

NATIVE COARCTATION OF AORTA: IS SURGERY STILL THE GOLD STANDARD?

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CONFLICT OF INTEREST

- NONE RELATED TO THIS PRESENTATION

OBJECTIVES

- Review the clinical history, pathophysiology, investigations and management of native coarctation
- Identify the indications for surgical and catheter based interventions.
- Discuss the pros and cons of surgical and catheter based treatment strategies.

DEFINITION

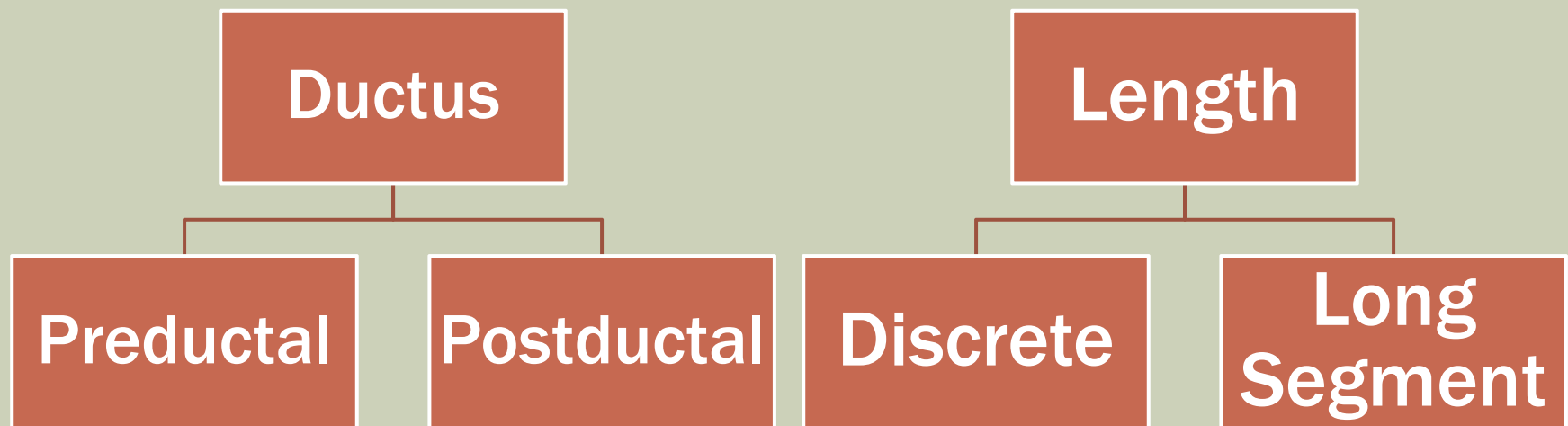
- Constricted aortic segment that comprises localized medial thickening, with some infolding of the medial and superimposed neointimal tissue.
- Shelf-like structure with an eccentric opening
- Membranous curtain-like structure with a central or eccentric opening

INCIDENCE & PREVALENCE

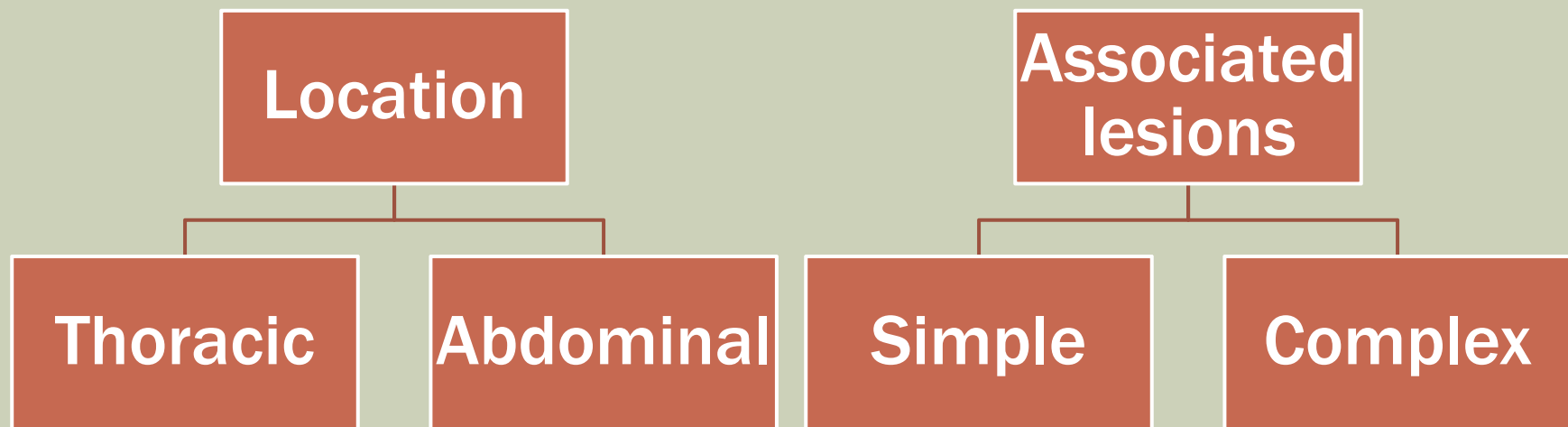
- 5-10% of cases of congenital heart disease.
- Birth prevalence in 1980-1994 was 0.32 case per 1000 live births.
- Usually isolated
- Males (59%)

Grech V. Diagnostic and surgical trends, and epidemiology of coarctation of the aorta in a population-based study. *Int J Cardiol.* Feb 28 1999;68(2):197-202

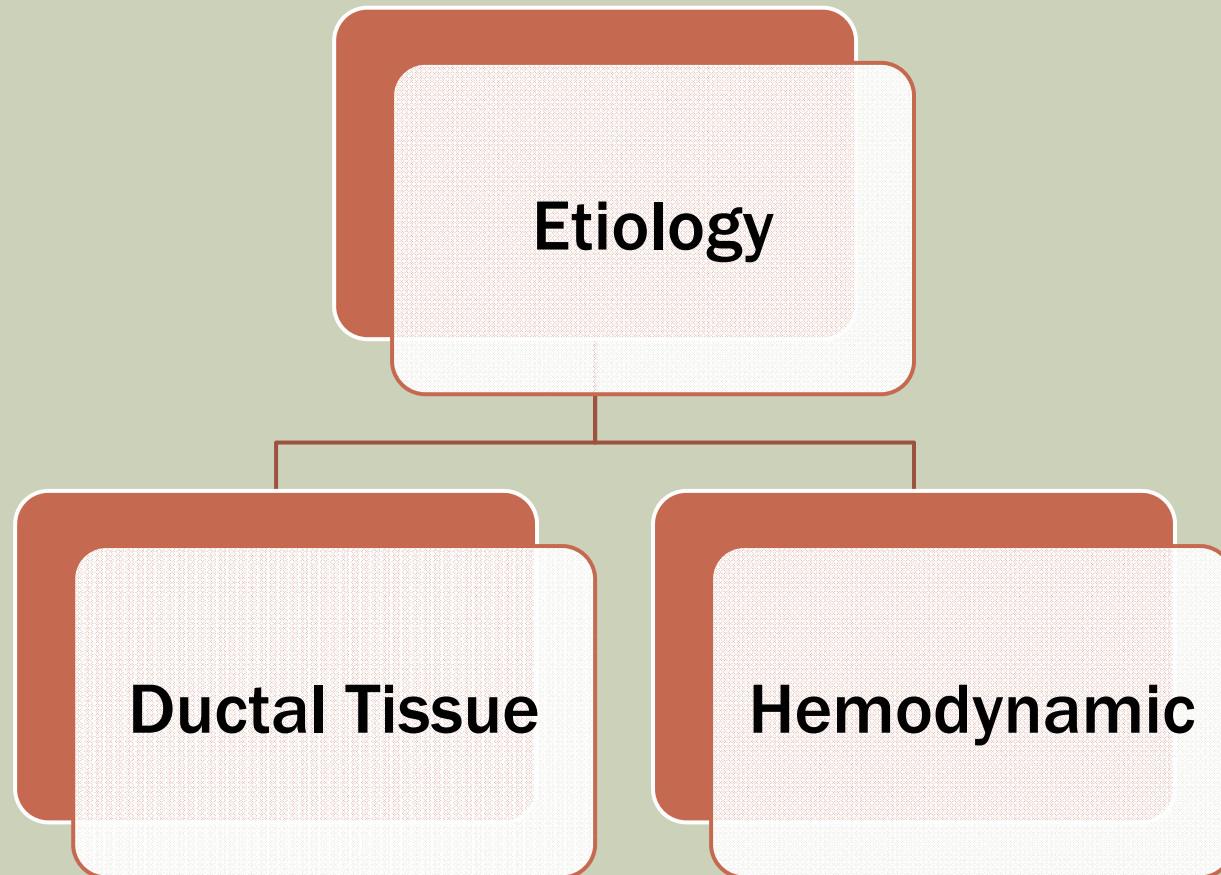
CLASSIFICATION



CLASSIFICATION (CONT'D)



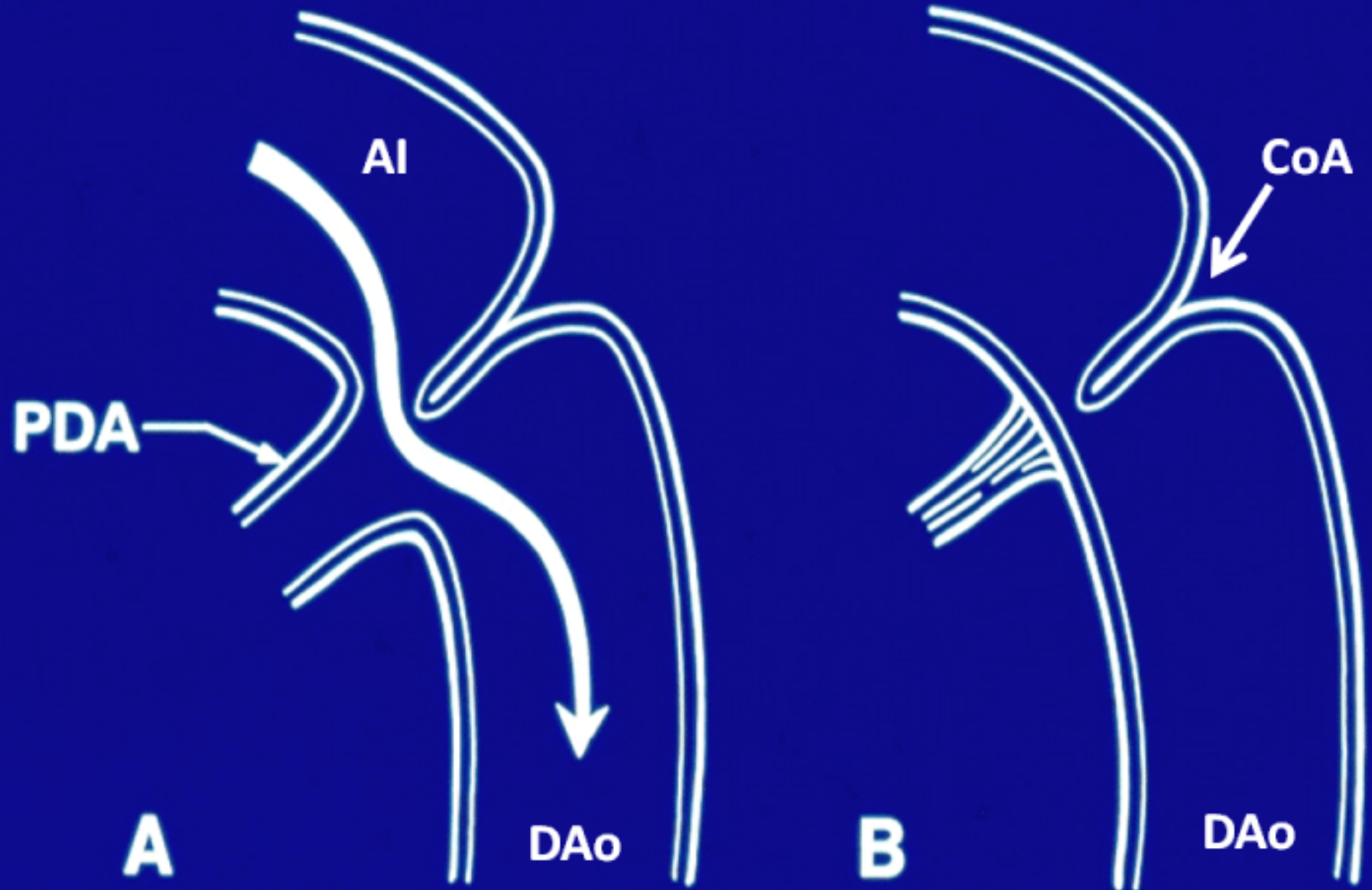
TWO THEORIES



DUCTAL TISSUE THEORY

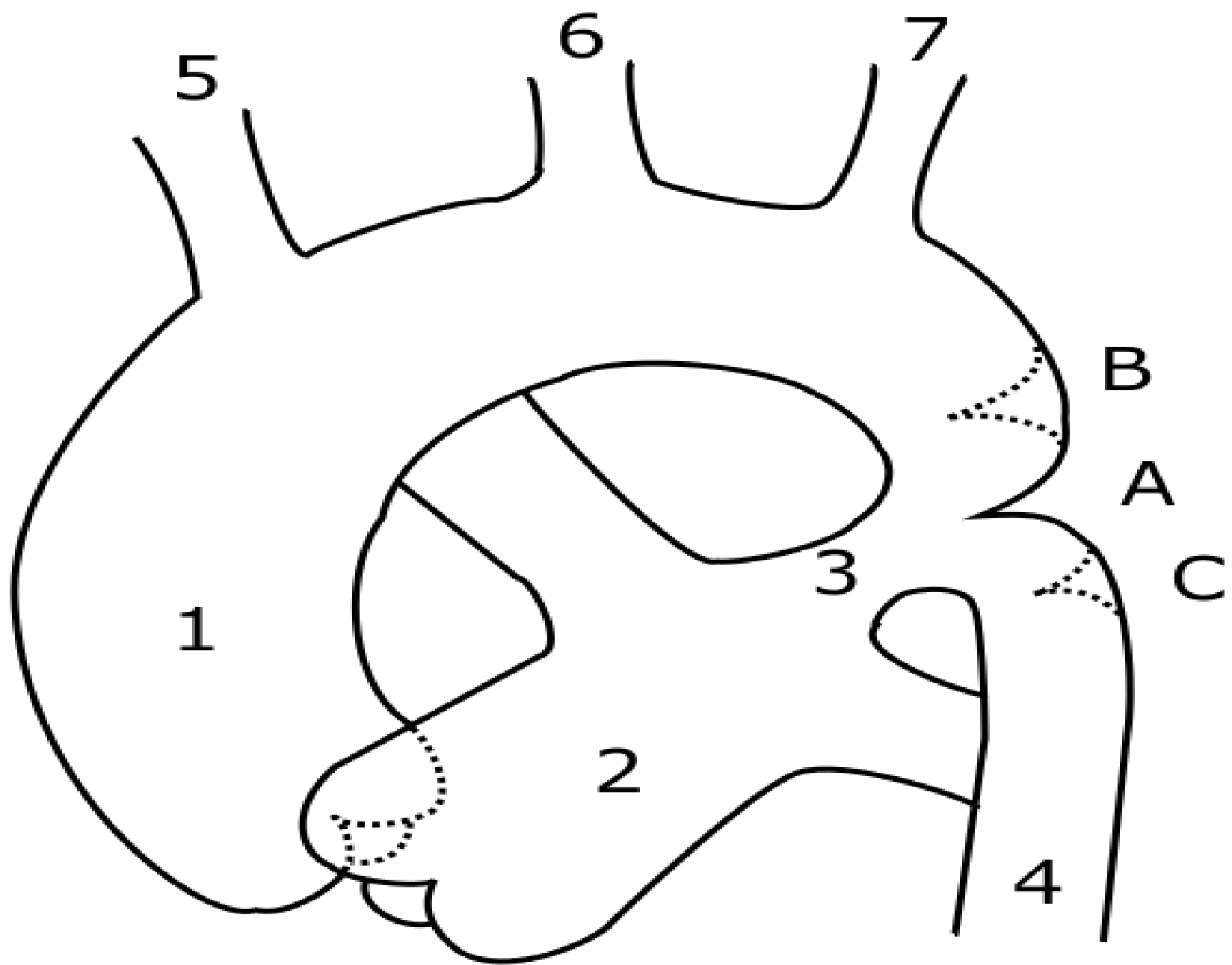
- Extension of tissue from the ductus arteriosus (a muscular artery) extends into the aorta (an elastic artery) during development.
- When ductus contracts and fibroses at birth, it leads to a narrowing of the aortic lumen.
- Limitation: ectopic tissue growth is not present in all patients.
- Coarctation is not a result of abnormal tissue growth, but rather, a result of abnormal fetal blood-flow patterns. *

* Rudolph AM, Heymann MA, Spitznas U. Hemodynamic considerations in the development of narrowing of the aorta. *Am J Cardiol.* Oct 1972;30(5):514-25



HEMODYNAMIC THEORY

- Prenatally, the aortic isthmus is a watershed area.
- So, the initial diameter of the isthmus is small, and if the proper molecular cues are not present, it may not grow sufficiently.



ASSOCIATIONS

- Bicuspid aortic valve (50-70%)
- VSD (20%)
- More than 70% of VSDs close spontaneously.^[1]
- 50% of patients with the Taussig-Bing anomaly.^[2]
- Complex cardiac defects (30%)
- 15% of children with Turner syndrome: genetic or familial

[1] Moene RJ, Gittenberger-de Groot AC, Oppenheimer-Dekker A, Bartelings MM. Anatomic characteristics of ventricular septal defect associated with coarctation of the aorta. *Am J Cardiol.* Apr 15 1987;59(9):952-5.

[2] Parr GV, Waldhausen JA, Bharati S, et al. Coarctation in Taussig-Bing malformation of the heart. Surgical significance. *J Thorac Cardiovasc Surg.* Aug 1983;86(2):280-7.

NATURAL COURSE

- Average age of survival: 35 years
- Mortality rate: 75% at age 46 years
- Common complications: hypertension; accelerated coronary artery disease; aortic dissection; stroke; heart failure
- 3 major causes of death:
 - spontaneous rupture of the aorta
 - bacterial endocarditis
 - cerebral hemorrhage

Campbell M. Natural history of coarctation of the aorta. *Br Heart J.* Sep 1970;32(5):633-40.

CLINICAL FEATURES

NEONATES

- Asymptomatic if PDA present or mild COA
- Absent/Delayed femoral pulses
- Murmur
- Systolic click
- Differential cyanosis
- Heart failure/Shock

OLDER INFANTS AND CHILDREN

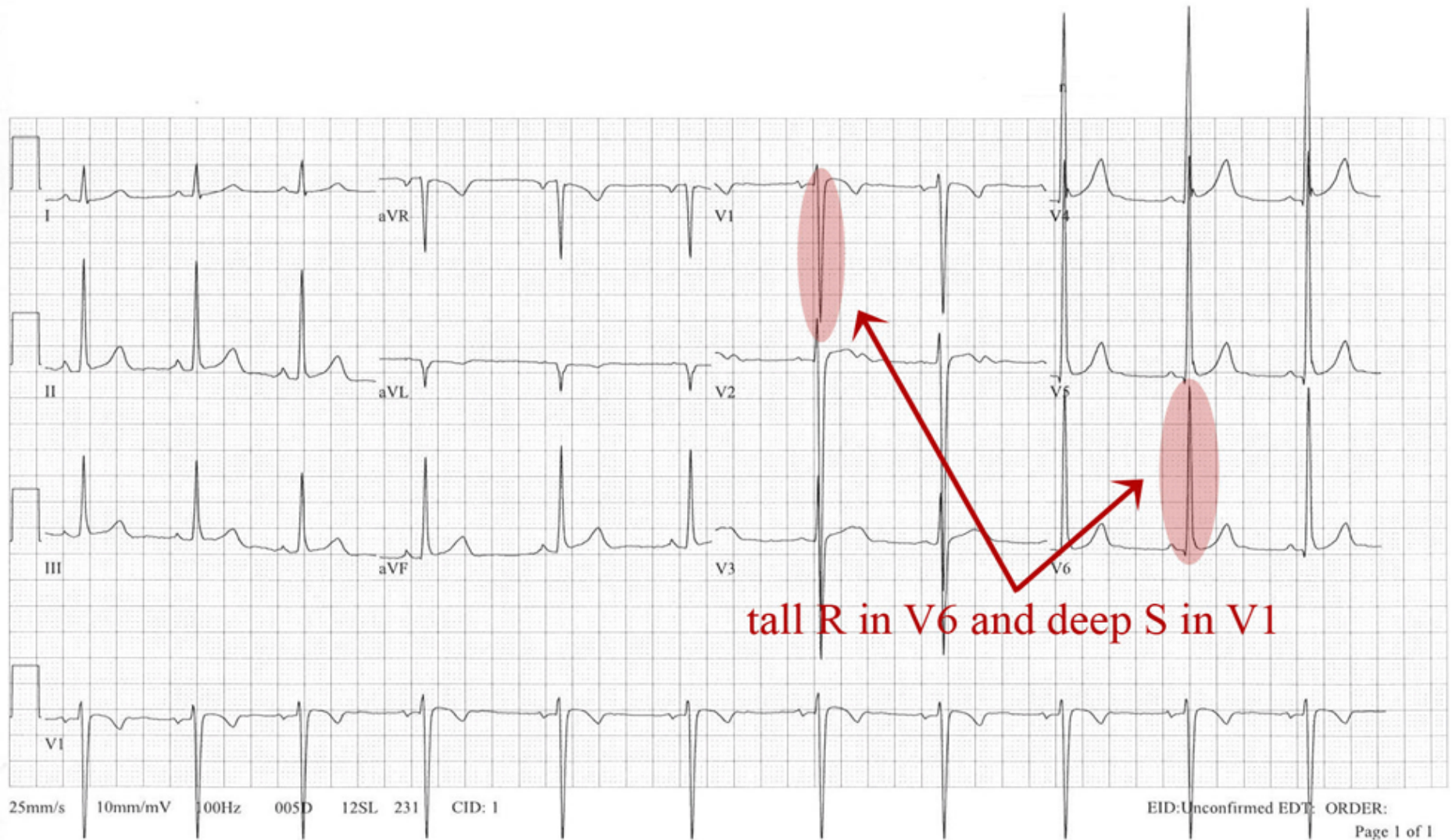
- Mostly asymptomatic
- Radiofemoral delay
- Murmur
- Symptoms with exertion: chest pain, cold extremities and claudication
- BP difference between extremities
- Hypertension

ADULTS

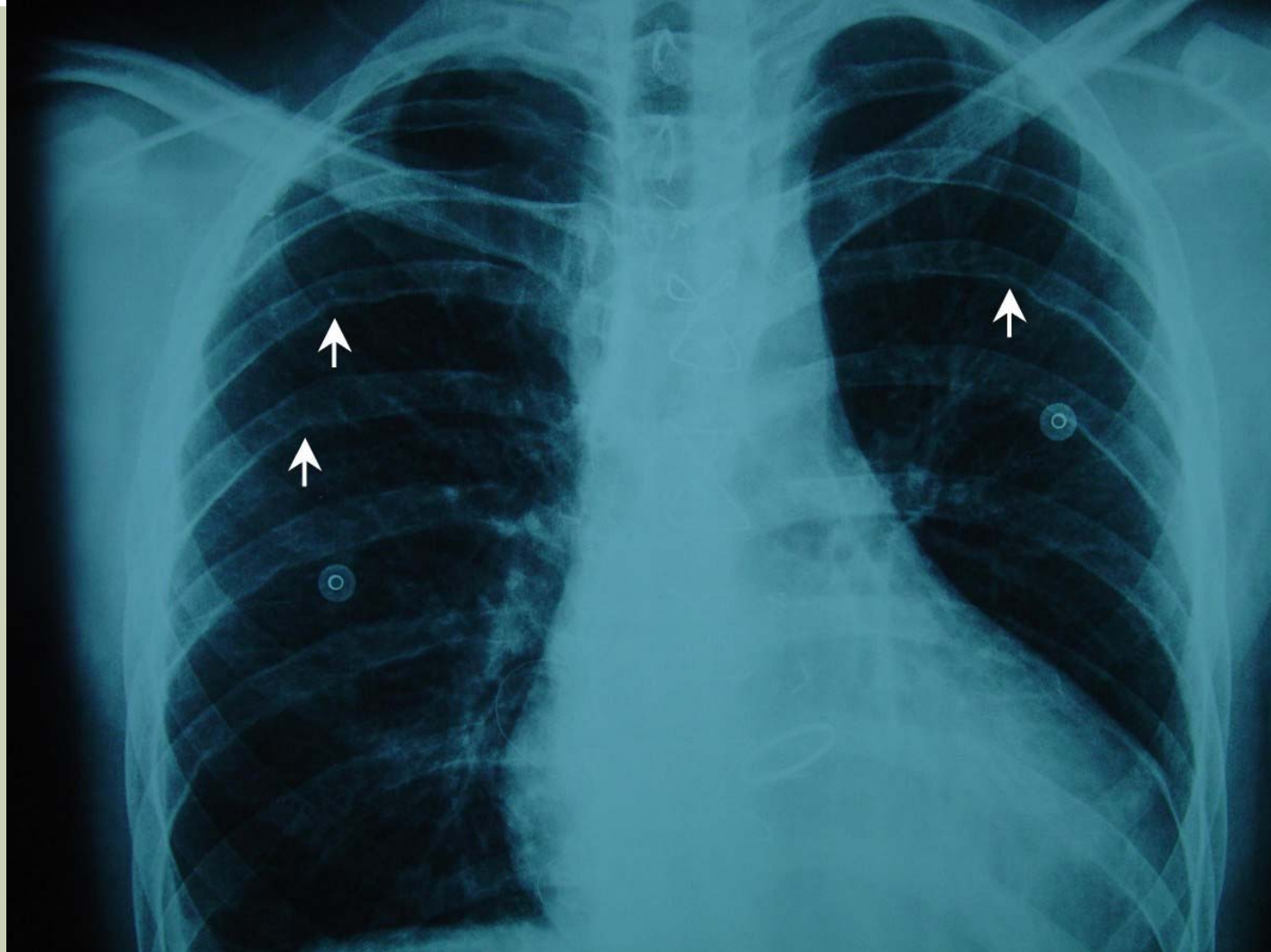
- Mostly asymptomatic
- Hypertension: classic
- Severe HTN: epistaxis, headaches, heart failure, dissection, claudication in lower extremities

INVESTIGATIONS

EKG

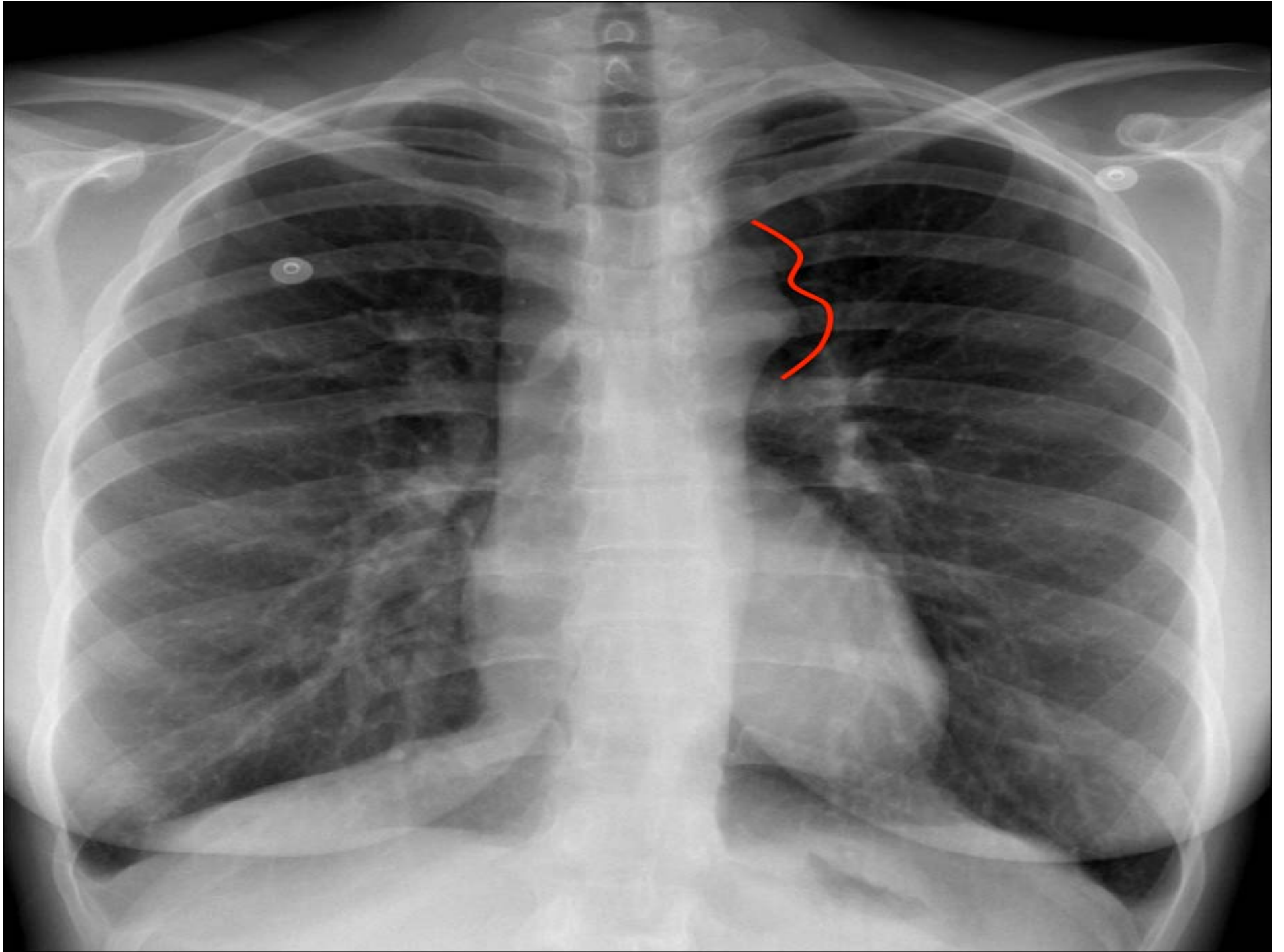


CXR

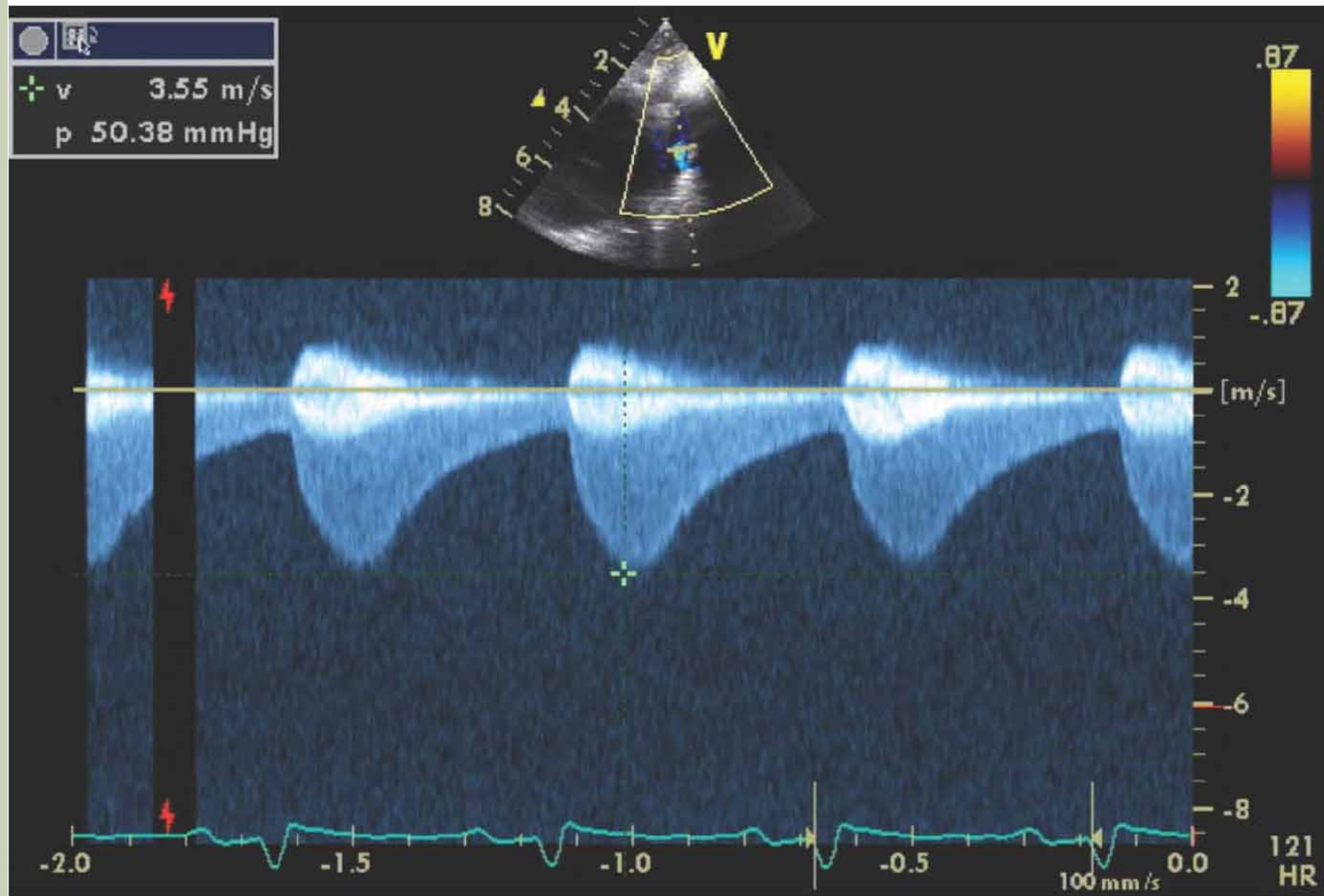


CXR

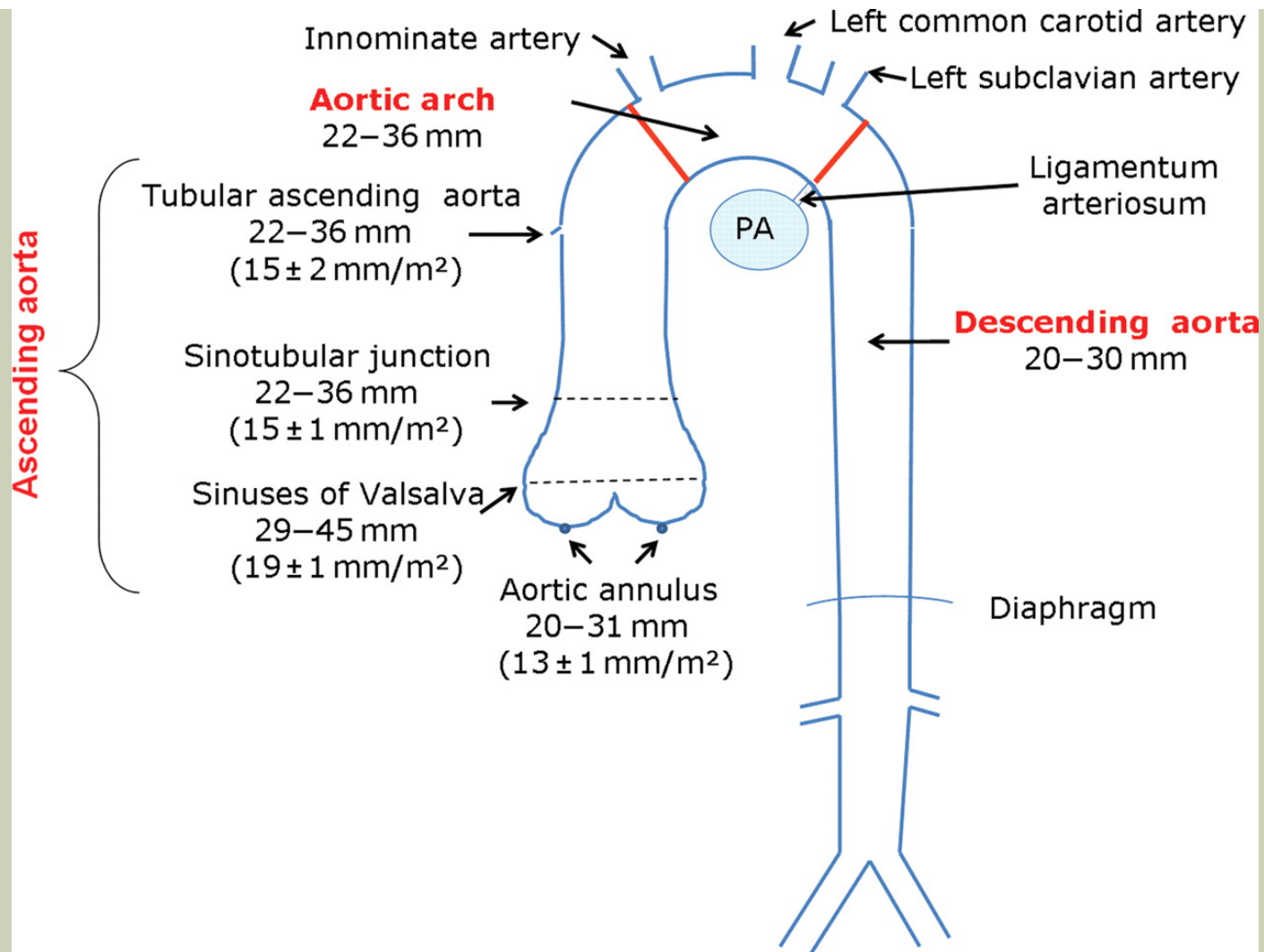
- Neonates: Cardiomegaly; increased pulmonary vascular markings
- Older children & Adults:
 - Classic sign of rib notching (by Meckel in 1827)
 - Notching increases with age
 - More prominent in the posterior third of the 3-8 ribs.
 - 3 sign



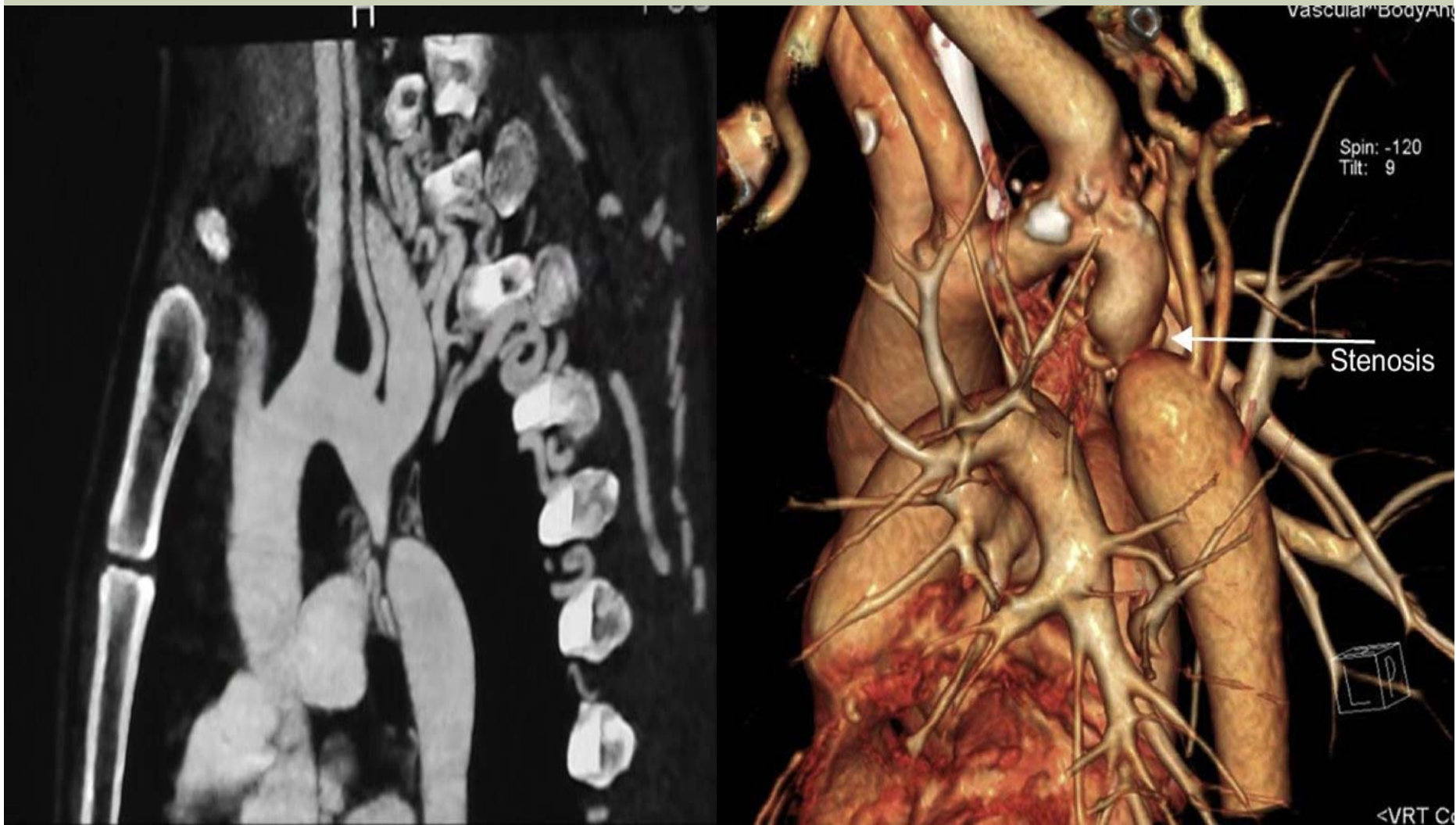
ECHO



MEASUREMENTS



CT



CMR



- 2008 ACHD AHA/ACC guidelines
- MRI recommended for complete evaluation of thoracic aorta in adults (Class IIa; Level C).
- No radiation

CARDIAC CATHETERIZATION

- Usually performed as part of therapeutic intervention
- Association with complex cardiac defects
- Coronary artery disease in adults

EXTRACARDIAC FINDINGS

- Intracranial aneurysms: 10% versus 2% in general population.

- Connolly HM et. al, Intracranial aneurysms in patients with coarctation of the aorta: a prospective magnetic resonance angiographic study of 100 patients. Mayo Clin. Proc. 2003 Dec;78(12):1491-9.

PRENATAL DIAGNOSIS

- Difficult
- Only 10% of fetal cardiac output flows across it.
- Presence of PDA limits the pressure gradient across the coarctation site.
- As early as 16-18 weeks.

MANAGEMENT

MEDICAL THERAPY

- **IV Prostaglandin E1**
 - Limited to critical coarctation in neonates.
 - Preoperative temporization to optimize the patient's hemodynamic status before surgery/balloon.
- **Inotropic support for CHF**
- **Supportive care**

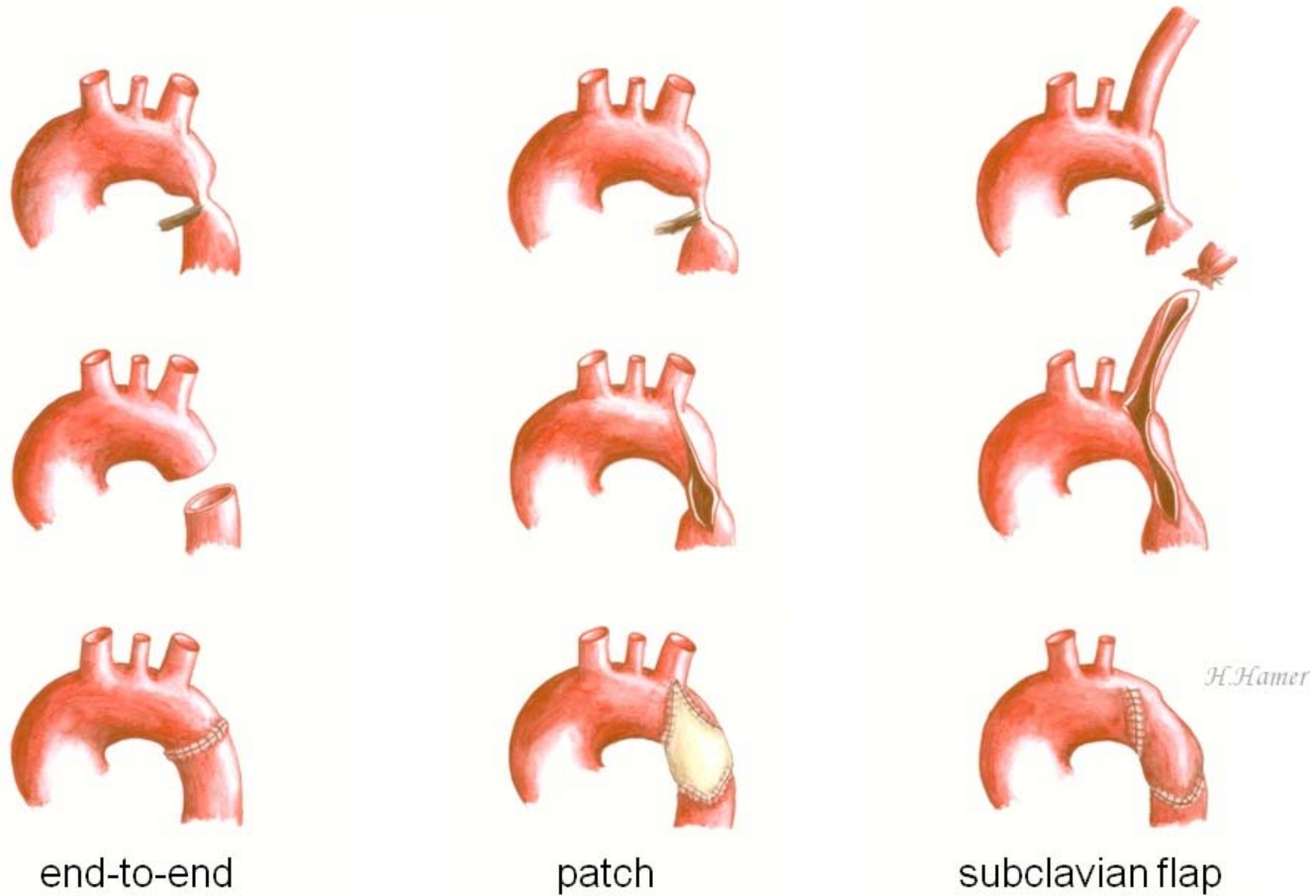
INDICATIONS

- Peak to peak gradient ≥ 20 mm Hg
- Peak to peak gradient < 20 mm Hg (anatomic evidence of significant coarctation + significant collateral flow +/- low cardiac output)

CONTRAINDICATIONS

- Coexisting conditions: NEC
- Sepsis

SURGICAL THERAPY



Surgical procedures for coarctation (1). Type of procedure depends on anatomy

Courtesy: ACHD learning center

SURGICAL THERAPY



Interposition graft

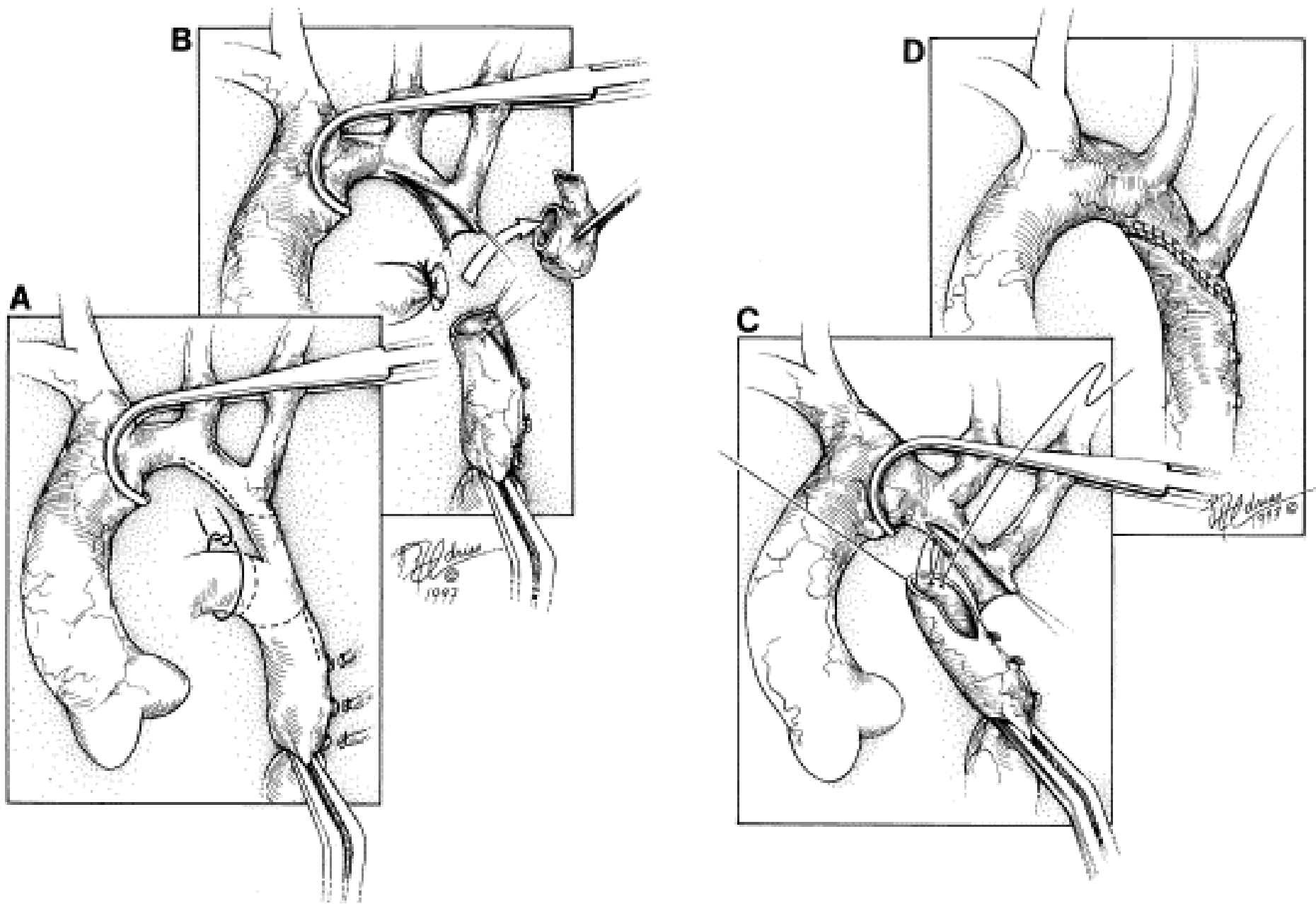
extended aortic arch repair

extra-anatomical bypass

Surgical procedures for coarctation (2). Type of procedure depends on anatomy

END-TO-END REANASTOMOSIS

- Reserved for infants and small children because of absence of enlarged collaterals and short distance for reanastomosis
- Benefits:
 - avoidance of prosthetic materials
 - excision of ductal tissue
 - preservation of the left subclavian artery
 - total relief of left ventricular outflow obstruction
 - growth potential of the aortic anastomosis.



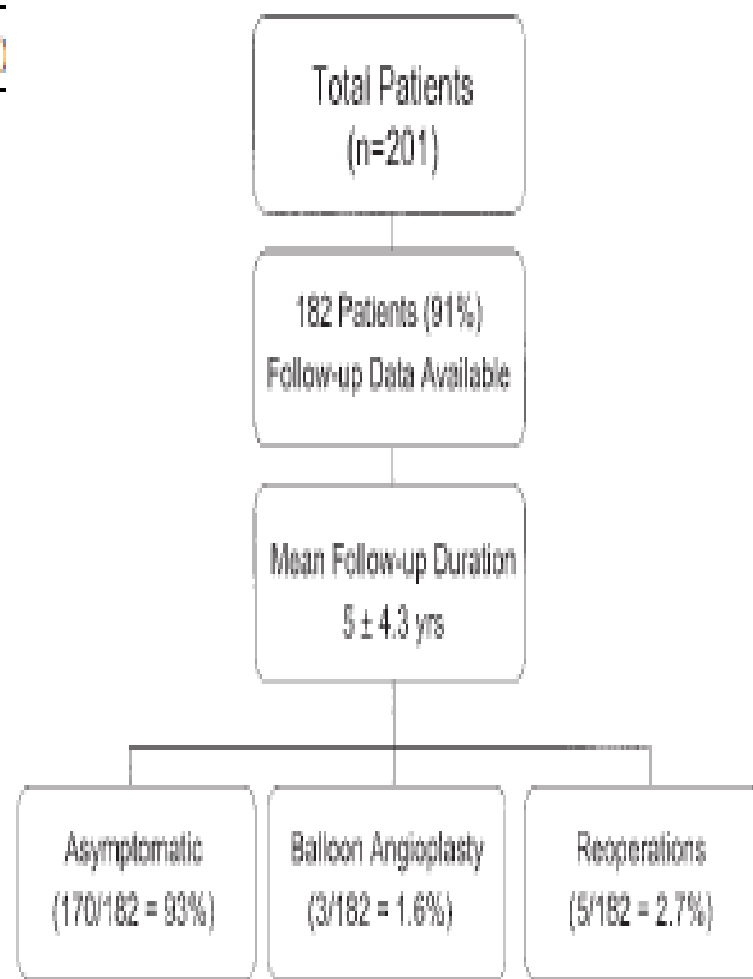
Backer, CL et al., Repair of coarctation with resection and extended end-to-end anastomosis, *The Annals of Thoracic Surgery* ; Volume 66, Issue 4, Pages 1365-1370 (October 1998)

Coarctation of the Aorta: Midterm Outcomes of Resection With Extended End-to-End Anastomosis

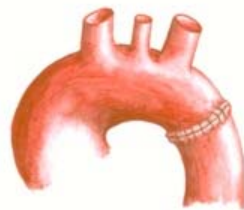
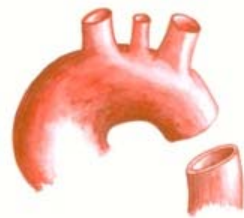
Sunjay Kaushal, MD, Carl L. Backer, MD, Jay N. Patel, BA, Shivani K. Patel, BS, Brandon L. Walker, MS, Thomas J. Weigel, MD, Guy Randolph, MD, David Wax, MD, and Constantine Mavroudis, MD

(Ann Thorac Surg 2009;88:1932–8)

Characteristics	Number (%)
Early mortality	
Yes	4 (2.0%)
No	197 (98%)
Reoperations (in 30 days)	
Yes	3 (1.5%)
No	198 (98.5%)
Complications	
Septicemia	8 (4%)
Recurrent laryngeal temporary paresis	6 (3%)
Chylothorax	5 (3%)
Pulmonary hypertensive crisis	2 (1%)
Seizures	3 (1.5%)
Length of stay (days)	
Mean	13.8 ± 15
Median	7
Range	3–169



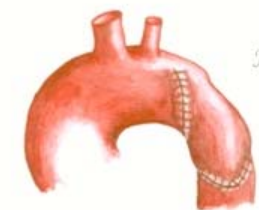
SURGICAL THERAPY



end-to-end



patch



subclavian flap

Surgical procedures for coarctation (1). Type of procedure depends on anatomy

Courtesy: ACHD learning center

SUBCLAVIAN-FLAP AORTOPLASTY

■ Benefits:

- avoidance of prosthetic material
- decreased cross-clamp time
- possibility that the anastomosis may grow as the child ages.

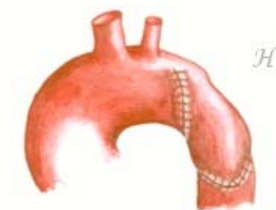
SURGICAL THERAPY



end-to-end



patch



subclavian flap

H. Hamer

Surgical procedures for coarctation (1). Type of procedure depends on anatomy

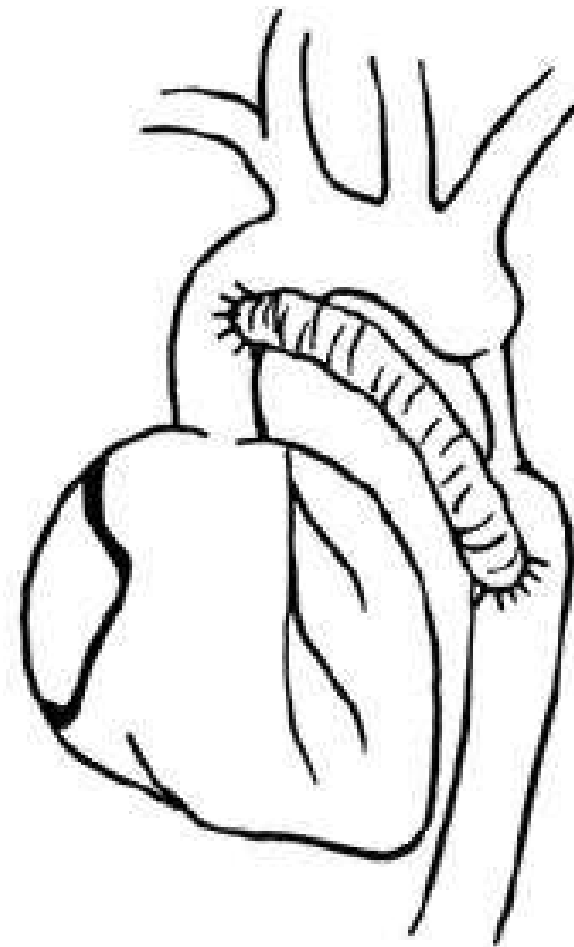
Courtesy: ACHD learning center

PROSTHETIC-PATCH ONLAY GRAFT

- First performed in 1957
- Out of favor due to the need for prosthetic implants and the risk of aneurysms and pseudoaneurysms
- Differential elasticity of Dacron grafts in relation to supple aorta is believed to increase the rate of aneurysm formation.
- Homograft patch material has not shown an increased rate of aneurysmal formation

INTERPOSITION GRAFTS

- Most often used in older patients who have exceeded their growth potential
- Useful when the narrowed segment cannot be completely excised without making primary reanastomosis impossible.



SURGICAL COMPLICATIONS

- Perioperative mortality (< 1%)
- Rebound hypertension (64%):
 - First phase: 24-36 hours after repair from activation of the sympathetic nervous system
 - Second phase: rise in diastolic blood pressure as a result of activation of the renin-angiotensin system.
- Left recurrent laryngeal nerve paralysis (3%)
- Phrenic nerve injury

SURGICAL COMPLICATIONS (CONT'D)

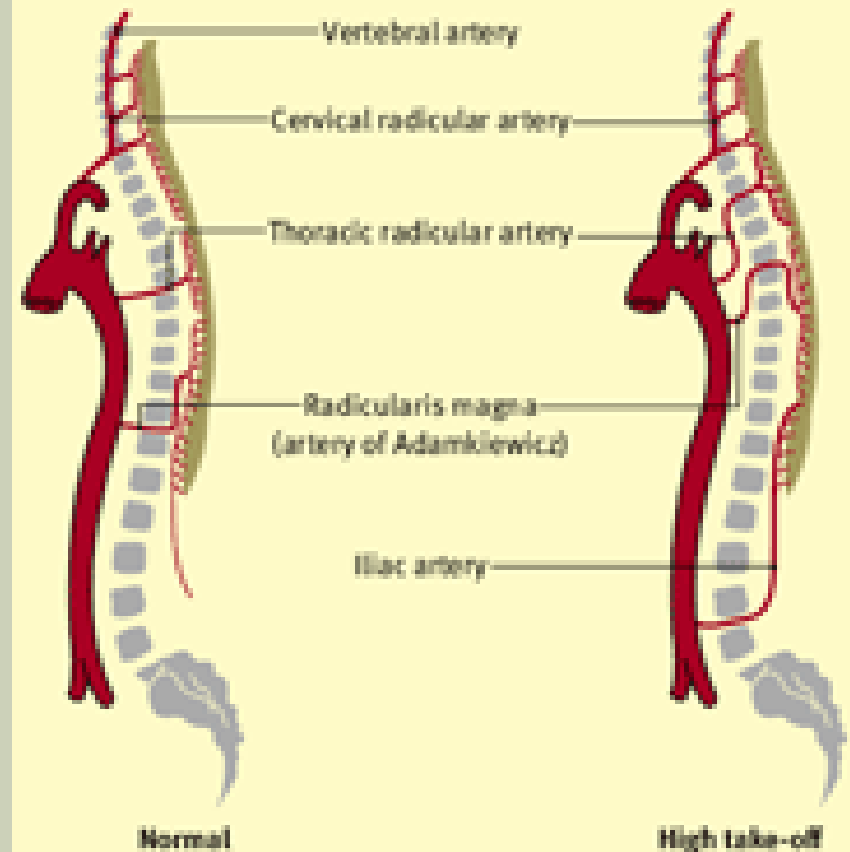
- **Mesenteric arteritis:**
 - Post-coarctectomy syndrome of abdominal pain and distension (first described by Sealy in 1957)
 - Seen in 20% of patients
 - Aggressive control of postoperative hypertension usually prevents the full-blown syndrome
 - Associated with coarctation in older patients, as this complication is rarely seen with infants and small children

SURGICAL COMPLICATIONS (CONT'D)

- **Recoarctation (5-10%):**
 - inadequate resection of ductal tissue
 - failure of anastomotic growth
 - suture-line thrombosis.
- **Chylothorax (3%)**
- **Subclavian steal**

PARAPLEGIA

- Incidence 0.1-1%.
- Incidence in adults rises proportionately with age: upto 2.6%.
- No definitive predisposing factors
- Poor collaterals, anomalies of the origin of the right subclavian artery, distal hypertension during cross-clamp, reoperation, or relative hyperthermia during the operation may contribute.
- Variation in spinal-cord blood supply, including the elusive artery of Adamkiewicz



BALLOON ANGIOPLASTY

- 422 procedures in patients with median age of 4.2 years (2 days-63 years) from 1982-1995. *
- 3 deaths
- 80 procedures (19%) had suboptimal outcome:
 - Residual gradient of >20 , death, tear, stroke

* McCrindle BW et al. Acute results of balloon angioplasty of native coarctation versus recurrent aortic obstruction are equivalent. Valvuloplasty and Angioplasty of Congenital Anomalies (VACA) Registry Investigators. J Am Coll Cardiol. 1996 Dec;28(7):1810-7.

BALLOON ANGIOPLASTY

- BA not successful in diffuse hypoplasia
- Not recommended for infants < 4mo as recoarctation occurs and reintervention required in 5-12 weeks.
- Palliative BA considered in critically ill patients

BALLOON ANGIOPLASTY

■ CLASS II A:

- Palliative measure to stabilize a patient irrespective of age when severely depressed ventricular function, severe mitral regurgitation, low cardiac output, or systemic disease affected by the cardiac condition (*Level of Evidence: C*).

■ CLASS II B:

- 1. Patients beyond 4 to 6 months of age when associated with a transcatheter systolic coarctation gradient >20 mm Hg and suitable anatomy (*Level of Evidence: C*).
 - 2. Complex coarctation anatomy or systemic conditions such as connective tissue disease or Turner syndrome but should be scrutinized on a case-by-case basis (*Level of Evidence: C*).
- Feltes TF et. al; on behalf of the AHA Congenital Cardiac Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Clinical Cardiology, and Council on Cardiovascular Radiology and Intervention. Indications for cardiac catheterization and intervention in pediatric cardiac disease: a scientific statement from the American Heart Association. *Circulation*. 2011;123:2607-2652.

COMPLICATIONS OF BA

- Residual pressure gradient ≥ 20 mm Hg (20%)
- Recoarctation (5-25%)
- Aneurysm formation (5-7%)
- Femoral arterial complications (15%)
- Aortic rupture and dissection (rare)

BA VERSUS SURGERY

- Risk of recoarctation and aneurysm greater in BA group than surgical group

- Shaddy RE et. al. Comparison of angioplasty and surgery for unoperated coarctation of the aorta. *Circulation*. 1993 Mar;87(3):793-9.
- Cowley CG et. Al. Long-term, randomized comparison of balloon angioplasty and surgery for native coarctation of the aorta in childhood. *Circulation*. 2005 Jun 28;111(25):3453-6.

STENTING: BENEFITS

- Minimal residual gradient
- Improves luminal diameter
- Sustains hemodynamic benefit
- Reduces complications
- Likely planned intervention in children < 25 kg

INDICATIONS

■ CLASS II A:

- Peak gradient of >20 mm Hg (*Level of Evidence: B*).
- Peak gradient of <20 mm Hg but with systemic hypertension due to anatomic narrowing (*Level of Evidence: C*).
- Long-segment coarctation with a peak gradient >20 mm Hg (*Level of Evidence: B*).
- Patients in whom balloon angioplasty has failed, as long as a stent that can be expanded to an adult size can be implanted (*Level of Evidence: B*)

■ CLASS II B

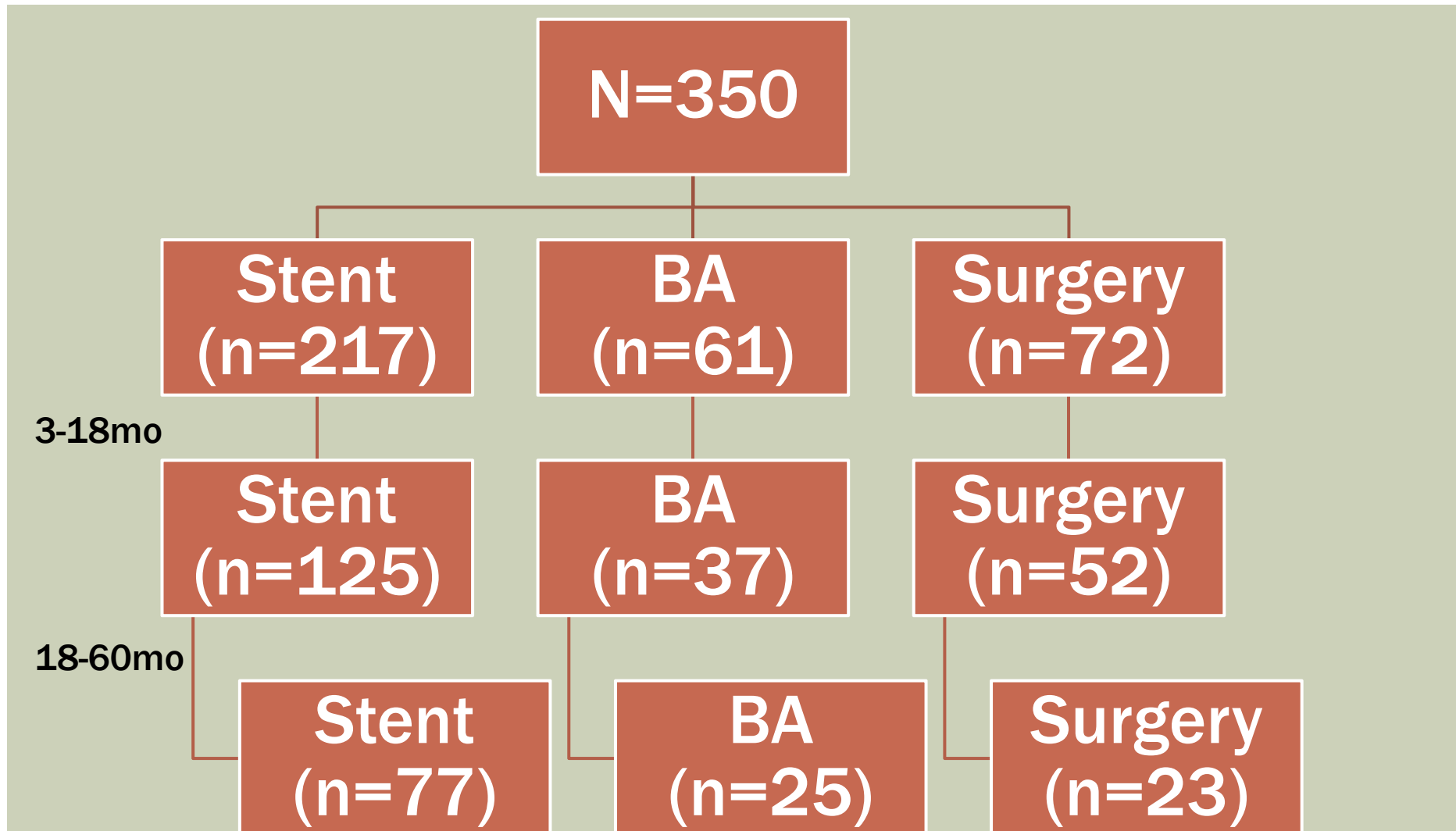
- High risk surgical candidates (*Level of Evidence: C*).
- Peak gradient of <20 mm Hg but with an elevated left ventricular end-diastolic pressure and significant aortic collaterals (*Level of Evidence: C*)

Feltes TF et. al; Indications for cardiac catheterization and intervention in pediatric cardiac disease: a scientific statement from the American Heart Association. *Circulation*. 2011;123:2607-2652

Comparison of Surgical, Stent, and Balloon Angioplasty Treatment of Native Coarctation of the Aorta

JACC Vol. 58, No. 25, 2011

December 13/20, 2011;2664-74



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INCLUSION & EXCLUSION CRITERIA

1. Inclusion criteria

- a. Body weight ≥ 10 kg
- b. Presence of significant native coarctation based on the following:
 1. ULG > 20 mm Hg
 2. ULG ≥ 10 mm Hg with either decreased LV function (EF $< 30\%$) or moderate to severe aortic insufficiency
 3. ULG ≥ 10 mm Hg plus significant collateral flow

2. Exclusion criteria

- a. Body weight < 10 kg
- b. Refusal to sign consent
- c. Requiring other surgical procedures that would entail correction of the coarctation segment in the same setting
- d. Known or suspected arteritis
- e. Recurrent coarctation of the aorta

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BASELINE CHARACTERISTICS

	Surgery (n = 72)	Balloon (n = 61)	Stent (n = 217)	p Value (2-Sided)
Age, yrs	10.0 ± 9.7	9.0 ± 8.0	16.6 ± 10.9	<0.001*
Age range, yrs	0.1/58.6	0.4/42.5	2.2/74.3	
Weight, kg	35 ± 24	30 ± 21	55 ± 24	<0.001*
Male	69%	64%	69%	0.750
Pre-intervention right-arm SBP, mm Hg	137 ± 19	138 ± 23	143 ± 21	0.061
Pre-intervention ULG	37 ± 21	43 ± 23	40 ± 24	0.399
Pre-intervention catheterization SBP gradient	NA	38.7	36.7	0.459
Pre-intervention coarctation measurement, mm/BSA	NA	5.8	4.3	0.001*
Coarctation location				0.212
Isthmus	86%	95%	90%	
Distal	56%	63%	71%	
Proximal	31%	32%	19%	
Transverse aorta	7%	2%	8%	
Complex	5%	2%	1%	
Abdominal/thoracic aorta	0%	2%	1%	
Bicuspid aortic valve	40%	46%	40%	0.708
Other CHD diagnosis	13%	5%	9%	0.308
Catheterization laboratory patient volume				0.090
Large	60%	67%	66%	
Medium	14%	23%	19%	
Small	26%	10%	15%	

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ACUTE OUTCOMES

	Surgery (n = 72)	Balloon (n = 61)	Stent (n = 217)	p Value (2-Sided)
Post-intervention right-arm SBP, mm Hg	123 ± 13	118 ± 15	125 ± 15	0.002*
Discharge ULG	7.7 ± 18.2	10.3 ± 12.9	4.9 ± 13.2	0.032
Discharge ULG ≤10 mm Hg	64%	56%	76%	0.011*
Discharge ULG ≤15 mm Hg	73%	69%	81%	0.101
Post-intervention catheterization SBP gradient	NA	12.4 ± 12.2	4.8 ± 8.6	<0.001*
% Increase in coarctation measurement post-intervention	NA	125%	172%	0.008*
Any complications	18.1%†	9.8%	2.3%	<0.001*
Aortic wall injury	UK‡	9.8%	0.0%	<0.001*
Dissection/intimal tear	UK‡	9.8%	0.0%	
Aneurysm	UK‡	0.0%	0.0%	
Balloon rupture	NA	0.0%	0.5%	
Stent migration	NA	n/a	1.4%	
Femoral	UK‡	0.0%	0.5%	
Atrial fibrillation	3%	0%	0%	
Severe/prolonged hypertension	3%	0%	0%	
Length of stay, days	6.4/5.0	3.6/1.0	2.4/1.0	<0.001*

Spinal injury (n=1)
A.Fib (n=2)
Severe HTN (n=2)
Pleural effusion (n=3)
Vocal cord Palsy (n=1)

Stent migration (n=3)
Groin hematoma (n=1)

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SHORT TERM OUTCOMES

Outcomes	Surgery (n = 52)	Balloon (n = 37)	Stent (n = 125)	p Value (2-Sided)
Age at follow-up, yrs	12.1 ± 10.9	10.4 ± 9.2	17.2 ± 10.1	<0.001†
Weight at follow-up, kg	41.3 ± 30.6	34.4 ± 22.4	59.3 ± 21.9	<0.001†
Normal SBP*	84.6%	72.2%	87.2%	0.096
Antihypertensive medications	40%	16%	41%	0.019†
Right-arm SBP, mm Hg	114 ± 17	118 ± 14	121 ± 13	0.005†
ULG	1.2 ± 21.5	9.9 ± 16.8	0.9 ± 13.9	0.011†
ULG ≤10 mm Hg	89%	35%	75%	<0.001†
ULG ≤15 mm Hg	91%	65%	86%	0.004†

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SHORT TERM OUTCOMES

Outcomes	Surgery (n = 26)	Balloon (n = 28)	Stent (n = 97)	p Value (2-Sided)
Any complications*	23.1%	32.1%	8.3%	0.003‡
Aortic wall Injury	11.5%	21.4%	3.1%	0.004‡
Dissection/intimal tear	0.0%	7.1%	0.0%	0.062
Aneurysm	11.5%	14.3%	3.1%	0.040‡
Coarct:Dao ratio, mean	0.91	0.73	0.82	0.003‡
Coarct:Dao ≥ 0.6	87.0%	79.0%	90.0%	0.247
Any reobstruction	19.2%	32.1%	15.4%	0.057
Mild†	7.7%	17.9%	11.3%	
Moderate	7.7%	3.6%	4.1%	
Severe	3.9%	10.7%	0%	

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INTERMEDIATE OUTCOMES

	Surgery (n = 23)	Balloon (n = 25)	Stent (n = 77)	p Value (2-Sided)
Age at follow-up	15.0 ± 11.2	12.9 ± 6.5	18.3 ± 9.8	0.035†
Weight at follow-up	57.4 ± 22.4	40.3 ± 16.2	60.8 ± 18.6	<0.001†
Normal SBP*	96%	72%	82%	0.092
Antihypertensive medications	13%	16%	31%	0.130
Right-arm SBP, mm Hg	115 ± 9	122 ± 13	123 ± 13	0.044†
ULG	-1.4 ± 13.9	5.5 ± 14.3	1.9 ± 13.7	0.118
ULG ≤10 mm Hg	90%	55%	75%	0.032†
ULG ≤15 mm Hg	95%	82%	85%	0.421

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INTERMEDIATE OUTCOMES

	Surgery (n = 16)	Balloon (n = 16)	Stent (n = 56)	p Value (2-Sided)
Any complications*	25.0%	43.8%	12.5%	0.020‡
Aortic wall injury	12.5%	43.8%	7.1%	0.003‡
Dissection/intimal tear	0.0%	6.3%	1.8%	0.598
Aneurysm	12.5%	43.8%	5.4%	<0.001
Coarct:Dao ratio, mean	0.98	0.79	0.80	0.011‡
Coarct:Dao ratio ≥0.6	88%	93%	89%	1.000
Any reobstruction	18.8%	18.8%	14.3%	0.923
Mild†	6.3%	18.8%	12.5%	
Moderate	6.3%	0%	1.8%	
Severe	6.3%	0%	0%	

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REINTERVENTION RATE

	Surgery (n = 72)	Balloon (n = 61)	Stent (n = 217)
Patients with reintervention	4	6	44
Patients with planned procedures	0	2	35
Patients with unplanned procedures	4	4	9
Time to first planned reintervention, yrs	NA	1.43 ± 1.70	1.14 ± 1.15
Time to first unplanned reintervention, yrs	2.24 ± 2.23	1.28 ± 1.43	2.84 ± 1.43

Comparison of Surgical, Stent, and Balloon Angioplasty Treatment of Native Coarctation of the Aorta

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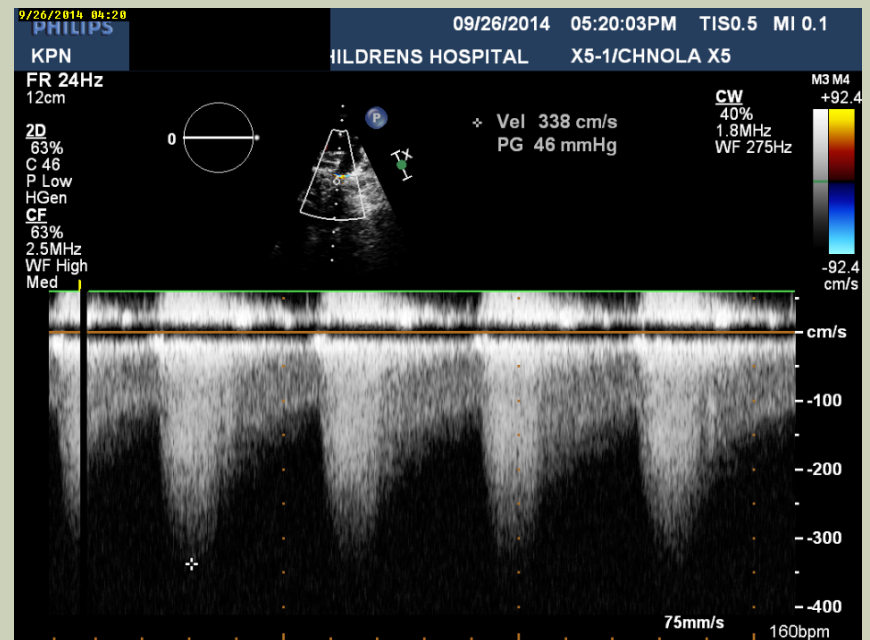
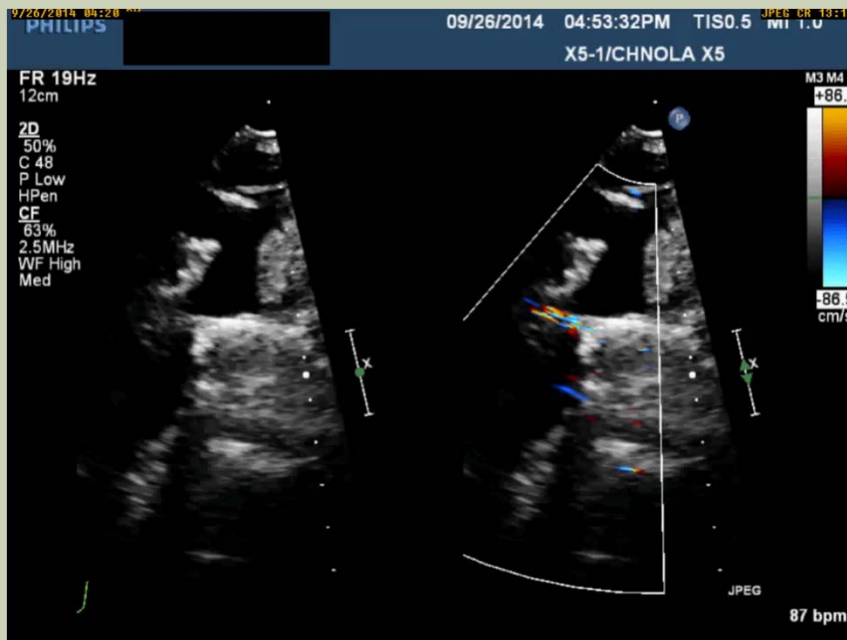
SUBGROUP ANALYSIS

	Surgery	Balloon	Stent	p Value (2-Sided)
Acute outcome	n = 23	n = 23	n = 57	
Age, yrs	8.7 ± 2.1	8.6 ± 2.1	9.3 ± 1.8	0.301
Discharge ULG	2.3 ± 12.9	10.1 ± 14.3	2.9 ± 10.6	0.050†
Any complications	13.0%	13.0%	1.8%	0.035†
Aortic wall complications	NA	13.0%	0.0%	0.022†
Short-term follow-up outcome	n = 18	n = 14	n = 32	
Antihypertensive medications	44%	0%	28%	0.018†
Normal SBP*	89%	79%	84%	0.728
ULG	-4.9 ± 14.3	10.2 ± 16.5	2.0 ± 11.3	0.016†
Any complications	25%	38%	4%	0.010†
Aortic wall complications	13%	31%	0%	0.009†

OPTIMAL APPROACH

- Neonates and young infants < 4mo:
 - Medical supportive therapy + surgery
- Older infants (4 months - 5 years) and young children < 25 kg:
 - Balloon angioplasty if discrete and no arch hypoplasia
- Older children and adults (>25 kg):
 - Stenting

13 YO MALE WITH HYPERTENSION & DECREASED PULSES



MRI

