Anomalous Origin of the Coronary Arteries

(Congenital Anomalies of the Coronary Arteries)

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Classification of Coronary Artery Anomalies (1)

Anomalies of Origin and Course

I Anomalous location of coronary ostium:
- a High ostium
- b Commissural ostium

II Anomalous origin of coronary artery from opposite sinus with one of four courses:
- a Interarterial
- b Transseptal
- c Retroaortic
- d Prepulmonic

III Anomalous origin of coronary artery from pulmonary trunk:
- Type 1: Left coronary artery
- Type 2: Right coronary artery
- Type 3: Circumflex coronary artery
- Type 4: Left and right coronary arteries

IV Single coronary artery

V Multiple ostia

VI Anomalous origin of coronary artery from noncoronary sinus

VII Duplication of coronary arteries

Source: Kirklin/Barrat- Boyes Cardiac Surgery 2013, 4th ed, Elsevier, P.1644-1666

Classification of Coronary Artery Anomalies (2)

Anomalies of Intrinsic Coronary Arterial Anatomy

I Congenital ostial stenoses
II Coronary artery ectasia or aneurysm
III Myocardial bridging

Anomalies of Termination

I Congenital coronary artery fistula
II Extracardiac termination

Modified from Kayalar and colleagues.[K5]

Source: Kirklin/Barrat- Boyes Cardiac Surgery 2013, 4th ed, Elsevier, P.1644-1666

Origin or Connection

- Embryologic information: proximal coronary arteries grow from the peritruncal area into the aorta
- Therefore: “the anomalous artery arises from” is inappropriate
- Anomalous connection of LCA to Pulmonary trunk (instead of Anomalous origin of LCA from PT)

Source: Kirklin/Barrat- Boyes Cardiac Surgery 2013, 4th ed, Elsevier, P.1644-1666

Benign or potentially serious isolated coronary artery anomalies (1)

	No.	Incidence (%)	Anomalies (%)
Benign			
Separate origin of LAD and CX in LSV	513	0.41	30.4
CX from RSV or RCA	467	0.37	27.7
Coronary artery from PSV			
LMT from PSV	1	0.0008	0.06
RCA from PSV	4	0.003	0.24
Anomalous origin from ascending aorta			
LMT from aorta	16	0.013	0.95
RCA from aorta	188	0.15	11.2
Absent CX (''super-dominant RCA'')	4	0.003	0.24
Intercoronary communication	3	0.002	0.18
Small coronary artery fistulae	163	0.12	9.7
Total	1,359	1.07	80.6

*LAD, left anterior descending; CX, circumflex; RCA, right coronary artery; LMT, left main trunk; LSV, left sinus of Valsalva; RSV, right sinus of Valsalva; PSV, posterior sinus of Valsalva.

Source: Yamanaka O, Hobbs RE. Cathererization and Cardiovascular Diagnosis (1990);21:28-40

Benign or Potentially serious isolated coronary artery anomalies (2)

Potentially serious			
Coronary artery from pulmonary artery			
LMT from pulmonary artery	10	0.008	0.59
LAD from pulmonary artery	1	0.0008	0.06
RCA from pulmonary artery	2	0.002	0.12
Coronary origin from opposite aortic sinus			
LMT from RSV	22	0.017	1.3
LAD from RSV	38	0.03	2.3
RCA from LSV	136	0.107	8.1
Single coronary artery[a]			
R-I	1	0.0008	0.06
R-II	19	0.015	1.1
R-III	5	0.004	0.30
L-I	20	0.016	1.2
L-II	11	0.009	0.65
Multiple or large sized fistulae	62	0.05	3.7
Total	327	0.26	19.4

*LAD, left anterior descending; CX, circumflex; RCA, right coronary artery; LMT, left main trunk; LSV, left sinus of Valsalva; RSV, right sinus of Valsalva; PSV, posterior sinus of Valsalva.

Source: Yamanaka O, Hobbs RE. Cathererization and Cardiovascular Diagnosis (1990);21:28-40

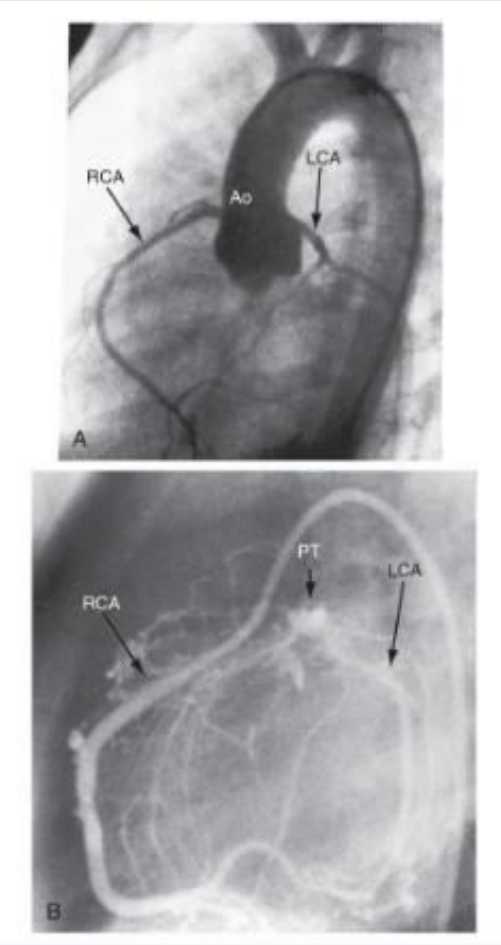
Anomalous connection of Left Coronary Artery (LCA) to Pulmonary Trunk

(Anomalous origin of LCA from the PT: ALCAPA)

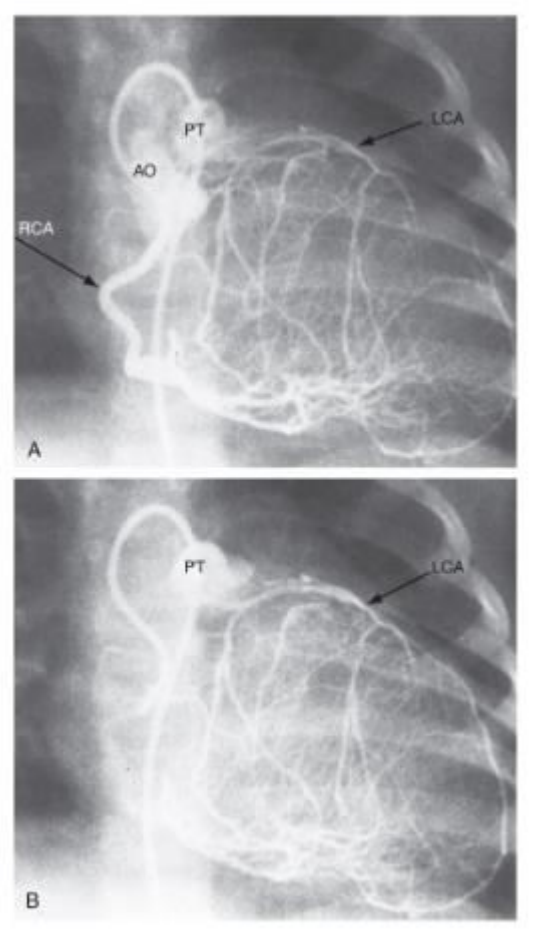
Pathology of ALCAPA

- Anomalous LCA: thin-walled, resembles a venous channel
- RCA: originates from aorta; dilated and tortuous

Aortogram: normal case and ALCAPA case



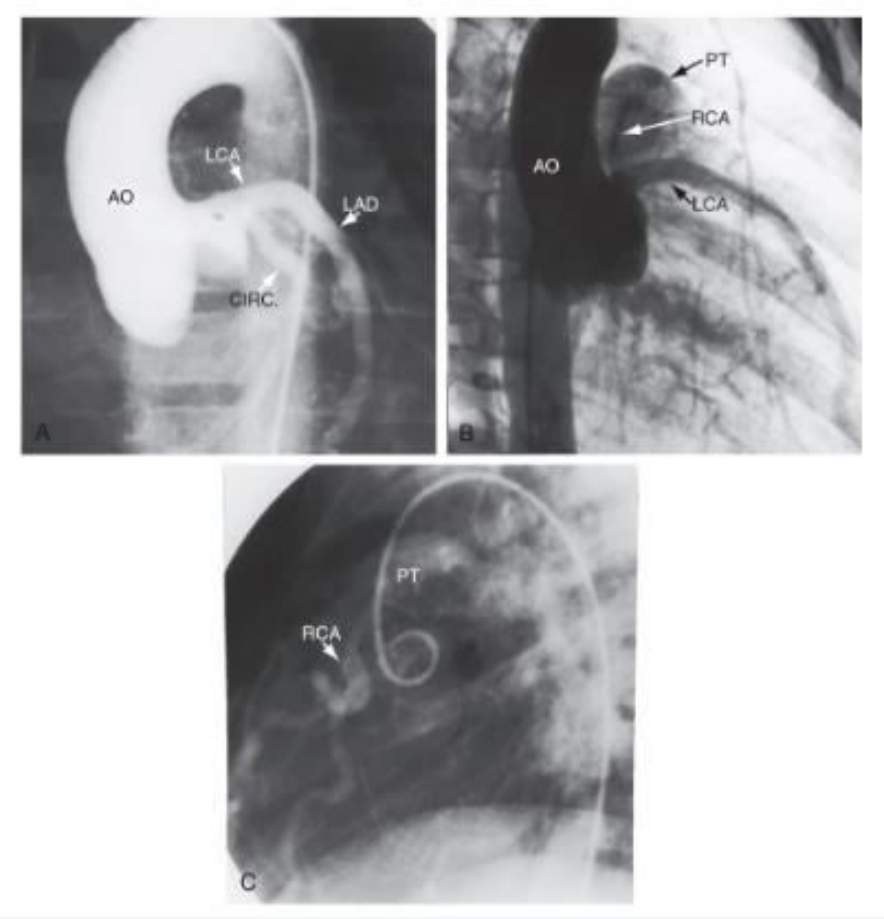
(A) a healthy 5-year-old girl: normal LCA and RCA
(B) A 4-year-old girl: LCA from PT



(A) a 4-year-old girl: LCA arises from PT
(B) Intercoronary anatomoses from LCA, filled by RCA, and was into PT

Source: Perloff's Clinical Recognition of Congenital Heart Disease, 4th ed, 2012. Elsevier, p. 394- 405.

Aortogram: 4-year-old boy/ALCAPA



A: LCA originates from Ao, divides into LAD and LCx.
B: Intercoronary anastomoses.
C: RCA enters the PT

Source: Perloff's Clinical Recognition of Congenital Heart Disease, 4th ed, 2012. Elsevier, p. 394-405.

History of ALCAPA (1)

- Both coronary arteries from PT: most severe
- 3 general patterns:
  - Serious symptoms in early infancy, death < 1 yo
  - Early symptoms followed by gradual attenuation or disappearance
  - Absence or virtual absence of symptoms with survival to adulthood
- 15% anomalous of LCA survive to adulthood

History of ALCAPA (2)

- Symptoms of heart failure after 2 months of age: irritability, dyspnea, wheezing, cough, diaphoresis, aggravated by feeding, crying, poor growth, death by heart failure
- 1/3 death by sudden death
- Hoarseness: by impingement of dilated PA on the recurrent laryngeal nerve

Physical exam of ALCAPA

- Holosystolic murmur of mitral regurgitation caused by ischemic papillary muscle dysfunction
- Short apical mid-diastolic murmur
- Continuous murmur by flow through intercoronary anastomoses; site similar to PDA murmur

ECG/ ALCAPA

- Deep narrow q waves in DI, aVL
- LVH
- Left axis deviation

A 4-year-old girl with ALCAPA. The QRS axis is indeterminate; q wave in 1 and aVL; and LVH (deep S wave in V3, prominant R wave in V6).

Source: Perloff's Clinical Recognition of Congenital Heart Disease, 4th ed, 2012. Elsevier, p. 394- 405.

Chest X-ray/ALCAPA (1)

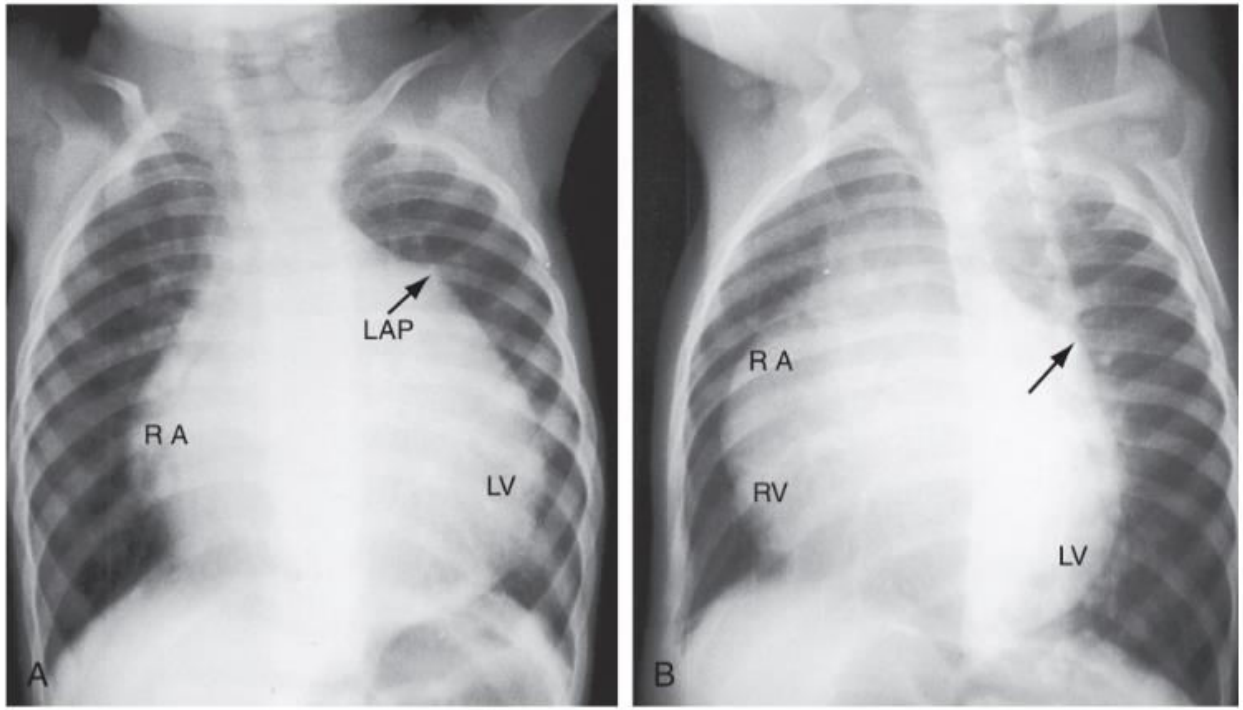


FIGURE 21-14 X-rays from a 10-month-old female with anomalous origin of the left coronary artery from the pulmonary trunk. **A,** The left atrial appendage (LAP) is conspicuous, a dilated left ventricle (LV) occupies the apex, and an enlarged right atrium (RA) occupies the right cardiac border. **B,** Left anterior oblique projection. The anterior border of the heart is formed by the right atrium and right ventricle (RV), the posterior border is formed by the dilated left ventricle, and a large left atrium lies beneath the left bronchus (arrow).

Source: Perloff's Clinical Recognition of Congenital Heart Disease, 4th ed, 2012. Elsevier, p. 394- 405.

Chest X-ray/ALCAPA (2)

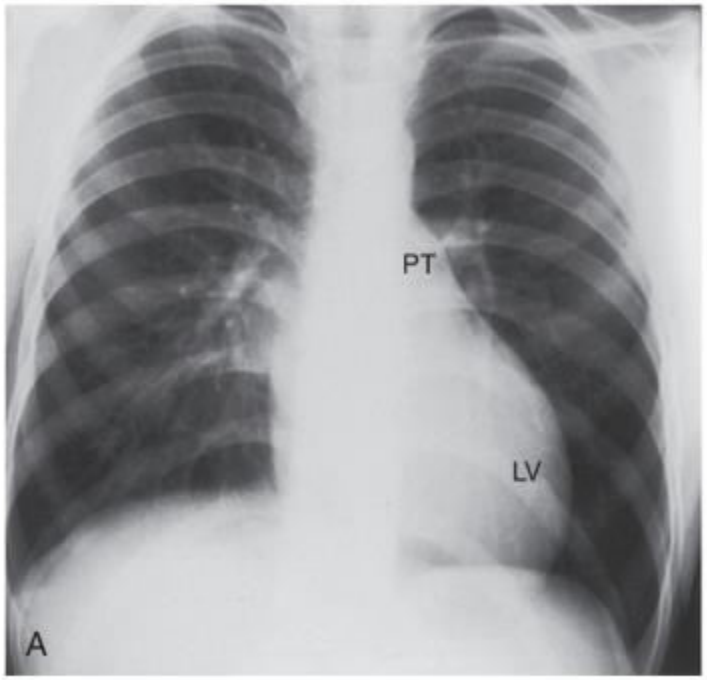
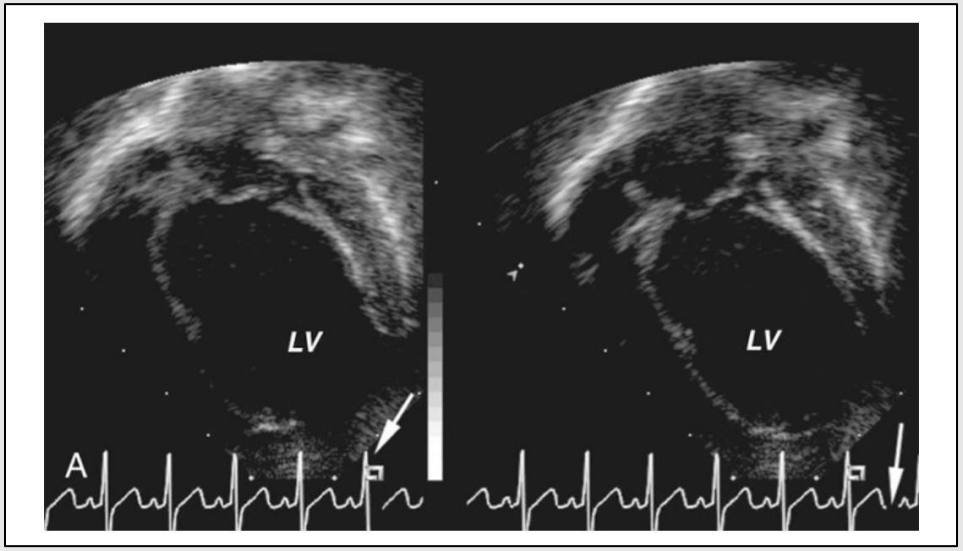
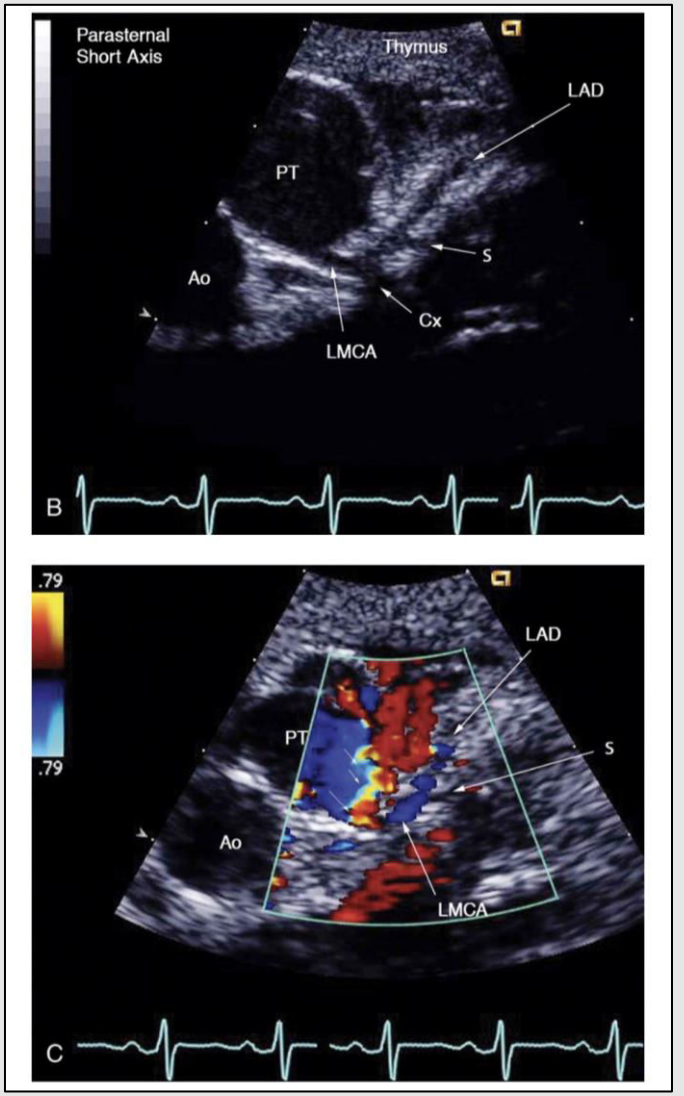


FIGURE 21-15 X-rays from the 13-year-old boy whose phonocardiogram is shown in Figure 21-7. A convex left ventricle (LV) occupies the apex, and there is mild prominence of the pulmonary trunk (PT). The lateral view shows displacement of the barium-filled esophagus (arrows) by a moderately enlarged left atrium.

Source: Perloff's Clinical Recognition of Congenital Heart Disease, 4th ed, 2012. Elsevier, p. 394-405.

Echocardiography/ ALCAPA





A, apical 4-chamber view: dilated LV cavity, reduced LV systolic function
B, LMCA arising from PA
C, Color Doppler image shown in B

Source: Kirklin/Barrat- Boyes Cardiac Surgery 2013, 4th ed, Elsevier, P.1644-1666

Comparison of anomalous Right versus Left Coronary Artery Connection to Pulmonary Trunk

	Right Coronary Artery	Left Coronary Artery
Prevalence	0.002%	0.008%
Age at presentation	>2 years	<1 year
Heart failure	No	Yes
Ischemia	No	Yes
Sudden death	Rare	Yes
Physical exam	Murmur	Heart failure, ±systolic murmur
ECG findings	Nonspecific	Ischemia, Q waves in I and aVL >80%
Reimplantation	Yes	Yes

Source: Kirklin/Barrat- Boyes Cardiac Surgery 2013, 4th ed, Elsevier, P.1644-1666

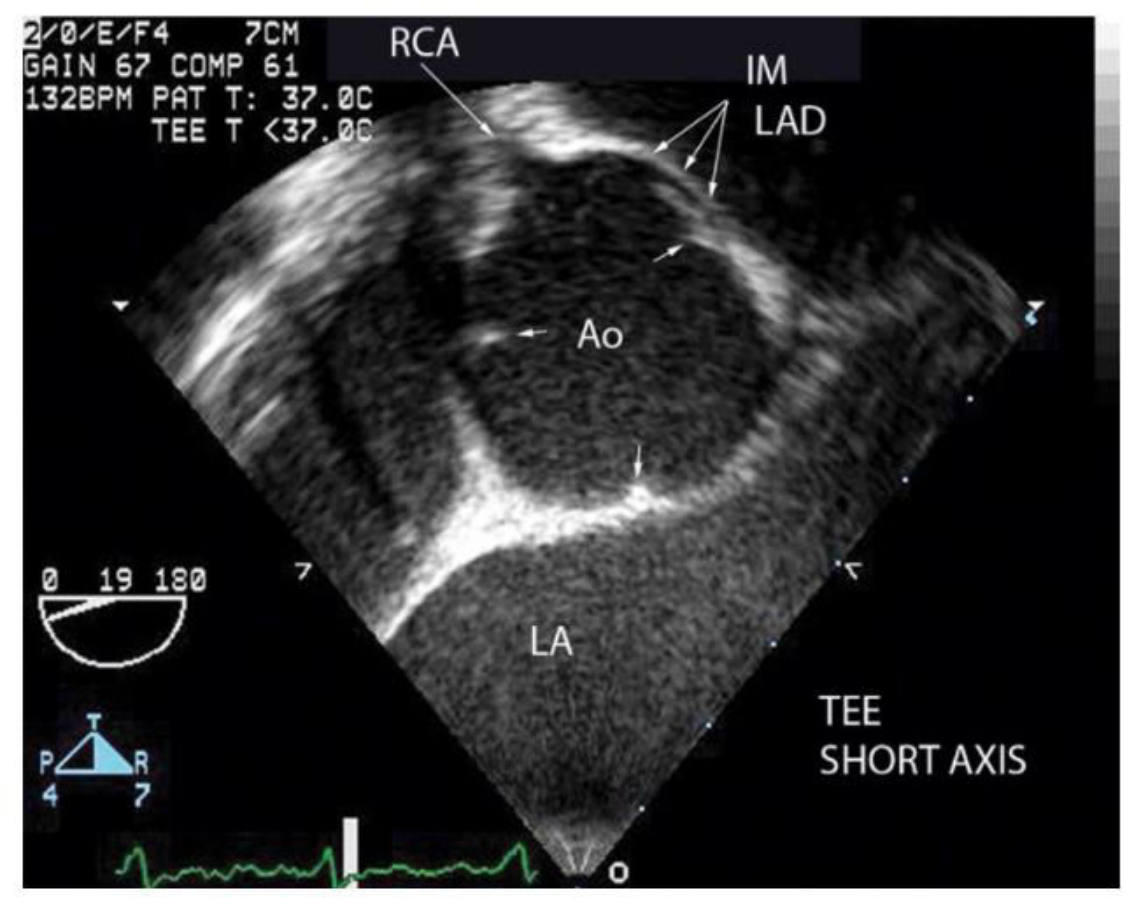
Anomalous Connection of a Main Coronary Artery to Aorta

(Anomalous origin of a main Coronary Artery from Aorta)

Anomalous Connection of a main Coronary Artery to Aorta

- LMCA connects to the aorta in site other than the left coronary sinus or sinotubular junction
- RCA connects to a site other than right coronary sinus
- Consequences:
  - Interarterial course
  - Intramural course
  - Ischemia
  - Clinical events

TEE of anomalous aortic origin of LMCA from right aortic sinus



Source: Kirklin/Barrat- Boyes Cardiac Surgery 2013, 4th ed, Elsevier, P.1644-1666

Morphology

- Sites of anomalous origin of LMCA
  - 2 coronary ostia close side by side
  - Single enlarged ostium in the right sinus with normal right RCA and anomalous LMCA
- Sites of anomalous origin of RCA
  - More common than anomalous origin of LMCA
  - Same ostial abnormalities as anomalous origin of LMCA

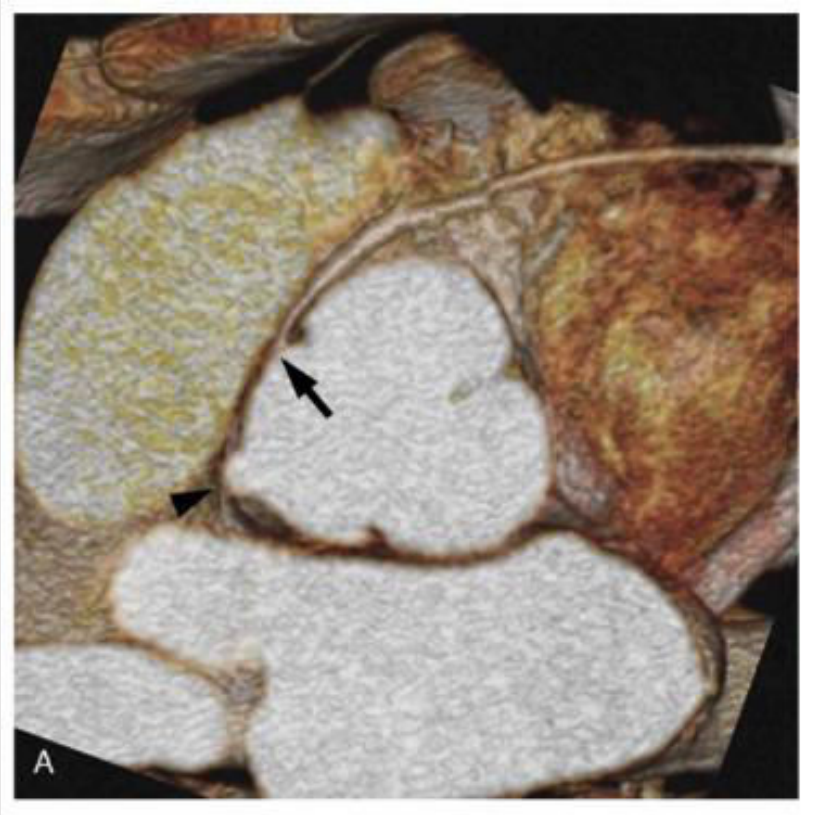
Clinical features of Anomalous connection of a main coronary artery to aorta

- Prevalence: 0.17%/ children with normal hearts
- Frequent first clinical manifestation: 2nd or 3rd decade of life
  - Angina
  - Syncope
  - Sudden death
- Rarely symptoms exist in neonates/infants or first decade of life

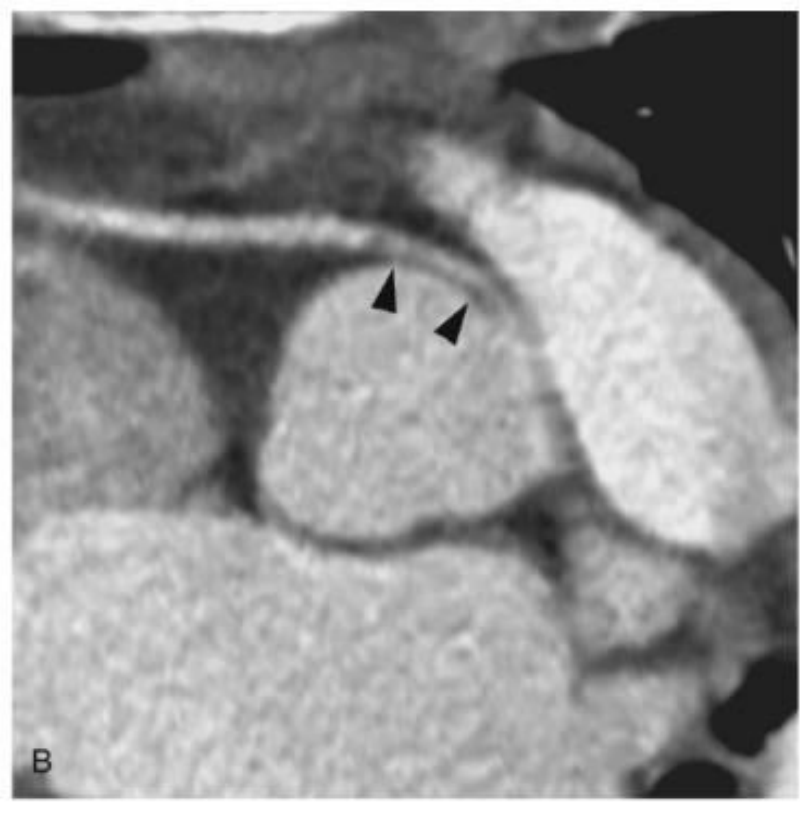
Imaging of anomalous connection of a main coronary artery to aorta

- Echocardiography, MSCT, MRI, Angiography
- Purposes of imaging:
  - Presence of a single ostium or separate ostia
  - Exact position of the ostium within or near the sinus
  - Presence of an intramural course
  - Identification of a slitlike or angulated ostium
  - Identification of an interarterial course and determination of whether the calibre of the artery is narrowed in thin area

MSCT/case of intramural course of RCA



A 13-year-old boy: RCA (arrow) and LMCA (arrowhead)



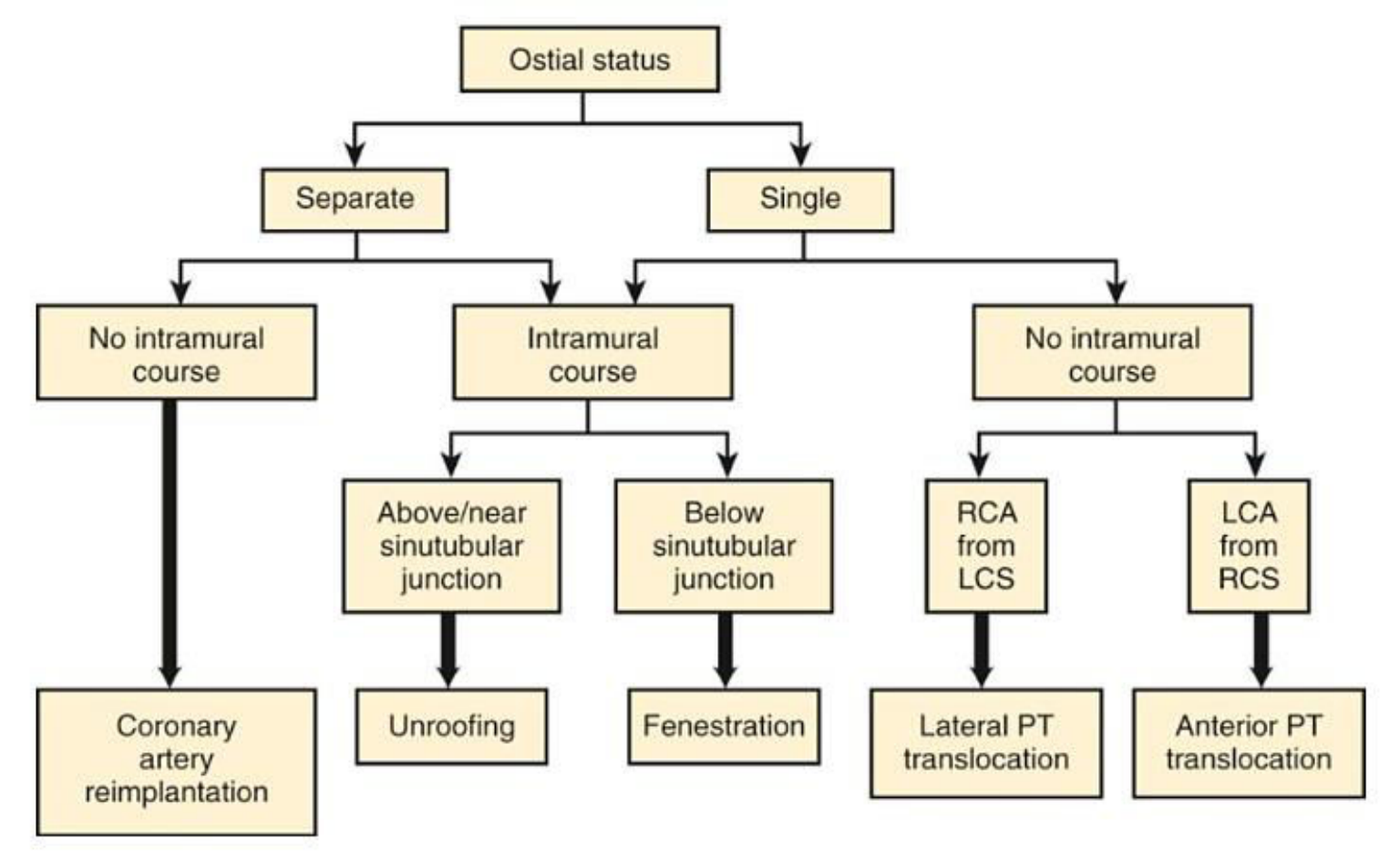
A 8-year-old boy: RCA from left coronary sinus

Source: Kirklin/Barrat- Boyes Cardiac Surgery 2013, 4th ed, Elsevier, P.1644-1666

Natural history of Anomalous Connection of a MCA to Aorta

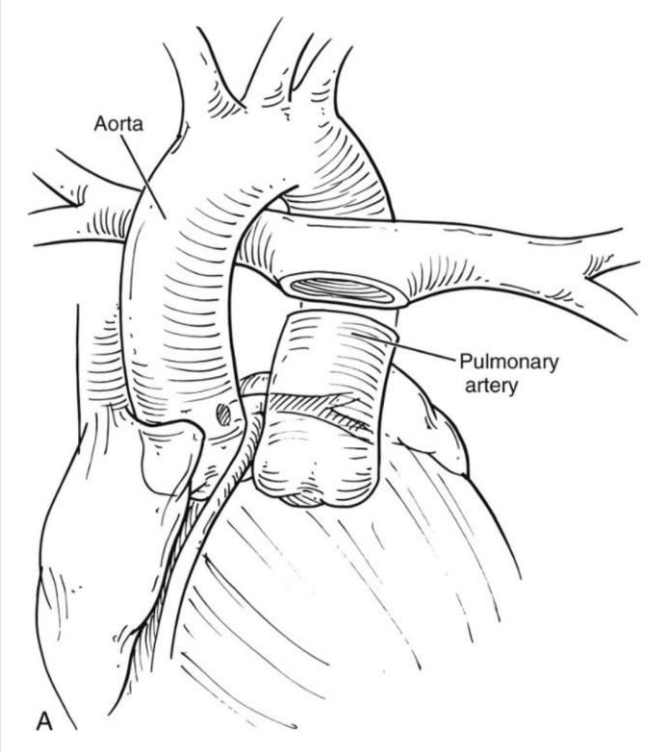
- Clinical events in neonatal period: rare
- Frequently ≥ 2nd decade of life
- Prevalence of sudden death: 2/1.000.000; competitive athletes due to coronary anomaly

Surgical management protocol

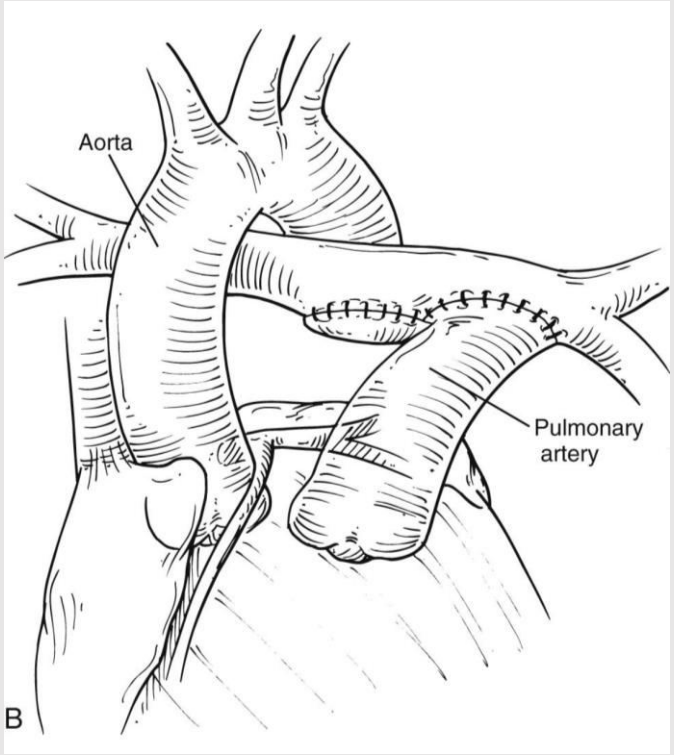


Source: Kirklin/Barrat- Boyes Cardiac Surgery 2013, 4th ed, Elsevier, P.1644-1666

Translocation of Pulmonary trunk for single coronary ostium without intramural element



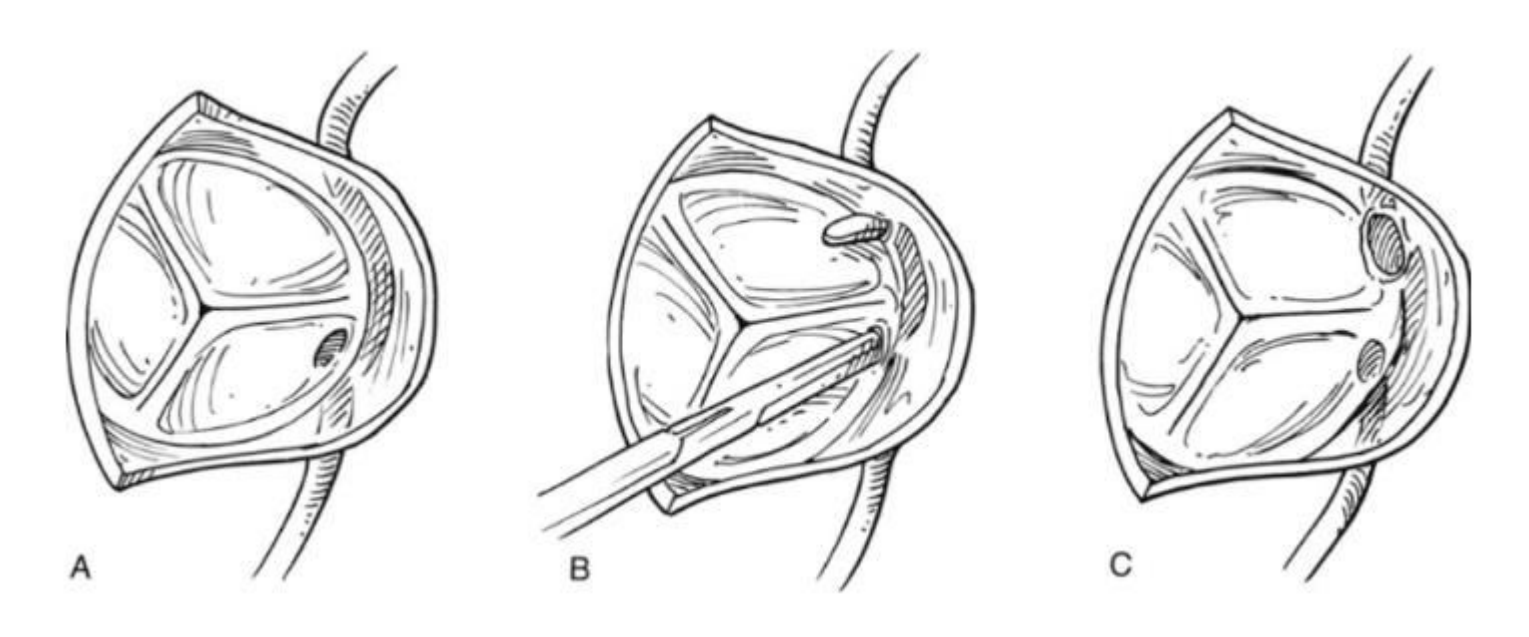
A: PT is dissected off its bifurcation. Patch augmentation of RPA; LPA is opened toward hilum. PT is translocated toward left hilum and reanastomosed.



B: Completed translocation of PT toward left hilum to create additional space b/w it and aorta

Source: Kirklin/Barrat- Boyes Cardiac Surgery 2013, 4th ed, Elsevier, P.1644-1666

Unroofing procedure/Anomalous origin of LCA from right sinus



A: RCA and anomalous LCA orifice arising from right sinus
B: intramural segment of the anomalous coronary is unroofed
C: create a neo-orifice in left sinus

Source: Kirklin/Barrat- Boyes Cardiac Surgery 2013, 4th ed, Elsevier, P.1644-1666

Conclusion

- Anomalous origin of coronary artery from PT (ALCAPA):
  - Clinical examination:
    - ✓ Infant heart failure; sudden death
    - ✓ Holosystolic murmur; continuous murmur
  - Diagnosis: imaging
    - ✓ Echocarddiography, MSCT, MRI, Angiogram
- Anomalous origin of a main coronary artery from aorta:
  - angina; syncope; sudden death (2^{nd} or 3^{rd} decade of life)
  - Diagnosis: imaging
    - ✓ Echocarddiography, MSCT, MRI, Angiogram