

Congenital Cardiac Surgery

Arch obstruction

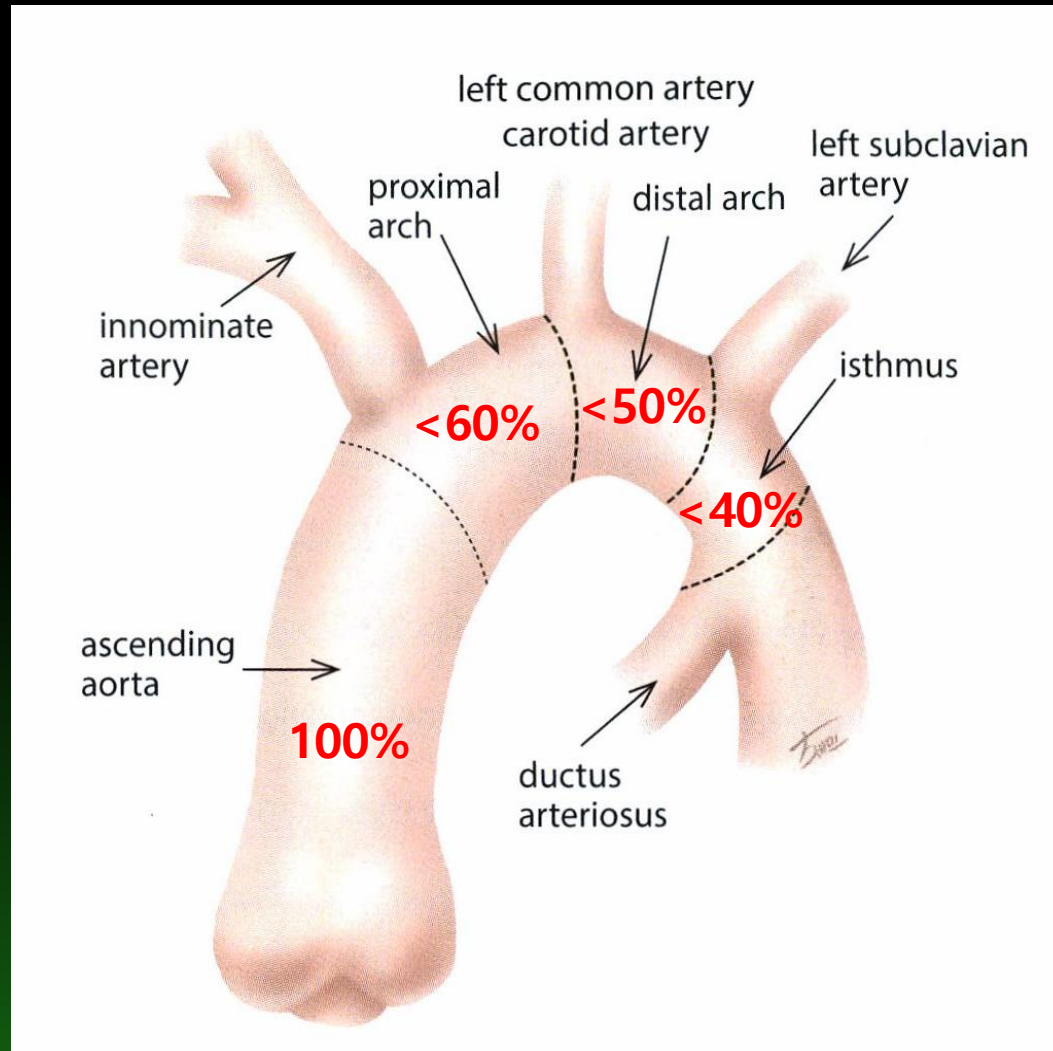
Kyungpook National University Hospital
Department of Thoracic & Cardiovascular Surgery

Youngok Lee

Arch Obstruction

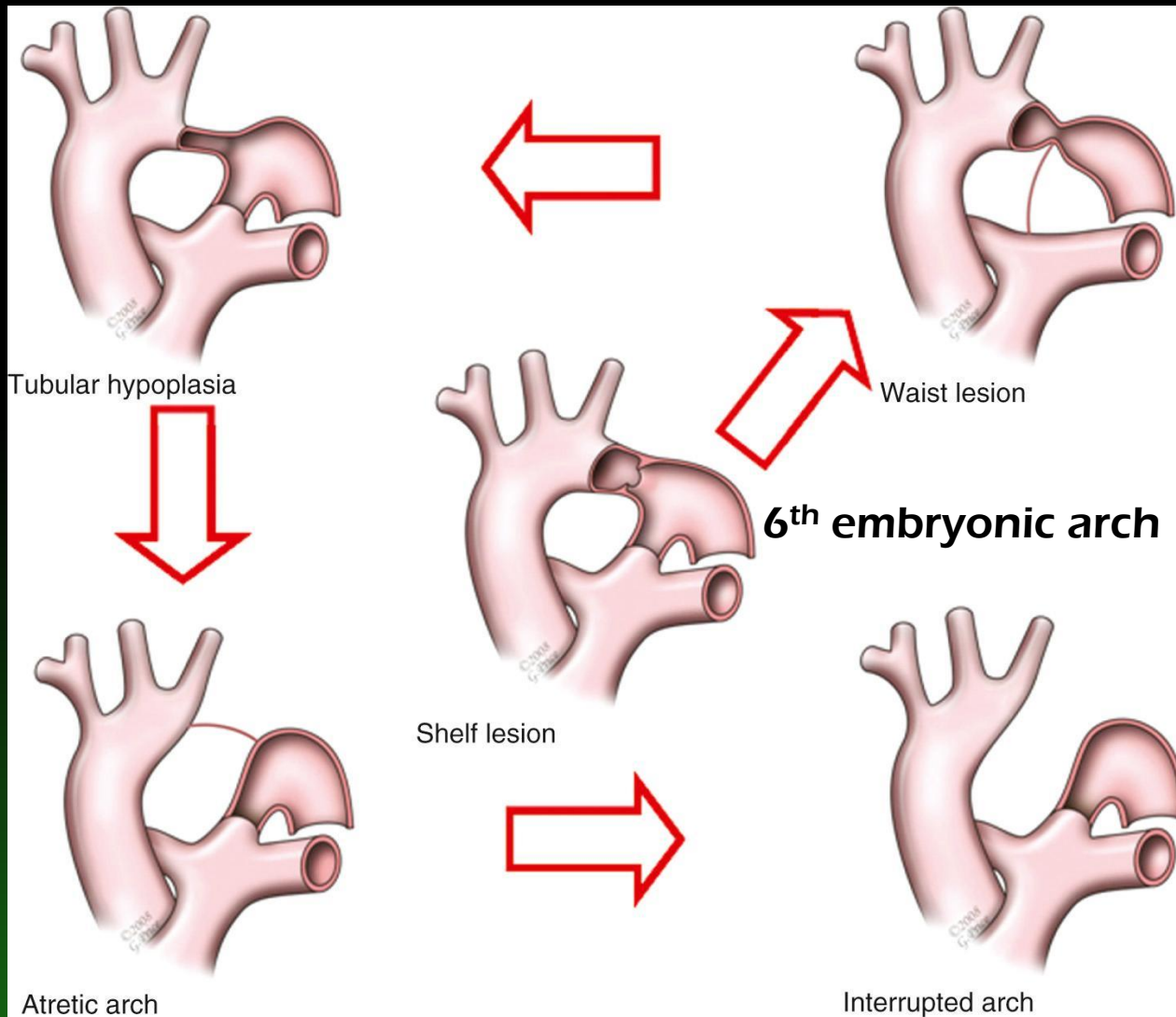
1. Coarctation of the Aorta (CoA) 대동맥 축착
2. Interrupted aortic arch (IAA) 대동맥 단절
3. Hypoplastic left heart syndrome (HLHS)
(좌측 심장 형성 부전증)

Aortic Arch hypoplasia - Anatomy

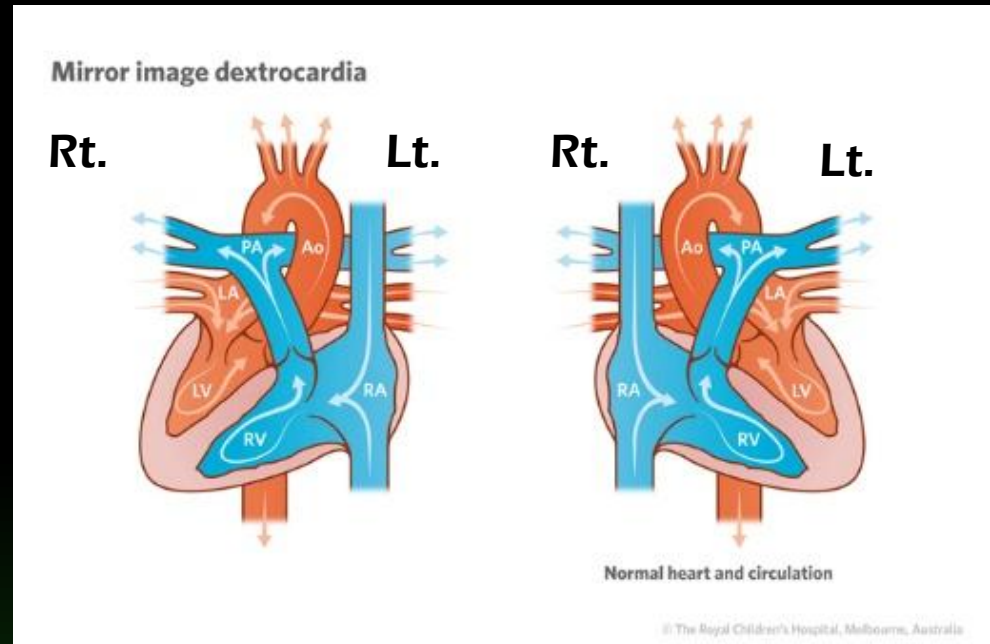
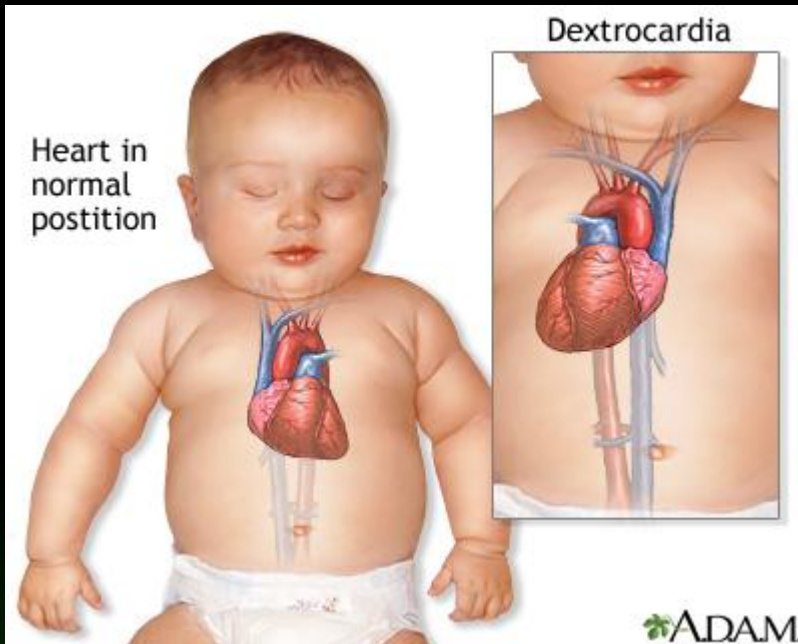


Arch Obstruction

Morphological Spectrum

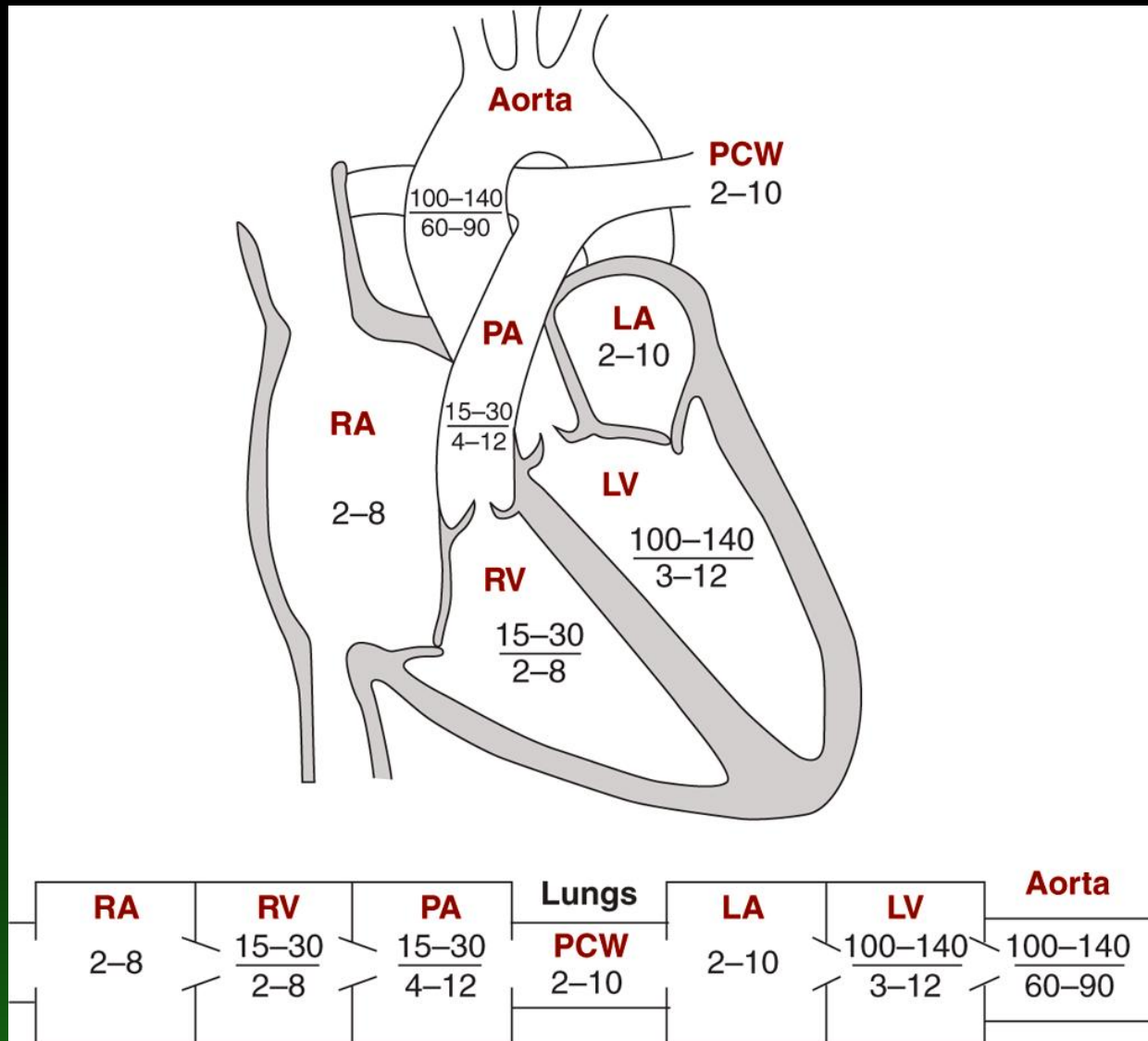


Normal Heart : CHD



Venous blood (Rt.) vs **Arterial blood (Lt.)**
Lt. to Rt. shunt

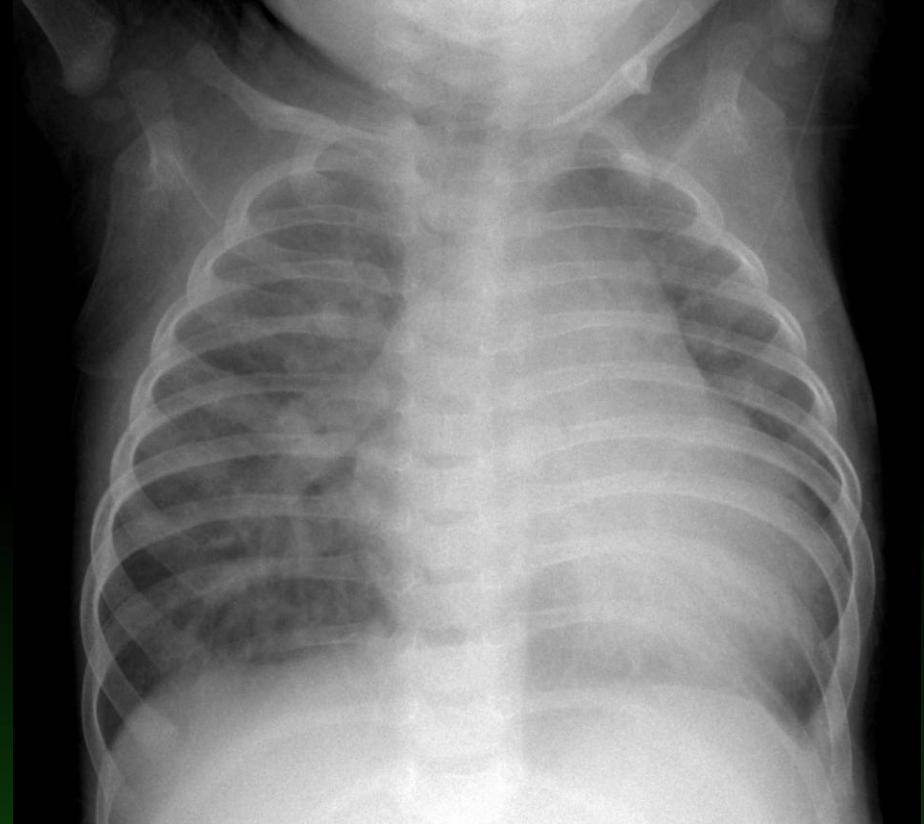
Normal pressures in the cardiac chamber



CHF



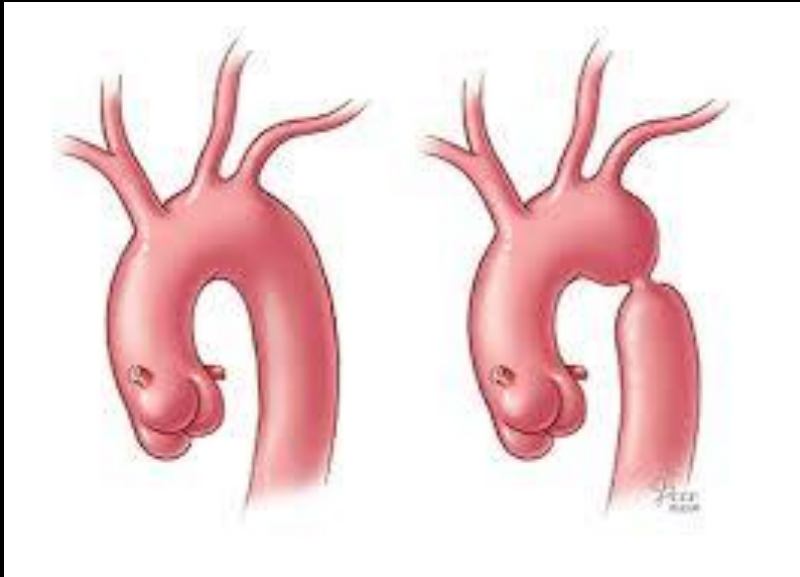
Normal



**pulmonary congestion
cardiomegaly**

Coarctation of aorta (CoA)

Coarctation of aorta (CoA)



Definition

a congenital narrowing of the aorta, usually situated between the origin of the LSCA proximally and the junction of the aorta and the ductus arteriosus distally



Coarctation of aorta (CoA)

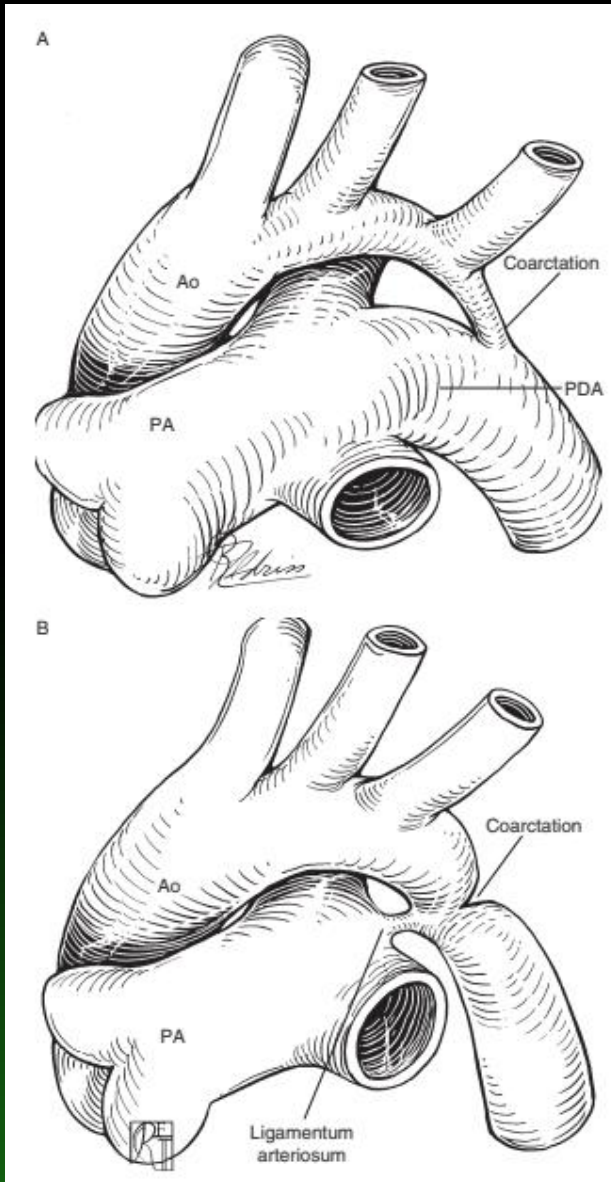
대동맥 축착

In infancy vs in adulthood

Coarctation of aorta (CoA)

A. preductal or infantile
: PDA provides the majority of blood flow to the descending aorta
: often associated with transverse arch hypoplasia

B. postductal or Adult
: juxtaductal narrowing and post. Ridge projecting into the lumen



Natural history

Neonate or infant

- blood flow to the lower body by PDA (PGE1)

- introduction of PGE1 by Elliott *et al.* in 1976

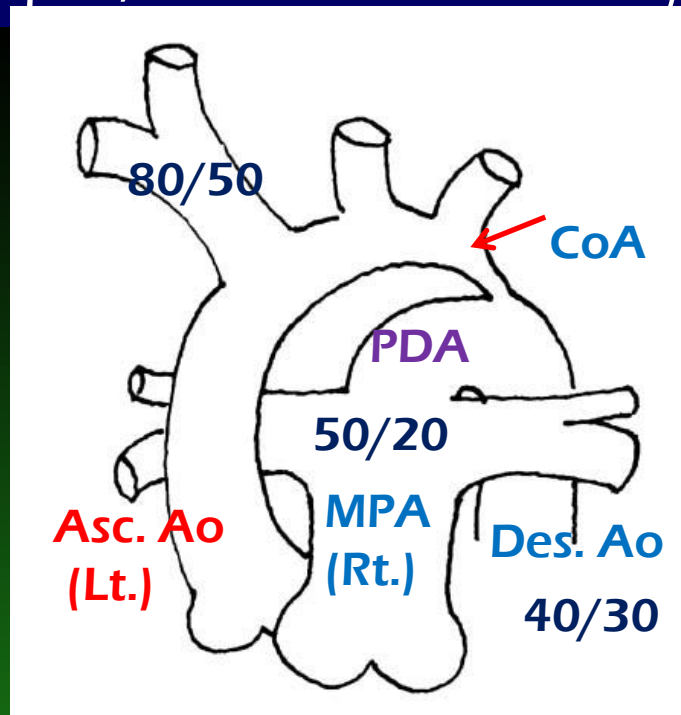
- after ductal closure :

ischemia of organ distal to the coarctation, shock

(poor urine output, metabolic acidosis)

상지 혈압 높다
상지 sat. 높다

하지 혈압 낮다
하지 sat. 낮다.



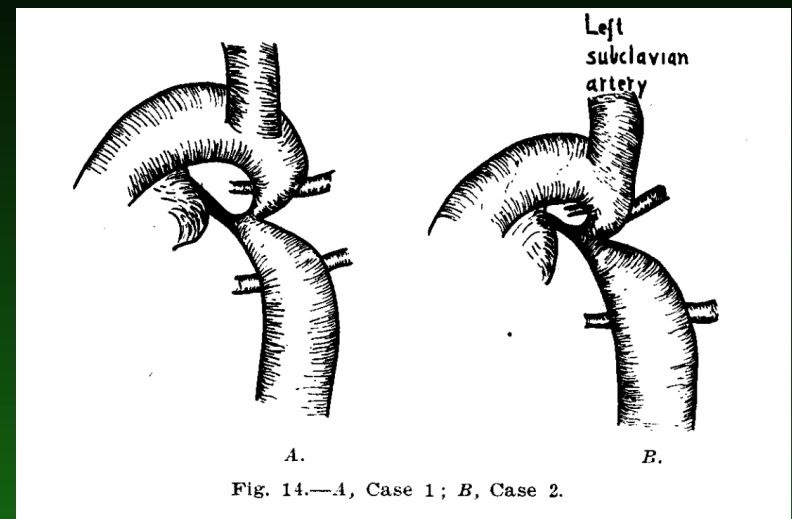
*Rt. to Lt. shunt
through PDA
(mainly systolic phase)*

Operation - history

The first repair was performed by Crafoord and Nylin in October 1944 (Sweden)

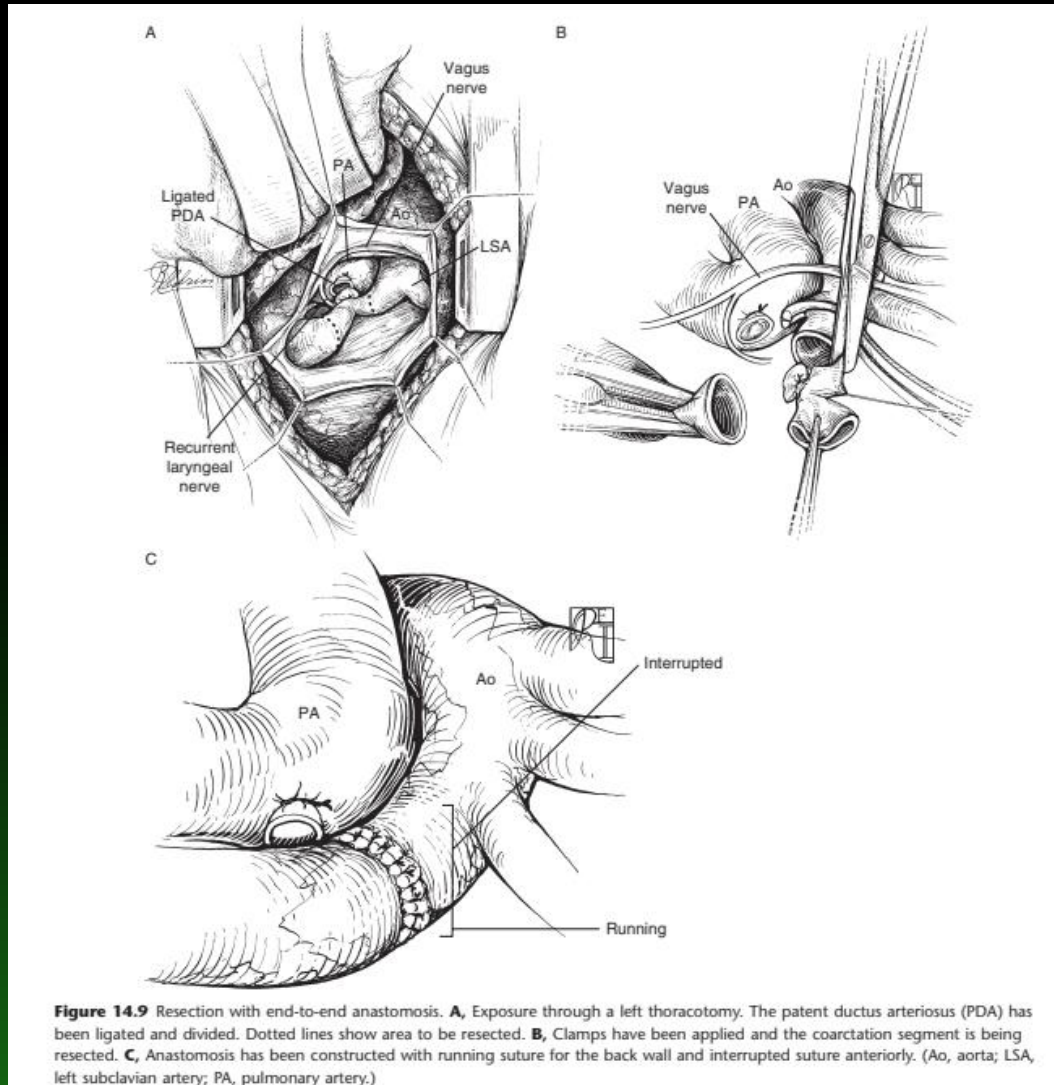
J Thorac Surg 1945;14:347-361

: resection of the coarctation with end-to-end anastomosis (Carrel technique), in a 12 year old boy and a 27 year old man with severe HTN



Operation

Resection with end to end anastomosis



Operation

Resection with end to end anastomosis

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

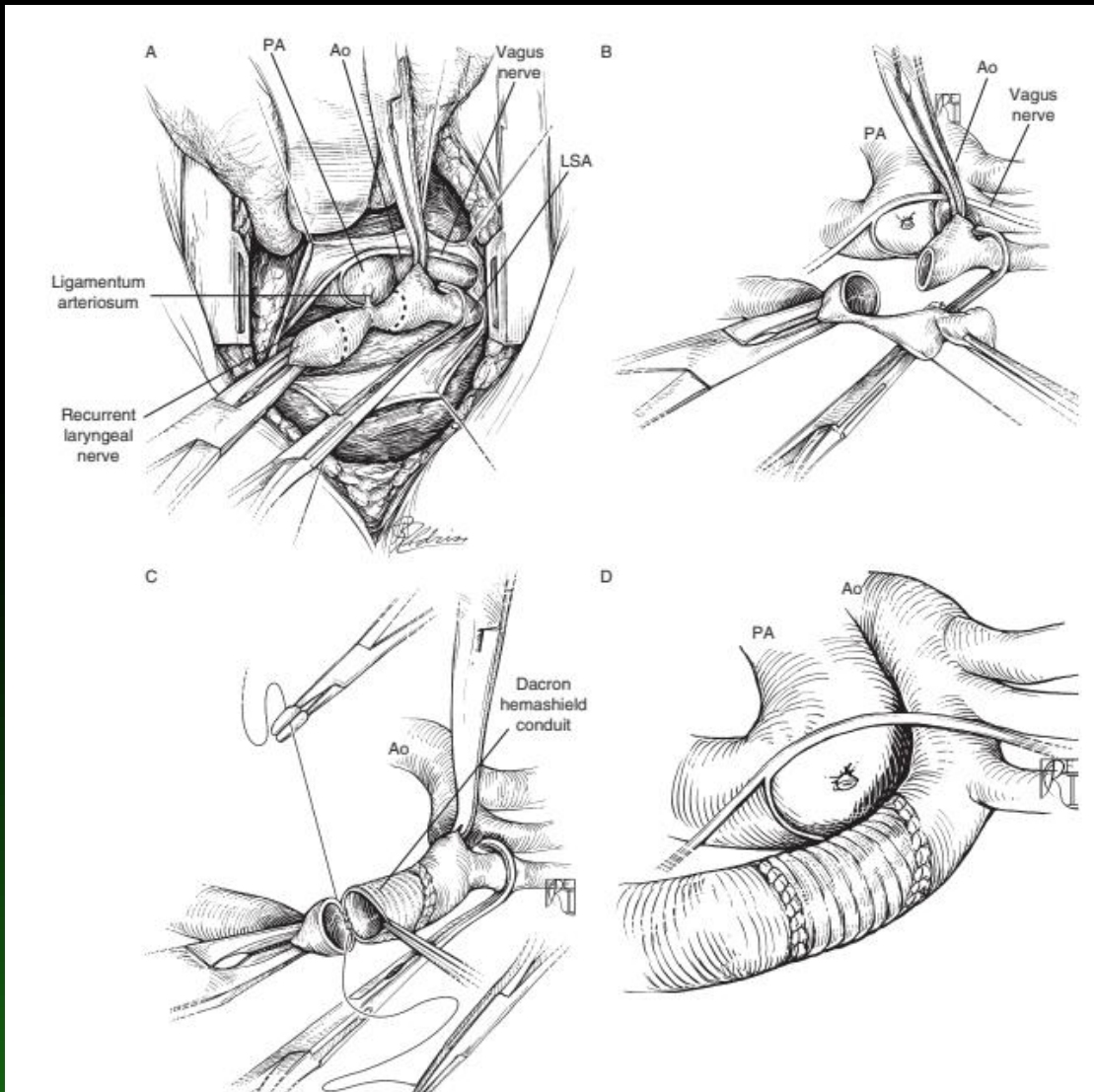
Author	Age	Year	Patients	Mortality	Recoarctation
Williams [66]	<1 yr	1980	176	66 (38%)	39 (33%)
Cobanoglu [67]	<3 mo	1985	55	16 (29%)	3 (8%)
Körfer [68]	<4 mo	1985	55	2 (4%)	3 (6%)
Ziemer [69]	<1 mo	1986	24	8 (33%)	4 (25%)
Brouwer [70]	<2 yr	1991	32	2 (6%)	4 (13%)
Kappetein [71]	<3 yr	1994	48	5 (10%)	41 (86%)*
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%)

*Kaplan-Meier estimate at 30 years.

^bCongenital Heart Surgeon's Society.

Operation

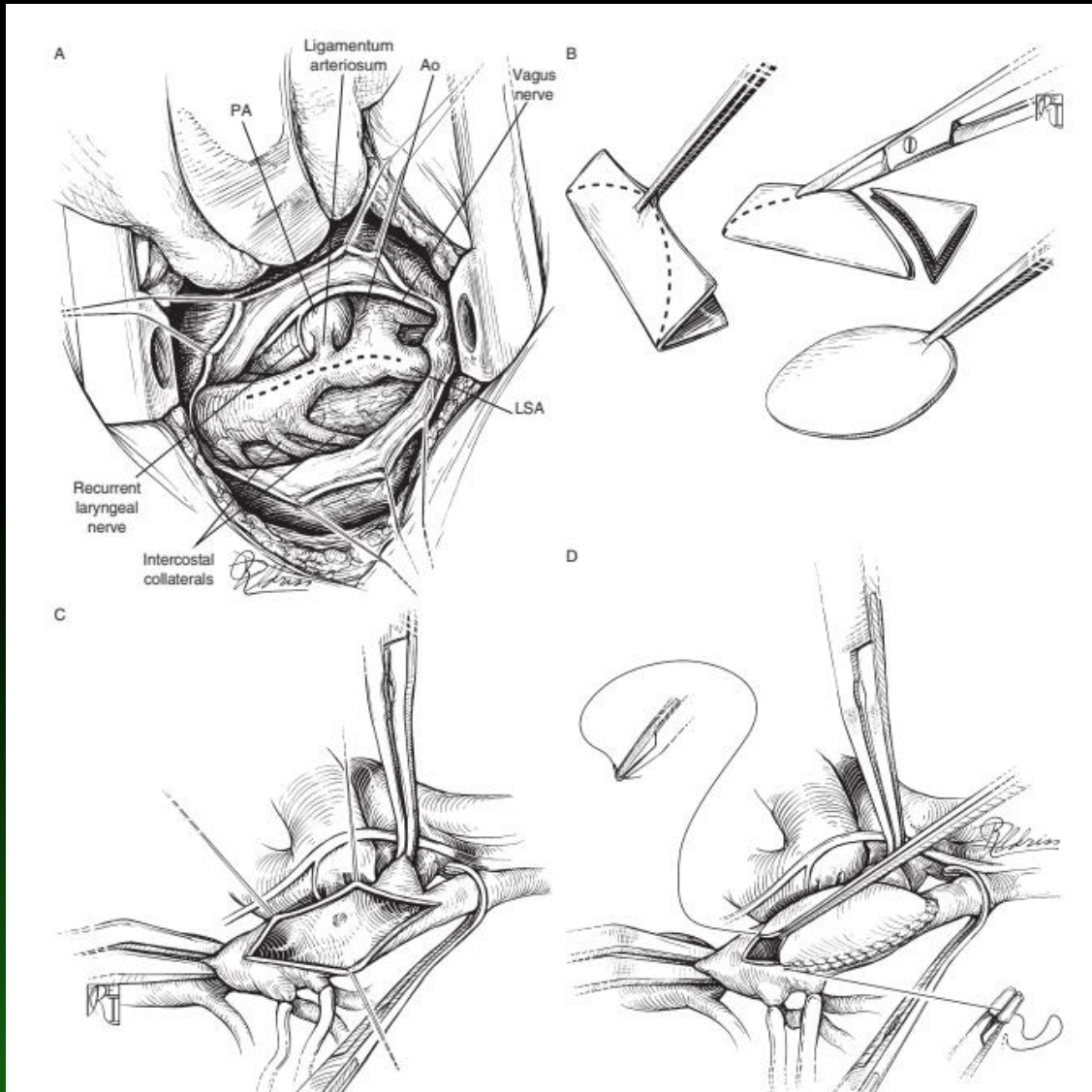
Prosthetic interposition Graft



First described
by Robert Gross
in 1951
: using homograft
over 10 yrs of age
(adult sized graft)

Operation

Prosthetic Patch Aortoplasty

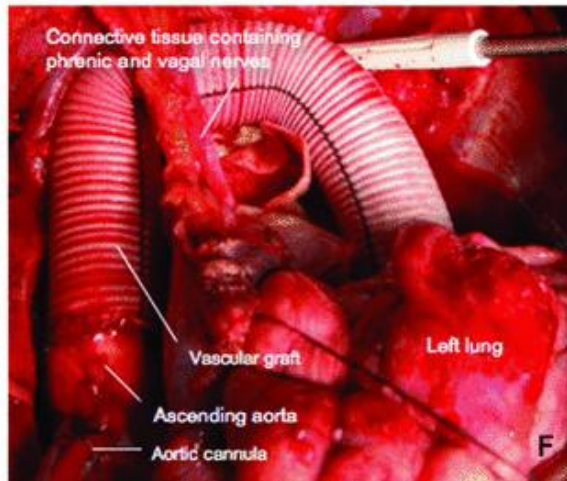
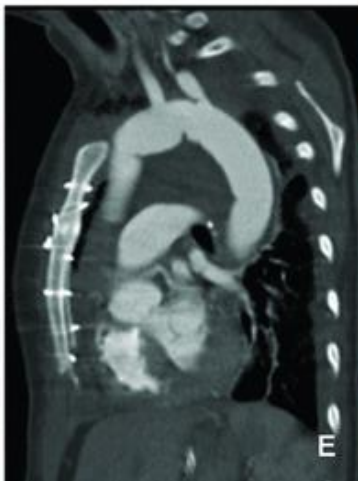
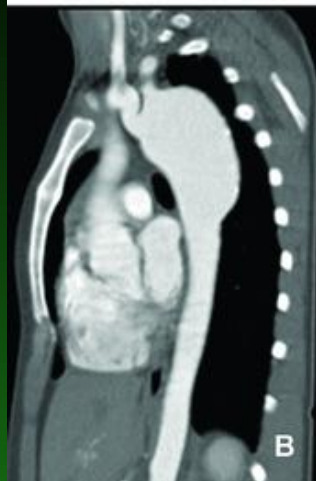
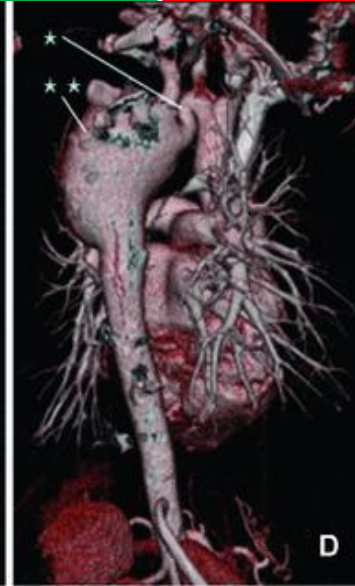


Vosschulte in 1957
Germany
“isthmusplastic” procedure

Operation

Prosthetic patch aortoplasty

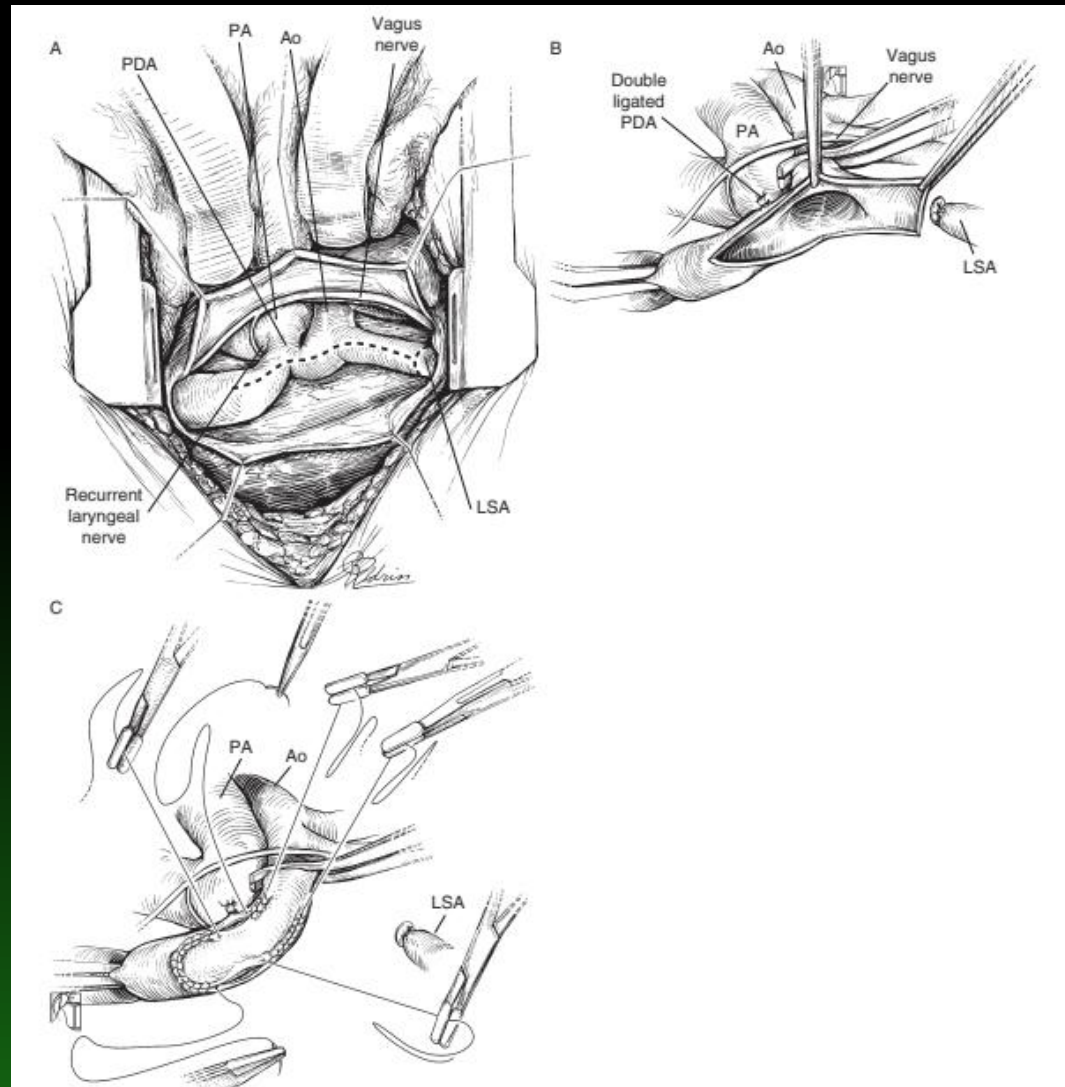
Table 14.3 Results of patch aortoplasty. (NS, not specified; PTFE, polytetrafluoroethylene,)



on	Aneurysm	Patch
	0	PTFE
	5 (13%)	Dacron
	18 (6%)	Dacron
	3 (5%)	Dacron
	0	PTFE
	0	PTFE
	8 (7%)	PTFE
	34 (4%)	

Operation LSCA flap aortoplasty

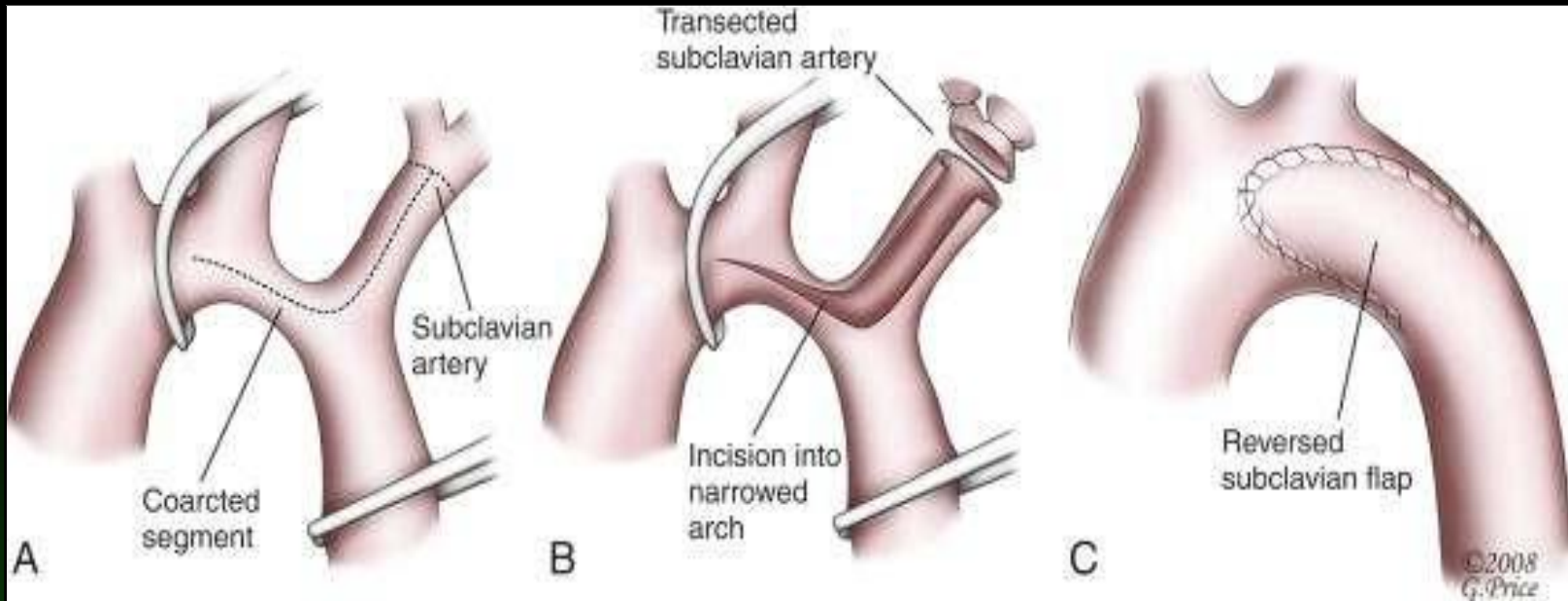
by Waldhausen and Nahrwold,
USA
in 1966



Operation

Reversed LSCA flap aortoplasty

By Hart and Waldhausen,
in 1983



Operation

LSCA flap aortoplasty

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

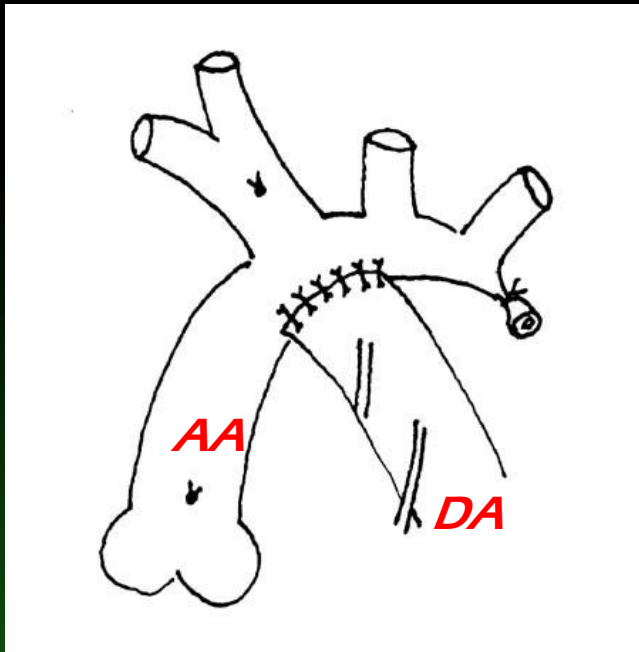
Author	Age	Year	Patients	Mortality	Recoarctation
Metzdorff [106]	<2 mo	1985	60	11 (18%)	10 (17%)
Ziemer [69]	<1 mo	1986	70	8 (11.4%)	9 (15%)
Ehrhardt [108]	<1 mo	1989	45	14 (31%)	7 (23%)
Milliken [97]	<1 mo	1990	123	11 (9%)	20 (16%)
Van Heurn [13]	<3 mo	1994	15	1 (7%)	6 (42%)
Quaegebeur [72] ^a	<1 mo	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%)

^aCongenital Heart Surgeon's Society.

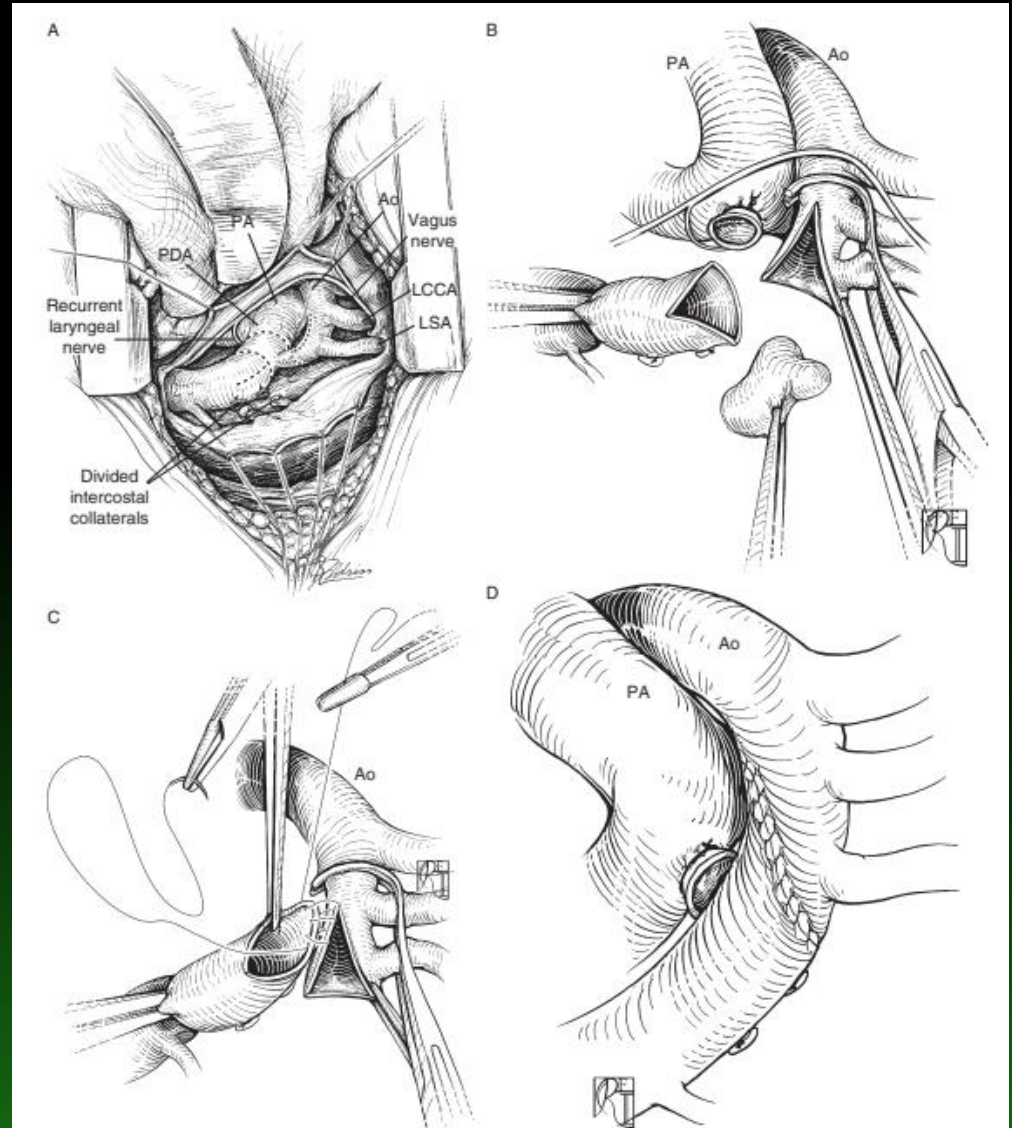
Operation

Resection and Extended end to end anastomosis

By Amato,
USA
in 1977



Extended end to side A.



Operation

Resection and Extended end to end anastomosis

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

Author	Age	Year	Patients	Operative mortality	Recoarctation
Lansman [9]	6mo	1986	17	1 (6%)	2 (12%)
Vouhé [19]	3mo	1988	80	21 (26%)	6 (10%)
Zannini [112]	3mo	1993	21	4 (19%)	4 (23%)
Van Heurn [13]	3mo	1994	77	5 (6%)	8 (11%)
Kappetein [71]	3yr	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%)

Operation

Sternotomy vs Thoracotomy

Median Sternotomy

Complete aortic arch exposure
Extensive arch reconstruction
Concomitant intracardiac repair
Antegrade cerebral perfusion

Requires CPB

Longer ICU and hospital stay
Potential CPB-related complications

Severe arch hypoplasia

Intracardiac defects requiring repair

Bovine arch with distal arch hypoplasia

Left Thoracotomy

Avoidance of CPB
Shorter operative time and recovery
Fewer CPB-related complications

Limited proximal arch exposure

Risk of spinal cord ischemia

Risk of residual arch obstruction

Isolated CoA

Mild arch hypoplasia

Favorable arch anatomy

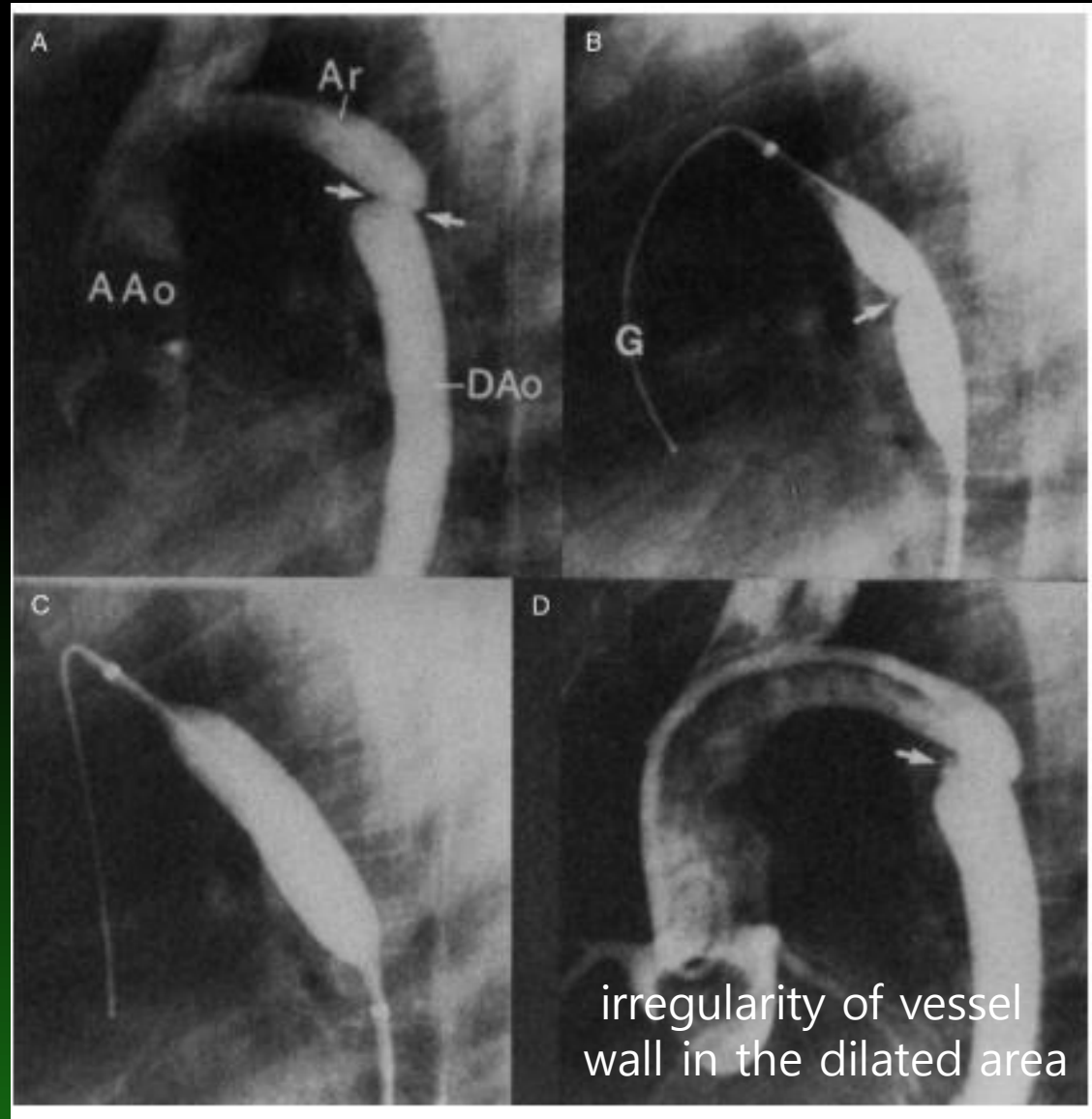
Advantages

Disadvantages

Preferred lx.

Balloon Dilatation Angioplasty

By Sos,
in 1979,
recurrent CoA



Potential complication of surgery

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

Complication

Persistent or Recurrent CoA

- Resting peak pressure gradient exceeding 20 mmHg across the repair area
- Possible causes of true recoarctation
 - 1) Lack of growth of the suture line
 - 2) Presence of abnormal mesodermal tissue (proliferates and produces marked intimal and medial hypertrophy)
 - 3) Remnants of ductal tissue
 - 4) Damage to the aorta from the vascular clamps used at repair

Complication

Persistent or Recurrent CoA

- Technical factors of persistent coarctation

- 1) Insufficient resection of a long, narrow segment
- 2) Excessive tension due to inadequate mobilization
- 3) Incorrect fashioning of a subclavian flap or polyester on lay patch
- 4) Failure to resect an obstructing intimal ridge
- 5) Use of a too-small tube graft in a child

Complication

Paradoxical postop. HTN

■ Two phases

- Phase 1 (immediate, <24 h): ~50% of patients
 - Baroreceptor resetting (in the carotid a.) →
 - sympathetic activity ↑ (norepinephrine ↑)
- Phase 2 (48-72 h): ~ 1/3 of Phase 1 patients; diastolic predominant
 - Renin-angiotensin activity ↑

Complication

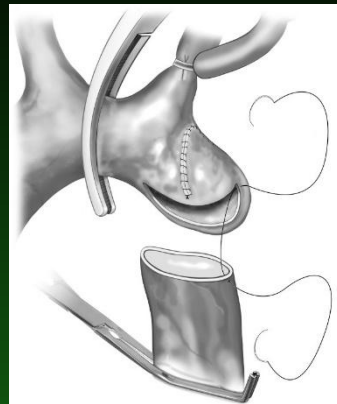
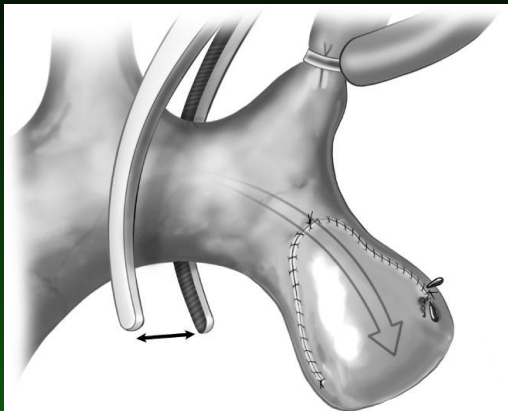
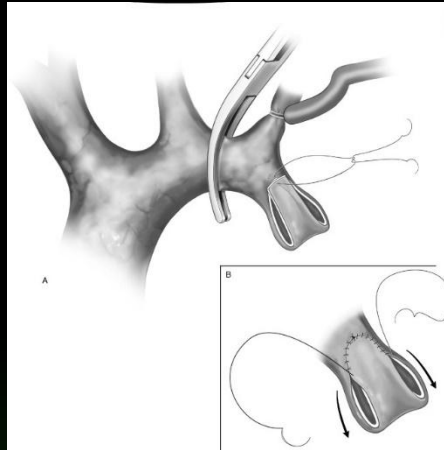
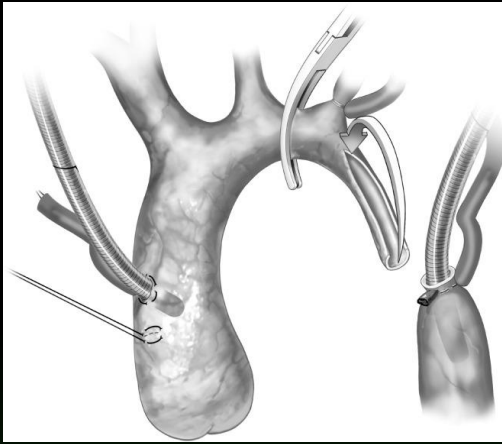
Late HTN

- Coarctation should be regarded not as a localized abnormality but as a **diffuse arteriopathy** deserving long-term follow-up.
- **diffuse arteriopathy**
 - 1) Structural vascular changes
 - 2) Increased arterial stiffness
 - 3) Alterations in baroreceptor function and blood pressure regulation
 - 4) Diffuse vascular abnormalities
 - 5) Recurrent coarctation of the aorta
 - 6) Gothic arch geometry

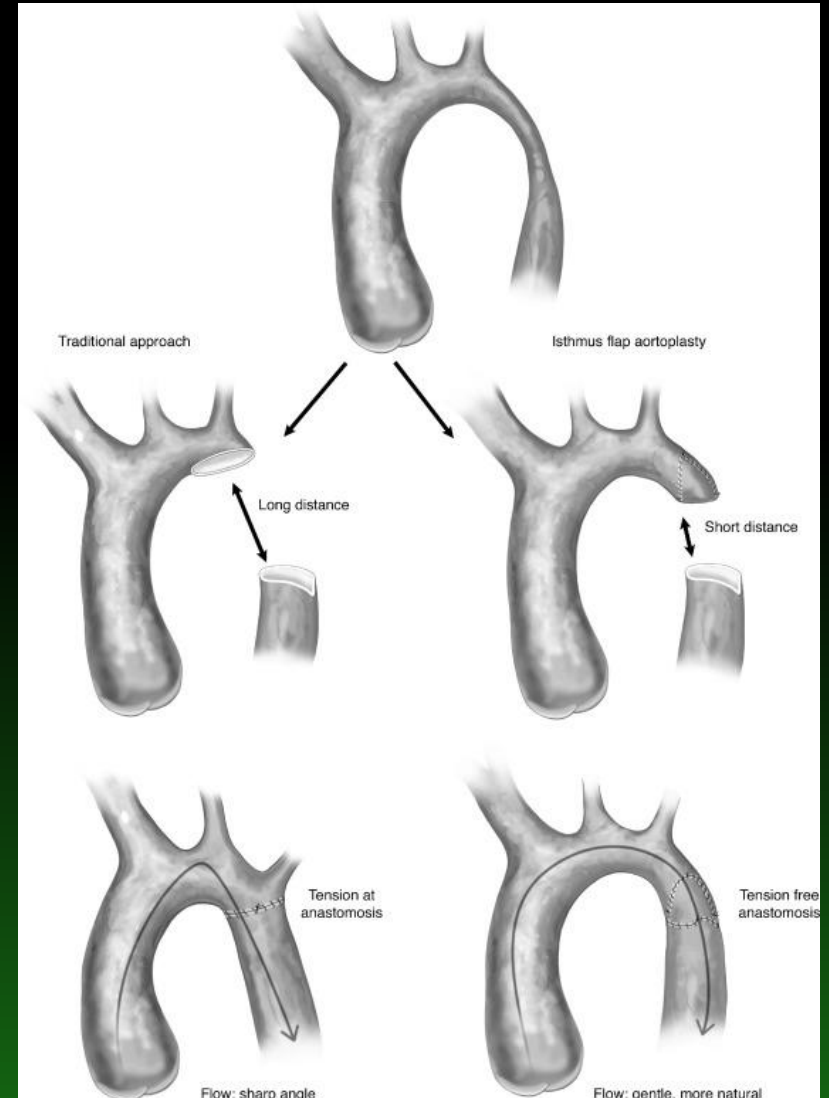
Complication

Late HTN

- Gothic arch geometry

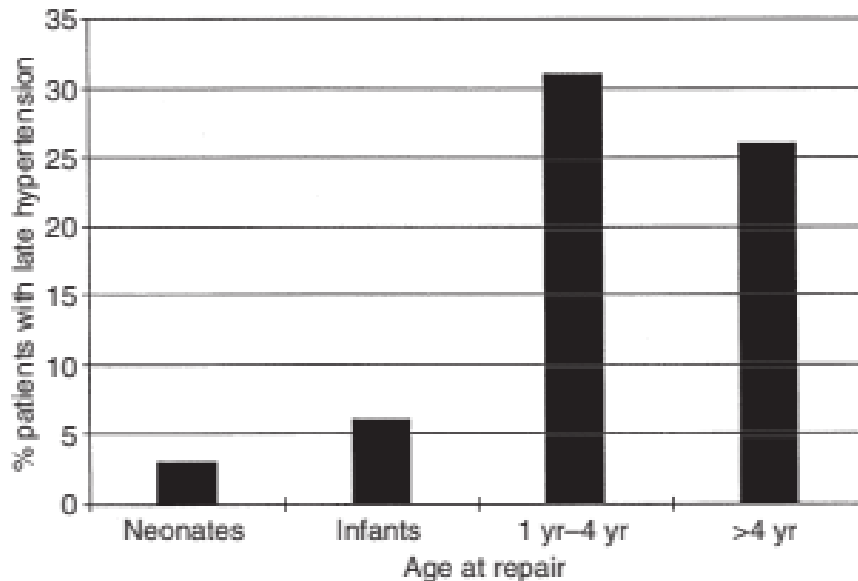


Operative Techniques in Thoracic and Cardiovascular Surgery, Volume 26, Issue 3, Autumn 2021, Pages 536



Complication

Late HTN



*Seirafi PA, et al. (1998)
Ann Thorac Surg 66, 1378-1382.*

Percentage of patients with persistent or late hypertension listed according to age at operation.

The percentage of patients with late hypertension was significantly less in the neonatal and infant groups ($n = 48$, 4.2%) than in toddlers and older children ($n = 59$, 27.1%; $p < 0.002$). YR, years

Complication

Abdominal pain – Mesenteric arteritis

- In 10% to 20% of cases,
- Mesenteric arteries are accustomed to low pressure, so a sudden rise can cause mesenteric arteritis and ischemia during the first 5 or 6 postoperative days (severe abdominal pain, distension, tenderness, GI bleeding, sometimes requiring bowel resection).

Complication

Paraplegia

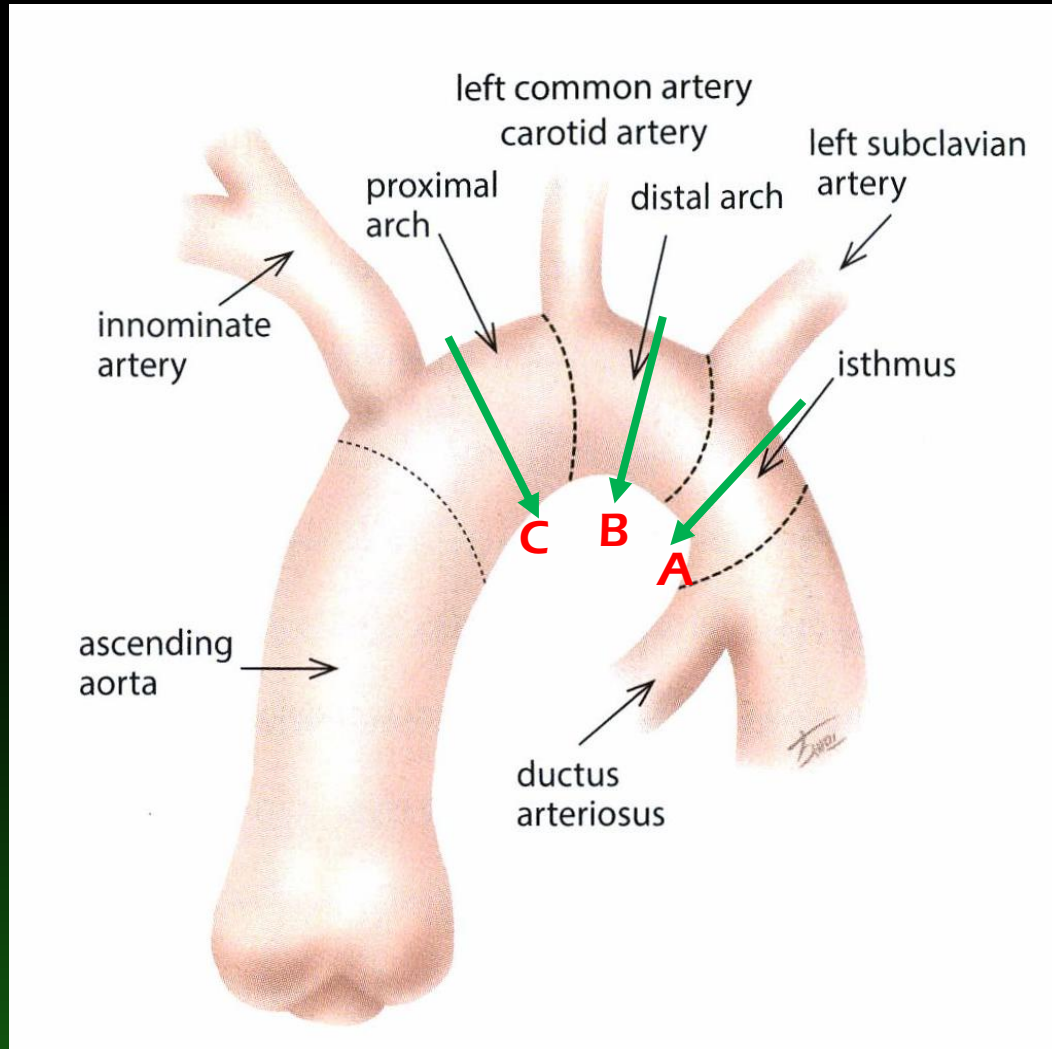
■ *Prevention*

1. Aortic cross-clamp time 최소화
2. Mild hypothermia (34–35°C) 유지
3. Proximal blood pressure 유지 및 acidosis 예방
4. Distal mean arterial pressure >45 mmHg 유지
5. Test clamping 시 distal MAP <45 mmHg이면
partial left heart bypass 시행

Interrupted Aortic Arch (IAA)

대동맥 단절

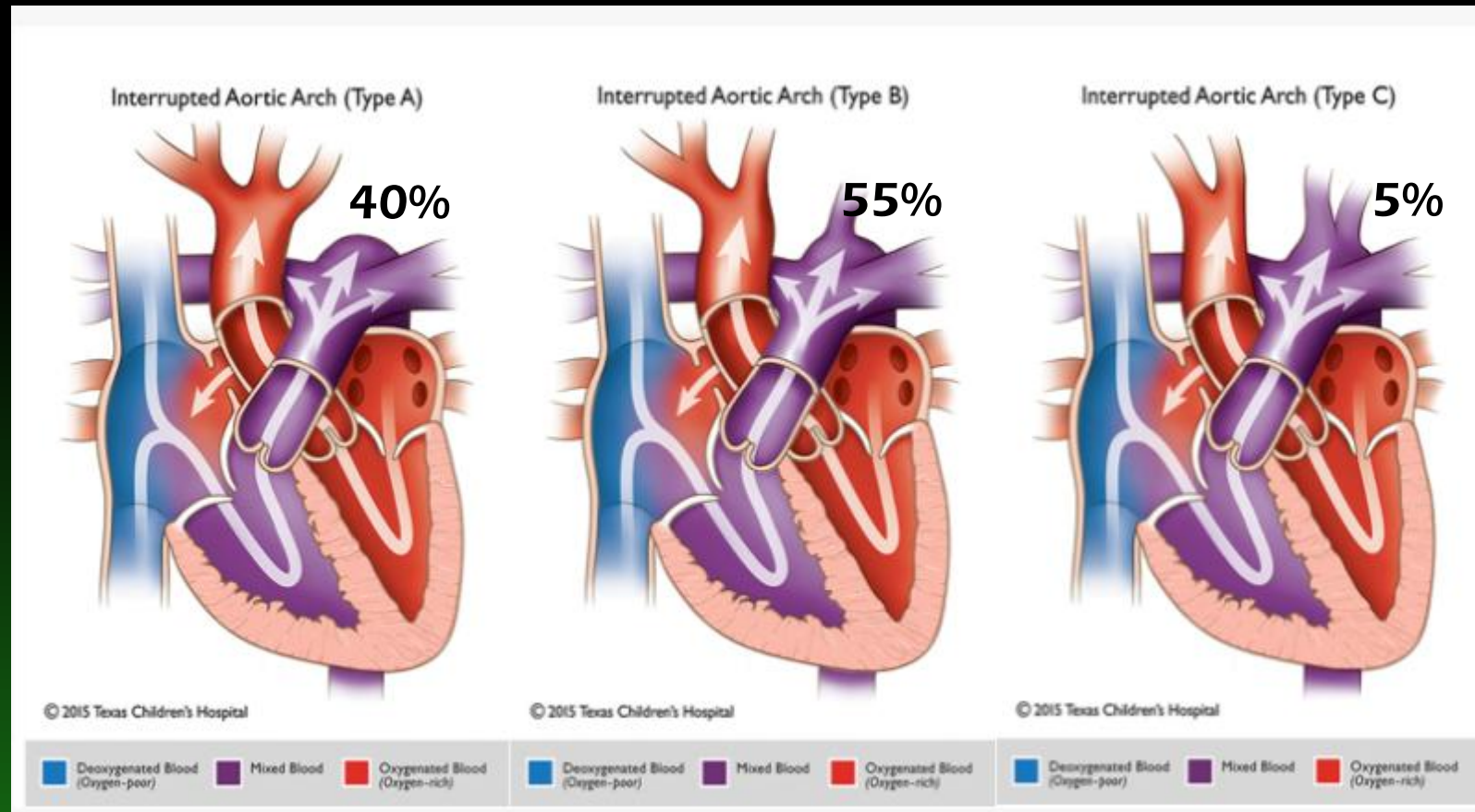
Aortic Arch Anatomy



Interrupted Aortic Arch (IAA)

Definition

Complete luminal and anatomic discontinuity between two segments of the aortic arch



IAA – coexisting anomaly

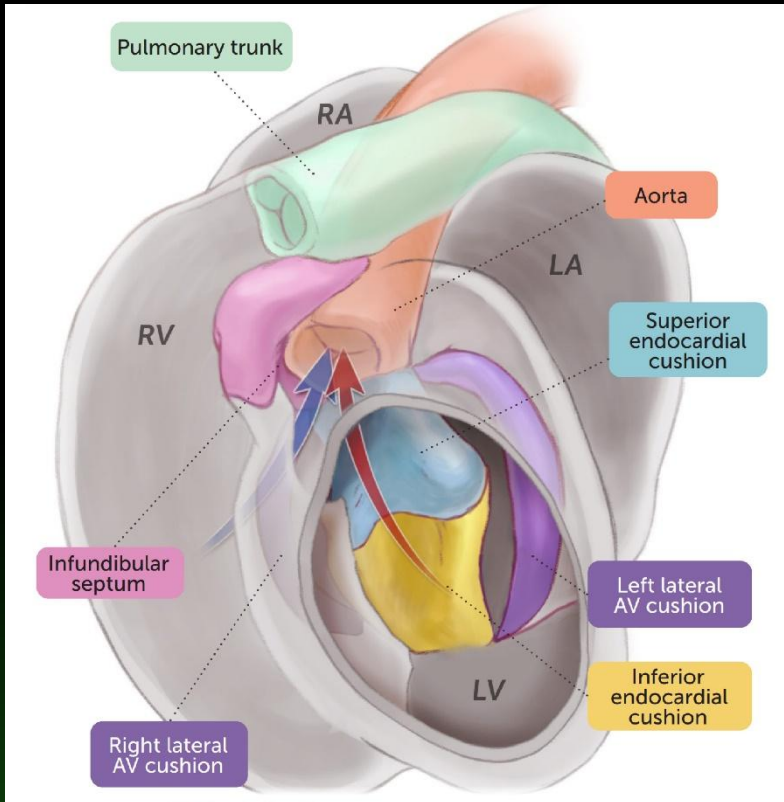
- *Left Ventricular Outflow Anomalies*

- : **Bicuspid aortic valve** in 30% to 50% of patients
- : Subaortic stenosis may be present or develop
- : Small aortic annulus (half size)

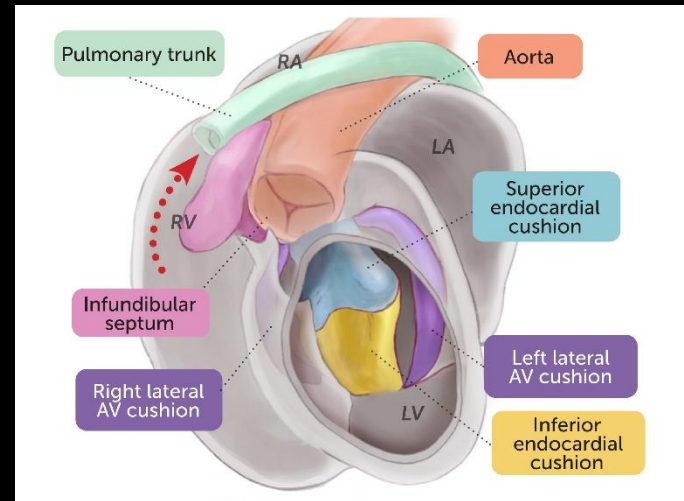
- *Coexisting Cardiac Anomalies*

- : **A large VSD in over 95%**, Mostly conoventricular type
- : Posteriorly malaligned and displaced infundibular septum usually produces subaortic obstruction of variable severity
- : Truncus arteriosus, AP window, Taussig-Bing anomaly
- **DiGeorge syndrome** is frequent
(up to half of type B IAA)

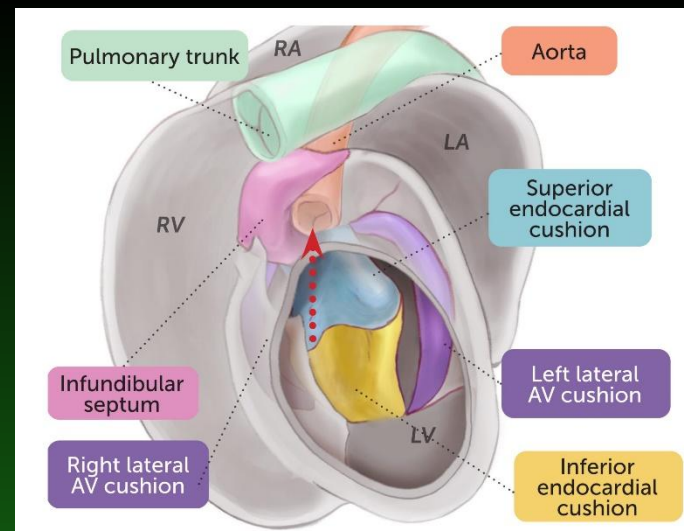
Malaligned and displaced infundibular septum



Normal



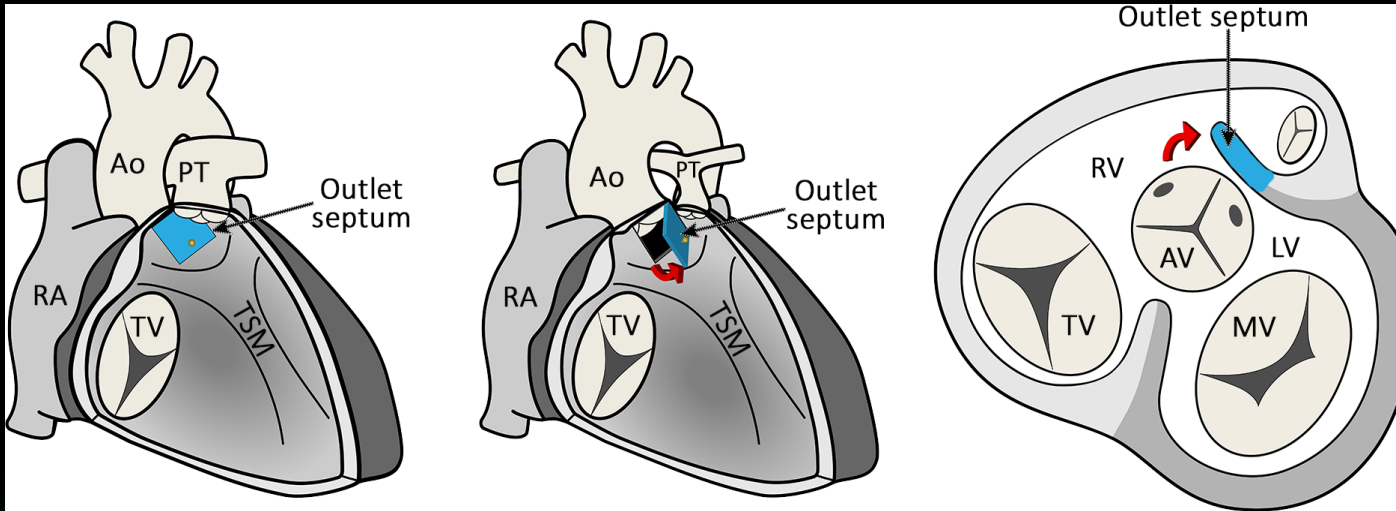
Anterior malalignment defect



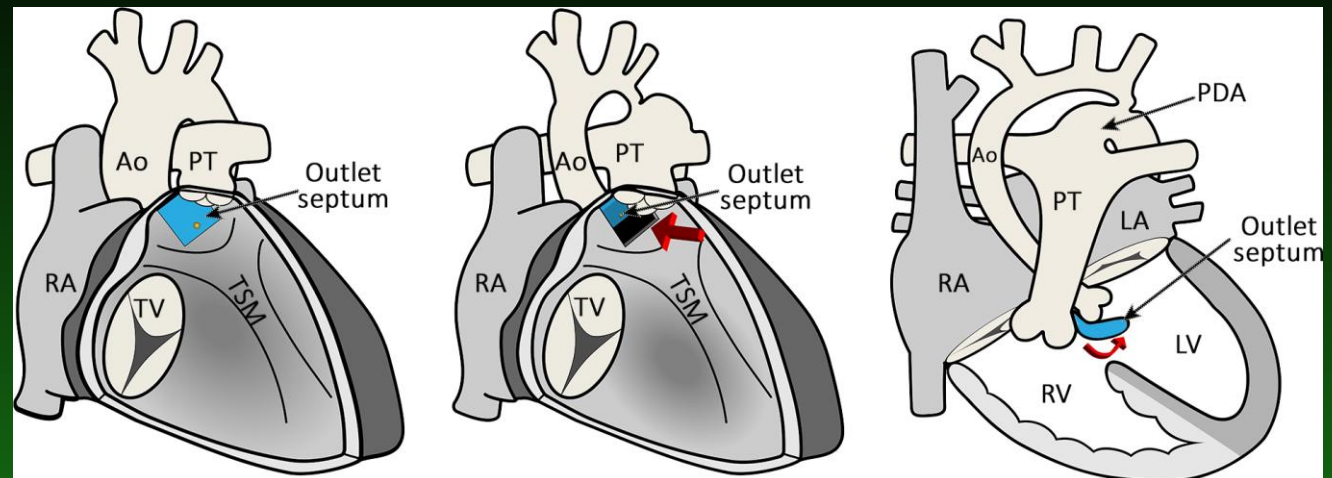
Posterior malalignment defect

Malaligned and displaced infundibular septum

Anterior malalignment defect

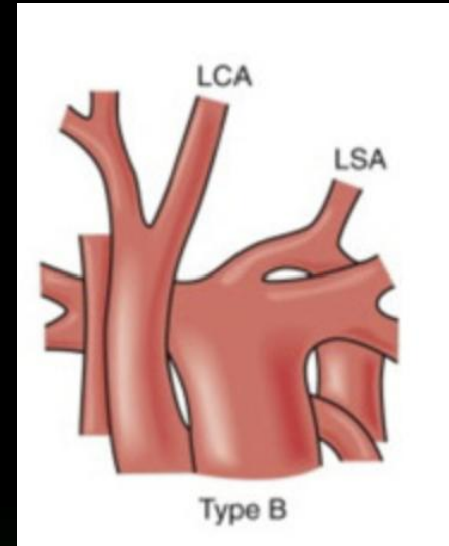


Posterior malalignment defect



Clinical Features, Management

- Large Intracardiac shunt, high afterload
→ Severe heart failure
- PDA closes
→ Metabolic acidosis, Anura,
Femoral pulse ↓

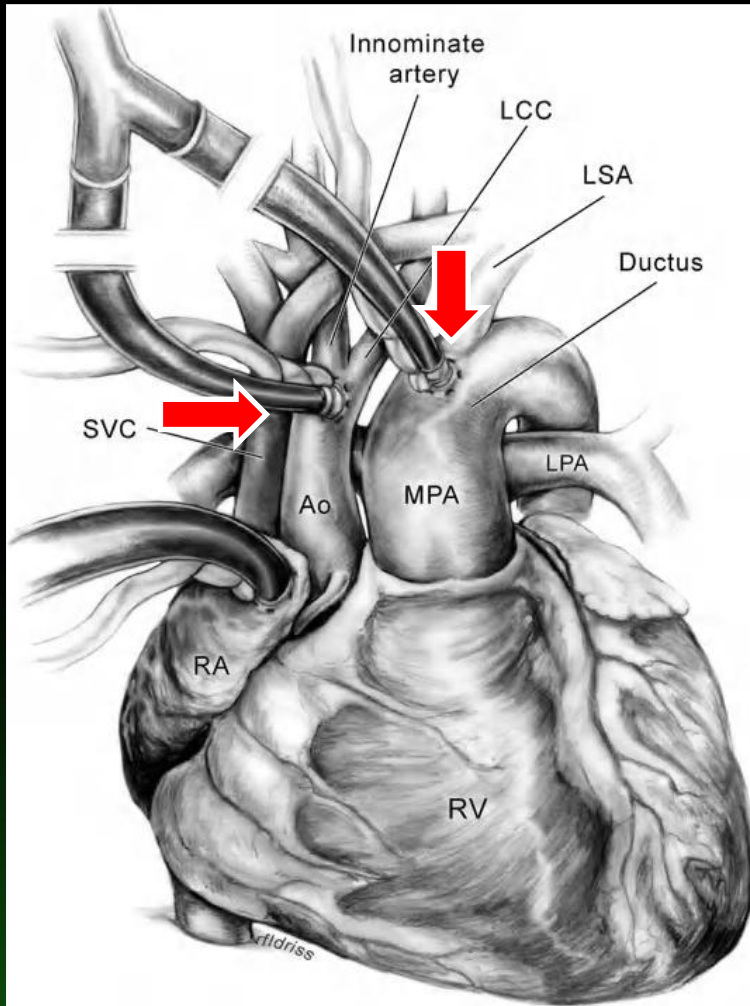


- Prostaglandin E1 keep (through a secure IV line)
- It is important that **pulmonary vascular resistance be maximized** : avoiding a high level of inspired oxygen, avoiding respiratory alkalosis (hyperventilation), PaCO₂ level 40-50mmHg

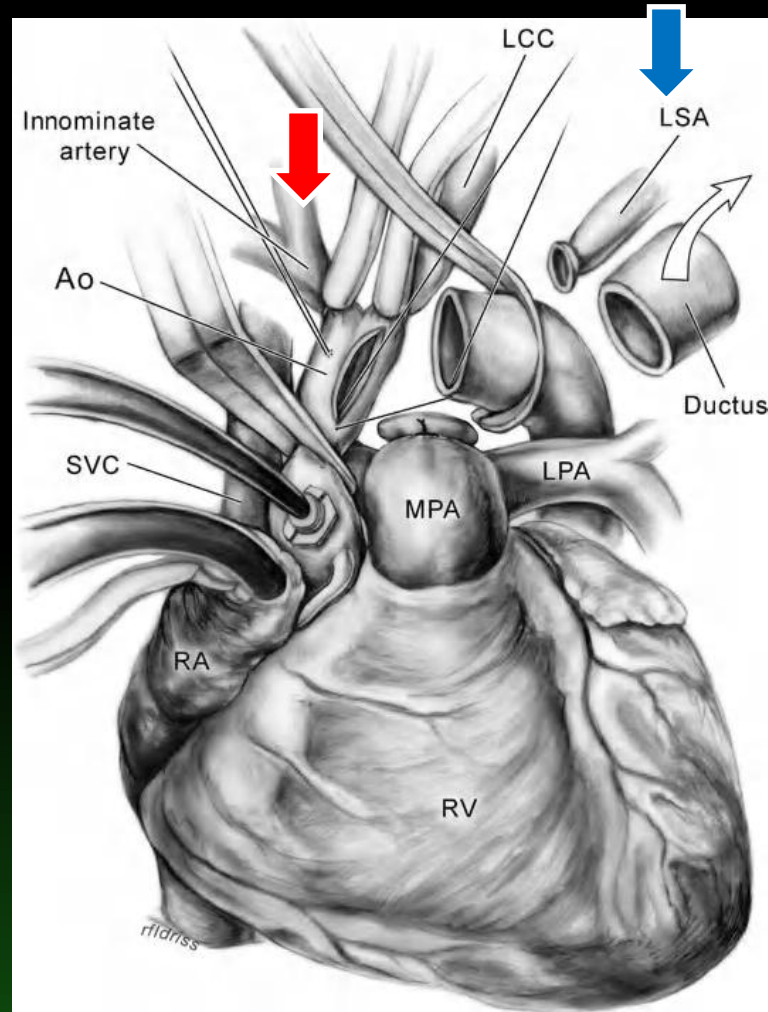
IAA – History

- **1955, First Successful Repair**
by Merrill et al. (Type A IAA, via direct anastomosis).
Associated VSDs were left unclosed
- **1975**
by Trusler (1975): First, one-stage repair via direct arch anastomosis.
- **1976, Revolution in Survival (PGE1)**
 - Elliott et al. introduced Prostane (PGE1).
 - Radically reduced the historically extreme mortality risk of IAA.
- **1996 | Cerebral Protection**
 - By Sakurada et al. & Asou et al. selective cerebral perfusion to minimize/eliminate circulatory arrest.

IAA – Operation

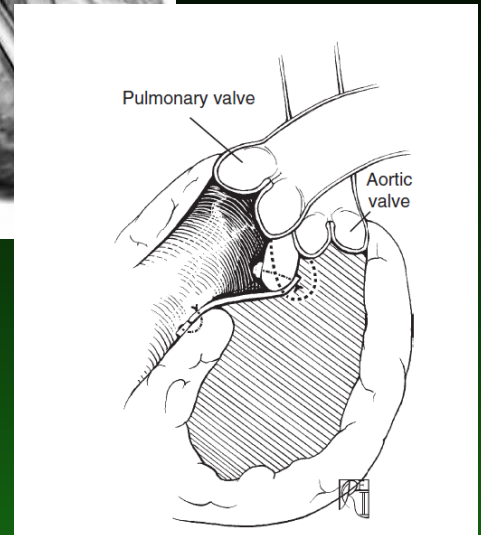
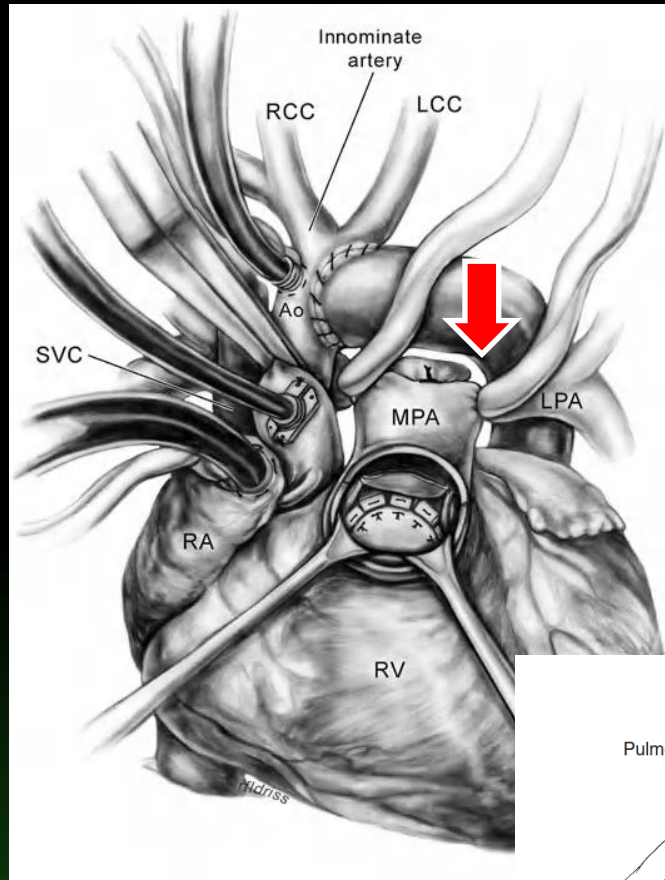
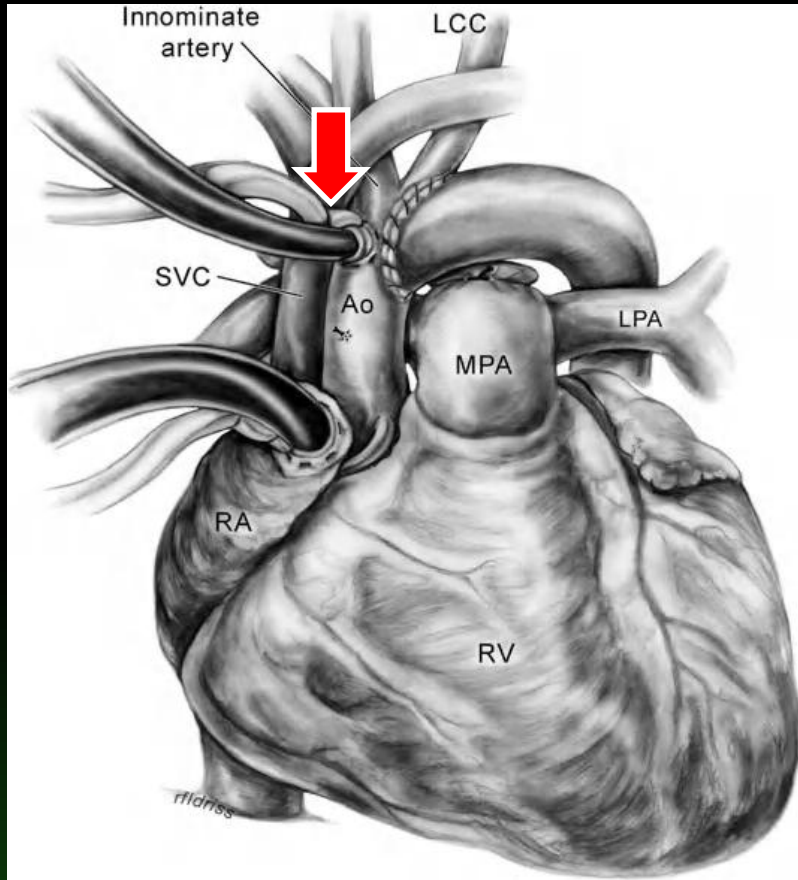


Two arterial cannulations



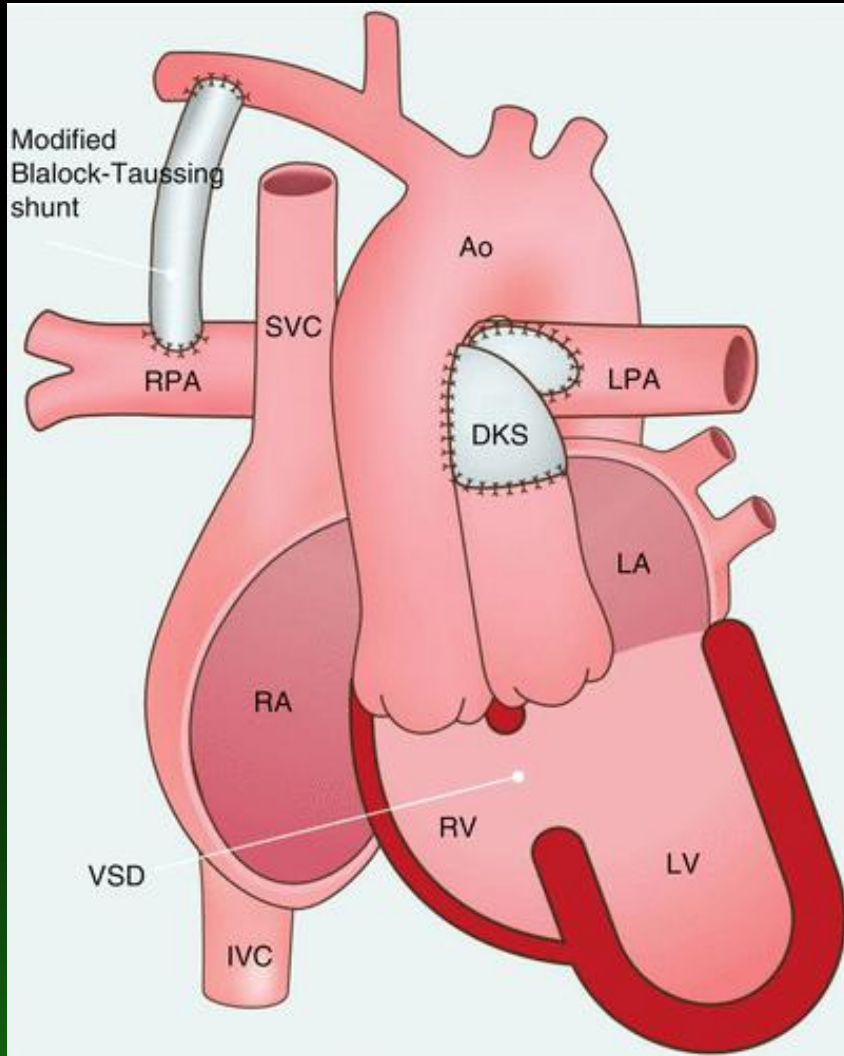
Hypothermic circulatory arrest
vs selective cerebral perfusion

IAA – Operation

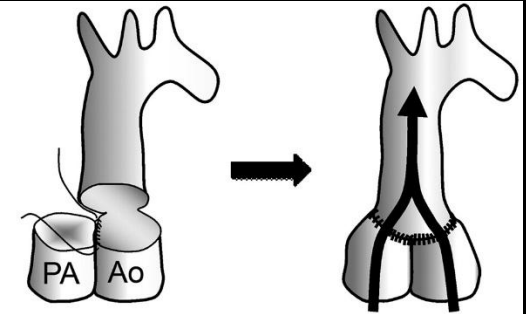


Damus–Kaye–Stansel operation

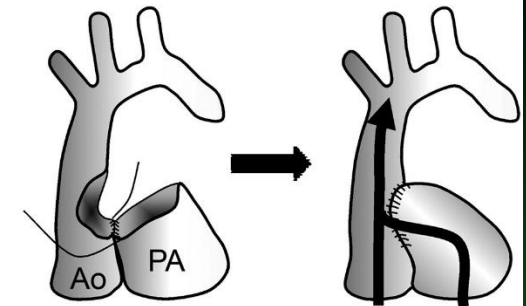
: systemic ventricular outflow tract obstruction



Double Barrel DKS

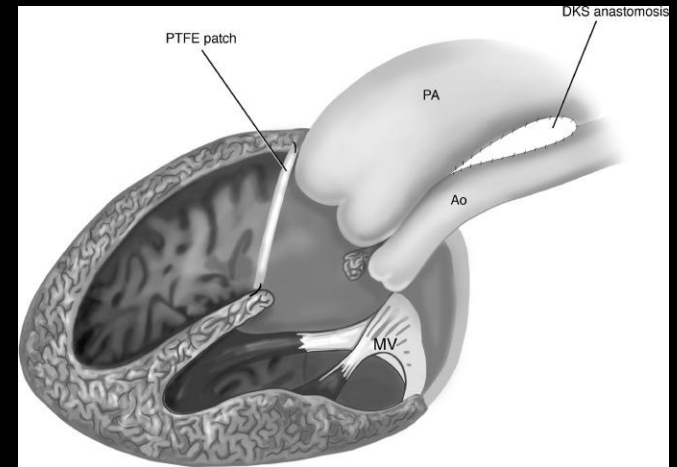
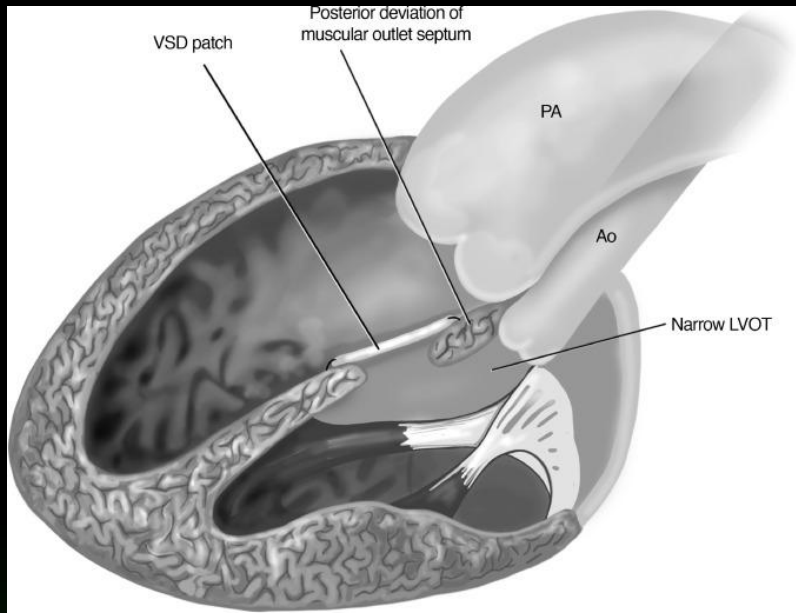


End-to-Side DKS



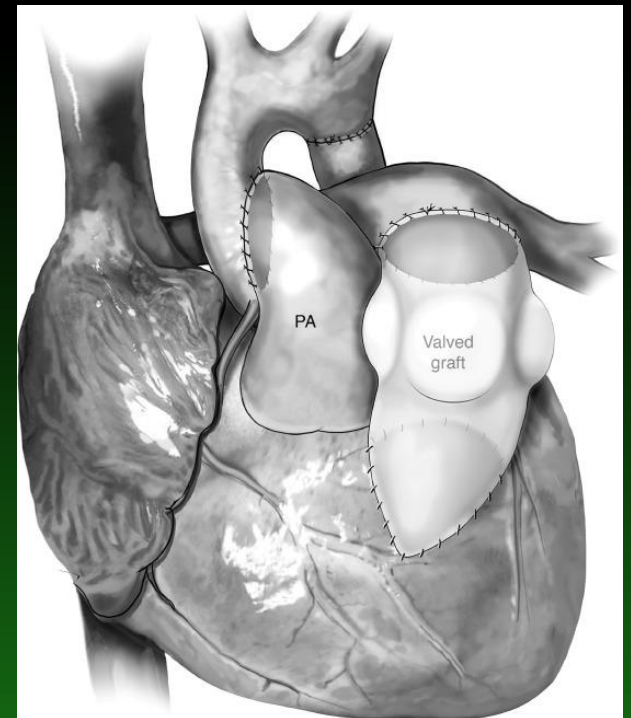
Single ventricular repair

IAA, VSD, severe subaortic stenosis

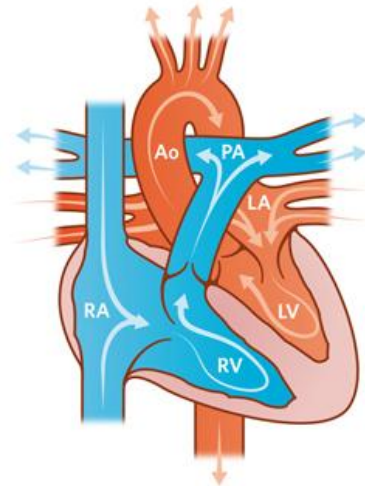


Biventricular repair

Modification of the Damus-Kaye-Stansel procedure described by Yasui *et al.*

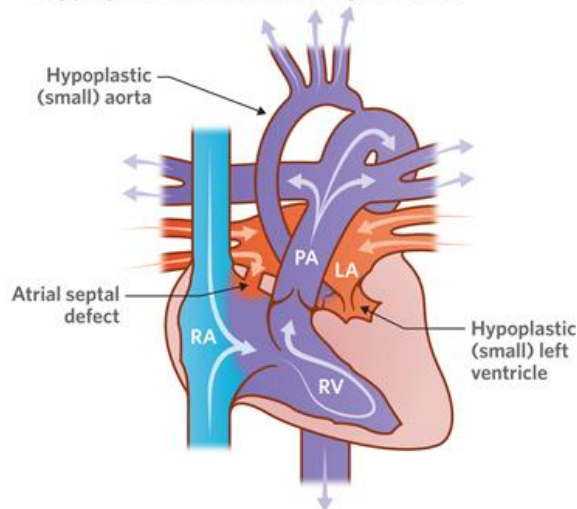


Norwood operation

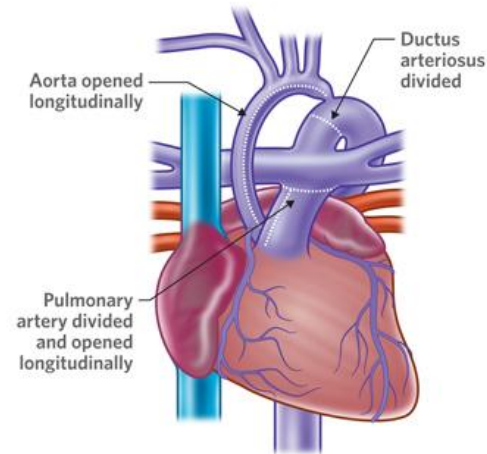


Normal heart and circulation

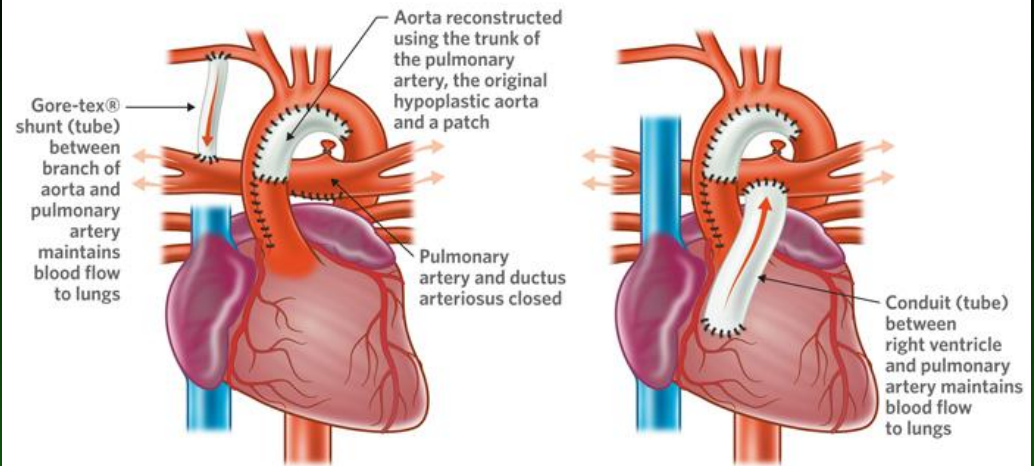
Hypoplastic left heart syndrome



Norwood operation



Pre-operative anatomy



Norwood operation

Norwood operation (Sano)

Potential complication of surgery

Recurrent or persistent LVOT obstruction

- 1) Posterior malalignment of infundibular septum
- 2) Hypertrophy of muscle of Moutaert
- 3) Bicuspid dysplastic aortic valve
- 4) Narrowness of aortic anulus
- 5) Hypoplasia of ascending aorta and aortic arch

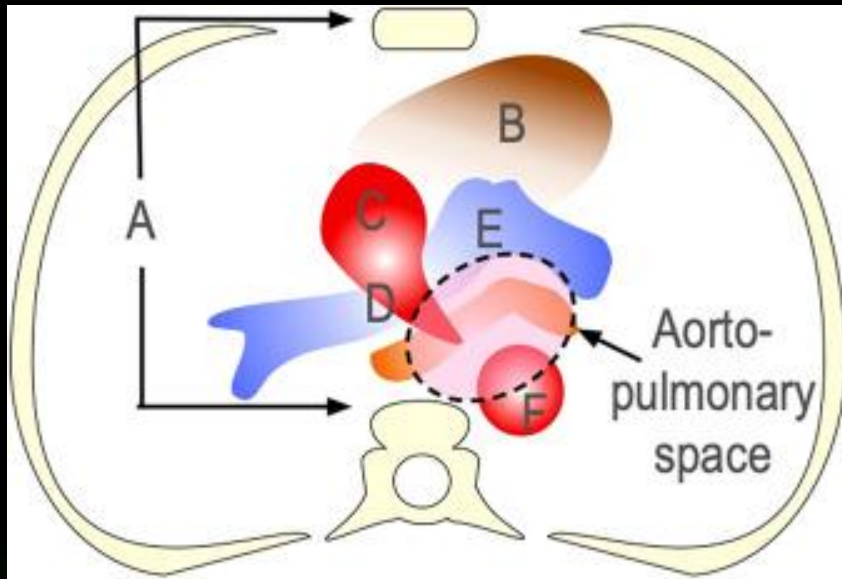
High reoperation rate due to these factors

Potential complication of surgery

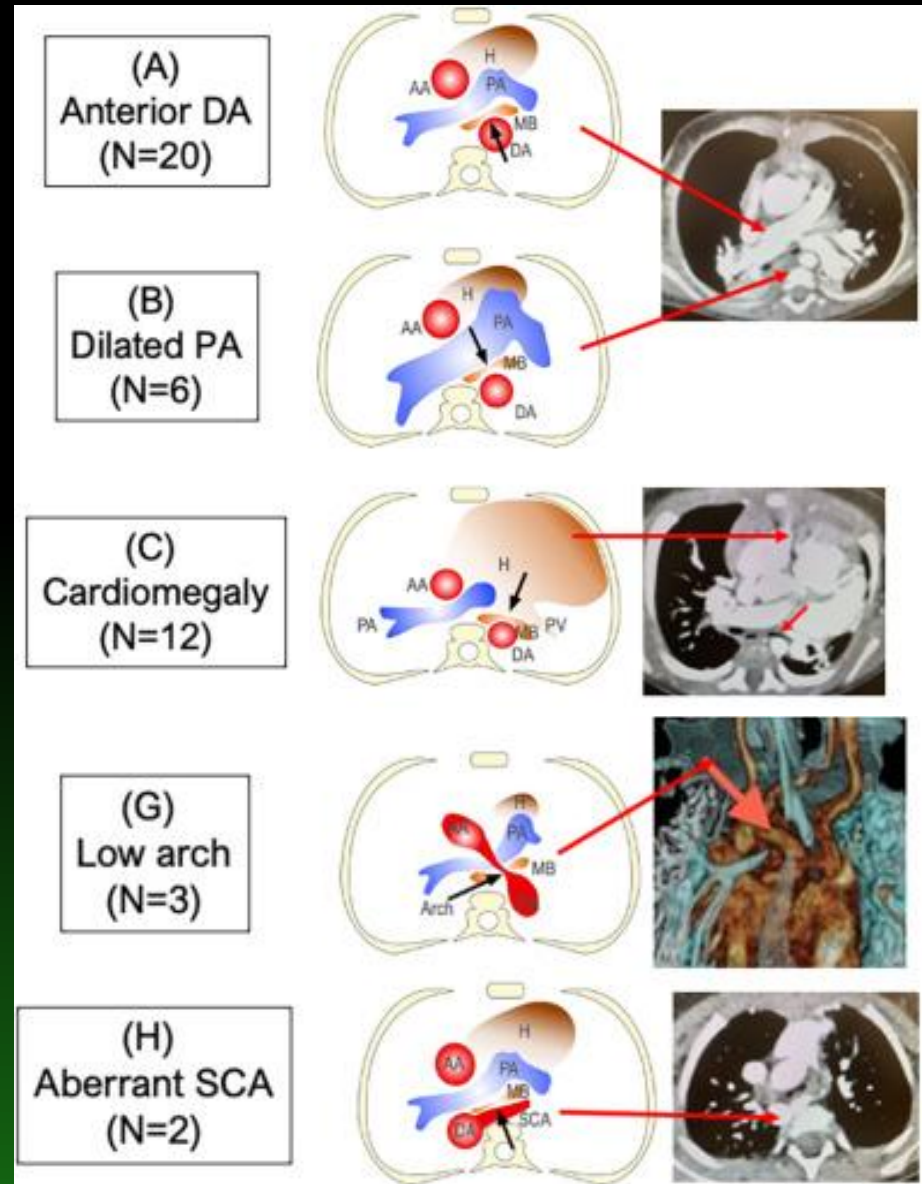
Bronchial compression

- Anterior & Downward Displacement of the Aortic Arch
: Surgical tension or arch reconstruction pulls the aorta downward.
- Sandwich Effect on Left Main Bronchus (LMB)
: LMB is compressed between the overlying reconstructed aortic arch and the underlying pulmonary artery.
- Tracheobronchomalacia
: Continuous pulsatile pressure on weak neonatal airway cartilage leads to localized airway collapse.
- Clinical Result
: Significant airway obstruction, stridor, and recurrent atelectasis.

Bronchial compression

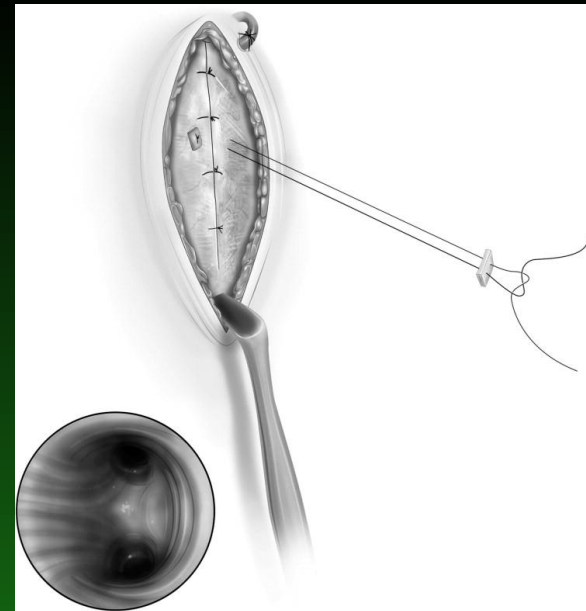
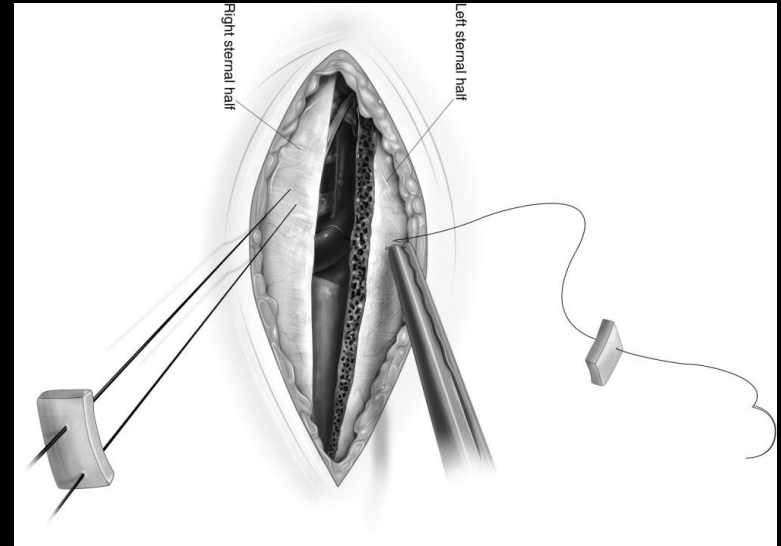
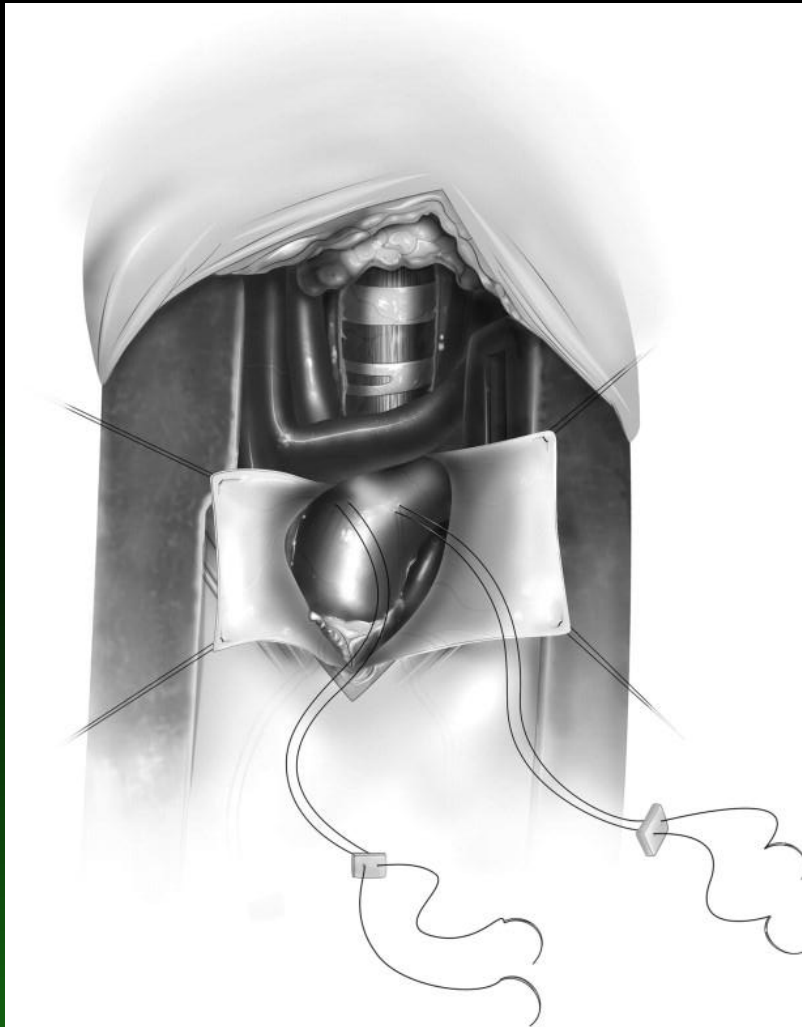


Sternum-vertebra distance (A),
heart (B), ascending aorta (C),
aortic arch (D),
pulmonary artery (E),
descending aorta (F)



Bronchial compression

Aortopexy



Thank you for your attention !



"I hope you become a warm-hearted cardiothoracic surgeon who truly cares for patients."