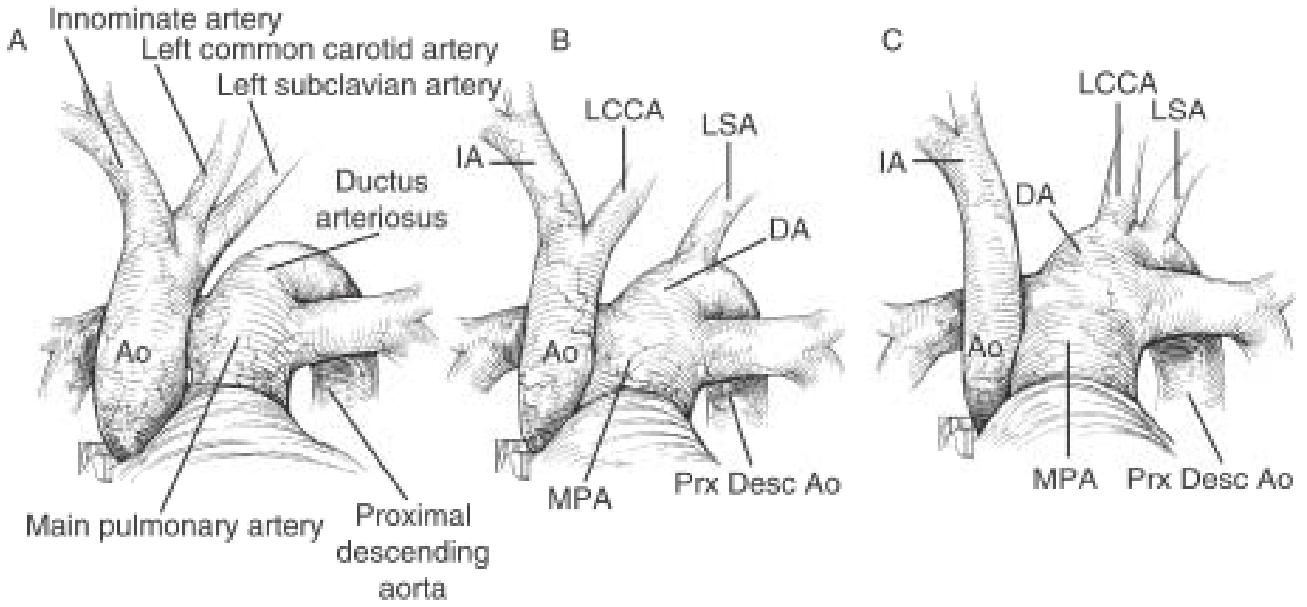
IAA

- Def.
  - Complete luminal and anatomic discontinuity between two segments of the aortic arch
  - Fibrous strand connecting two widely separated ends are also included IAA
- 1% of CHD
- 80% of die within 1st month without Tx.



Anatomy





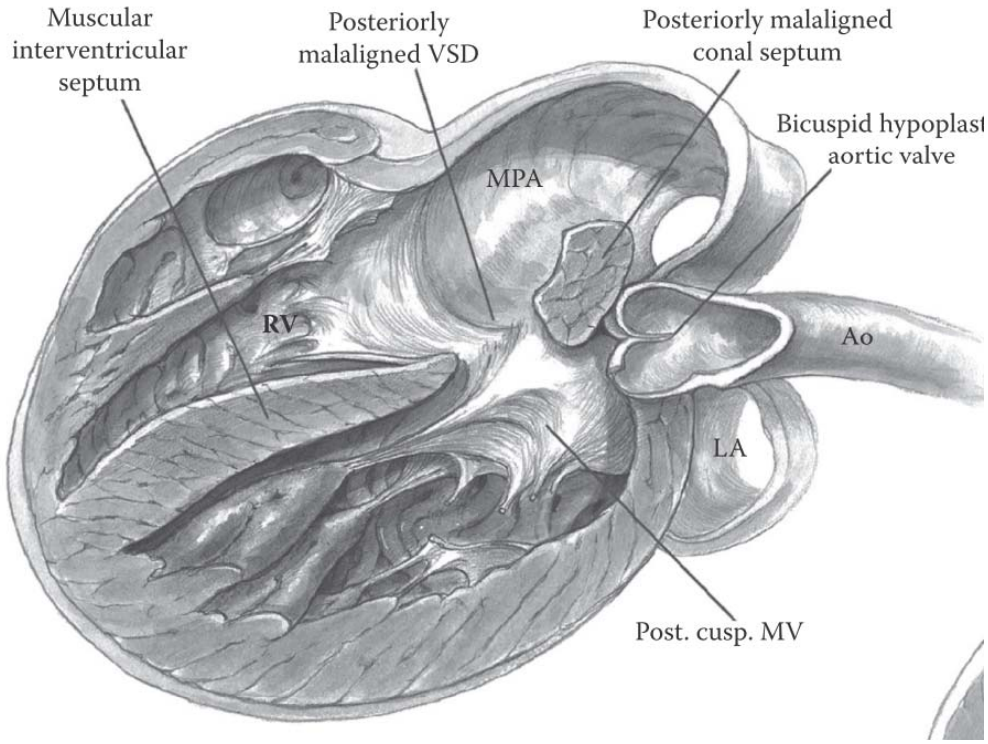
(40%) **(55%)** **(5%)**

Pathophysiology

- Distal blood flow dependent on patent ductus arteriosus
- Spontaneous ductal closure → systemic hypoperfusion, metabolic acidosis, end-organ-failure
- Upper ext. saturation > lower ext. saturation
- Severe pulmonary HTN in patients with VSD, ASD

Associated condition

- Commonly combined with LVOTO, bicuspid AV
- LVOTO
  - Posterior deviation of infundibular septum
  - Small aortic annulus
- VSD : 70%
- Truncus arteriosus : 10%
- TGA, DORV : 4~5%
- AP window : 3%
- DiGeorge syndrome, Berry syndrome

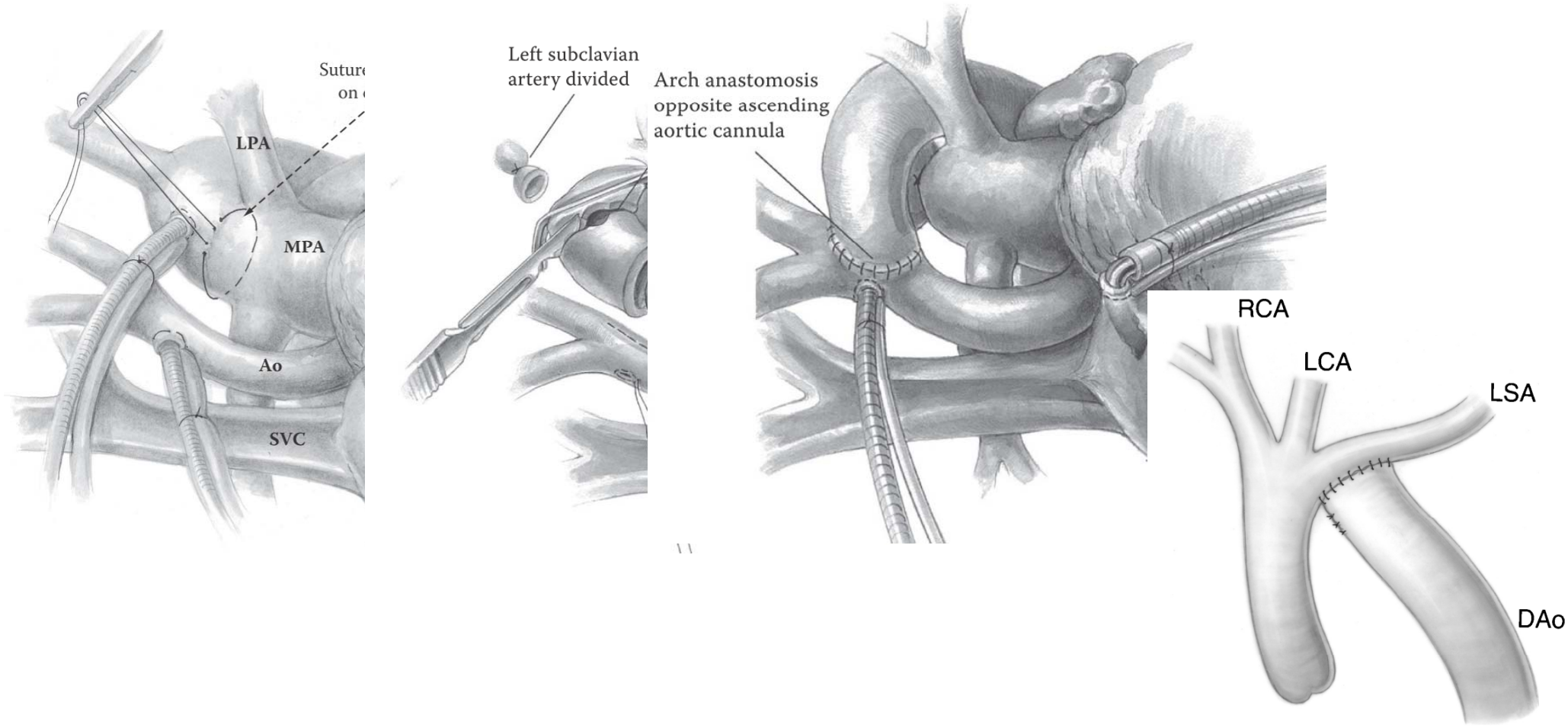


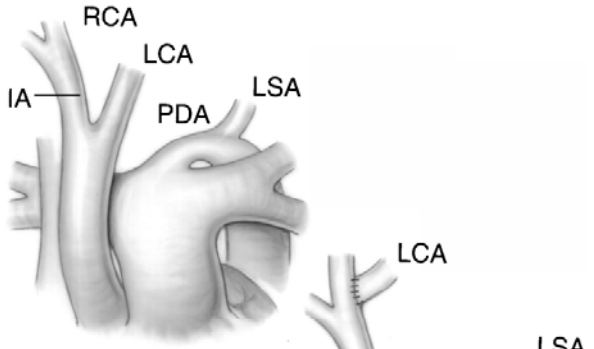
Medical treatment

- PGE1
- CHF management
- Mechanical ventilator if needed
- iCa level check

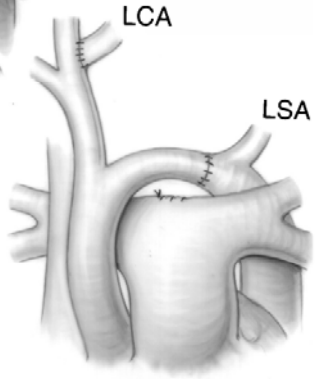
Surgical treatment

- Indication
  - Urgent operation is advisable
- Technique
  - Direct anastomosis
    - Resection and end to end anastomosis
    - Extended resection and end to end anastomosis
  - Interposition of graft
- One stage repair

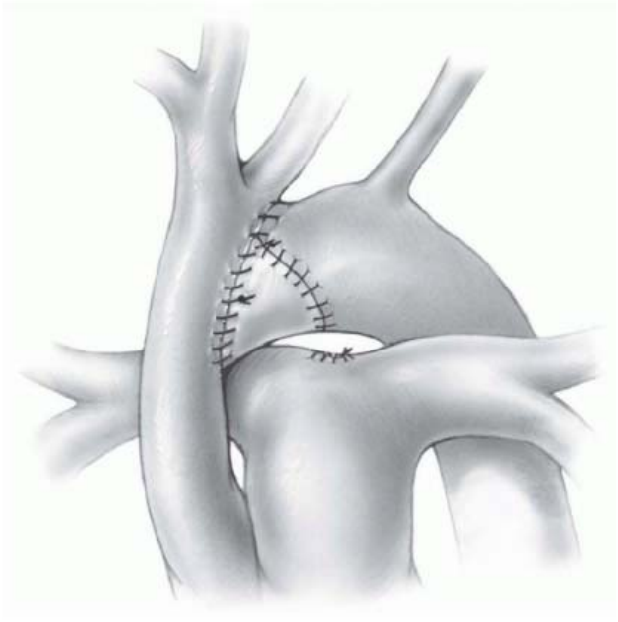

Suture
on
LPA
MPA
Ao
SVC
Left subclavian
artery divided
Arch anastomosis
opposite ascending
aortic cannula
RCA
LCA
LSA
DAo

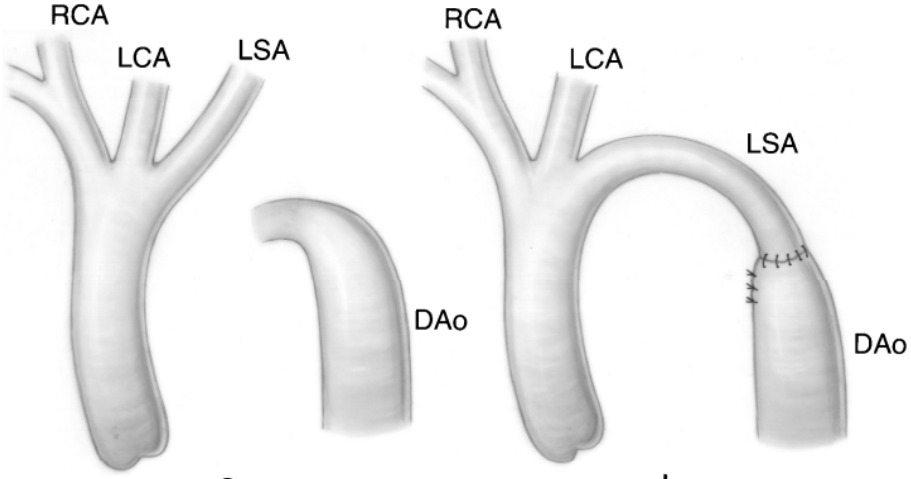

RCA
LCA
IA
PDA
LSA

a

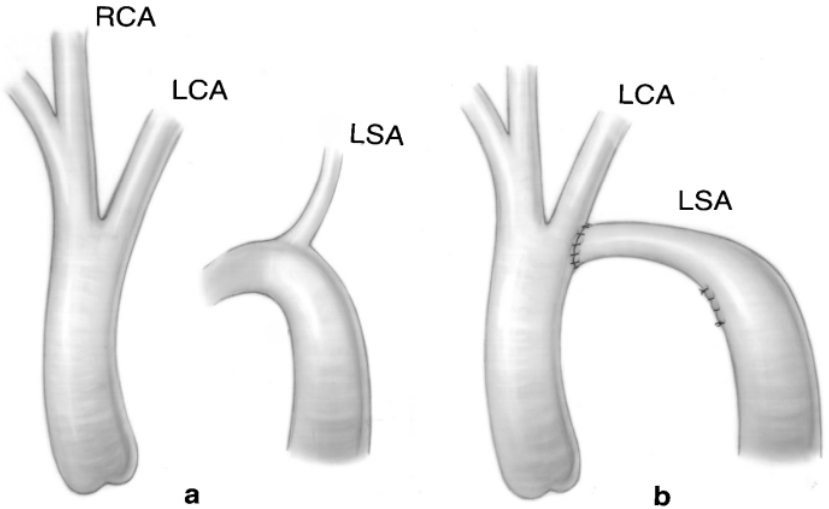

LCA
LSA

b




RCA
LCA
LSA
DAo
RCA
LCA
LSA
DAo

a b


RCA
LCA
LSA
LCA
LSA

a b

Results

- Mortality
- Early death
- Acute or subacute HF with or without multisystem failure
- One stage repair < 10%
- Techniques other than direct anastomosis are associated with higher risk of reintervention

Complication

- Early
  - Bleeding : inadequate mobilization, friability of tissue
  - Restenosis : ductal tissue
  - Left recurrent laryngeal and phrenic nerve injury
  - Paraplegia
  - Compressed left main bronchus
- Late
  - Pressure gradient across arch : balloon dilatation
  - LVOTO