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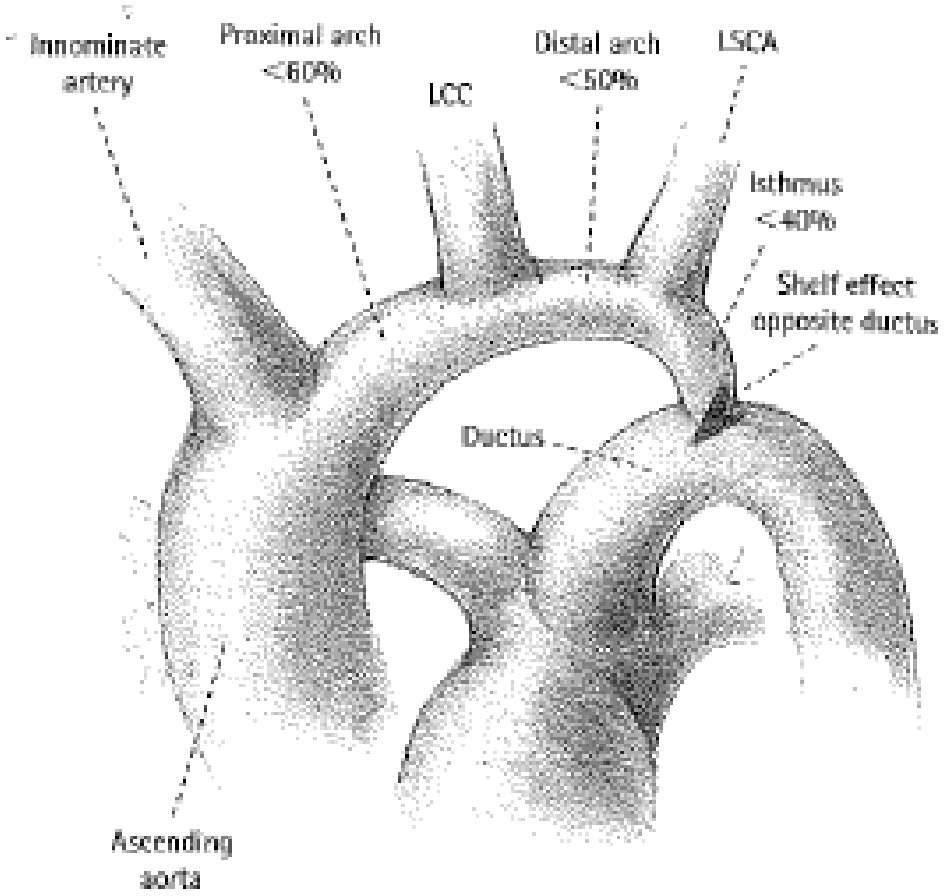
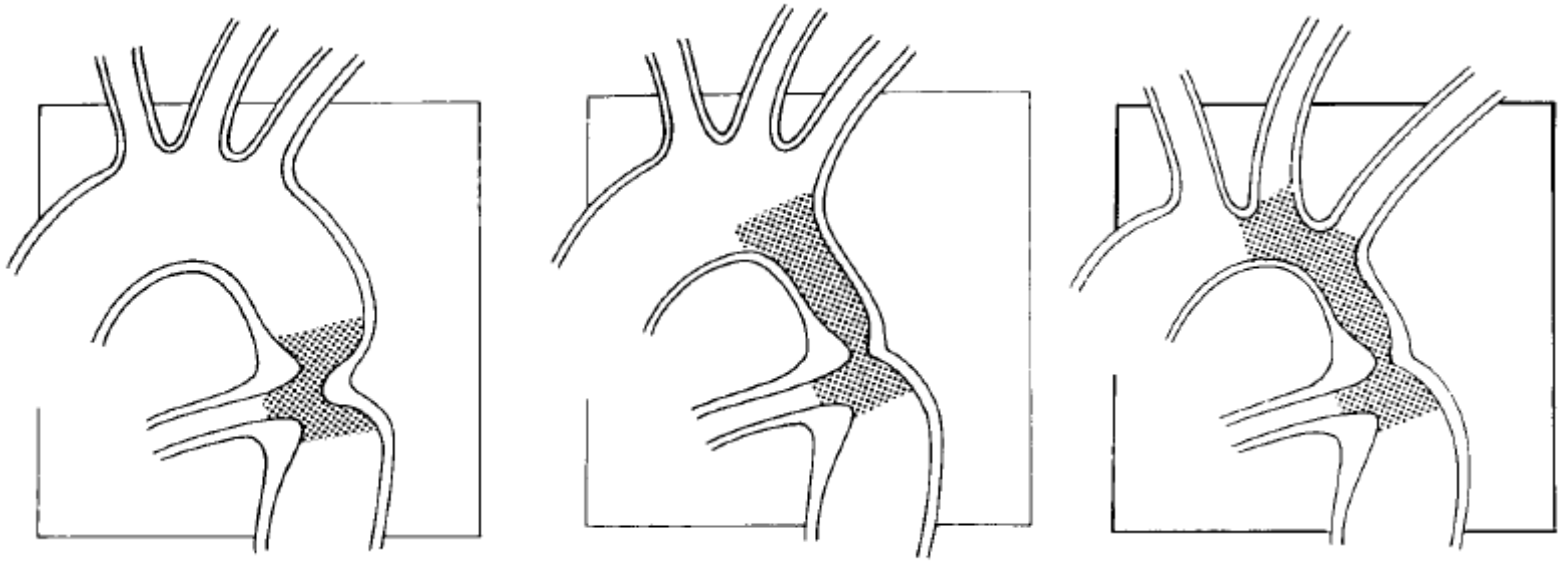
Hypoplastic arch: When and How to intervene

Aortic dimensions in coarctation



Extent of coarctation/hypoplasia



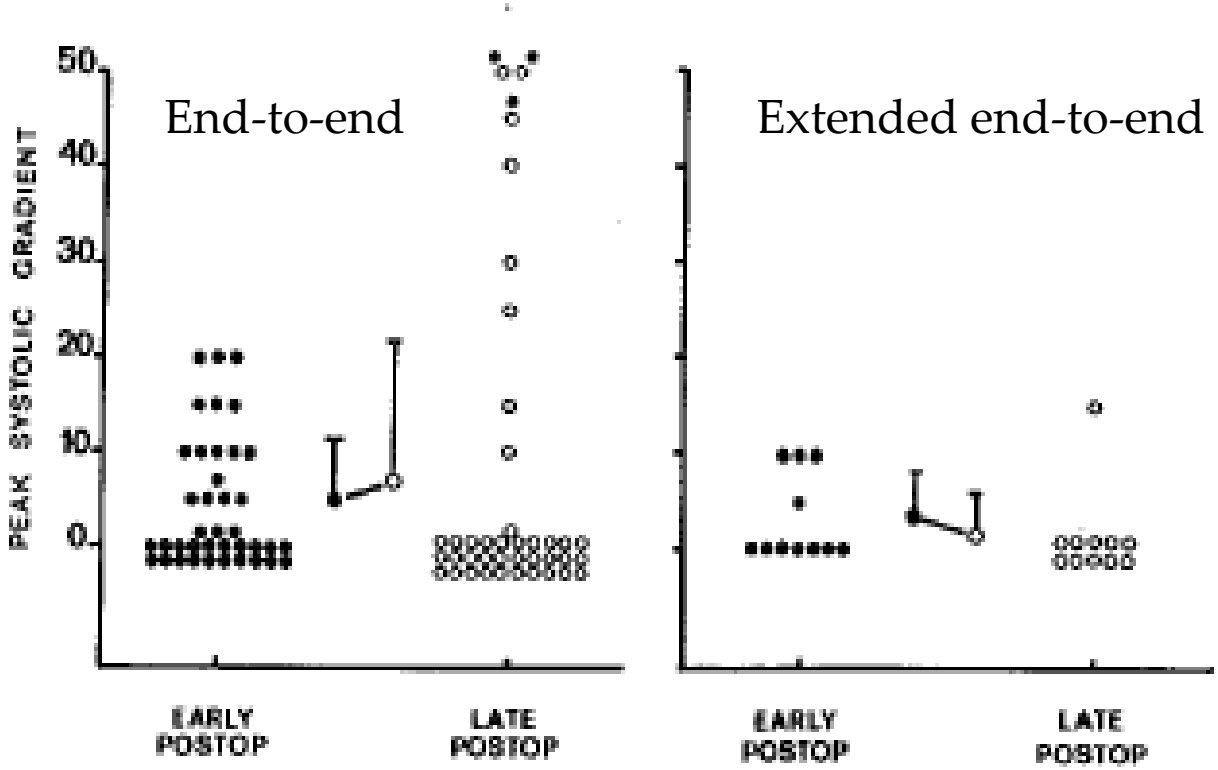
Juxta-ductal coarctation

Narrow isthmus

Arch hypoplasia

Amato et al. Ann Thorac Surg 1991;52:615-20

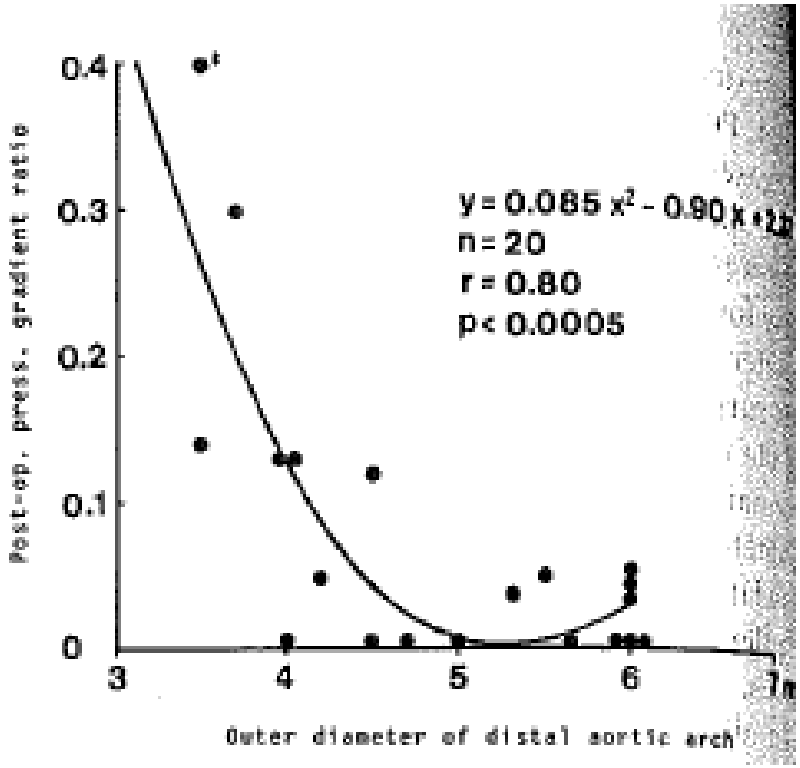
Hypoplastic arch? – 50% rule

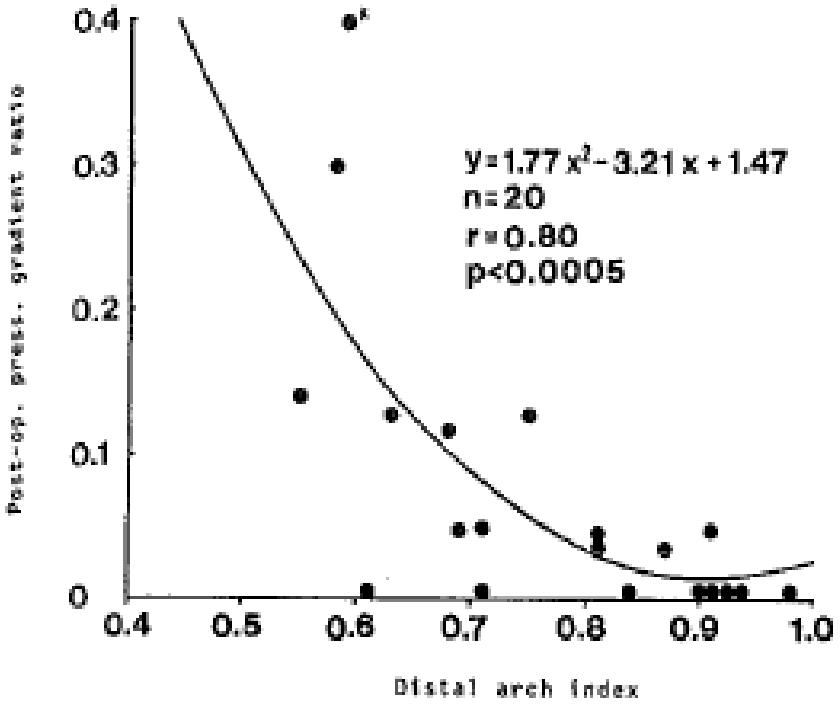


Increased gradient after end-to-end repair when arch <50% ascending aorta size

Vouhe et al. J Thorac Cardiovasc Surg 1988;96:557-63

Hypoplastic arch? – 3.9mm rule





Increased gradient ratio correlates with distal arch size < 3.9 mm or arch index < 0.63

Qu et al. J Cardiovasc Surg 1990;31:796-800

Hypoplastic arch? – more rules

- Some degree of arch hypoplasia present in all coarctations
- Transverse arch diameter < body wt. (kg.) + 1
  (Karl et al. J Thorac Cardiovasc Surg 1992;104:688–95)
- Transverse arch diameter < Z -2 for BSA
  (Brouwer et al. J Thorac Cardiovasc Surg 1992;104:426–33)
- Distal transverse arch < diameter of L carotid
  (Swartz et al. Congenit Heart Dis 2011;6:583-91)

Incidence of hypoplastic arch

Distribution of arch anatomy in 151 patients

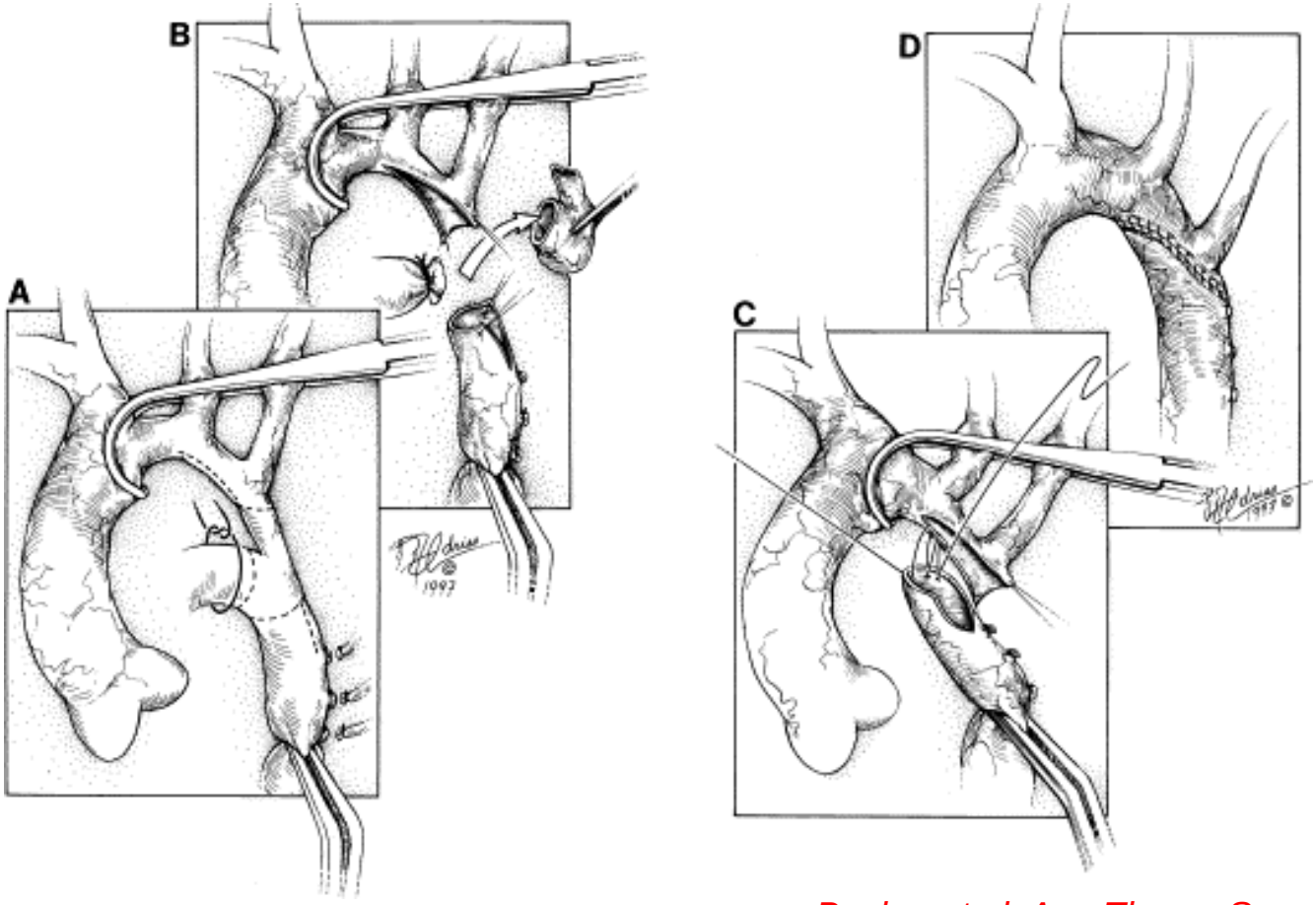
Associated defect	Normal Arch No. (%)	Hypoplastic isthmus No. (%)	Hypoplastic transverse arch No. (%)	Total No. (%)
Simple coarctation	42 (58.3)	14 (19.4)	16 (22.2)	72 (100)
Coarctation with VSD	10 (22.7)	17 (38.6)	17 (38.6)	44 (100)
Coarctation with major heart defect	12 (34.3)	6 (17.1)	17 (48.6)	35 (100)
Total	64 (42.4)	37 (24.5)	50 (33.1)	151 (100)

van Heurn et al. J Thorac Cardiovasc Surg 1994;107:74-86

Repair options

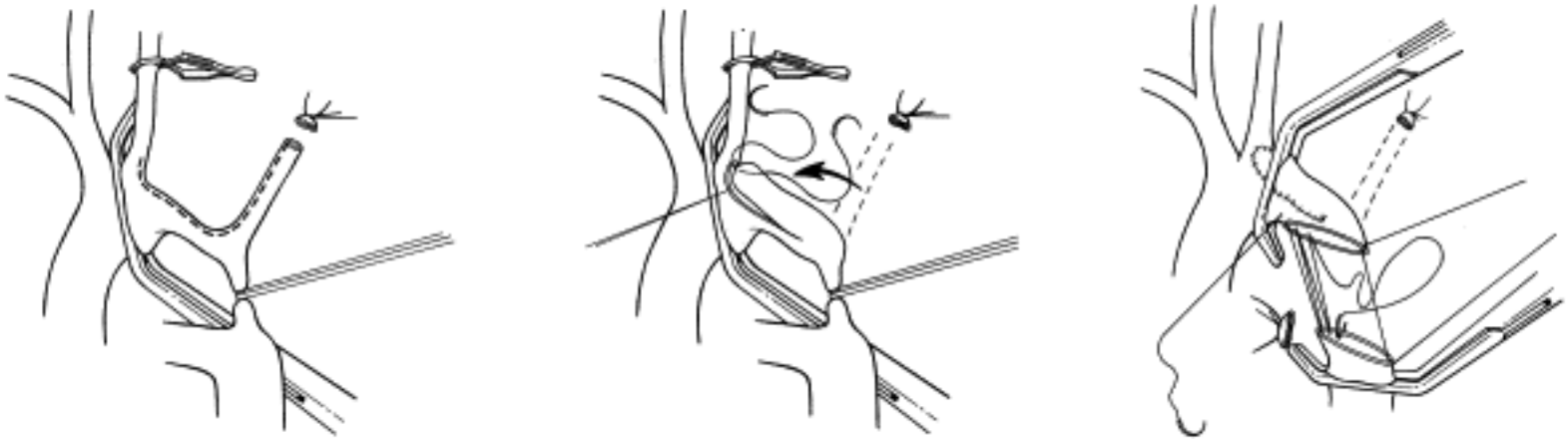
- Thoracotomy vs. sternotomy
- Definite indications for sternotomy
  - VSD needs closure
  - Aortic valve intervention
  - Need for bypass for safe proximal cross clamp (Bovine arch)
  - Other cardiac interventions
- Hypoplastic arch?

Thoracotomy – Radical resection, end-end



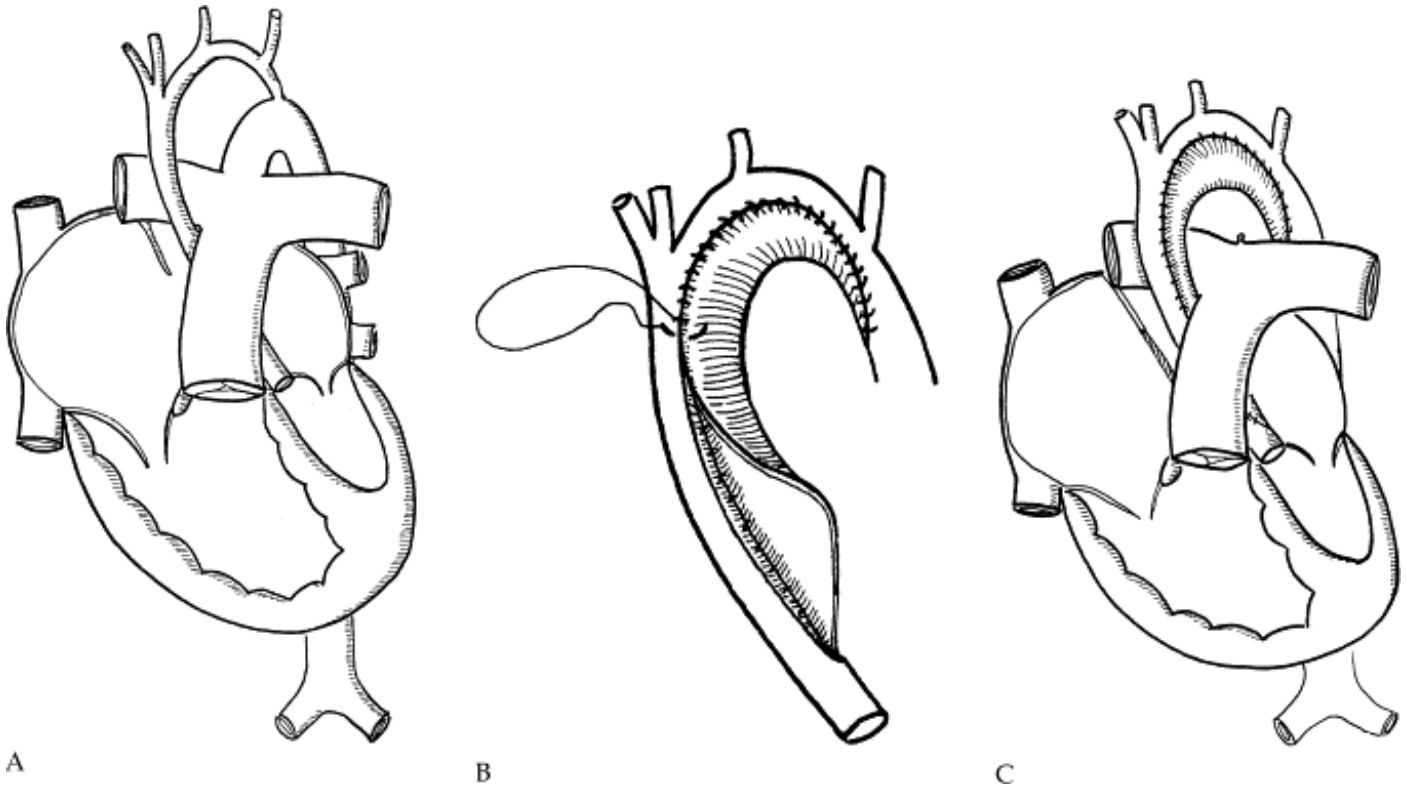
Backer et al. Ann Thorac Surg 1998;66:1365-70

Thoracotomy – Reverse subclavian flap



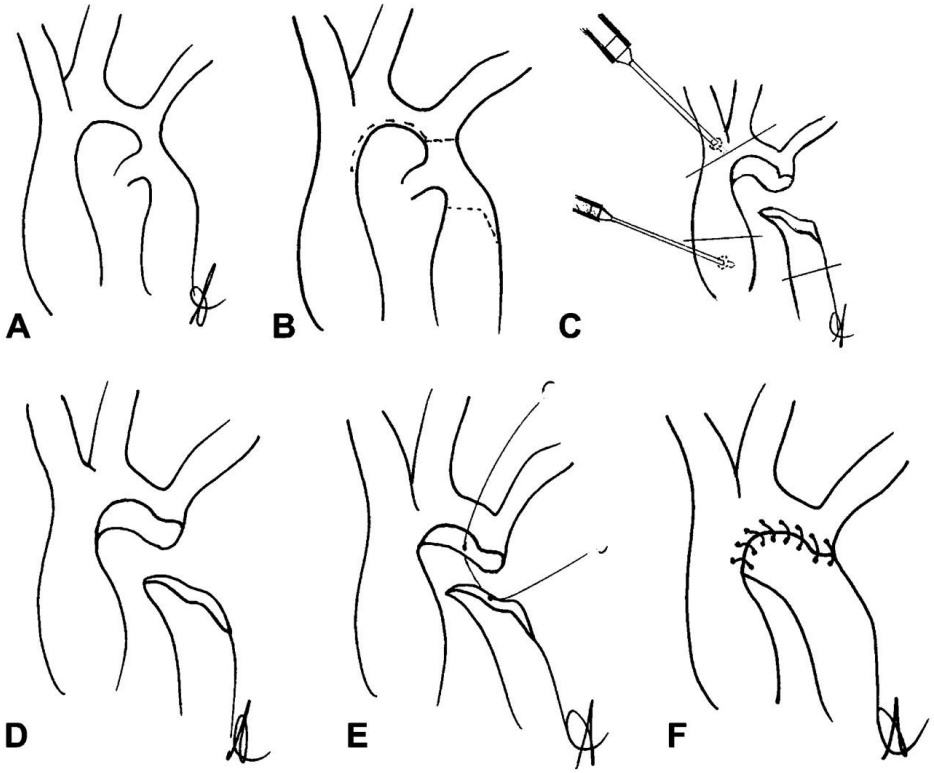
Kanter et al. Ann Thorac Surg 2001;71:1530-6

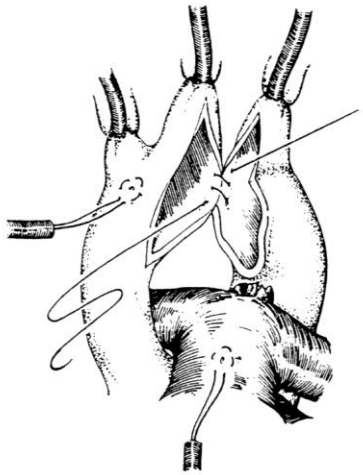
Sternotomy – On lay patch augmentation

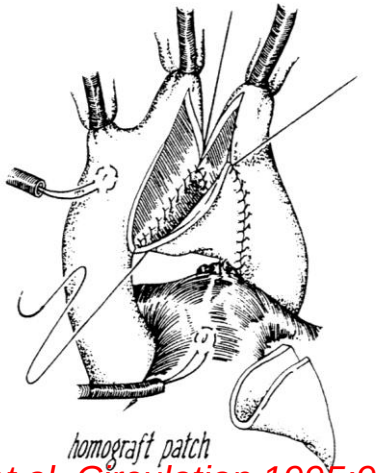


Tchervenkov et al. Ann Thorac Surg 1998;66:1350-6

Sternotomy – Resection and reconstruction

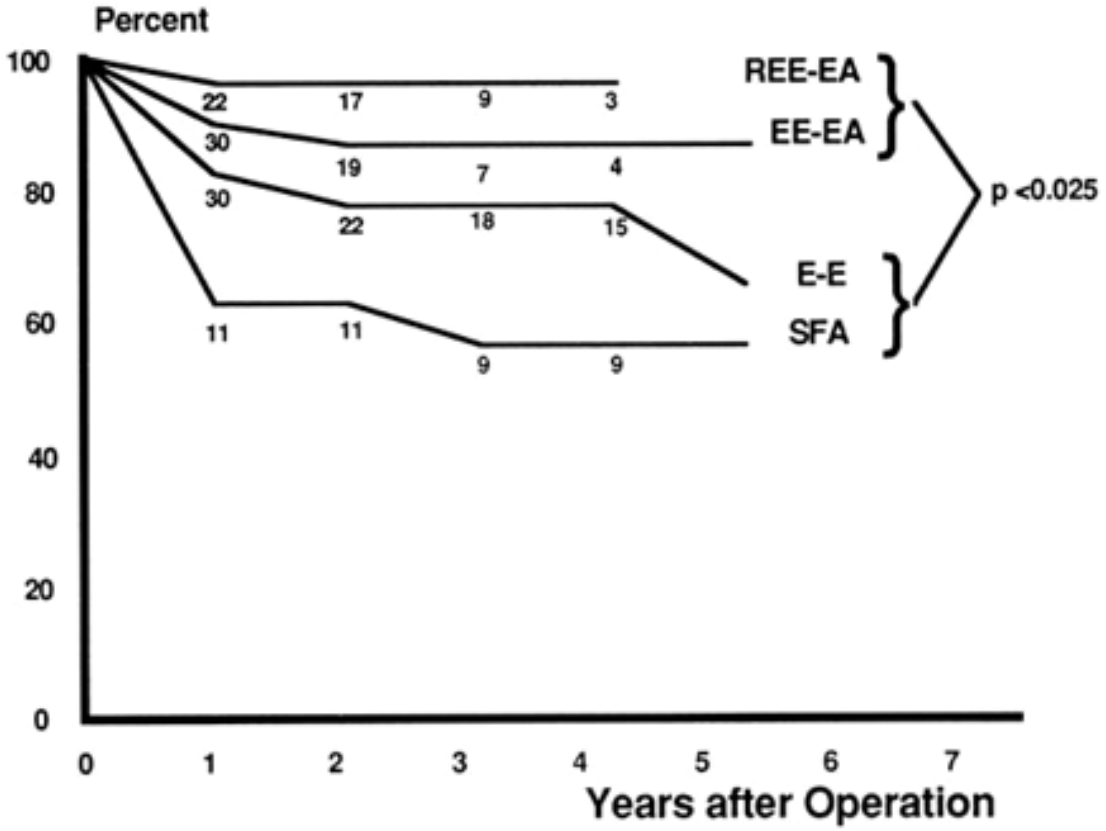






Jacobs et al. Circulation 1995;92:128-31

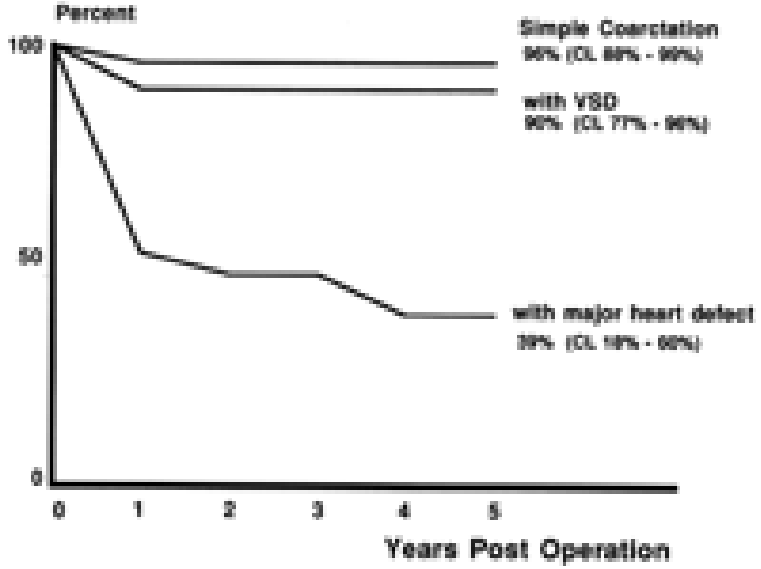
Long-term outcome after arch repair



REE-EA – Radically extended end-end
EE-EA – extended end-end
E-E – end-end
SFA – subclavian flap angioplasty

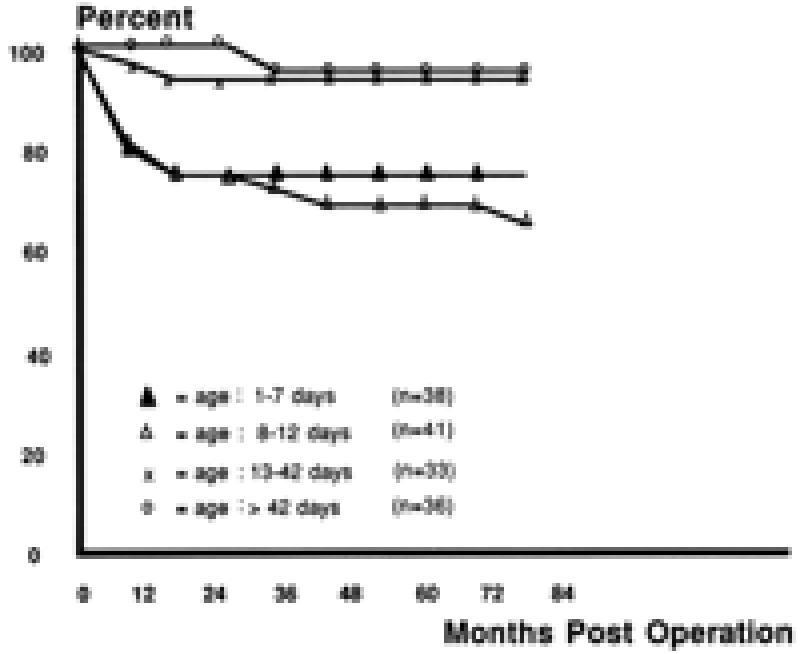
van Heurn et al. J Thorac Cardiovasc Surg 1994;107:74-86

Prognosis following arch repair



Associated major heart defect impacts survival

Lower age at repair predicts re-coarctation



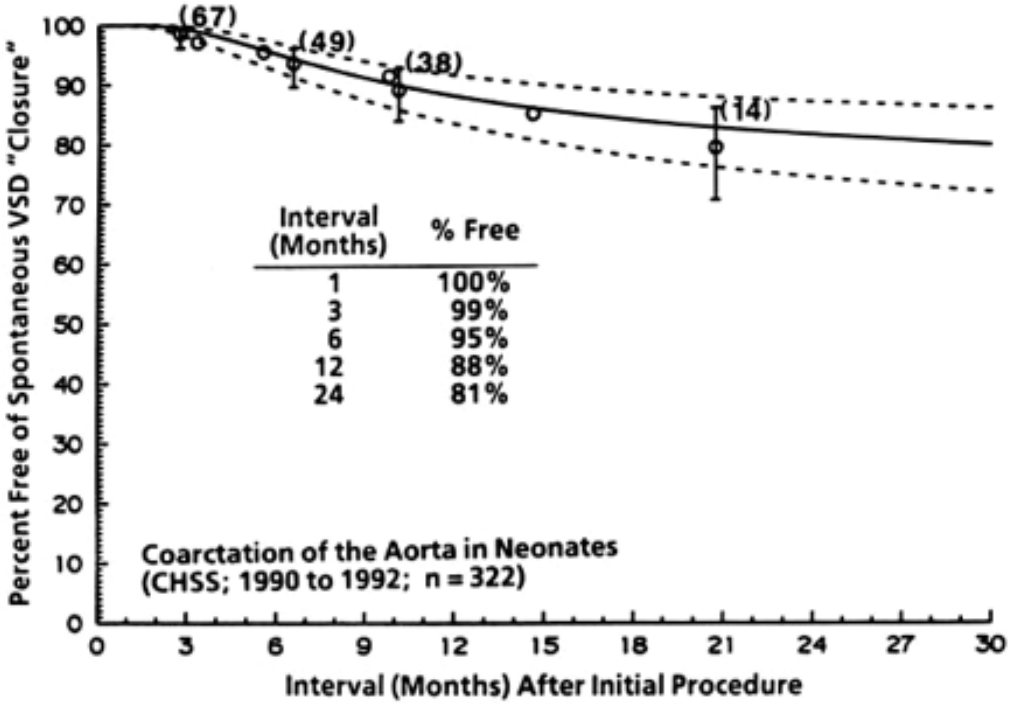
van Heurn et al. J Thorac Cardiovasc Surg 1994;107:74-86

Associated VSD

- VSDs associated with about 30% arch abnormalities
- Can be muscular, outflow posterior mal-aligned, peri-membranous or multiple
- Options for treatment
  - Repair arch via thoracotomy ± PA band
  - Repair arch via sternotomy ± PA band
  - Repair arch and close VSD – one-stage repair

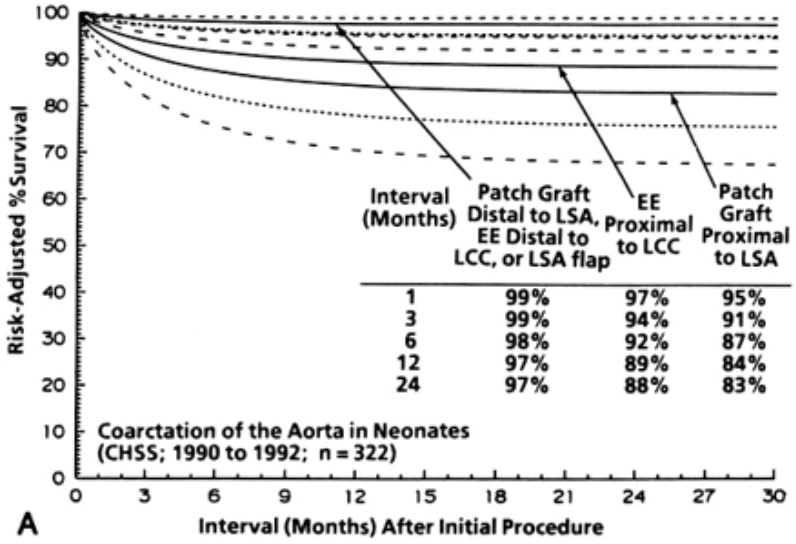
VSD rarely close spontaneously

- 19% VSDs close within 24 months
- Muscular VSDs most likely to close
- Outflow VSDs least likely to close



Quaegebeur et al. J Thorac Cardiovasc Surg 1994;108:841-54

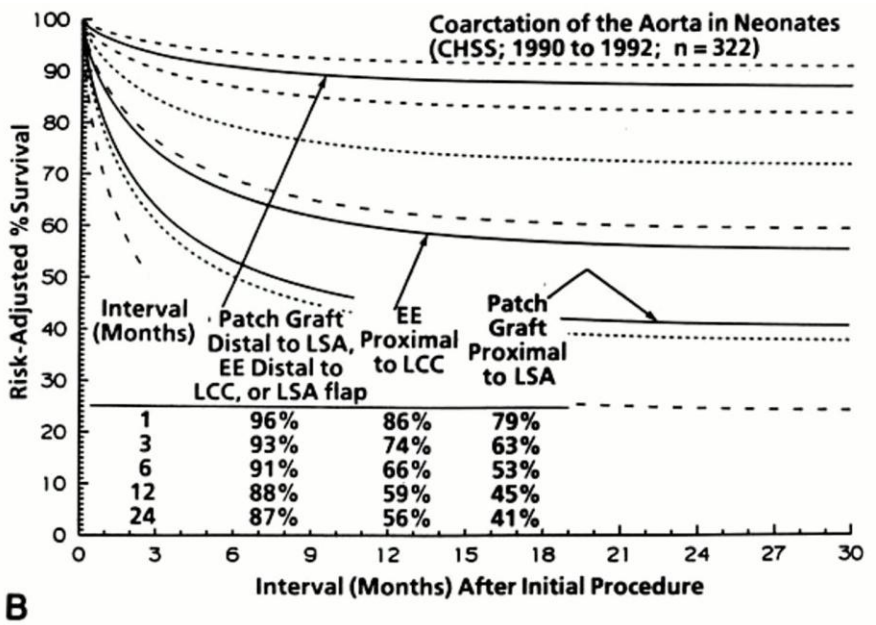
Arch strategy (not VSD) determines survival



Arch repair + PA banding

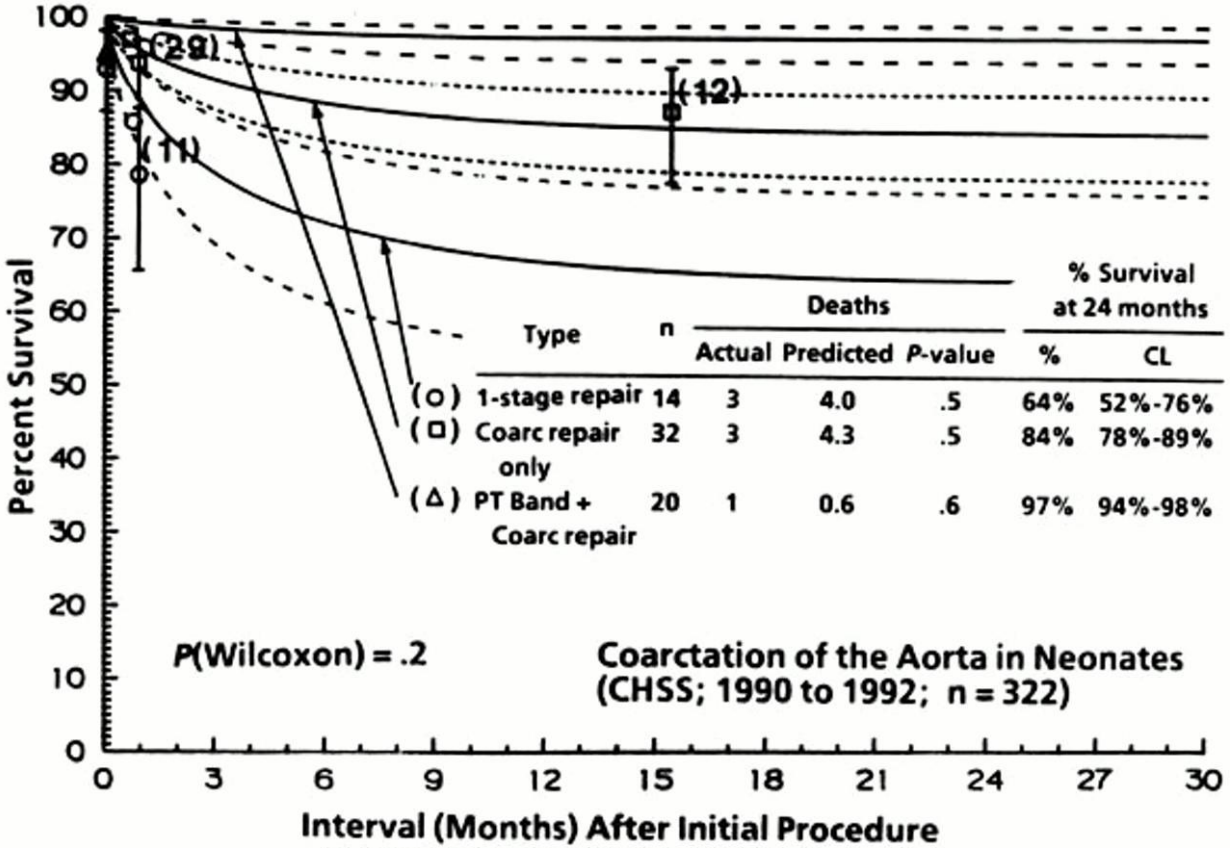
Need to carry repair proximal to L carotid increases mortality

Arch repair alone or with VSD closure



Quaegebeur et al. J Thorac Cardiovasc Surg 1994;108:841-54

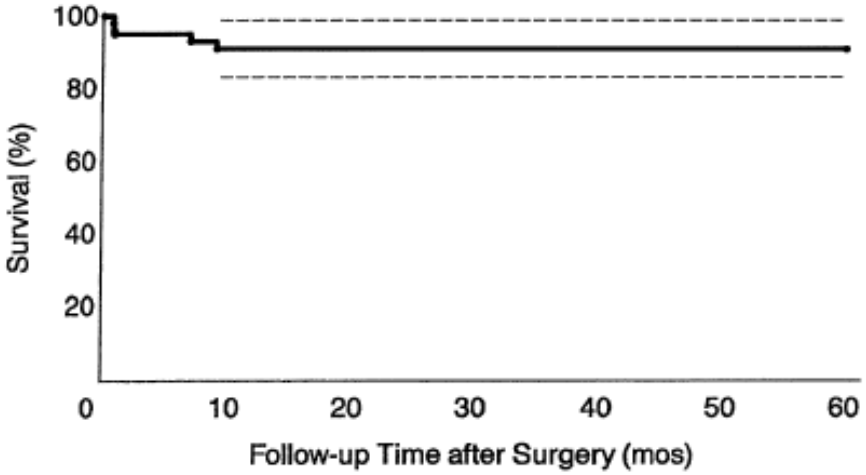
One-stage repair is safe



No significant difference in survival with different approaches

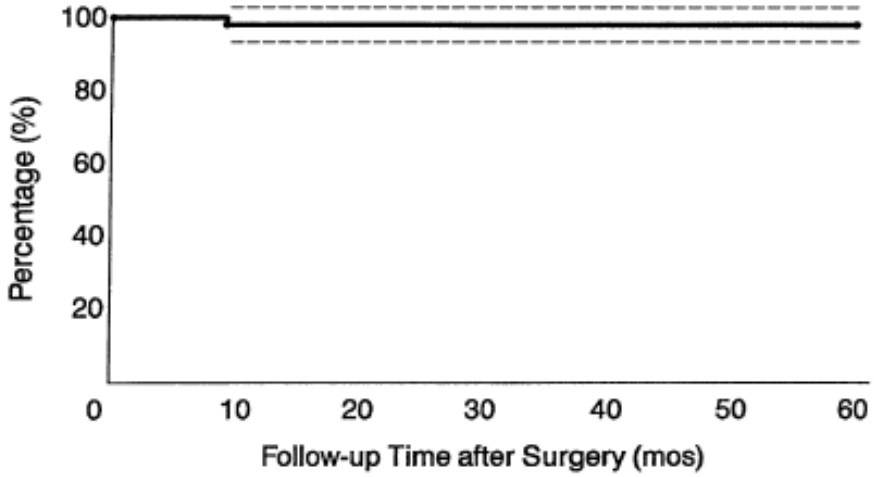
Quaegebeur et al. J Thorac Cardiovasc Surg 1994;108:841-54

One-stage repair is durable



Survival

Re-coarctation



Elgamal et al. Ann Thorac Surg 2002;73:1267-73

Late post-operative death

TABLE 3 Causes of Late Postoperative Death

Cause of Late Death	No.
Coronary artery disease	10
Second cardiac operation	7
Aortic dissection	6
Sudden unexplained	7
Automobile accident	3
Other*	7
Unknown	5

*Suicide, endocarditis, cardiomyopathy, alcohol, cancer, autoimmune deficiency syndrome; and prostate surgery.

Toro-Salazar et al. Am J Cardiol 2002;89:541-7

Hypoplastic arch management – CHLA experience

- Jan 2001 – June 2012 – 190 arch repairs
- 104 (55%) repaired via sternotomy – on lay patch, or resection and reconstruction
- 86 (45%) repaired via thoracotomy - extended end-to-end anastomosis for narrow isthmus/distal arch
- 78 (41%) had associated VSD
  - 41 (53%) underwent one-stage repair
  - 11 (14%) multiple muscular VSDs managed with PA band
  - 26 (33%) isolated small muscular VSDs left alone

In summary…

- Need clarity in pre-op decision of sternotomy vs. thoracotomy – sternotomy preferred if any question
- Aggressive approach to arch augmentation
  - Resect all ductal tissue
  - Autologous tissue or on lay homograft patch
  - Carry augmentation from distal arch to ascending aorta
  - Particularly in setting of bicuspid aortic valve
- One-stage repair preferred
- Arch management (not VSD) key to long-term survival