Aortic Arch Obstruction
- Morphology, Surgical Repair, and Postoperative Management -

COARCTATION of the AORTA

Definition

- A congenital narrowing of the upper descending aorta adjacent to the site of attachment of the ductus arteriosus
- Aortic lumen may be atretic, but aortic walls above and below the atresia are in continuity
- Aortic arch interruption; short distance separation between the aortic ends
- Uncommonly, coartaction occurs more proximally, between LCCA and LSCA
- Without major associates cardiac anomalies; Primary, Pure, or Isolated coartation

Historical Note

- Morgagni is credited in 1760 with the first description of an aortic coarctation found at autopsy
- The first coarctation repair in a patient was performed by Crafoord and Nylin in October 1944
- Repair of coarctation in neonates became more successful after documentation of prostaglandin E_1 (PGE_1) in these sick small babies, achieved by maintaining patency of the ductus arteriosus until time of repair

Historical Note

Interrupted aortic arch and ventricular septal defect

Direct repair through a median sternotomy incision in a 13-day-old infant

A type B interrupted aortic arch was successfully repaired in a 13-day-old infant during profound hypothermia and circulatory arrest. Through a median sternotomy incision it was possible to resect a patent ductus arteriosus and mobilize the descending thoracic aorta for anastomosis to the side of the ascending aorta. At the same time a large ventricular septal defect (VSD) and a small atrial septal defect were closed through the right atrium. Cardiac catheterization 5 months after operation showed a small persistent VSD with a pulmonary to systemic blood flow ratio of 1.1/1. The systolic gradient between the ascending and descending aorta was 20 mm. Hg.

G. A. Trusler, M.D., and T. Izukawa, M.D., *Toronto, Ontario, Canada*

The Hospital for Sick Children, Toronto, on June, 1973

J Thorac Cardiovasc Surg 1975;69:126

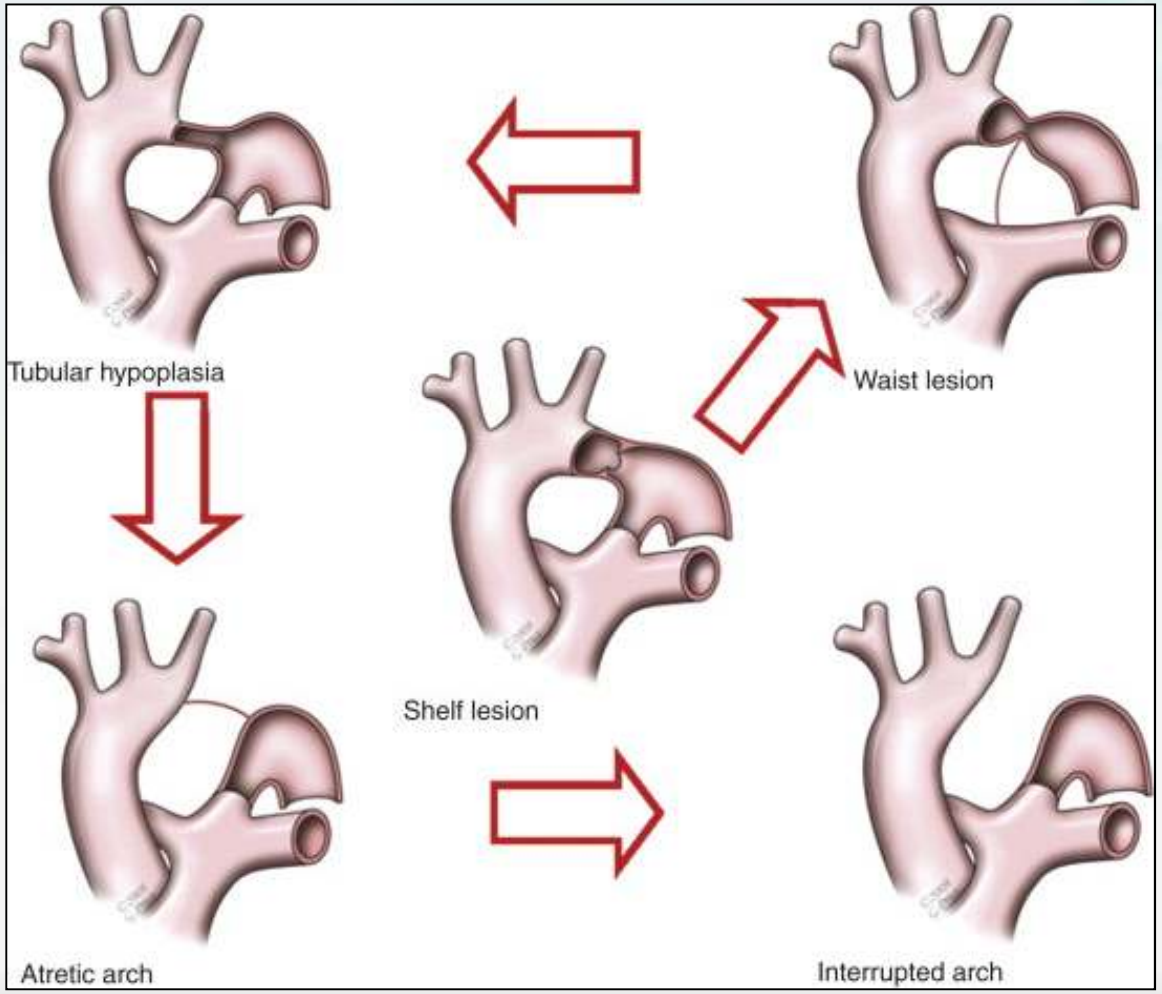
Morphology and Morphogenesis

Coarctation

- Vary in severity
- In localized lesion, reduction of cross-sectional area by more than about 50% → hemodynamically important pressure gradient
- *Pseudocoarctation ;* occasionally the adult aorta may be redundant and severely kinked opposite the ligamentum arteriosum, without any pressure gradient

Morphology and Morphogenesis

Morphological spectrum of aortic arch obstruction



Morphology and Morphogenesis

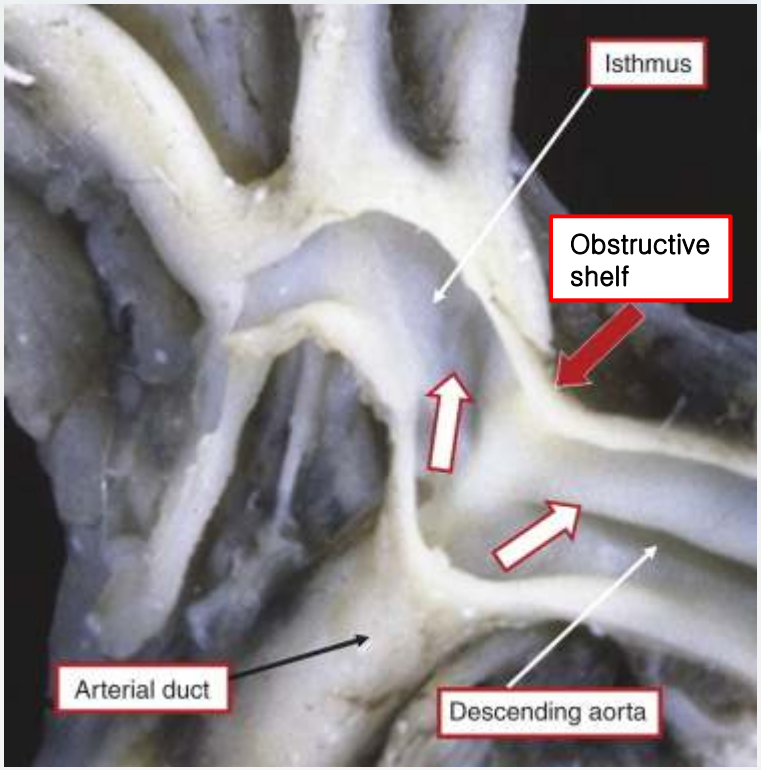
Coarctation

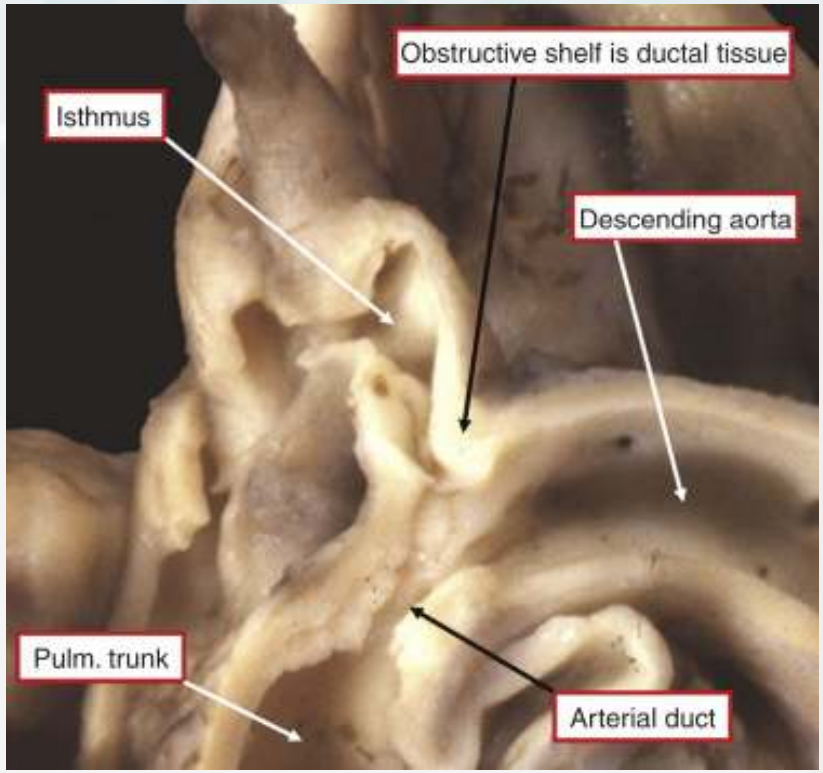
- Shelf, projection, or infolding of the aortic media into the lumen (most prominent in the opposite of the ductus arteriosus)
- External narrowing may be absent in the young infant
- Poststenotic dilatation
- The shelf may be preductal or postductal but is usually periductal
- Localized ridge of intimal hypertrophy that further narrows the lumen

Morphology and Morphogenesis

Coarctation

- Shelf, projection, or infolding of the aortic media





Morphology and Morphogenesis

Coarctation

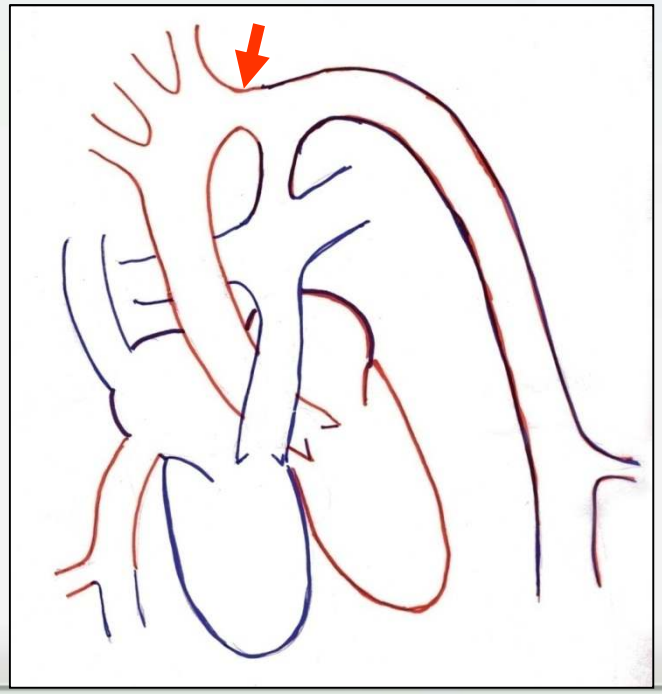
- **Ductal tissue forms a sling that completely surrounds the periductal aorta, which may progressively proliferate after birth and cause restenoses after repair of coarctation in neonates and young infants**

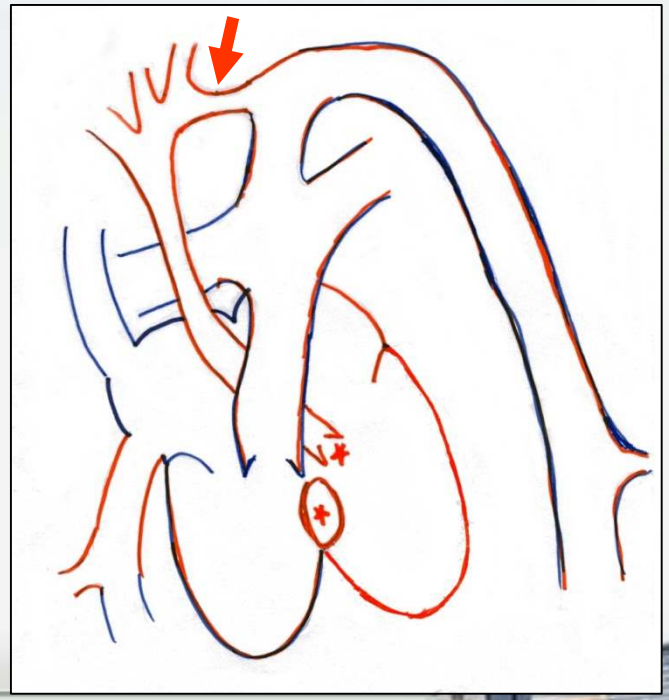
- **Prevalence and type of coarctation are related to fetal flow patterns through the ductus and aorta**

Morphology and Morphogenesis

Distal Aortic Arch Narrowing

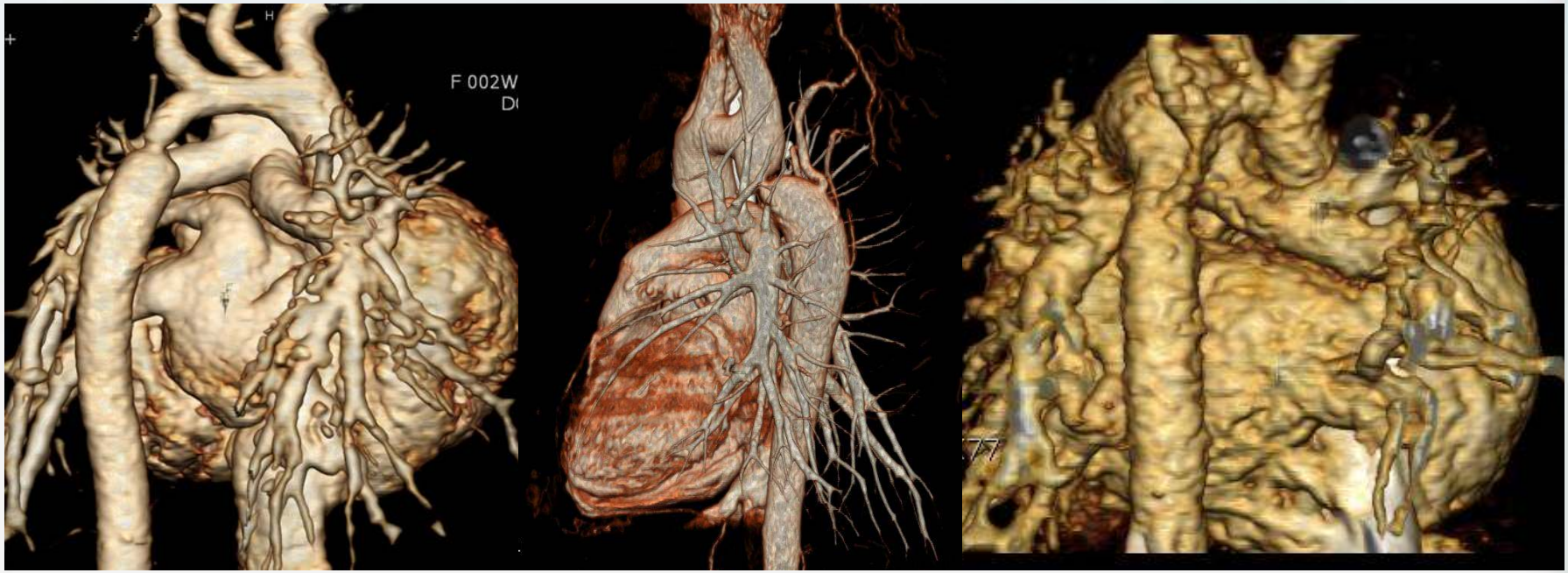
- This narrowing appears in some cases to be a transient finding related to prenatal flow pattern
- There is evidence that unrepaired arch hypoplasia, at least in some cases, does not grow adequately, requiring repeat surgery





Morphology and Morphogenesis

Coarctation



Morphology and Morphogenesis

Proximal Aortic and Arterial Walls

- **The wall of the entire aorta proximal to the coarctation is abnormal**
- **The abnormalities extend out to all major arteries supplied by the aorta proximal to the coarctation.**
- **These abnormalities may be primary ones that have developed in utero**
  - ✓ Impaired flow-mediated vasodilatation and increased intima media thickness were apparent in a study group with a mean age of 12 years

Morphology and Morphogenesis

Collateral Circulation

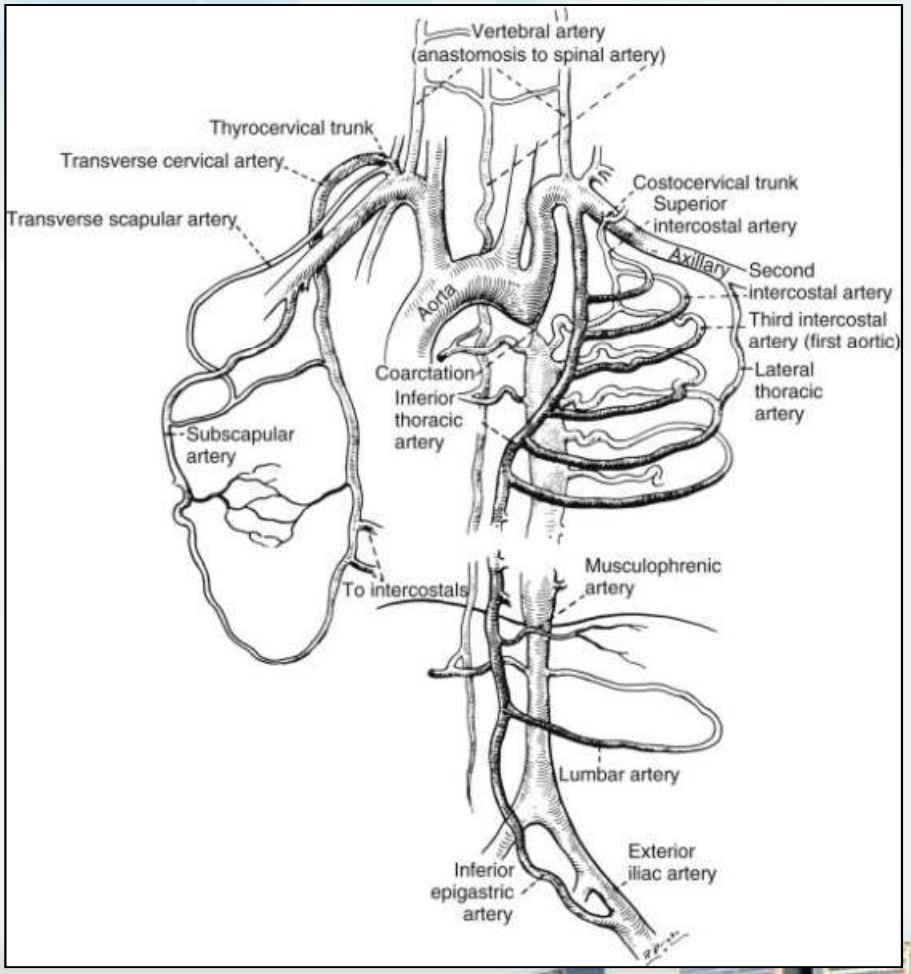
- Between aorta proximal to the coarctation and that distal to it
- Classic signs, such as parascapular pulsations and rib notching
- It is usually present to some extent in newborns but increases in size and extensiveness as the patient ages

Morphology and Morphogenesis

Collateral Circulation

From; branches of both subclavian arteries, particularly internal thoracic, vertebral, costocervical, and thyrocervical trunks

To; first two pairs of intercostal arteries distal to the coarctation (third and fourth intercostal arteries)



Morphology and Morphogenesis

Aneurysm Formation

- **Enlarged, tortuous third and fourth intercostal arteries may become aneurysmal**

- **The aorta itself may become aneurysmal adjacent to the site of maximal narrowing as a result of hemodynamic effects, aortic dissection, or mycotic aneurysm**

  about 10% by the end of the second decade of life,
  20% by the end of the third decade,
  probably even higher in older patients

Morphology and Morphogenesis

Atria

- Foramen ovale to be prolapsed, causing left-to-right shunting
- In about 10% of patients with ASD, however, intractable heart failure will develop in infancy following coarctation repair, requiring ASD closure. The best predictor of development of heart failure when ASD coexists with coarctation is small mitral valve diameter, not the size of the ASD itself

Morphology and Morphogenesis

Left Ventricle

- Left ventricular hypertrophy
- Left ventricular outflow tract may be abnormal
- Reduced interpapillary distance

Aortic Valve

- Bicuspid aortic valve is common
- When aortic regurgitation appears in coarctation, it is usually based on a bicuspid aortic valve combined with persistent hypertension

Morphology and Morphogenesis

Intracranial Aneurysm

- Coarctation and berry-type intracranial aneurysm coexist in some patients

Coarctation as Part of Hypoplastic Left Heart Physiology

- Hypoplasia of ascending aorta
- Supravalvar, valvar, subvalvar, and annular aortic stenosis or hypoplasia
- Aortic atresia
- Left ventricular hypoplasia or hypertrophy
- Endocardial fibroelastosis
- Mitral stenosis with or without a single papillary muscle (parachute mitral valve)
- Supravalvar mitral ring

Morphology and Morphogenesis

Coexisting Cardiac Anomalies

- **Coexisting Cardiac Anomalies in Severely Symptomatic Neonates with Coarctation**

Coexisting Cardiac Anomaly	*n*	% of 432
None	**171**	**40**
VSD (isolated)	**155**	**36**
Single ventricle	32	7
TGA	27	6
AV septal defect	16	4
DORV	9	2
Taussig-Bing heart	12	3
CCTGA	6	1
Truncus arteriosus	1	0.2
Anomalous origin of LCA from PT	1	0.2
TAPVC (with VSD)	1	0.2
PAPVC	1	0.2

J Thorac Cardiovasc Surg 1994; 108:841

Morphology and Morphogenesis

PDA

- PDA is present in almost 100% of neonates and in most infants with a preductal type of coarctation.
- PDA and tubular arch hypoplasia are considered part of isolated coarctation rather than an additional anomaly
- Tubular hypoplasia of the distal aortic arch is considered to be part of the anomaly of coarctation rather than an associated anomaly

Clinical Features and Diagnostic Criteria

Depend on; coexisting cardiac anomalies
patient's age at presentation

Neonates and Infants

- ✓ Severe heart failure
- ✓ Femoral pulses are absent or reduced in volume and delayed compared with radial or brachial pulses
- ✓ Blood pressure is higher in the arms than in the legs (by >20 mmHg)
- ✓ Delay in onset of heart failure is probably related to time to ductal closure
- ✓ Right-to-left shunt through PDA → differential cyanosis (uncommon)

Clinical Features and Diagnostic Criteria

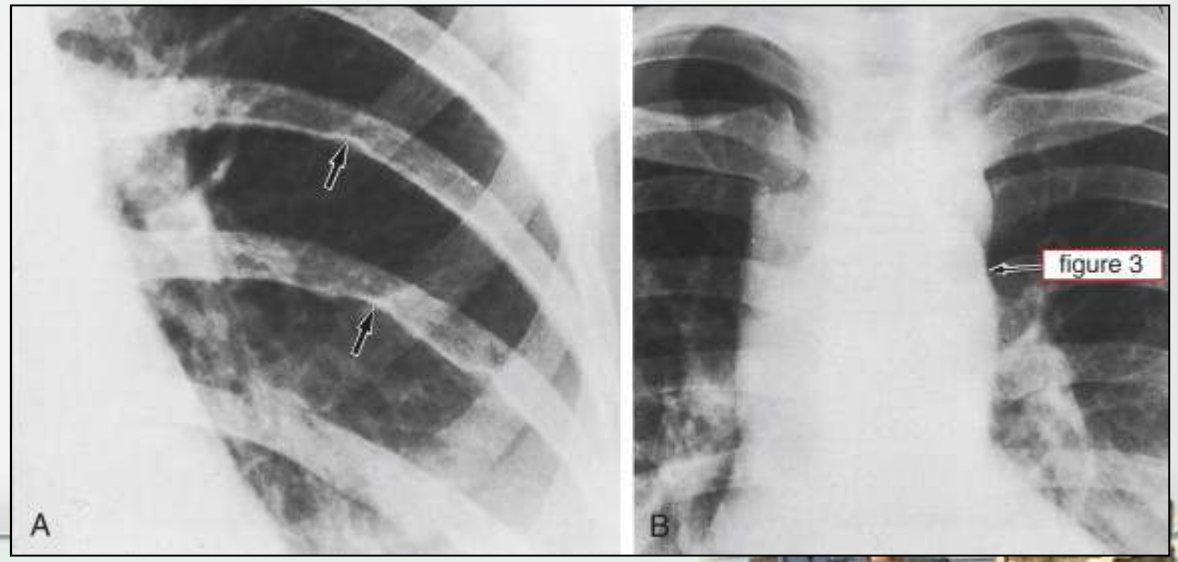
Childhood (Age 1 to 14 Years)

- ✓ Asymptomatic unless they have important associated anomalies
- ✓ Heart failure in 7 of 65 children (11%) age 1 to 14 years (Patel et al)
- ✓ Hypertension occurs in almost 90% of patients

Clinical Features and Diagnostic Criteria

Adolescence (Beyond 14 Years) and Adult Life

- ✓ Hypertension is common and more severe than in younger patients, and heart failure may occur after about age 30 years
- ✓ Effort dyspnea, cardiomegaly, and important left ventricular hypertrophy on ECG → Heart failure
- ✓ Headache, nose bleeds, fatigue, and calf claudication occasionally occur
- ✓ Almost always, rib notching



Clinical Features and Diagnostic Criteria

Associated Syndromes

Turner syndrome
von Recklinghausen disease
Rarely, Noonan syndrome or congenital rubella

Clinical Features and Diagnostic Criteria

Special Diagnostic Methods

Two-dimensional echocardiography

In moderate or mild coarctation, presence of an open ductus may obscure a coarctation at echocardiographic examination

MRI and CT

Imaging modalities of choice for COA in patients beyond infancy

Hemodynamic data can be assessed by MRI (measure of collateral flow

Clinical Features and Diagnostic Criteria

Special Diagnostic Methods

Cardiac catheterization and aortography

- Now play a secondary role
- Used mainly when hemodynamic data are important
- Severity of the coarctation can be better assessed on aortography than by catheter withdrawal pressures

Natural History

- ✓ About 6.5% of CHD
- ✓ About 50 of 100,000 live births
- ✓ About 40 of these can have isolated COA
- ✓ M/F > 2 in isolated COA
- ✓ No gender difference in those with important coexisting cardiac anomalies

Natural History

Isolated Coarctation

- ✓ Before the era of surgical correction (beyond ; 2 years of age), median age of death is 31 years
- ✓ 10% of isolated COA may die of acute cardiac failure during 1st month of life if untreated
- ✓ 20% may die later during the first year of life of heart failure or its sequelae

→ true median age of death may be closer to 10 years

Natural History

Isolated Coarctation

1) Heart Failure in Infancy

is related to

- **Ductal closure**
- **Degree of collateral development**
- **Presence of major noncardiac anomalies**

Left ventricular stroke volume and ejection fraction severely depressed due to afterload mismatch → L to R shunt through PFO → RV enlargement

Cause of death of isolated COA

- ✓ **heart failure**
- ✓ **infective endocarditis**
- ✓ **aortic rupture or dissection (each in about 20% of cases)**
- ✓ **rupture of an intracranial aneurysm in about 10%**

Natural History

Isolated Coarctation

2) Heart Failure in Childhood and Adult Life

- ✓ Most such deaths occurred in the fourth and fifth decades
- ✓ Associated valvar heart disease, usually aortic
- ✓ Congenitally abnormal aortic valve (bicuspid valves were present in 42% of the hearts) was the usual cause of stenosis or regurgitation

3) Infective Endocarditis or Endarteritis

4) Aortic Rupture (dissection)

5) Intracranial Lesions

Natural History

Coarctation Associated with VSD

- ✓ Most of these babies die within a few months without surgical treatment
- ✓ In many the VSD rapidly becomes small, and the natural history then becomes essentially that of isolated

Coarctation Associated with Other Major Cardiac Anomalies

- ✓ Early heart failure

Technique of Operation

- ✓ In general, resection of the coarctation and reconstruction of the aorta should be considered the ideal method of repairing coarctation. For a number of reasons, however, this cannot always be achieved, and alternative methods must be used.

Technique of Operation

Isolated Coarctation

Preparation, Incision, and Dissection

Neonates and Infants

Nasopharyngeal temperature of about 35°C
Left posterolateral thoracotomy, with the entry through the fourth intercostal space

Technique of Operation

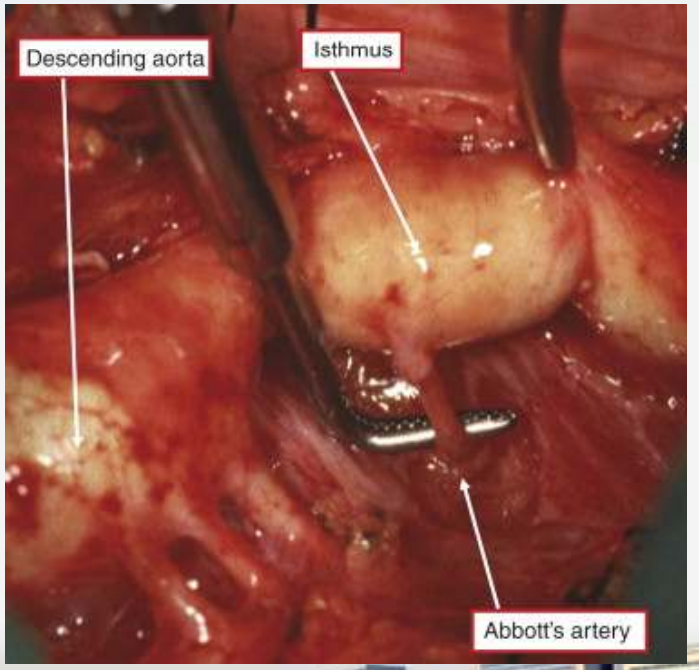
Isolated Coarctation

Preparation, Incision, and Dissection

Children

The operation is technically more demanding in children than in neonates and infants because collateral circulation is much larger

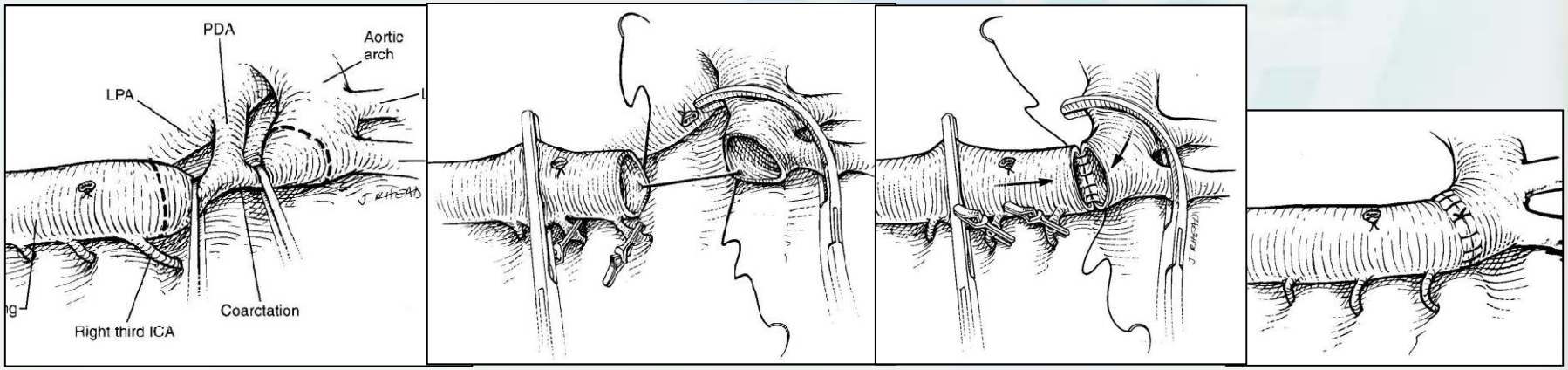
"The Abbott artery" occasionally arises from the medial aspect of the isthmus and, when present, should be ligated and divided



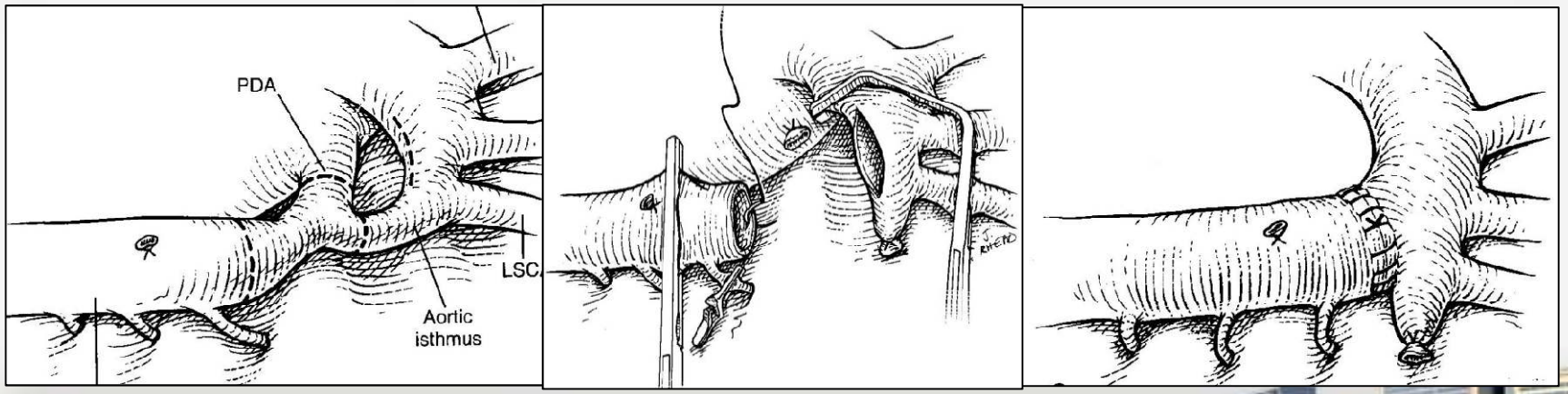
Technique of Operation

Isolated Coarctation

Resection and End-to-End Anastomosis



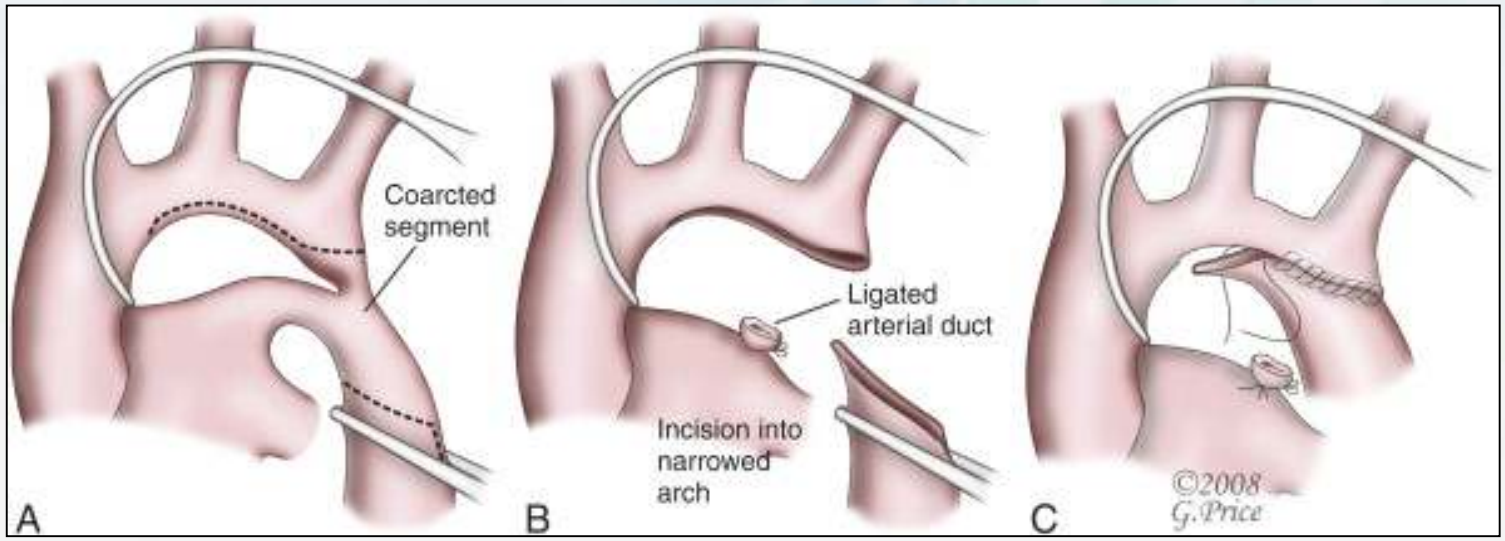
Resection and End-to-Side Anastomosis



Technique of Operation

Isolated Coarctation

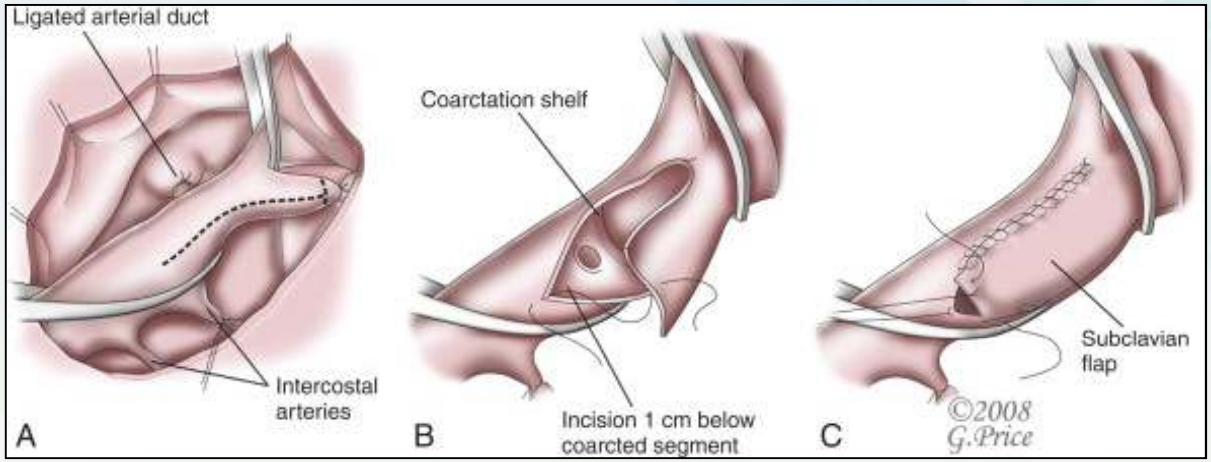
Extended End-to-End Repair



Technique of Operation

Isolated Coarctation

Subclavian Flap Repair



✓ Vertebral artery is ligated to prevent any subsequent subclavian steal leading to cerebral ischemia

Benefits

Use of exclusively native material (decreased risk of infection)
Improved potential for growth (no circumferential anastomosis)
Less extensive dissection
Less tension on the suture

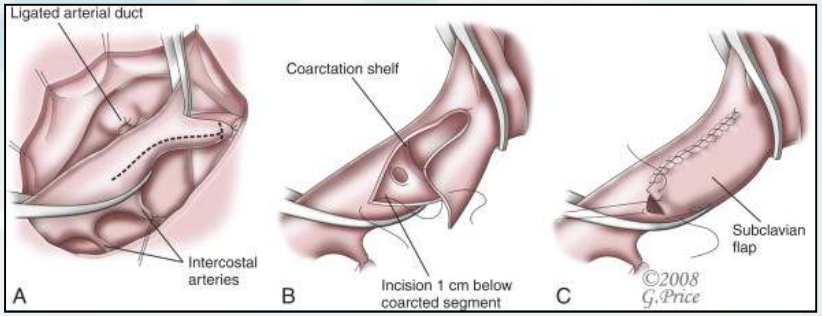
Disadvantage

Loss of the main arterial supply to the left arm

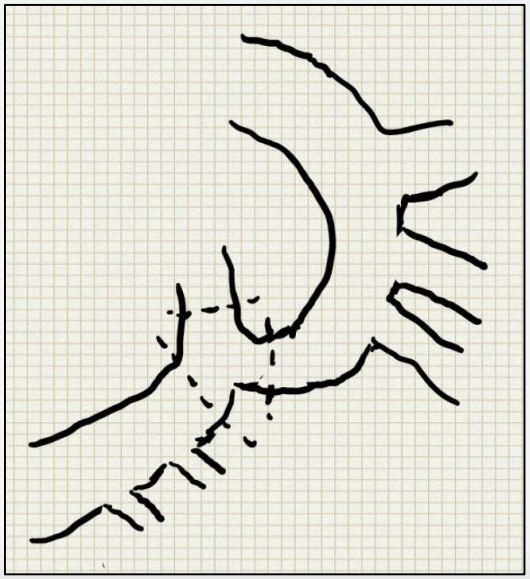
Technique of Operation

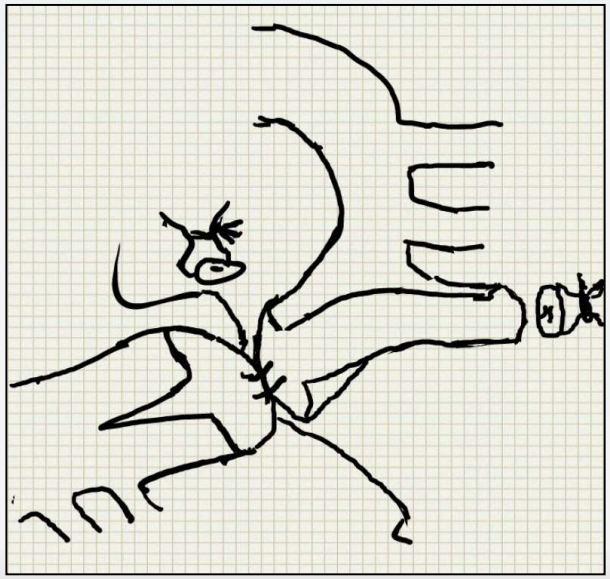
Isolated Coarctation

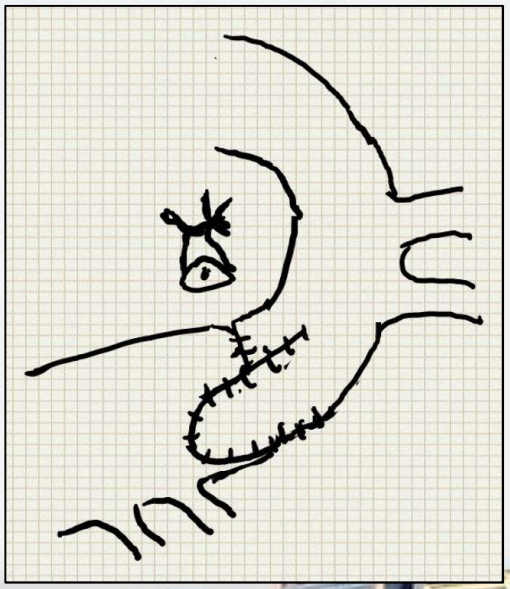
Resection and Subclavian Flap Repair



Complete excision of ductal tissue



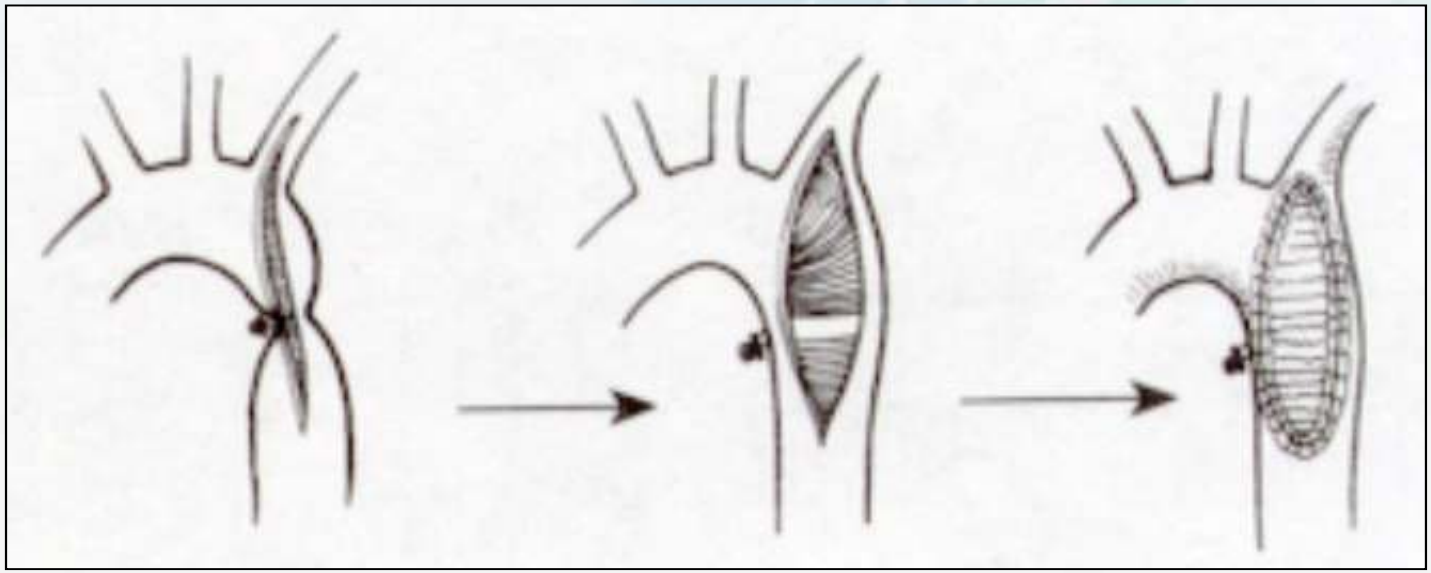




Technique of Operation

Isolated Coarctation

Patch Aortoplasty



Aneurysmal formation as complication

Technique of Operation

Isolated Coarctation

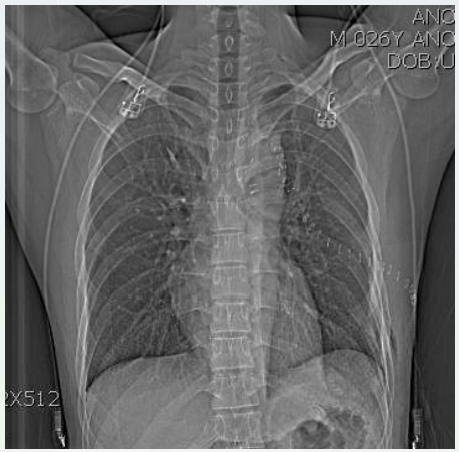
Children and Adults

- ✓ Vessels are much more friable
- ✓ Intercostal arteries larger and more easily damaged
  → dissection potentially more hazardous
- ✓ Use of controlled hypotension by the anesthesiologist is important
- ✓ Once the aortic clamps are in place, upper body blood pressure is allowed to increase to moderately hypertensive levels (to promote collateral blood flow)
- ✓ The junction of the enlarged intercostal artery with the aorta is the most fragile and easily damaged point
- ✓ Occasionally, because of immobility of the aortic structures in an older patient or because of a long-segment coarctation, end-to-end anastomosis is not possible, and either an interposed polyester tube graft or an augmentation patch is necessary

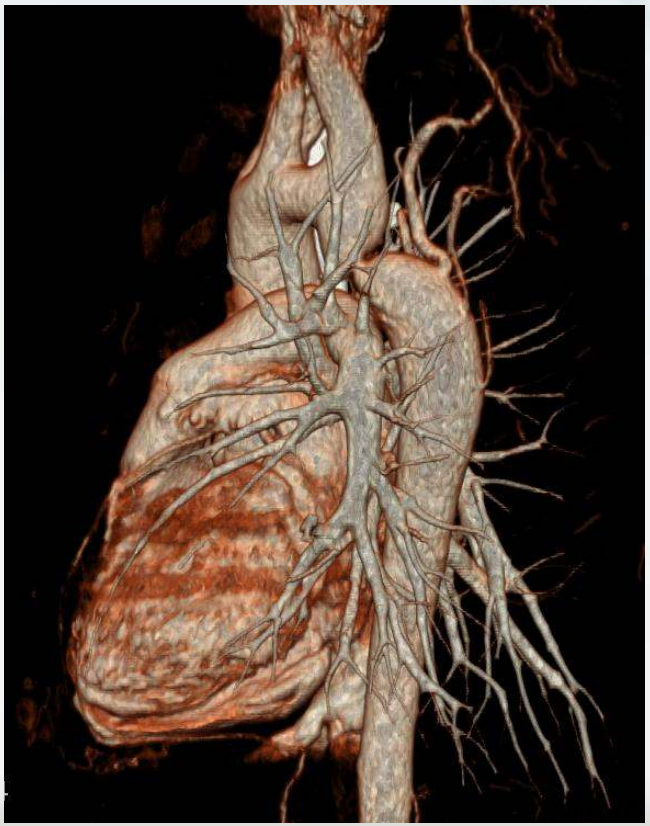
Technique of Operation

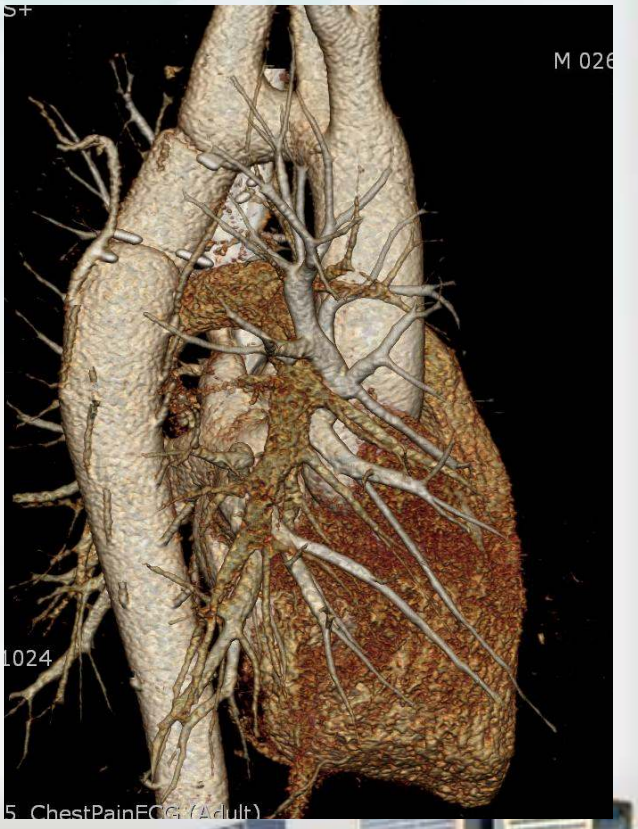
Isolated Coarctation

31y / M, Upper body hypertension



Resection and graft interposition without bypass





Technique of Operation

Immediate Post-Repair Management

- ✓ Distal clamp is removed first
- ✓ Maintain proper ventilation and baseline systemic blood pressure for at least the next 5 minutes as a precaution against sudden development of intractable ventricular fibrillation 3 to 4 minutes after release of the clamp (de-clamping syndrome)
- ✓ Give sodium bicarbonate or an infusion of a pure peripheral vasoconstrictor (or both) just before clamp removal in particularly unstable infants or in those with prolonged clamp times
- ✓ After repair, if there is a systolic gradient of greater than 10 mmHg, clamps are reapplied, sutures removed, and the repair refashioned. In neonates, the residual gradient may reside in the hypoplastic distal aortic arch between left carotid and subclavian arteries.

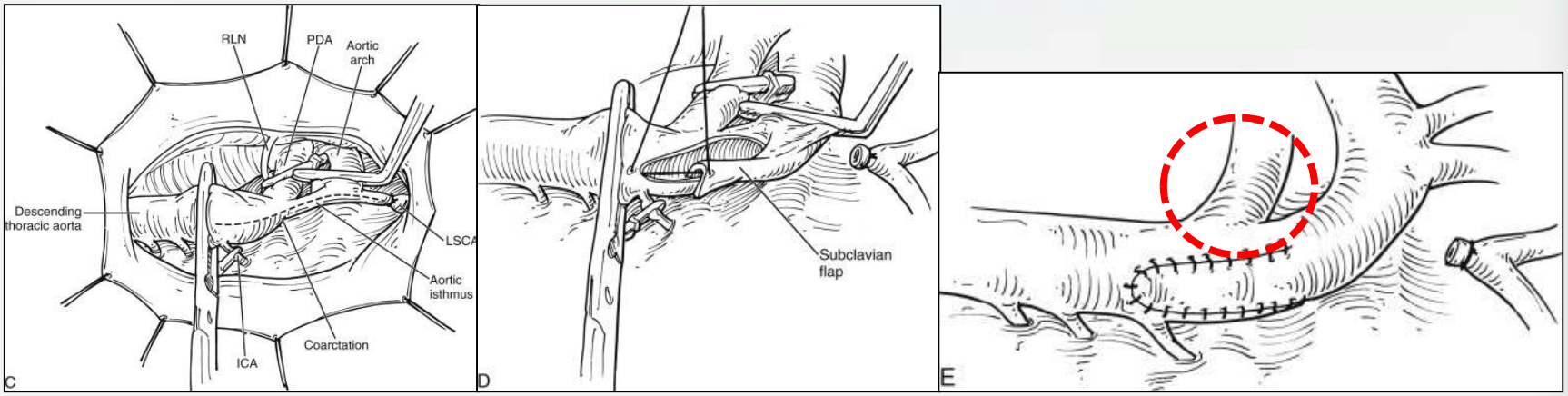
Technique of Operation

Isolated Coarctation

Special situation

Subclavian Flap Aortoplasty

- ✓ When it is advantageous to preserve the ductus in the setting of a borderline left ventricle (PGE1 infusion is maintained throughout the procedure)



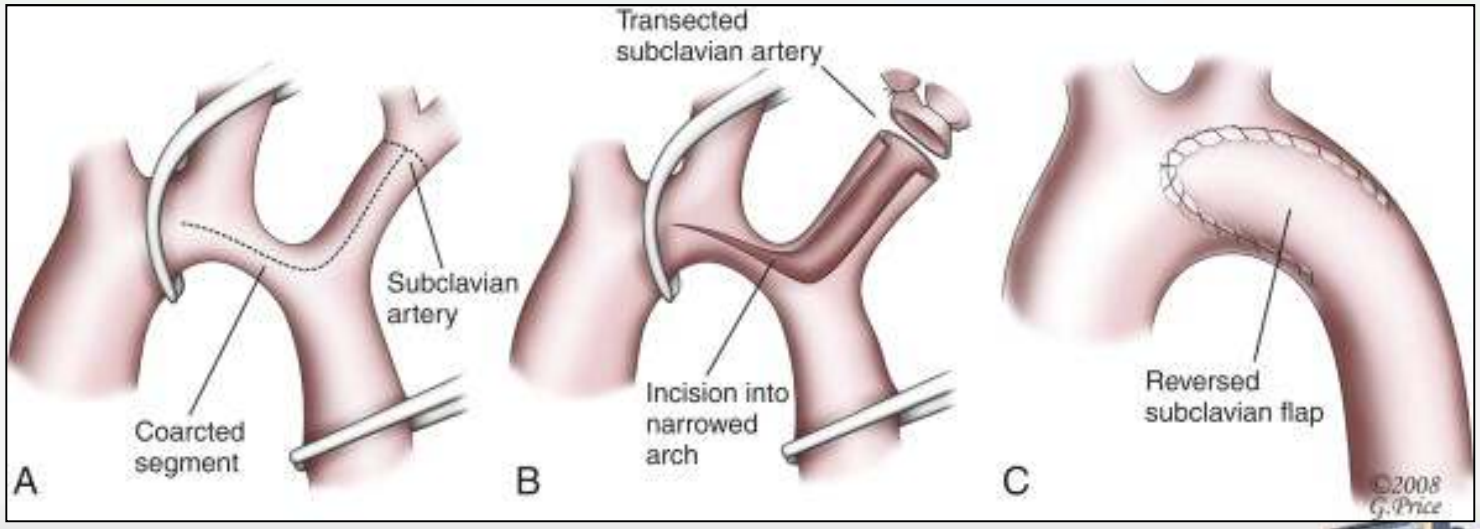
Technique of Operation

Isolated Coarctation

Special situation

Repair of Coarctation Proximal to Left SCA

Reversed subclavian flap aortoplasty
Resection with end-to-side anastomosis
Reverse subclavian flap and end-to-end anastomosis



Technique of Operation

Isolated Coarctation

Special situation

Repair When Aneurysm Is Present

Repair of Persistent or Recurrent Coarctation with Aneurysm

- ✓ Aneurysm following coarctation repair is more likely when transverse arch hypoplasia is present

- ✓ Surgical management can vary but typically requires CPB, either via median sternotomy or left thoracotomy, with resection of the aneurysm and obstructive segment and interposition graft insertion. These procedures carry a mortality risk of 14% to 23%, and therefore endovascular management should be considered when anatomic details are favorable

Technique of Operation

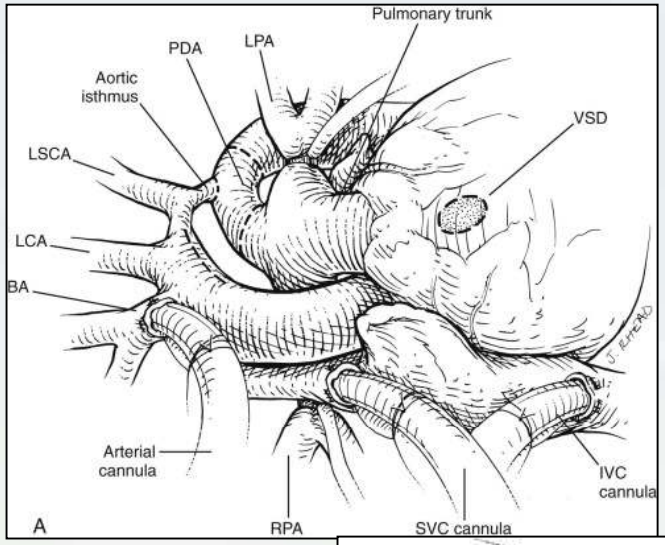
Repair from an Anterior Midline Approach

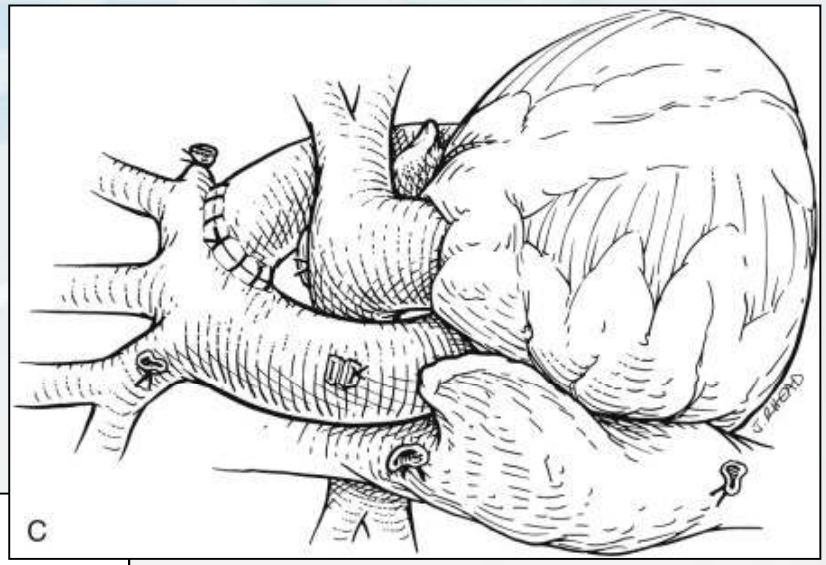
- ✓ Particularly in neonates and young infants, coarctation of the aorta can be well repaired from an anterior midline approach using CPB.
- ✓ Although use of hypothermic circulatory arrest is advocated by some, continuous CPB with **antegrade cerebral perfusion** can be used routinely for this repair

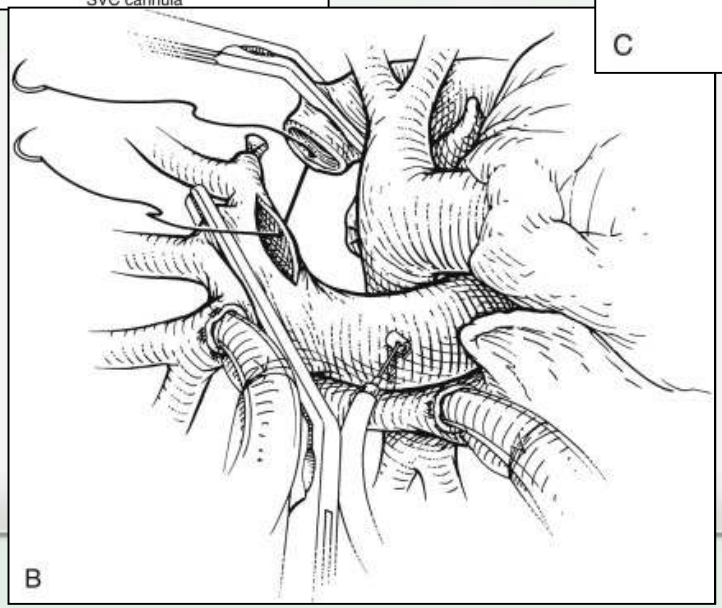
- ✓ **The midline approach is particularly useful for**
  1) concomitant repair of intracardiac defect
  2) severe proximal transverse arch hypoplasia
  3) “bovine” trunk with distal arch hypoplasia

Technique of Operation

Repair from an Anterior Midline Approach







Brain Protection During Arch Repair

- Hypothermic circulatory arrest

  18 ℃

- Regional cerebral perfusion

  Through 3.0 or 3.5mm Gore-Tex tube graft connected to innominate artery.

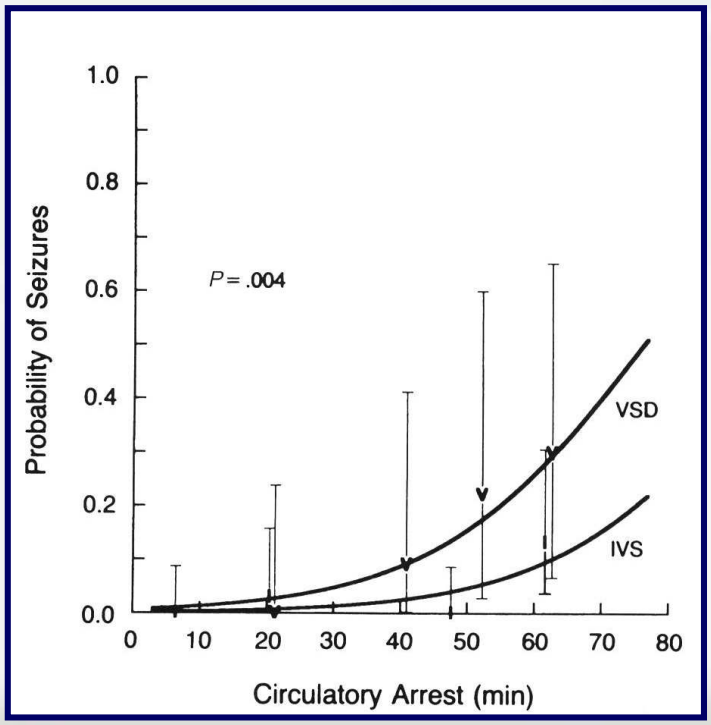
  Flow rate; 50ml/kg, Right radial artery pressure > 30mmHg

  Perfusate temperature; 24℃ (at rectal temperature 25℃)

Brain Protection During Arch Repair

"Boston circulatory arrest trial"

4-year enrollment period up to March 1992
171 TGA < 3 months of age, IVS/VSD
DHCA (mean 55minutes) versus low-flow bypass (50cc/kg/min)



- Perioperative findings in DHCA group

- Clinical seizure 12%, seizure by EEG 26%
- No seizure in the patient < 35minututes
- Strong correlation between duration of DHCA and occurrence of seizure
- Greater release of creatine kinase BB
- Longer recovery to first EEG activity

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Brain Protection During Arch Repair

❑ Findings at 1 year of age

- **Lower fine and gross motor ability in DHCA group**
- **Correlation between longer duration of DHCA and lower score**
- **Neurological abnormality tended to be more common in DHCA group**
- **Clinical seizure predicted increased neurological abnormalities at 1 year of age**

❑ Findings at 4 year of age

- **Neurodevelopmental performance as a whole was below normal**
- **DHCA group had significantly worse motor function and speech**
- **Perioperative seizure predicted significantly increased risk of both lower IQ scores and neurological abnormality**

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Brain Protection During Arch Repair

HOW TO DO IT

Selective Cerebral Perfusion Technique During Aortic Arch Repair in Neonates

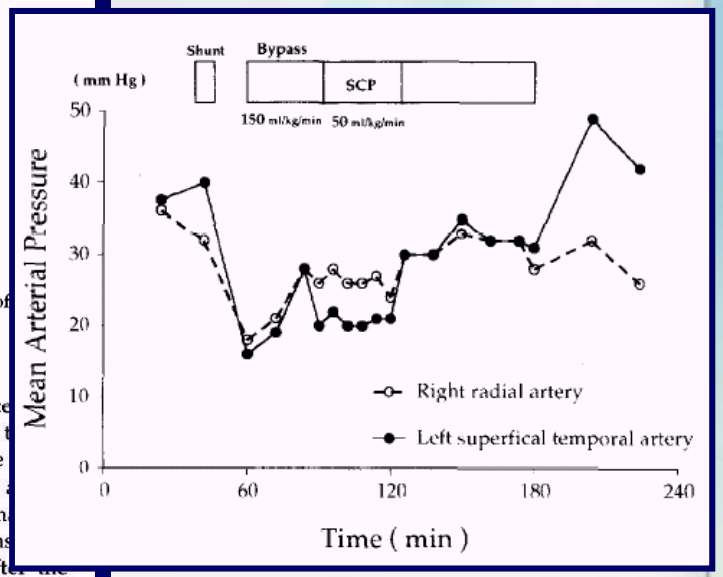
Toshihide Asou, MD, Hideaki Kado, MD, Yutaka Imoto, MD, Yuichi Shiokawa, MD, Ryuji Tominaga, MD, Yoshito Kawachi, MD, and Hisataka Yasui, MD

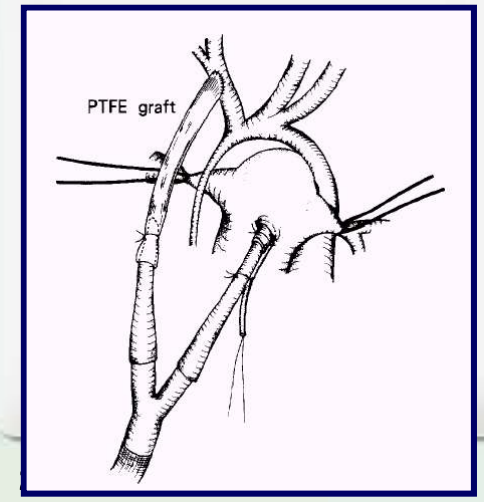
Department of Cardiovascular Surgery, Fukuoka Children's Hospital, and Division of Cardiac Surgery, Research Institute of Angiocardiology, Kyushu University, Fukuoka, Japan

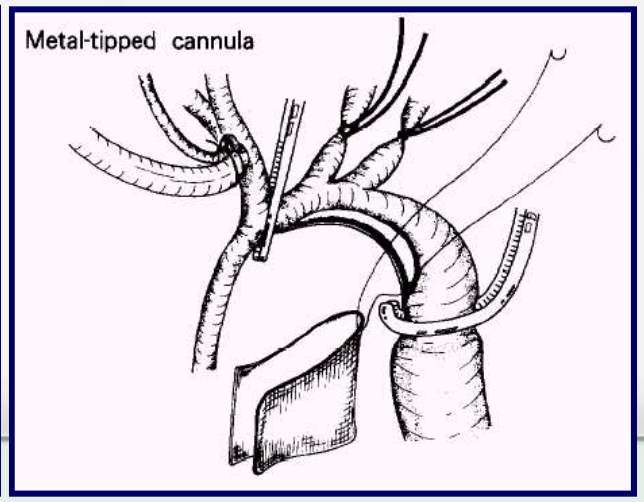
We describe selective cerebral perfusion techniques for repair of the aortic arch in neonates. These techniques may help protect the brain from ischemic injury caused by a cessation of cerebral perfusion for aortic arch reconstruction in patients with hypoplastic left heart syndrome or interrupted aortic arch.

(Ann Thorac Surg 1996;61:1546–8)

with both the innominate artery and the desce aorta unclamped. The perfusion rate returns t mL · kg^{-1} · min^{-1} to perfuse the upper and the part of the body via the graft of the innominate a The pericardial roll is anastomosed to the proxim of the main pulmonary artery conjoined with the as ing aorta to create a systemic outflow tract. Afte







Rectal temperature ; 22℃

Flow rate ; 50mL/kg/min

Right radial a. pr; 28mmHg

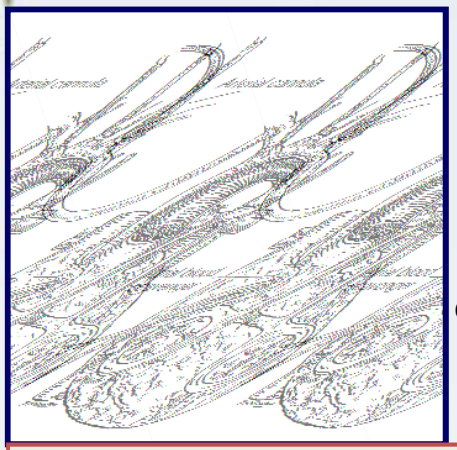
Lt sup temporal a. pr; 20mmHg

Neonate brain ;

1/7 –1/10 of body weight

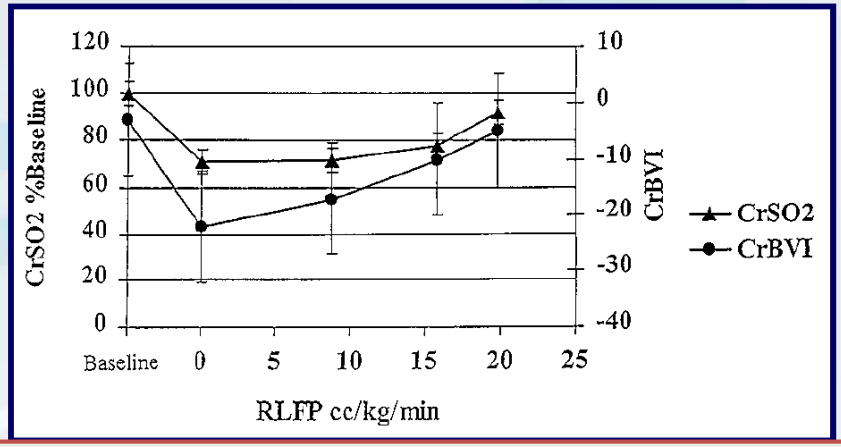
Adult ; 1/15 of body weight

Brain Protection During Arch Repair

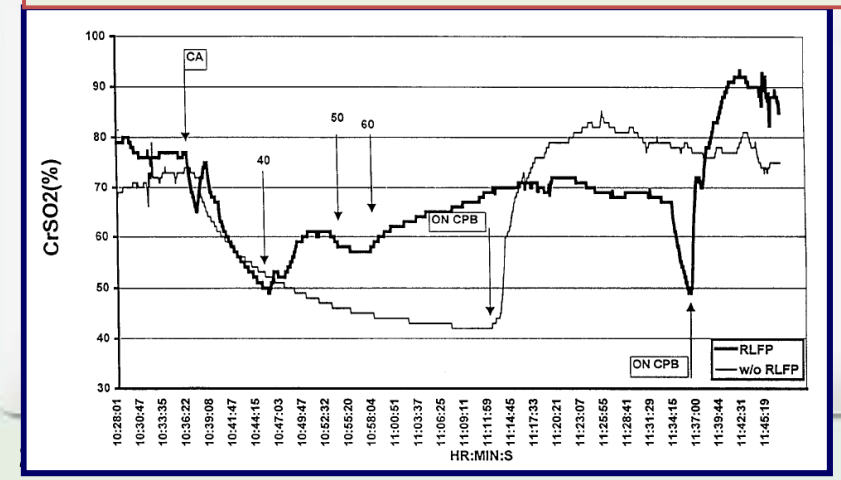


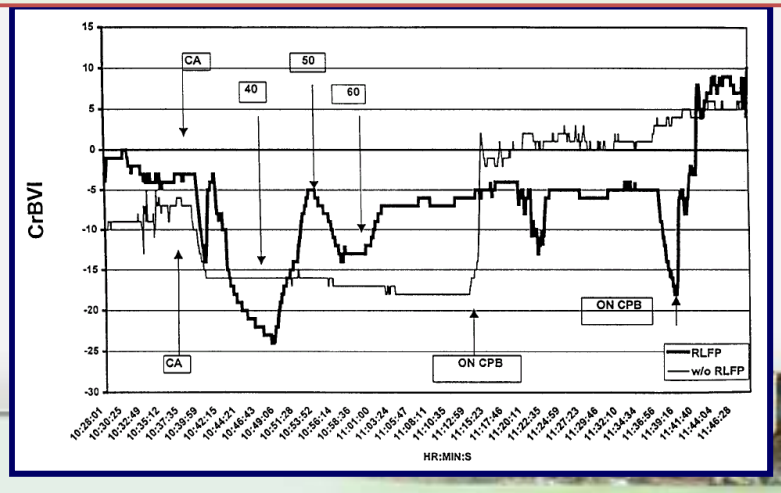
Frank A. Pigula
Children's hospital
of Pittsburgh

J Thorac Cardiovasc Surg
2000;119:331
Seminars in Thoracic and
Cardiovascular Surgery 2002



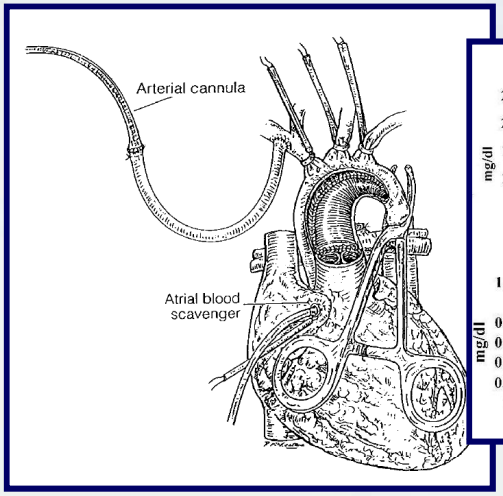
- RLFP is used when expected circulatory arrest time > 20min
- Rectal temperature; 18 ℃
- Near infrared spectoscopy is used to regulate regional blood flow
- At reacquisition of baseline CrBVI and Cr SO_2, mean left radial artery pr was 22mmHg(16-28mmHg)
- At flow rate of 20 to 30cc/kg/min, mean left radial artery pr was 22mmHg





Brain Protection During Arch Repair

Ann Thorac Surg 2001;72:401



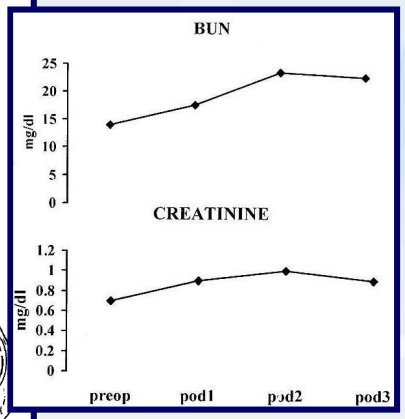
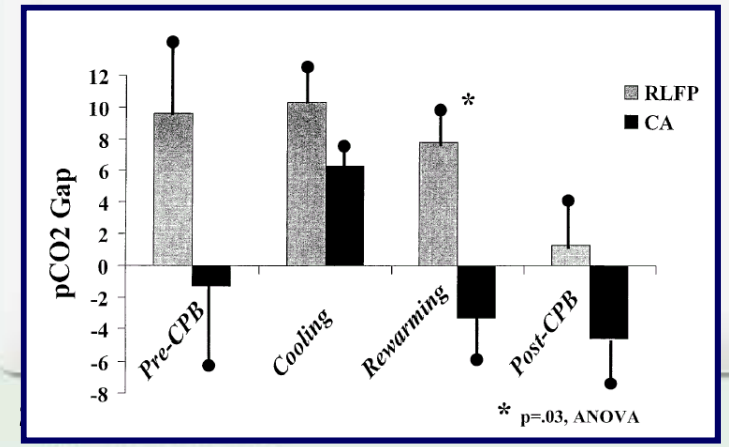
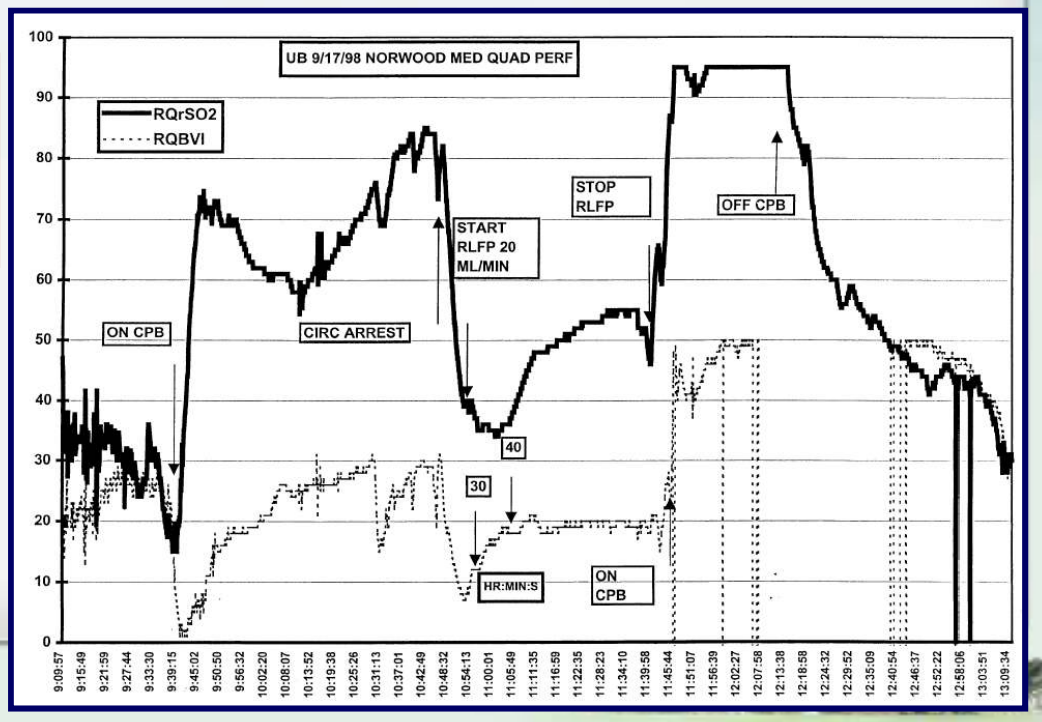


Table 2. Abdominal Aortic Blood Pressure and Quadriceps Near-Infrared Spectroscopic Data[a]

Variable	CPB	DHCA	RLFP	*p* Value[b]
Abdominal aortic blood pressure (mm Hg)	29 ± 5	0 ± 0	12 ± 3	0.0001
QrBVI	10 ± 27	−17 ± 26	5 ± 24	0.0001
$QrSO_2$	68 ± 20	33 ± 12	57 ± 25	0.0001

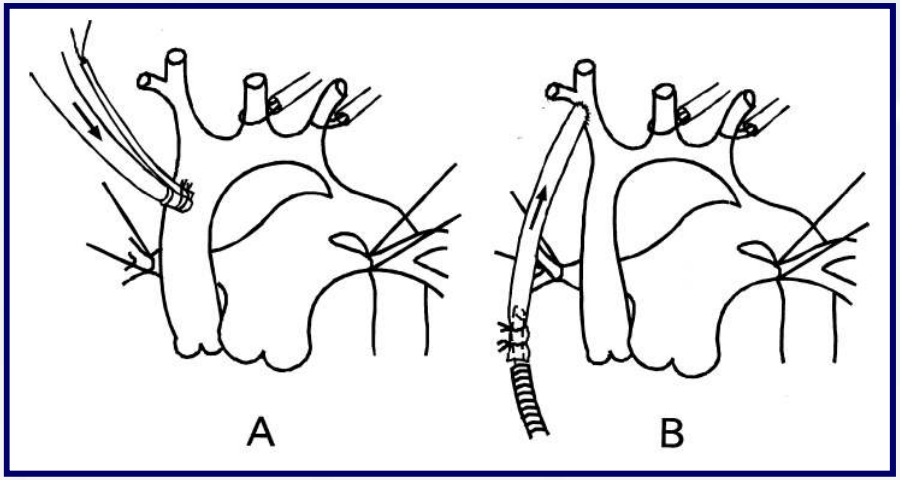
- Regional low-flow perfusion provides **somatic circulatory support** during neonatal aortic arch surgery

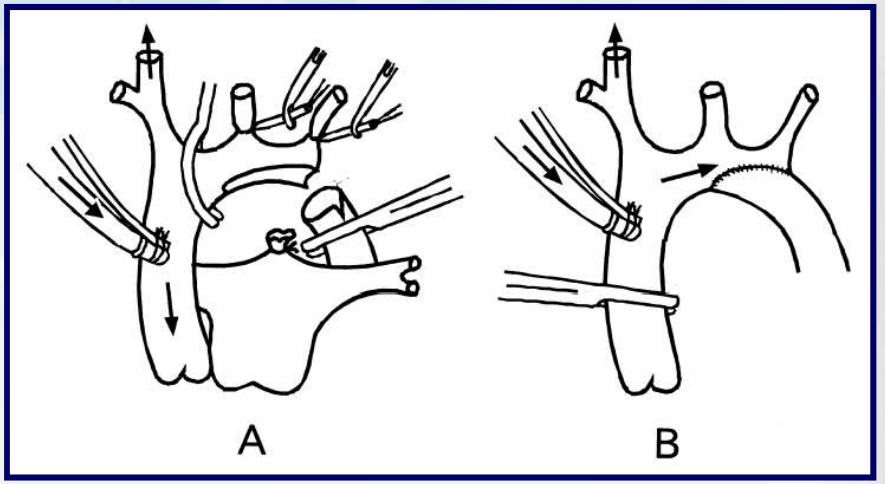


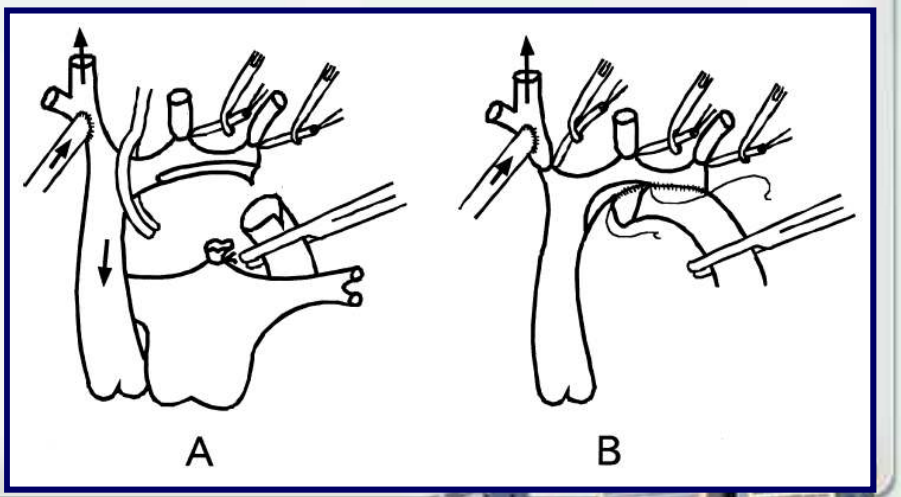


Brain Protection During Arch Repair

Ishino K, et al.
Okayama University Medical School
Eur J Cardiothorac Surg 2000;17:538



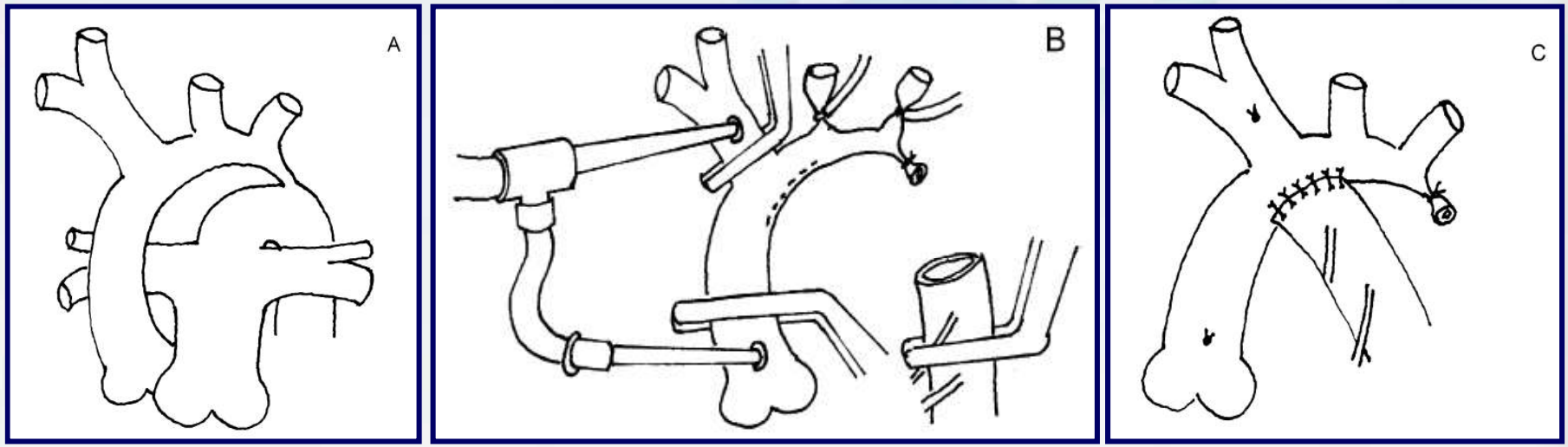




- **30 - 50% of full flow (150ml/kg/min)**
  at 28 ℃ of rectal temperature
  to keep radial pressure of 30- 45mmHg

Brain Protection During Arch Repair

Cheong Lim, et al.
Bucheon Sejong hospital
Eur J Cardiothorac Surg 2003;23:149

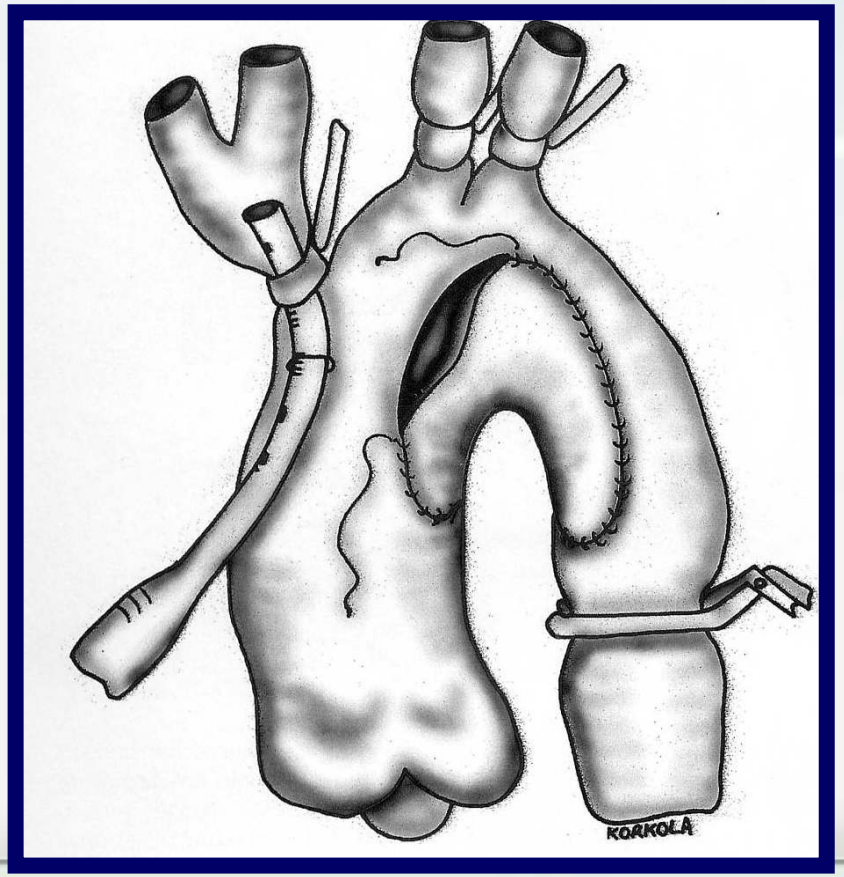


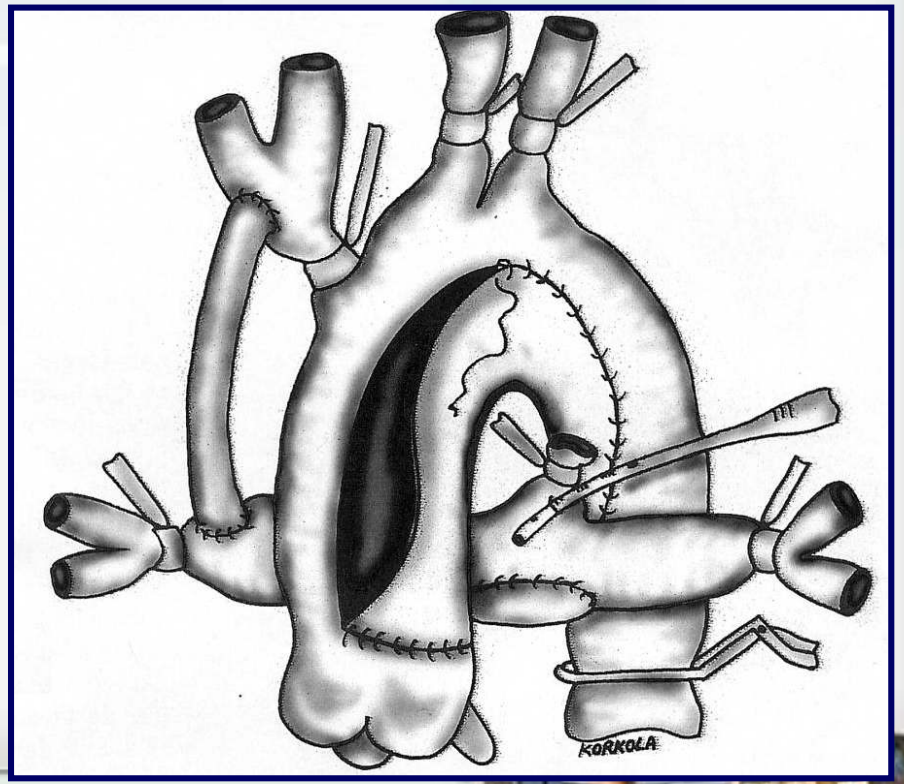
- **Rectal temperature; 18 ℃**
- **Flow rate; 50-100ml/kg/min**
- **Right radial artery pressure; 40- 50mmHg**
- **Mean Hematocrit; around 20%**

Brain Protection During Arch Repair

Tchervenkov CI, et al.
The Montreal Children's Hospital
Ann Thorac Surg 2002;47:1730
Seminars in Thoracic and
Cardiovascular Surgery 2002

- **Flow rate; 0.3 – 0.4 L/min/m^2**
- **Minimum temperature; 18 ℃**





Perfusion during arch repair

At PNUYH

- ***In mild COA or no PDA***
  One arterial cannulation (through Graft to innominate artery)
  Cardiac procedure (VSD closure) first, and then arch repair

- ***In severe COA***
  One arterial cannulation (through Graft to innominate artery)
  Arch repair first and VSD repair

- ***In IAA or very severe COA***
  Two arterial cannulations (through Graft to innominate artery and through PDA to descending aorta)
  Cardiac procedure first, and then arch repair

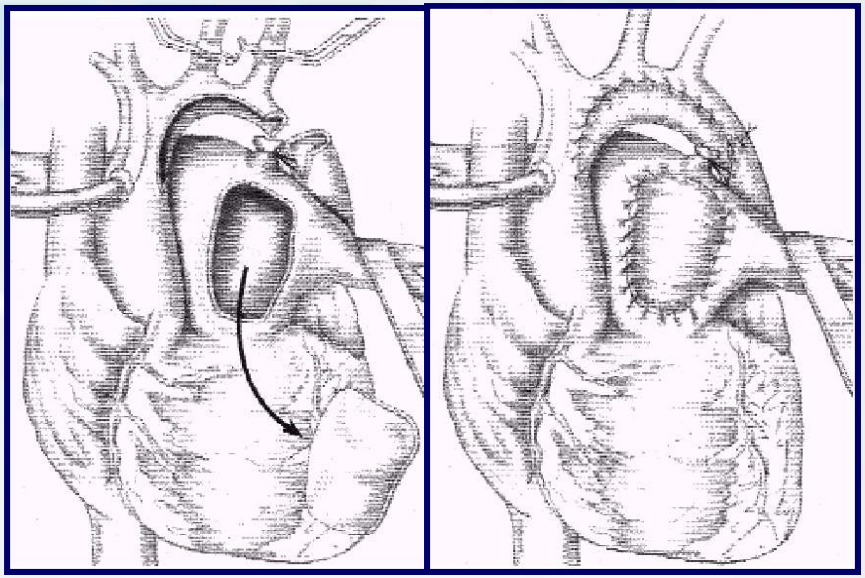
Special Technique

Pulmonary autograft patch aortoplasty

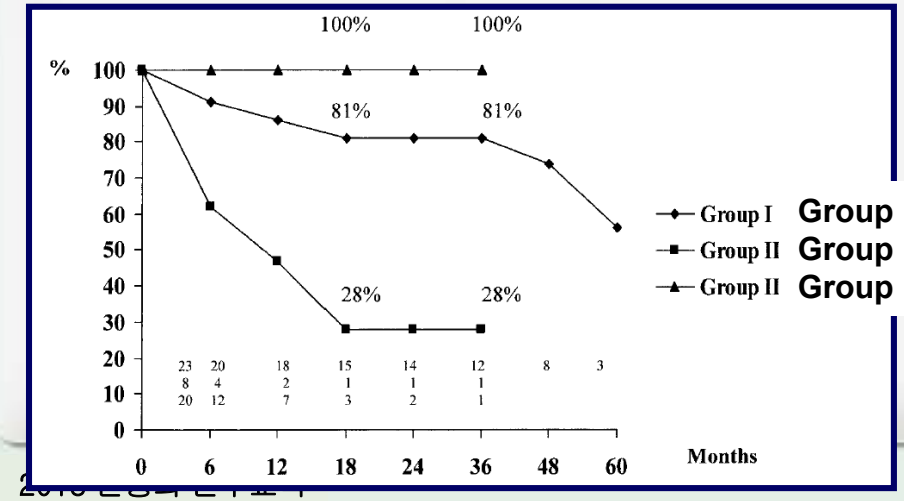
Roussin R, et al.

Marie Lannelongue Hospital

J Thorac Cardiovasc Surg 2002;123:443



Actuarial freedom from recurrent arch



Group I; direct anastomosis

Group II; homograft or pericardial patch aortoplasty

Group III; pulmonary autograft patch aortoplasty

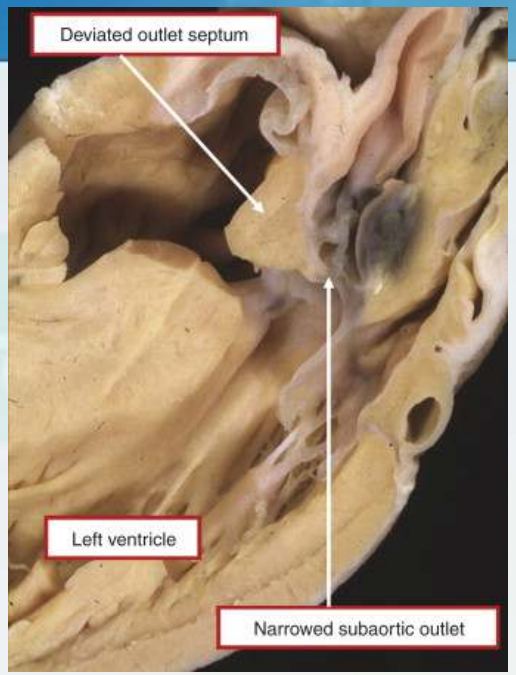
Special Technique

❑ Anterior deflection of conal septum

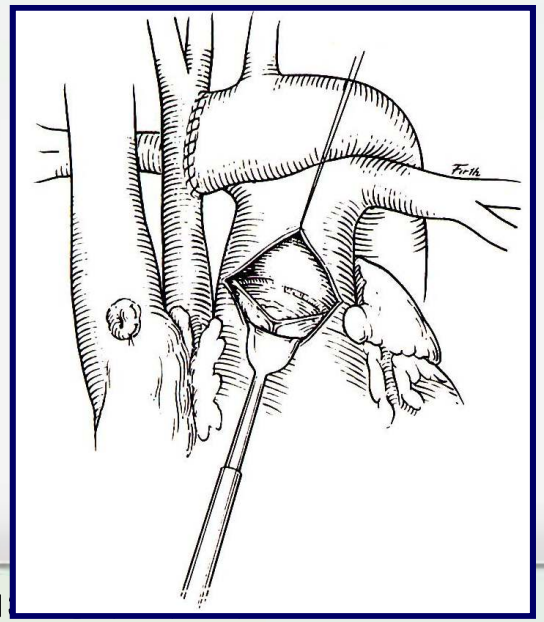
Luciani GB, et al.

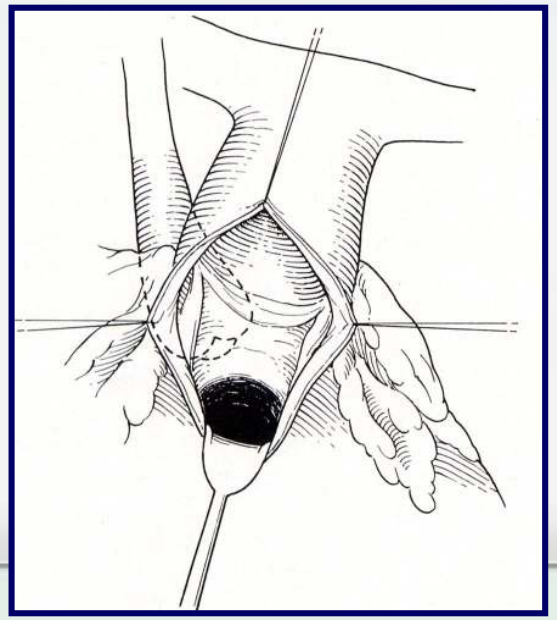
Children's Hospital Los Angeles

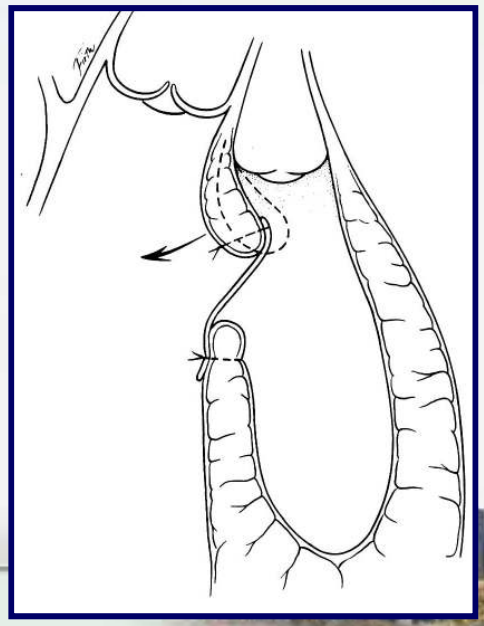
J Thorac Cardiovasc Surg 1996;111:348



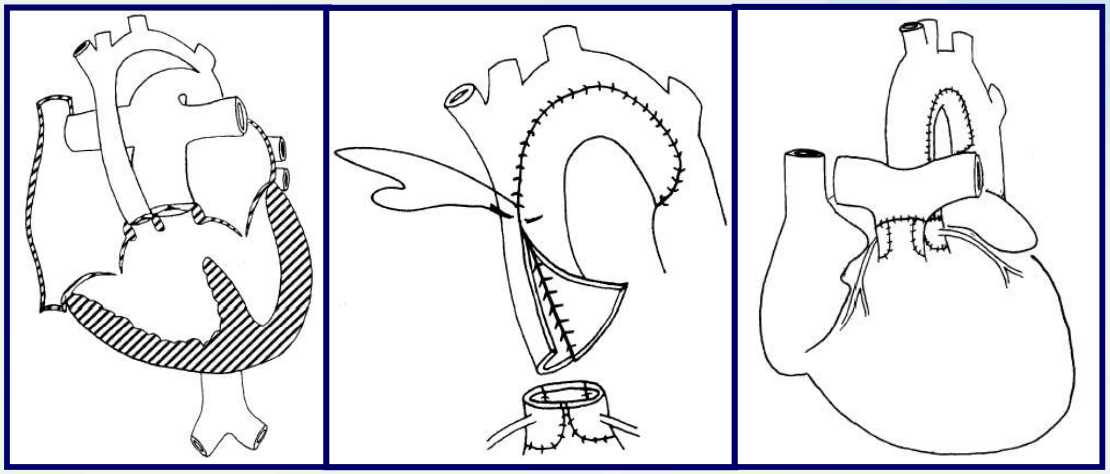
▪ **Ventricular septal patch was placed on the left side of the septum to deflect the conal septum anteriorly and away from the subaortic area without resection of the conal septum**



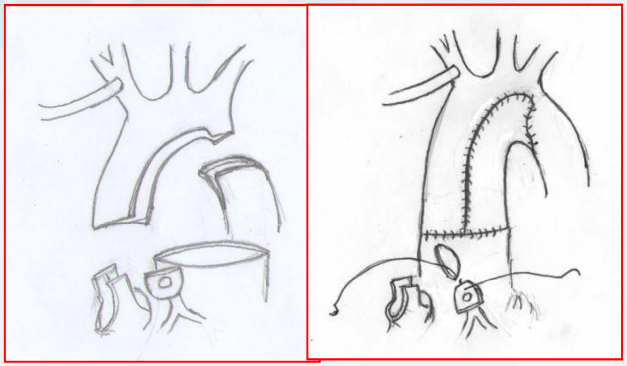


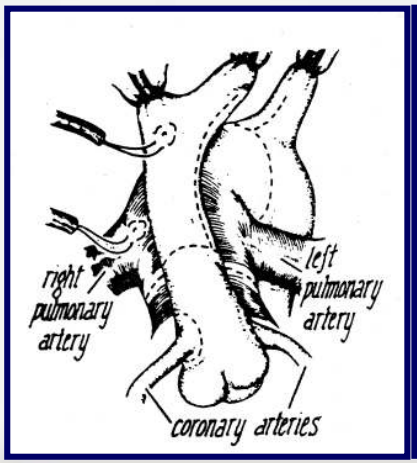


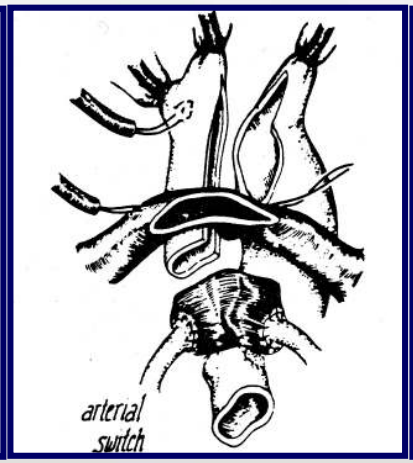
Aortic arch repair in TGA or TB anomaly

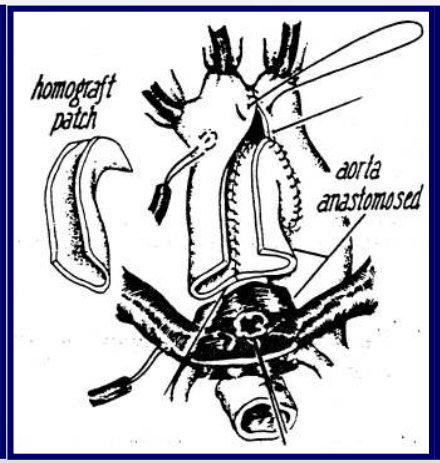


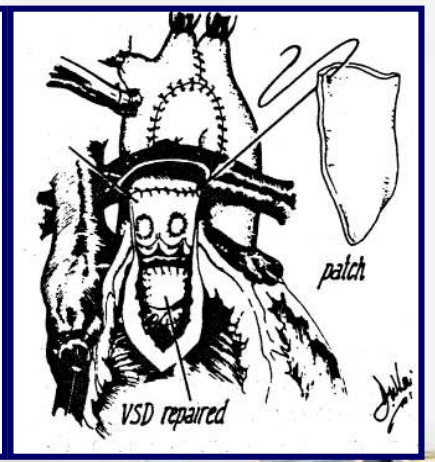
CA transfer after aortic arch repair and neoaortic reconstruction











Special Features of Postoperative Care

Managing Systemic Arterial Hypertension

- ✓ Systemic arterial hypertension is usually present after operation
- ✓ In older patients, if systolic blood pressure is greater than 150 mmHg, a β-adrenergic receptor blocking agent or captopril is administered for a few weeks
- ✓ In infants and young children, treatment is given less routinely for postoperative hypertension. Intravenous nitroprusside is the treatment of choice; however, esmolol is also effective

Special Features of Postoperative Care

Abdominal Pain

- ✓ Most have mild abdominal discomfort for a few postoperative days
- ✓ In 5% to 10% of cases, this is prominent, and abdominal distention with hypoactive bowel sounds may develop
- ✓ Treatment consists of bowel decompression via a nasogastric tube and antihypertensive drugs
- ✓ In current practice, the need for laparotomy for abdominal crisis is rare

Special Features of Postoperative Care

Chylothorax

- ✓ Copious serous or milky drainage is probably chyle, a finding in about 5% of patients
- ✓ If it continues profusely until the sixth or seventh postoperative day, reoperation is indicated

Results

Repair of Isolated Coarctation

Survival

Early (Hospital) Death

In neonates, 0% to 2%

In older infants, children, adolescents, and young adults, 1%

Time-Related Survival

12- and 24-month survivals were both 95%

Survival at 2, 5, and 10 years was 92%, 88%, and 88% (from Kaushal et al)

Survival was better for patients with isolated coarctation

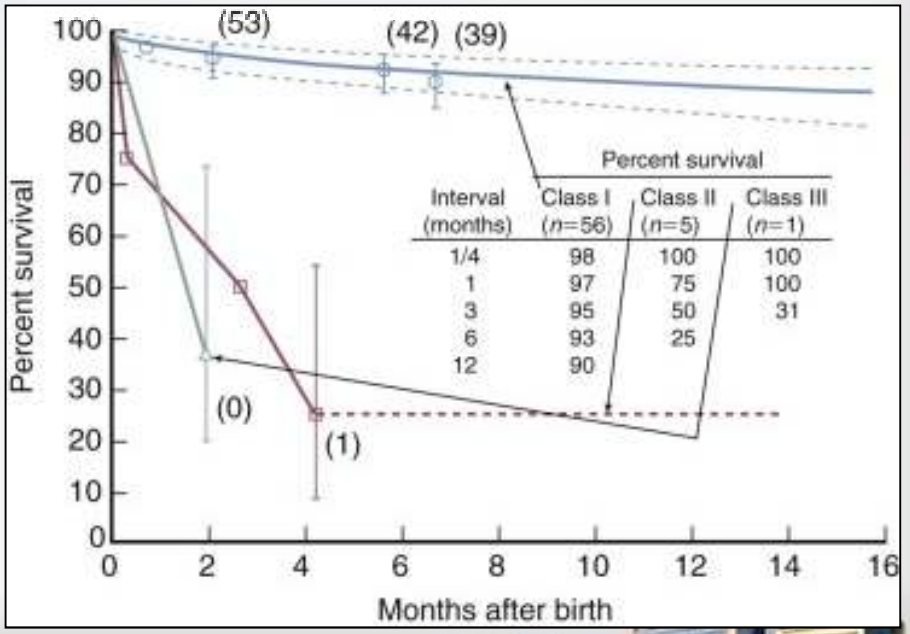
Results

Repair of Isolated Coarctation

Survival

Incremental Risk Factors for Death after Repair

- Older age at operation
- Low birth weight (?)
- Transverse arch hypoplasia
- Hypoplastic left heart class



Results

Repair of Isolated Coarctation

Late Postoperative Exercise Capacity

Exercise capacity is lower than normal (80% of predicted)

Results

Repair of Isolated Coarctation

Late Postoperative Upper Body Hypertension

Resting Values

About 50% of patients who have undergone coarctation repair have an upper body resting systolic blood pressure higher than the mean value for normal individuals

Results

Repair of Isolated Coarctation

Late Postoperative Upper Body Hypertension

Time course

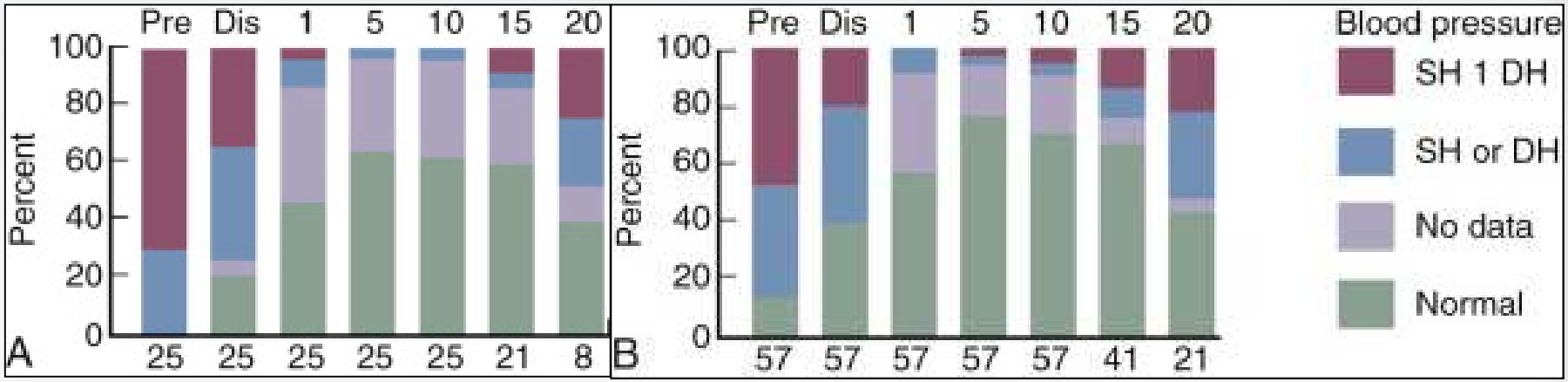
- ✓ Often considerably elevated early postoperatively.
- ✓ Normalize in most patients such that by 5 years after repair 80% to 90% of patients have normal upper body systolic and diastolic blood pressures at rest
- ✓ After 5 years, prevalence of patients with normal blood pressure begins to decline,
- ✓ 20 years after operation, only 40% to 50% have normal blood pressure. Prevalence declines still further after that.

Results

Repair of Isolated Coarctation

Late Postoperative Upper Body Hypertension

Time course



Repair at age of 5–9 years

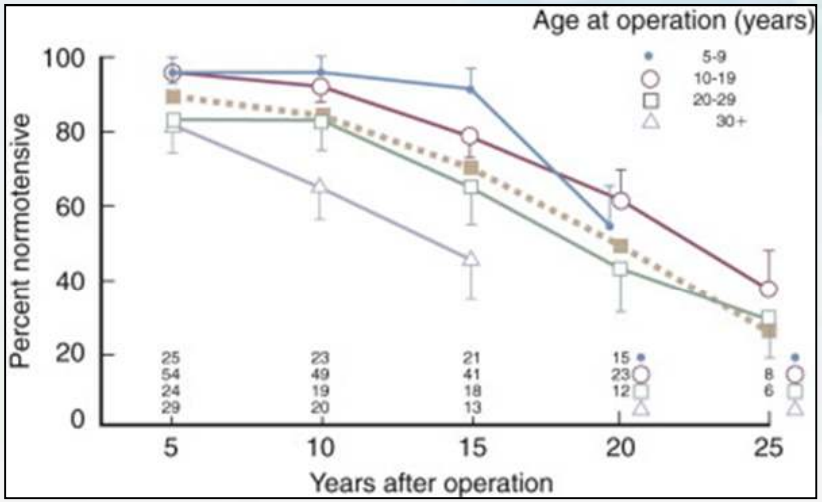
Repair at age of 10–19 years

Results

Repair of Isolated Coarctation

Late Postoperative Upper Body Hypertension

Time course



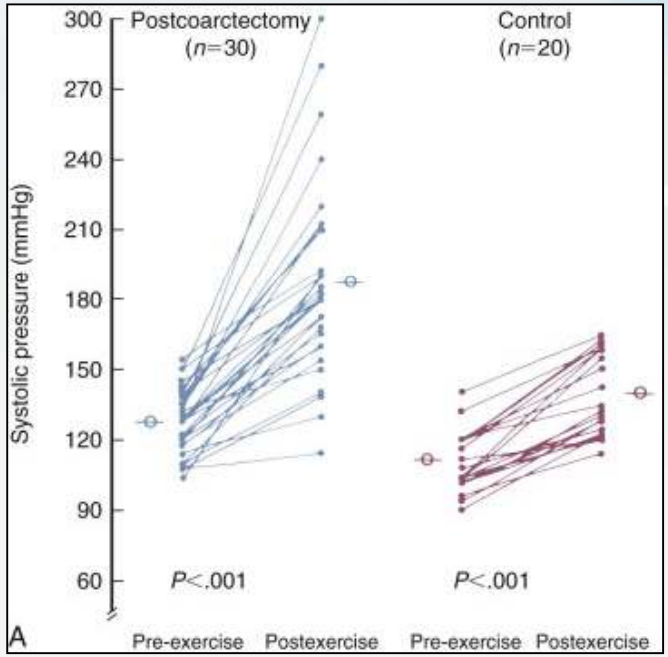
- ✓ Late hypertension is less prevalent in patients who undergo coarctation repair within the first year of life rather than at an older age
- ✓ In patients undergoing coarctation repair as adults (age ≥ 16 years), generally more than half are normotensive; the remainder are on antihypertensive medication

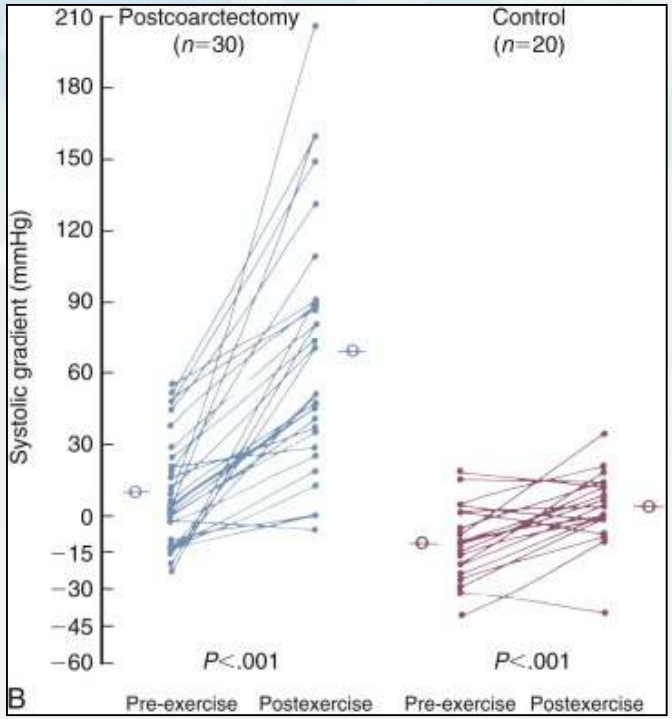
Results

Repair of Isolated Coarctation

Late Postoperative Upper Body Hypertension

Values with Exercise





- ✓ **Patients who have undergone coarctation repair experience a considerable increase in upper body blood pressure during exercise**
- ✓ **Patients operated on after age 1 year having a greater chance of developing exercise-induced hypertension**

Results

Repair of Isolated Coarctation

Late Postoperative Upper Body Hypertension

Risk Factors of Upper Body Hypertension

- Endocrine factors
- Abnormal compliance or reactivity of upper body small blood vessels
- Poorly compliant aorta proximal to coarctation repair
- Morphologically persistent or recurrent coarctation
- Presence of an angulated or "gothic-shaped" arch

Results

Repair of Isolated Coarctation

Persistent or Recurrent Coarctation

- ✓ Resting peak pressure gradient exceeding 20 mmHg across the repair area
- ✓ It is uncertain whether this represents a persistent or recurrent coarctation

● 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

Results

Repair of Isolated Coarctation

Persistent or Recurrent Coarctation

- Technical factors of **persistent coarctation**

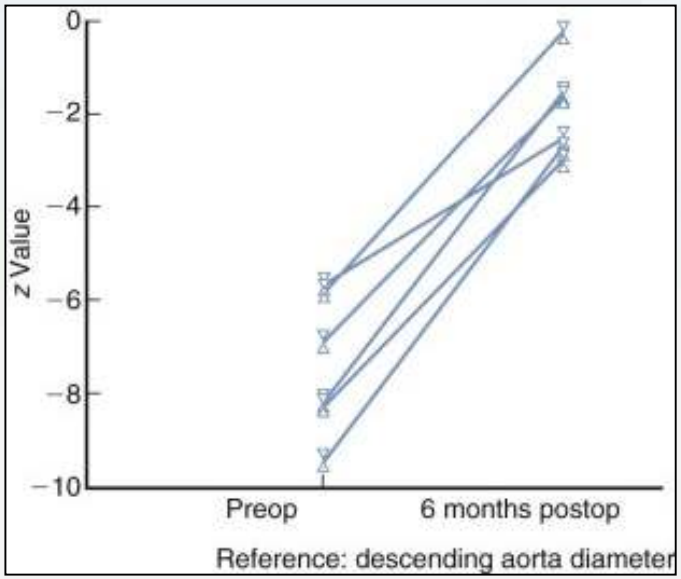
1) Insufficient resection of a long, narrow segment
2) Excessive tension due to inadequate mobilization
3) Incorrect fashioning of a subclavian flap or polyester onlay patch
4) Failure to resect an obstructing intimal ridge
5) Use of a too-small tube graft in a child

Results

Repair of Isolated Coarctation

Persistent or Recurrent Coarctation

Residual arch hypoplasia, **usually between the left subclavian and common carotid arteries**



The z value of diameter of aortic arch between left common carotid and left subclavian arteries in infants younger than age 3 months at time of coarctation repair

Results

Repair of Isolated Coarctation

Persistent or Recurrent Coarctation

Prevalence after End-To-End Anastomosis

as high as about 20% in patients operated on before age 2 years

- ✓ **Less than 5% has been demonstrated in neonates and infants younger than age 3 months using the technique described by Hanley (end-to-side primary anastomosis)**
- ✓ **End-to-side repair using median sternotomy has also been reported, and outcomes compare favorably with the extended end-to-end technique**

Results

Repair of Isolated Coarctation

Persistent or Recurrent Coarctation

Prevalence after Subclavian Flap Aortoplasty

0 ~ 6%

✓ 6% (CL 3%-10%) of 81 infants repaired by the subclavian flap
27% using end-to-end at the Toronto Hospital for Sick Children

Prevalence after End-To-End Anastomosis with Subclavian Flap Aortoplasty

✓ Lower than subclavian flap aortoplasty

Prevalence after Patch Aortoplasty

✓ Late results of polyester or polytetrafluoroethylene (PTFE) patch aortoplasty have been variable

Results

Repair of Isolated Coarctation

Paraplegia after Repair

0.4% among 12,532 coarctectomies (collective review by Brewer)

- ✓ Wherever the collateral circulation typical of coarctation has not developed, risk of paraplegia is increased

Situations of poor development of collateral circulation

1) Coarctation in infants
2) Coarctation proximal to the left subclavian artery
3) Coarctation with patent ductus arteriosus supplying the descending thoracic aorta
4) Coarctation associated with stenosis at the origin of the left subclavian artery
5) Aberrant right subclavian artery arising from descending aorta
6) Less severe coarctation
7) Re-repair

Results

Repair of Isolated Coarctation

Early Postoperative Hypertension and Abdominal Pain

- ✓ Nearly all patients, including infants, have some systolic and diastolic hypertension for a variable period after coarctation repair.
- ✓ Many patients, if observed carefully, have mild abdominal discomfort and distention during the first 5 or 6 postoperative days.
- ✓ In 10% to 20% of cases, this becomes sufficient to produce important discomfort and distention

- ✓ Early systolic hypertension within the first 36 postoperative hours
- ✓ Delayed mainly diastolic hypertension after 48 hours that lasted 7 to 14 days (related to abdominal pain)
- ✓ Sympathetic nervous system is responsible for the early phase
- ✓ Renin–angiotensin system plays a major role in the later phase, (more recent information would indicate that the renin–angiotensin system also plays a role in the early phase)

Results

Repair of Isolated Coarctation

Left Arm Function after Subclavian Flap Aortoplasty

- ✓ Rarely (<1% of patients) does actual gangrene develop
- ✓ The affected arm is smaller later in life, but uncommonly is it perceptibly so

Late Aneurysm Formation

- ✓ A true aneurysm from progressive deterioration of the aortic wall opposite a prosthetic onlay patch has been reported
- ✓ False (suture line) aneurysms can be mycotic when they occur early postoperatively

Results

Repair of Isolated Coarctation

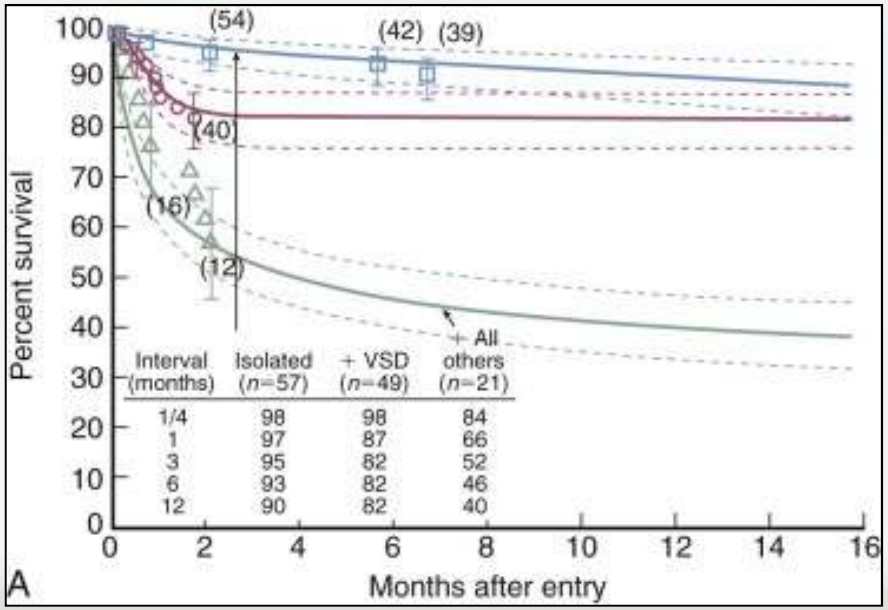
Other Events

- ✓ Heart failure may occasionally persist postoperatively
- ✓ Infective endocarditis occasionally occurs on the aortic or mitral valve
- ✓ Cerebrovascular accidents are more common in patients with persistent hypertension

Results

Repair of Coarctation and Coexisting VSD

- ✓ VSDs have the same tendency to close as when coarctation is not present
- ✓ Some single-institution studies indicate that presence of a VSD has no effect on either early or late risk, with excellent early and midterm outcomes reported



Congenital Heart Surgeons Society, 1990 to 1991

Results

Repair of Coarctation and Coexisting VSD

Treatment Options

1) Repair of coarctation via thoracotomy with medical management of the VSD
2) Repair of coarctation via thoracotomy with PA banding
3) Coarctation repair via median sternotomy with VSD closure (either hypothermic circulatory arrest or continuous perfusion is required)

- ✓ **Natural history of VSD associated with COA is not established**
- ✓ **Another approach involves coarctation repair with pulmonary trunk banding using absorbable material (polydioxanone). Band reabsorption occurred over approximately 6 months in an experience with 11 selected patients. VSD closure was necessary in only one patient following band reabsorption.**

Results

Repair of Coarctation and Other Major Coexisting Intracardiac Anomalies

- ✓ Early and intermediate-term survival is less good in this group, particularly when patients are in hypoplastic left heart classes II
- ✓ This was a highly heterogeneous group of patients and experience with any one group is small.
- ✓ Individual institutions have recently demonstrated substantially better outcomes in these patient groups, with 5-year survival of more than of more than 70%

Indications for Operation

Isolated Coarctation

- ✓ Diagnosis of isolated coarctation is an indication for operation
- ✓ In critically ill neonates with coarctation, intravenous PGE1 (0.1 mg · kg−1 · min−1) is begun immediately and continued until the situation is remedied at operation

→ Response is dramatic in about 80% of infants, with reappearance of femoral pulses and disappearance of metabolic acidosis from hypoperfusion of the lower body

(Operation is delayed until the baby's condition has stabilized)

- ✓ No benefit from delay of operation

In occasional patients with a mild to moderate degree of left heart hypoplasia in whom it may not be clear that the left heart is adequate to sustain the systemic circulation, the **subclavian flap operation can be particularly useful**. With this operation, the ductus arteriosus can be preserved, and the PGE1 can then be weaned slowly to test the adequacy of the left heart once the coarctation is repaired.

Indications for Operation

Coarctation and Coexisting VSD

- ✓ When the VSD is small or moderate in size, and particularly if it is muscular and occasionally perimembranous, spontaneous reduction in size, and closure, are real possibilities. In this situation the option of coarctation repair alone, with subsequent observation of the VSD and patient, is the preferred treatment protocol.

Indications for Operation

Coarctation and Other Major Coexisting Intracardiac Anomalies

- ✓ Candidates for single ventricular repair;
  COA repair + PA band
- ✓ Candidates for biventricular repair;
  One-stage total repair
- ✓ Coarctation and atrioventricular septal defect;
  the preferred approach in most cases is to repair the coarctation and defer intracardiac repair for several months.

Special Situations and Controversies

Coarctation Proximal to Left Subclavian Artery

- ✓ Coarctation proximal to the left subclavian artery is rare (≈1% of all cases).
- ✓ Stenosis is localized, and femoral pulses are usually only slightly decreased and systolic pressure gradient across the coarctation is mild (<20 mmHg) or moderate.
- ✓ Coarctation is often not detected until young adult life, at which time upper body hypertension is often present.
- ✓ A collateral circulation is generally not well

Special Situations and Controversies

Mild and Moderate Coarctation in Classic Position

- ✓ Uncommonly in infants and older patients, moderate coarctation is present in the classic position.
- ✓ Collateral vessels are absent.
- ✓ The natural history of this entity is not clear and thus neither are indications for operation.
- ✓ Degenerative changes are, however, prone to occur in the region of the coarctation
- ✓ When calcification is apparent, resection and replacement of this area with a tube graft may be recommended

Special Situations and Controversies

Preventing Paraplegia as a Complication of Repair

Preoperative identification of patients with potentially inadequate collateral circulation

1) Absence of rib notching on the chest radiograph
2) Absence of palpable parascapular pulsations
3) Only mildly diminished femoral pulsations
4) Diminished left radial pulse (stenosis of the origin of the LSCA)
5) Diminished right radial pulse (aberrant RSCA from distal to the coarctation)

If uncertainty remains, pressure can be measured at thoracotomy in the descending aorta with the proximal aortic clamp temporarily in place; if this pressure is less than 50 mmHg, special measures are needed

Special Situations and Controversies

Preventing Paraplegia as a Complication of Repair

- ✓ Use of mild hypothermia (35°C nasopharyngeal or tympanic membrane temperature) is a good precaution for in infant and older patients. (probably allows 30 minutes of safe aortic clamping). Ice-cold saline lavage may need to be prolonged to about 10 minutes

- ✓ Temporary bypass shunt, usually from left subclavian to descending thoracic aorta, or femorofemoral CPB may be used during the period of aortic clamping

Special Situations and Controversies

Reintervention for Persistent or Recurrent Coarctation

- ✓ Current indication for reintervention is demonstrating a reduced luminal diameter of greater than 50% at the anastomosis. Under this circumstance, heart failure or upper body hypertension (systolic pressure greater than 140 mmHg in infants and children) is an indication for reintervention.

- ✓ Currently, percutaneous balloon aortoplasty is generally the treatment of choice

Special Situations and Controversies

Balloon Aortoplasty and Stenting for Coarctation

- ✓ Percutaneous balloon aortoplasty as primary treatment is controversial, particularly in small patients
- ✓ Higher rate of aneurysm formation
- ✓ Higher rate of recoarctation
- ✓ Arterial occlusion at the site of catheter intervention
- ✓ Percutaneous stenting of both native and recurrent coarctation is currently well established, particularly in older patients, but longer-term outcome is lacking

The engineering of stents continues to evolve. Covered stents, growth stents, and self-expanding nitinol stents have recently been introduced into clinical practice for coarctation management

Special Situations and Controversies

Coarctation in Adults

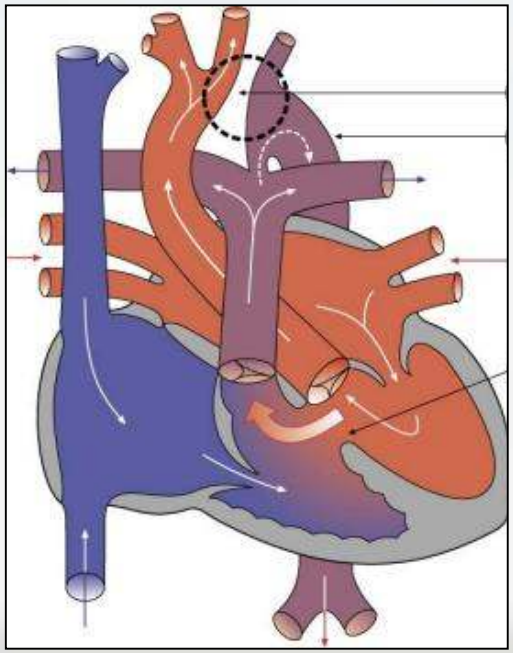
- ✓ Excellent results have been achieved with operation in patients older than 50 years (up to 73 years) without mortality and with important improvement of hypertension
- ✓ Effective results can be achieved with balloon angioplasty and stenting for uncomplicated native coarctation in adults
- ✓ When operation is considered for more complex arch obstructions in adults, extra-anatomical bypass grafting has proved to be a good surgical option
- ✓ Adults with particular risk of spinal cord injury may benefit from partial left heart CPB when a left thoracotomy approach is taken

Interrupted Aortic Arch

Definition

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

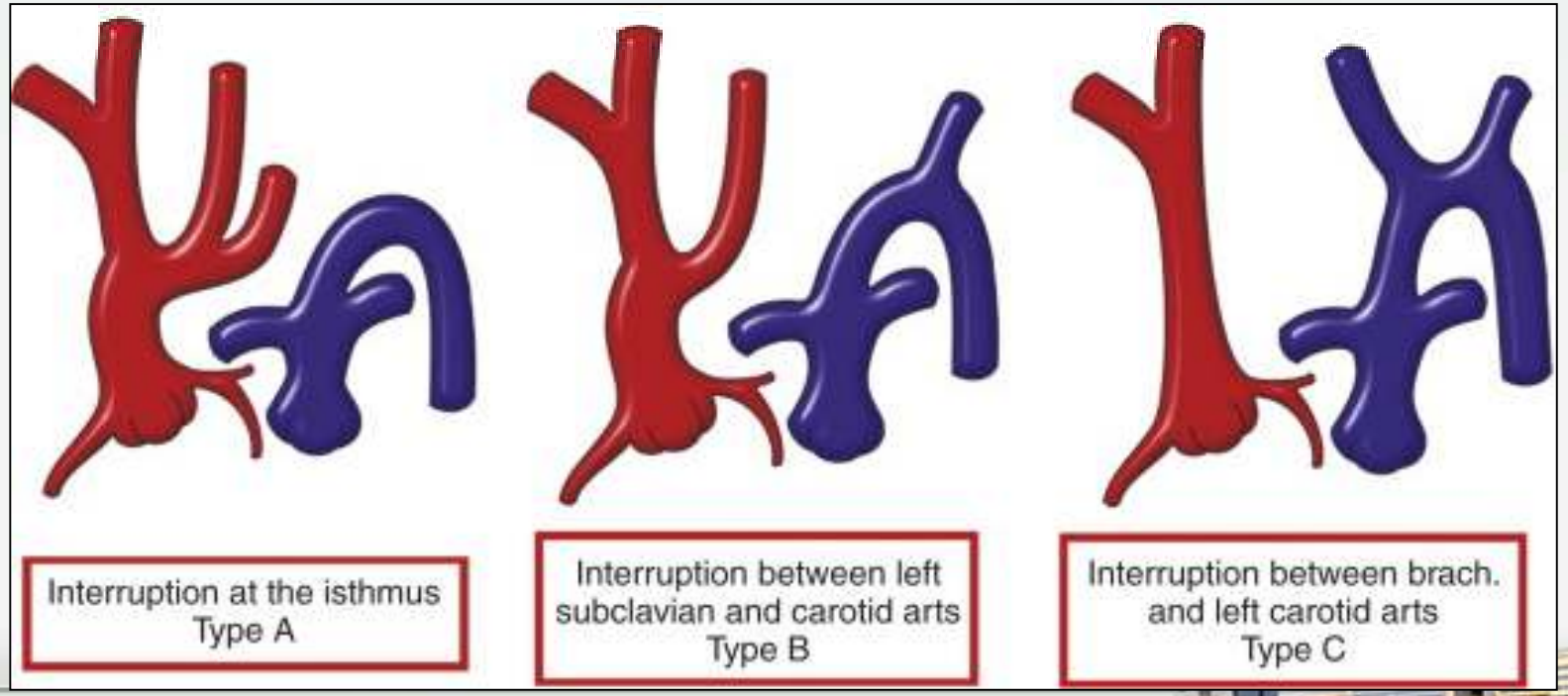
Those rare specimens that exhibit a fibrous strand connecting two widely separated ends are also included under aortic arch interruption rather than coarctation



Morphology and Morphogenesis

Types of Interrupted Aortic Arch

Type A; Interruption located just distal to left subclavian artery (40%)
Type B; Interruption located between left subclavian and left common carotid arteries (55%)
Type C; Interruption located between left common carotid and brachiocephalic artery (5%)



Morphology and Morphogenesis

Aortic Arch

- ✓ Anomalies of the origins of brachiocephalic vessels are frequent (aberrant right subclavian artery)
- ✓ Characteristically, the ascending aorta is about half normal diameter and is straight, dividing into two branches of about equal size (the V sign)
- ✓ Pulmonary trunk is huge
- ✓ Descending aorta is a direct continuation of the ductus arteriosus

Morphology and Morphogenesis

Left Ventricular Outflow Anomalies

- ✓ Bicuspid aortic valve in 30% to 50% of patients
- ✓ Subaortic stenosis may be present or develop
- ✓ **Left ventricular outflow tract obstruction**;
  conal septal posterior malalignment,
  muscle of Moulaert,
  small aortic annulus

Coexisting Cardiac Anomalies

- ✓ A large VSD in over 95%, Mostly conoventricular in type
- ✓ Posteriorly malaligned and displaced infundibular septum usually produces subaortic obstruction of variable severity
- ✓ Truncus arteriosus, AP window, Taussig-Bing anomaly

Morphology and Morphogenesis

Morphogenesis and Associated Syndrome

Microdeletion of q11 of chromosome 22,
velocardiofacial syndrome are common
DiGeorge syndrome is frequent (up to half of type B IAA)

Clinical Features and Diagnostic Criteria

- Intracardiac shunt, high afterload
  → Severe heart failure
- PDA closes
  → Metabolic acidosis, Anura, Femoral pulse ↓
- Differential cyanosis is usually not visible
  reversed differential cyanosis can be seen in TGA

✓ Echocardiography usually provides all the necessary diagnostic information relating to both aortic arch and intracardiac anomaly

✓ Cardiac CT angiography provides excellent spatial resolution

Natural History

- 1 ~ 4% of autopsy cases of CHD
- 1.3% of infants presenting critical CHD
- 0.06 per 1000 births
- Male predominance
- Highly lethal, median age of death, 4-10 days
  75% die within 1 month of birth
- When the ductus is obliterated in fetal life, collateral circulation already present at birth and survival is usual

Technique of Operation

Repair of IAA and VSD

One-stage gave the best 5-year survival

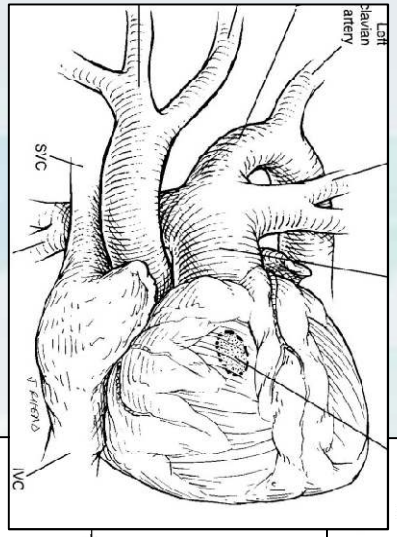
Repair using continuous perfusion

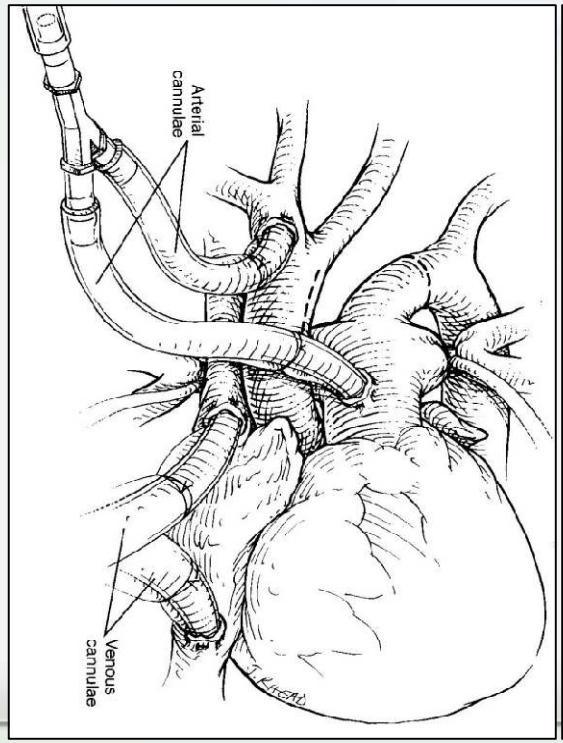
Repair using circulatory arrest

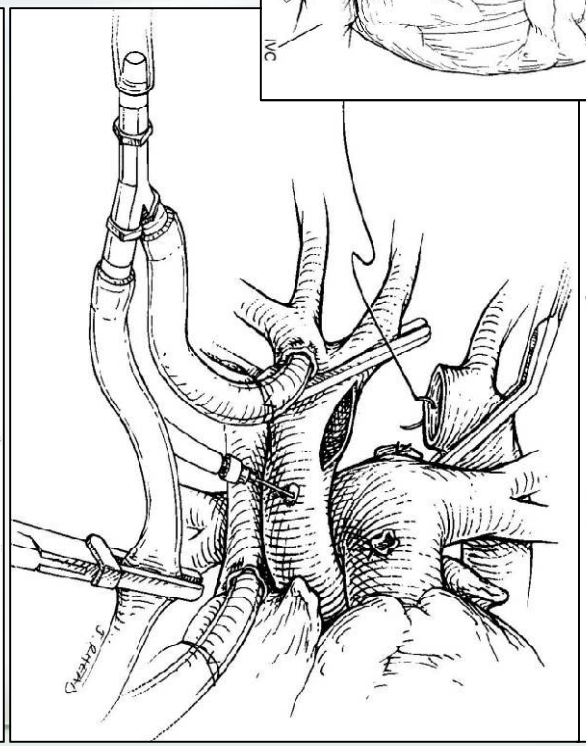
Aberrant RSCA is usually divided
Sometimes LSCA is divided for tensionless anastomosis between ascending aorta and descending aorta

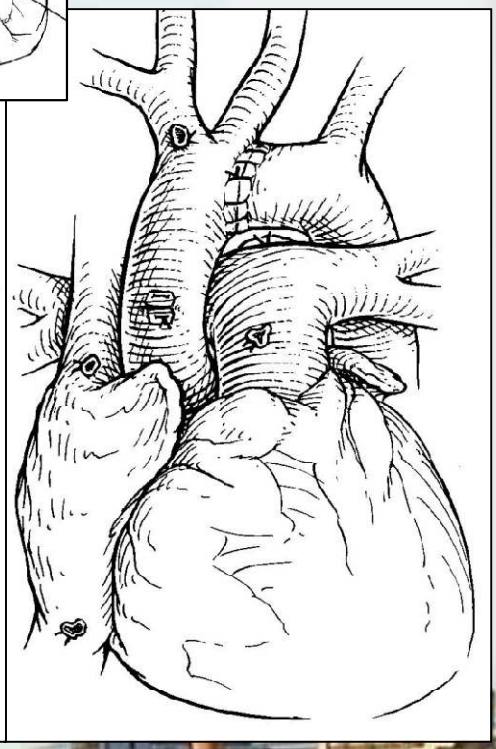
Technique of Operation

Repair of IAA









Technique of Operation

Repair of IAA and VSD and LVOTO

Surgical options

1) Direct muscular or fibromuscular LVOTO resections along with arch and VSD repair
2) Norwood operations with Rastelli septation
3) Norwood operations, followed by staged Rastelli septation
4) Ross-Konno operation

Repair of IAA and Other Coexisting Cardiac Anomalies

Several techniques have been described for one-stage repair of IAA and other coexisting cardiac anomalies

Special Features of Postoperative Care

- Special attention is paid to the possible hypocalcemia if DiGeorge syndrome is present
- Left lung atelectasis caused by **left main bronchus compression**, prevalence is low, surgical intervention is needed
- Postoperative **LVOTO > 30mmHg** systolic pressure gradient, particularly if there is LV dysfunction or hemodynamic instability, reoperation is required

Results

Survival

Early Death

In IAA + VSD by CHSS study
Type A; 4%
Type B; 11%

Time-related survival

Optimal repair in type A ; 5-year survival of 93%
in type B; 5-year survival of 83%

Modes of Death

- ✓ Most deaths are with acute or subacute heart failure
- ✓ Some are related to late reoperations for LVOT reoperation or aortic arch obstruction

Results

Incremental Risk Factors for Premature Death

Demographic

- **Lower Birth weight**
- **Younger Age at repair**

Morphologic

- **IAA type B**
- **Outlet or trabecular VSD**
- **Smaller Size of VSD**
- **Smaller Dimension (z) of LV-aortic junction**

Data from Jonas and colleagues

Results

Left Ventricular Outflow Obstruction

Patients with interrupted aortic arch often have at least some degree of hypoplastic left heart physiology as well

The specific features all relate to the LVOT

1) Posterior malalignment of infundibular septum
2) Hypertrophy of anterolateral muscle bundle of the left ventricle (muscle of Moulaert)
3) Bicuspid dysplastic aortic valve
4) Narrowness of aortic anulus
5) Hypoplasia of ascending aorta and aortic arch; occasionally this is apparent before operation, but more often it becomes evident after repair

Results

Left Ventricular Outflow Obstruction

- ✓ LVOT obstruction has important implications for both short- and long-term prognosis

- ✓ In about 40% of patients with a conoventricular VSD, or with a VSD in the outlet portion of the right ventricle, evidence of LVOT obstruction develops at midterm or late follow-up, even when the LVOT was adequate after initial operation

- ✓ Risk factors for late LVOT intervention were
  1) single ventricle
  2) aberrant right subclavian artery
  3) bicuspid aortic valve.

Results

Persistent or Recurrent Aortic Arch Obstruction

- ✓ Late reoperation is frequent after neonatal repair of IAA

- ✓ There are multiple reasons for reoperation;
  1) Recurrent arch obstruction
  2) LVOT obstruction
  3) Residual VSD
  4) Bronchial compression
  5) Diaphragm palsy
  6) Complete heart block, and bicuspid aortic valve

- ✓ Reoperation from all causes was 40% at 15 years in a single center study

Indications for Operation

- Diagnosis of interrupted aortic arch is an indication for operation, no matter what the coexisting cardiac anomaly. Severe chromosomal abnormalities may contraindicate surgical intervention

Managements

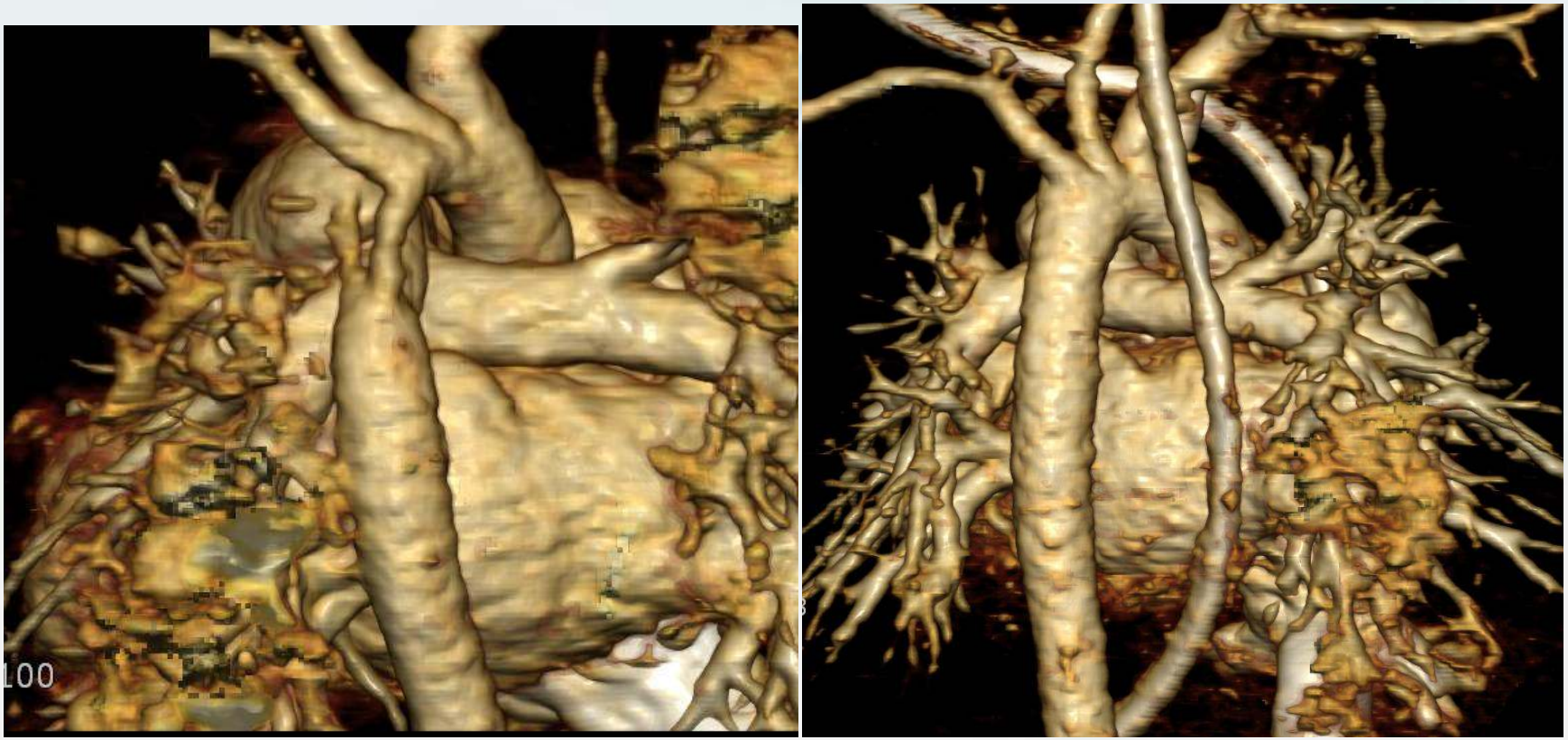
An infusion of PGE1 is begun
infant is intubated and appropriately ventilated (high FiO2 is avoided)
If the infant's condition is good, operation is undertaken
If the infant's condition is not good,

1) Right-to-left shunting into the descending aorta for augmenting systemic blood flow by increasing PVR
   increasing CO2 in the inspired gas mixture
   mildly hypoventilating the patient so that PaO2 is about 40mmHg
2) Cardiac output and renal blood flow are increased by infusing dopamine
3) Acidosis is corrected by intravenous sodium bicarbonate

Special Situations and Controversies

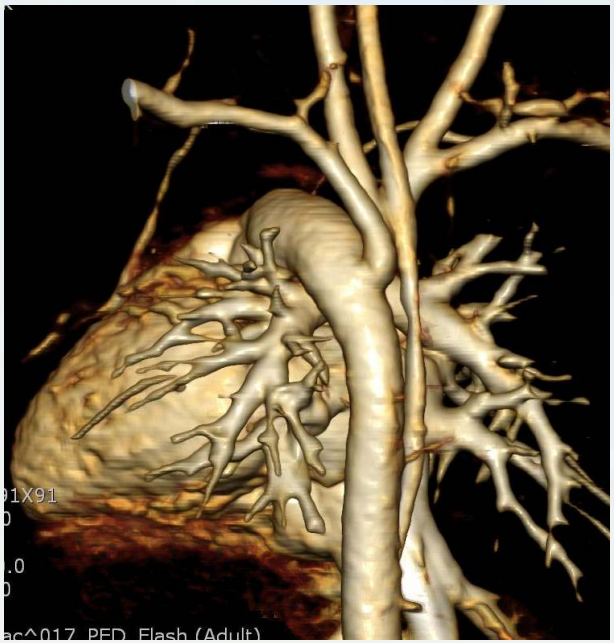
- Preoperative assessment of the LVOT is difficult because preoperative physiologic measurements are unreliable predictors of postoperative physiology
- Using echocardiography, if the smallest diameter of the subaortic region (measured in millimeters) is equal to or greater than body weight (measured in kilograms), the LVOT will be adequate
- Others advocate directly resecting the obstruction in the LVOT at the time of the arch repair and VSD closure; however, the 1994 CHSS data suggest that attempting to do this is a risk factor for death

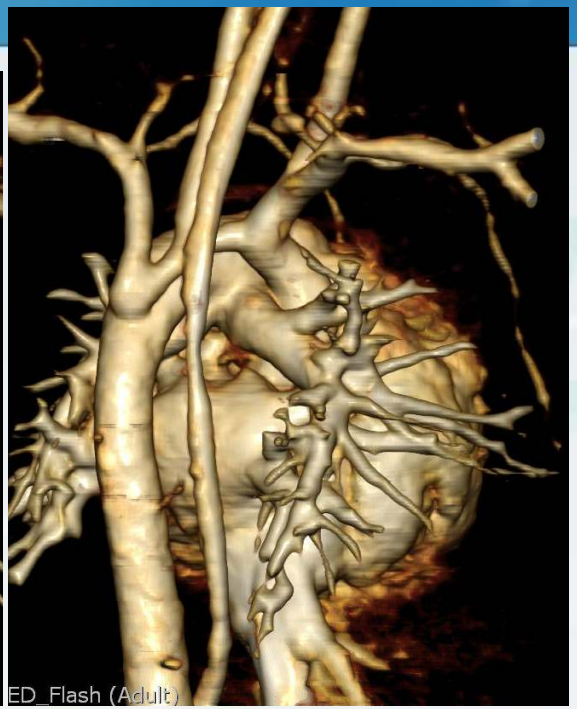
Cases



COA + VSD **One-stage repair of COA/VSD**

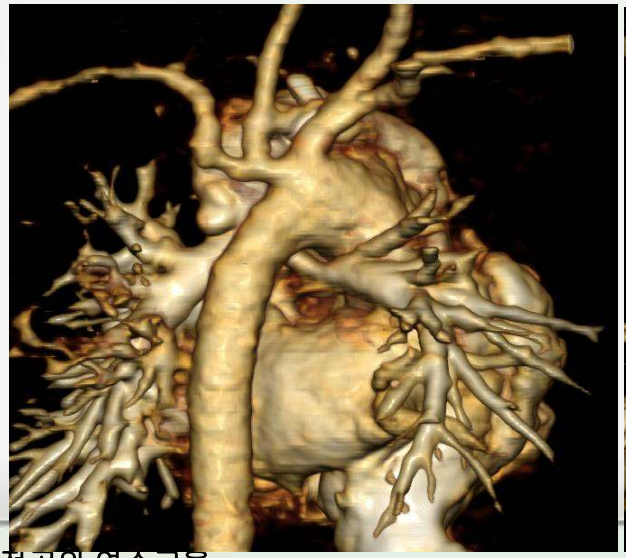
Cases

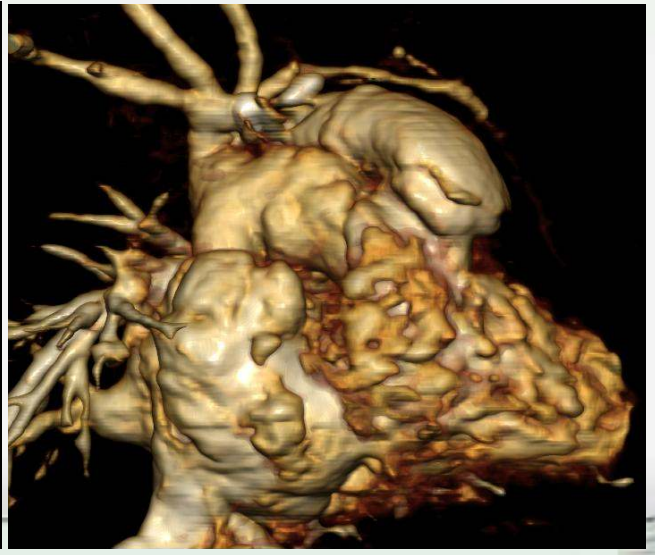




COA + VSD
+ Severe AS

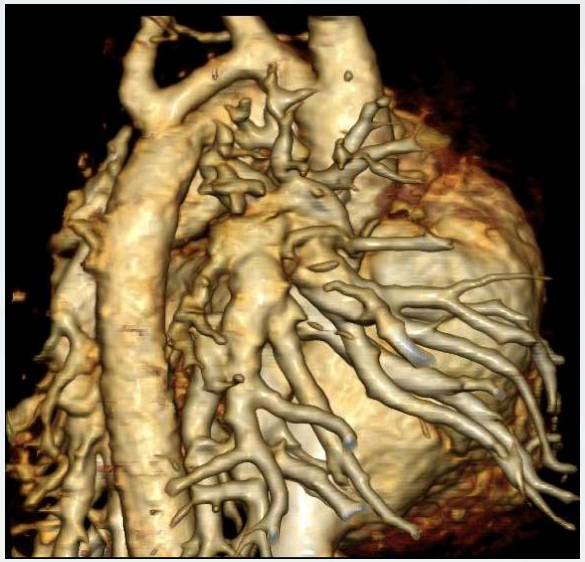
Norwood-Rastelli procedure

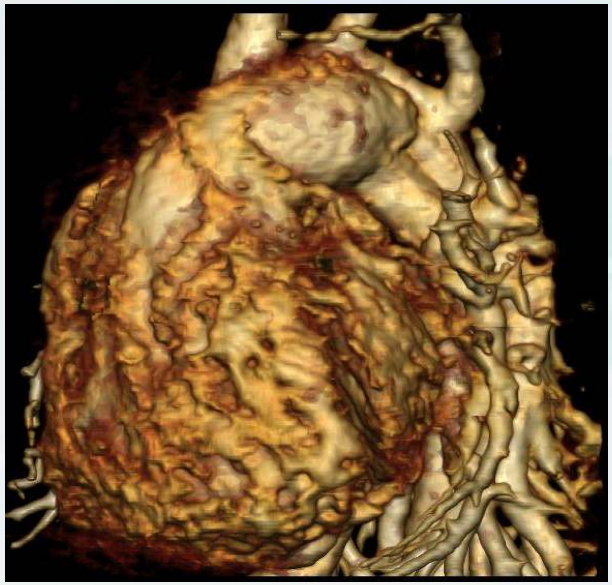




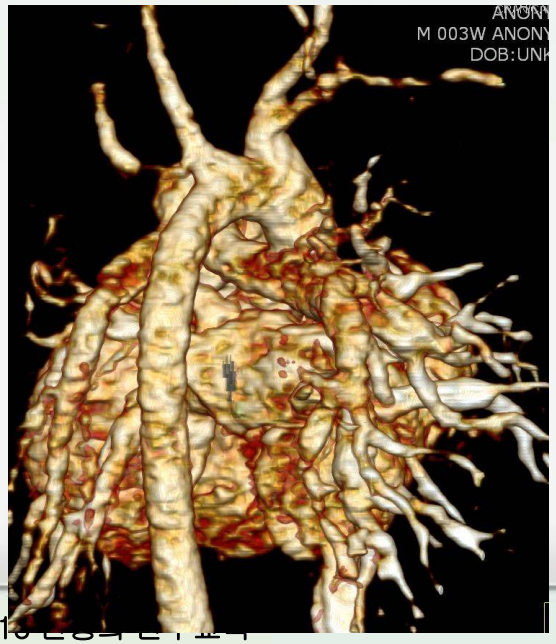
Postop.

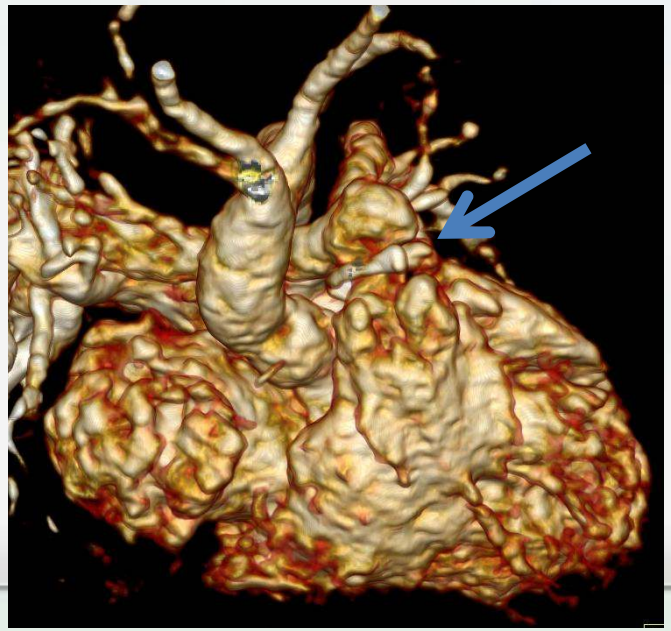
Cases





COA + VSD
+ Hypoplastic LV

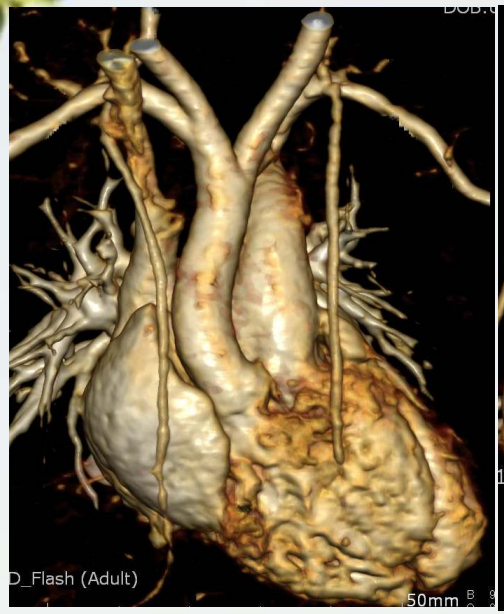


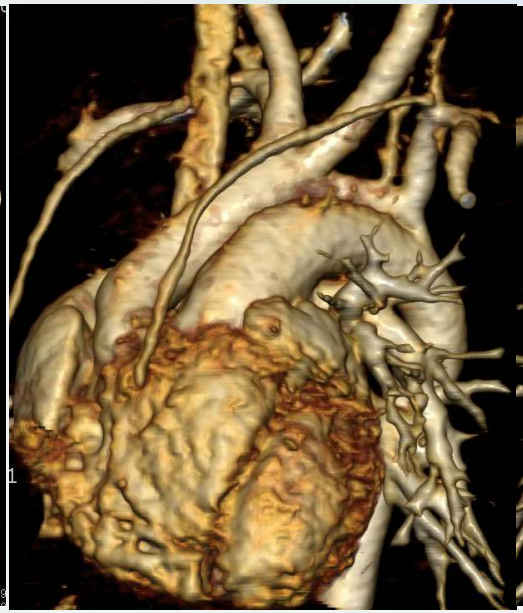


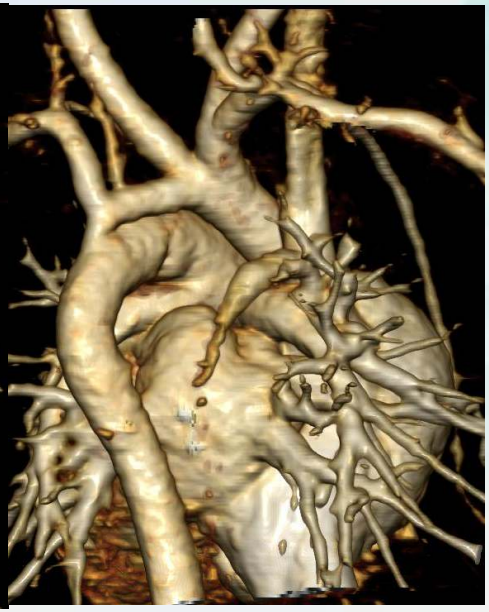
COA repair +
PA banding

Postop.

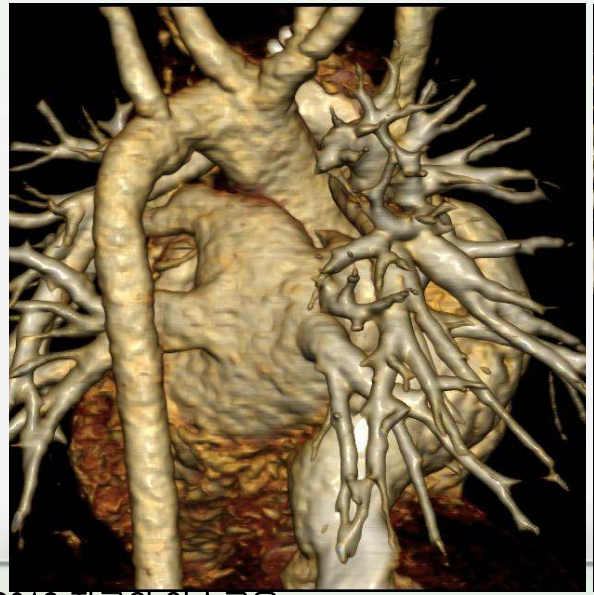
Cases

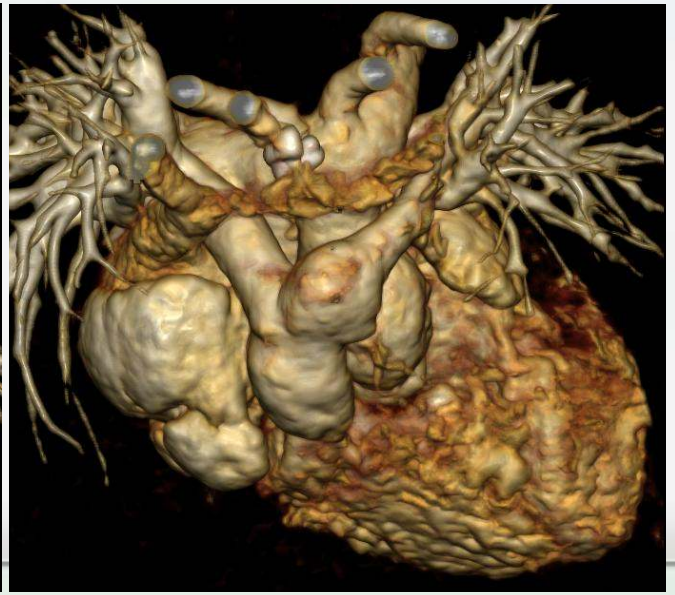






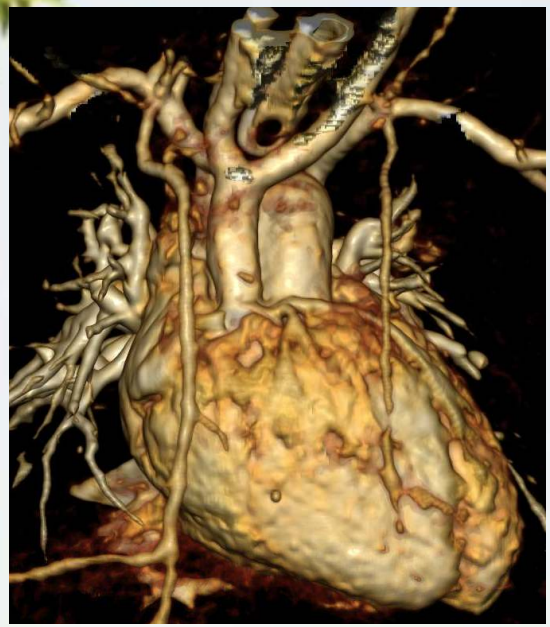
COA +
Taussig
-Bing
anomaly

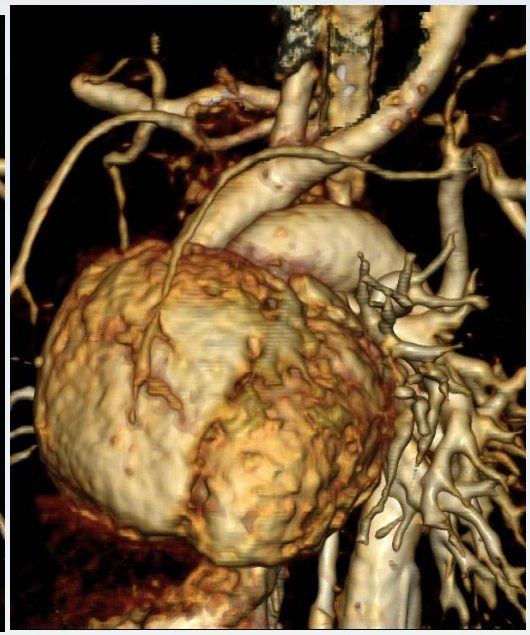




One-stage Repair
(Arterial switch op
+ intraventricular
baffling
+ Arch repair)

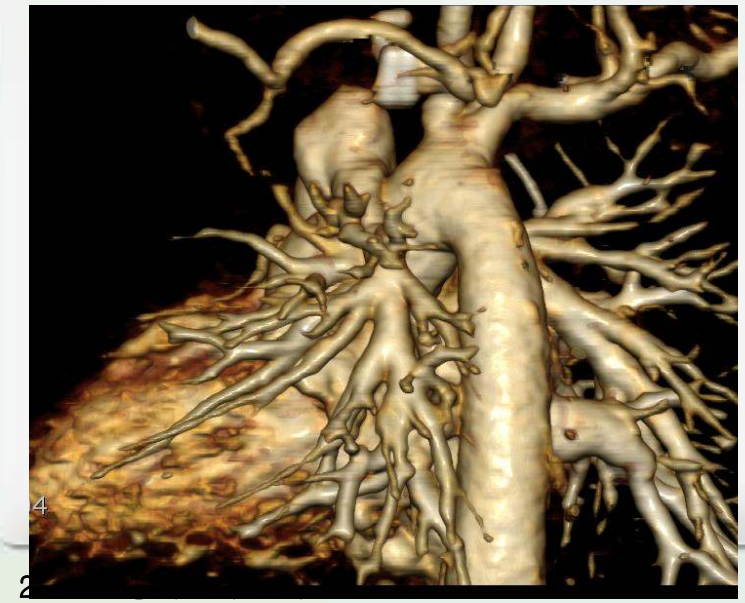
Cases

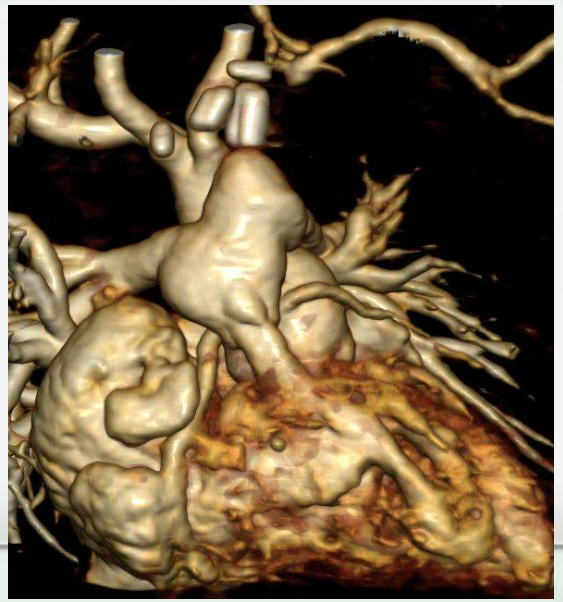




Type B IAA +
Taussig-Bing anomaly
(DORV with subpulmonary VSD)

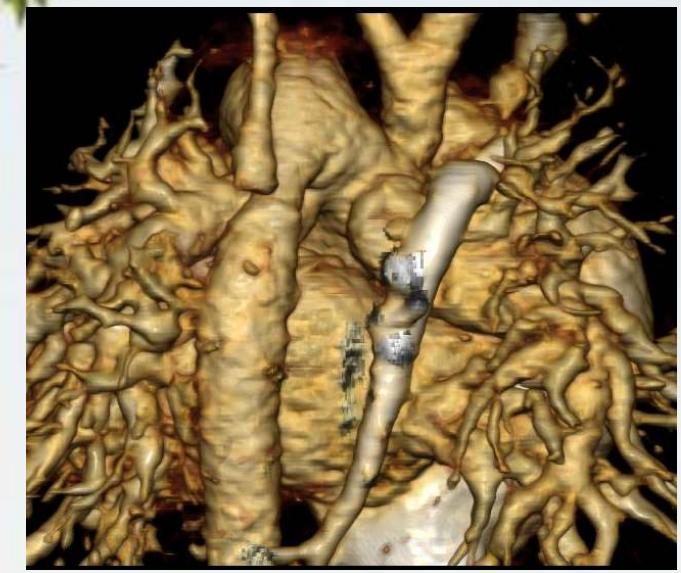
One-stage Repair
(Arterial switch operation
+ intraventricular baffling
+ Arch repair)

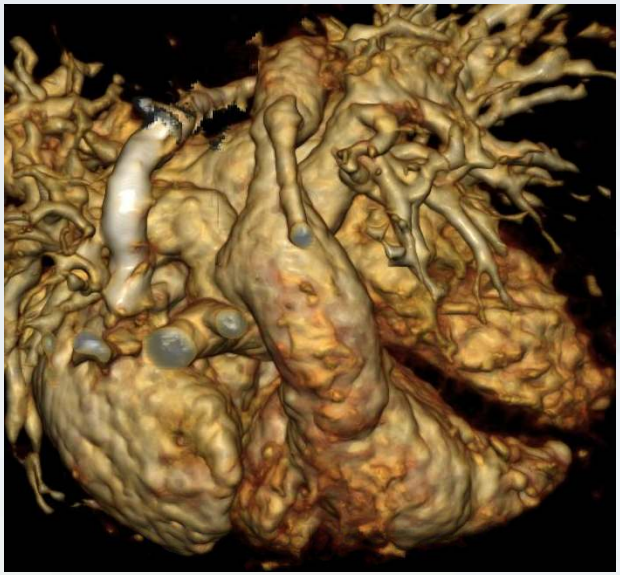


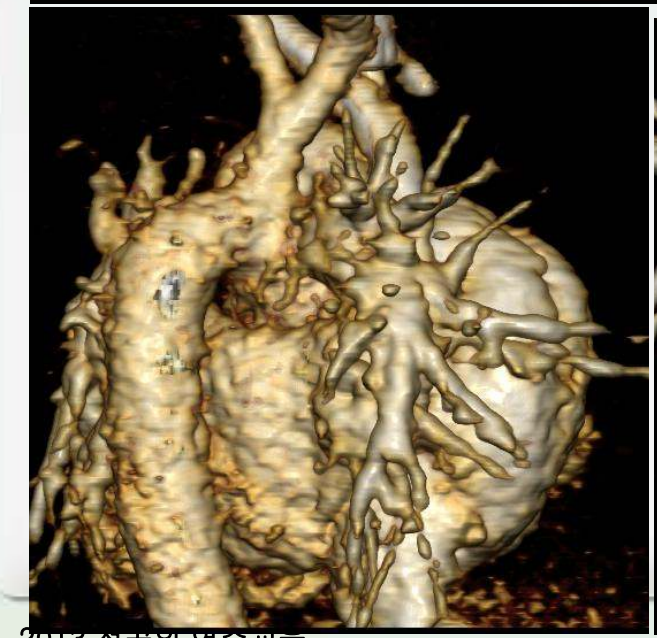


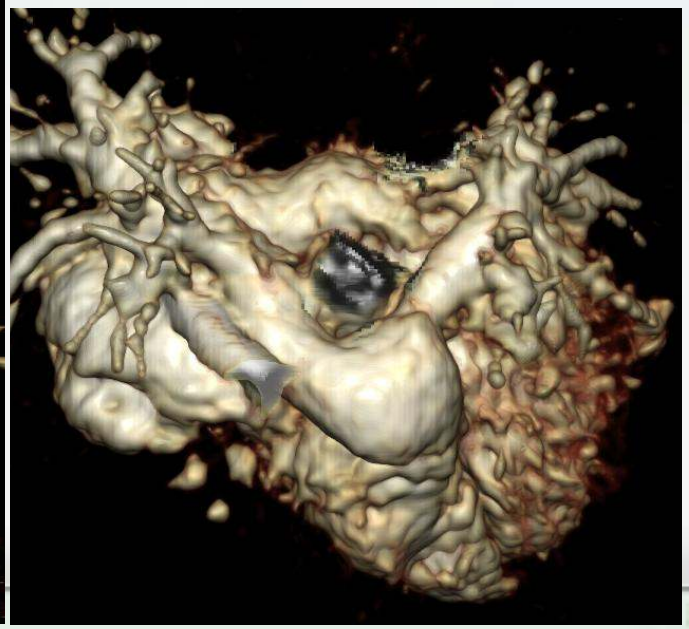
Postop.

Cases







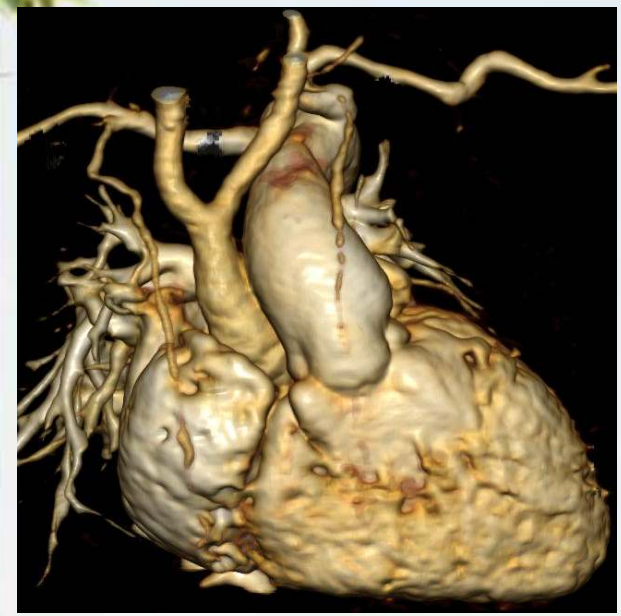


Type B IAA
+ AP window
+ RPA from ascending aorta

One-stage total Correction +
RPA anterior translocation

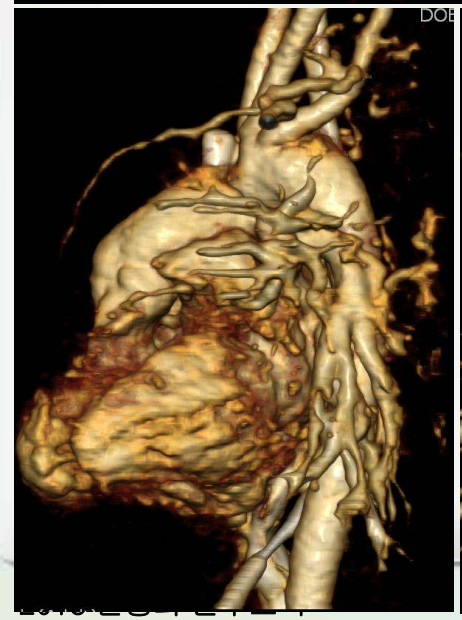
Postop.

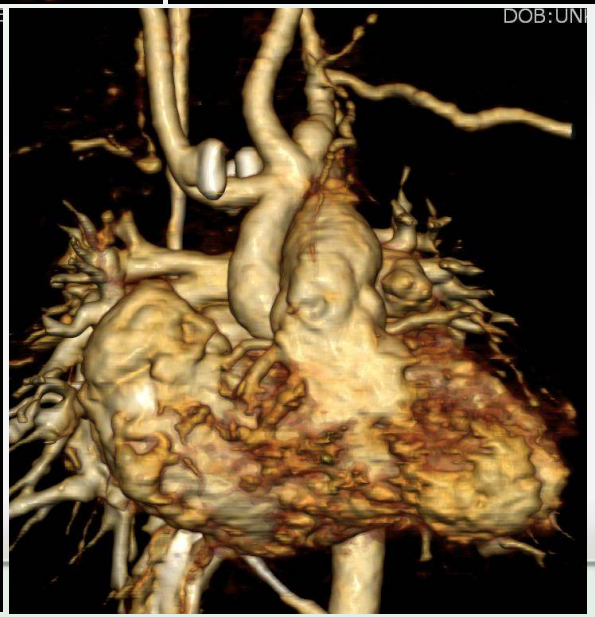
Cases



Type B IAA
+ VSD
+ Aberrant RSCA

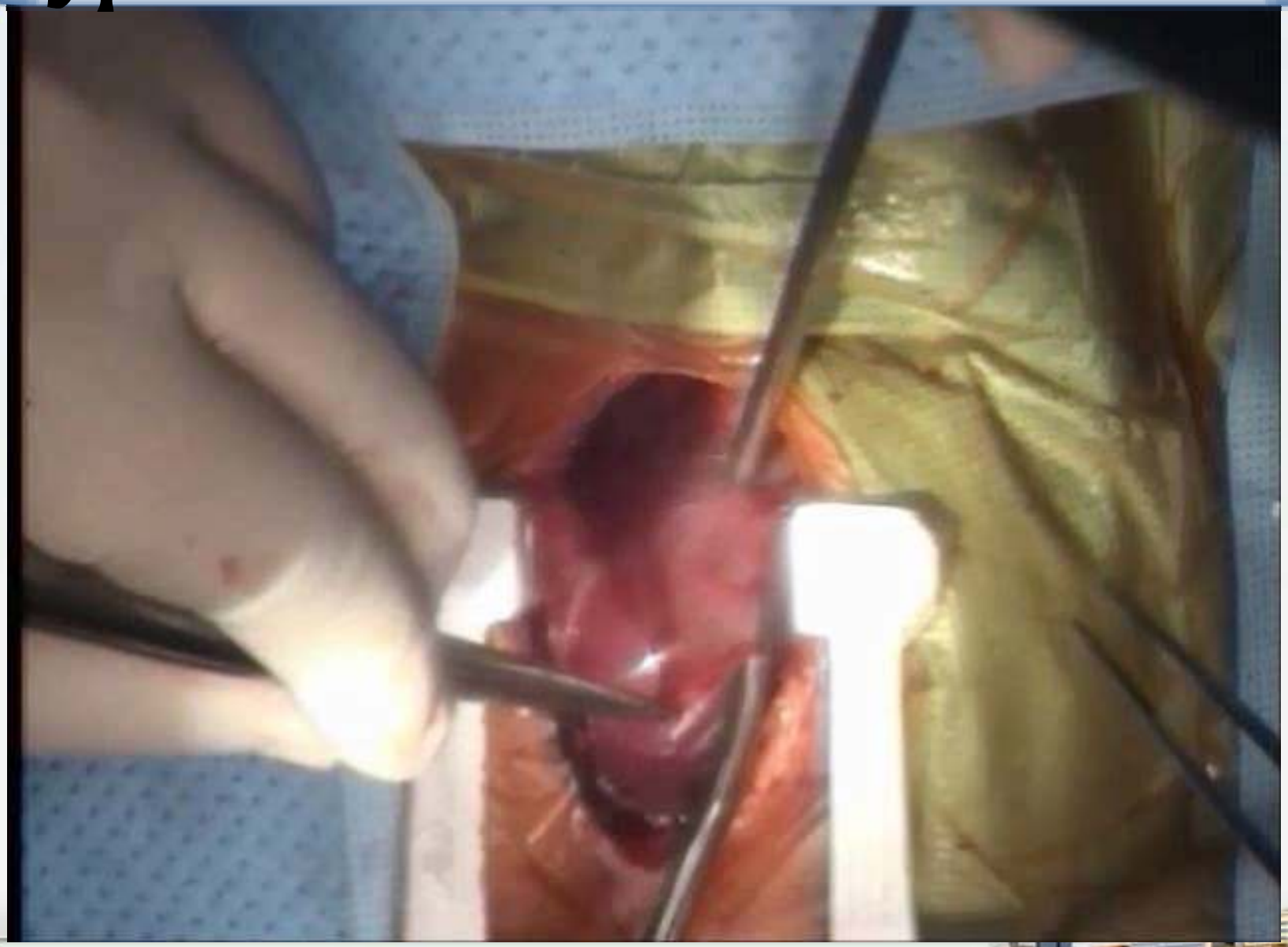
One-stage Repair



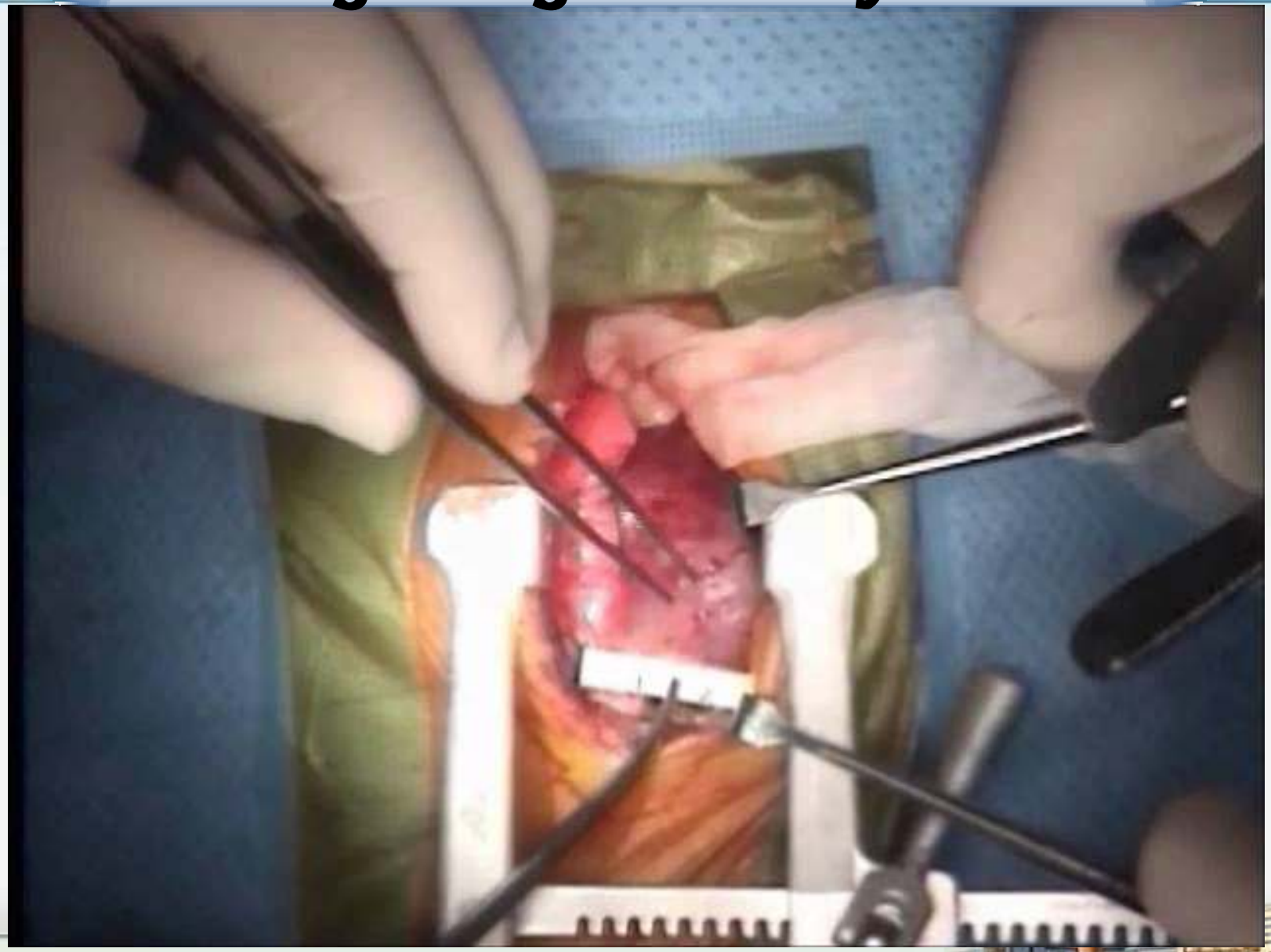


Postop.

Type A IAA + VSD + Aberrant RSCA



Taussig-Bing anomaly + COA



Morphology and Morphogenesis

Coronary Arteries

- In the intimal layer; degenerative and proliferative changes of the elastic fibers and excess collagenous tissue
- The media thickens to about twice normal with a rich elastic fiber network and often hyaline changes
- As a result of prolonged hypertension, arteriosclerotic changes are apt to occur more often and at a younger age