



# *Coarctation of the aorta - Clinical decision making* **When is the arch too small ?**

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*Disclosure : None*

A man with a mustache, wearing a white clerical shirt, stands in a church with his arms raised. The background shows wooden pews and a dark interior.

**GOD HAS A RIVAL.**

**#AtTheNecker**

**Coarctation  
repair is not a  
problem for me !**

**Any type  
Any weight  
Any time  
I'll fix it in 10  
minutes**

**R.Gaudin**



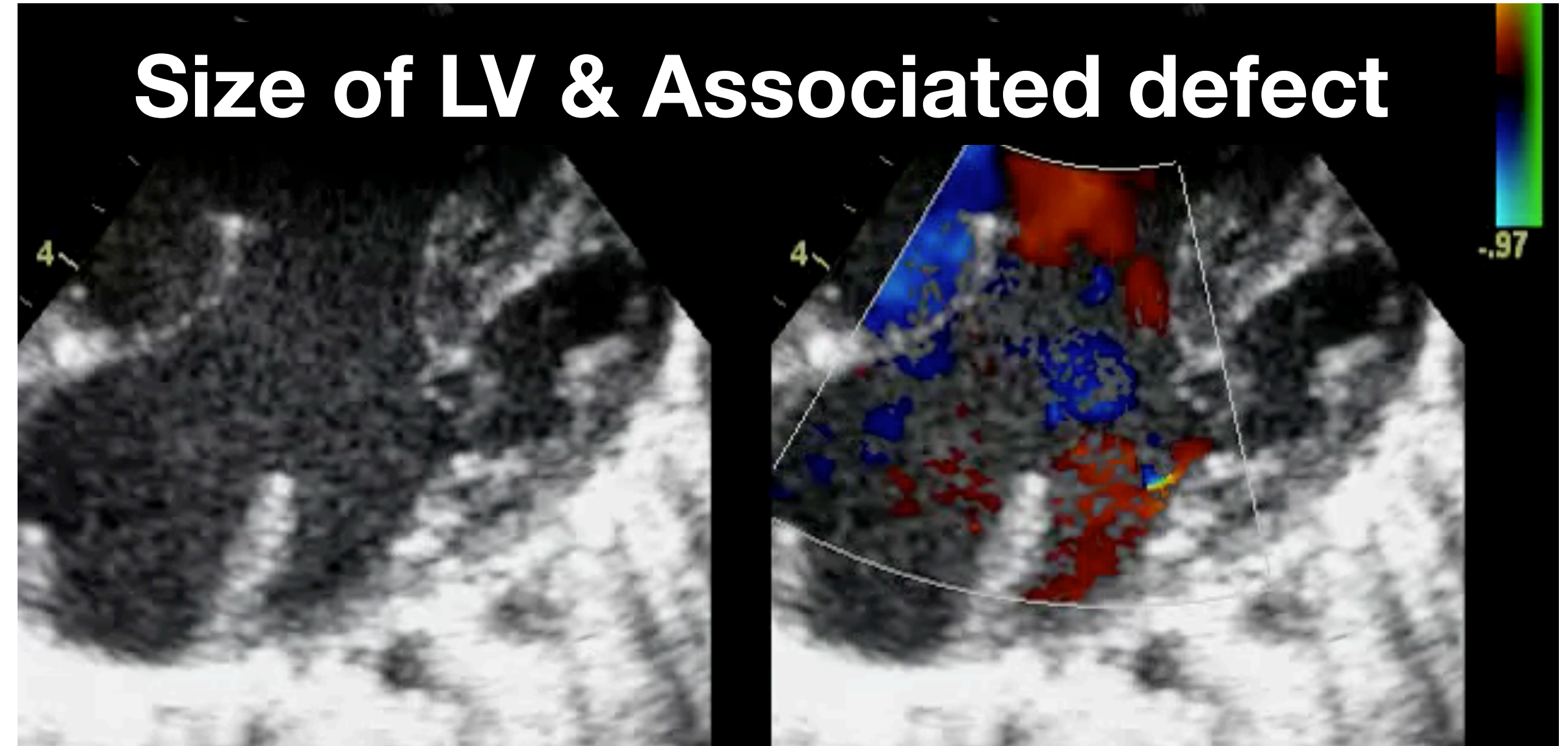
**O.Raisky**



**How small is too small ?**

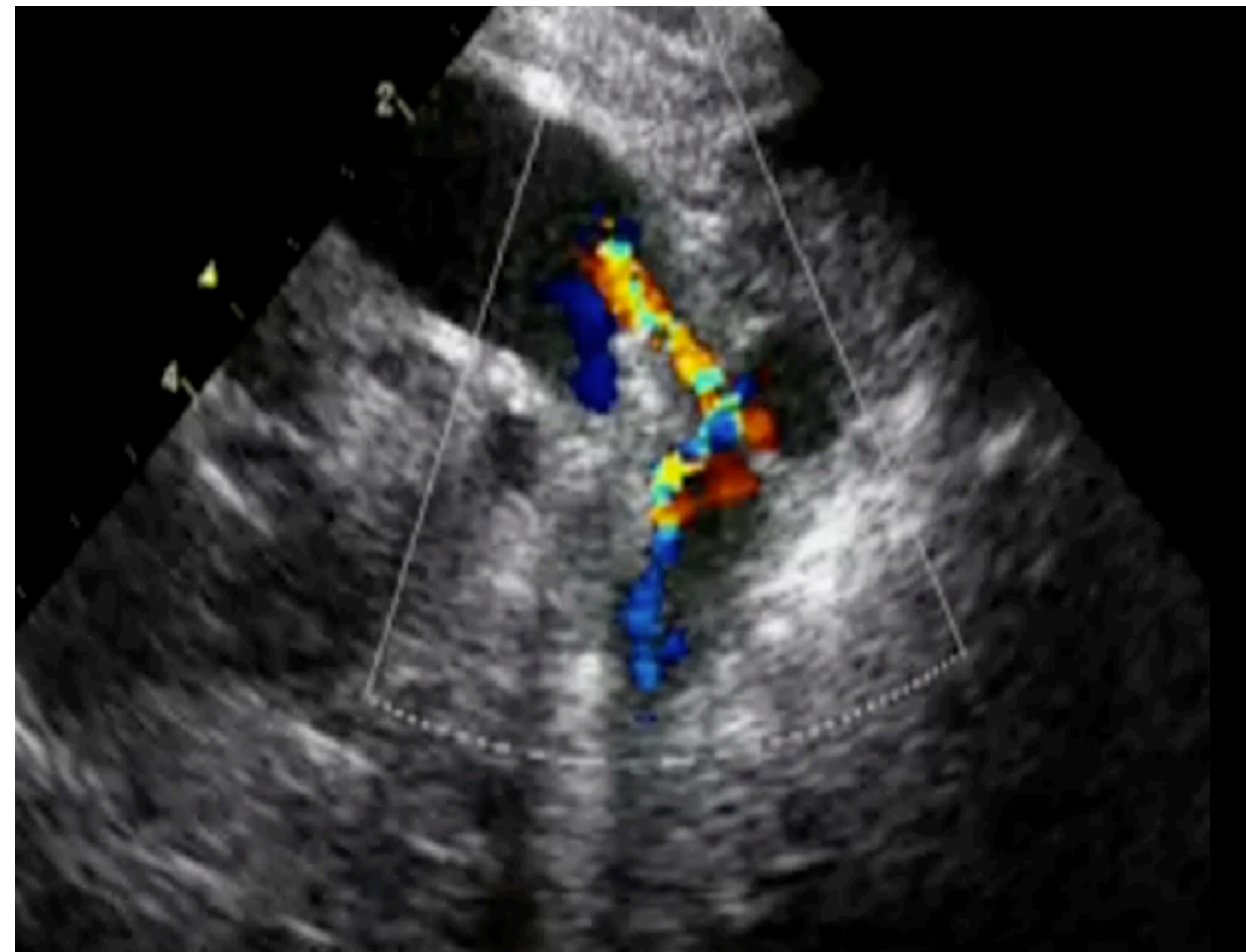
# What are the questions for decision making ?

1- Non anatomical / non aortic arch related factors contributing to decision

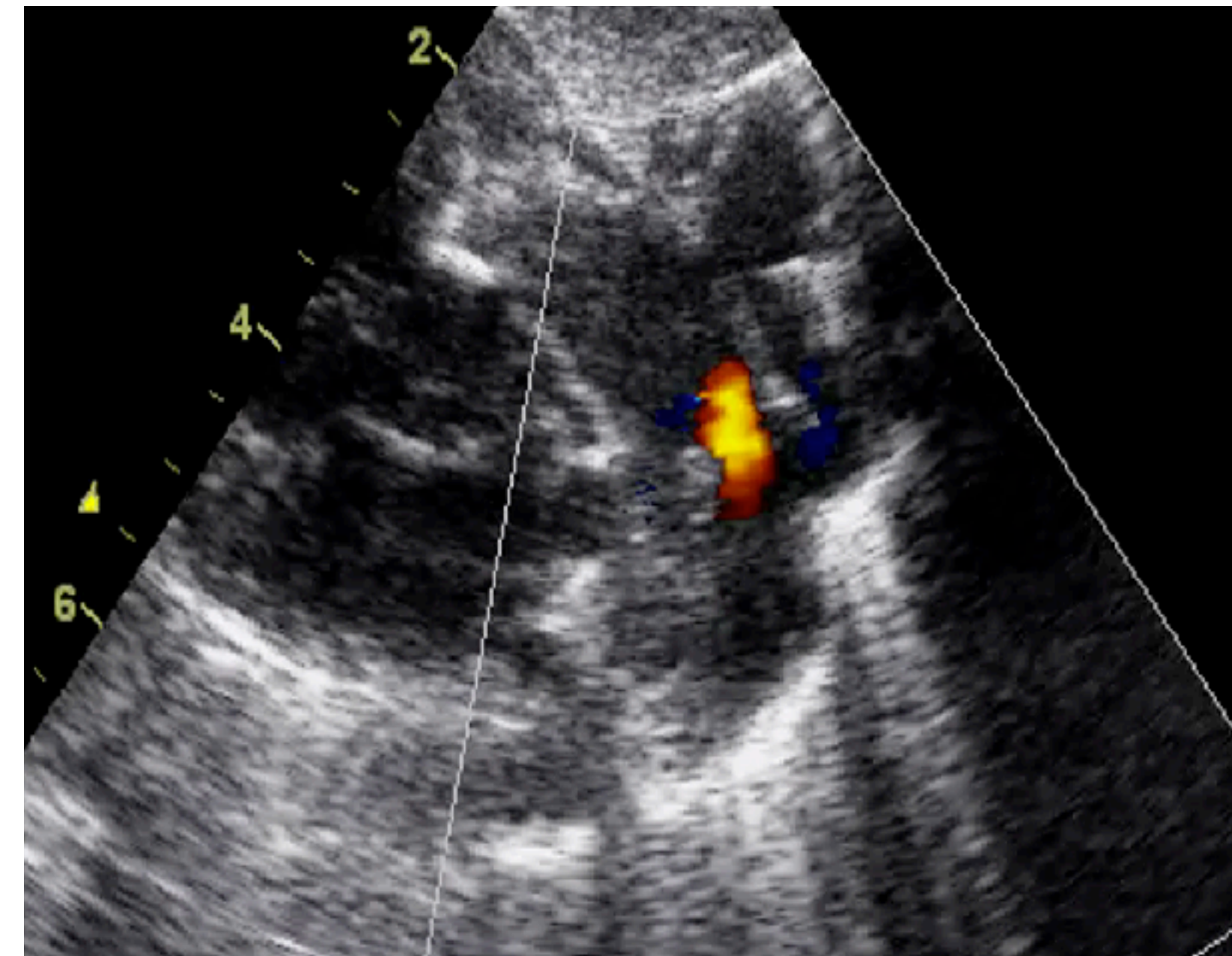


# What are the questions for decision making ?

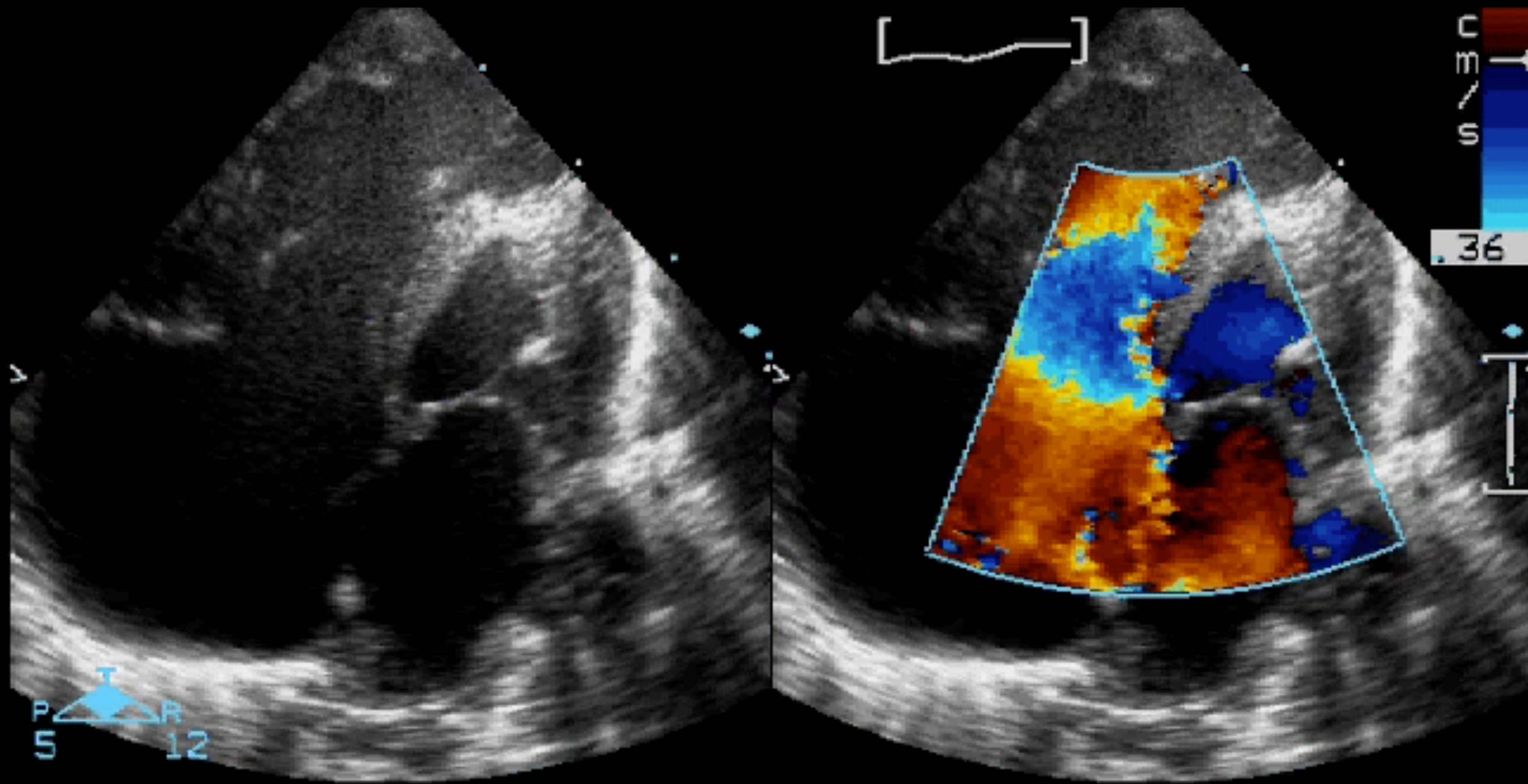
- 1- Non anatomical / non aortic arch related factors contributing to decision  
Physiology of neonatal coarctation



**LV perfuses Aorta + DA**

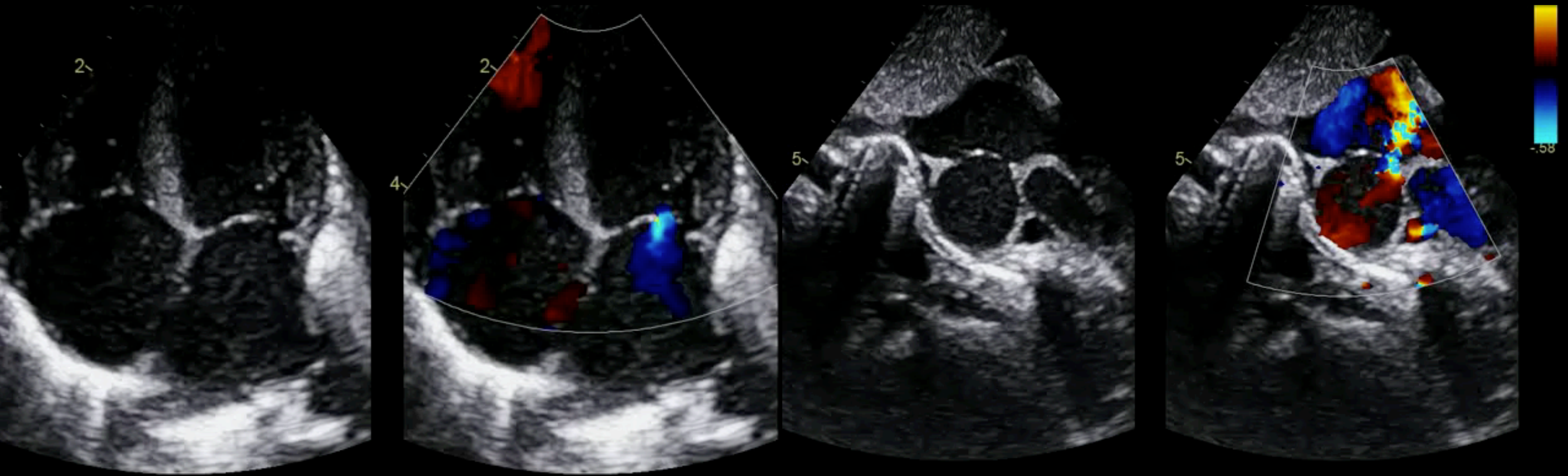


**LV does not perfuse totally aorta - why?**



Too small ?





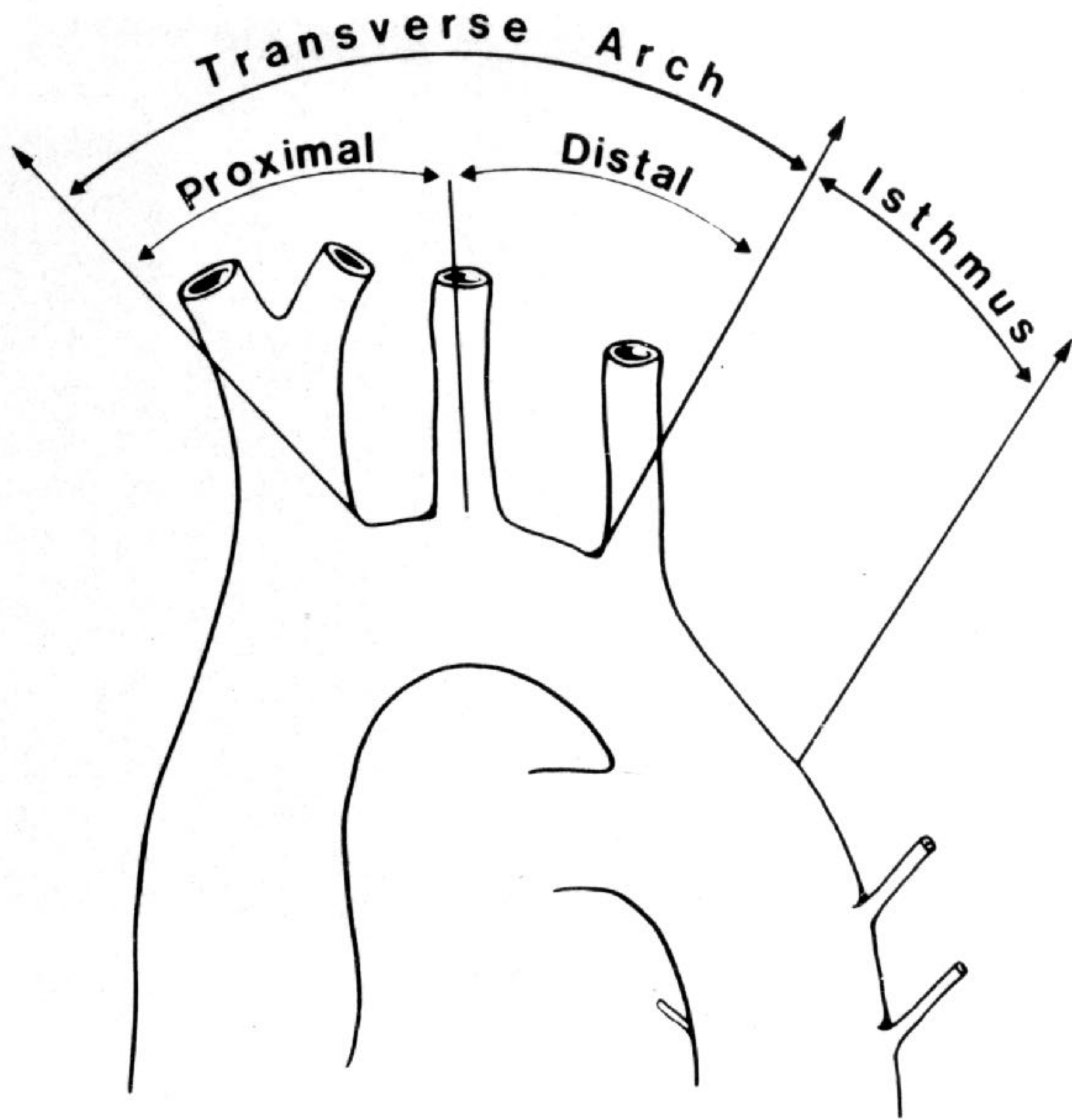
**Failing**



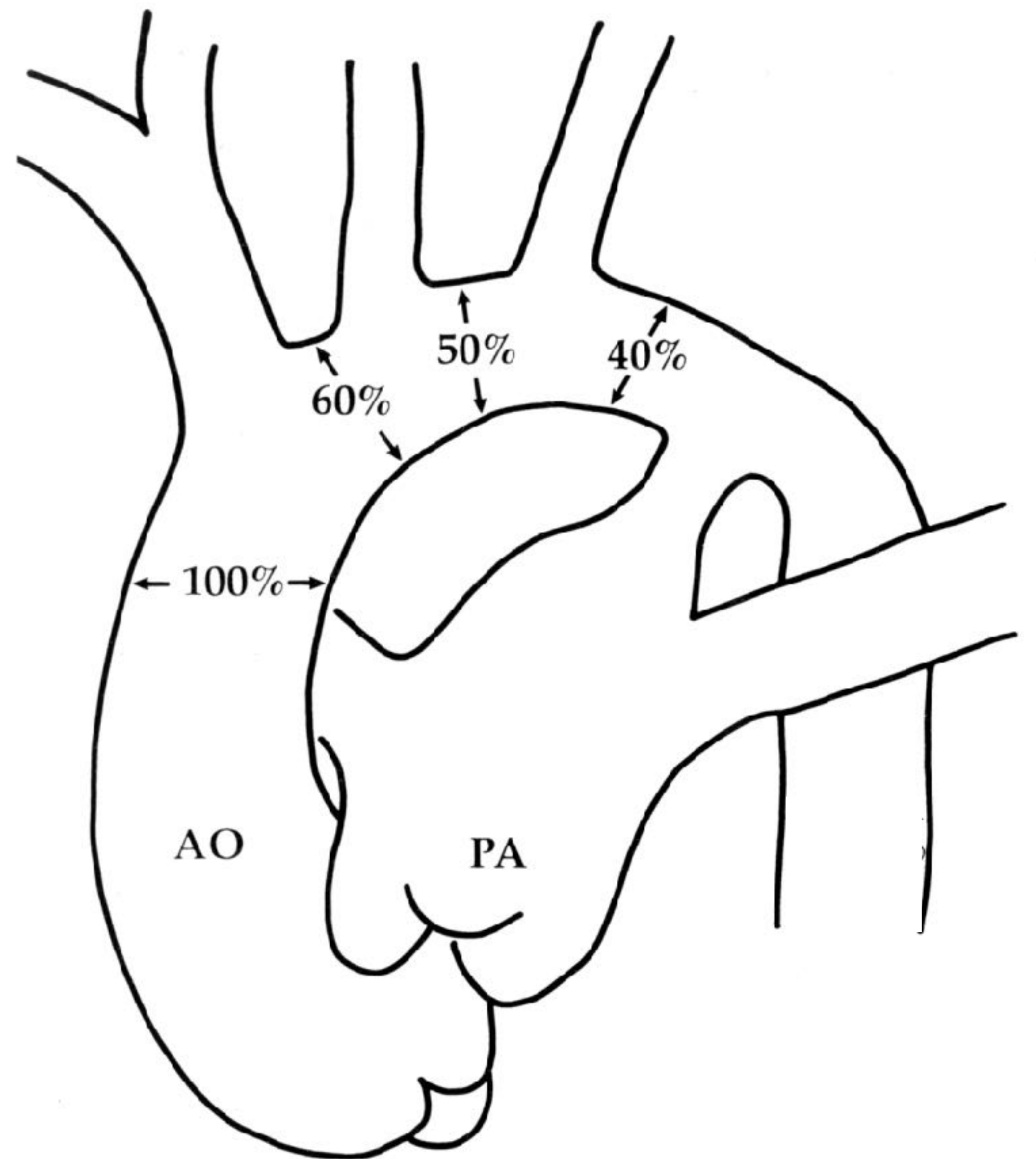
# What is the question for decision making ?

**3- Does size/anatomy of the aortic arch allow repair through thoracotomy ?**

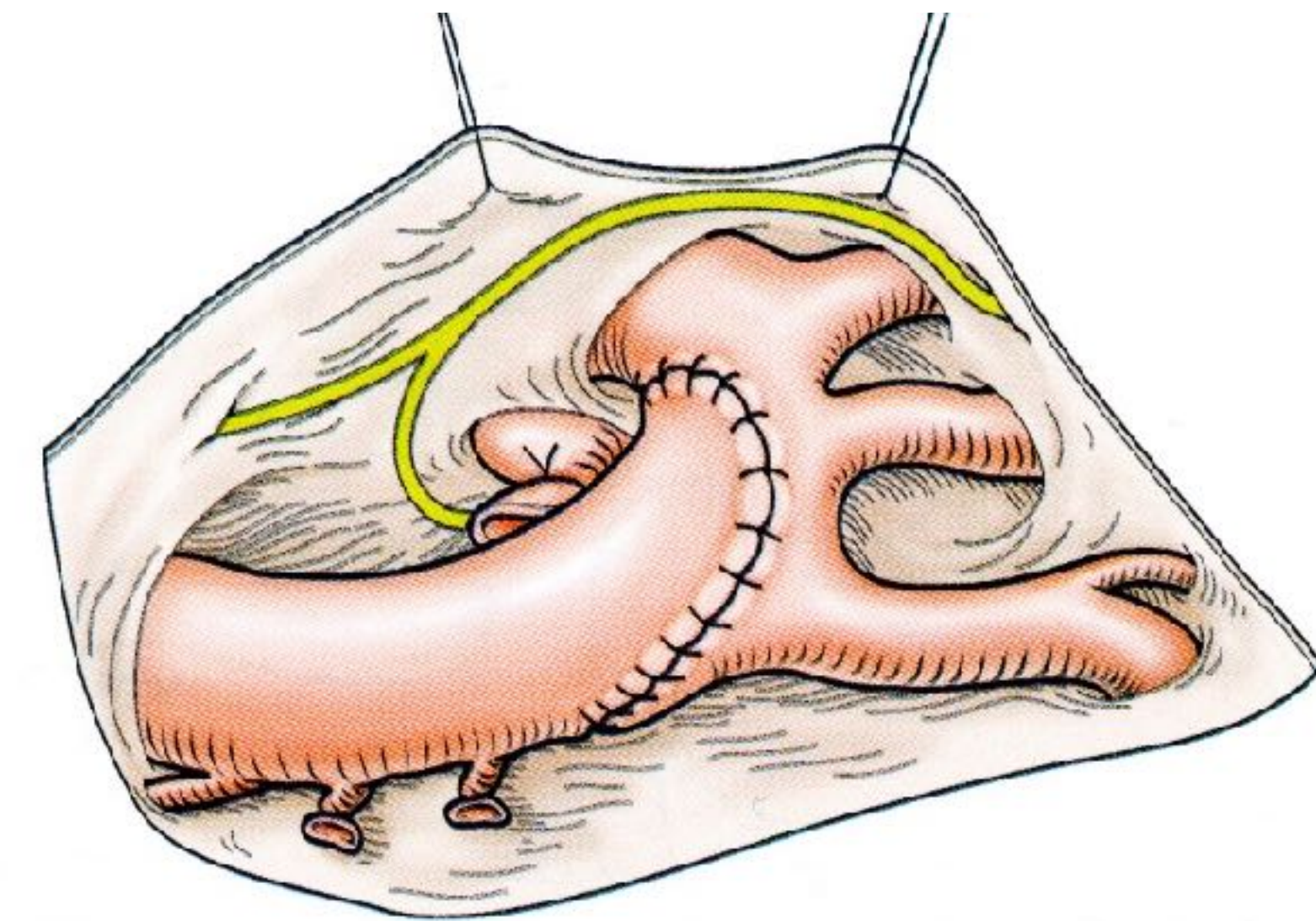
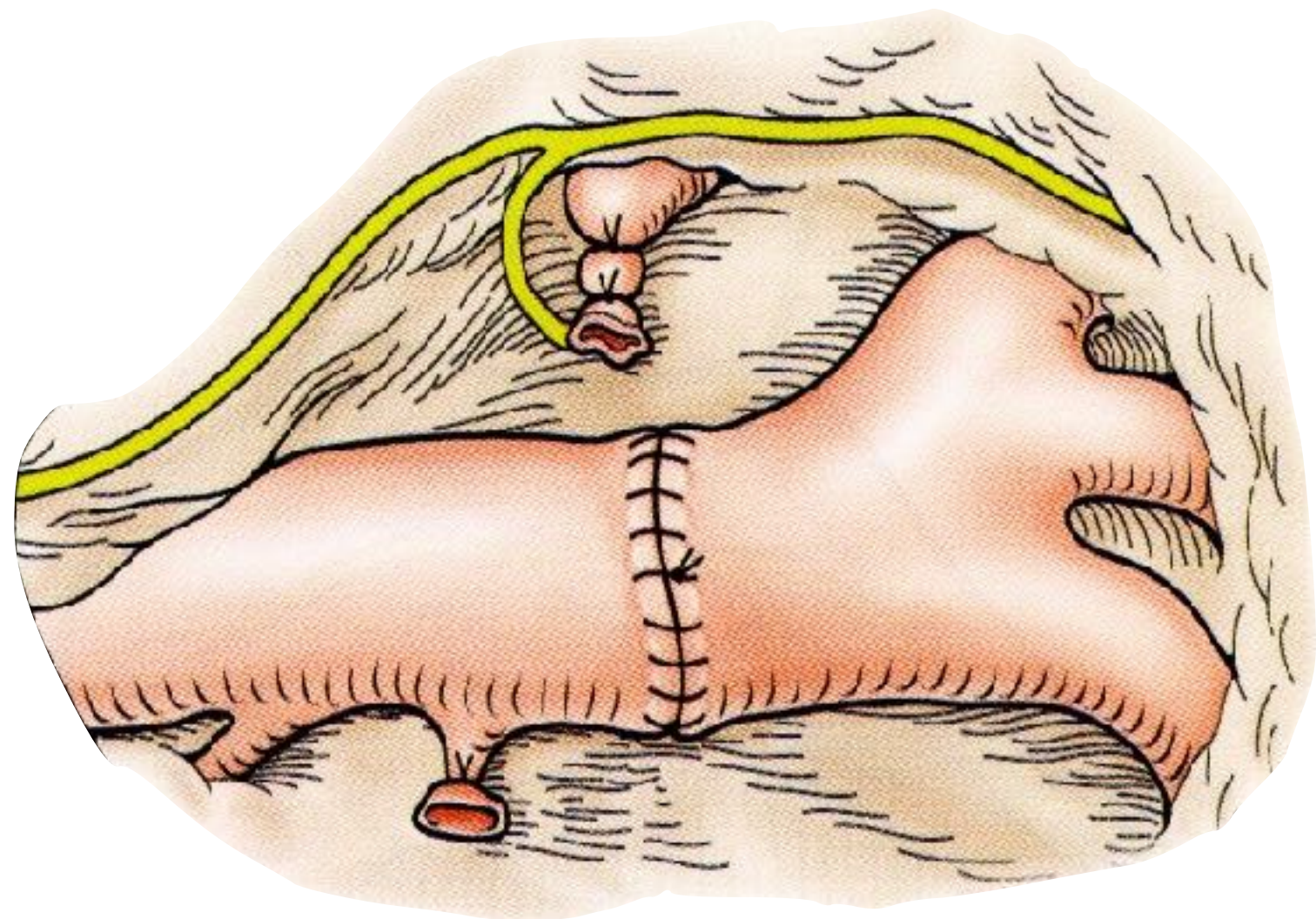
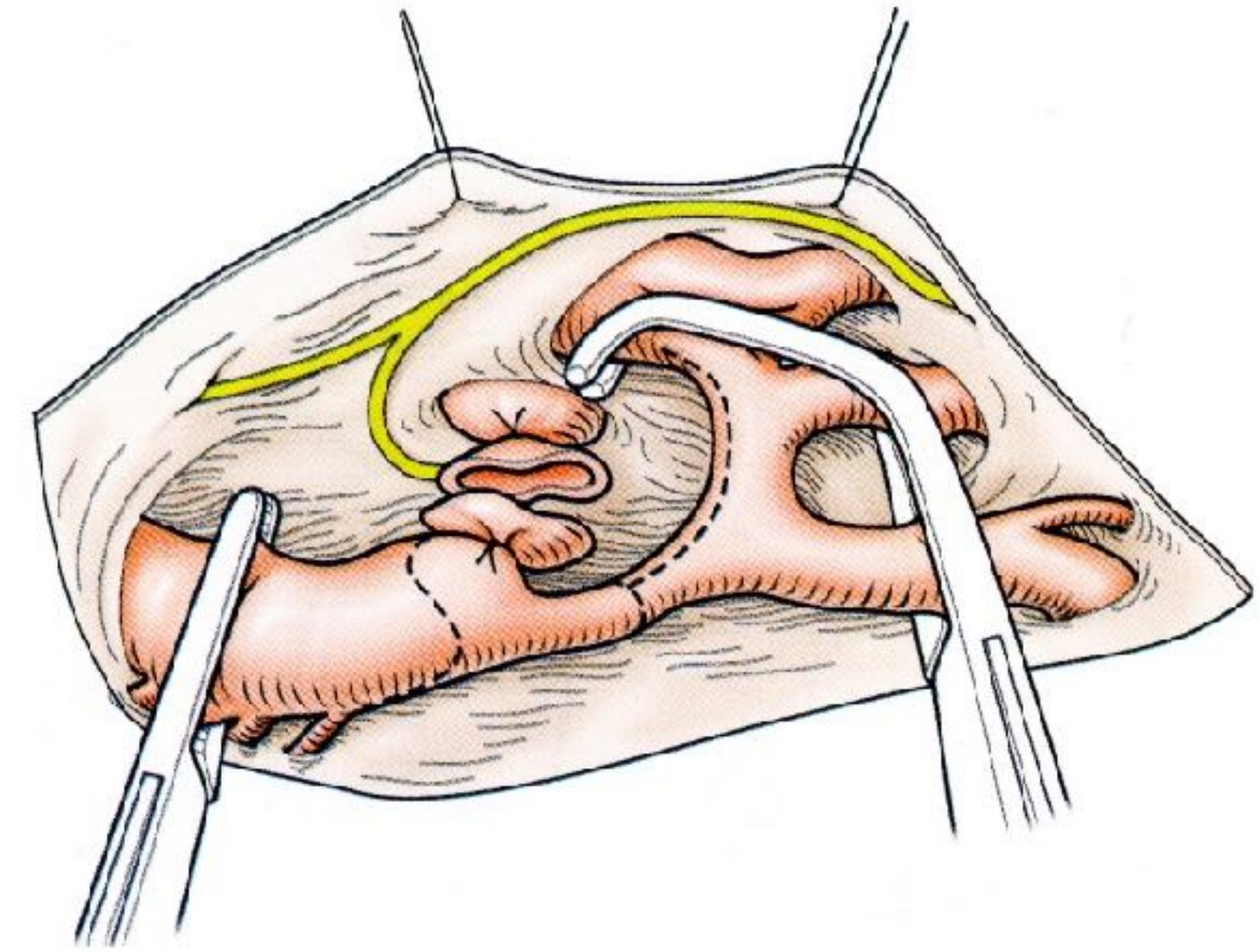
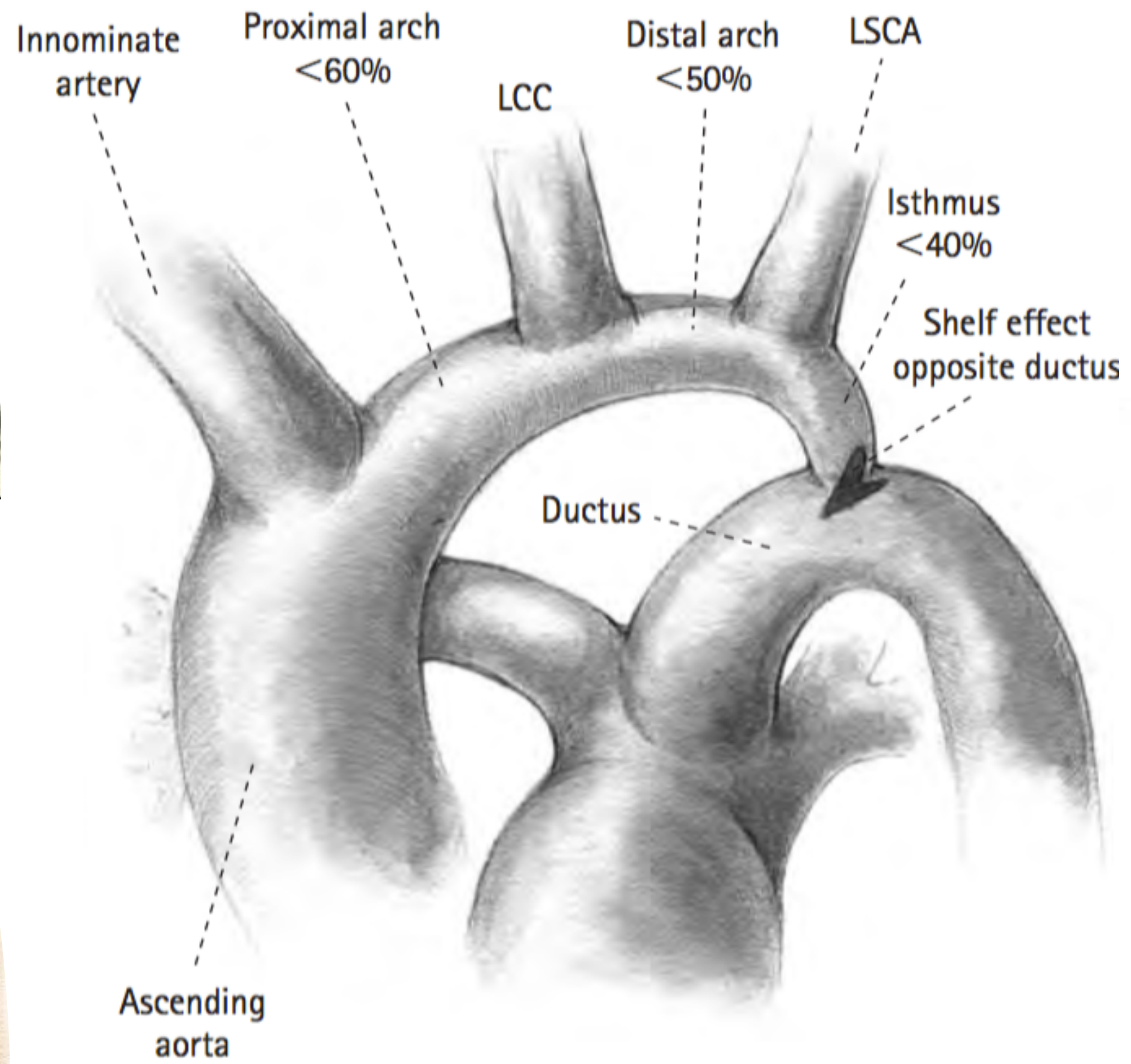
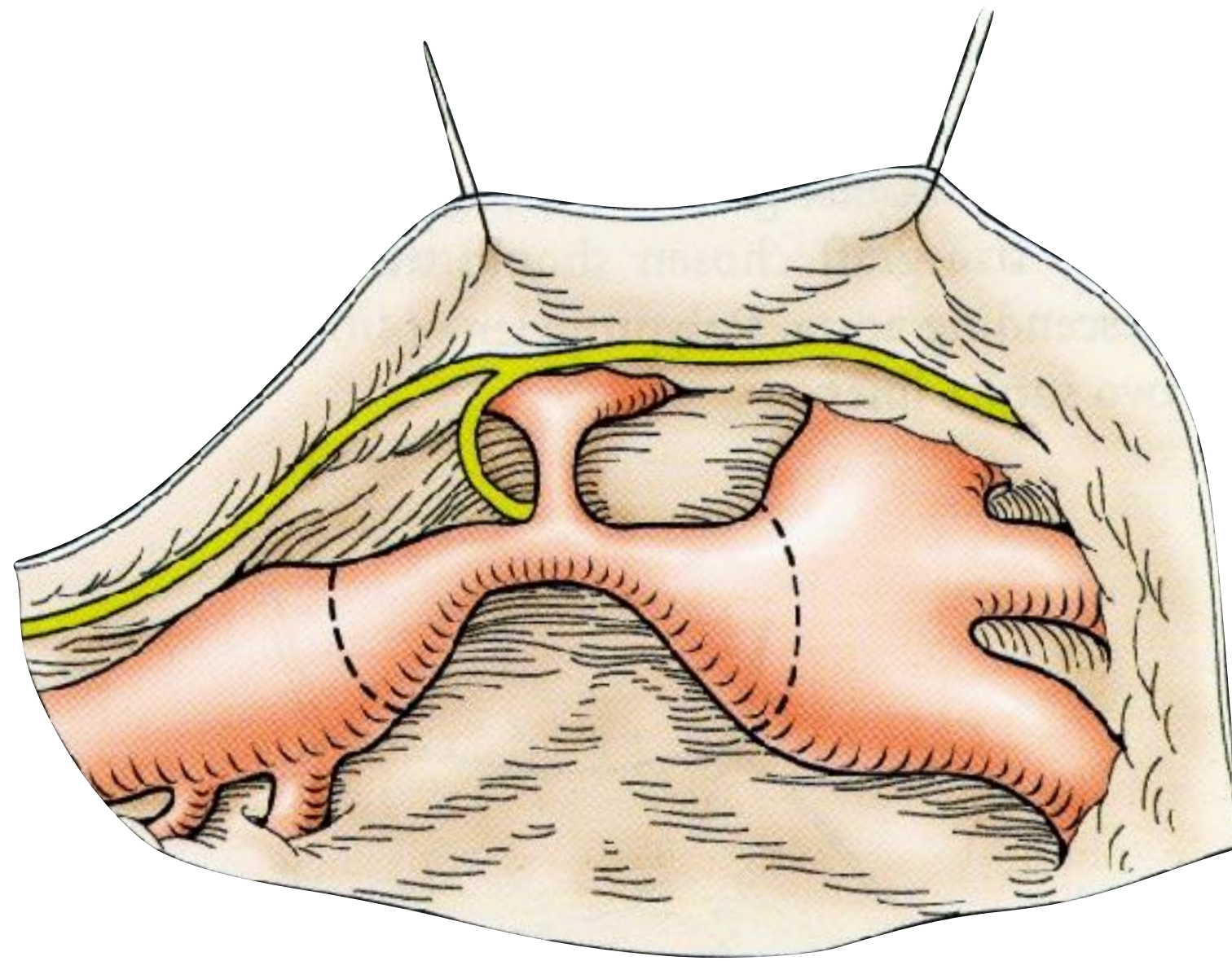


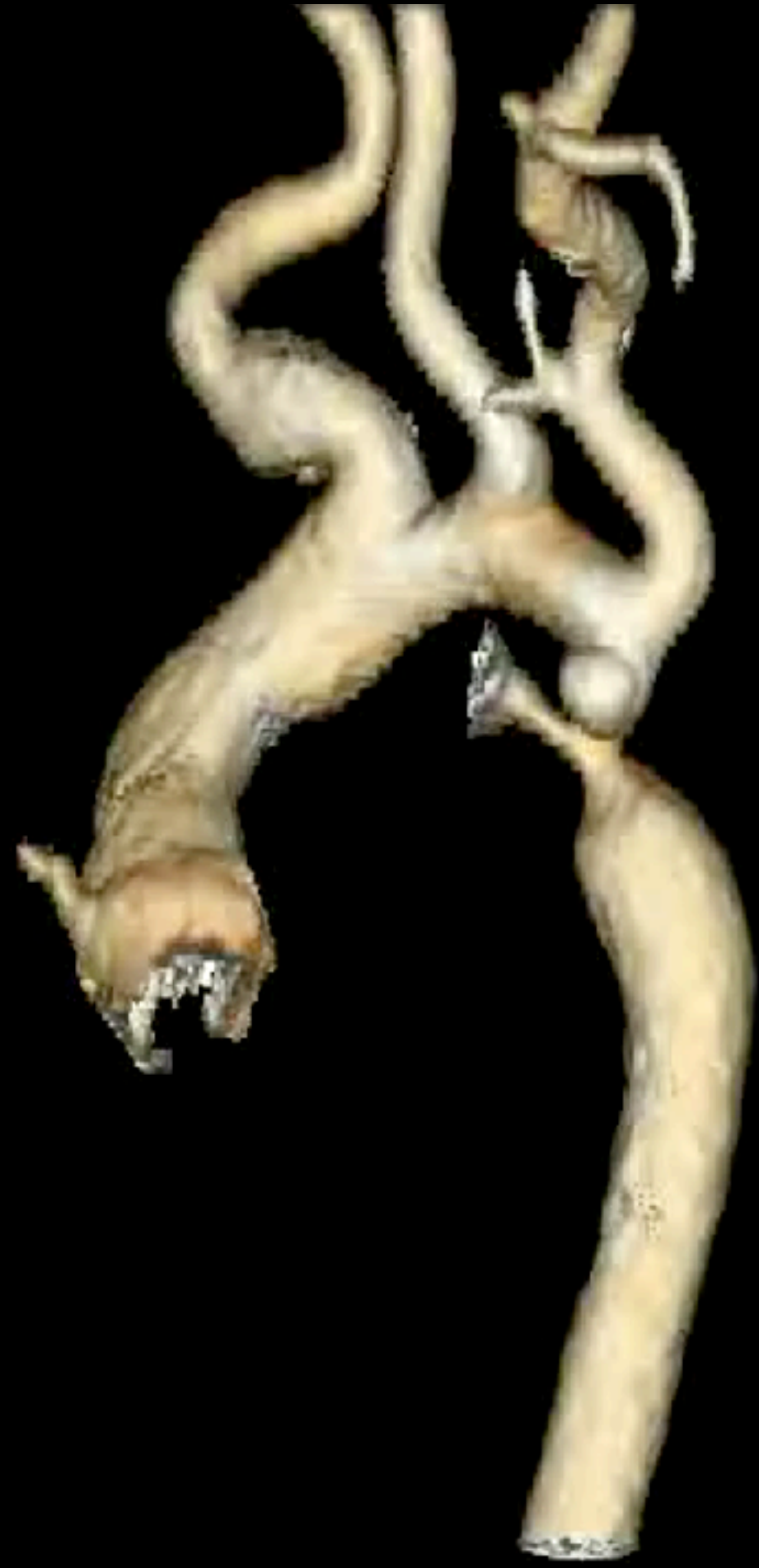


**Fig. 1.** Aortic arch anatomy in neonates.



*Fig 1. Minimal relative diameters of the normal thoracic aorta (AO) in infancy. (PA = pulmonary artery.) (Adapted from Moulaert AJ, Bruins CC, Oppenheimer-Dekker A. Anomalies of the aortic arch and ventricular septal defects. Circulation 1976;53:1101-5, by permission.)*





**What are the useful characteristics of the aortic arch to do/  
predict optimal repair ?**

**3 bis - Is there a need to extend the repair to the horizontal aorta ?**

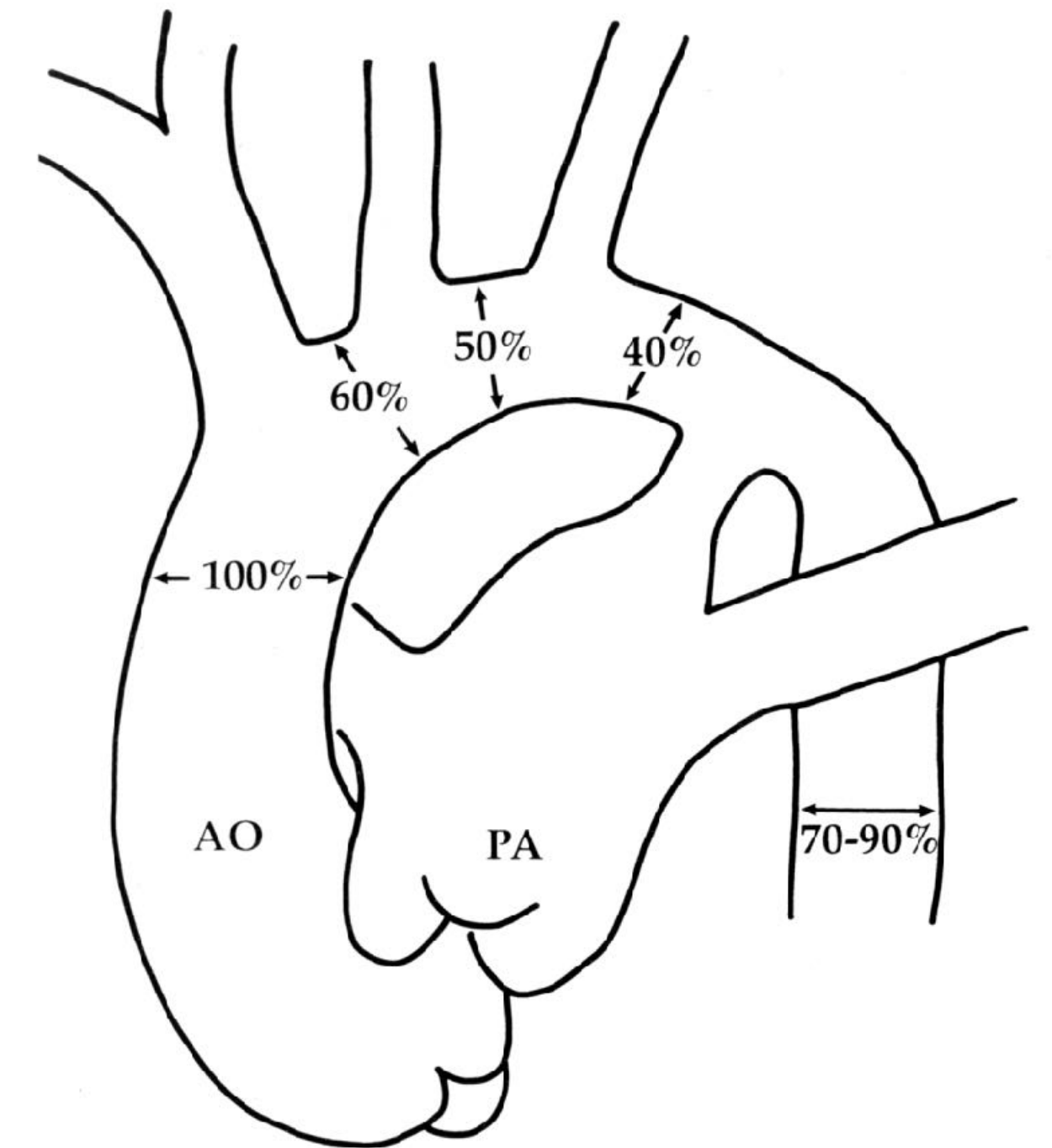
What is the definition of hypoplastic aortic arch/segment ?

Will hypoplasia of the aortic arch create flow obstruction if not treated ?

Which segment is really important to measure ?

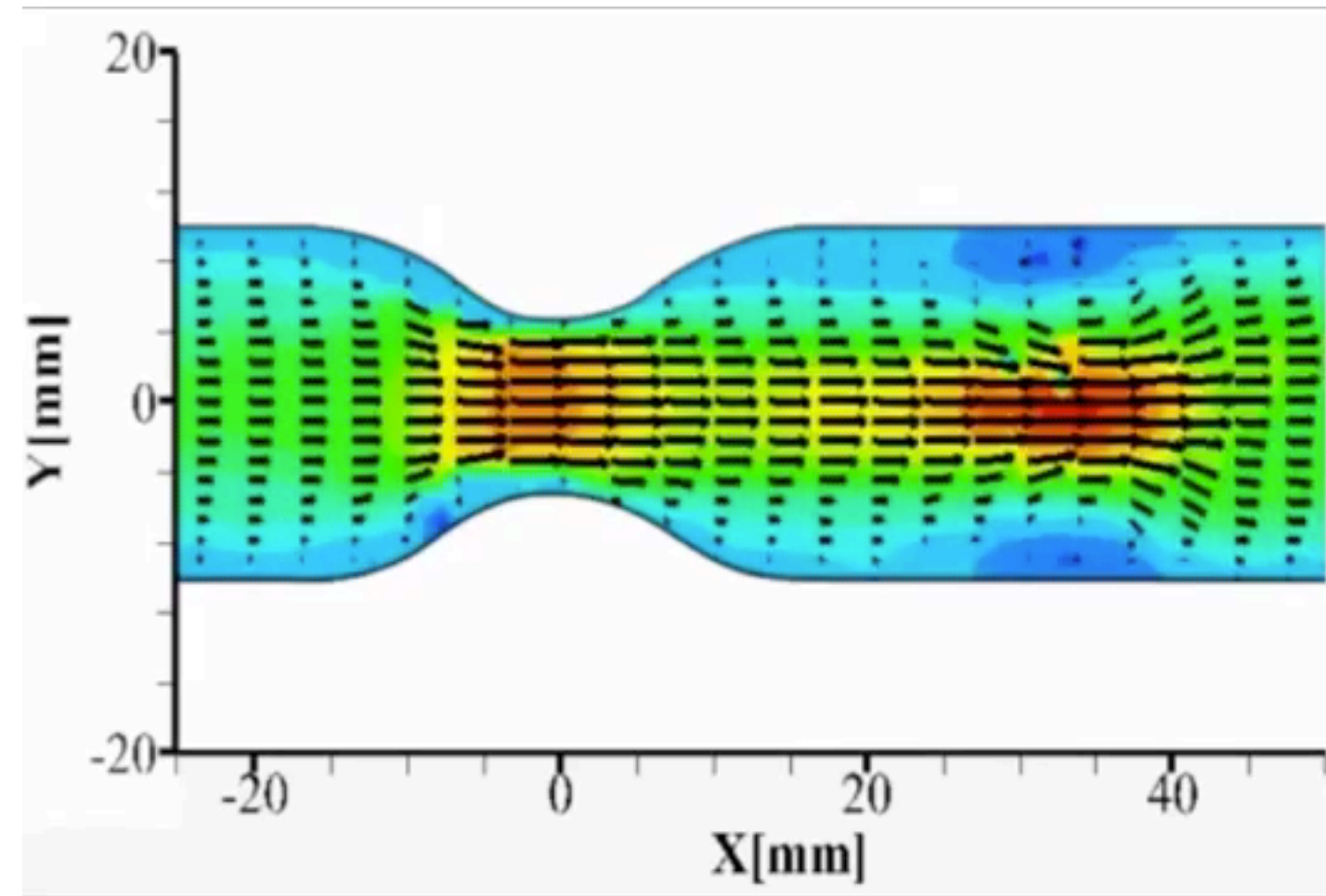
# Definition(s) of hypoplastic aortic arch

1. The ratio of transverse arch to ascending aorta  $< 50\%$   
because of the decreased aortic blood flow, the size of the ascending aorta itself may be smaller than normal in newborns presenting with coarctation.
2. Transverse arch diameter is less than the patient's body weight in kilograms plus 1
3. The aortic arch is hypoplastic if the Z value is  $-2$

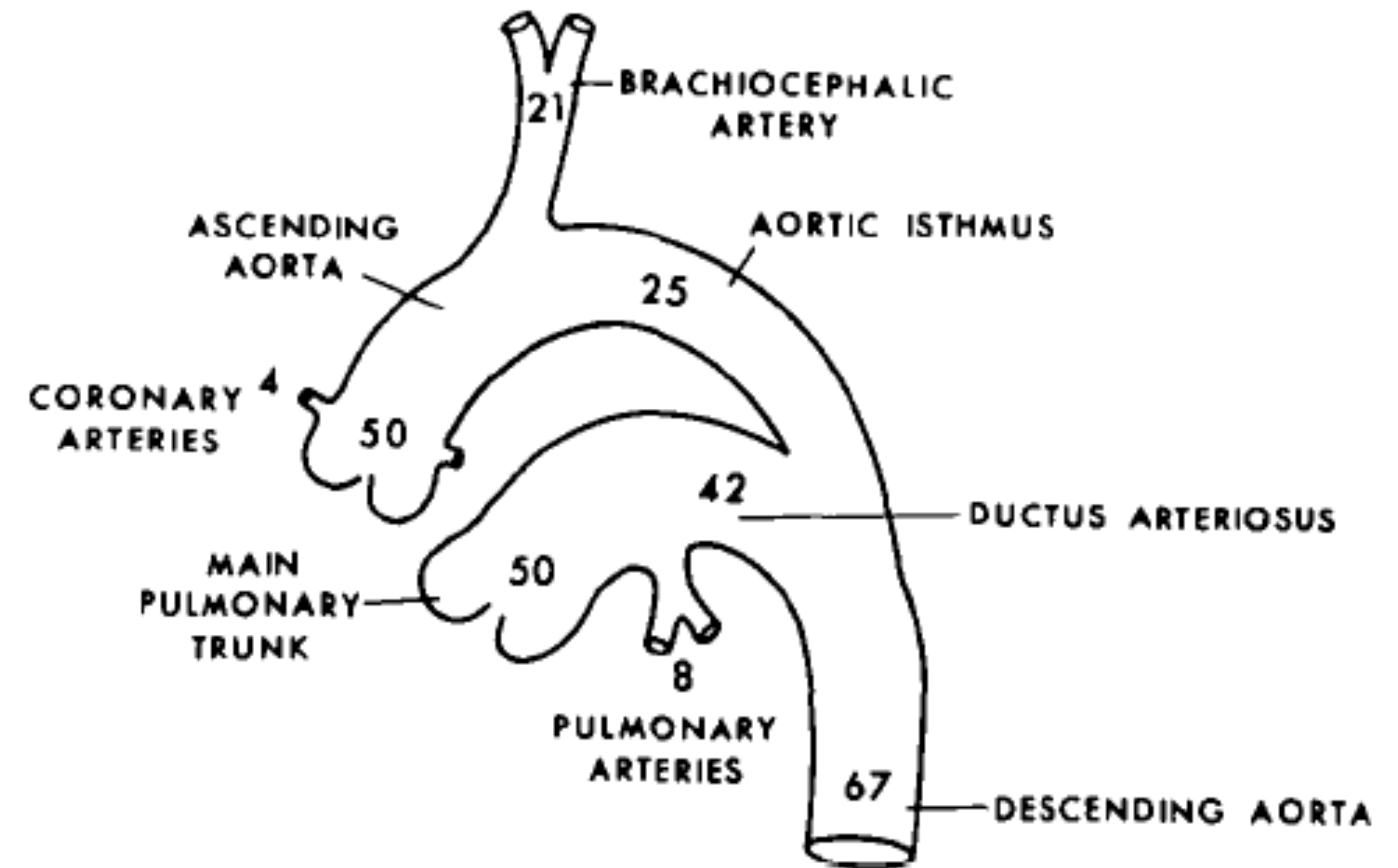


Lacour-Gayet F et al. J Thorac Cardiovasc Surg. 1990;100:808-16.  
Karl TR, et al. J Thorac Cardiovasc Surg 1992;104:688-95.  
Cantinotti M, et al. J Am Soc Echocardiogr. 2014;27: 179-191.

# Will hypoplasia of the aortic arch create flow obstruction ?



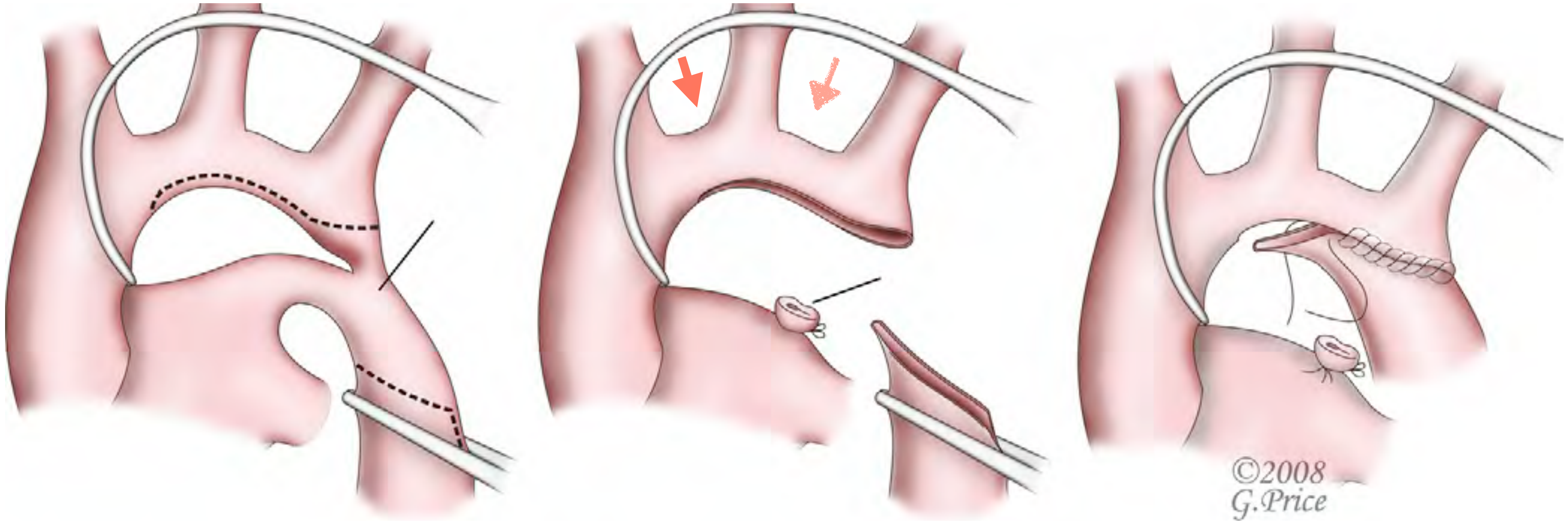
1. The hemodynamic molding theory predicts dimension of the aortic arch
2. The majority (> 50%) of newborn with hypoplastic aortic arch will not have obstruction after coarctation repair





# Extended end-to-end anastomosis : Which segment is important ?

Is it technically feasible ?



**Only one very precise question for size.**

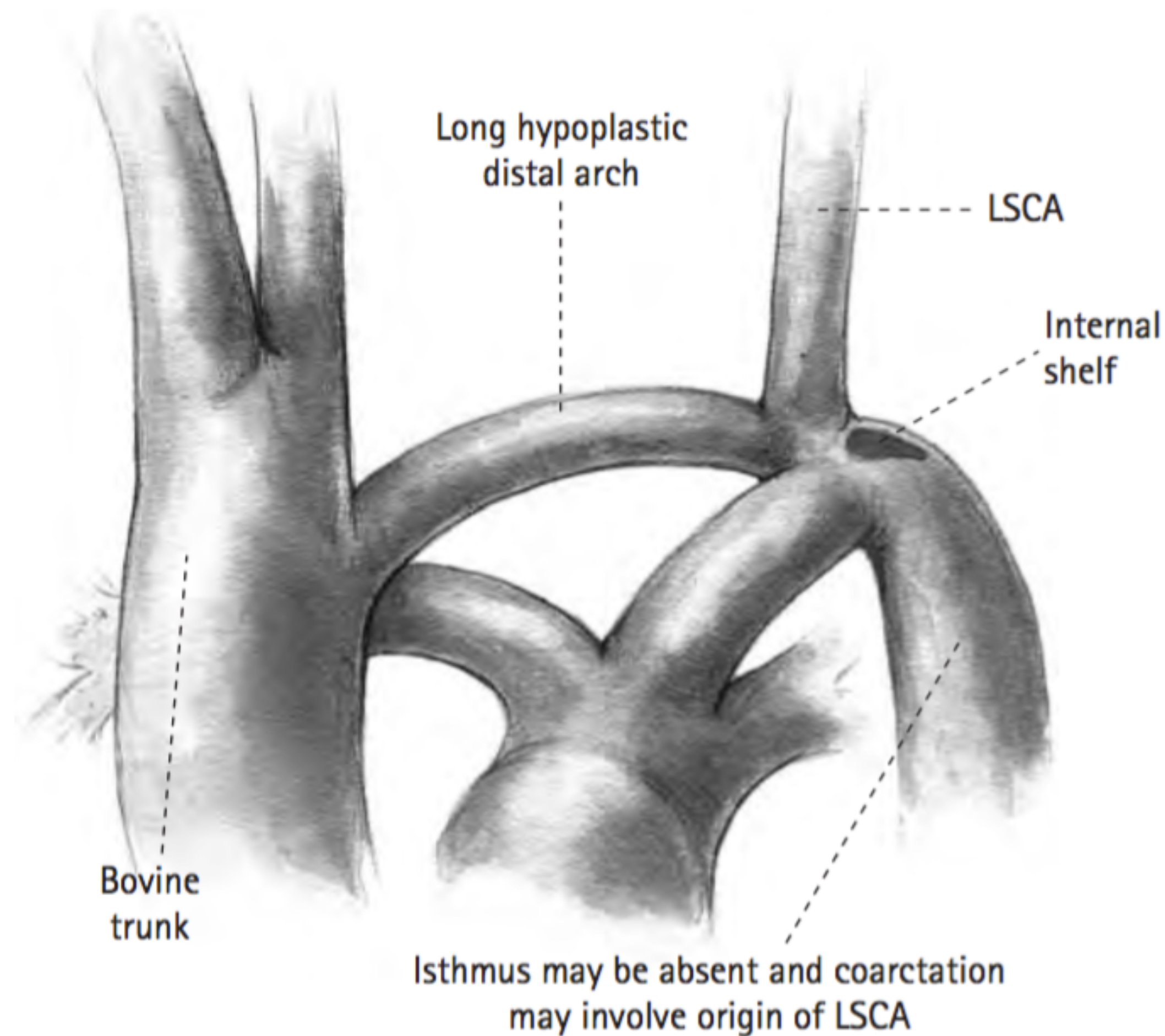
**What is the size of the proximal segment of the aortic arch ?**

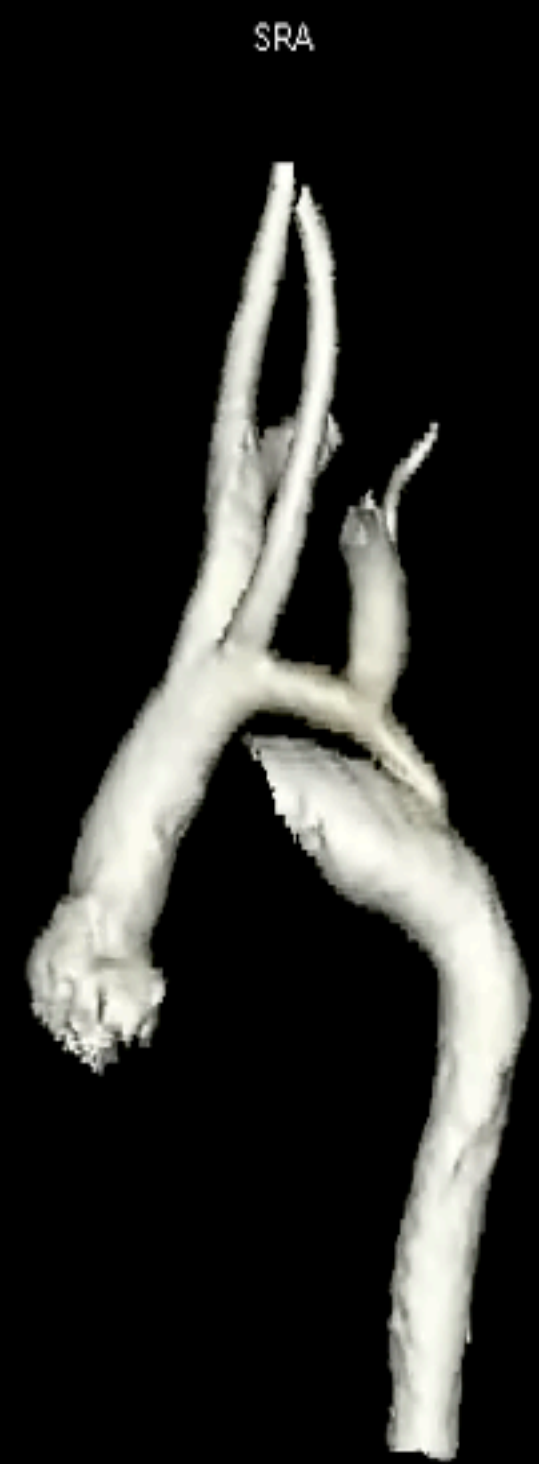
1. Proximal segment of the transverse arch diameter is less than the patient's body weight **in kilograms**
2. The proximal segment is too small to be adequately repaired through a thoracotomy if the **Z value is -4.5 to -6**

**Is the size sufficient to take the decision ?**

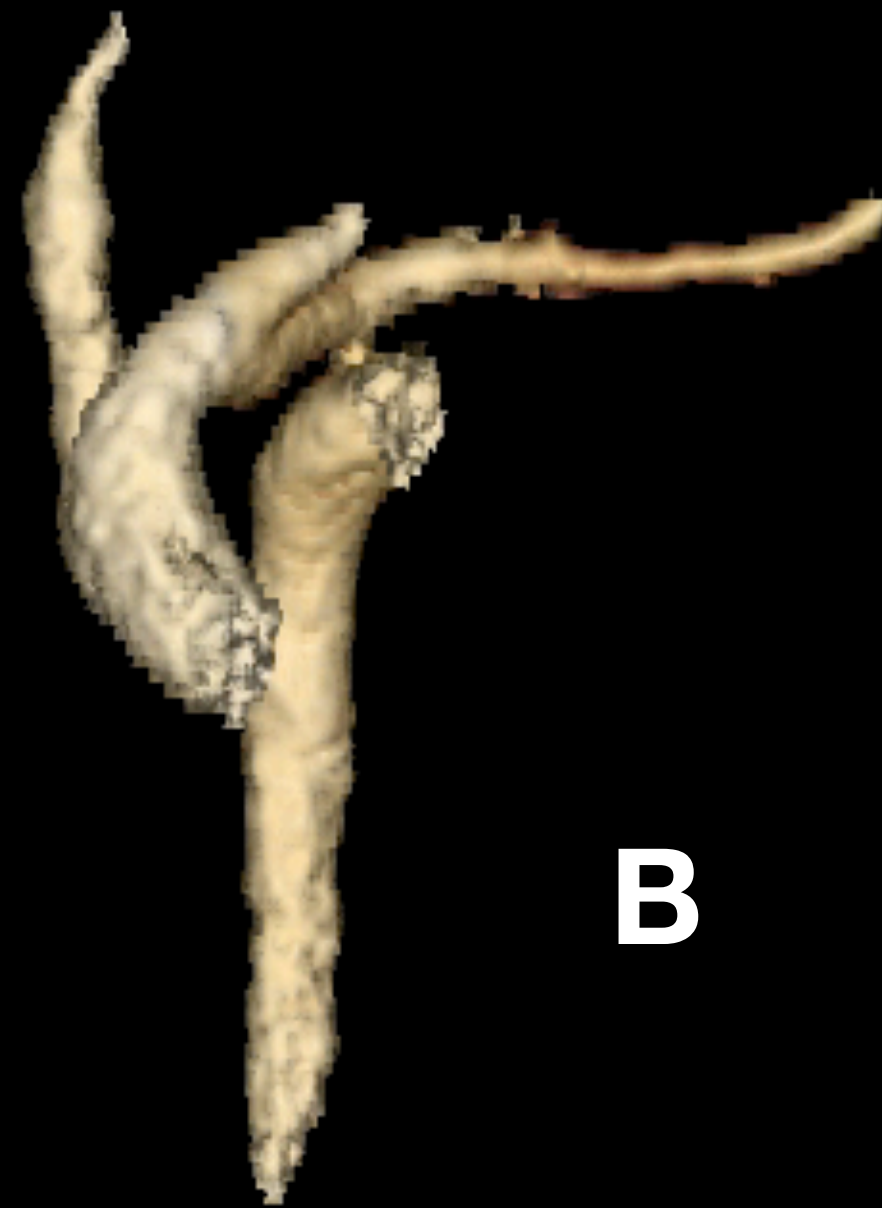
**No**

**Distribution of the brain vessels influences surgical strategy**

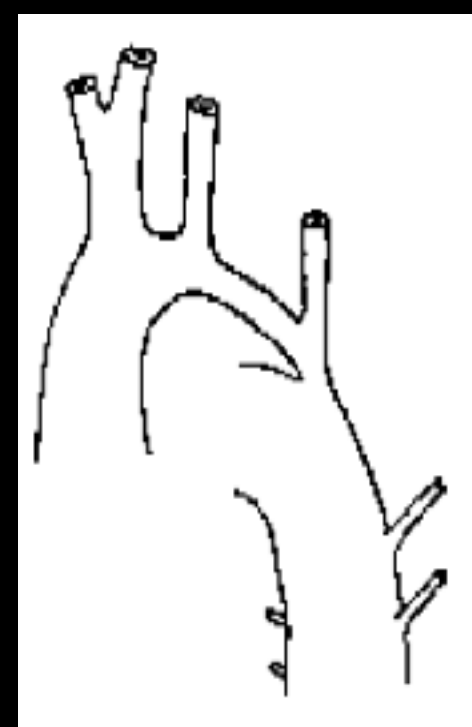




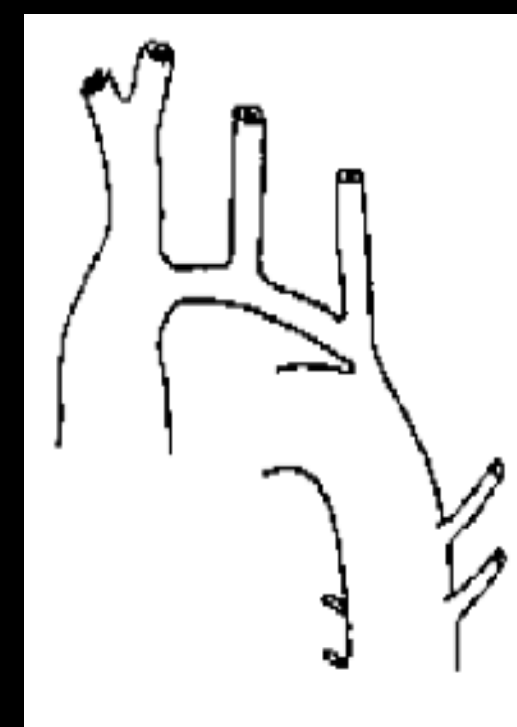
**A**



**B**



**C**

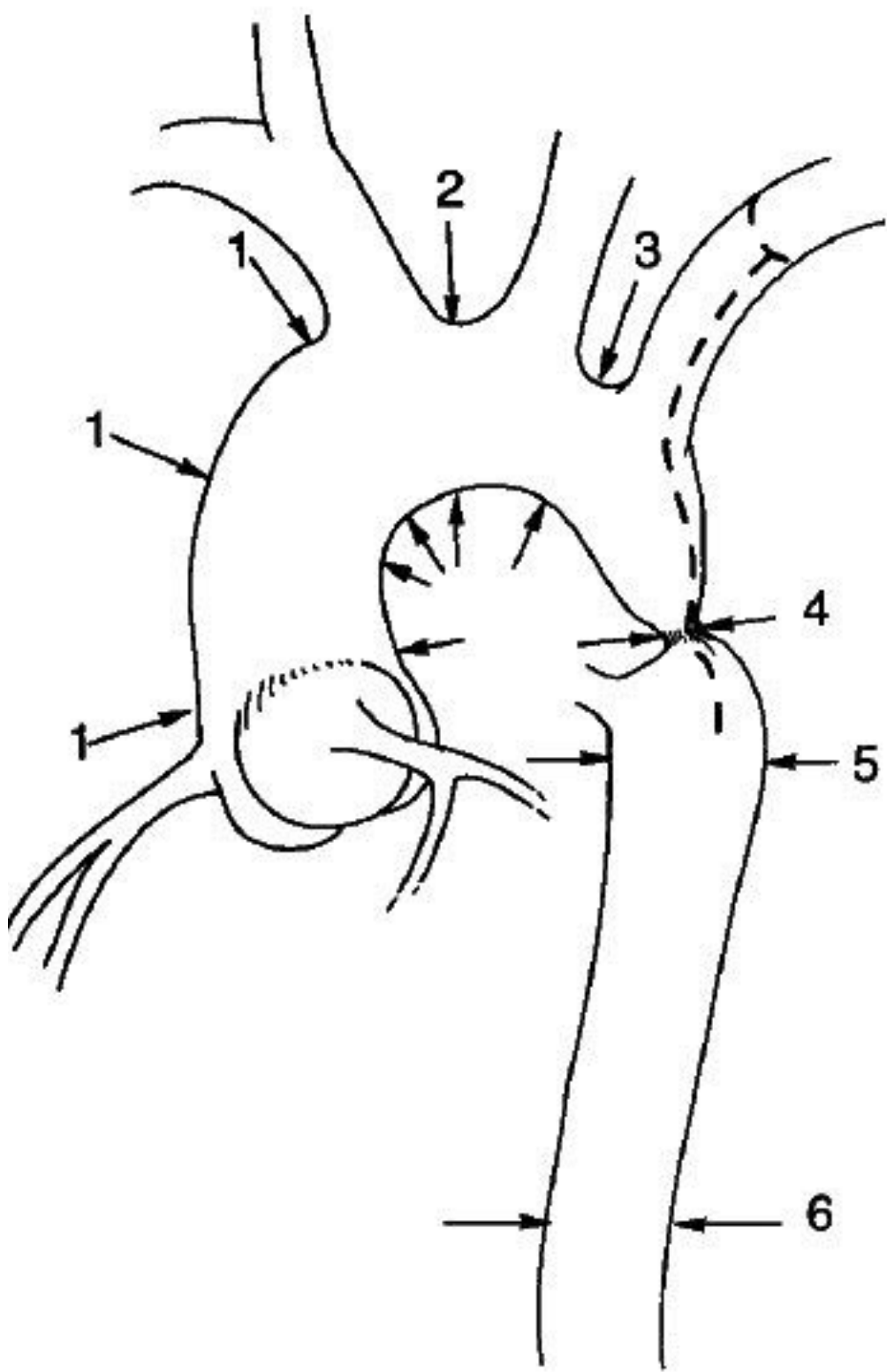


L  
7  
0

# What is (are) the objective(s) of choosing the best technique for repair ?

*No residual obstruction - No recurrent obstruction*

**4 - If the proximal segment is hypoplastic and is not enlarged, will it grow ?**



*Table 4. Aortic Growth in Patients Less Than One Month of Age at Time of Surgical Intervention (Group I)*

Region	Preoperative	Postoperative	% Change	p Value (pre vs post)
2	0.59 ± 0.10	0.68 ± 0.07	+13 ± 16	0.05
3	0.49 ± 0.06	0.67 ± 0.09	+23 ± 14	<0.001
4	0.23 ± 0.08	0.65 ± 0.24	+57 ± 17	<0.001
5	0.98 ± 0.11	0.82 ± 0.13	-21 ± 14	<0.02
6	0.81 ± 0.11	0.70 ± 0.06	-14 ± 14	<0.02

A

# What is (are) the objective(s) of choosing the best technique for repair ?

## No residual obstruction - No recurrent obstruction

### 4 - If the proximal segment is hypoplastic and is not enlarged, will it grow ?

Table 4. Changes in Left-Sided Heart Structure Measurements in the THAA Group

Measurement	Preoperative	At Discharge	Follow-Up	<i>p</i> <sup>a</sup>
Transverse aortic arch				
PAA z-score	-4.63 (-6.07 to -3.77) <sup>b</sup>	-2.58 (-3.73 to -0.75) <sup>c</sup>	-1.17 (-2.46 to -0.15) <sup>d</sup>	<0.001
DAA z-score	-4.84 (-6.34 to -4.02) <sup>b</sup>	-2.67 (-4.19 to -0.91) <sup>e</sup>	-1.04 (-2.65 to 0.42) <sup>d</sup>	<0.001
Ascending aorta z-score	-0.25 (-1.32 to 0.73) <sup>f</sup>	0.82 (-0.28 to 1.66) <sup>g</sup>	1.31 (0.48 to 2.10) <sup>d</sup>	0.003
Isthmus z-score	-6.16 (-7.54 to -4.67) <sup>b</sup>	-1.42 (-2.67 to 1.15) <sup>e</sup>	0.40 (-0.93 to 1.92) <sup>d</sup>	<0.001
Descending aorta z-score	-1.82 (-3.27 to -0.26) <sup>b</sup>	-0.57 (-1.83 to 0.49) <sup>h</sup>	0.75 (-0.59 to 1.59) <sup>d</sup>	<0.001
Aortic valve z-score	-4.00 (-5.20 to -2.41) <sup>i</sup>	-3.02 (-4.40 to -1.39) <sup>e</sup>	-0.41 (-1.15 to 0.70) <sup>d</sup>	<0.001
Mitral valve z-score	-2.97 (-4.43 to -1.62) <sup>j</sup>	-2.58 (-4.08 to 1.28) <sup>k</sup>	-1.98 (-2.87 to -0.83) <sup>d</sup>	0.004
LVEDD z-score	-3.41 (-5.26 to -1.61) <sup>b</sup>	-1.61 (-3.33 to -0.31) <sup>e</sup>	0.29 (-0.50 to 1.66) <sup>d</sup>	<0.001

<sup>a</sup> Post-hoc tests conducted if *p* < 0.05. <sup>b</sup> *p* < 0.001 vs discharge. <sup>c</sup> *p* = 0.002 vs follow-up. <sup>d</sup> *p* < 0.001 vs preoperative. <sup>e</sup> *p* < 0.001 vs follow-up. <sup>f</sup> *p* = 0.002 vs discharge. <sup>g</sup> *p* < 0.014 vs follow-up. <sup>h</sup> *p* = 0.011 vs follow-up. <sup>i</sup> *p* = 0.001 vs discharge. <sup>j</sup> *p* = 0.034 vs discharge. <sup>k</sup> *p* = 0.102 vs follow-up.

DAA = distal aortic arch; LVEDD = left ventricular end-diastolic dimension; PAA = proximal aortic arch; THAA = transverse hypoplastic aortic arch.

**What is (are) the objective(s) of choosing the best technique for repair ?**

***No residual obstruction - No recurrent obstruction***

**4 - If the proximal segment is hypoplastic and is not enlarged, will it grow ?**

**Yes, but not in all patients**

Patients with moderately hypoplastic arch (Z-score -2 to -5) treated by conventional coarctation repair have **adequate growth of the distal arch** demonstrated at long-term follow-up, **but one-third of them keep a small proximal arch.**

# What are the useful characteristics of the aortic arch to do/ predict optimal repair ?

## 5 - Do anatomical characteristics predict recoarctation ?

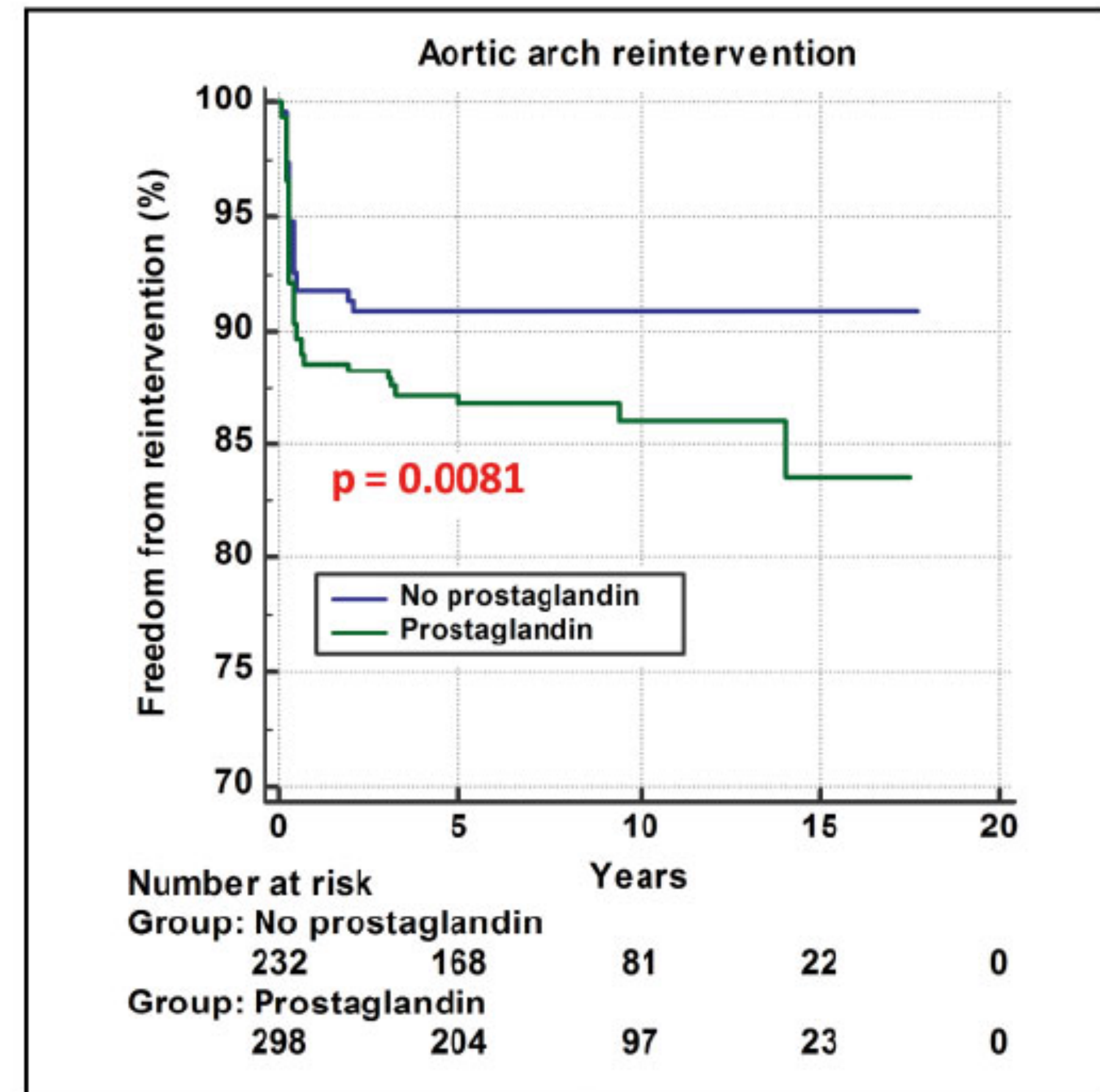
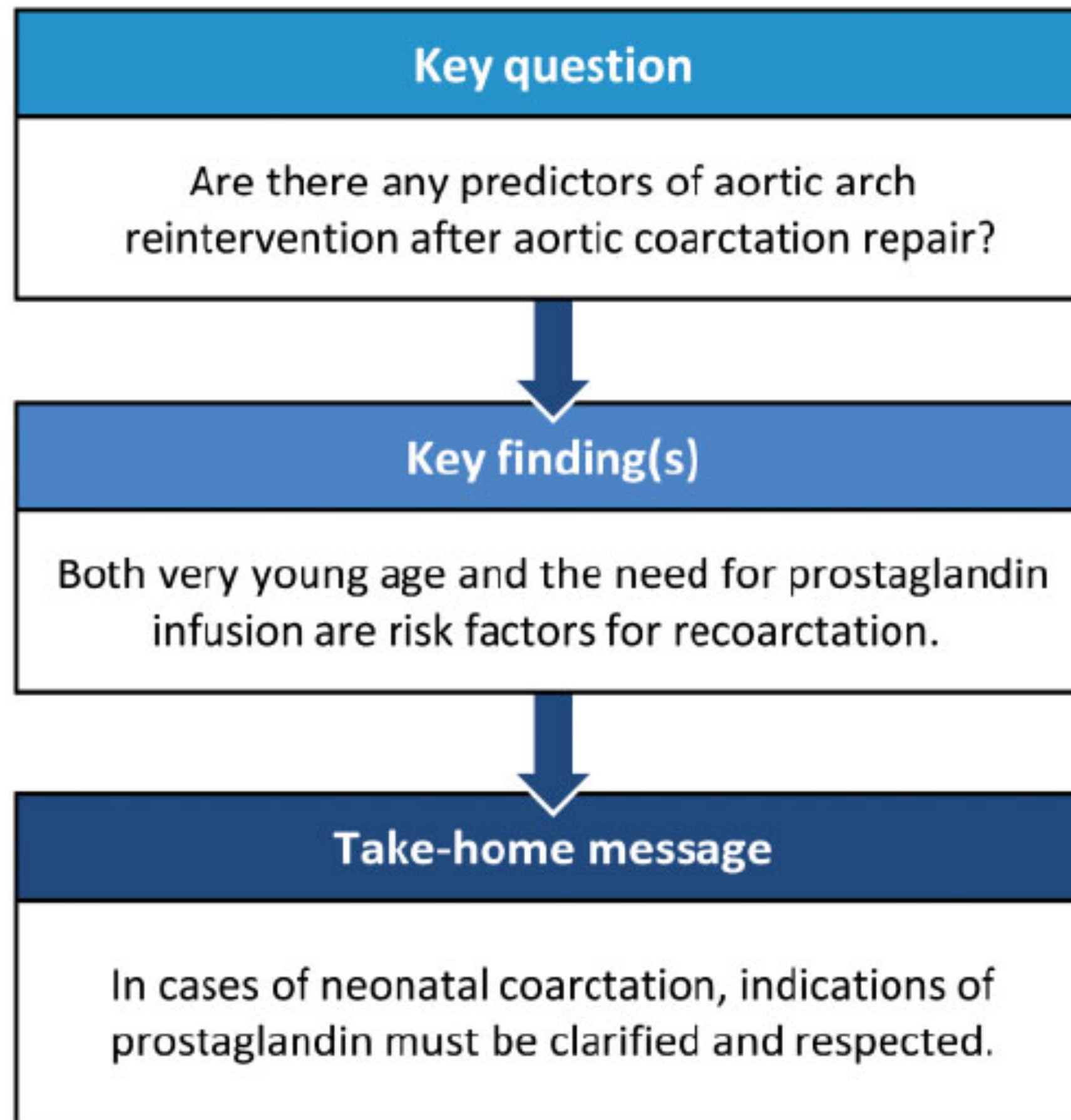
1. Proximal arch diameter below than the patient's body weight in kilograms plus 1  
**AND**
2. The aortic arch Z value below  $< -4$

**does not predict re-coarctation**



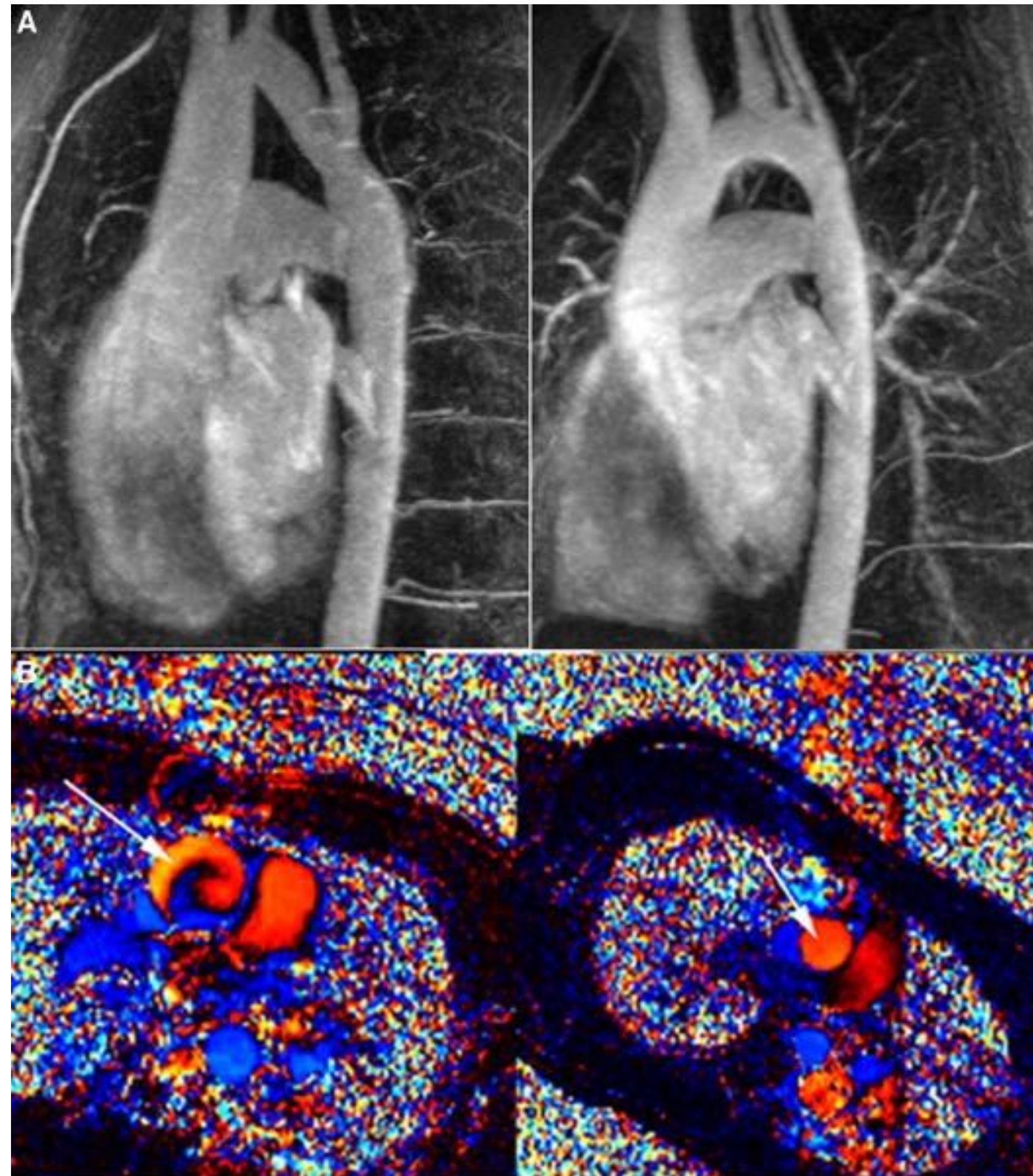
# Use of PGE1 in coarctation of the aorta

## Repair of coarctation with PGE1 increases the risk of re-coarctation



# What are the useful characteristics of the aortic arch to do/ predict optimal repair ?

## 5 - Do anatomical characteristics predict cardiovascular outcomes ?

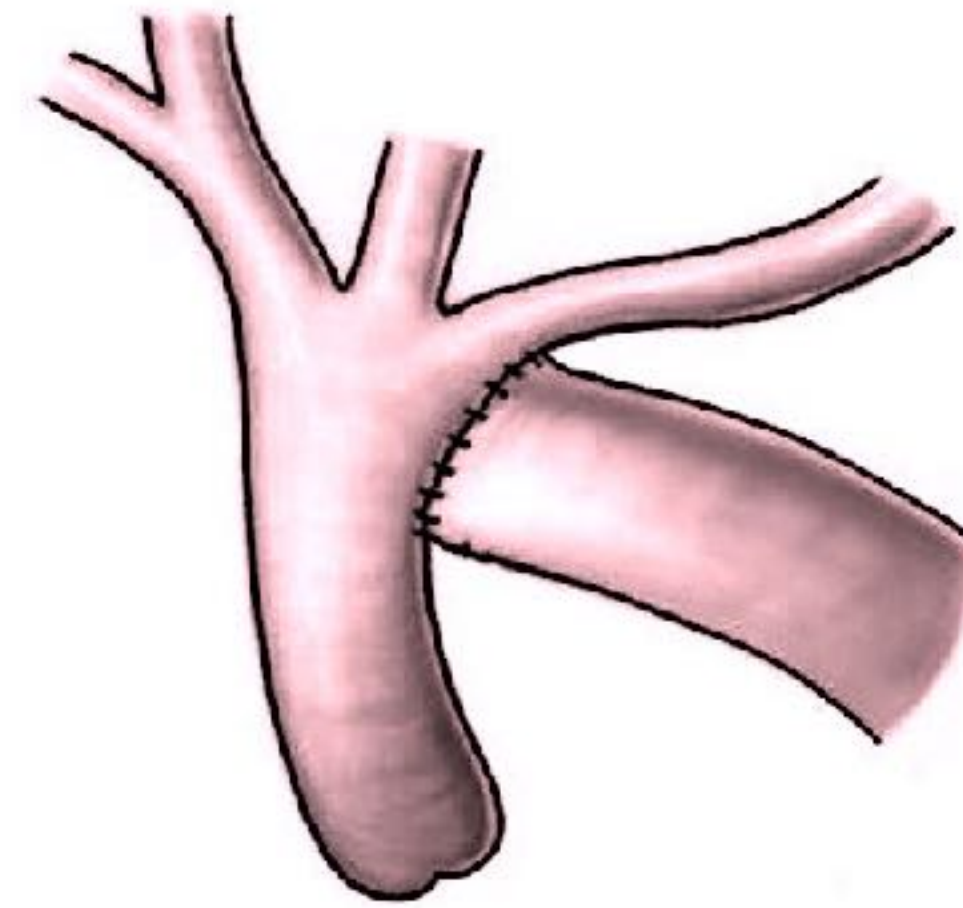
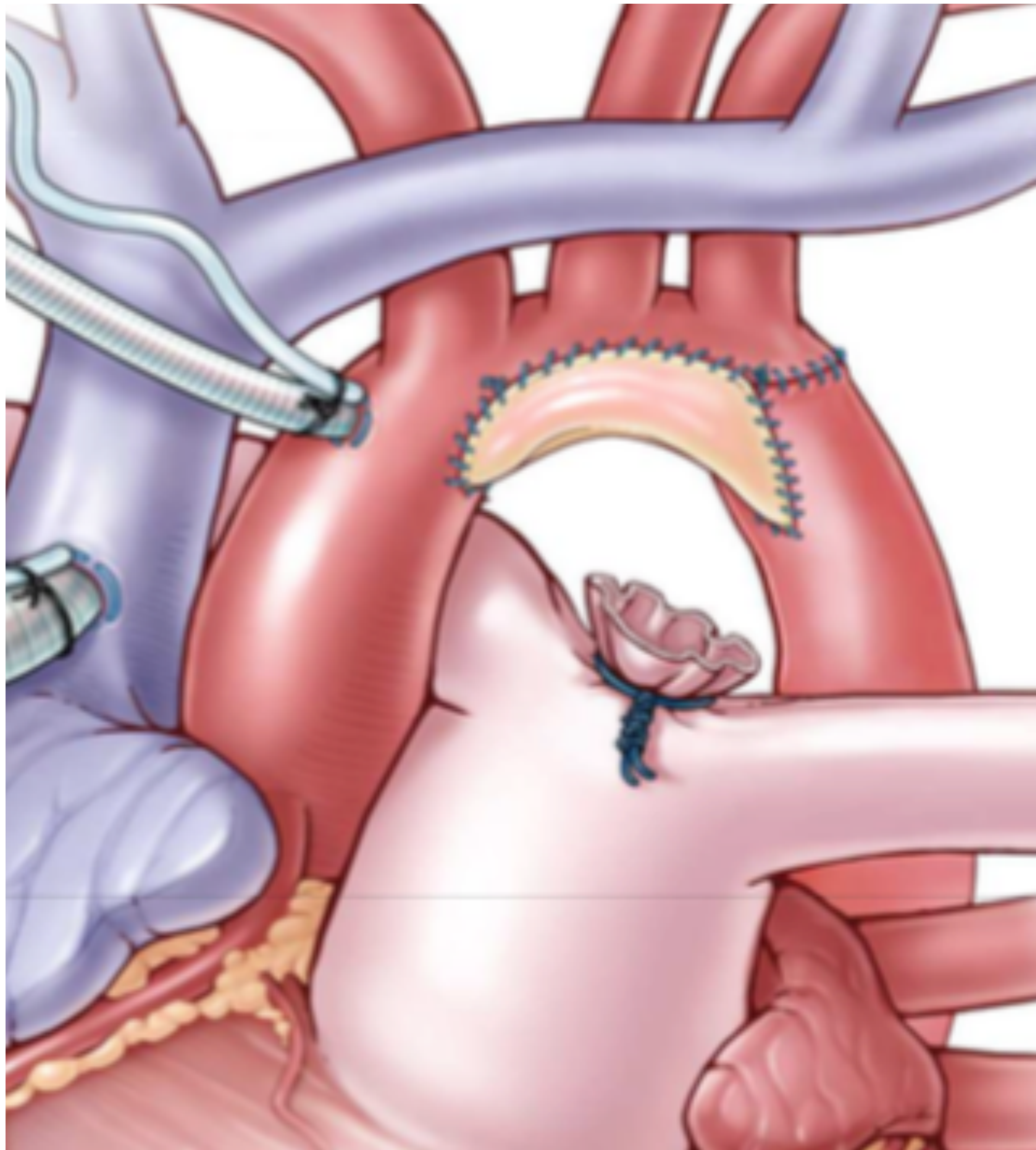


### **Gothic vs. Romanesque aortic arch geometry**

- Risk of Systemic hypertension
- Increases afterload for Left ventricle
- Favors aortic root dilatation

Ou P, et al. J Thorac Cardiovasc Surg 2008;135:62-68

Szopos M et al. J Thorac Cardiovasc Surg. 2014;148:1572-82.



No available data to compare the postoperative geometry with the different technique for repair

**Mix of**

- preoperative geometry (size, distribution of vessels)
- immediate postoperative anatomy
- growth

# What are the useful characteristics of the aortic arch to do/ predict optimal repair ?

## **6 - In case of doubt, should aortic arch be repaired through sternotomy with DHCA or Selective cerebral perfusion ?**

Re-intervention rate is comparable between sternotomy and thoracotomy. Thoracotomy is considered a risk factor for reoperation probably because « non-perfect » evaluation of aortic arch anatomy.

# Take-home messages

- 1- The vast majority of isolated coarctation of the aorta can be adequately repaired through thoracotomy using extended end-to-end anastomosis with excellent short term results.
- 2- "Very small" proximal segment of aortic arch and distribution of brain vessels (length of segments) not allowing adequate aortic cross-clamping indicate repair through sternotomy.
- 3- Residual coarctation is mainly related to inadequate evaluation of anatomy and/or poor growth of proximal segment of aortic arch.
- 4- Postoperative anatomy has an important role in predicting cardiovascular outcomes after coarctation repair. Improving aortic arch geometry is the next challenge.



Collective ignorance is our motivation  
Curiosity is our strength  
Research is our path

Individual experience is the brake  
Indifference is the weakness  
Argument from authority is the threat