전공의 연수강좌
2017.05.26

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Aortic arch obstruction

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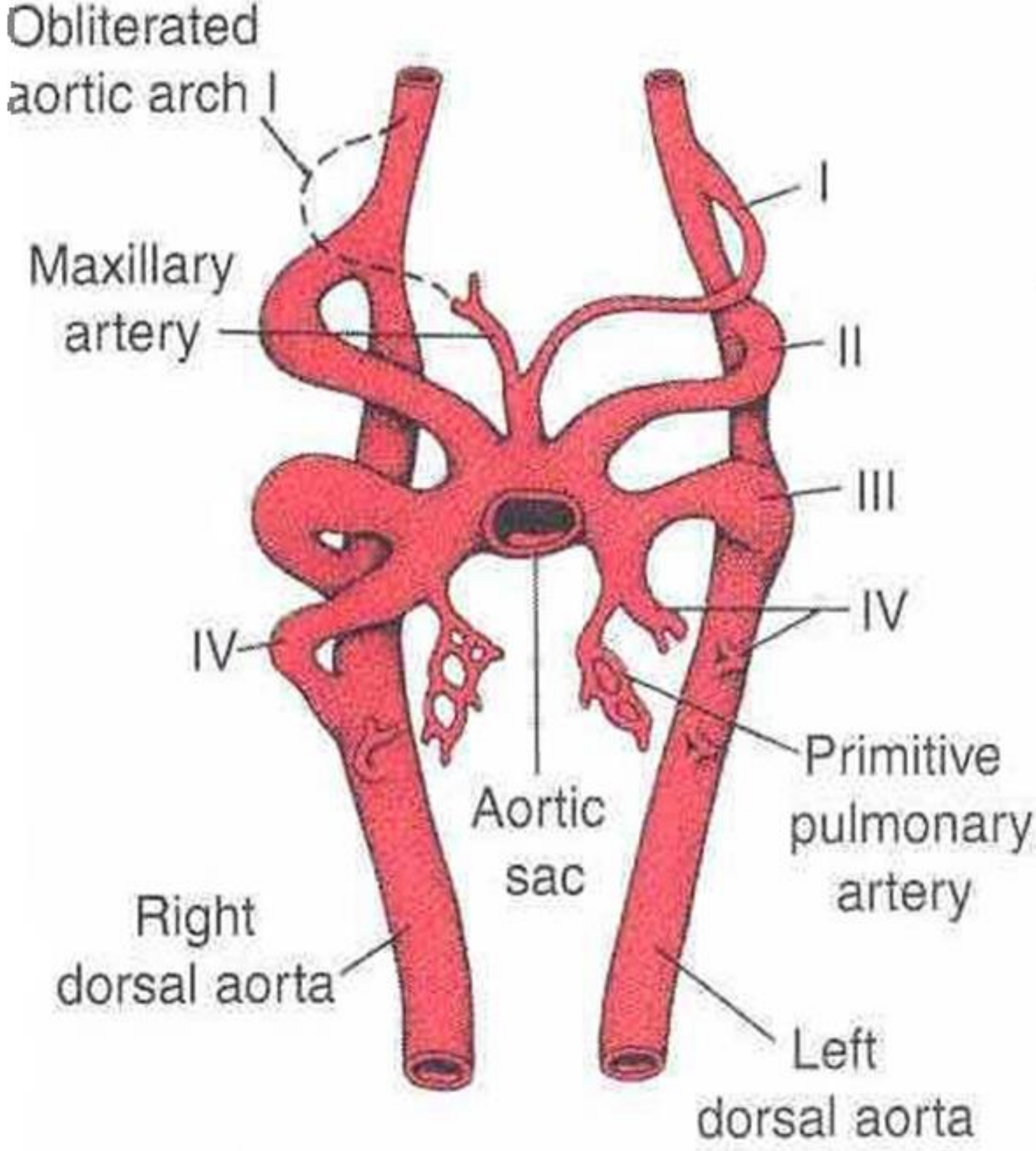
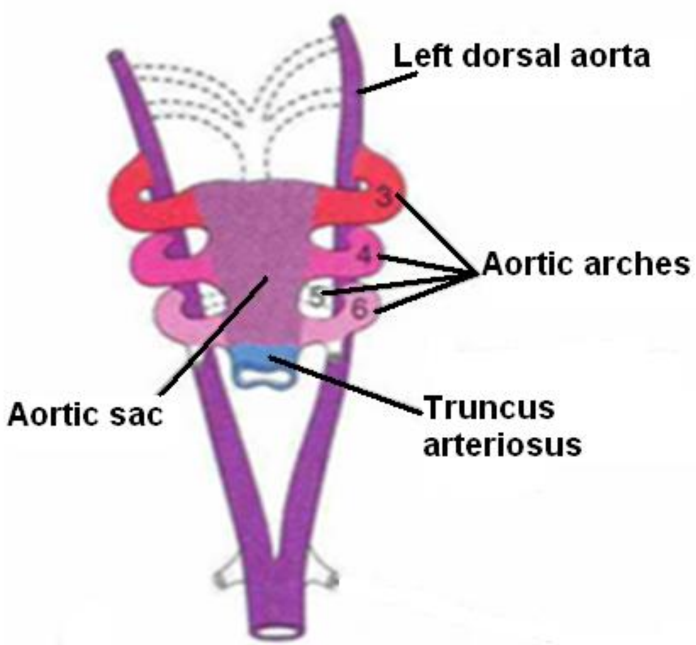
Derivatives of Aortic Arches

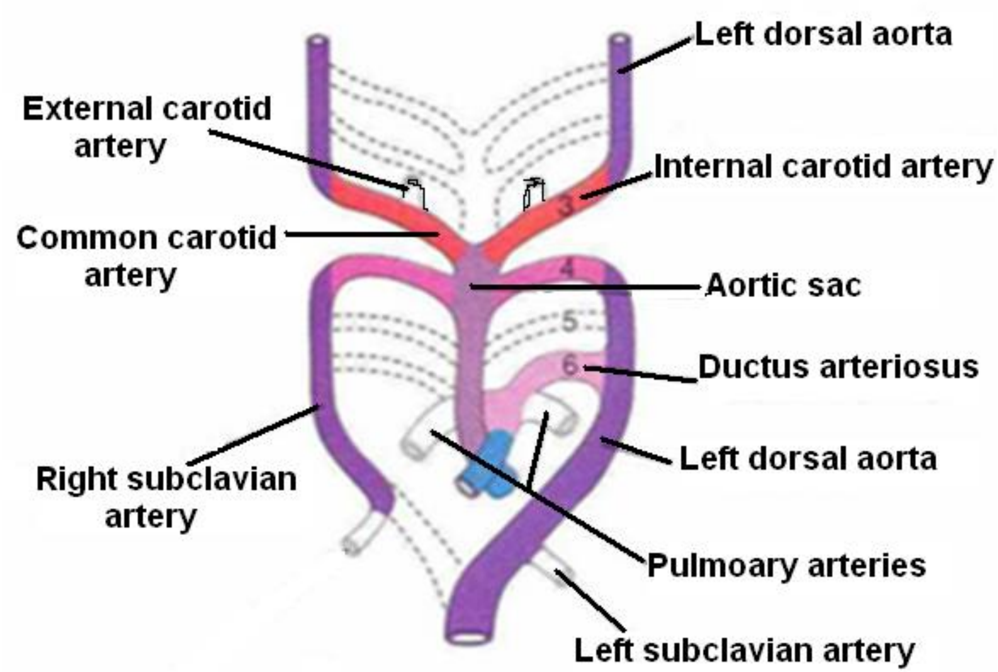


Obliterated aortic arch I
Maxillary artery
I
II
III
IV
IV
Primitive pulmonary artery
Aortic sac
Right dorsal aorta
Left dorsal aorta

Third Pair

- Proximal part:
  - Forms the common carotid arteries
- Distal part:
  - Joins the dorsal aorta to form the internal carotid arteries

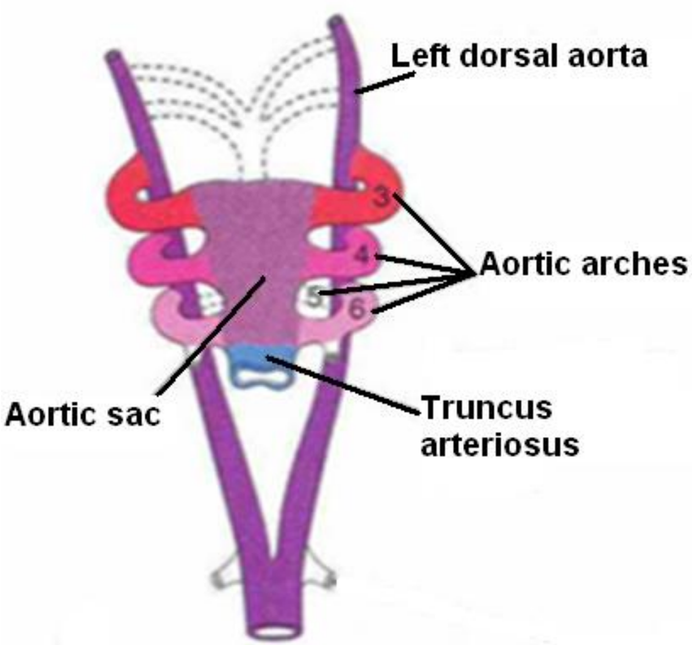


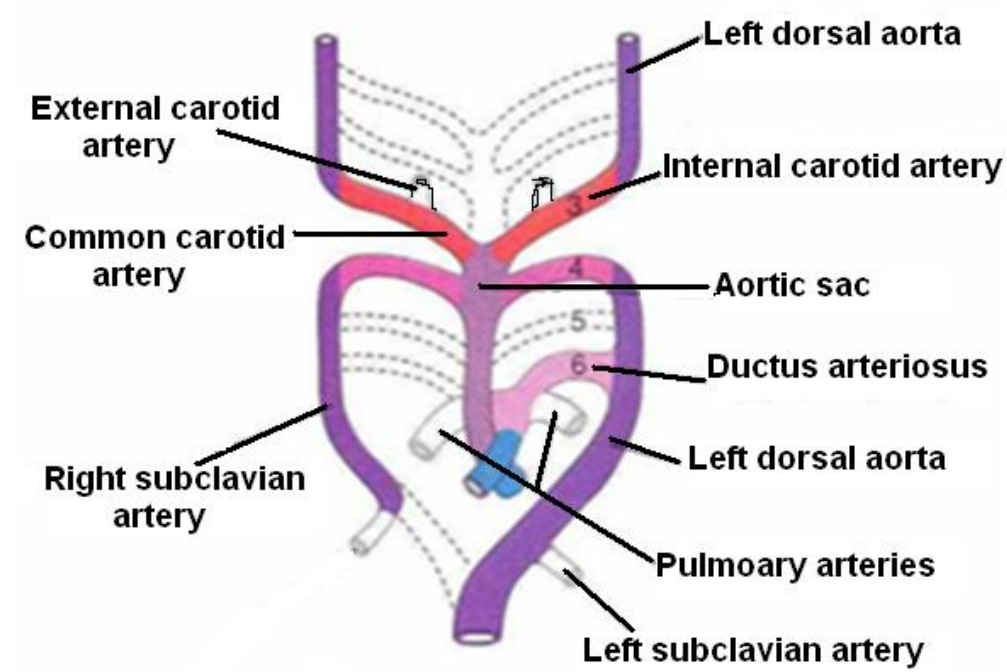


The fate of 4 & 6th pairs of aortic arches differs on the right and left side

Fourth Pair

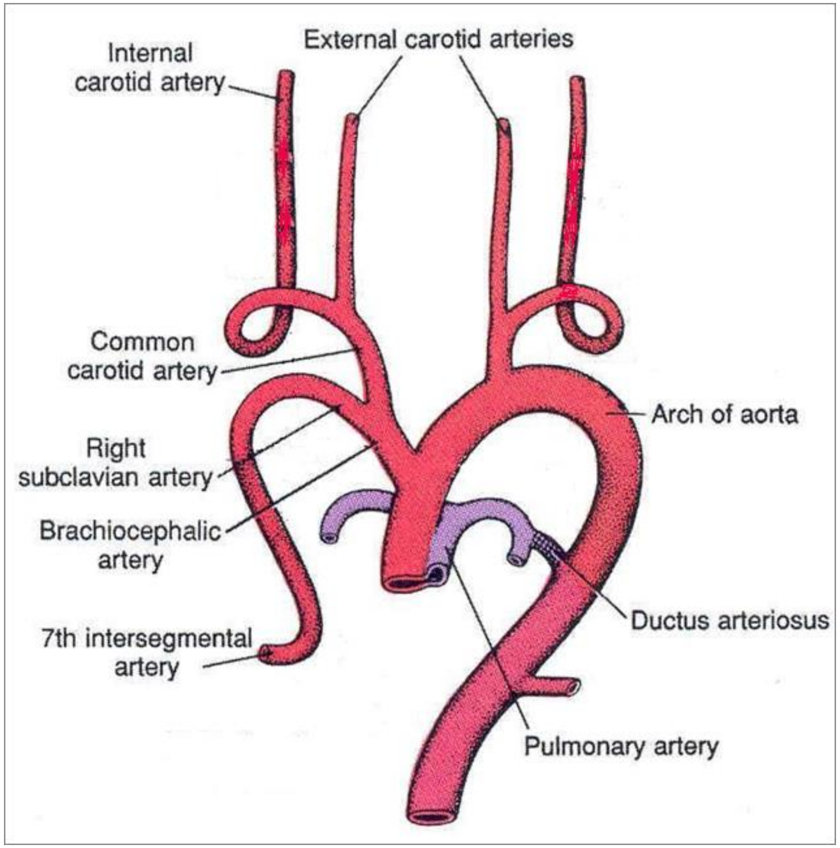
- Right:
  - Becomes the proximal part of the right subclavian artery
- Left:
  - Forms of the aortic arch





Sixth Pair

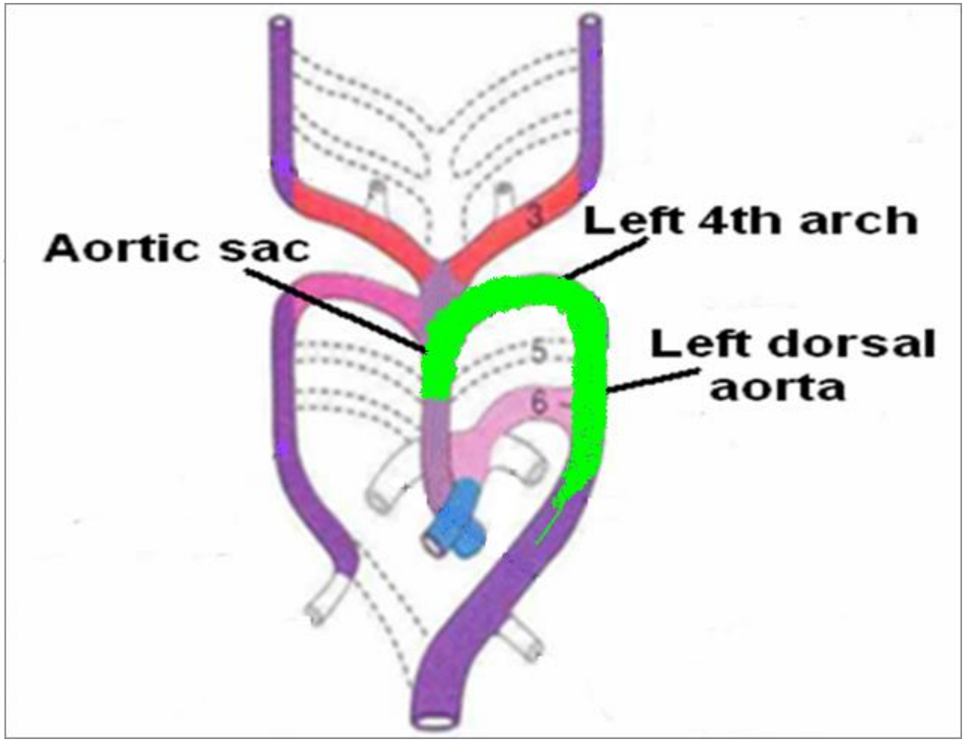
- **RIGHT**:
  - Proximal part: persists as the proximal part of the right pulmonary artery
  - Distal part: degenerates
- **LEFT**:
  - Proximal part: persists as the proximal part of the left pulmonary artery
  - Distal part: forms ductus arteriosus, a shunt between pulmonary artery and dorsal aorta



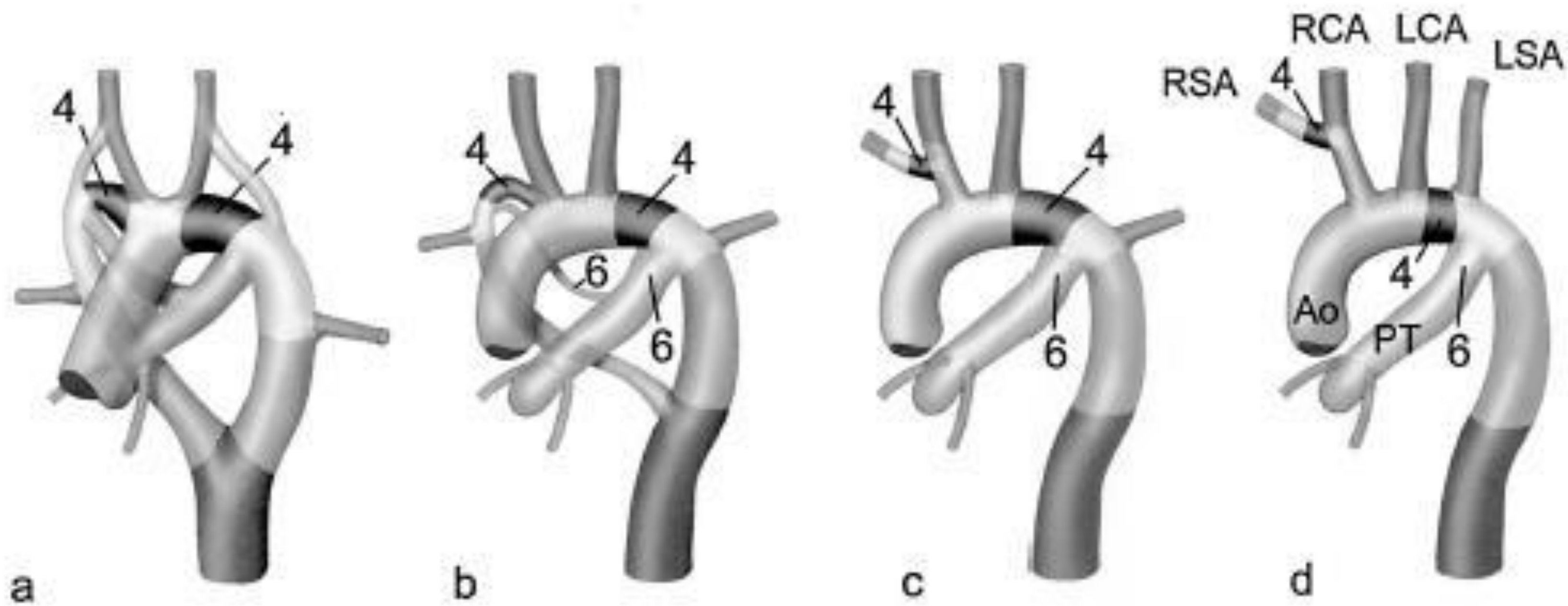
Arch of Aorta

Derived as:

- Proximal segment
  from aortic sac
- Middle segment
  from the left 4th aortic arch
- Distal segment
  from the left dorsal aorta



Development continued



Molin, D. DeRuiter, M.C, Wisse, L.J, Azhar, M., Doetschman, T., Poelmann, R. E., Gittenberger-de Groot, A. C. Altered apoptosis pattern during pharyngeal arch artery remodeling is associated with aortic arch malformations in Tgfβ2 knock-out mice. *Cardiovascular Research*. 2002; 56: 312-322.

Gittenberger-De Groot, A.C. Bartelings, M.M. Deruiter, M.C. Poelmann, R.E. Basics of Cardiac Development for the Understanding of Congenital Heart Malformations. *Pediatric Research.* 2005; 57 (2): 169-176.

Coarctation of aorta

Coarctation of Aorta

- Definition
  - Congenital narrowing of upper descending thoracic aorta **adjacent to** the site of attachment of **ductus arteriosus**
- Abnormal development of **left 4th and 6th aortic arches**
- 4~8% of CHD, Male dominant
- 30~35%, Turner syndrome

- 50~80%, Bicuspid aortic valve
- VSD
  - Posterior malalignment of conal septum
- LVOTO, Subaortic stenosis, AS, MS,
- TGA, DORV, UVH

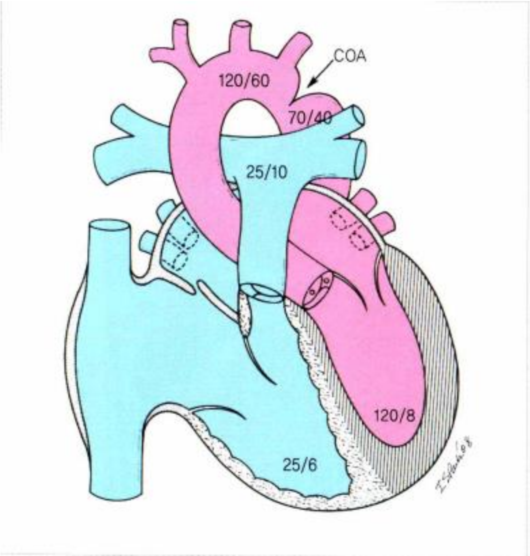
Clinical features

- **Infancy**
  - Heart Failure
    - Mostly < 3 months of life
    - A significant number < 1st week of life
  - Circulatory collapse
    - Ductal closure: ↓in lower body perfusion
    - Falling PVR: preferential flow of blood to the pulmonary circulation
    - Development of acidosis and shock

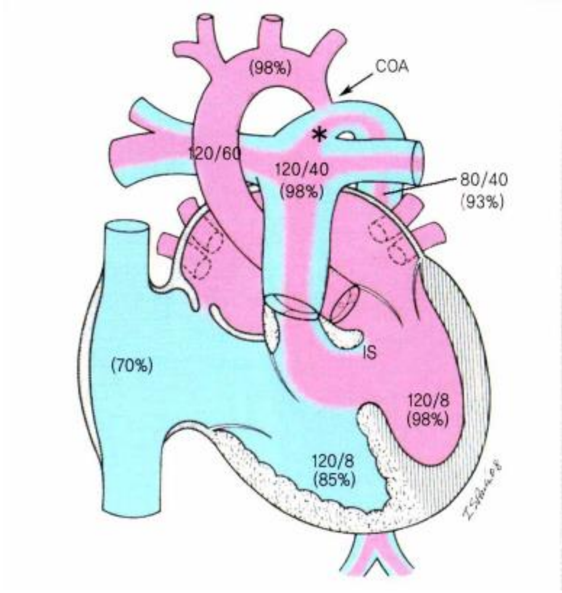
Clinical features

- **Childhood**
  - Asymptomatic
  - HTN (90%), Cardiomegaly (30%), Rib notching (15%)
- **Adolescence and adult**
  - HTN
  - Vascular heart disease
  - HF at 30 years of age

- Simple or isolated CoA



- Complex CoA, CoA with VSD, DORV, TGA



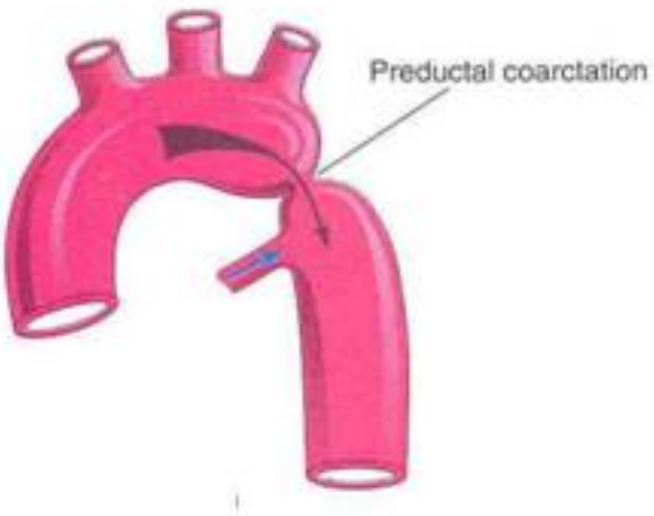
- Shone Complex

- **Simple or Isolated CoA**
  - LVH, CHF
  - Weak femoral pulse
  - Collateral vessels
  - HTN d/t Renin-angiotensin system abnormality
  - Claudication
  - Headache
  - Hypertensive encephalopathy
  - Rupture of aneurysm of cerebral vessel,
  - Dissection
  - Infective endocarditis. Related with bicuspid aortic valve

- **Complex CoA**
  - CHF d/t LR shunt lesion
  - Severe Pul. HTN
  - AKI, NEC
- Tx
  - PGE1
  - Diuretics, DGX d/t CHF
  - Ventilator
    - O2 (x) ,   PaCO2 : 40~50mmHg

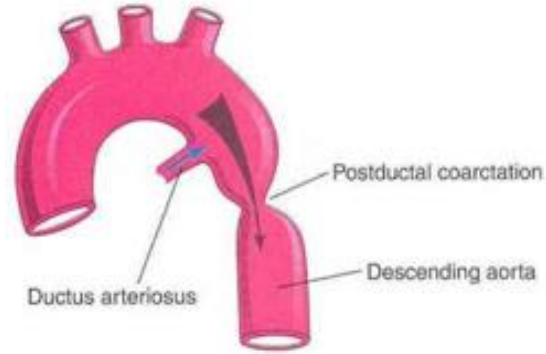
Classification

- Preductal type
  - Less common
  - The narrowing is proximal to the DA
  - If severe, blood flow to the aorta distal to the narrowing depends on a PDA



- **Postductal type**
  - Most common, Adult type
  - Distal to the DA
  - Ductus usually remains open to communicate PA with DAo.
  - Blood flow to the lower body can be impaired with open DA
  - Collateral circulation
  - Hypertension in the upper ext.

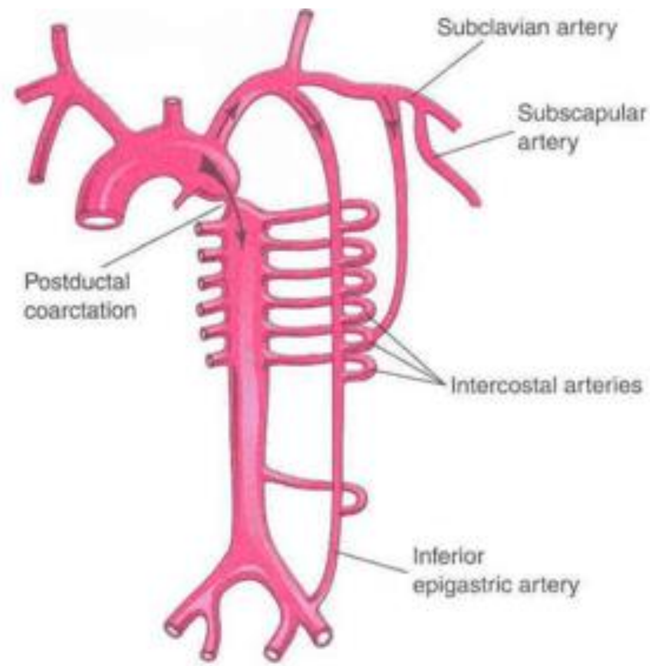
  and weak pulses in the lower ext.



But, mostly the constriction lies

distal to the origin of subclavian art

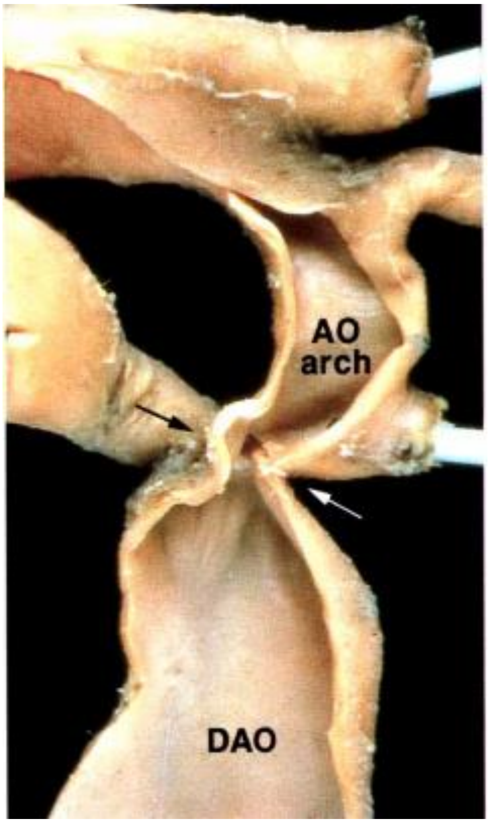
opposite the DA (Juxtaductal)



Etiology

- **Ductus Tissue Theory**
  - Due to a **migration of ductus smooth muscle cells** into the periductal aorta with subsequent constriction and narrowing of the aortic lumen
- **Hemodynamic Theory**
  - **Reduced intrauterine blood flow** causes underdevelopment of aortic arch
  - Results from reduced volume of blood flow through the fetal aortic arch and isthmus

Discrete CoA



CoA Shelf

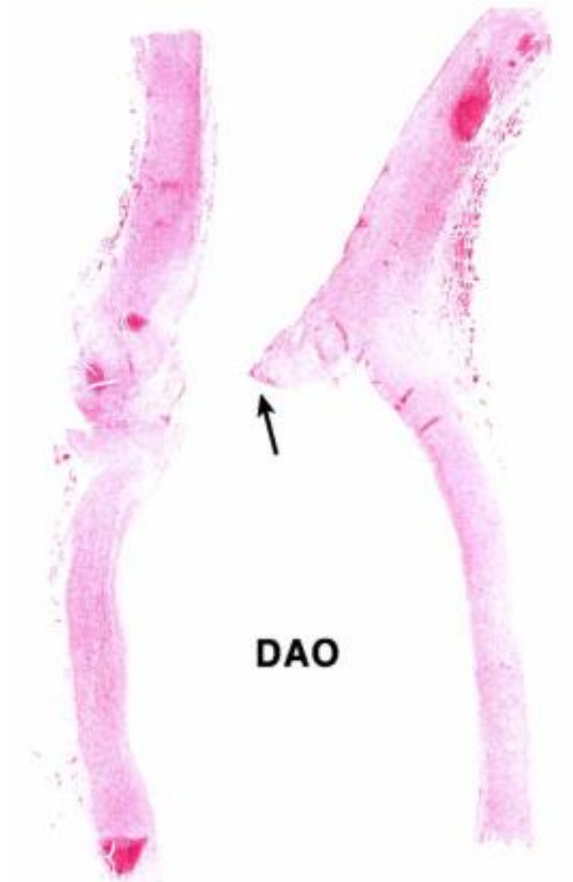
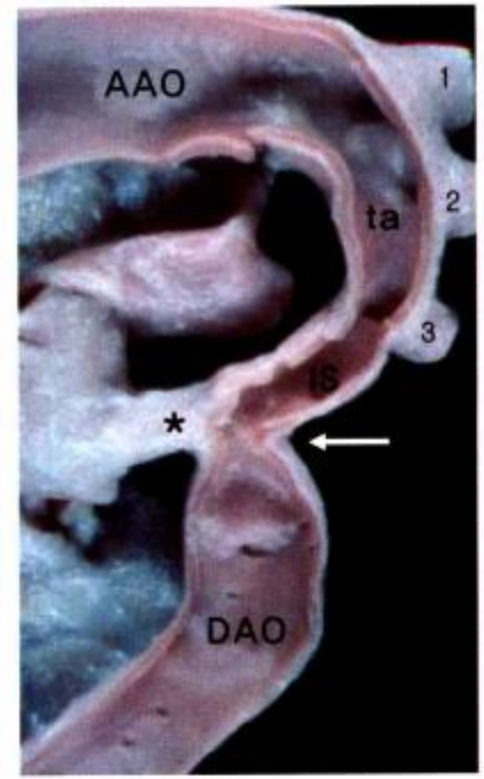
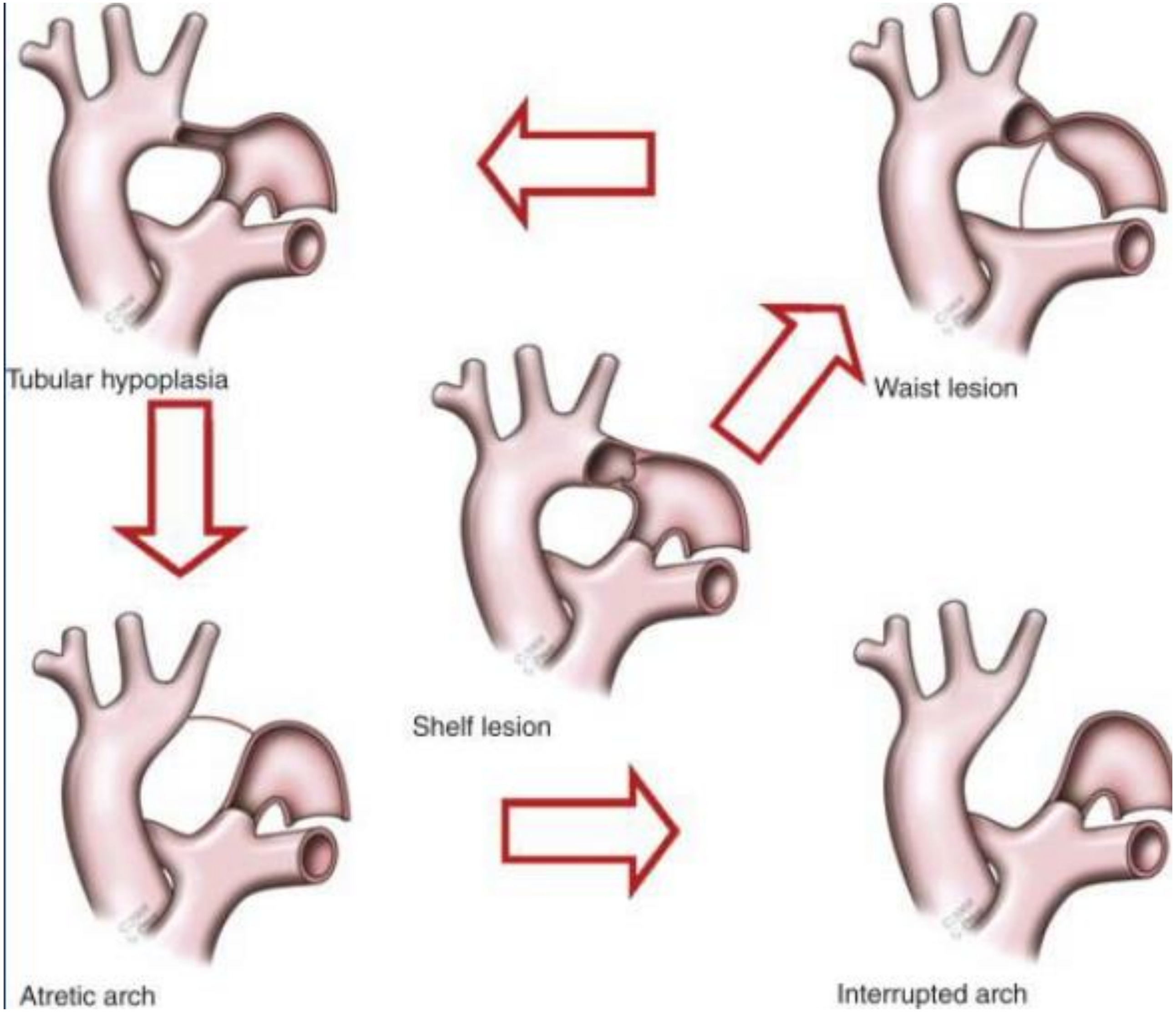


Figure of “3” sign

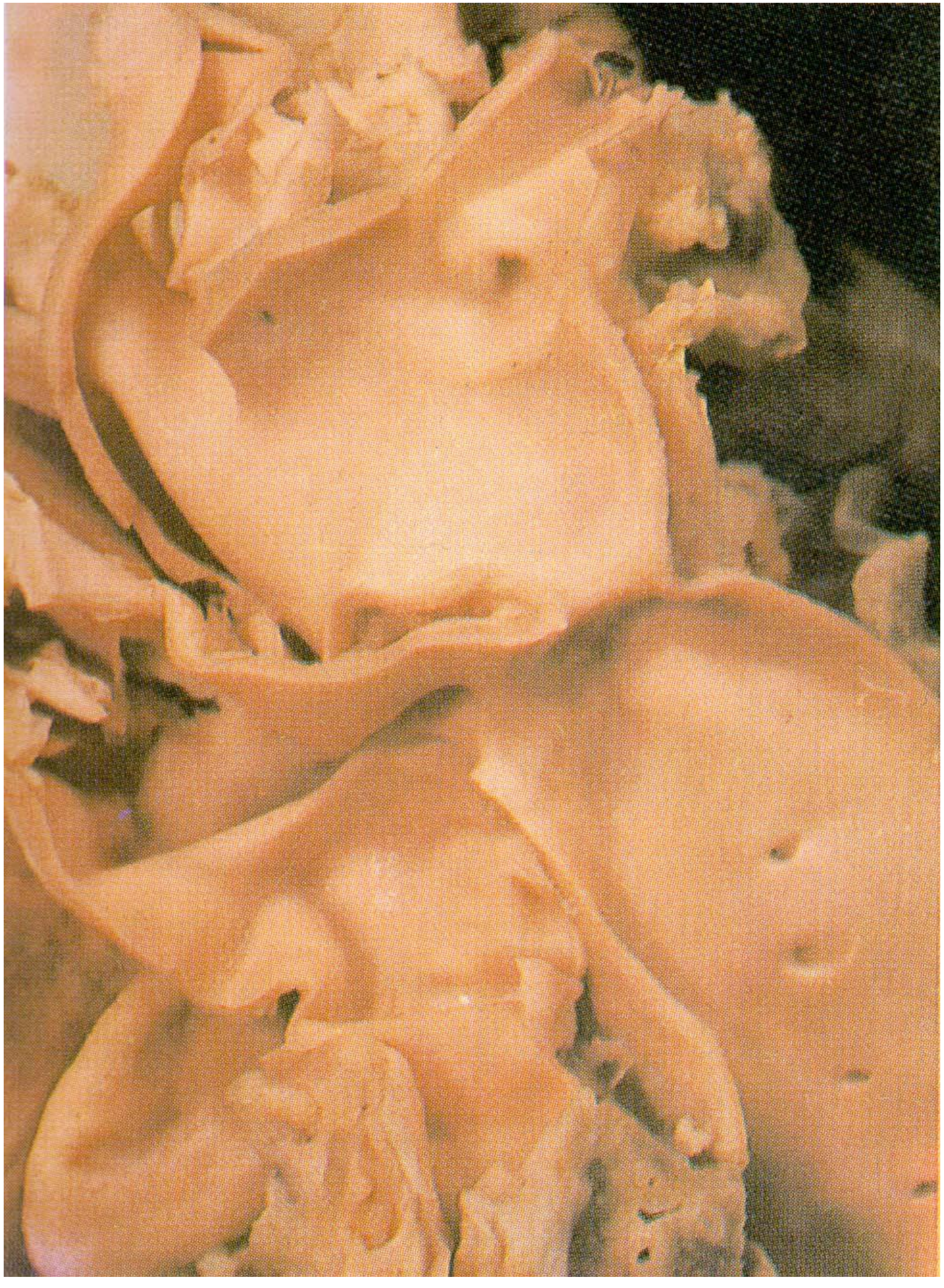


Morphologic Spectrum



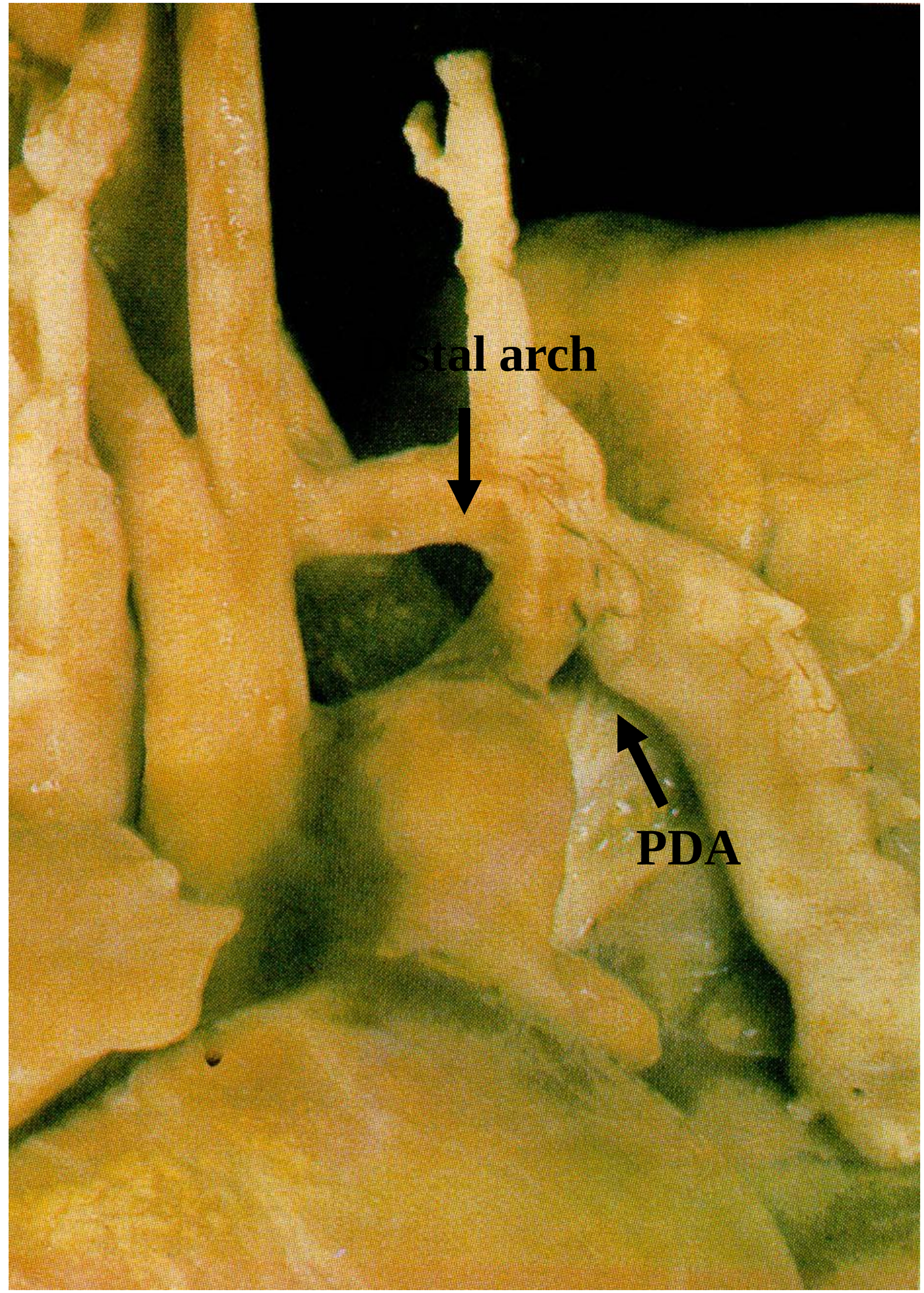
CoA

Localized

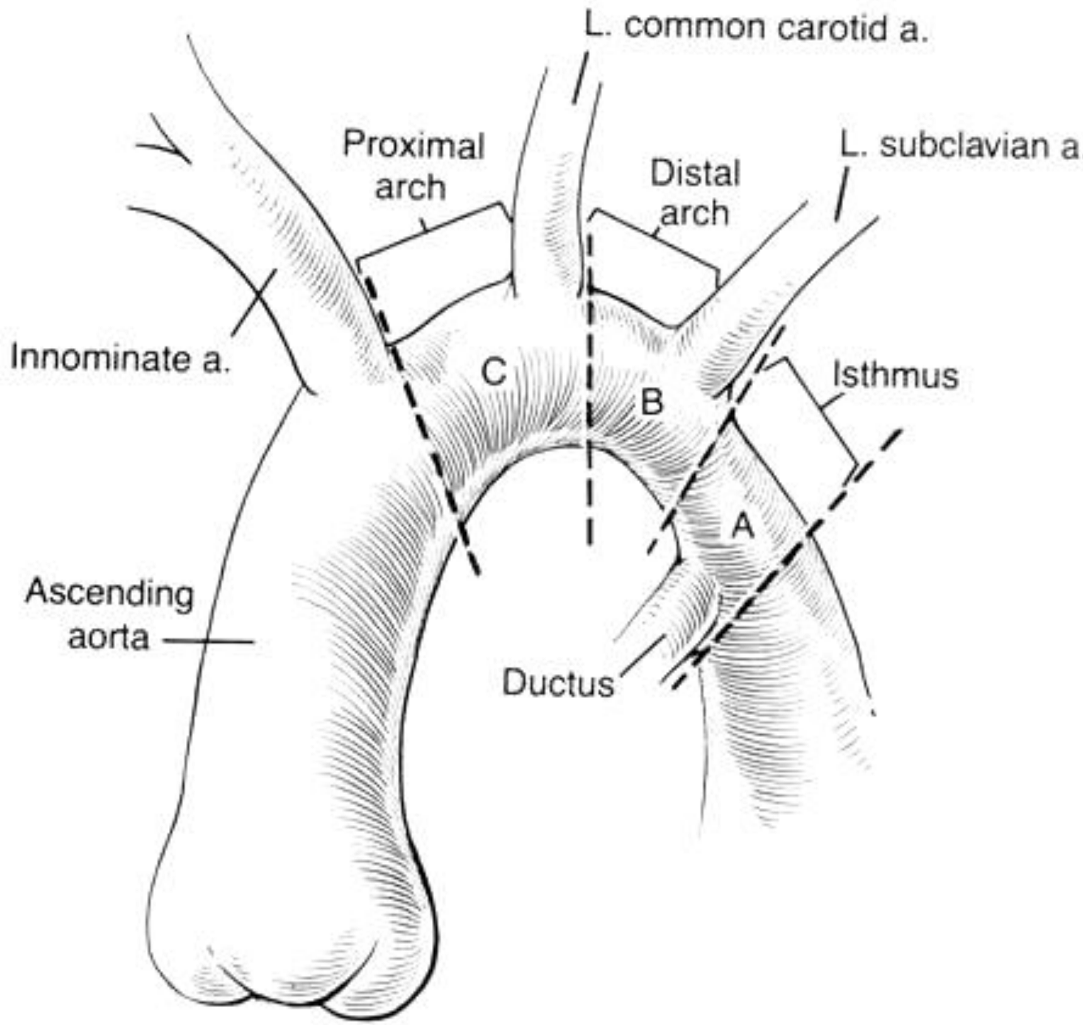


CoA

Tubular Hypoplasia

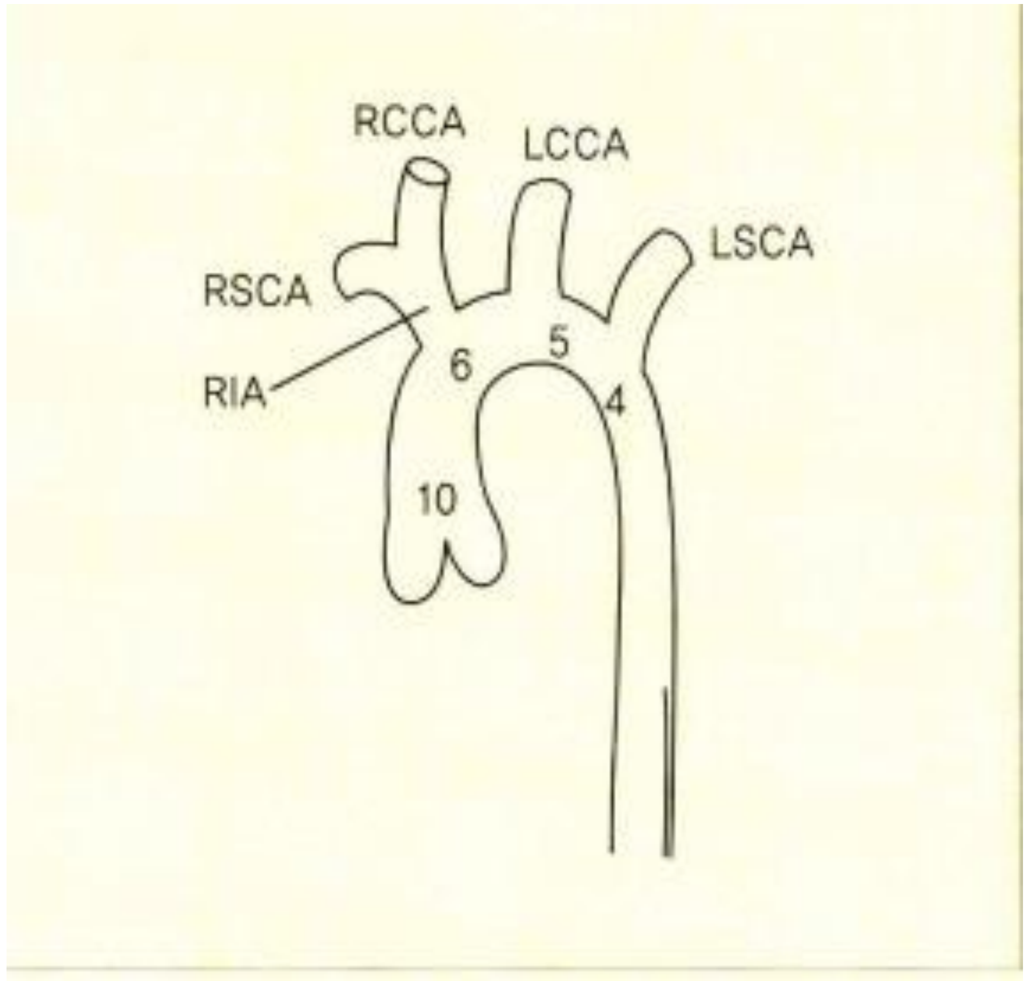


3 Components of Aortic Arch



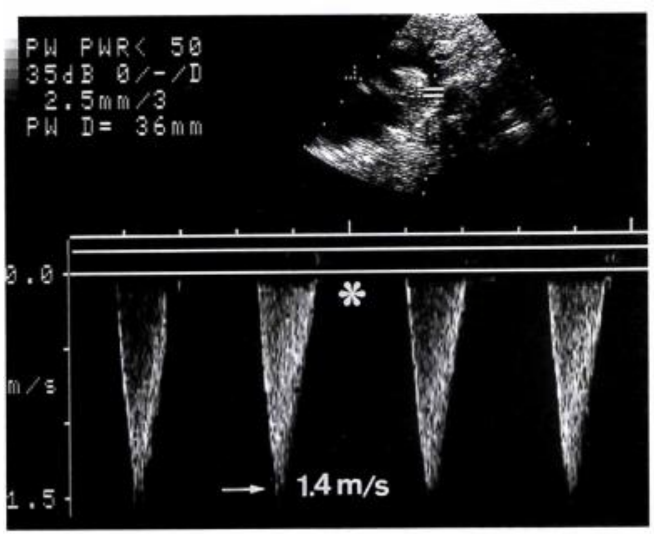
Aortic arch Hypoplasia

- Arch/Aao diameter < 50%
- Arch < Bwt. +1 kg
- Z-score < -2

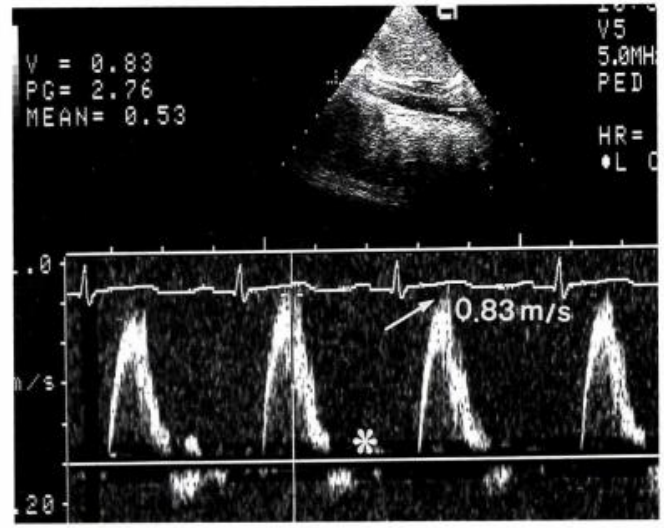



ta
isthmus
DAO

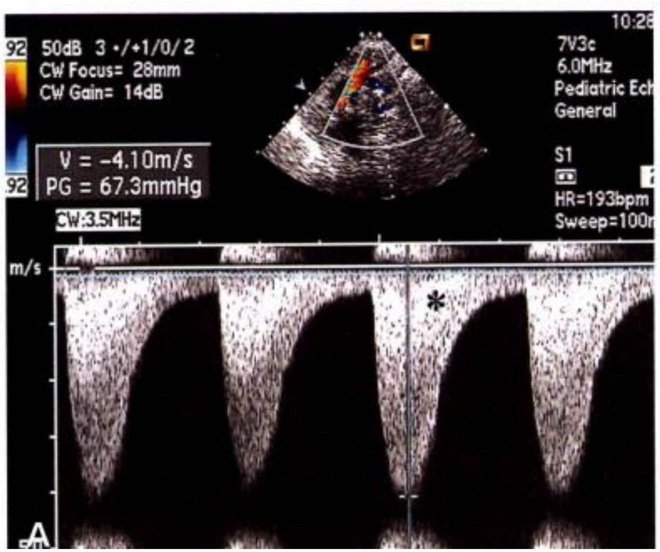
Descending thoracic aorta



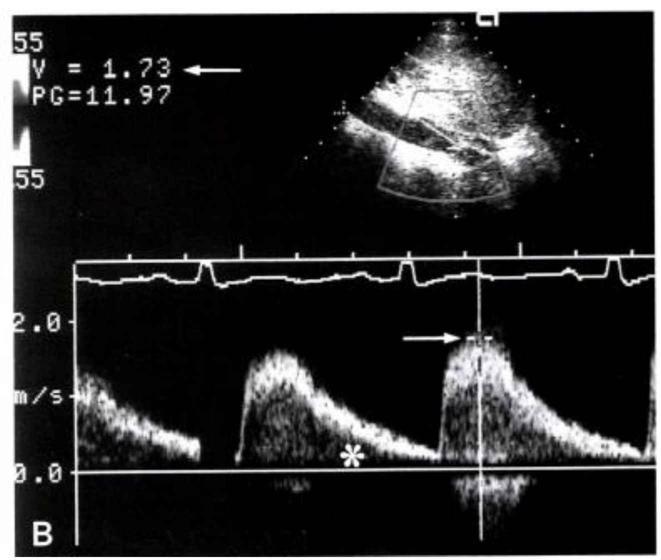
Abdominal aorta



Descending thoracic aorta



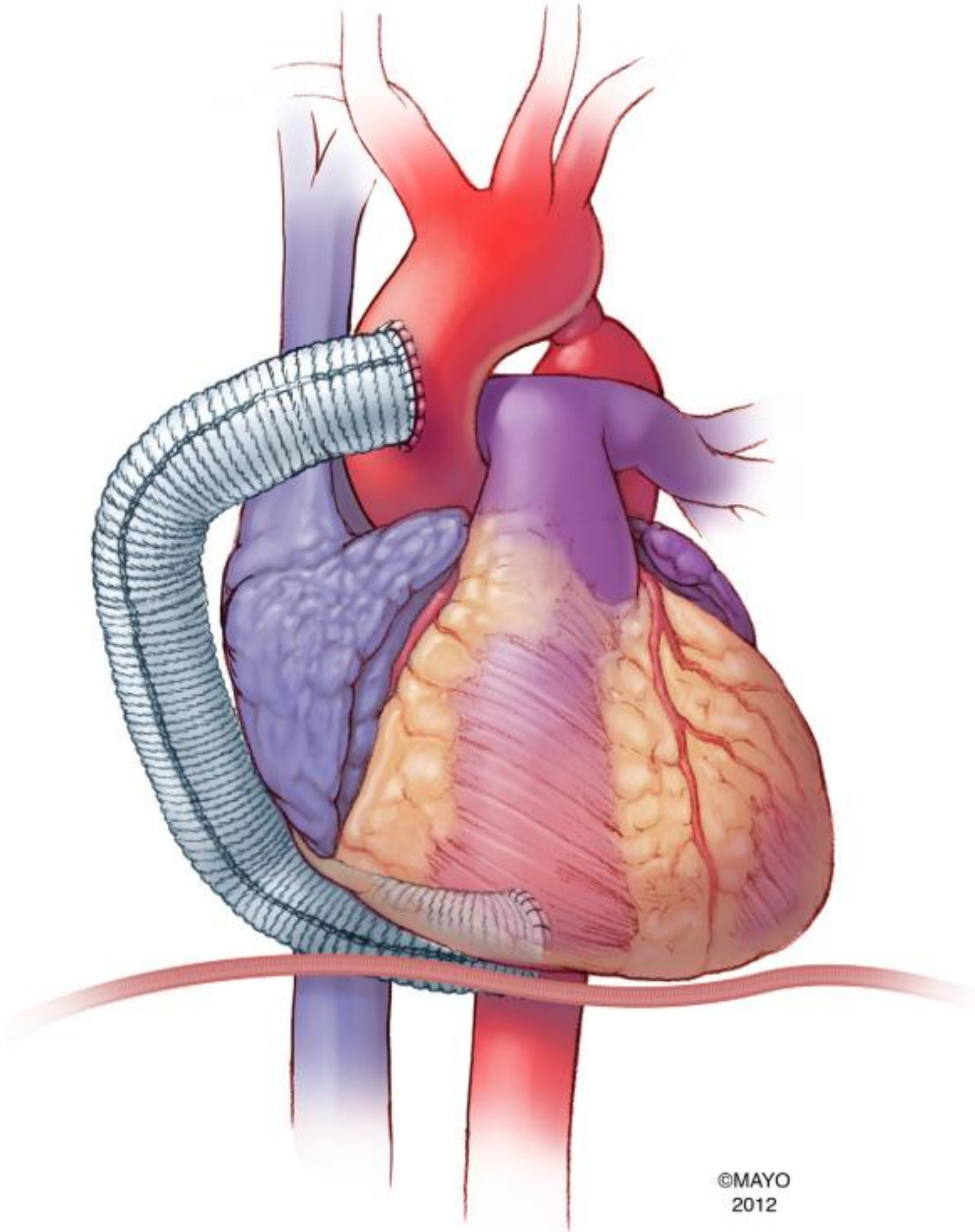
Abdominal aorta



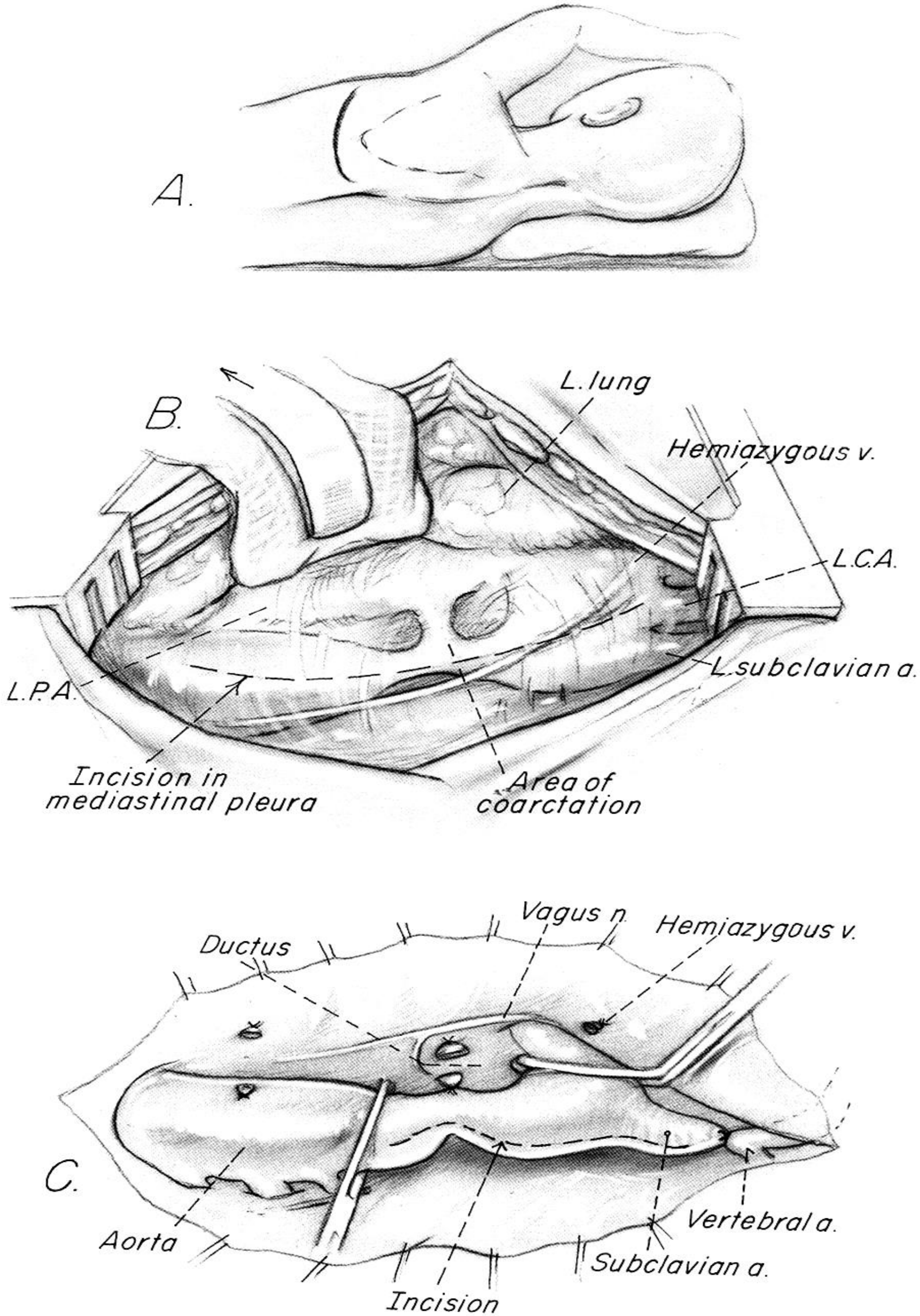
Indications for operation

- Reduction of luminal diameter greater than 50% at any age
- Upper body hypertension over 150 mmHg in young infant (not in HF)
- CoA with CHF at any age
- BP difference > 20 mmHg
- Increase of BP difference at exercise

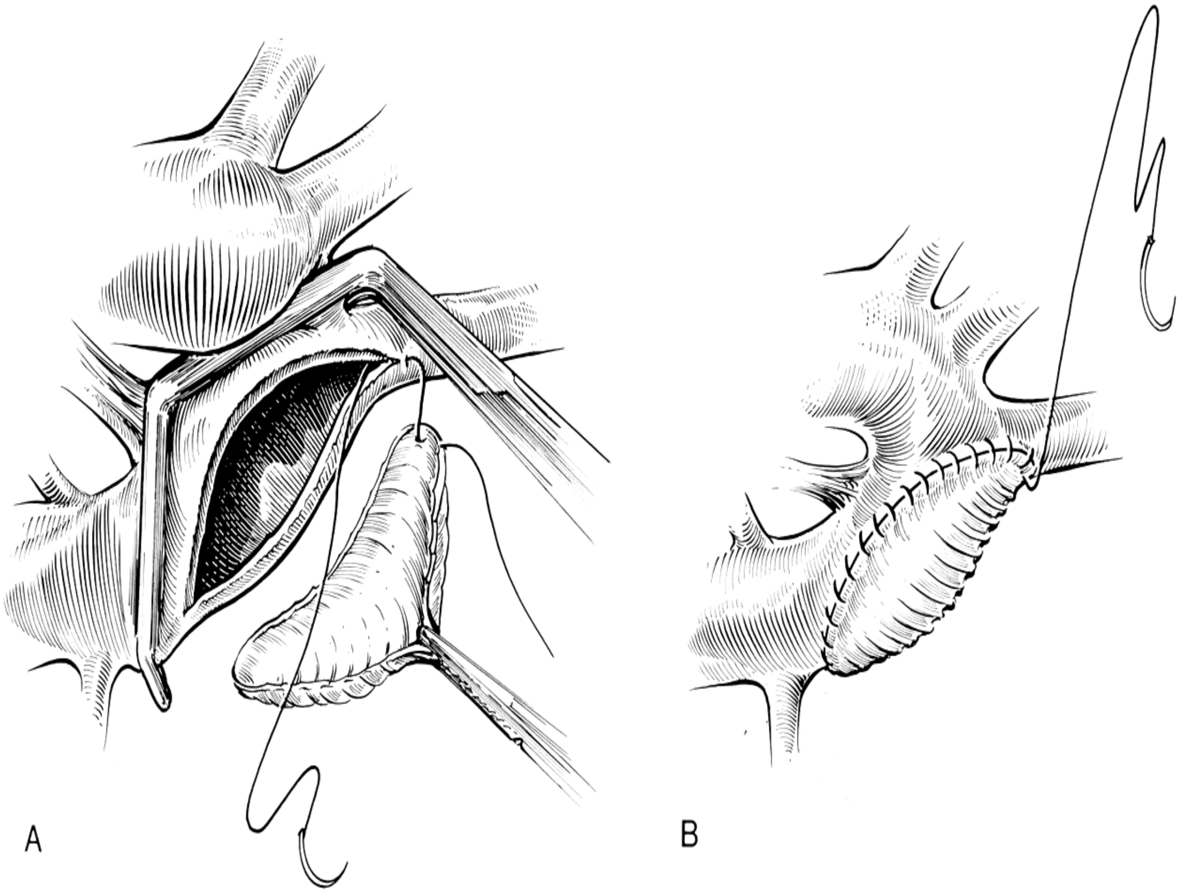
- Res
  - E
  - E
  - E
    -
- Patc
- Left
- Byp nd
  desc



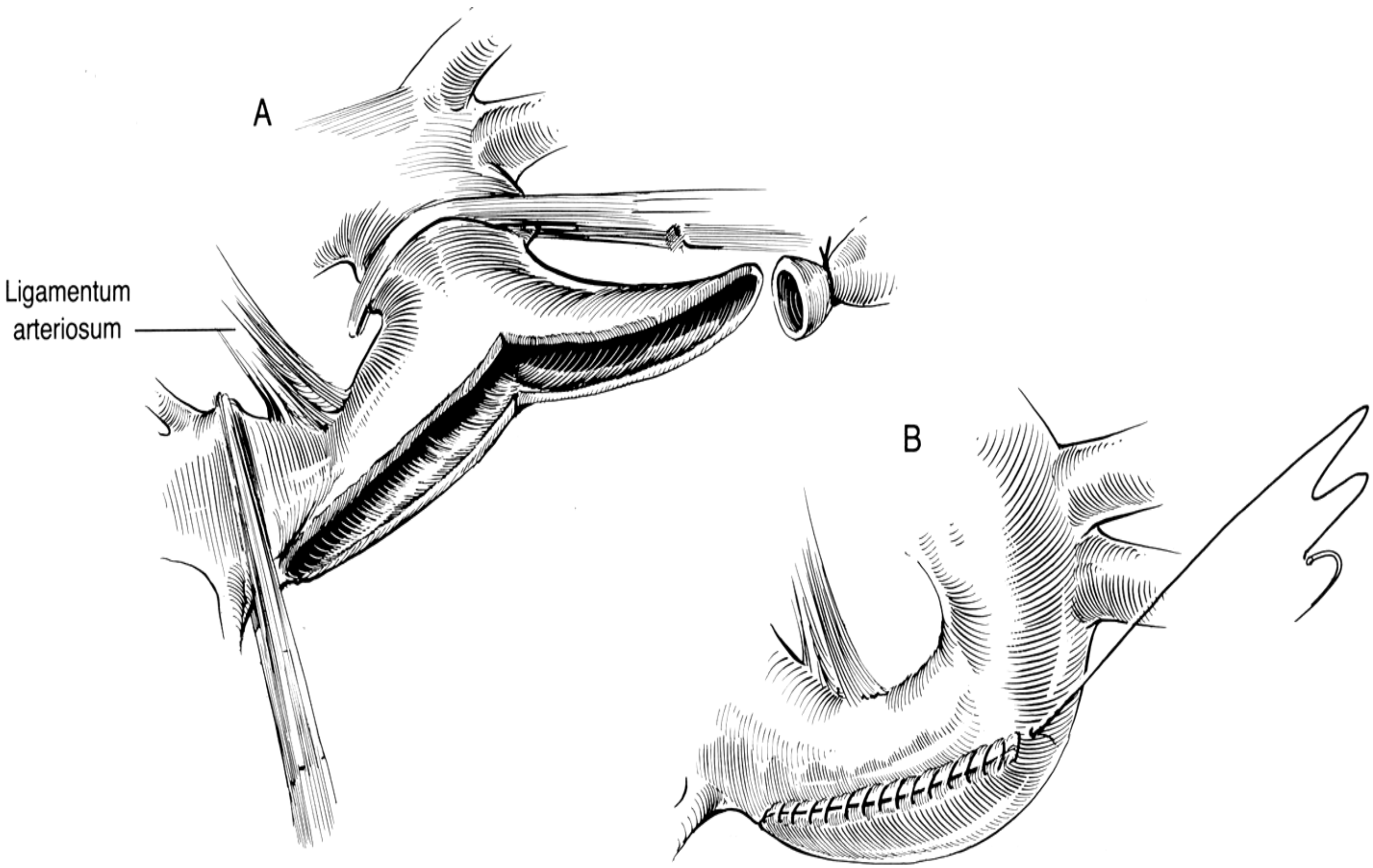
CoA
Exposure



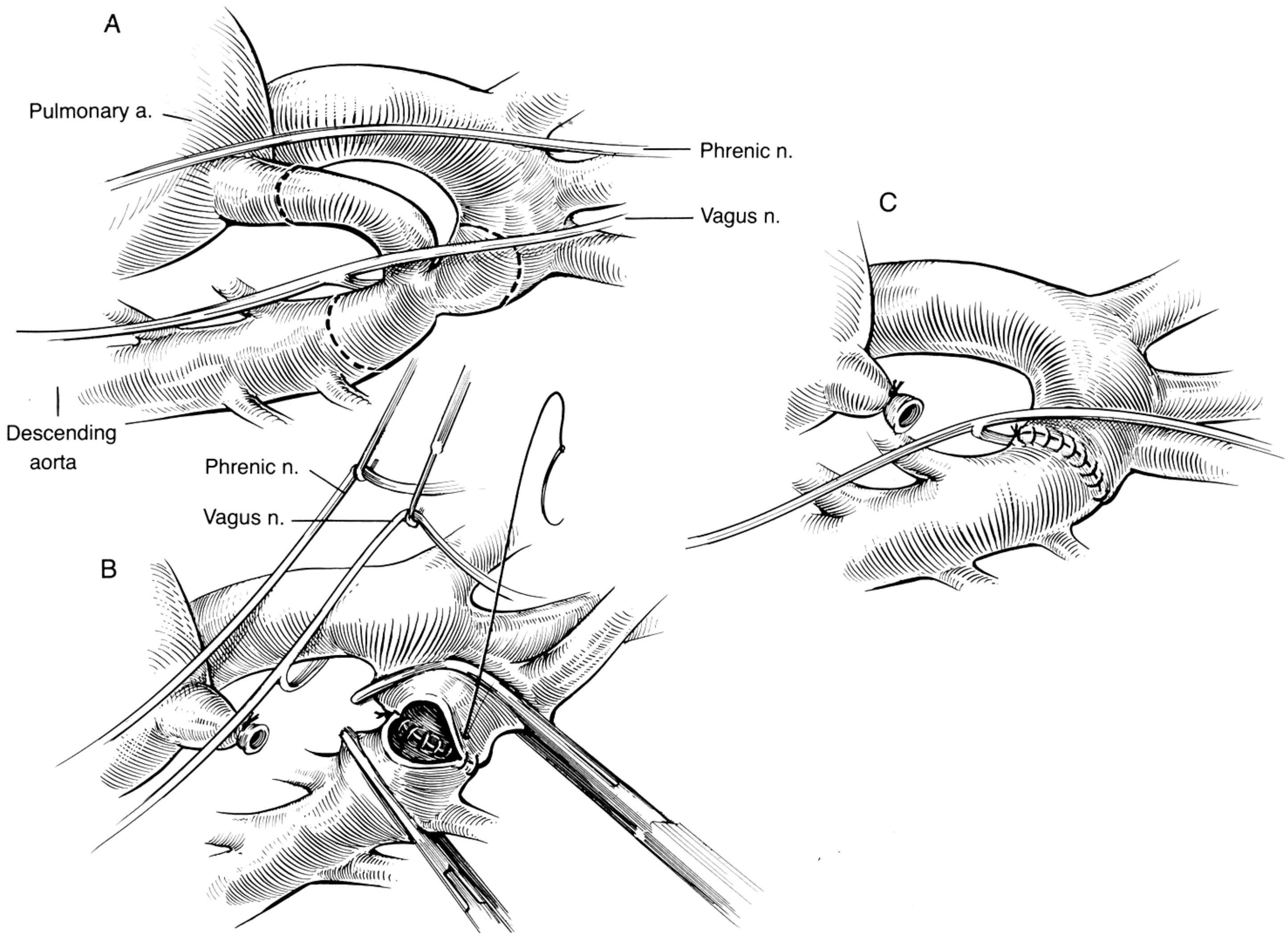
CoA Patch Augmentation



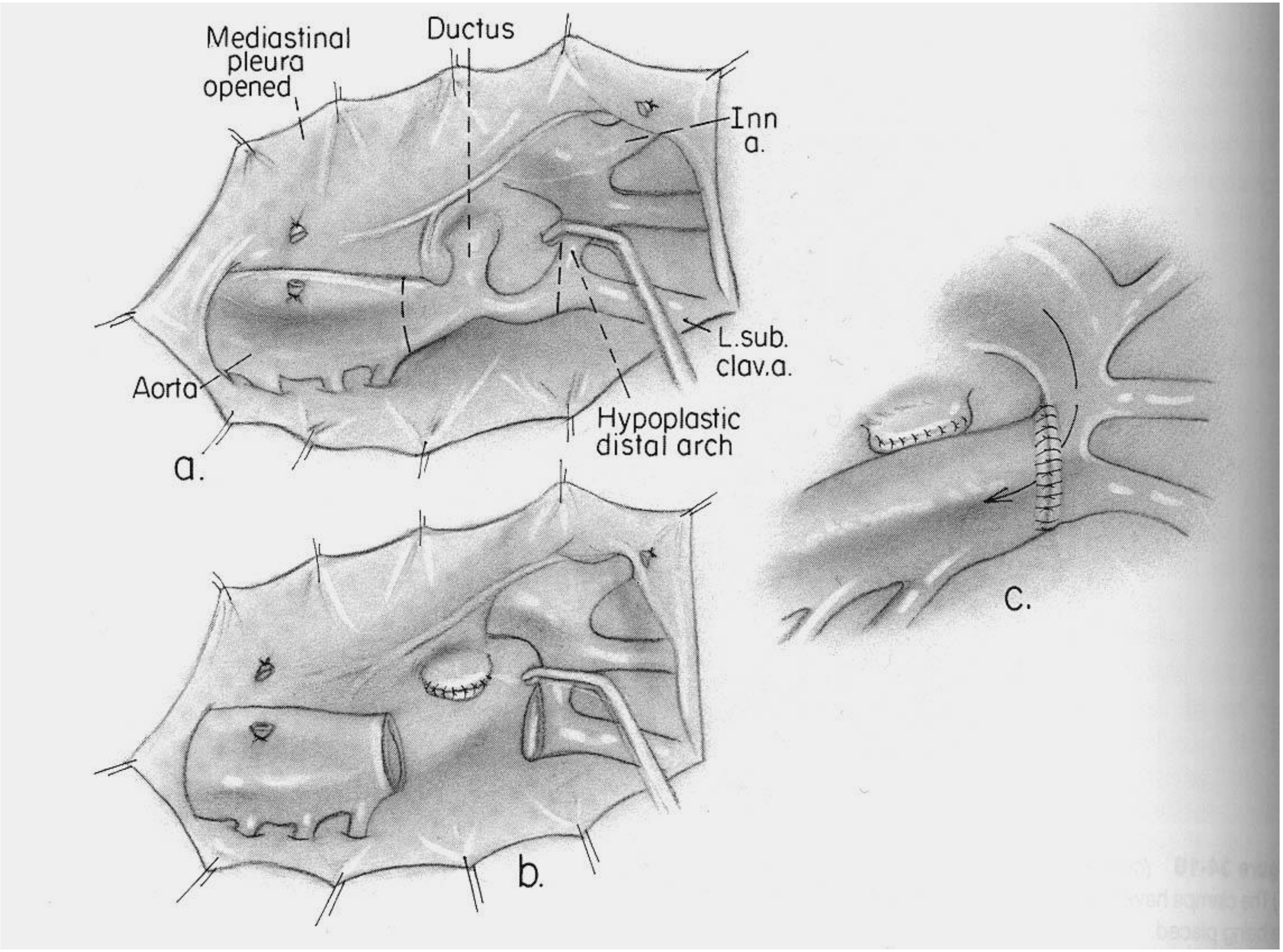
CoA Subclavian Artery Flap



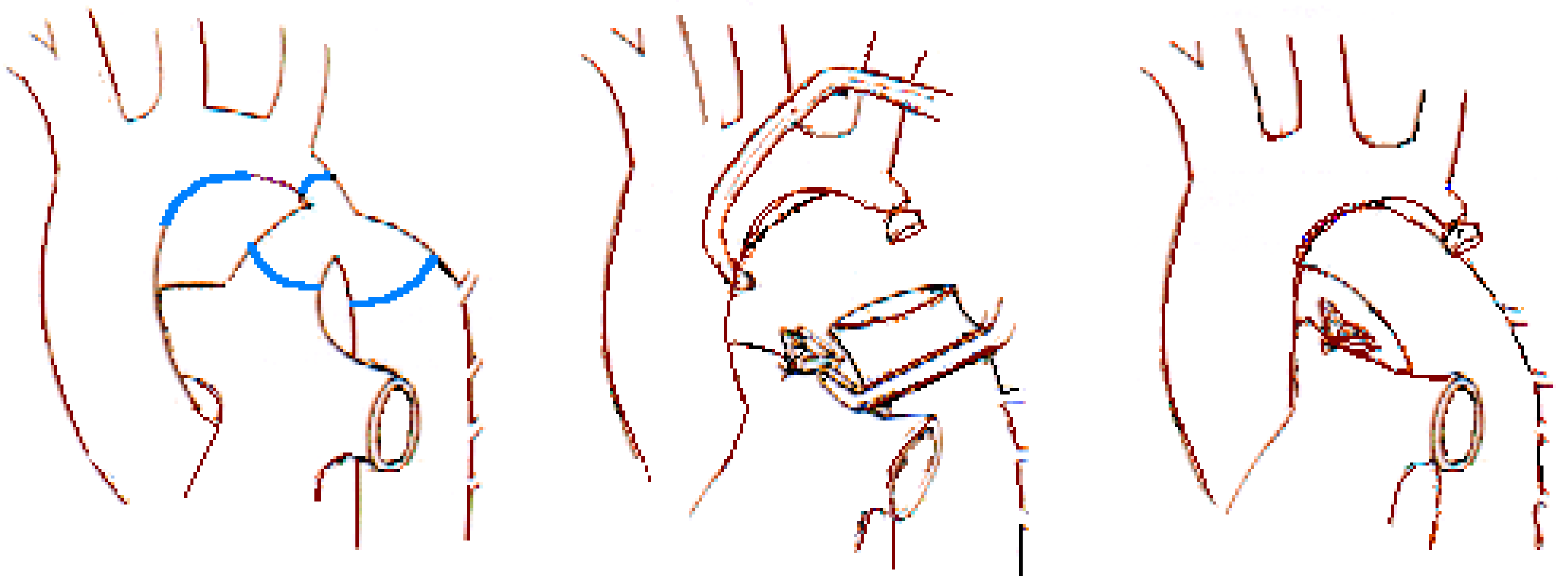
CoA End-to-End Anastomosis



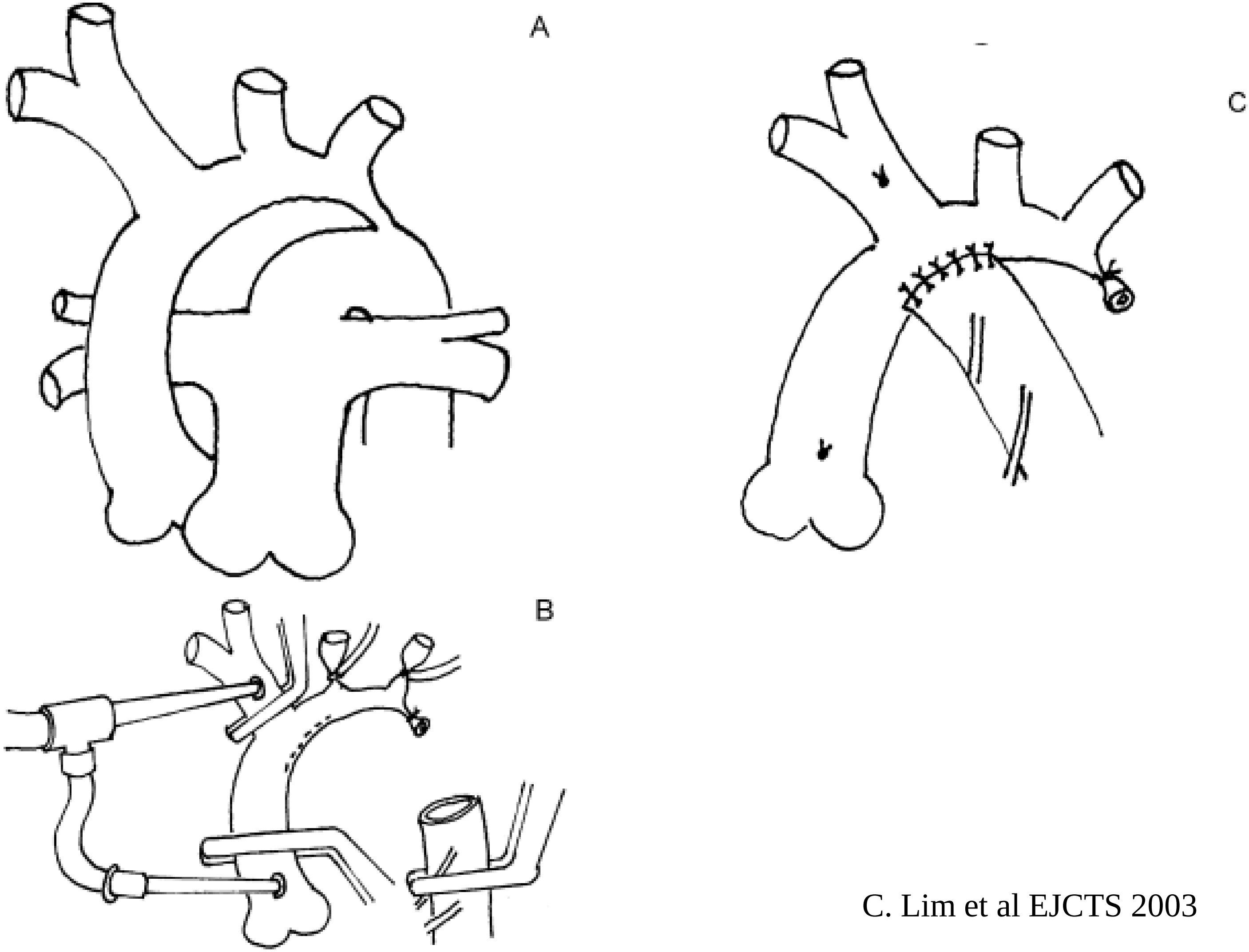
CoA Extended end-to-end Anastomosis



CoA End-to-side Anastomosis



CoA End-to-side Anastomosis



C. Lim et al EJCTS 2003

Surgical Consideration

- DHCA vs. regional perfusion

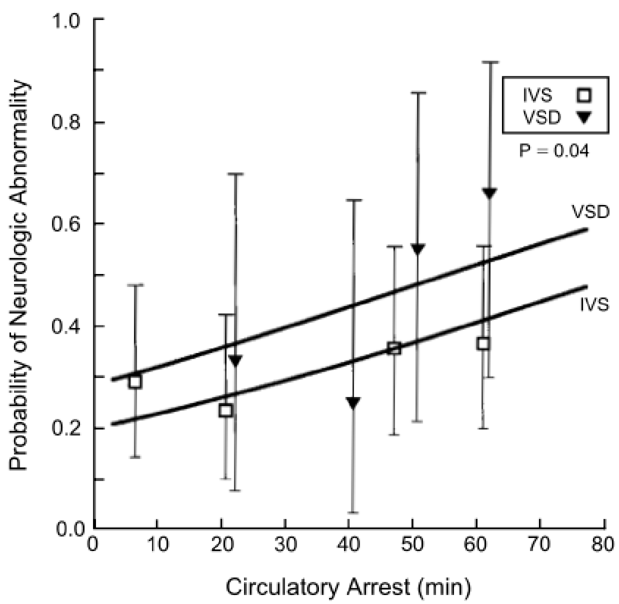
	DHCA	SCP
Advantage	Clear operation f More accurate co small babies	
Disadvantage	Poor neurodevelopmental outcome → Safe duration of circulatory arrest ???	Crowded operative field Technically demanding Lack of randomized trial

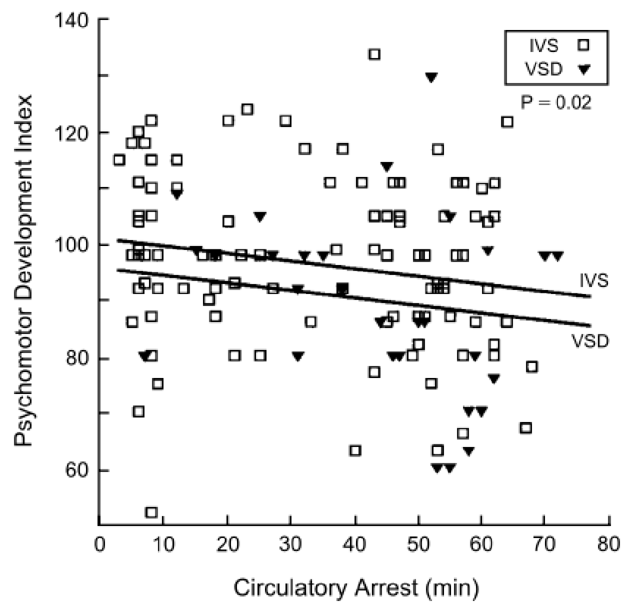
Optimal perfusion flow ??
Optimal perfusion pressure??
Is it really neuroprotective??

Brain protection during arch surgery

- Boston Circulatory Arrest Trial
- 4 year enrollment period up to 1992
- 171 TGA < 3 monthd of age, IVS/VSD
- DHCA (mean 55 min) vs low-flow bypass (50 cc/kg/min)

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





NEJM 1995;332:549-55

Brain protection during arch surgery

The effect of duration of deep hypothermic circulatory arrest in infant heart surgery on late neurodevelopment: The Boston Circulatory Arrest Trial

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Jane W. Newburger, MD, MPH[a,f]
Leonard A. Rappaport, MD[b,f]
Adre J. duPlessis, MBChB[c,g]
Richard A. Jonas, MD[d,h]
Gil Wernovsky, MD[a,f]
Ming Lin, MS[e,i]
David C. Bellinger, PhD, MSc[c,g]

Objectives: Despite the technical advantages of total circulatory arrest for vital organ support during infant heart surgery, many centers have moved away from its use because of the demonstrated effects of circulatory arrest of long duration on neurodevelopmental outcomes. Our goal was to determine the functional form of the association between duration of circulatory arrest and risk of neurodevelopmental dysfunction.

Methods: From 1988 to 1992, in a single-center trial, infants with D-transposition of the great arteries underwent the arterial switch operation after random assignment to circulatory arrest or low-flow bypass. The alpha-stat method was used, and hematocrit on bypass was maintained at 20%. Developmental, neurologic, and speech outcomes were assessed at 8 years of age in 155 of 160 eligible children (97%). Outcomes selected for analysis were Full-Scale, Verbal, and Performance IQ, Reading and Mathematics Composite, time to complete the Grooved Pegboard (dominant hand), and the Mayo Test for Apraxia.

Results: Nonparametric regression and piecewise linear models indicated that neurodevelopmental outcomes were generally not adversely affected unless the duration of circulatory arrest exceeded a threshold of 41 minutes (95% 1-sided lower confidence limit of 32 minutes).

Conclusions: We found that the effect of duration of total circulatory arrest on later neurodevelopmental outcomes is nonlinear, with little influence at shorter durations and with steadily worsening outcomes after longer durations of circulatory arrest. Because the effects of duration of circulatory arrest may vary according to diagnosis, age at surgery, and other bypass and perioperative variables, this study cannot ascertain a universally "safe" duration of total circulatory arrest.

J Thorac Cardiovasc Surg 2003;126:1397-403

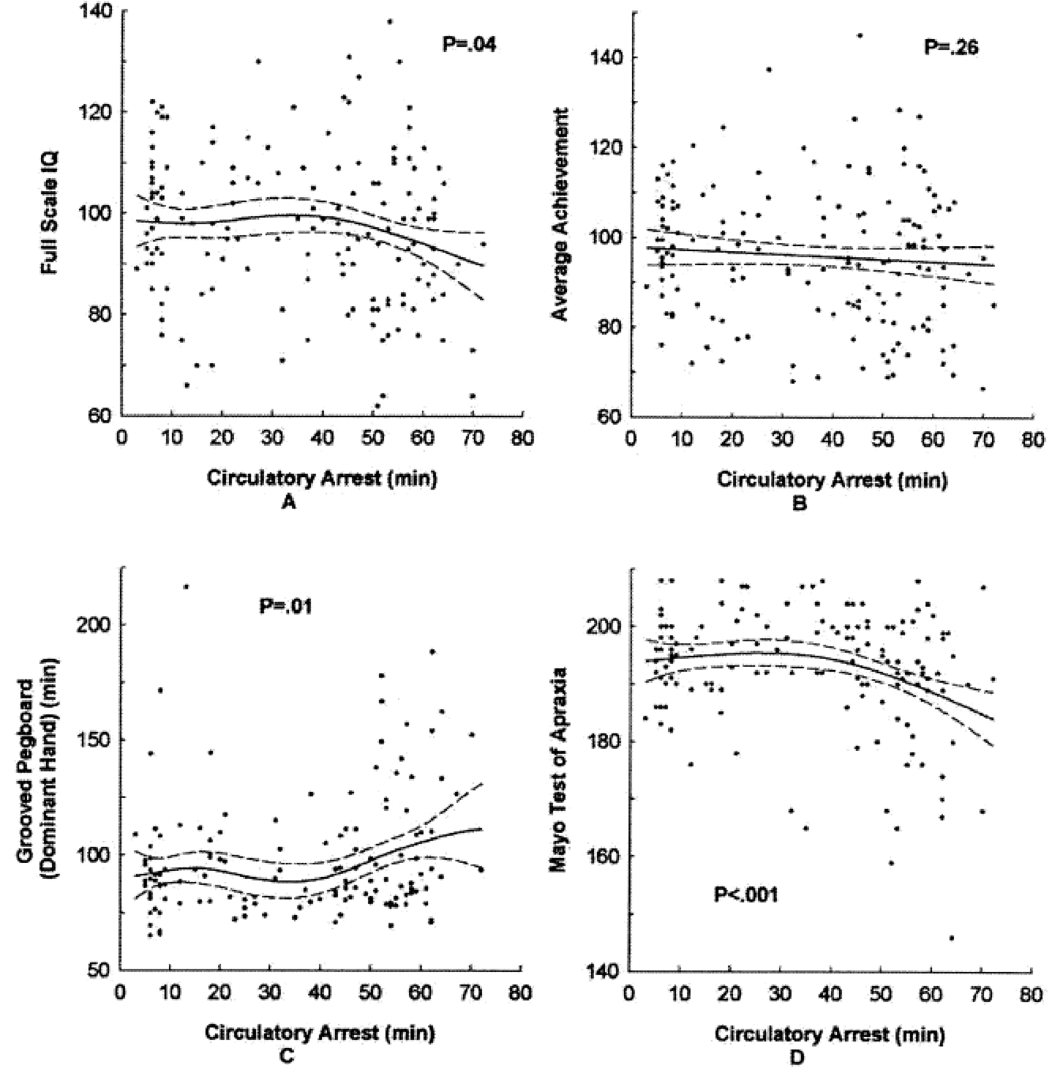


Figure 1. Neurodevelopmental outcome as a function of duration of CA. The *solid lines* were derived from generalized additive models as a smooth function of duration of CA, with adjustment for diagnosis and family social class. The *dashed lines* present 95% confidence intervals, with the scatter plots denoting the raw data values. The *P* values assess whether there is any effect of duration of CA on outcomes, with adjustment for diagnosis and family social class. The panels represent Full-Scale IQ (A), Average Achievement (of Wechsler Reading and Mathematics Composite scores) (B), Grooved Pegboard (Dominant Hand) (C), and Mayo Test of Apraxia of Speech (D). The natural logarithm of the time to complete the Grooved Pegboard was used because this transformation yielded scores that were approximately normally distributed. The graph then exponentiates the results to return to the raw scale.

TABLE 2. Estimation of threshold effects of duration of circulatory arrest based on piecewise linear models*

Variable	Cut-point estimate (min)	95% lower confidence limit (min)
Full-Scale IQ	42	27
Verbal IQ	41	23
Performance IQ	47	31
Average Achievement	43	4
Grooved Pegboard	35	13
Mayo Test for Apraxia	40	29
Combined analysis (over all 6 outcomes)	41	32

*All models were based on piecewise linear regression, with adjustment for diagnosis and social class.

Brain protection during arch surgery

HOW TO DO IT

Selective Cerebral Perfusion Technique During Aortic Arch Repair in Neonates

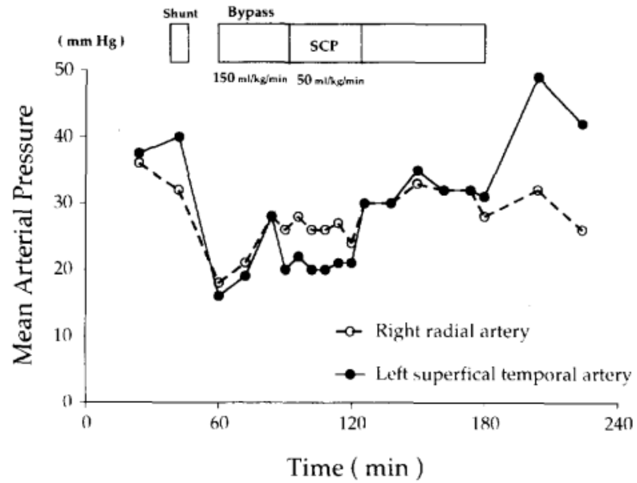
Toshihide Asou, MD, Hideaki Kado, MD, Yutaka Imoto, MD, Yuichi Shiokawa, MD, Ryuji Tominaga, MD, Yoshito Kawachi, MD, and Hisataka Yasui, MD

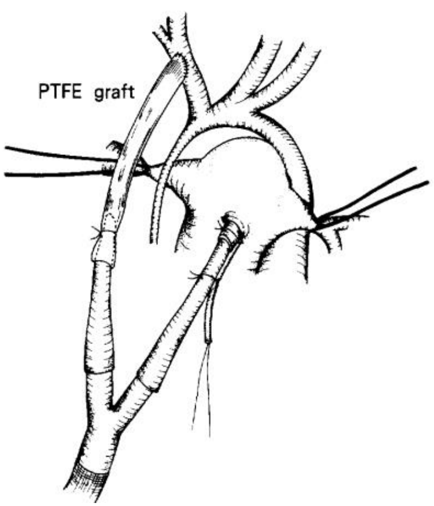
Department of Cardiovascular Surgery, Fukuoka Children's Hospital, and Division of Cardiac Surgery, Research Institute of Angiocardiology, Kyushu University, Fukuoka, Japan

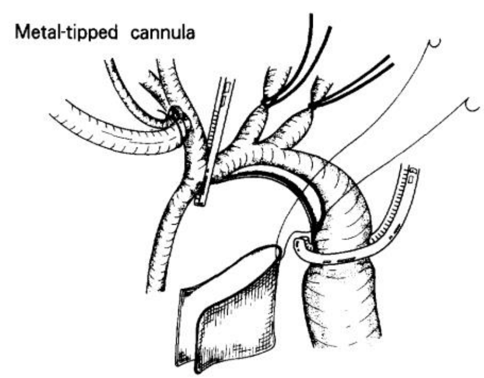
We describe selective cerebral perfusion techniques for repair of the aortic arch in neonates. These techniques may help protect the brain from ischemic injury caused by a cessation of cerebral perfusion for aortic arch reconstruction in patients with hypoplastic left heart syndrome or interrupted aortic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

(Ann Thorac Surg 1996;61:1546–8)

with both the innominate artery and the descending aorta unclamped. The perfusion rate returns to 150 $mL \cdot kg^{-1} \cdot min^{-1}$ to perfuse the upper and the lower part of the body via the graft of the innominate artery. The pericardial roll is anastomosed to the proximal end of the main pulmonary artery conjoined with the ascending aorta to create a systemic outflow tract. After the atrial septal defect is enlarged, another arterial cannula is then inserted into the pericardial roll to perfuse the whole body while the cannula inserted into the polytetrafluoroethylene graft is removed. Finally, the graft is trimmed and anastomosed to the pulmonary artery as a systemic–pulmonary arterial shunt.







Rectal temperature: 22℃
Flow rate: 50mL/Kg/min
Right radial a. pr: 28 mmHg
Lt. sup temporal a. pr: 20mmHg
Neonate brain: 1/7~1/10 of Bwt.
Adult: 1/15 of Bwt.

Brain protection during arch surgery

Table 5. Antegrade Selective Cerebral Perfusion and Selective Cerebral and Myocardial Perfusion: Management Protocols Reported

Author	Year	Pts	Flow[a]	Temp °C	Hematocrit	Gas Management	Reference
ASCP							
Asou	1996	14	50	22		alpha stat	ATS 1996;61:1546–8
McElhinney	1997	7	30	18		alpha stat	JTCS 1997;114:718–26
Pigula	2001	15	20	18		alpha stat	ATS 2001;72:401–7
Tchervenkov	2001	18	18–76	18			ATS 2001;72:1615–20
Andropoulos	2003	34	63	17–22	25	pH stat	JTCS 2003;125:491–9
Hoffman	2004	9	30–70	18–20	25–30	pH stat	JTCS 2004;127:223–33
Dent	2006	22	30	18	28–30	pH alpha stat	JTCS 2006;131:190–7
Amir	2005		20–40	22–25	25–30	pH/alpha stat	ATS 2005;80:1955–64
SCMP							
Kostelka	2004	24	30–40	18–28	>30		ATS2004;78:1989–93
Lim	2002	48	50–100	18	20		EJCTS 2003;23:149–55

[a] Flow rates are mL (kg · min).

ASCP = antegrade selective cerebral perfusion; SCMP = selective cerebral and myocardial perfusion.

Ann Thorac Surg 2006:82:2233-9

Brain protection during arch surgery

Outcomes after aortic arch reconstruction for infants: deep hypothermic circulatory arrest versus moderate hypothermia with selective antegrade cerebral perfusion

Igor A. Kornilov[a,*], Yuri S. Sinelnikov[b], Ilya A. Soinov[c], Dmitry N. Ponomarev[a], Marina S. Kshanovskaya[c], Aleksandra A. Krivoshapkina[d], Artem V. Gorbatykh[c] and Alexander Y. Omelchenko[c]

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EJCTS 2015;48:e45-50

Selective anterograde cerebral perfusion has lower risk of neurological complications and higher incidence of renal complications.

Brain protection during arch surgery

Changes of Brain Magnetic Resonance Imaging Findings After Congenital Aortic Arch Anomaly Repair Using Regional Cerebral Perfusion in Neonates and Young Infants

Jae Gun Kwak, MD, Woong-Han Kim, MD, Jin Tae Kim, MD, In-One Kim, MD, and Jong-Hee Chae, MD

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time:
patien

brain MRI, and these were acute focal subdural hemorrhage (n = 1) and acute focal infarction (n = 1). However, they were without clinical significance. Periventricular leukomalacia was not observed on brain MRI. There was no significant change between the preoperative and postoperative findings on brain sonography and electroencephalograms. All the patients showed normal neurologic growth for a mean follow-up duration of 175.3 days (range: 25 to 497 days).

Brain protection during arch surgery

Neurological Injury After Neonatal Cardiac Surgery
A Randomized, Controlled Trial of 2 Perfusion Techniques

Selma O. Algra, MD; Nicolaas J.G. Jansen, MD, PhD; Ingeborg van der Tweel, PhD;
Antonius N.J. Schouten, MD; Floris Groenendaal, MD, PhD; Mona Toet, MD, PhD;
Wim van Oeveren, MD, PhD; Ingrid C. van Haastert, PhD; Paul H. Schoof, MD, PhD;
Linda S. de Vries, MD, PhD; Felix Haas, MD, PhD

Circulation 2014;129:224-33

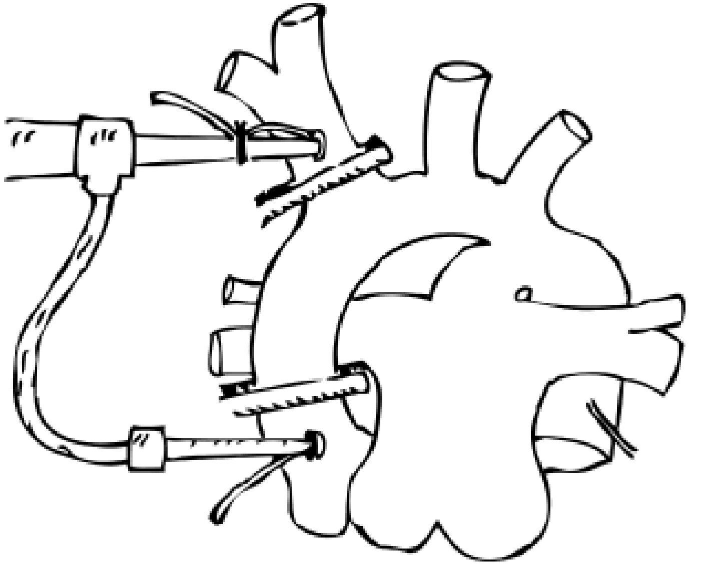
Nasopharyngeal Temp: 18℃, Hct: 24~28%, 25% of full flow, <40mmHg

- MRI: Preop and 1 wk after operation
- Preop: 50% had evidence of cerebral injury
- **No difference in DHCA and ACP groups**
- **78% of DHCA and 72% of ACP group had new injury**
- **White matter injury** : m/c type in both groups
- Early motor and cognitive outcome at 2 year were similar
- Central infarction occurred exclusively after ACP

Selective Cerebral perfusion

- 50~100 ml/kg/min
- Right arm a-line monitoring
- Transcranial Doppler
- Cerebral oximeter
- Lactic acid

- ± myocardial perfusion



JG Kwak et al. Eur J Cardio Thorac Surg 2007;32:751-5

CONTROVERSIES IN PEDIATRIC/CONGENITAL HEART SURGERY

Perioperative Neuroprotective Strategies

David P. Nelson,[a] Dean B. Andropoulos,[b] and Charles D. Fraser, Jr[c]

Long-term neurodevelopmental impairment is common in newborns and infants undergoing corrective or palliative congenital heart surgery. The etiologies of neurodevelopmental morbidity in these children are multifactorial and include prenatal, preoperative, intraoperative, and postoperative factors. Perioperative neurologic monitoring is thought to be integral to prevention or rescue from adverse neurologic events. Recent advances in perfusion techniques for congenital heart surgery now ensure adequate cerebral O_2 delivery during *all* phases of cardiopulmonary bypass. Periventricular leukomalacia and other serious neurologic injury can be minimized by an optimized perfusion strategy of continuous high-flow, high hematocrit cardiopulmonary bypass, minimal use of deep hypothermic circulatory arrest, antegrade cerebral perfusion during aortic arch reconstruction, pH-stat blood gas strategy, and cerebral monitoring with NIRS and trans-cranial Doppler. Because there is evidence that brain injury can also occur in the prenatal, preoperative, and postoperative periods, improved strategies to prevent injury in these arenas are much needed. Extensive further clinical investigation is warranted to identify neuroprotective management strategies for the operating room and intensive care unit to preserve neurologic function and optimize long-term neurodevelopmental outcomes in children with congenital heart disease.
Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 11:49-56

Neurologic Monitoring

Near infrared spectroscopy

prolonged periods of low cerebral O2 sat (<45%) have higher incidence of PVL and poor neurological outcome

Transcranial Doppler

EEG

Perioperative Neuroprotective Strategies

David P. Nelson,[a] Dean B. Andropoulos,[b] and Charles D. Fraser, Jr[c]

Long-term neurodevelopmental impairment is common in newborns and infants undergoing corrective or palliative congenital heart surgery. The etiologies of neurodevelopmental morbidity in these children are multifactorial and include prenatal, preoperative, intraoperative, and postoperative factors. Perioperative neurologic monitoring is thought to be integral to prevention or rescue from adverse neurologic events. Recent advances in perfusion techniques for congenital heart surgery now ensure adequate cerebral O_2 delivery during *all* phases of cardiopulmonary bypass. Periventricular leukomalacia and other serious neurologic injury can be minimized by an optimized perfusion strategy of continuous high-flow, high hematocrit cardiopulmonary bypass, minimal use of deep hypothermic circulatory arrest, antegrade cerebral perfusion during aortic arch reconstruction, pH-stat blood gas strategy, and cerebral monitoring with NIRS and trans-cranial Doppler. Because there is evidence that brain injury can also occur in the prenatal, preoperative, and postoperative periods, improved strategies to prevent injury in these arenas are much needed. Extensive further clinical investigation is warranted to identify neuroprotective management strategies for the operating room and intensive care unit to preserve neurologic function and optimize long-term neurodevelopmental outcomes in children with congenital heart disease.
Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 11:49-56

Alkalosis and hypocarbia should be avoided
Near infrared spectroscopy monitoring is useful

Special features of postoperative care

- Systemic arterial HTN
  - Usually
- Abdominal pain
  - 5~10%
  - Mild abdominal discomfort for a few days
  - Increase in blood flow and pressure in the mesenteric arteries
  - Delay feeds, control blood pressure, nasogastic decompression
- Chylothorax
  - 5%

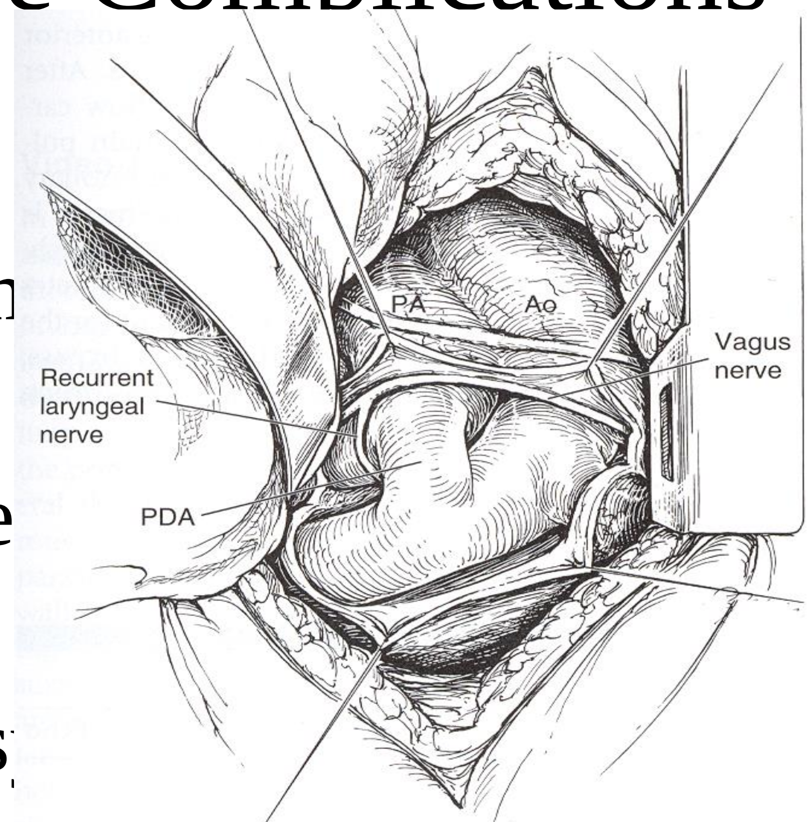
Postoperative hypertension

- Altered baroreceptor response with increased excretion of epinephrine or norepinephrine
- **Sympathetic nervous system** in early phase (24~36hr after operation), and **renin-angiotensin system** in late phase

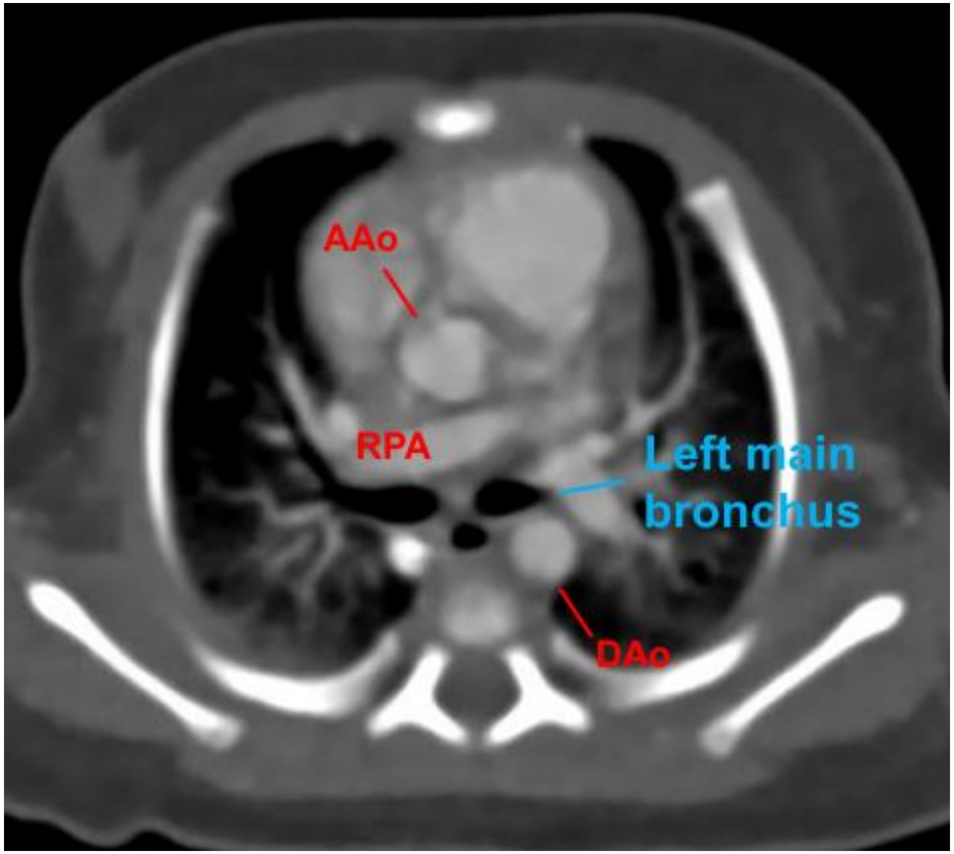
- Beta blocker & Nitroglycerin, Nitroprusside

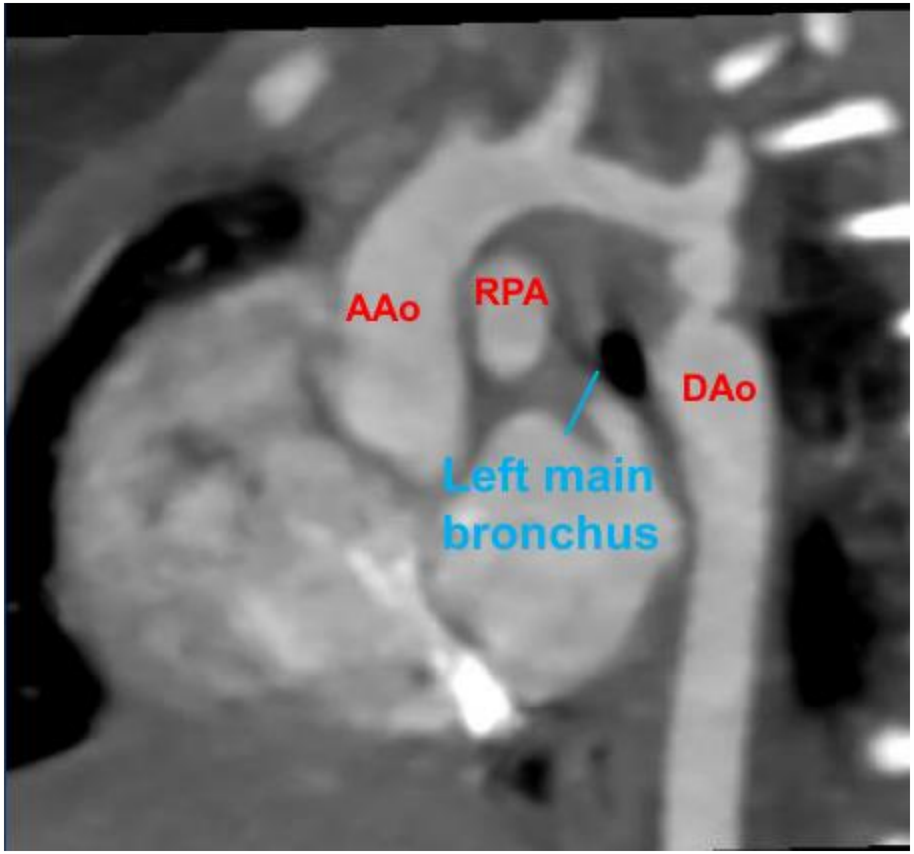
Postoperative Complications

- Hoarseness
- Ipsilateral diaphragn
- Chylothorax
- Vessel injury & blee
- Rebound HTN
- Post-coarctectomy sy
- Left main bronchus compression
- Paralysis due to spinal cord ischemia



Left main bronchus compression!!





To avoid airway problem

- Extensive dissection arch vessels and descending aorta

- Arch repair using autologous MPA patch

Augmentation of the Lesser Curvature With an Autologous Vascular Patch in Complex Aortic Coarctation and Interruption

Heemoon Lee, MD, Ji-Hyuk Yang, MD, PhD, Tae-Gook Jun, MD, PhD, Yang Hyun Cho, MD, PhD, I-Seok Kang, MD, June Huh, MD, PhD, and Jinyoung Song, MD, PhD

Departments of Thoracic and Cardiovascular Surgery and Pediatrics, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea

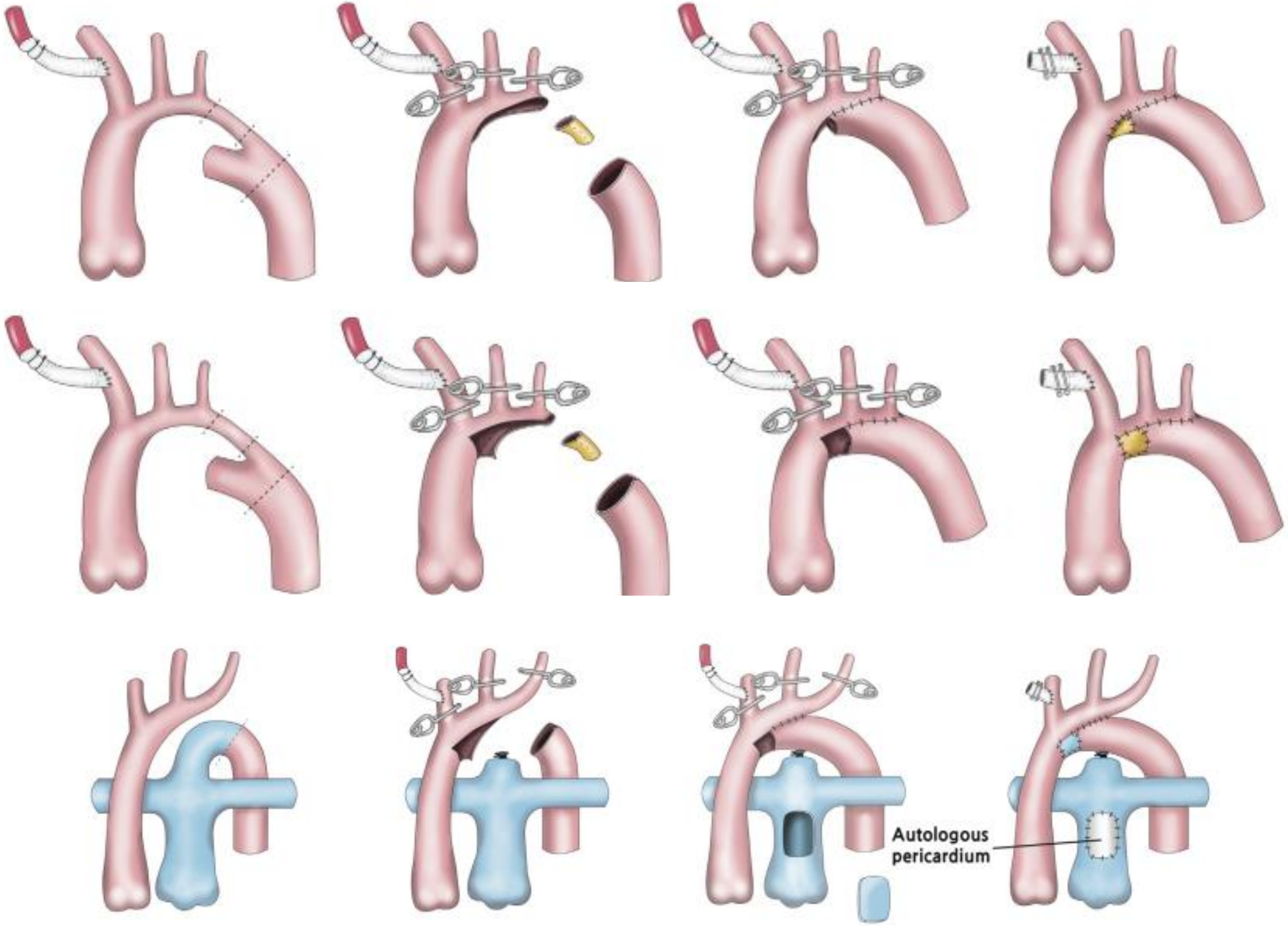
Background. Reconstruction of the aortic arch in patients with complex aortic coarctation or interruption continues to be a challenge because of early left main bronchial compression or recoarctation and late Gothic arch formation. We propose a modified arch reconstruction technique augmenting the lesser curvature with an autologous vascular patch, which can relieve tension on the anastomosis without a prosthetic material.

Methods. We retrospectively reviewed 33 patients with coarctation and arch hypoplasia (n = 31) or arch interruption (n = 2) who underwent arch reconstruction with an autologous vascular patch from 2007 to 2012. Median age at the operation was 17 days (range, 5 to 200 days). Median body weight was 3.7 kg (range, 2.3 to 7.0 kg). Cardiopulmonary bypass was used for all operations. Median antegrade selective cerebral perfusion time was 35 minutes (range, 23 to 59 minutes). Combined intracardiac anomalies in 29 patients (88%) were corrected simultaneously. The reconstructed arch was supplemented in the lesser curvature with an autologous vascular patch that was harvested from aortic isthmus (n = 25), pulmonary artery (n = 4), left subclavian artery (n = 2), aberrant right subclavian artery (n = 1), or distal arch (n = 1).

Results. One patient (3%) died of acute respiratory distress syndrome. All survivors were discharged at 15 days (range, 7 to 58 days) postoperatively without neurologic complications or bronchial obstructions. Median follow-up was 24.8 months (range, 0.2 to 48.5 months). No recoarctation was observed during follow-up, and no patient needed reoperation.

Conclusions. Augmenting the lesser curvature with an autologous vascular patch during arch reconstruction resulted in excellent midterm outcomes. Not only can a more natural shape of arch and less tension on the anastomosis be obtained, but complications, such as left main bronchial obstruction or recoarctation, can also be minimized. Long-term follow-up is needed to evaluate late development of recoarctation, hypertension, or aneurysm formation.

(Ann Thorac Surg 2016;101:2309–14)



Lee et al. ATS 2016

Airway problem management

- Aortopexy
- RPA anterior translocation

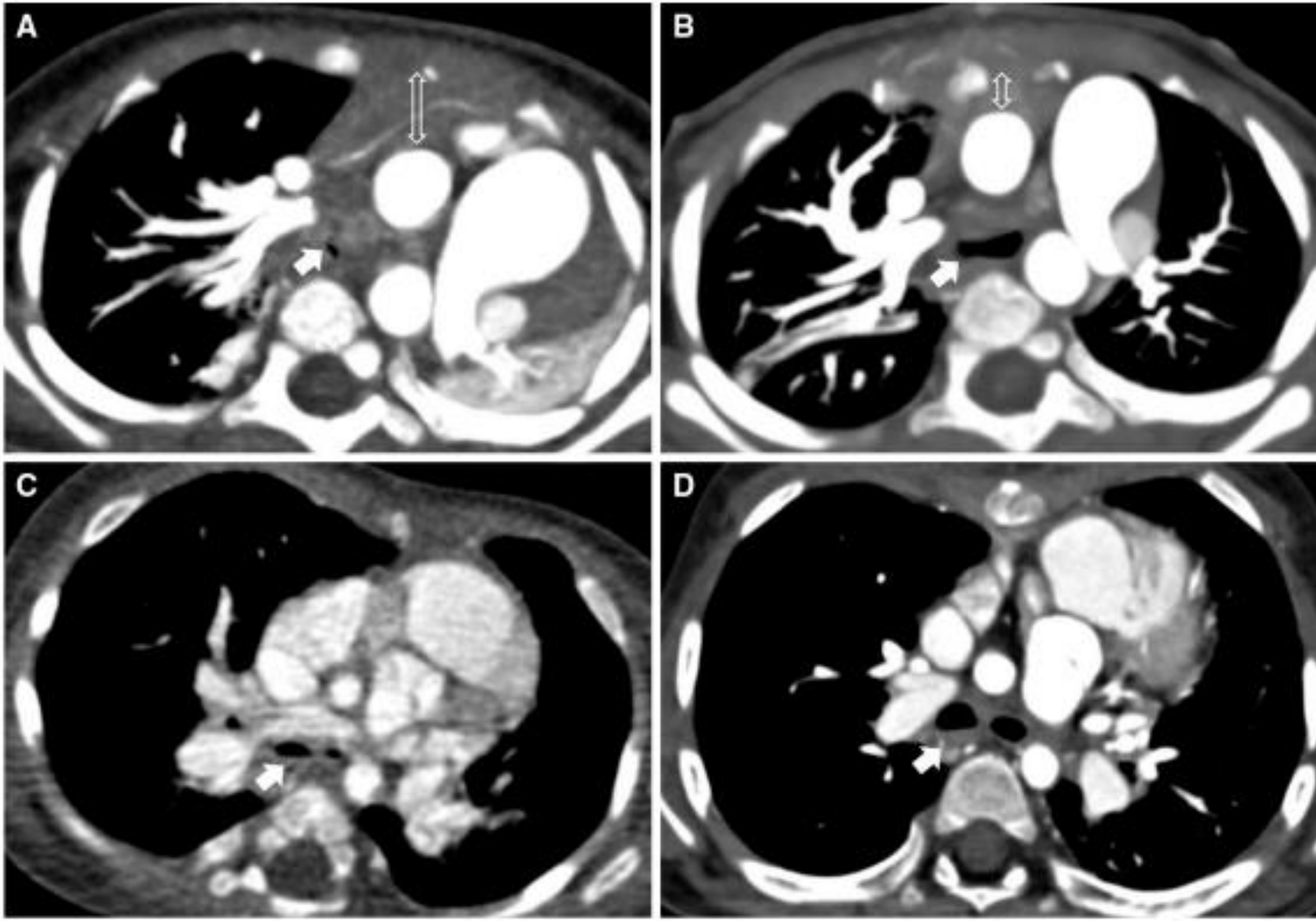
Aortopexy With Preoperative Computed Tomography and Intraoperative Bronchoscopy for Patients With Central Airway Obstruction After Surgery for Congenital Heart Disease: Postoperative Computed Tomography Results and Clinical Outcomes

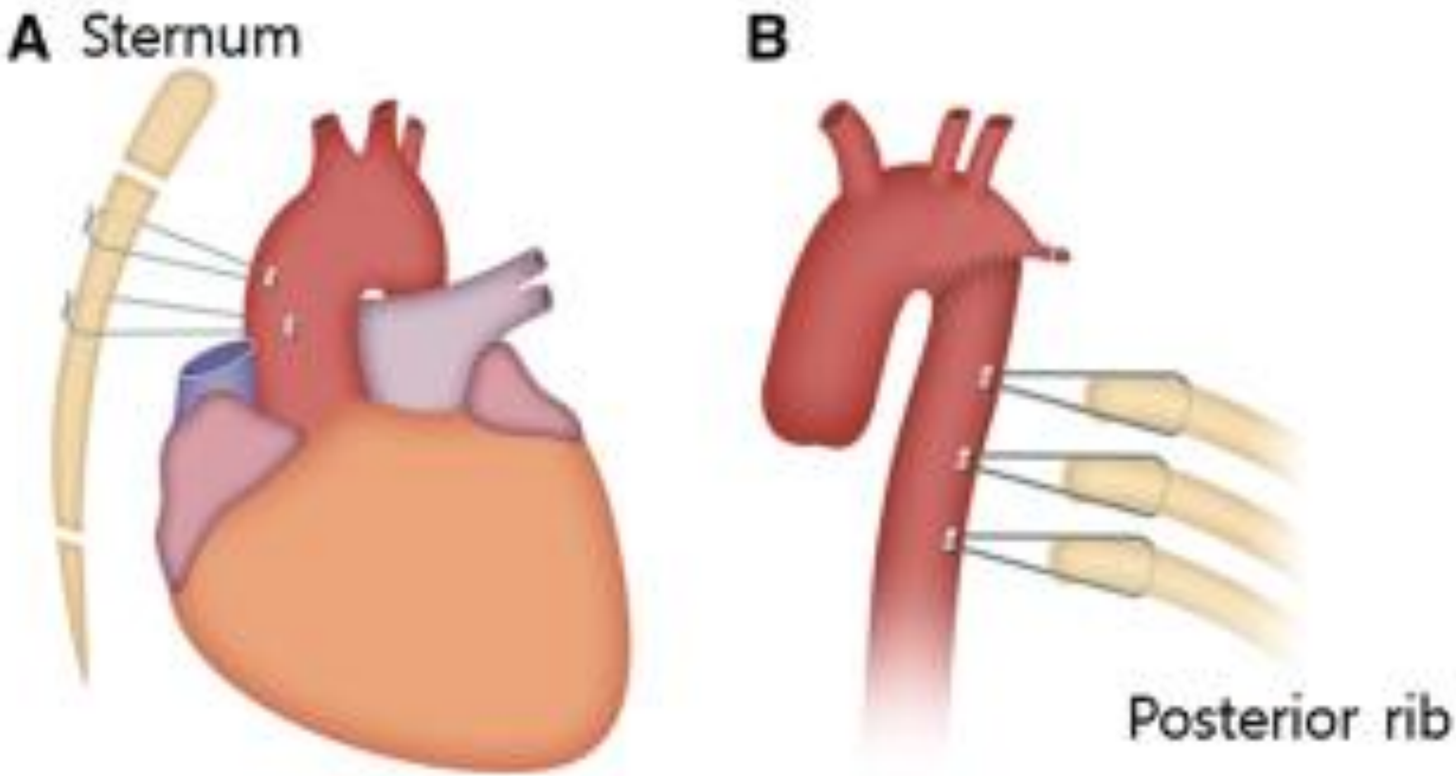
Woo Sung Jang · Woong-Han Kim · Kwangho Choi · JinHae Nam · Jin-Tae Kim · Jeong Ryul Lee · Yong Jin Kim · Gi Beom Kim

Received: 16 October 2013 / Accepted: 23 January 2014

Abstract Bronchoscopy-guided aortopexy is a surgical management option for patients with central airway obstruction after congenital heart surgery. This study aimed to evaluate the usefulness of bronchoscopy-guided aortopexy based on midterm follow-up evaluation with computed tomography (CT) and clinical outcome. From January 2004 to August 2011, bronchoscopy-guided aortopexy was performed for 16 patients (median age 0.5 years, M:F = 10:6) who had central airway obstruction caused by extrinsic compression (13 in the left main bronchus, 2 in the trachea, 1 in the diffuse trachea and bronchus) after congenital heart surgery. The surgical site for aortopexy was determined by the anatomic relationship between the aorta and the compressed bronchus according to preoperative CT and intraoperative bronchoscopy. The median follow-up period was 2.3 years. The ratios of the diameter and area of stenosis at the narrowed point were estimated using pre- and postoperative CT. Almost all the patients (15/16) showed relief of their preoperative symptoms. The median extubation time was 18 h. The stenosis diameter and area ratios significantly improved, as shown by with the immediate postoperative CT (7.7–48.5 %, $p = 0.003$; 54.8–80.5 %, $p = 0.006$). Airway stenosis of more than 75 % ($p = 0.013$), immediate diameter ratio improvement of <50 % ($p = 0.015$), preoperative severe respiratory insufficiency ($p = 0.038$), and male sex ($p = 0.024$) were associated with recurrent minor respiratory susceptibility. Bronchoscopy-guided aortopexy is a safe and reliable surgical management choice for central airway obstruction after congenital heart surgery. Furthermore, airway improvement after aortopexy was maintained during the midterm follow-up evaluation, according to CT measurements.

Keywords Aortic operation · Airway obstruction · Bronchoscopy · Computed tomography


A
B
C
D


A
Sternum
B
Posterior rib

Anterior Translocation of the Right Pulmonary Artery to Avoid Airway Compression in Aortic Arch Repair

Yun Hee Chang, MD, Si Chan Sung, MD, Hyungtae Kim, MD, Ki Seuk Choo, MD, Hyoung Doo Lee, MD, and Ji Ae Park, MD

Department of Thoracic and Cardiovascular Surgery, Seoul St. Mary's Hospital, College of Medicine, The Catholic University of Korea, Seoul; Thoracic and Cardiovascular Surgery, Research Institute for Convergence of Biomedical Science and Technology, Busan National University Yangsan Hospital, Busan; and Departments of Diagnostic Radiology, and Pediatrics, Busan National University Yangsan Hospital, Busan, Korea

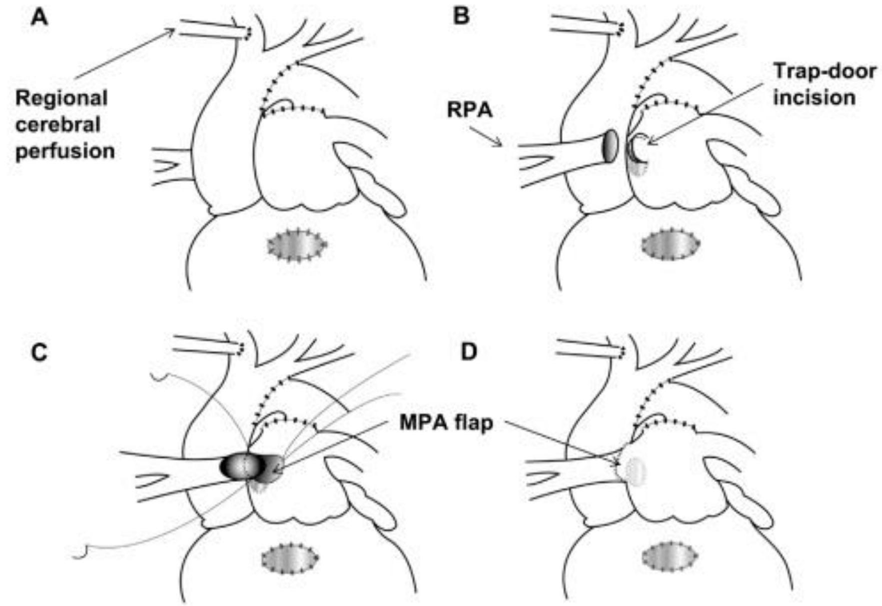
Background. Airway compression caused by an enlarged right pulmonary artery (RPA) in patients with a large shunt can usually be managed with intracardiac repair and concomitant anterior aortopexy. However, anterior aortopexy can be less effective or even dangerous in patients with coexisting arch anomaly due to excessive tension at the arch repair site. We have adopted anterior translocation of RPA without aortic transection in the group of patients with a high risk of postoperative airway compression. We reviewed the early and midterm results of this technique.

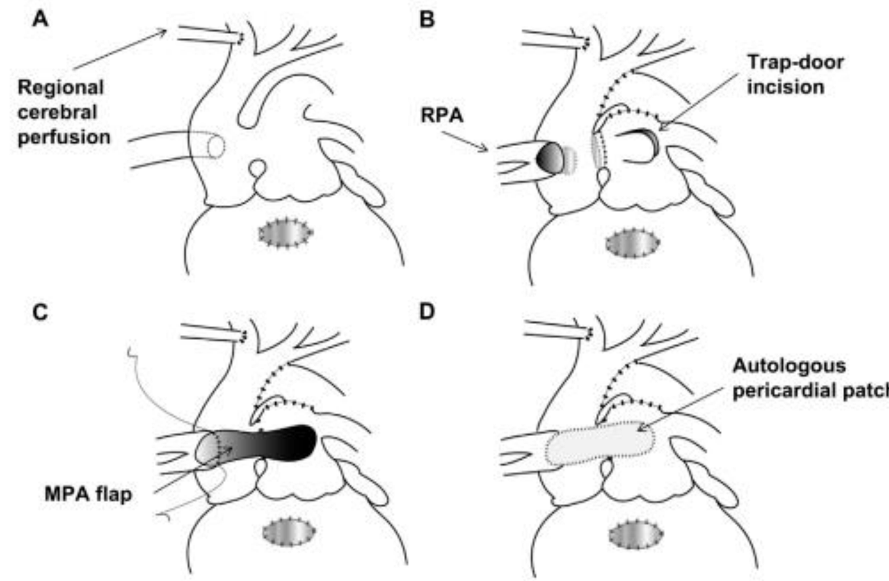
Methods. From February 2006 to January 2013, 8 patients underwent RPA anterior translocation as a concomitant procedure in one-stage repair of ventricular septal defect (VSD) and aortic arch anomaly to avoid postoperative airway problems. The enlarged RPA was disconnected from the main pulmonary artery (MPA) at its origin and was relocated anterior to the ascending aorta, and subsequently reimplanted to the U-shaped trapdoor incision at the anterolateral MPA wall. The mean age at operation was 34 days (median, 14 days, 6 to 77 days), and the mean body weight was 3.6 kg (2.15 to 5.5 kg). All patients had coarctation of the aorta and VSD except 1 who had aortic arch interruption. Five patients were dependent on a ventilator preoperatively. Six patients had evidence of preoperative bronchial compression (left; 4, right and left; 2), and 2 had a high probability of postoperative bronchial compression due to unusual anterior location of the descending aorta.

Results. There was no early or late death. There were no postoperative airway problems such as reintubation or left lung atelectasis. Widely patent RPA was confirmed on postoperative computed tomographic angiography in all patients. The mean follow-up duration was 54.0 ± 17.1 months. One patient required balloon angioplasty for mild stenosis at the clamping site 3 years after the operation. All patients had no RPA stenosis at the latest follow-up evaluation.

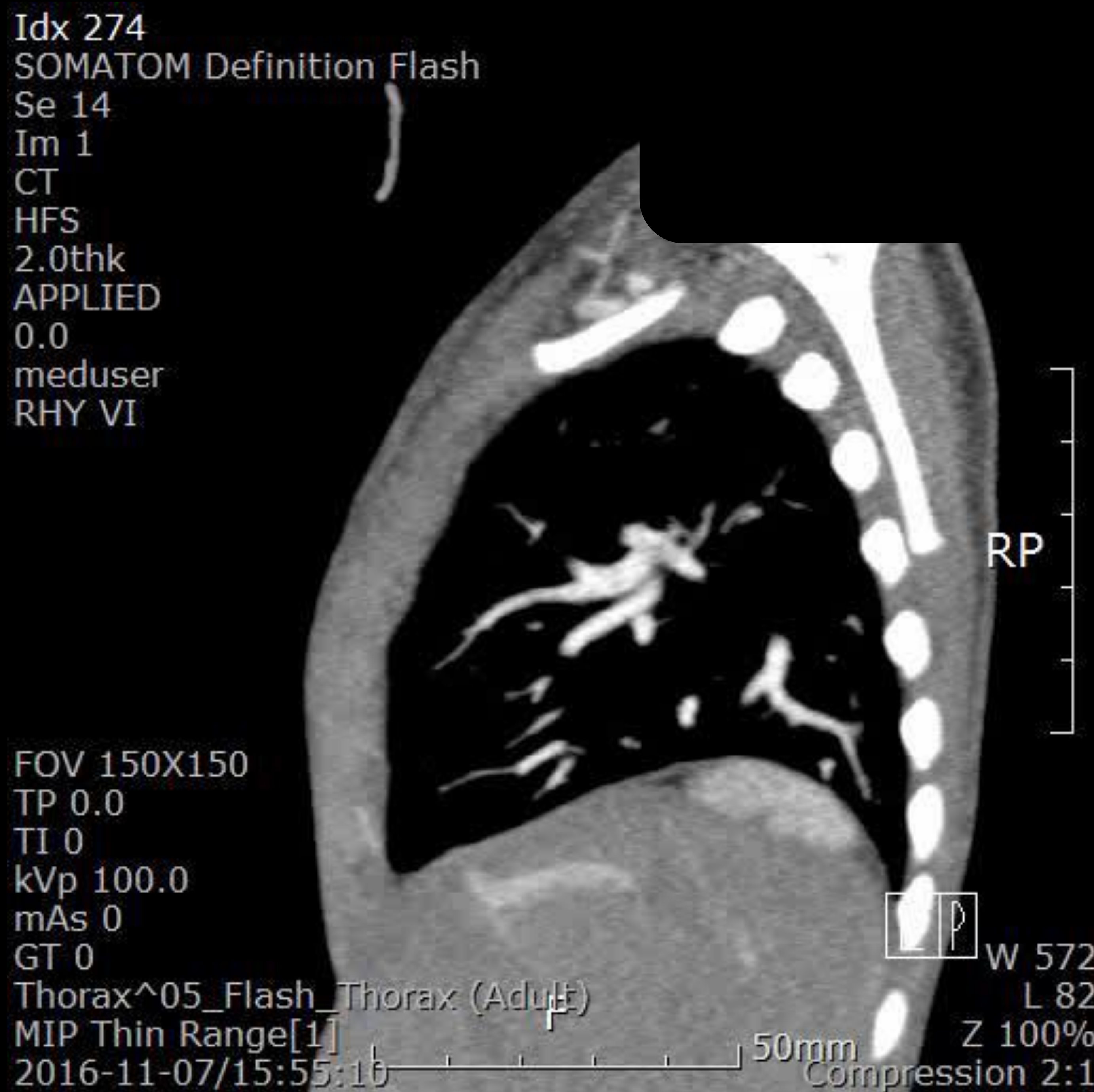
Conclusions. Anterior translocation of the RPA as a concomitant procedure in one-stage repair of VSD and arch anomaly is a safe and effective procedure to avoid postoperative airway problems in high-risk patients.

(Ann Thorac Surg 2013;96:2198–202)


A
Regional cerebral perfusion
B
RPA
Trap-door incision
C
MPA flap
D


A
Regional cerebral perfusion
B
RPA
Trap-door incision
C
MPA flap
D
Autologous pericardial patch


Idx 172
SOMATOM Definition Flash
Se 11
Im 1
CT
HFS
2.0thk
APPLIED
0.0
DELAY
RHY VI
L
FOV 140X140
TP -1222.0
TI 285
kVp 100.0
mAs 28(257 mA)
GT 0
Thorax^05_Flash_Thorax (Adult)
DELAY 2.0 B30f
2016-11-07/15:46:06
P
50mm
W 400
L 40
Z 100%
Compression 3:1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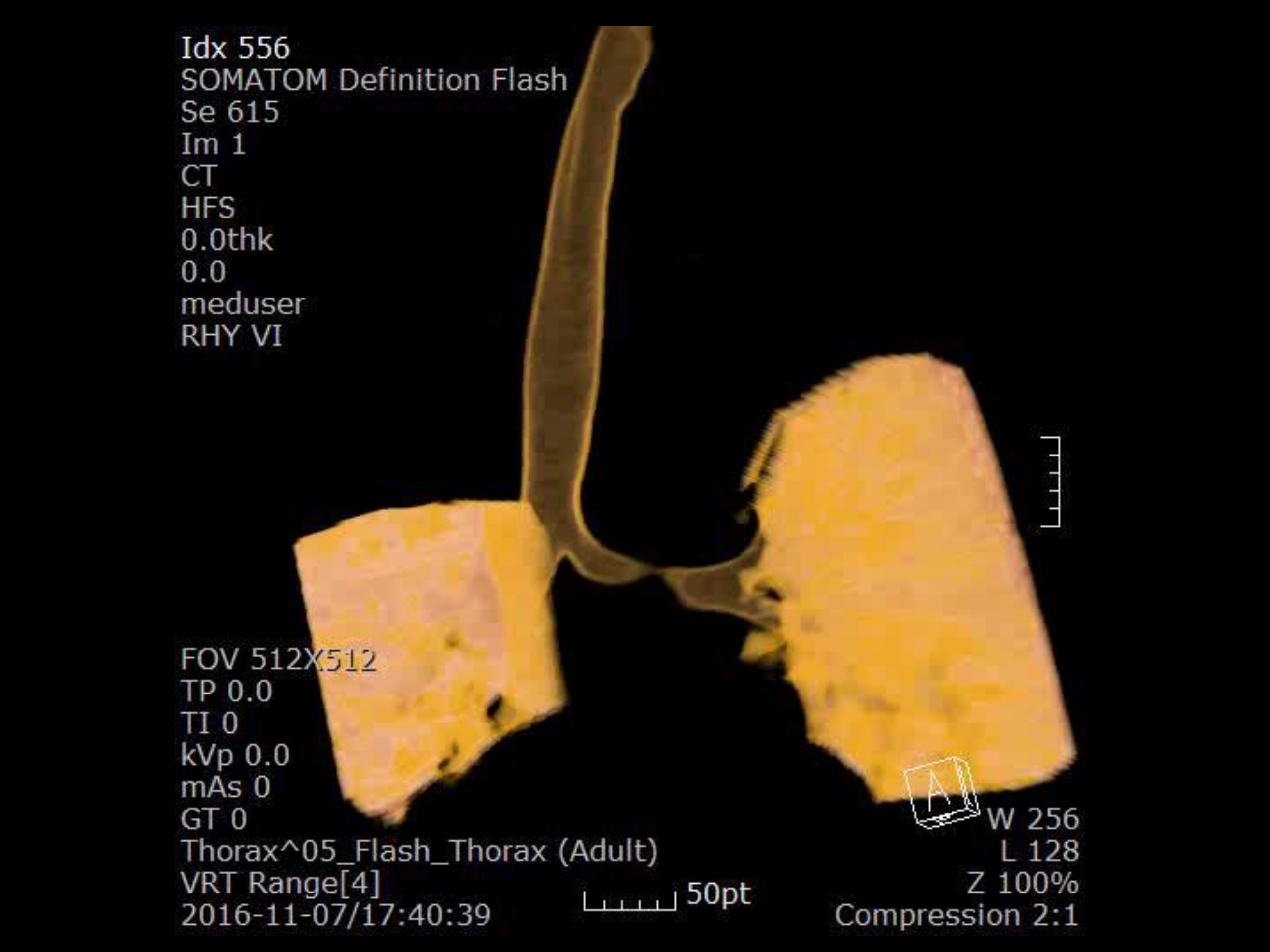
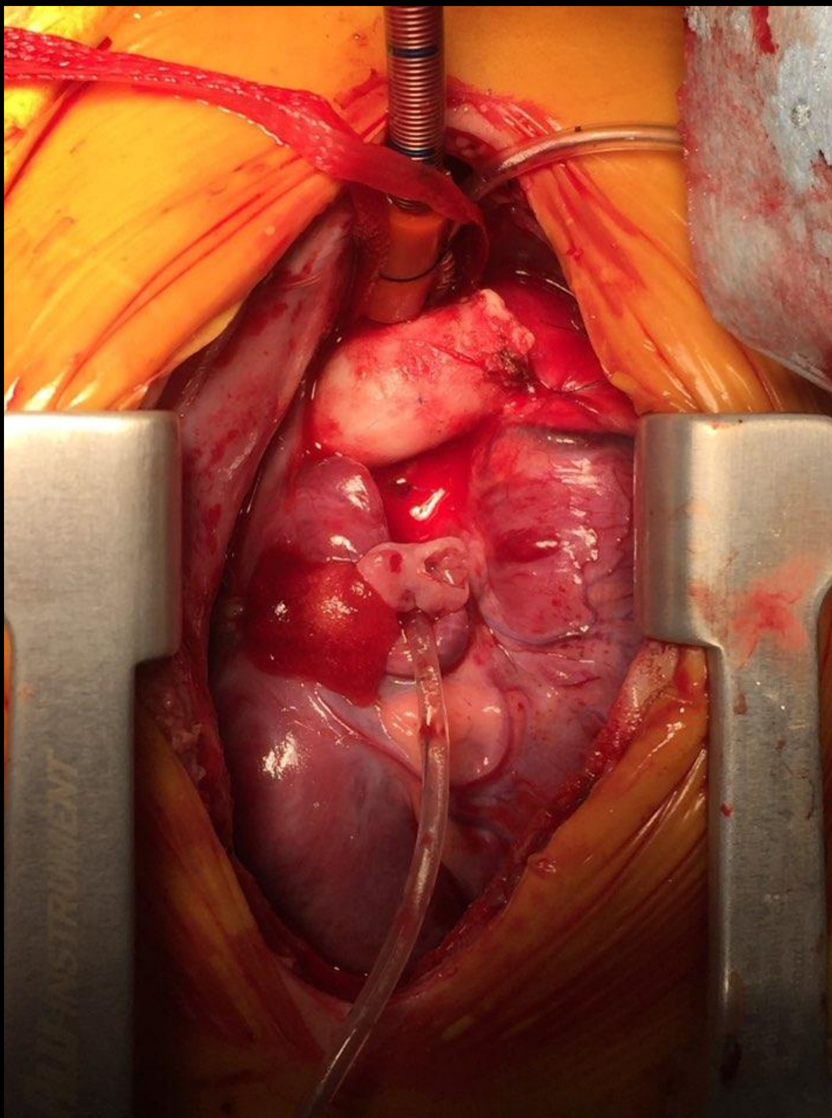
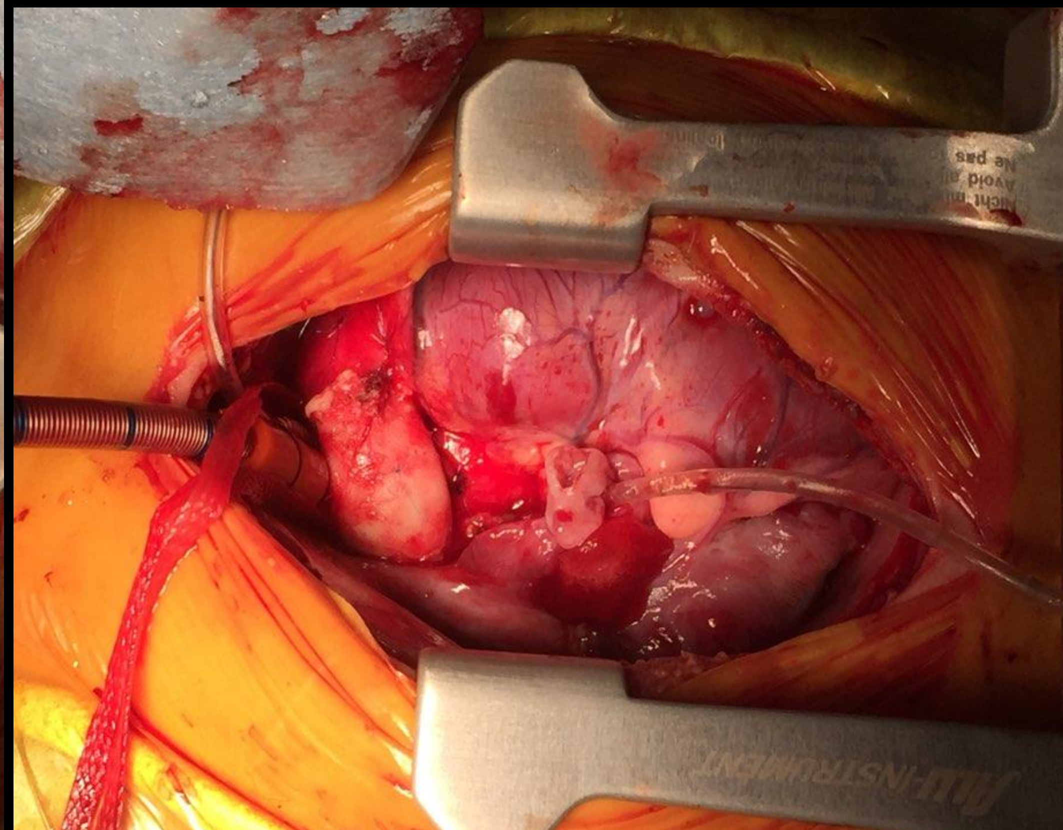
SOMATOM Definition Flash
Se 14
Im 1
CT
HFS
2.0thk
APPLIED
0.0
meduser
RHY VI
[redacted]
RP
FOV 150X150
TP 0.0
TI 0
kVp 100.0
mAs 0
GT 0
Thorax^05_Flash_Thorax (Adult)
MIP Thin Range[1]
2016-11-07/15:55:10
50mm
W 572
L 82
Z 100%
Compression 2:1


Idx 556
SOMATOM Definition Flash
Se 615
Im 1
CT
HFS
0.0thk
0.0
meduser
RHY VI
FOV 512X512
TP 0.0
TI 0
kVp 0.0
mAs 0
GT 0
Thorax^05_Flash_Thorax (Adult)
VRT Range[4]
2016-11-07/17:40:39
50pt
A
W 256
L 128
Z 100%
Compression 2:1

- **Paraplegia, 0.1~1%**
  - Related to
    - Prolonged distal clamp time
    - Low distal aortic pressure
      - Related to collateral vs.
      - Dramatic drop in the distal arterial pressure with the test clamp
      - Anomalous origin of SCA
    - Intraoperative hyperthermia
    - Hypotension/ Acidosis
  - To avoid
    - Reducing clamp time
    - Local hypothermia
    - Hypertension
    - Somatosensory potential (SEP)

Table 1. Reported Incidence of Paraplegia After Coarctation Repair

Author	Year	Patients	Paraplegia	Percent (%)
Brewer et al. [1]	1972	12,532	51	0.4
Pennington et al. [3]	1979	164	1	0.6
Lerberg et al. [4]	1982	334	5	1.5
Anyanwu et al. [5]	1984	253	1	0.4
Palantianos et al. [6]	1985	107	1	0.9
Keen [2]	1987	5,492	16	0.3
Backer et al.[a]	2006	384	1	0.3
Total		19,266	76	0.4

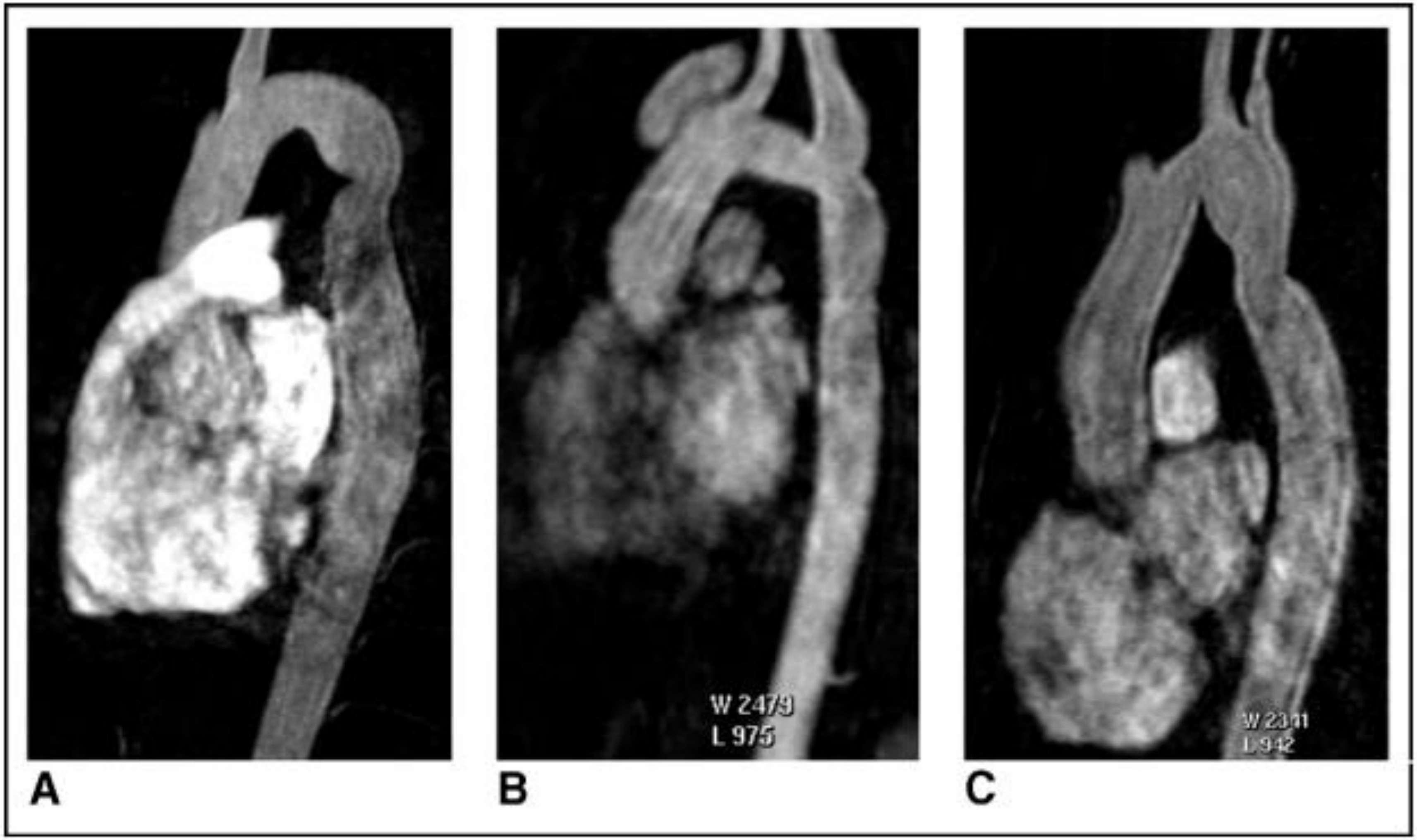
[a] Current report.

Backer et al. ATS 2006;82:964-72

Long term complications

- Re-stenosis
  - Influenced **by presence of residual ductal tissue** within the aorta
- HTN
  - More likely in repair at a later age
- Aortic aneurysm
  - Higher in onlay patch technique
- Bacterial endocarditis
- Neurologic abnormalities
  - Completeness of the circle of Willis

Long term complications



Normal Crenel Gothic

How successful is successful? Aortic arch shape after successful aortic coarctation repair correlates with left ventricular function

CrossMark

Jan L. Bruse, MSc,[a] Abbas Khushnood, MD,[a] Kristin McLeod, PhD,[b,c] Giovanni Biglino, PhD,[a] Maxime Sermesant, PhD,[c] Xavier Pennec, PhD,[c] Andrew M. Taylor, MD,[a] Tain-Yen Hsia, MD,[a] and Silvia Schievano, PhD,[a] for the Modeling of Congenital Hearts Alliance Collaborative Group*

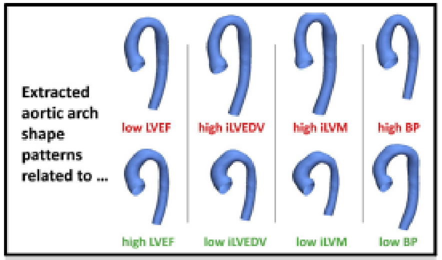
ABSTRACT

Objectives: Even after successful aortic coarctation repair, there remains a significant incidence of late systemic hypertension and other morbidities. Independently of residual obstruction, aortic arch morphology alone may affect cardiac function and outcome. We sought to uncover the relationship of arch 3-dimensional shape features with functional data obtained from cardiac magnetic resonance scans.

Methods: Three-dimensional aortic arch shape models of 53 patients (mean age, 22.3 ± 5.6 years) 12 to 38 years after aortic coarctation repair were reconstructed from cardiac magnetic resonance data. A novel validated statistical shape analysis method computed a 3-dimensional mean anatomic shape of all aortic arches and calculated deformation vectors of the mean shape toward each patient's arch anatomy. From these deformations, 3-dimensional shape features most related to left ventricular ejection fraction, indexed left ventricular end-diastolic volume, indexed left ventricular mass, and resting systolic blood pressure were extracted from the deformation vectors via partial least-squares regression.

Results: Distinct arch shape features correlated significantly with left ventricular ejection fraction ($r = 0.42$, $P = .024$), indexed left ventricular end-diastolic volume ($r = 0.65$, $P < .001$), and indexed left ventricular mass ($r = 0.44$, $P = .014$). Lower left ventricular ejection fraction, larger indexed left ventricular end-diastolic volume, and increased indexed left ventricular mass were identified with an aortic arch shape that has an elongated ascending aorta with a high arch height-to-width ratio, a relatively short proximal transverse arch, and a relatively dilated descending aorta. High blood pressure seemed to be linked to gothic arch shape features, but this did not achieve statistical significance.

Conclusions: Independently of hemodynamically important arch obstruction or residual aortic coarctation, specific aortic arch shape features late after successful



SSM extracted aortic arch shape features related to cardiac function.

Central Message

Worse left ventricular function and increased left ventricular volume and mass are associated with unique aortic arch shape features late after coarctation repair.

Perspective

Despite successful aortic coarctation repair, 3D SSM analysis revealed associations of abnormal arch geometry with lower LVEF, increased left ventricular volume, and higher left ventricular mass. 3D morphologic shape features may provide a tool for risk stratification in patients after aortic coarctation repair.

See Editorial Commentary page 428.

See Editorial page 415.

Bruse et al. JTCS 2017


COA113
COA099
COA089
COA081
COA074_2
COA049
COA043
COA037
COA031
COA025
COA018
COA013
COA007
COA117_2
COA108
COA098
COA086
COA080
COA072
COA048
COA040
COA034
COA030
COA023
COA017
COA011_2
COA004_2
COA106
COA093
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COA076
COA054
COA045
COA039_2
COA033
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COA003_2
COA105
COA090_2
COA083
COA075
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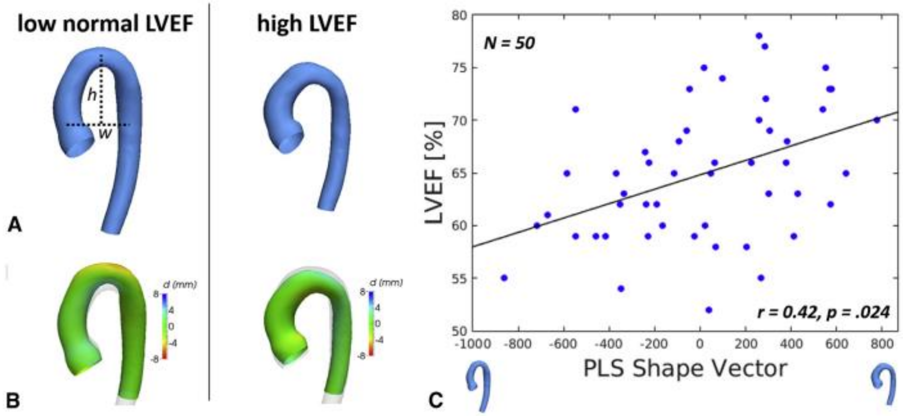
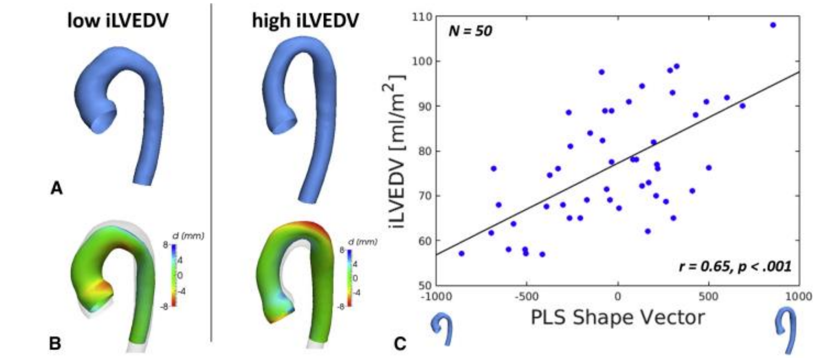
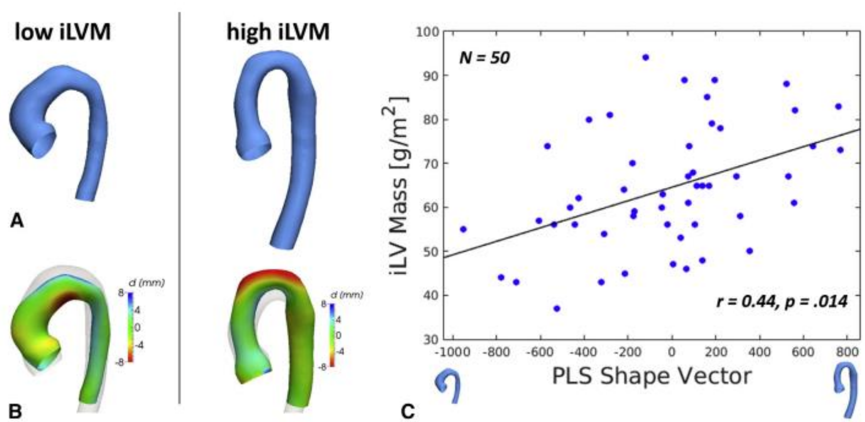
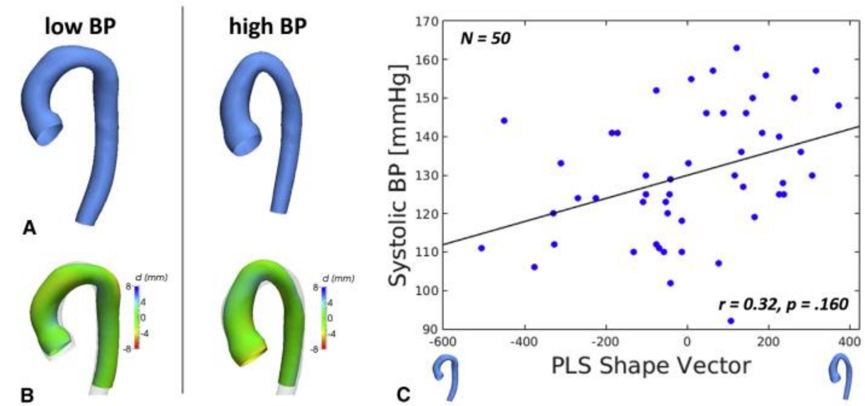
COA019
COA015_2
COA005_2
COA002
Template,
3D mean
shape


low normal LVEF
high LVEF
h
w
A
B
d (mm)
8
4
0
-4
-8
N = 50
LVEF [%]
r = 0.42, p = .024
PLS Shape Vector
C


low iLVEDV
high iLVEDV
A
B
d (mm)
N = 50
iLVEDV [ml/m²]
r = 0.65, p < .001
PLS Shape Vector
C


low iLVM
high iLVM
A
B
d (mm)
N = 50
iLV Mass [g/m²]
r = 0.44, p = .014
PLS Shape Vector
C

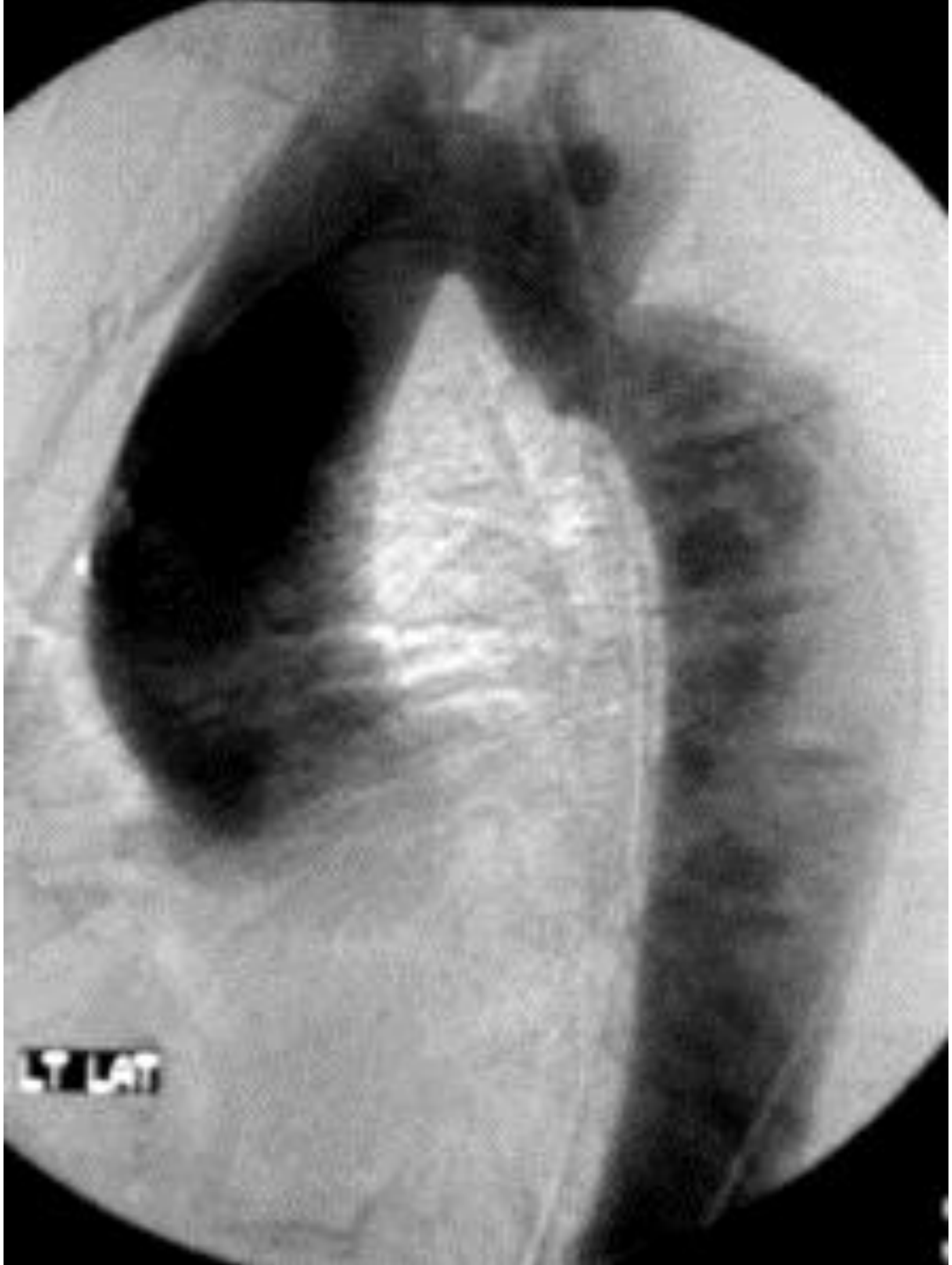

low BP
high BP
A
B
d (mm)
N = 50
Systolic BP [mmHg]
r = 0.32, p = .160
PLS Shape Vector
C

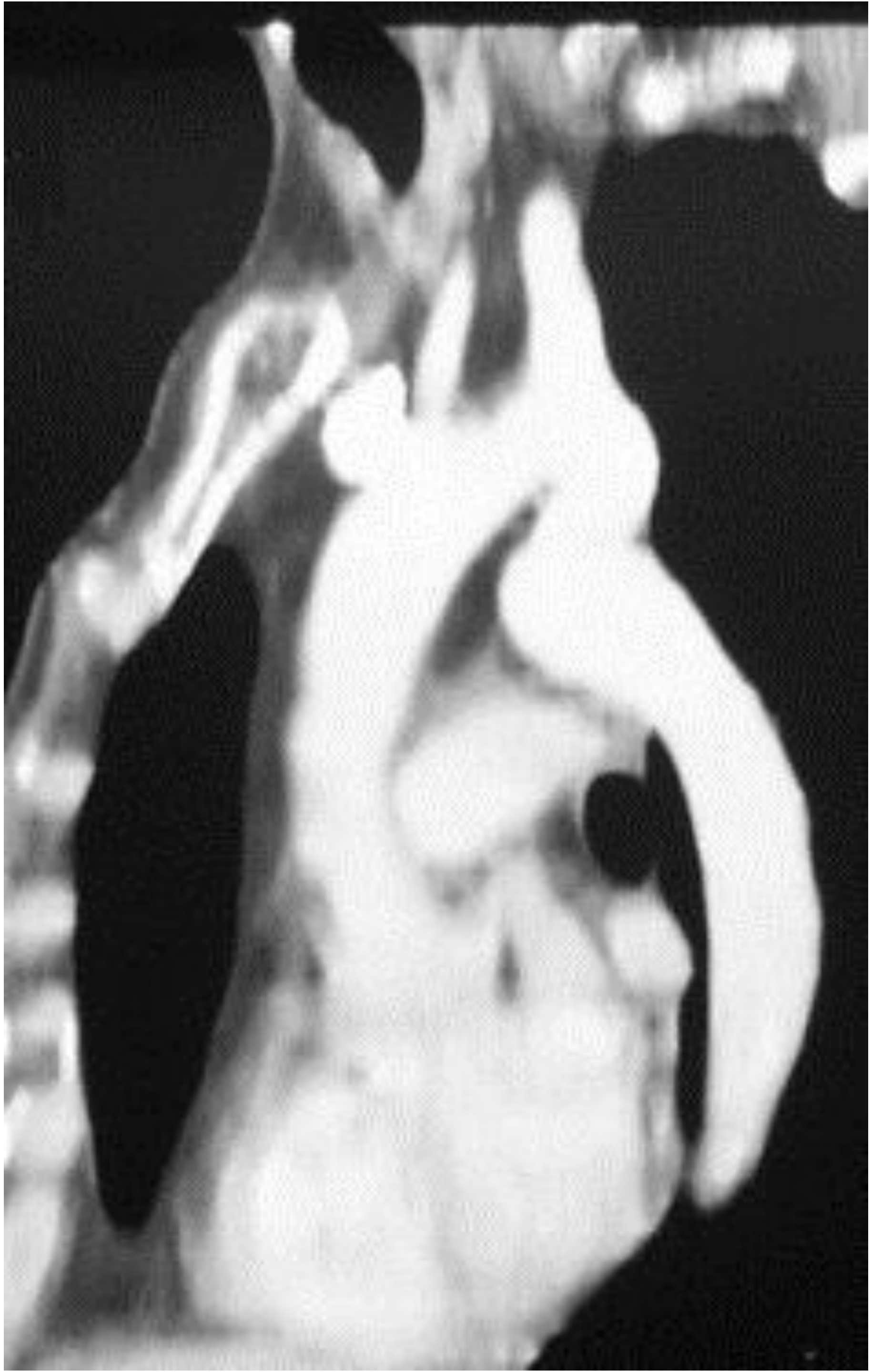
Prevention of recoarctation

- Ideal operative procedure
  - Successfully address transverse arch hypoplasia (if present)
  - **Resection of all ductal tissue**
  - Prevention of residual circumferential scarring at the aortic anastomosis site
  - **Tension free anastomosis**
- Factors
  - Younger age at operation
  - Presence of aortic arch hypoplasia remain risk factors for recoarctation

Pseudocoarctation

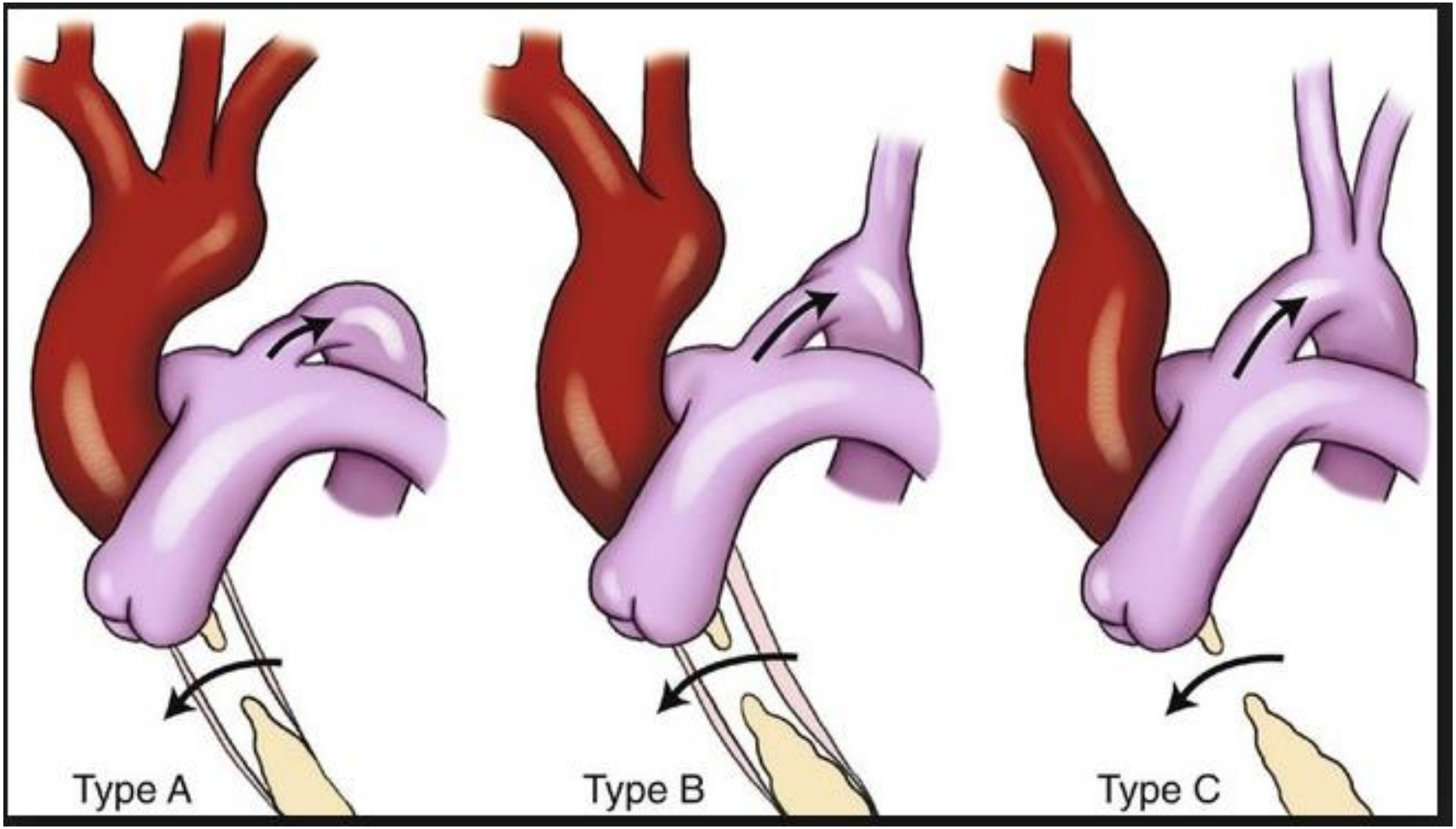
- Kink at insertion site of ligamentum arteriosum
- Minimal gradient across deformity


LT LAT



Interrupted aortic arch

Interrupted Aortic Arch

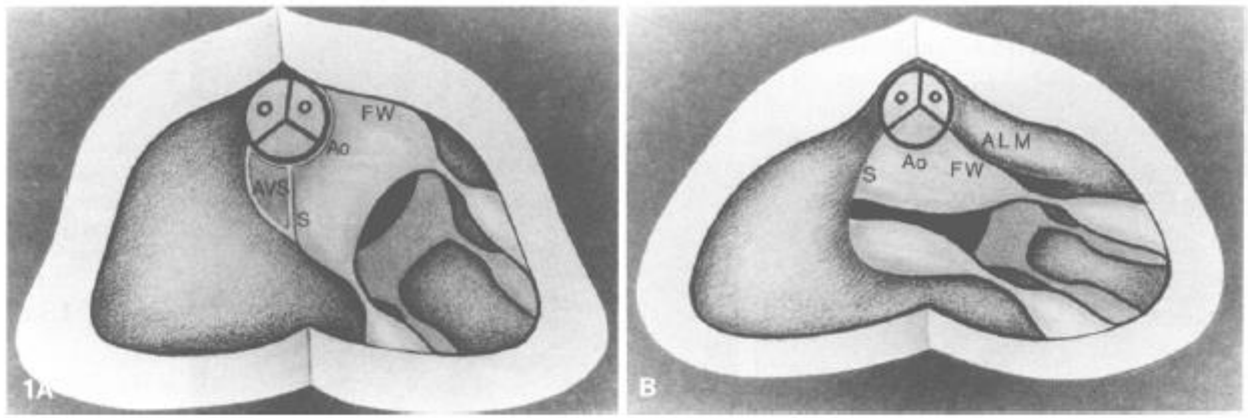


(40%) (55%) (5%)

Pathophysiology of IAA

- Distal blood flow is dependent on a patent ductus arteriosus.
- Spontaneous ductal closure results in systemic hypoperfusion, metabolic acidosis, and end-organ failure
- $Sat_{\text{upper ext.}} > Sat_{\text{lower ext.}}$
- Critically ill neonate in severe CHF
- Severe pulmonary HTN in patients with VSD, ASD

- Commonl
- LVOTO
  - Posterio
  - Promine leaflet o
- VSD : 70
- Truncus a
- TGA, DO
- AP windo
- 1/3 of DiC
- Berry syn



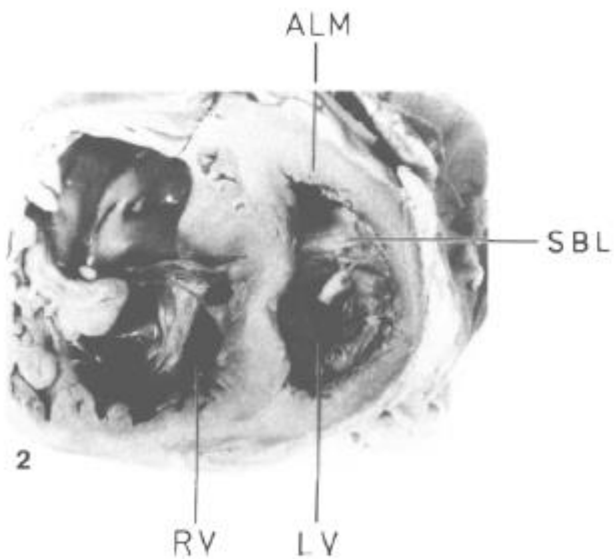
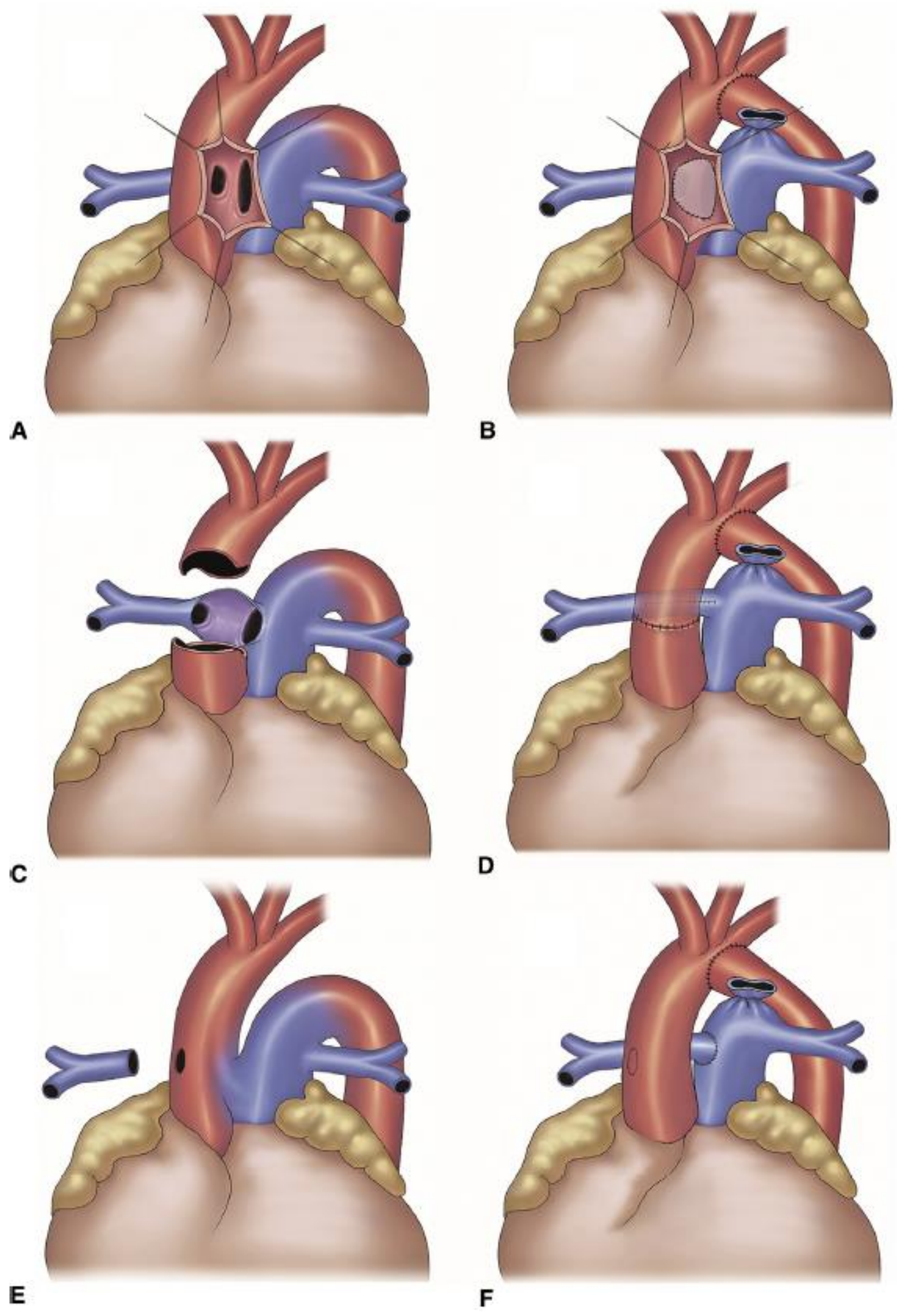


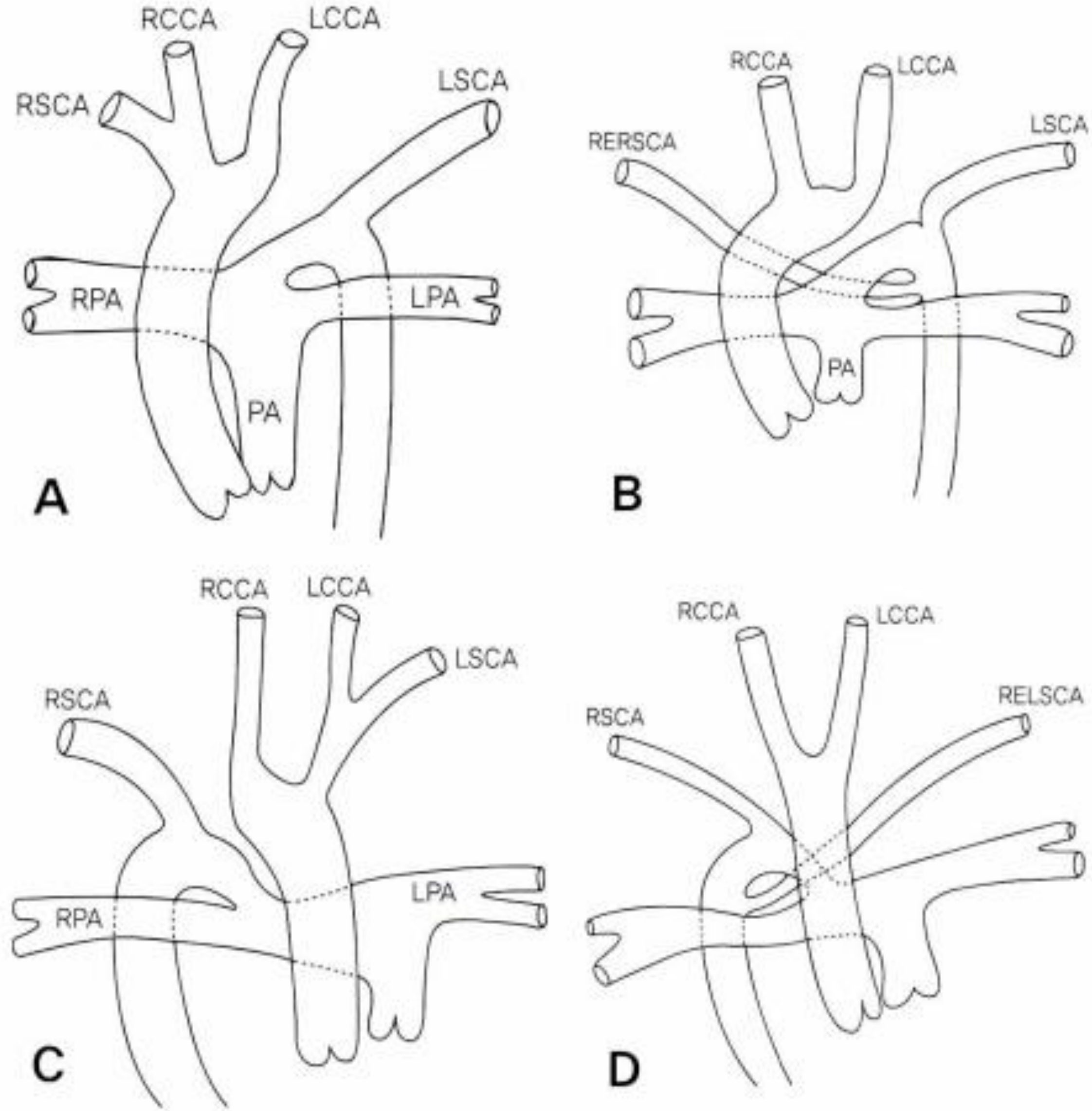
Fig. 1. LVOT and AV valve attachment. **(A)** Normal heart: The LVOT gives wide access to the aortic orifice. *AO*, aortic attachment of the aortic mitral leaflet; *AVS*, atrioventricular septum; *FW*, free-wall attachment; *S*, septal attachment. **(B)** AVSD: The LVOT is bounded laterally by the anterolateral muscle bundle (*ALM*), inferiorly by the superior bridging leaflet, and medially by the subaortic part of the ventricular septum.

Fig. 2. Short-axis cross-section of AVSD showing the clockwise rotation of the superior bridging leaflet (*SBL*), resulting in its position being perpendicular to the ventricular septum.

- Interrupt
- Distal ac
- RPA from
- Intact ve



Anatomic Variants



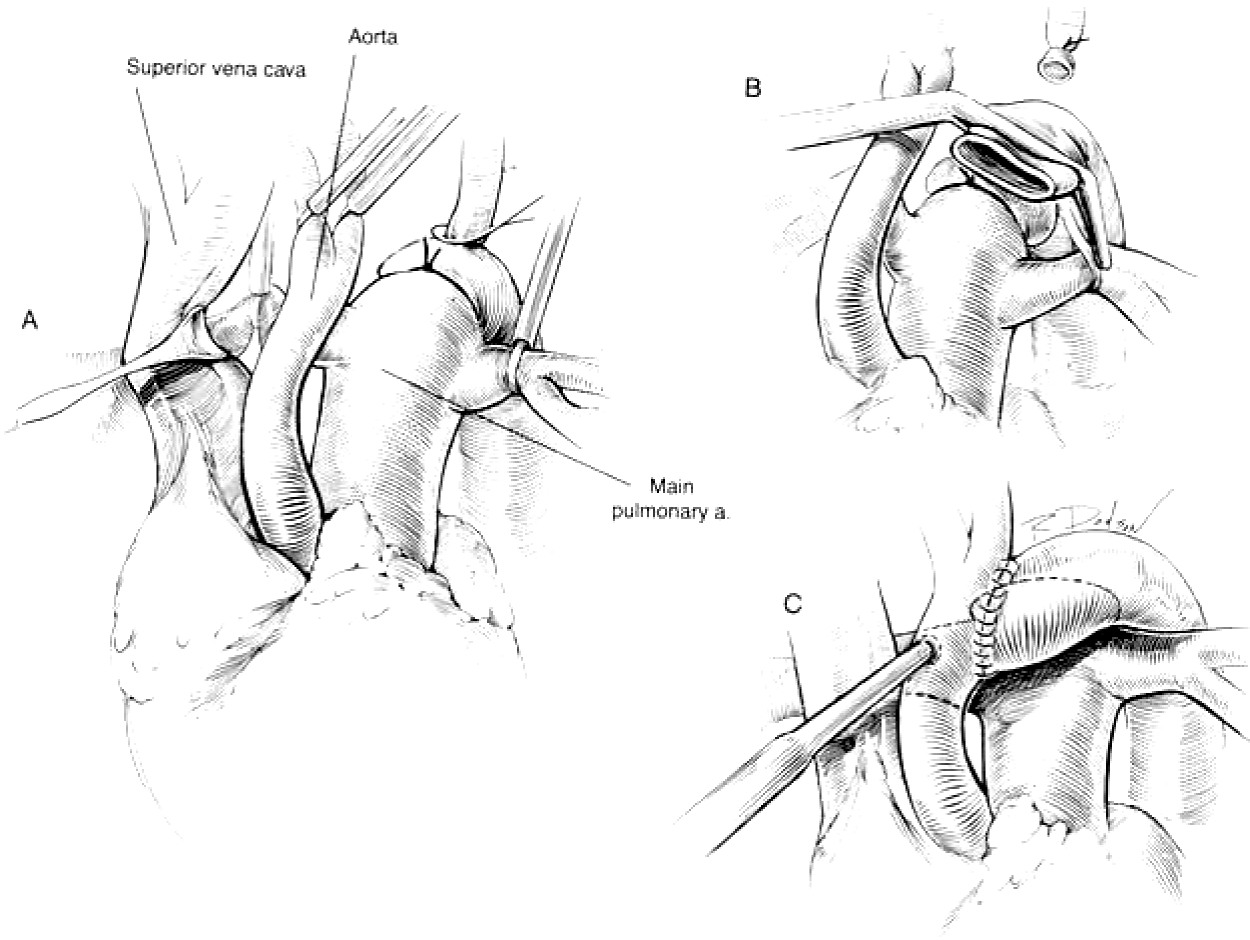
Medical treatment

- PGE1
- CHF management
- Consider mechanical ventilator Tx.
  - $PaCO_2$ 40~50 mmHg
- iCa level check

Surgical Treatment

- Indication
  - Urgent operation is advisable
- Technique
  - Direct anastomosis
    - Resection and end to end anastomosis
    - Extended resection and end to end anastomosis
  - Interposition of graft
- One stage repair

IAA Type B Operative Procedure



Operative results

- Mortality
  - Early death
    - Acute or subacute HF with or without multisystem failure
    - One stage reapir: Under 10%
    - IAA repair & second repair : 12~47%
  - Late death
    - Survival
      - Good

- Incremental risk factors for premature death
  - Coexisting cardiac anomalies
  - Location of interruption
  - Preoperative Condition
  - Earlier date of operation

Operative results

- Morbidity
  - Early period
    - Recurrent laryngeal n or phrenic n injury
    - Left main bronchus compression
    - Paraplegia
  - Late Period
    - Restenosis
    - Development of subaortic stenosis