

Thida Yingchutrakul. M.D.

History

- An 80-year-old Thai female
- CC : progressive dyspnea for 1 month
- PI: 3 months ago: She reported of dyspnea on exertion and orthopnea but denied palpitation or chest pain. She was admitted to private hospital and diagnosed CHF. Her symptoms were slightly improved after treatment.

History

 PI: 1 month later, She became worsening dyspnea with generalized edema. She was admitted at a private hospital for a few days and referred to King Chulalongkorn Memorial Hospital.

Past History

• 10 years PTA: Hypertension and dyslipidemia

• 1 months PTA: Hyperthyroidism

mg/day

Ourrent Medication:

1. Warfarin2.5mg/day2. Digoxin0.125mg/day

- 3. Aspirin 81 mg/day
- 4. Bisoprolol 10 mg/day

5. Valsartan 160

6. Furosemide 20 mg/day
7. Trimetazidime 70 mg/day
8. Isosorbide mononitrate 20 mg/day
9. PTU 100 mg/day
10. Addi-K 750 mg/day

Physical examination

An elderly Thai female, good consciousness, looked chronically ill, no respiratory distress

- Vital signs:
- BP 140/70 mmHg, PR 80 bpm (regular), RR 18/min
- HEENT : mild pale conjunctivae, anicteric sclerae JVP 3 cm above sternal angle
- Heart : Apical impulse at left 5th ICS, MCL, no heave/thrill normal S1, S2

Continuous murmur gr. III/VI at LUPSB

Pan systolic murmur gr. II/VI at apex radiate to axiila

- Lungs : no crepitation, no wheezing
- Extremities : no pitting edema







Echocardiography



















































Stress CMR

Negative stress inducible ischemia
No myocardial hyper-enhancement

ALCAPA or Bland White Garlan syndrome Anomalous of Left Coronary Artery from Pulmonary Artery

Anomalous of Left Coronary Artery from Pulmonary Artery



Incidence

- 1/250 1/400 of all congenital malformed hearts
- Overall incidence : 1/300,000 children
- Frequency in boys (2.3 : 1)



Figure 45-3 In this specimen, the left coronary artery takes its or circumflex arteries in normal fashion.

Paediatric Cardiology, 3rd edition: Robert H. Anderson.

Pathophysiology

- Fetus
- : diastolic PAP = systemic pressure
- : coronary a. perfusion same as in normal
- After birth
- $: PAP \downarrow \rightarrow coronary steal$
- : LV ischemia
- : develop collateral connection from RCA



Paediatric Cardiology, 3rd edition: Robert H. Anderson.

http://nursingcrib.com/nursing-notes-reviewer/fetal-circulation/

Pathophysiology

- Typically
 - : RCA enlarge at its origin
 - : LCA turn to be small, relative thin-walled
 - : collateral may be diffusely large, well distributed

Presentation

- Majority present during infancy
- Myocardial ischemia -> CHF
- Anginal attack -> severe distress, grunting, or crying
- Unexplained heart murmur
- Myocardial ischemia in children/ young adult
- May be sudden unexplained death
- Progressive MR (fibrosis involve pappillary m, LV dilatation)

Clinical course and prognosis

- 10%–15% of patients survive to adulthood (extensive intercoronary collaterals)
- extent of collateral vascularisation -> clinical course
- may present later in childhood, adolescence, or adulthood with progressive MR, CHF, or myocardial ischaemia with exertion
- Untreated -> prognosis is poor



Figure 2. A 3-year-old boy with older type ALCAPA. (A) Modified parasternal short-axis view identified left main coronary artery from the main pulmonary artery and the red shunt from it to the pulmonary artery. (B) Parasternal short-axis view showed abundant septal collateral signals from right to left coronary artery. (C) Real time three-dimensional echocardiographic imaging showed retrograde flow from left coronary branches and then via main trunk into the pulmonary artery. (D) Real time three-dimensional echocardiographic imaging showed the dilated right coronary artery arising from the aorta. (E) Aortogram from ascending aorta in frontal projection showed normal origin of a single large right coronary artery. The posterior descending artery gives rise to numerous intercoronary collaterals. These course through the ventricular septum, opacifying in retrograde fashion, the left coronary system, and the main pulmonary artery. AO = aorta; LAD = left anterior descending artery; LCX



Figure 4. Multidetector-row computed tomography (CT). (A) Oblique coronary image of coronary CT angiography shows a dilated right coronary artery (RCA, arrow) which is 1.0 cm in diameter, arising from the right aorta (Ao) sinus. (B) Axial image of coronary CT angiography clearly shows a dilated left main coronary artery (LMCA, arrow) originating from the pulmonary artery (PA). (C) The 3-dimensional volume-rendered image reveals multiple well-developed collateral arteries (black arrowheads) between the RCA (black arrow) and the left anterior descending coronary artery (LAD, white arrowhead); the LMCA merges with the pulmonary trunk. LCx = left circumflex artery.

J Chin Med Assoc, September 2010: Vol 73, No 9

MRI

- Described coronary a. anatomy and origin of LCA from PA
- Assess venticular function and volume
- Identify MR (secondary to ischemia)
- RWMA/ myocardial ischemia
- Retrograde/ bidirectional flow in proximal LCA
- Post-repaired -> supravalve PS at site of PA patch



Figure 2 - Magnetic resonance angiography of the coronary arteries with 3D reconstructions showing the ostia of the coronary arteries. Note that the left coronary artery has its origin in the pulmonary trunk (TAP – trunk of pulmonary artery, Ao - ascending aorta, LA - left atrium, Cx - circumflex coronary artery, DA - left anterior descending coronary artery, CD - right coronary artery).

Managements

- Surgery
- reimplantation of the left coronary artery into the aorta



Thank you for your attention