תינוקת עם Pulmonary Artery Sling ועוד...

16/3/18

ענבל גולן טריפטו

הצגת המקרה

- בת 4 שבועות, נולדה בשבוע 40+3, מ.ל- 2830 גרם
- מעקב הריון חסר
- לאחר לידתה imperforated anus & recto-vestibular fistula, נותנת יציאות דרכה. הרחבות אנאליות
- אקו לב- ASD ו- PDA גדול
- סונר כליות- hydronephrosis של כליה ימנית
- סונר עמוד שדרה- תקין, ללא tethrhed cord

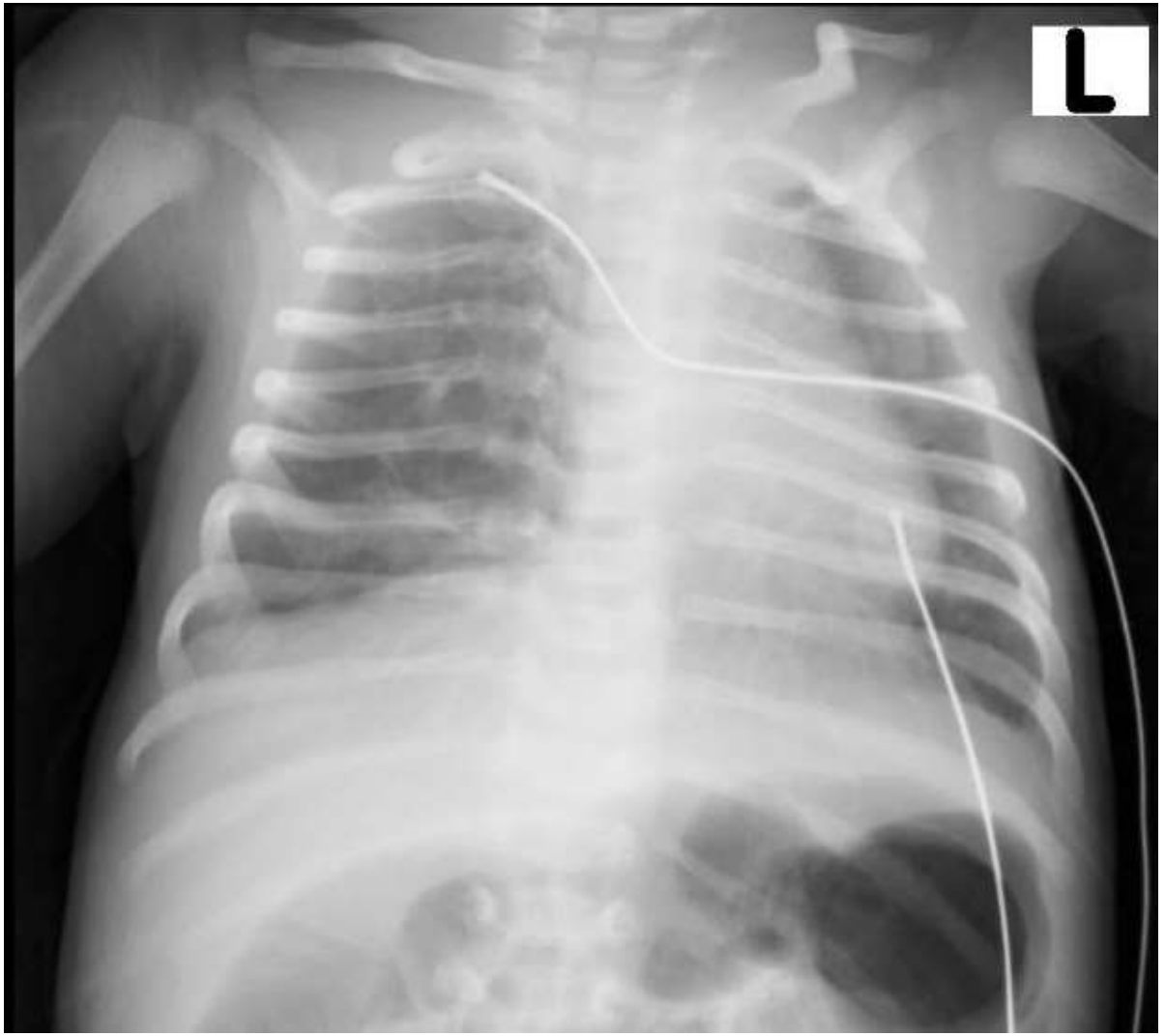
VACTER- L association

Vertebral anomalies
Anal atresia
Cardiovascular anomalies
Tracheoesophageal fistula
Esophageal atresia
Renal and/or radial anomalies
Limb defect

תלונה עיקרית

- מזה 3 ימים נזלת ושיעול, ללא עליית חום
- בקבלתה למיון:
  60-RR, דיספנאית, סטורציה-86% בא"ח, 93%
  במסיכת רזרוואר, חום-36.7
  בהאזנה- פקעים וחרחורים דו"צ
- גזים(וריידים)- 7.22-PH, PCO2-69, PO2-104, HCO3- 27
- הועברה לט"נ ילדים- שם הונשמה לא פולשנית NIPPV וכעבור יממה הועברה למחלקתנו
- משטף אף- חיובי לאנטרווירוס

צילום חזה בקבלתה

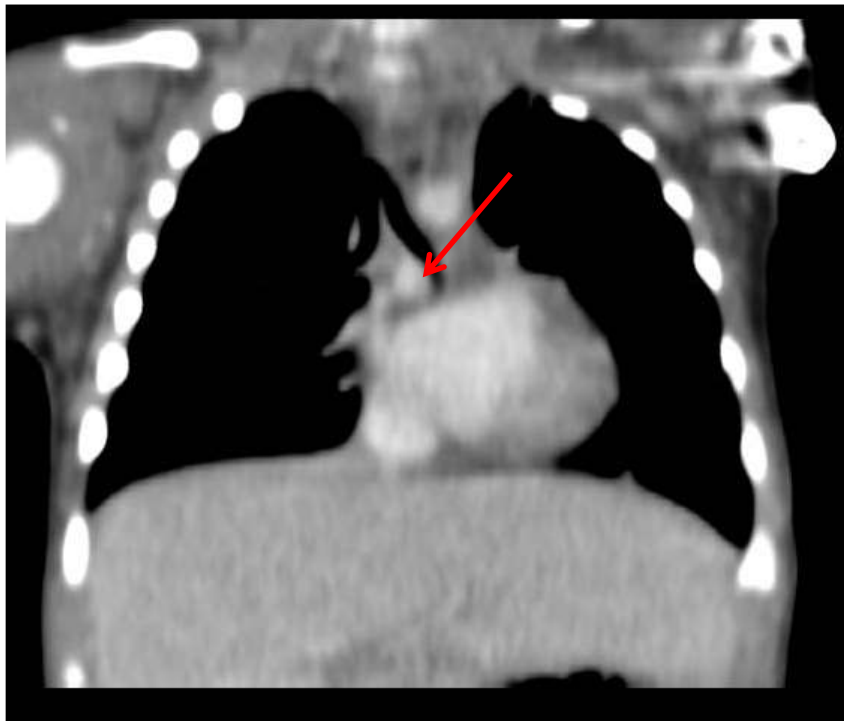


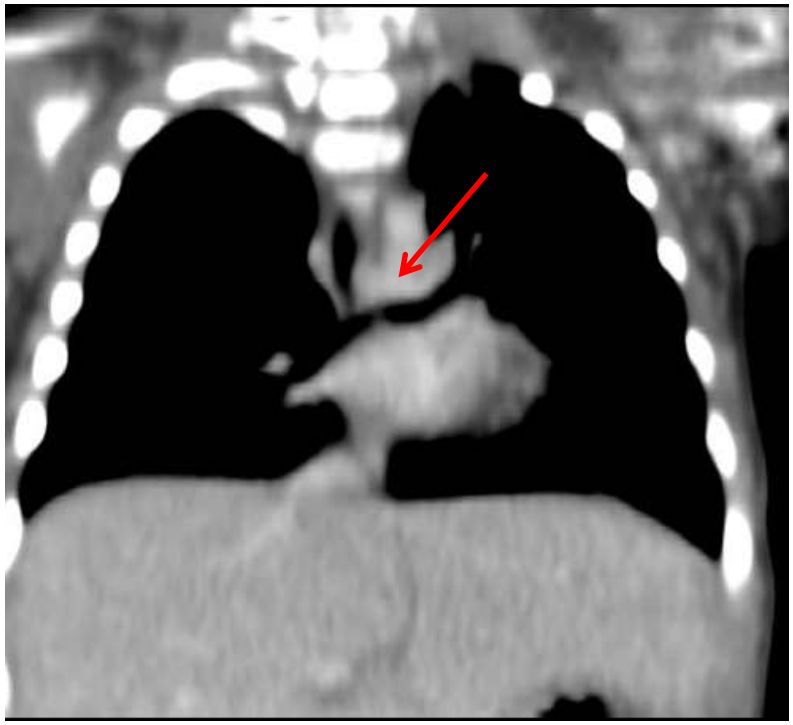
4 weeks – 18/11/2017

מהלך במחלקה

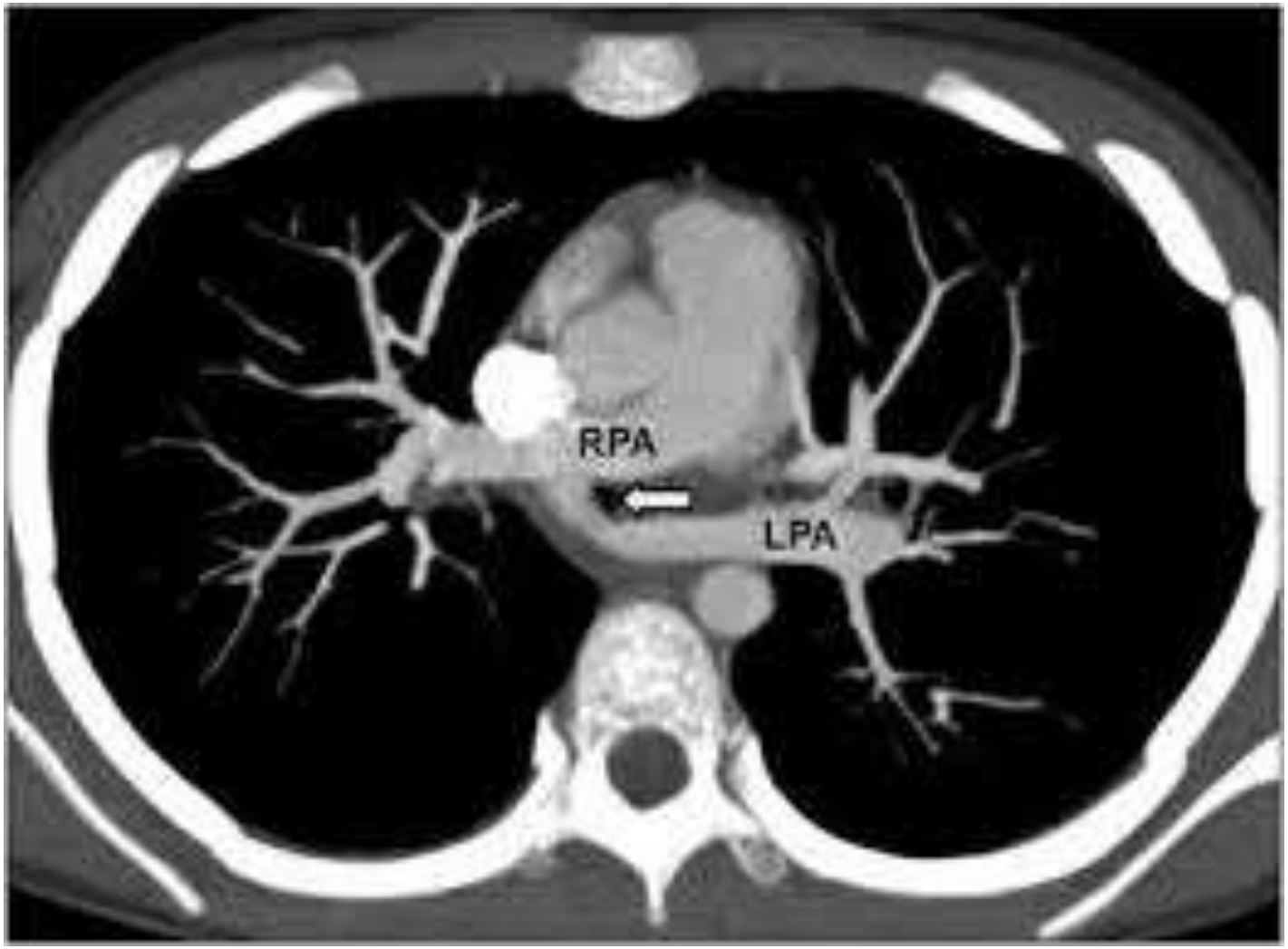
- טיפול במרחיבי סימפונות ובסטרואידים סיסטמיים
- היפוקסמיה ממושכת (10 ימים)
- בוצע אקו לב- הועלה חשד ל Pulmonary sling
- בוצע CTA

CTA

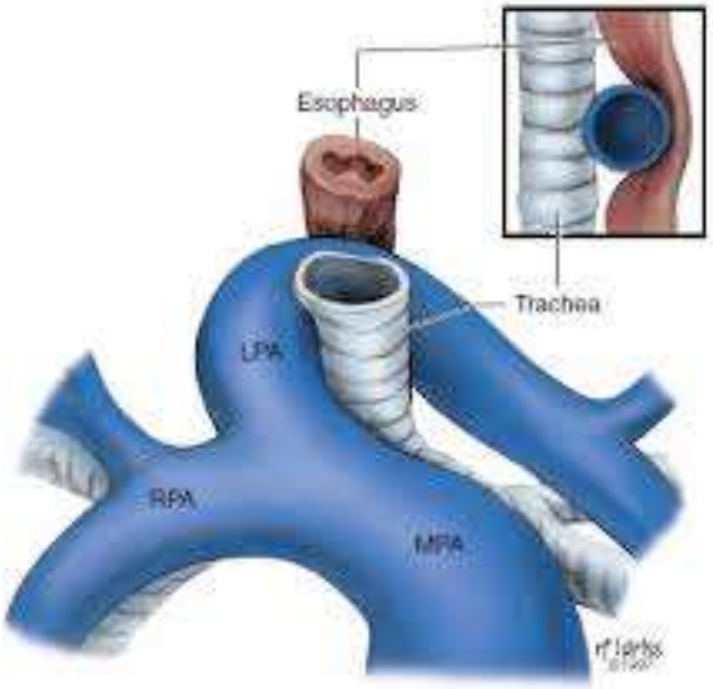


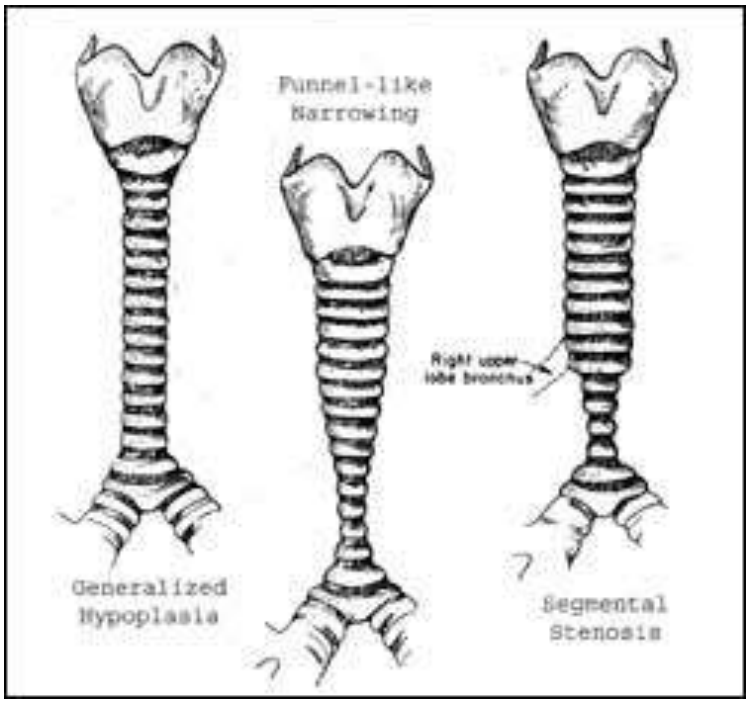


CTA



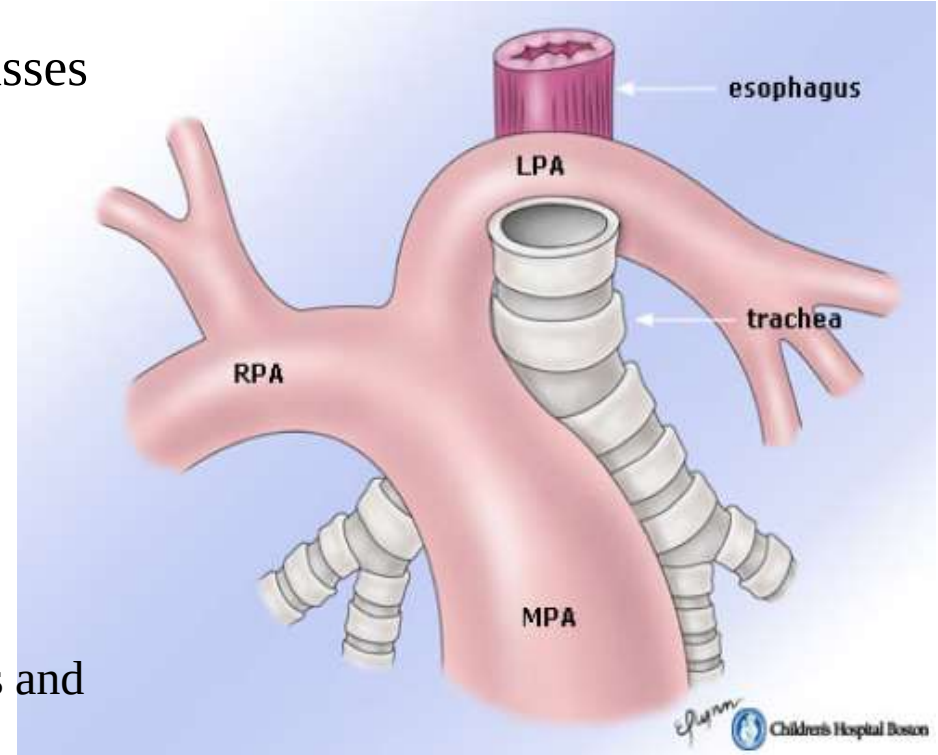
Pulmonary Ring- Sling Syndrome





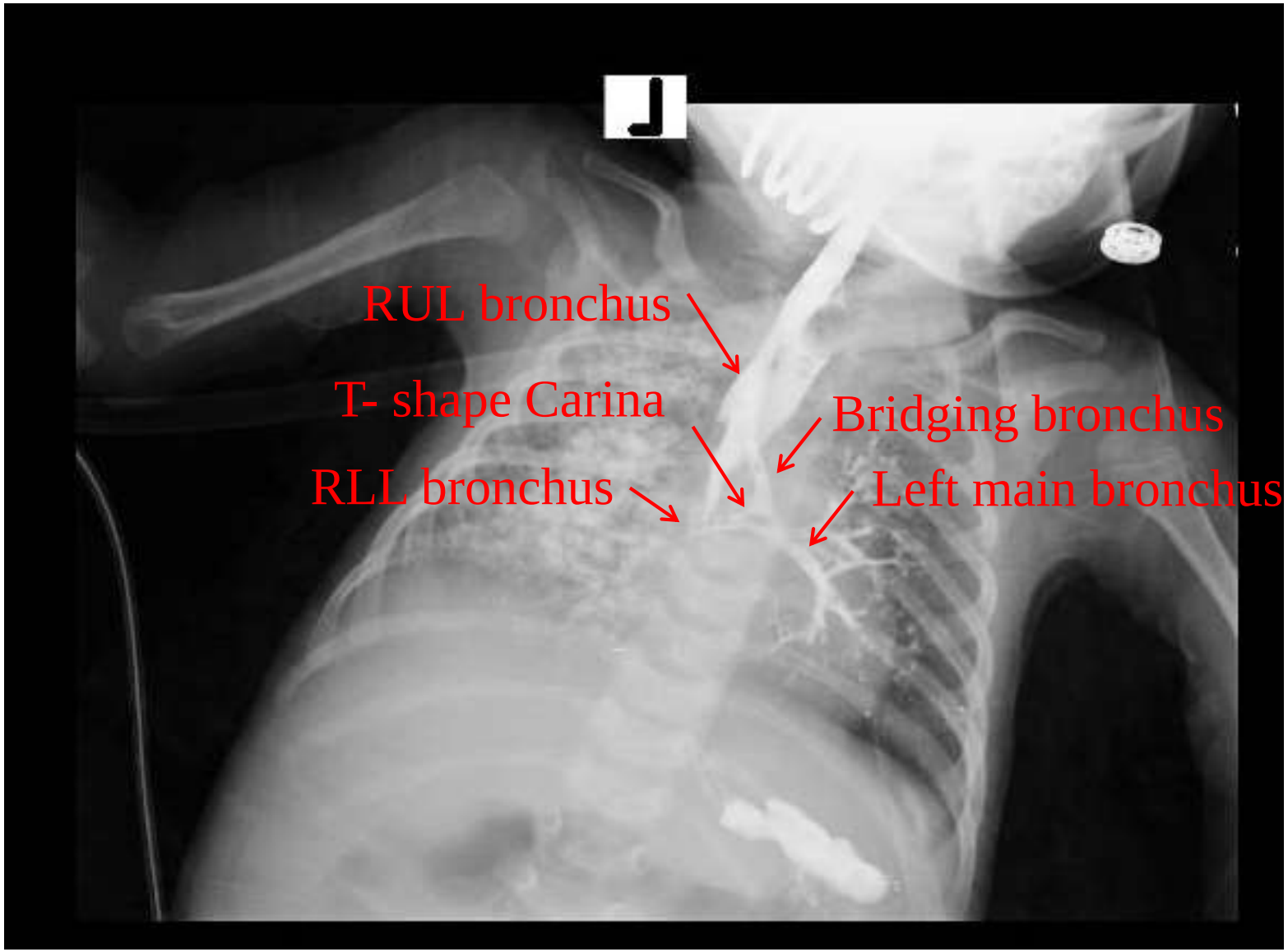
Anomalous Left Pulmonary Artery - Sling

- Rare
- LPA arises from the RPA, passes behind trachea and anterior to esophagus
- Frequently associated with tracheal ring: Ring – Sling Syndrome
- Associated with other heart defects, VSD , ASD
- Symptoms: feeding difficulties and respiratory symptoms
- Biphasic or expiratory stridor, "noisy breathing

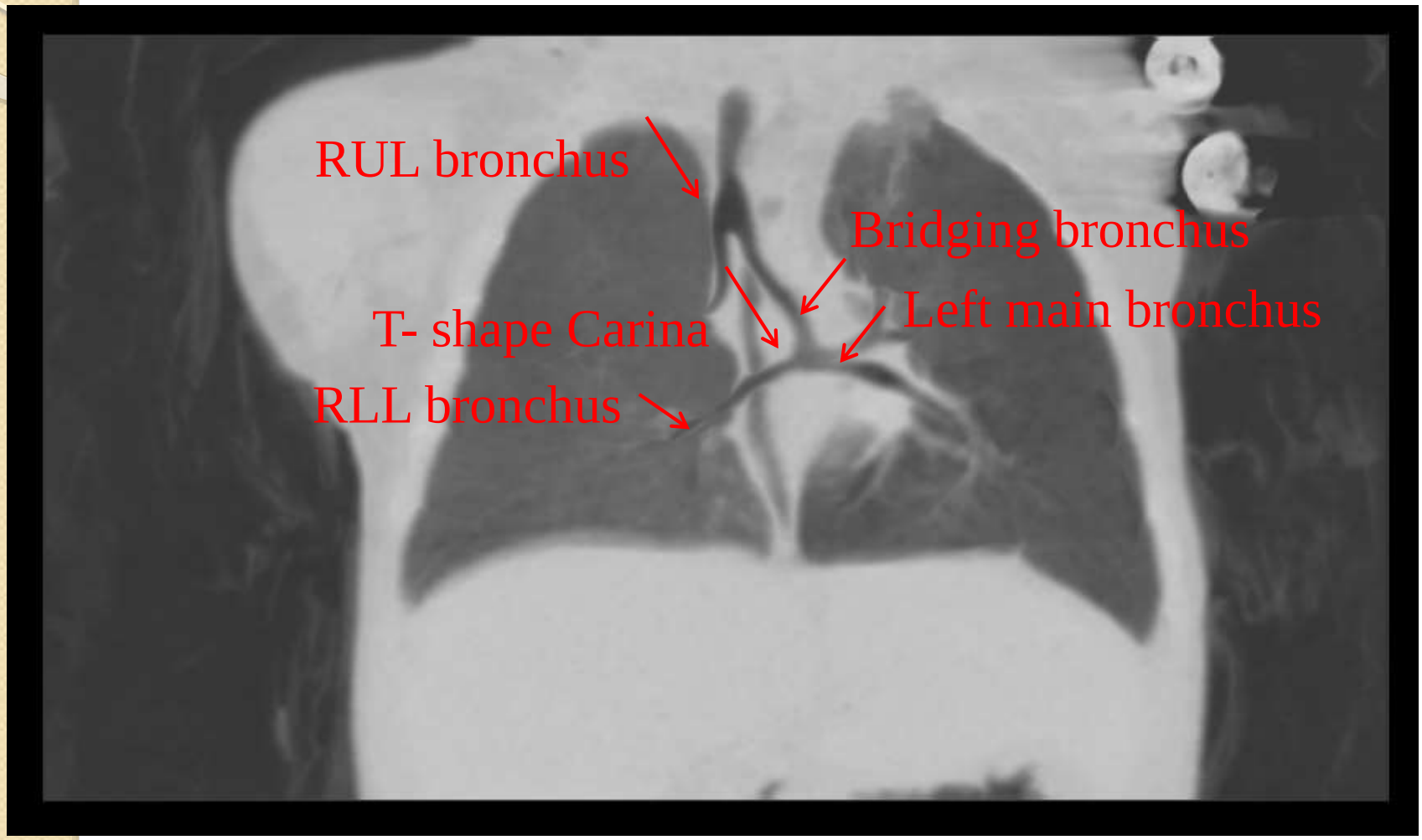


Bronchoscopy

Bronchography



CT


RUL bronchus
Bridging bronchus
Left main bronchus
T- shape Carina
RLL bronchus

Type 2 left pulmonary artery sling – Association with decreased right lung volume

Young ah Cho, M.D., Beverley Newman, M.D.
Department of Pediatric Radiology, Lucile Packard Children's Hospital, Stanford University School of Medicine

Introduction

Left pulmonary artery sling (LPAS) is a rare congenital anomaly in which the LPA originates from the posterior aspect of the right pulmonary artery and courses between the trachea and esophagus to reach the left lung. Approximately 50% of LPAS cases are associated with intrinsic tracheal stenosis.

There are two major types of left pulmonary artery sling. **Type 1 LPAS** usually compresses the distal trachea and right main stem bronchus resulting in hyperinflation of the right lung. A tracheal bronchus may be present. **Type 2 LPAS** is characterized by a more inferiorly located left pulmonary artery sling and abnormal bronchial branching with a low T-shaped carina. Long segment tracheal stenosis with complete cartilaginous rings is a common accompaniment often with bilateral hyperinflation. Other lung abnormalities may also occur, including right-side pulmonary hypoplasia and agenesis as well as scimitar syndrome and other foregut lesions.

We present 5 cases of Type 2 LPAS with diminished right lung volume in order to emphasize the association of this appearance with right lung/vascular abnormalities.

Anatomic type of LPAS

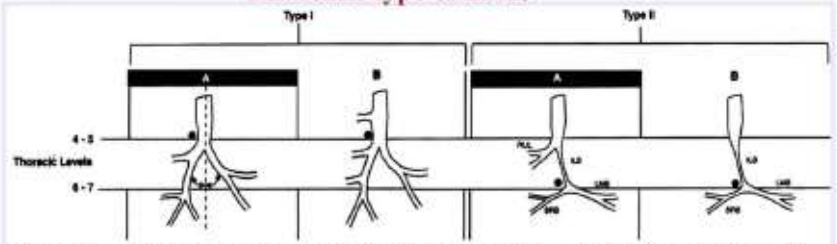


Figure 1. Anatomic types of PA sling (Wells, *et al.*) – solid circle denotes left pulmonary artery origin. RUL Right upper lobe bronchus, ILB intermediate left bronchus, BRB bridging right bronchus, LMB left main bronchus

Materials and Methods

5 Children (3 males and 2 females) aged 1 day to 6months presented with respiratory distress and decreased right lung volume with ipsilateral

Case 3. Type IIB LPAS in a 4-month-old girl

Fig.4 A. Poor tracheal visualization ; right lung decreased volume? patchy atelectasis/pneumonia.

Fig.4 B&C. Oblique coronal MR images demonstrate left pulmonary artery (arrow) arising from right pulmonary artery (arrowhead) just behind the airway. Note low T-shaped carina (arrow) with moderate long segment lower tracheal narrowing and bridging right bronchus (arrowhead).

Case 4. Type IIB with right lung agenesis in a 6-month-old boy

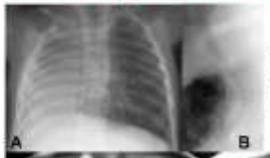


Fig.5 A-B. Poorly visualized trachea; probable absent/ hypoplastic right lung with ipsilateral mediastinal shift and herniation of left lung to right. Lateral airway high KV fluoroscopic image shows narrowed lower trachea bowed posteriorly.

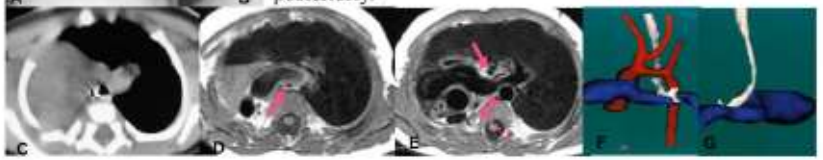


Fig.5 C-G. Noncontrast CT(C) and Sup to Inf axial MR images (D-G) confirm absent right lung. The lower trachea (arrow) is displaced posteriorly by the crossing aortic arch and is also stenotic. The LPA (arrowhead) passes between the airway (arrow) and the esophagus (dashed arrow). This represents left pulmonary artery sling anatomy but with absence of the distal right pulmonary artery. F and G - 3D frontal and lateral reconstructions of the anatomy. Note common origin of carotid artery.

Case 5. Type IIB with Scimitar syndrome in a newborn boy

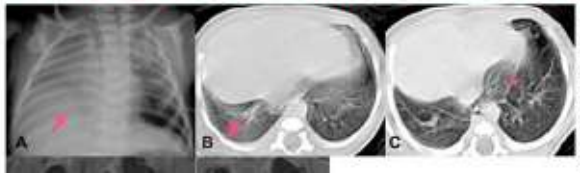
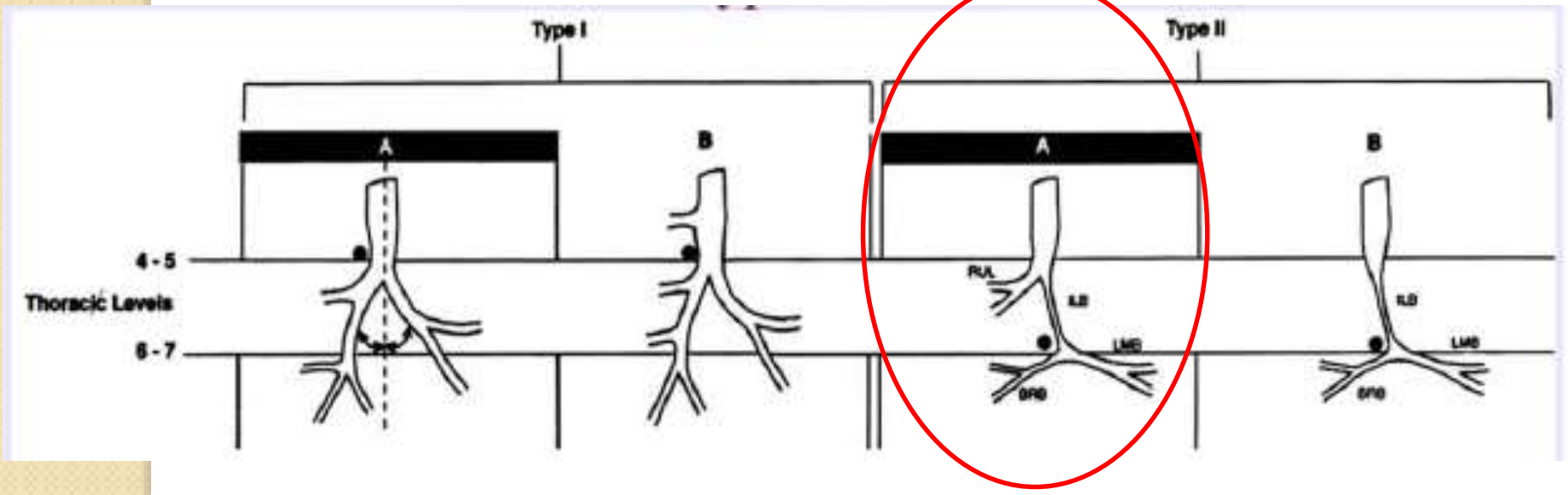


Fig.6 A. Plain chest AP- hypoplastic right lung with probable right scimitar vein (arrow). B-C. Scimitar vein (arrow) and horseshoe lung (arrowhead) shown on chest

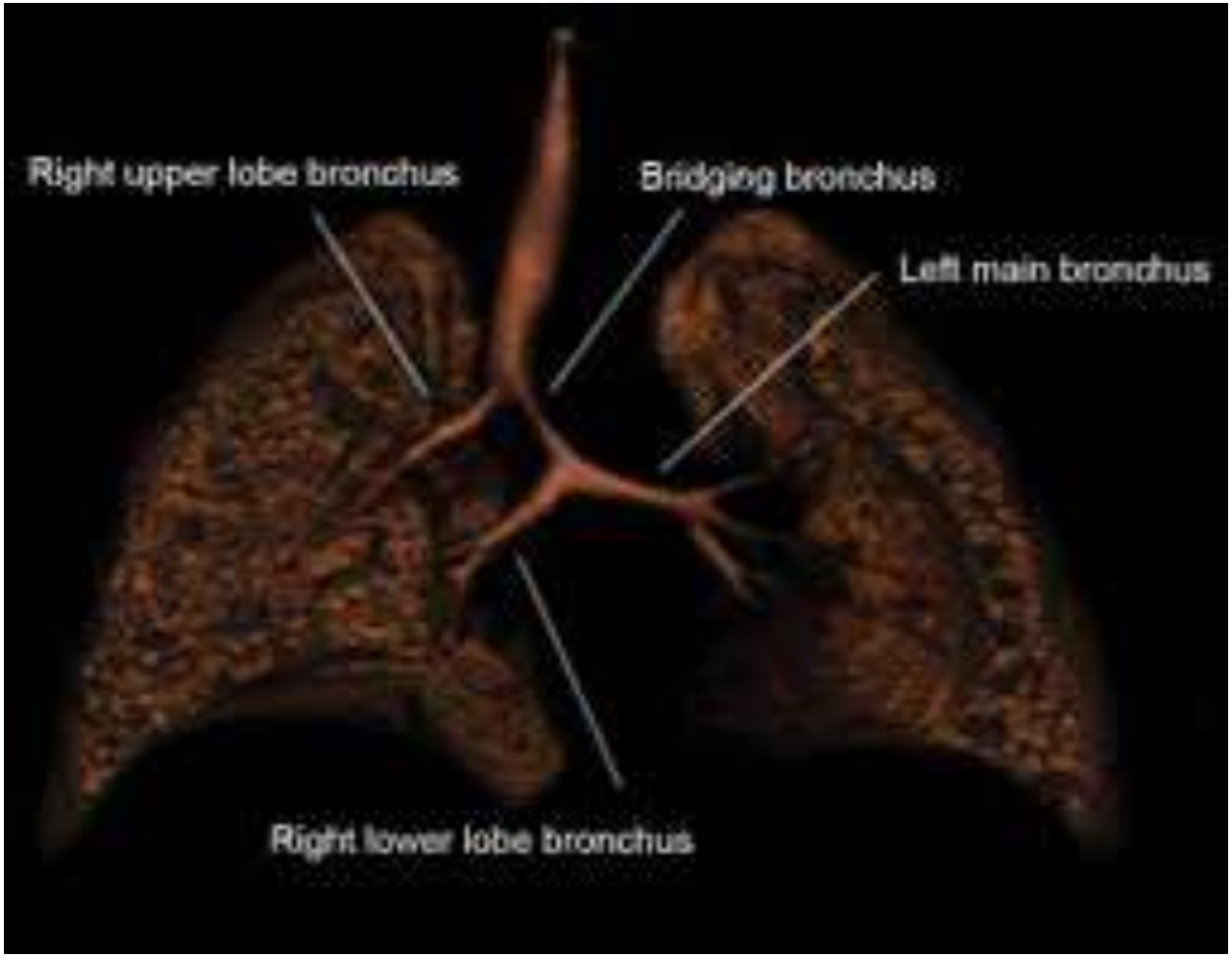
Type 1 vs Type 2



Type1: distal trachea & Rt main bronchus. Rt lung hyperinflation

Type 2: inferiorly located & abnormal bronchial branching T-shape carina. Long- segment tracheal stenosis with bilateral hyperinflation.
Other abnormalities.

Type 2 pulmonary artery sling



Pulmonary sling & VACTER

Journal List › J Tehran Heart Cent › v.12(3); 2017 Jul › PMC5643871

J Tehran Heart Cent. 2017 Jul; 12(3): 131–133. PMCID: PMC5643871

A Rare Case of Pulmonary Artery Sling with the VACTERL Association in a 20-Month-Old Infant

Yazdan Ghandi, MD,[1,*] Akbar Shafiee, MD, MSc,[2] Mehrazad Sharifi, MD,[3] and Najmeh Sadat Bolandnazar, MD[4]

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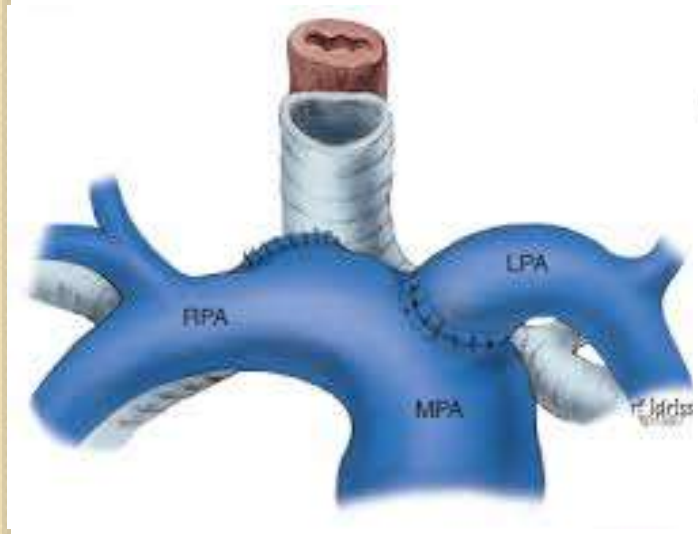
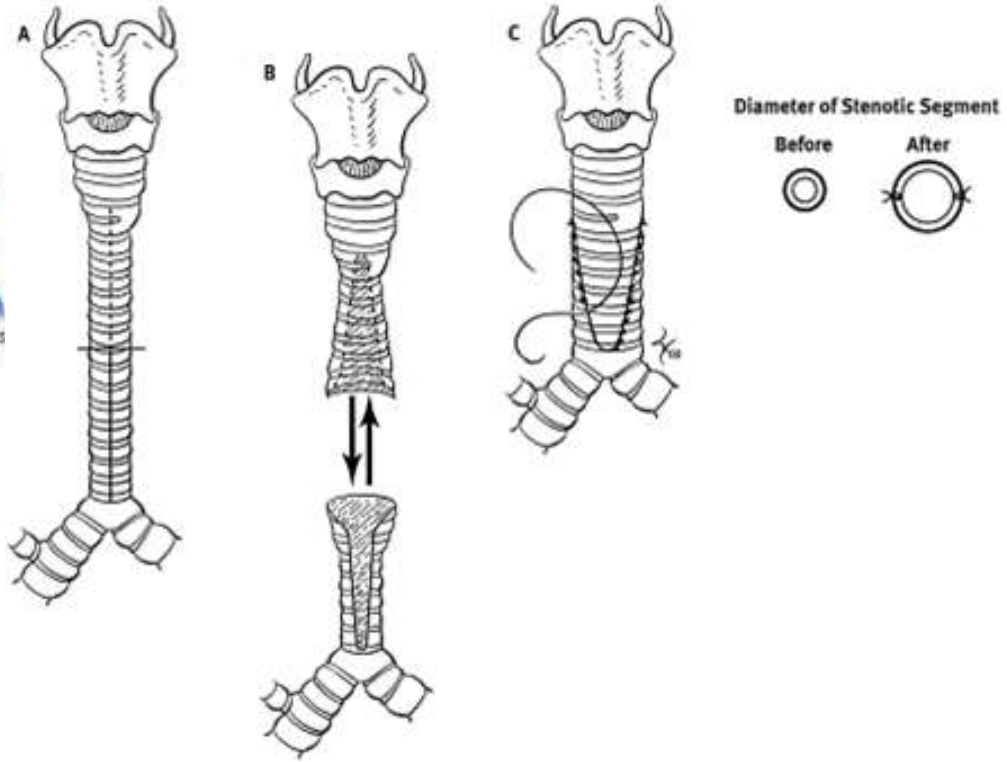
Abstract

Go to:

The VACTERL association, co-occurrence of vertebral, anorectal, cardiac, tracheoesophageal, genitourinary, and limb malformations, is a rare congenital anomaly. Several cardiac anomalies have been reported as a part of the VACTERL association, particularly ventricular and atrial septal defects. Pulmonary artery sling is a rare congenital abnormality in which the left pulmonary artery arises from the right pulmonary artery. This anomaly is not frequently observed in the VACTERL association and has been rarely reported. A 20-month-old girl was admitted to our hospital due to pneumonia in the right lung, which had pulmonary artery sling as a part of the VACTERL association. Barium meal X-ray showed pressure effects on the esophagus, and computed tomography angiography revealed pulmonary artery sling. Pneumonia management was done. However, the parents of our patient refused to give consent for the surgical correction of this vascular anomaly. Three months after discharge from the hospital, the patient was visited, at which time the parents again refused surgery and treatment for their daughter despite our recommendations.

Re-implantation & Slide tracheoplasty





Surgical treatment

J Thorac Cardiovasc Surg. 2012 Jan;143(1):144-51. doi: 10.1016/j.jtcvs.2011.09.038. Epub 2011 Nov 3.

Pulmonary artery sling: current results with cardiopulmonary bypass.

Backer CL[1], Russell HM, Kaushal S, Rastatter JC, Rigsby CK, Holinger LD.

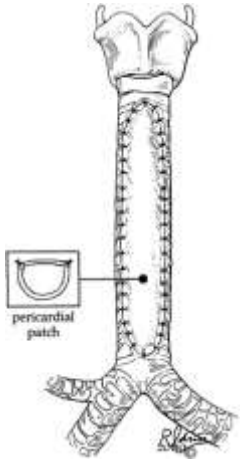
Author information

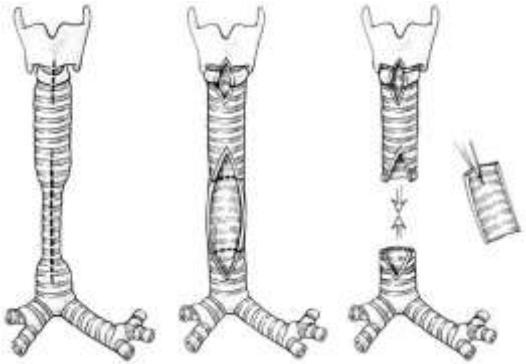
Pericardial patch tracheoplasty
1985 - 1994

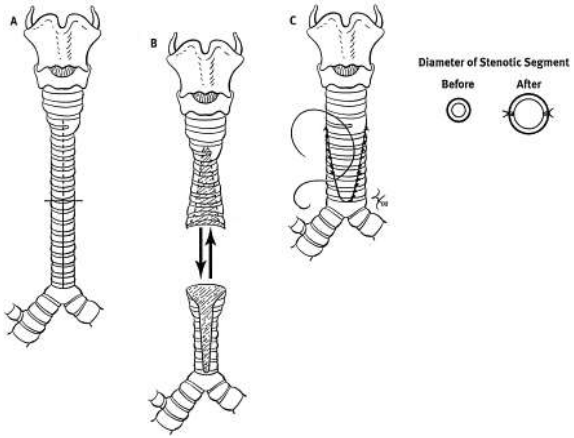
→ Tracheal autograft technique
1996 - 2001

→ Tracheal resection

→ Slide tracheoplasty technique
2002 to the present







Clinical outcomes of slide tracheoplasty in congenital tracheal stenosis FREE

Su Ryeun Chung, Ji-Hyuk Yang ✉, Tae-Gook Jun, Wook Sung Kim, Yong Han Kim, I-Seok Kang, June Huh, Jin Young Song, Joongbum Cho

European Journal of Cardio-Thoracic Surgery, Volume 47, Issue 3, 1 March 2015, Pages 537–542, https://doi.org/10.1093/ejcts/ezu196
Published: 12 May 2014 **Article history** ▾

- 18 patients (2004-2011), 2.5 months
- 8 had pulmonary sling

Results: 1 death (agenesis of rt lung)

- 2 re-operated for tracheal stenosis
- 13- symptoms-free
- 2- tracheostomy (tracheomalacia)

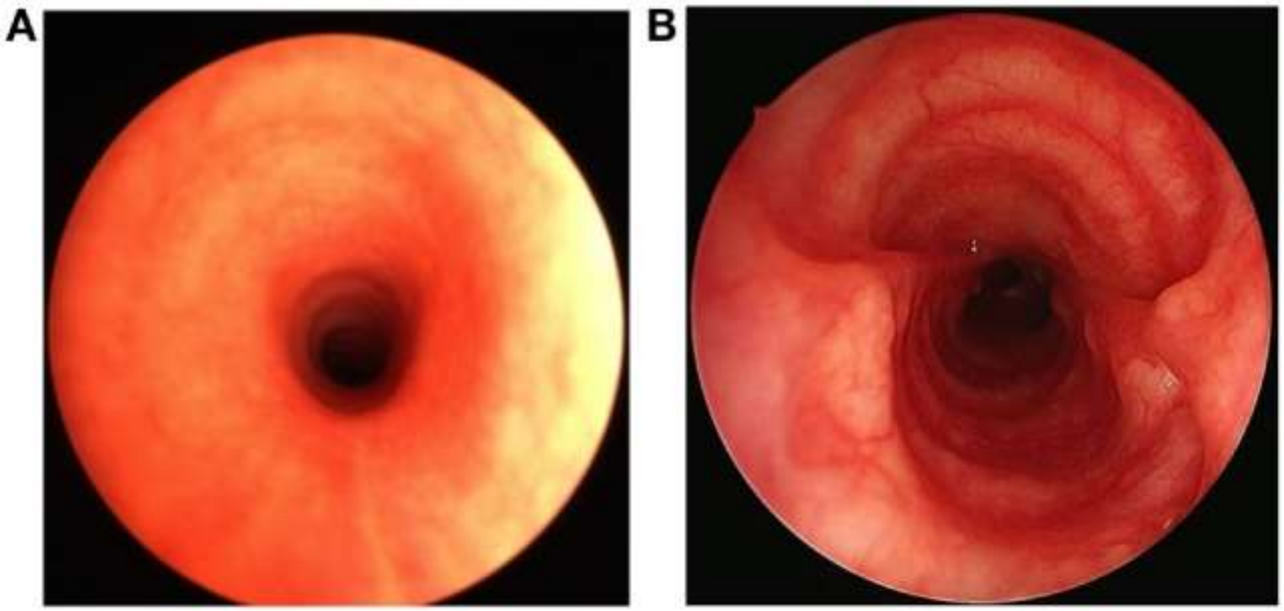
Conclusion: slide tracheoplasy is a effective technique. However, shortening of the trachea after reconstruction may give rise to recurrent obstruction.

Front Pediatr. 2017 Apr 10;5:67. doi: 10.3389/fped.2017.00067. eCollection 2017.

Complicated Postoperative Course after Pulmonary Artery Sling Repair and Slide Tracheoplasty.

Weber A[1], Donner B[2], Perez MH[3], Di Bernardo S[1], Trachsel D[4], Sandu K[5], Sekarski N[1].

Author information



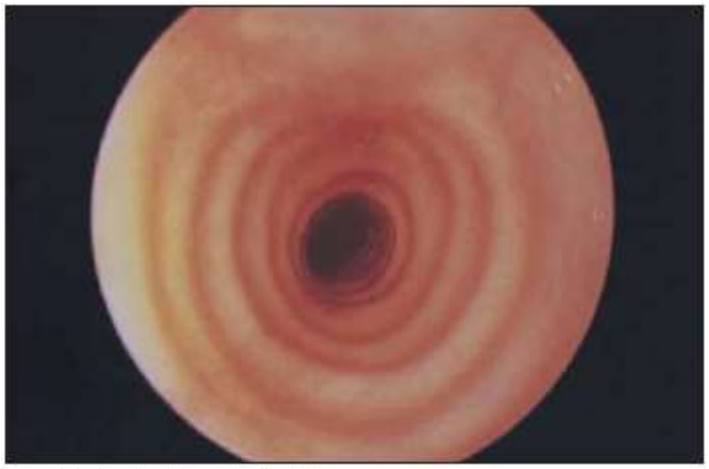
"Management of PAS and tracheal stenosis can still be challenging. However, good long-term outcome can be achieved if the initial postoperative phase is overcome".

Treatment

ORIGINAL ARTICLE

Nonoperative Management of Complete Tracheal Rings

Michael J. Rutter, FRACS; J. Paul Willging, MD; Robin T. Cotton, MD



Complete tracheal rings (patient 2).

Background: Children with complete tracheal rings are often challenging to manage. Most children will present early with a severely compromised airway and will require tracheal reconstruction.

Objective: To show that a small number of minimally symptomatic patients with complete tracheal rings experience airway growth over time and do not require tracheoplasty.

Design: A retrospective medical chart review over a 10-year period.

Setting: A tertiary care pediatric hospital.

Patients: Children (N = 10) with a diagnosis of complete tracheal rings, confirmed on bronchoscopy, who were observed for a minimum of 1 year prior to determining the need for tracheoplasty.

Main Outcome Measures: Patient symptoms, bronchoscopic findings, airway size, and the progression of these over time. Other congenital anomalies, the reason for initial diagnosis, and the need for tracheoplasty were documented.

Results: The 10 patients in our series fell into the following 3 categories: 5 patients were minimally symptomatic or asymptomatic, showed bronchoscopic evidence of progressive airway growth, and did not require tracheoplasty; 2 patients had worsening symptoms of exercise intolerance, showed minimal airway growth, and ultimately required tracheoplasty; and 3 patients are still being clinically observed and may eventually require tracheoplasty. Periods of observation have varied from 1 year to over 12 years.

Conclusions: Not all patients with complete tracheal rings require tracheoplasty. Some have satisfactory airway growth and do not require airway reconstruction. A period of observation to monitor airway growth and clinical symptoms is safe and may spare some patients from undergoing unwarranted airway reconstruction.

Arch Otolaryngol Head Neck Surg. 2004;130:450-452

בחזרה לחולה...

- <u>ביקורת מרפאת ריאות:</u>

2/18- בת 4 חודשים, ללא קליניקה נשימתית עלתה במשקל 1.3 ק"ג במהלך חודש וחצי מאז שחרורה (משקל עדכני 4.1 ק"ג)

- <u>אשפוז במרץ 18 :</u> בשל מצוקה נשימתית

שוקלת 3.5 ק"ג- ירידה של 600 ג' בחודש.

שיפור תחת סטראידים סיסטמיים, ללא תגובה למרחיבי סימפונות. RSV חיובי במשטף.

משקל בשחרור-3.850

- נמתין לביקורת הבאה..

פנינו לאן...?