

Extended End To End Repair of Coarctation with Arch Hypoplasia: Single Surgeon Experience

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Incidence

- Represents 5% to 8% of all congenital cardiovascular lesions.
- It has been quoted as the fifth to eighth most common form of congenital heart disease.
- It occurs 2 to 5 times more frequently in males than females.

Associated Lesions

- Eighty-two percent of coarctations occur as isolated lesions.
- Eleven percent are associated with ventricular septal defects,
- and 7% have major associated lesions.
- PDA and PFO are so prevalent that they are not considered accompanying defects.
- A bicuspid valve is common with coarctation of the aorta with an incidence of 27% to 46%.
- Two syndromes are associated with coarctation: Turner's syndrome will have coarctation in 15% to 36% of cases, and coarctation is considered a part of Shones complex.

Presentation

- Neonates often present with heart failure, acidosis, and shock with critical coarctation.
- Less severe coarctation often detected during evaluation for hypertension or murmur in the older child or adult
- Diminished or delayed lower extremity pulses and a systolic pressure gradient between the upper and lower extremities are the most useful exam findings.
- Transthoracic echocardiogram is initial test of choice; CT and MRI useful if echocardiogram inconclusive and for surgical planning.

Surgical classification of coarctation

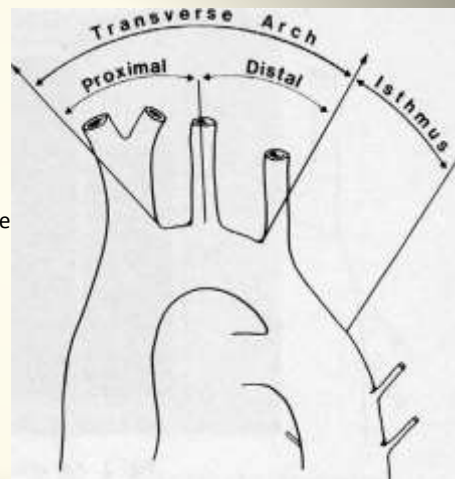
- (A) Type I, coarctation with or without patent ductus arteriosus with no other cardiac defect;
 (B) Type II, coarctation with isthmus hypoplasia with or without patent ductus arteriosus;
 (C) Type III, coarctation with tubular hypoplasia involving isthmus and segment of the arch.



Anatomy of aortic arch

Anatomy The aortic arch is divided into three parts:

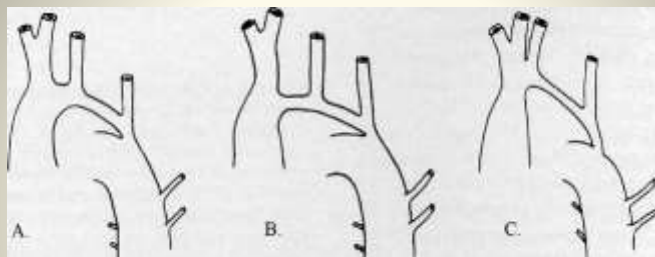
- The proximal transverse arch .
- The distal transverse arch .
- And the aortic isthmus.



Definition of Arch Hypoplasia

- The incidence of hypoplastic arch of varying severity in patients with coarctation is between 40% and 80%, depending on the definition used
- Because a precise and accepted definition of a hypoplastic aortic arch is unclear, a combination of the three definitions and parameters were found the most predictive.
- A neonatal patient with CoA will undergo enlargement of the aortic arch under the following circumstances:
When the:
 1. Size of the arch in mm is less than the patient's weight in kg.
 2. Aortic arch diameter z-score is more than -2.
 3. Ratio of the diameter of the transverse arch to the descending aorta is less than 50%.
 4. ??? The ratio of the transverse arch to the *ascending aorta* is less than 50% .

Types of transverse arch hypoplasia:



- (A) **hypoplasia of the distal transverse arch;**
 (B) hypoplasia of both the proximal and the distal transverse arches;
 (C) absence of the proximal and long-segment hypoplasia of the distal arch.

Therapeutic Options

- Surgical repair
 - Surgical repair typically preferred over transcatheter approaches in the infant and young child with native coarctation.
- Cath. interventions
 - Had been covered by our colleagues “interventional cardiologists” in their talks in this session.

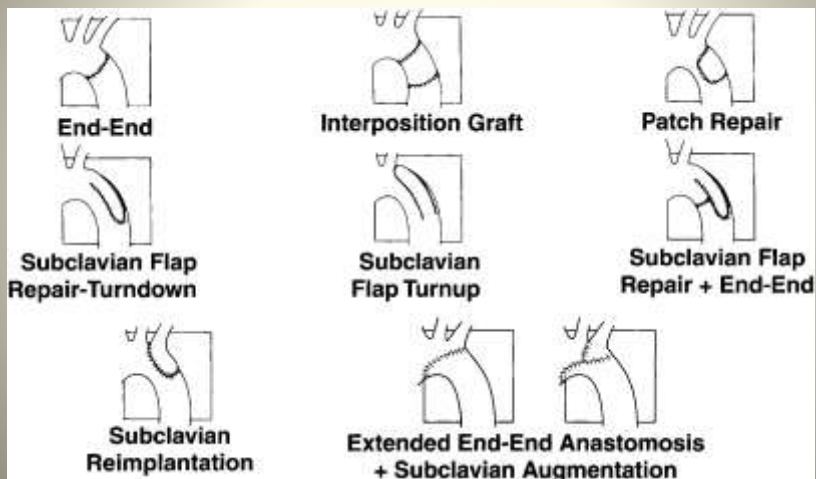
Milestones in Surgery of CoA

Surgical procedure	Author	Year	Country
Simple resection with end to anastomosis	Crawford	1944	Sweden
Interposition graft	Gross	1951	USA
Patch augmentation	Vosschulte	1957	Germany
SC flap aortoplasty	Waldhausen	1966	USA
Resection with EEEA	Amanto	1977	USA

Timing of Surgery

- Elective surgery for coarctation should be performed within the first year of life.
- Two factors justify performing elective repair in infancy:
 - good surgical results that are obtained with modern techniques of repair, especially with the types of extended end to end repairs.
 - The decreased risk of development of persistent hypertension.
 - An older age at the time of repair is a major predictor of late complications, thus surgery should preferably be performed before 1 years of age.

Various methods in the surgeon's armamentarium to repair the various types of coarctation.



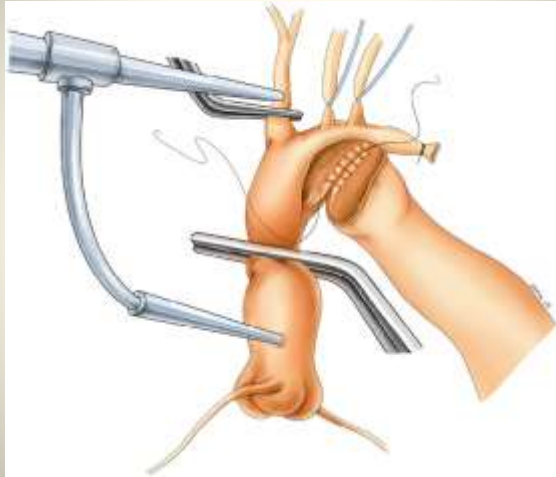


Illustration of coarctation repair with extended end-to-side anastomosis technique.
(AAA)

Complications of surgical Therapy of CoA

- Death.
- Bleeding.
- Paraplegia.
- Early and late recoarctation.
- Chylothorax.
- Nerve injury.

Our Experience

- From December 2005 to December 2017.
- 276 infants (under the age of one year) was diagnosed with CoA/Arch hypoplasia.
- Isolated CoA was present in 215 infants.
- Of those 215, 206 had Extended End to End Anastomosis (EEEE).
- The remaining 9 infants were managed by other surgical Techniques (4 by AAA, 3 SC flap and 2 by Vosschaulte technique).

Surgical Technique

- General anesthesia.
- Right radial arterial monitoring line.
- A femoral arterial line as well.
- Pulse oximetry in right hand and the right ear lobule.
- CVP line.
- We cool the room temperature to have the infant core around 34 degrees for spinal cord protection.

Surgical Technique

- The chest is entered through the 3rd IC space via a left posterolateral thoracotomy..
- The is retracted anteroinferiorly.
- Dissection of the mediastinal pleura, then dissection of the aortic arch till the origin of Rt, Brachiocephalic artery and upper portion of the descending aorta as far down as possible. We do not sacrifice any aortic intercostals.
- The 3 arch vessels are dissected as high as possible.

- After complete mobilization, Heparin is given and aortic clamps are applied.
- It is very crucial that the clamp on the arch should be as close as possible to ascending aorta on a portion on brachiocephalic artery as guided by the RT. Radial line and pulse oximetry on the Rt. ear lobule to make sure that the brain is perfused.
- Counter incisions are made on the under surface of the arch and posterior aspect of the descending aorta.
- Anastomosis is begun using 6/0 prolene suture in a continuous fashion.



Results

- 150 were males and 50 females.
- Age ranged from one week to 11 months (mean of 4 months).
- Weight ranged from 3 to 8.5 Kg. (mean of 4.5).
- Aortic clamp time ranged from 12 to 25 minutes (mean of 15 minutes).

Results

- Early mortality: 3
 - On table death in one because of acute severe LV failure.
 - Another one second PO day because of cerebral bleeding.
 - The third one on the 7th PO day because of sepsis.
- Late mortality one documented late death during surgery for Ross 6 years after CoA surgery.

Results

- Take back to OR for bleeding in one infant.
- Six cases of chylothorax. It our policy to do extensive dissection to mobilize the arch and descending aorta for better anastomosis.
- One case of paraplegia, resolved partially after 6 months and recovered completely after one year of physiotherapy.

Results

- Recoarctation:
- Defined as a peak gradient of more than 20 mmHg.
- Occurred in 5 patients (2.4%).
- All recoarctation were managed by cath intervention except one patient managed surgically through mediastinotomy.

Conclusion

- EEEA is a very simple, effective surgical technique for managing infants with CoA /arch hypoplasia.
- Associated with low mortality and morbidity.
- The recurrence rate is very low.
- No need for DHCA or selective cerebral perfusion that is required for anterior mediastinotomy approach.