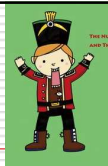


Aortic arch as a nutcracker

---A rare type vascular ring---



CMUH Pediatric Cardiologist Yi-Chin Peng /Chief Jeng-Sheng Chang



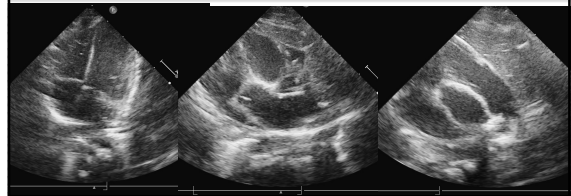
- A 1-month 8-days old female infant
- G2P2, GA=38+5 weeks, BBW=3130g, delivered via NSD
- Now 55 cm, 5.3 kg
- Well feeding 120 ml/Q3h

- First visit OPD due to abnormal prenatal echo examination r/o vascular ring (in China)
- PE: RHB G1-2/6 systolic murmur LUSB
- Smooth breathing, smooth feeding

→ Arrange Echocardiography

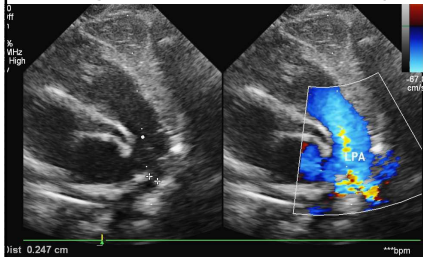
Echocardiography –

- Abnormal LV-LVOT-AAO arrangement (AS TGA pattern)
- Normal A-V, V-A relation



Echocardiography –

- LPA orifice discrete narrowing, PG: 33 mmHg
- Right arch, normal arch artery

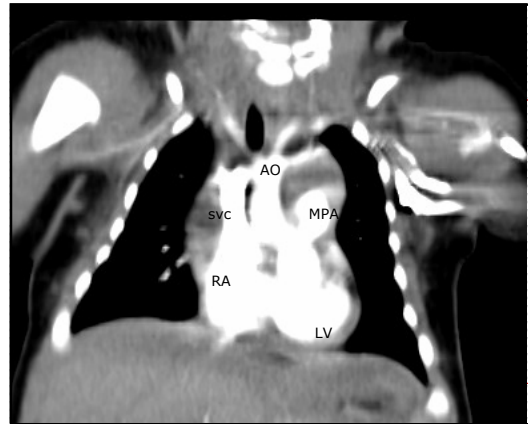


Admission and arrange CT



CT

- Levocardia
- **Left aortic arch** with left DAO , Normal arch artery
- Normal tracheobronchial tree ,**left main bronchus** compression by right main pulmonary artery and DOA with the minimal diameter: 1.6 mm
- LPA: 6.0 mm RPA: 6.2 mm
- **Elevation of RPA with tortuous course**
- Diameter of patent ductus arteriosus: 1.2 mm



Cardiac cath.

Cardiac cath.

- Counter-clockwise rotation of heart and great vessels
- PPS at LPA entry (25mmHg pressure gradient), partly due to a tight ligamentum ductus arteriosus
- Left main bronchus compression by vascular ring between the posteriorly displaced asc. AO, RPA and the desc. AO.

- LV cineangiogram (PA/Lat. views):
- AAO, arch and DAO cineangiogram (PA/Lat. views):
 - **The AAO was counterclockwise rotated and displaced leftward (totally left to the trachea) and posteriorly.**
 - The PDA was closed with a remnant of ampulla at descending AO.
 - Left main bronchus pulsating concomitantly with the surrounding RPA and aorta.

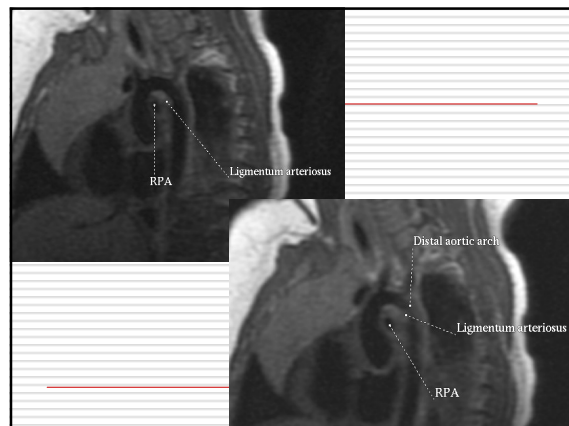
- RPA cineangiogram (PA/Lat. views):
 - counterclockwise rotation of the heart and great vessels make a stretched RVOT through the MPA and an extended RPA.
 - The orifice of the RPA located at about left 1/3 of the left clavicle.
- LPA cineangiogram (PA/Lat. views):
 - discrete narrowing, a tightened ligamentum ductus a.

Cardiac MRI

To delineate the ligamentum ductus art.

Cardiac MRI

- A tubular structure between the distal aortic arch (just distal to left subclavian artery) and right pulmonary artery (Se:8 Im:1-5, Se10:1-10, Reformation imaging), combination with CT and angiography, R/O ductus ligamentum from distal aortic arch tethering right pulmonary artery causing compression on left main bronchus.
- Patent foramen ovale (Se:13, Im:134)
- Left pulmonary artery stenosis due to the counterclockwise rotation of heart and great vessels and tethering of ductus ligamentum between distal aortic arch and right pulmonary artery.



- Document vascular ring between rotated MPA-LPA, AAO DAO & ductus ligamentum.
- Suggestion operation
 - relief the compression of left main bronchus and LAP

Operative Findings

- **"aortic arch as a nutcracker compression left bronchus"**
- **"aortic arch nutcracker syndrome"**
 - post displacement of aortic valve,
 - narrowed aortic angle, resulting in compression of L't bronchus by RPA below aortic arch.
 - LPA stenosis at PDA insertion.

- Division of PDA,
- Patch angioplasty of LPA,
- Translocation of RPA to ant. of ascending aorta

- Post OP smoothly except chylothorax
- Discharge at post OP day 13
- F/u Echo: still bilateral PPS mild

REVIEW

Bronchial Compression by Posteriorly Displaced Ascending Aorta in Patients With Congenital Heart Disease

Yang Min Kim, MD, Shi-Joon Yoo, MD, Woong Han Kim, MD, Tae Hoon Kim, MD, Joon Hee Joh, MD, and Soo Jin Kim, MD

Department of Radiology, Pediatrics, and Cardiac Surgery, Sejong General Hospital & Sejong Heart Institute, Pucheon, South Korea, and Department of Diagnostic Imaging, Hospital for Sick Children, Toronto, Ontario, Canada
(Ann Thorac Surg 2002;73:881 - 6)

Schematic drawing of the axial CT image at the level of the main stem bronchi for measurement of control patients (left panel) and schematic drawing for comparison with patients who have posterior malpositioning of ascending aorta (right panel)

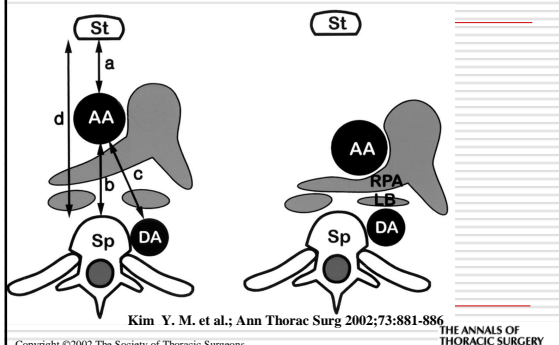
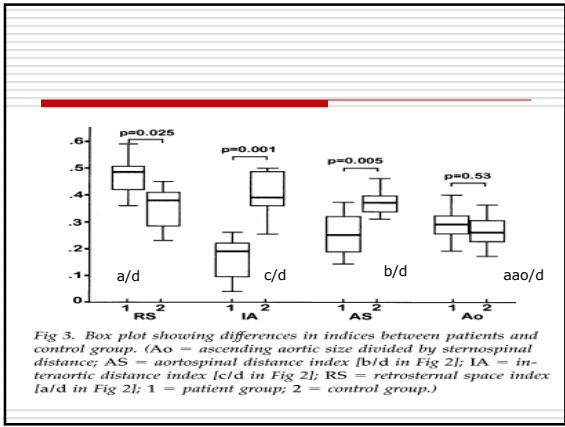
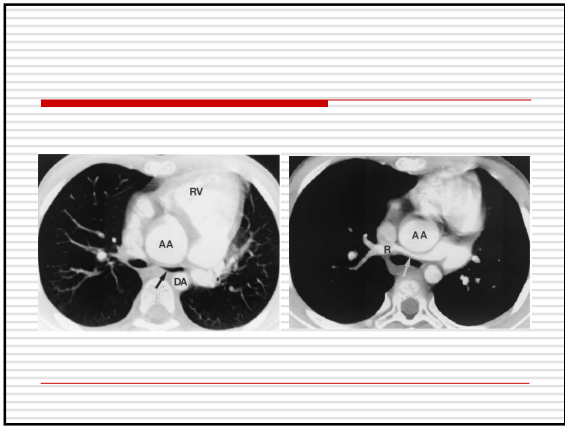


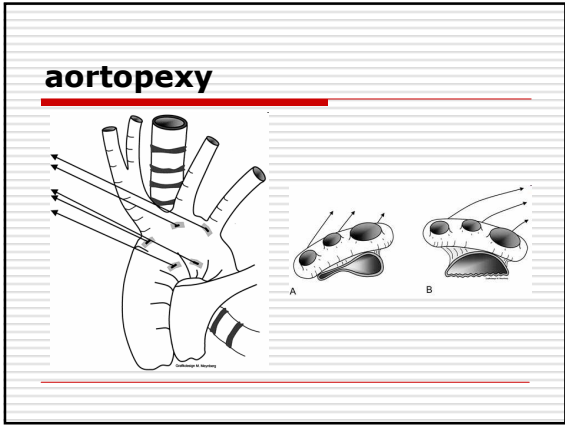
Table 1. Summary of Patient Data

Patient No.	Age at Scan	Sign	Airway Stenosis	Aortic Arch	RPA Compression	Diagnosis	Associated Finding	Aortopexy	RMB to Tr	LMB to Tr
1	15 mo	Stridor, grunting	LMB	Left	Yes	PDA	RVE		0.43	0.20
2	3 mo	Pneumonia	RMB	Right	No	PA VSD	...	Yes	0.25	0.54
3	7 mo	No	LMB	Left	No	COA PDA	...		0.59	0.11
4	7 y	No	LMB	Left	Yes	VSD	RVE, aorta dilatation		0.40	0.30
5	4 days	Pneumonia	RMB	Right	No	TOF	RVE, RVH		0.37	0.44
6	8 days	No	no	Left	...	Truncus arteriosus	RVE		0.45	0.75
7	19 y	No	LMB	Left	Yes	PDA	Midline descending aorta	Yes	0.50	0.20
8	6 mo	Pneumonia	RMB	Left	Yes	TOF	RVH	Yes	0.12	0.12

COA = coarctation of aorta; LMB = left main bronchus; LMB to Tr = area ratio between left main bronchus and trachea; MAPCA = major aortopulmonary collateral arteries; PA = pulmonary atresia; PDA = patent ductus arteriosus; RIB = right intermediate bronchus; RMB = right main bronchus; RMB to Tr = area ratio between right main bronchus and trachea; RVE = right ventricular enlargement; RVH = right ventricular hypertrophy; TOF = tetralogy of Fallot; VSD = ventricular septal defect.



- "pincer" effect (夾擊) of a malposed AAO and the DAO.
- compression of the **right pulmonary artery** due to a posteriorly displaced ascending aorta, **aortopexy** to re-lieve the compression of the main bronchus or the right pulmonary artery
- Bronchoscopy is thus required for evaluation of the dynamic component of



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**Left mainstem bronchial narrowing:
a vascular compression syndrome?**

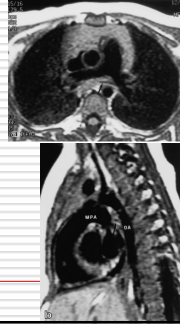
Evaluation by magnetic resonance imaging

Table 1 Descending aorta relative to vertebral body (Chi-square trend test comparing symptomatic and control patients and positions of the descending aorta: $P < 0.05$)

Descending aorta position	Symptomatic $n = 10$	Control $n = 40$
Paraspinal	0 (0%)	6 (15%)
25-50% anterior	2 (20%)	12 (30%)
50-75% anterior	4 (40%)	18 (45%)
Prespinal	4 (40%)	4 (10%)

Fig. 1 Composite drawing of DA positions. (1) Paraspinal 25-50% of circumference anterior to vertebral body, (2) 50-75% anterior to vertebral body, (4) prespinal

- ❑ LMSB narrowing is well-defined by MRI
- ❑ More common and more marked in children with LMSB narrowing at prespinal .



Compression of the Central Airways by a Dilated Aorta in Infants and Children With Congenital Heart Disease

Doff B. McElhinney, MD, V. Mohan Reddy, MD, Mark S. Plan, MD, Phillip Moore, MD, and Frank L. Hanley, MD

Divisions of Cardiothoracic Surgery, Pediatric Pulmonology, and Pediatric Cardiology, University of California, San Francisco, San Francisco, California

Table 1. Details of 5 patients who underwent surgery for airway compression by an enlarged aorta

Patient No.	Age	Primary Diagnosis	Preoperative		Affected Structures	Preoperative Imaging Studies	Treatment	Total Duration of Intubation	Follow-up
			Arch Laterality	Airway Symptoms					
1	7 mo	PA, VSD, MAPCAs	Left	No	Left MSB, esophagus	Bronchoscopy >3, CT, barium swallow	Aortopexy (49 d) ^a	51 d	28 mo, alive, no symptoms
2	3 y	PA, VSD, MAPCAs	Right	No	Distal trachea, carina, bilateral MSB	Bronchoscopy >2, MRI, CT	Aortopexy (17 d) ^a	Tracheostomy POD 60	24 mo, alive, moderate symptoms, tracheostomy removed 13 mo
3	3 mo	PA, VSD, MAPCAs	Right	Yes	Distal right trachea, carina, right MSB	Bronchoscopy	Aortopexy, reduction ascending aortoplasty	Tracheostomy POD 38	21 mo, alive, mild symptoms, tracheostomy removed 9 mo
4	12 d	Doubly-committed VSD, ASD, PDA	Right	Yes	Distal trachea, carina, bilateral MSB	Bronchoscopy, MRI, contrast tracheogram with arteriography	Aortopexy, reduction ascending aortoplasty	6 d	20 mo, alive, no symptoms
5	6 mo	Cervical arch, coarctation previously repaired	Right	Yes	Distal trachea, right MSB	Bronchoscopy, MRI, contrast tracheogram with arteriography	Aortopexy, reduction ascending aortoplasty	4 d	5 yr, alive, no symptoms

^a Duration after the initial operation.

ASD = atrial septal defect; CT = computed tomography; MAPCAs = major aortopulmonary collateral arteries; MSB = mainstem bronchus; PA = pulmonary atresia; PDA = patent ductus arteriosus; POD = postoperative day; VSD = ventricular septal defect.

❑ Thanks