

64 MDCT 3D Assessment of Complex Congenital Heart and Bilateral Tracheal Bronchi: A Case Report

ศุภิกา กฤษณีไพบุลย์^{1*}
สุภาพร โรยมณี²

64 MDCT 3D Assessment of Complex Congenital Heart and Bilateral Tracheal Bronchi: A Case Report.

Supika Kritsaneepaiboon¹, Supaporn Roymanee²

¹Department of Radiology, ²Department of Pediatrics, Faculty of Medicine, Prince of Songkla University, Hat Yai, 90110, Thailand.

*E-mail: supikak@yahoo.com

Songkla Med J 2015;32(2):103-107

บทคัดย่อ:

ภาวะแขนงหลอดลมออกจากท่อนลมใหญ่เป็นส่วนหนึ่งของความผิดปกติแต่กำเนิดของหลอดลมและท่อนลมใหญ่ นอกจากนั้นภาวะแขนงหลอดลมออกจากท่อนลมใหญ่ทั้งสองข้างพบได้น้อยมากและมักไม่มีอาการ รายงานผู้ป่วยเด็กหญิงอายุ 8 ปี มีโรคหัวใจพิการแต่กำเนิดชนิดซับซ้อน ซึ่งประกอบด้วย หัวใจห้องล่างหนึ่งห้อง ลิ้นหัวใจไตรคัสปิดไม่เจริญ และหลอดเลือดแดงใหญ่ของปอดตีบอย่างมาก ได้พบภาวะแขนงหลอดลมออกจากท่อนลมใหญ่ทั้งสองข้างและภาวะกลุ่มอาการไม่มีม้ามอย่างบังเอิญจากการตรวจโดยเครื่องเอกซเรย์คอมพิวเตอร์และฉีดสารทึบรังสี

สรุป: ภาวะแขนงหลอดลมออกจากท่อนลมใหญ่ทั้งสองข้างสามารถพบได้ในผู้ป่วยภาวะกลุ่มอาการไม่มีม้าม และเชื่อว่าเป็นแขนงหลอดลมข้างขวาสองข้าง

คำสำคัญ: กลุ่มอาการที่ไม่สามารถแยกซ้าย-ขวาได้, กลุ่มอาการไม่มีม้าม, ภาวะแขนงหลอดลมออกจากท่อนลมใหญ่

¹ภาควิชารังสีวิทยา ²ภาควิชากุมารเวชศาสตร์ คณะแพทยศาสตร์ มหาวิทยาลัยสงขลานครินทร์ อ.หาดใหญ่ จ.สงขลา 90110

รับต้นฉบับวันที่ 16 มกราคม 2558 รับลงตีพิมพ์วันที่ 2 เมษายน 2558

Abstract:

The tracheal bronchus manifests a variety of tracheobronchial anomalies. Bilateral tracheal bronchi are rare. Anomalous tracheal bronchus is usually asymptomatic.

Case report: We report an 8-year old girl who had underlying complex congenital heart diseases: uni-ventricular heart, tricuspid atresia, and severe pulmonary stenosis and who was incidentally found to have asplenia syndrome and bilateral tracheal bronchi from the contrast enhanced multidetector computed tomography (MDCT).

Conclusion: The bilateral tracheal bronchi are more often found in patients with asplenia syndrome and considered bilateral right bronchi.

Keywords: asplenia, heterotaxy syndrome, tracheal bronchus

Introduction

Tracheal bronchus is relatively rare and often incidentally discovered during bronchoscope or at chest computed tomography (CT).¹⁻³ The presentation can vary from asymptomatic clinical with conservative treatment or respiratory problems in intubated patients.²⁻⁴ Tracheal bronchus is more common in patient with associated condition such as congenital heart disease than in patients without congenital heart disease.² This case presented bilateral tracheal bronchi with heterotaxy syndrome.

Case report

An 8-year old girl presented with underlying complex congenital heart diseases: uni-ventricular heart, tricuspid atresia, and severe pulmonary stenosis. The patient underwent bilateral Blalock Taussig shunt and then bilateral Glenn operation at 1 year of age and lateral Fontan operation at 7 years of age. She had no history of recurrent pneumonia.

The chest CT was performed to evaluate causes of desaturation. Arterial and venous enhanced CT scan of the chest (Phillips Brilliance 64 slice, protocol: 110 mAs, 120 kVp, 2mm, 40 mL of Ultravist 370) showed tricuspid atresia, right sided aortic arch, severe supra-valvular pulmonary stenosis, multiple small aorto-pulmonary collaterals and hepatic congestion (Figures 1A and 1B). The lateral Fontan SVC and IVC pathways showed no thrombus or stenosis (Figures 2A and 2B) in delayed scan images. Asplenia syndrome and bilateral tracheal bronchi (displaced apical subtype) were incidentally found with no associated atelectasis (Figures 3A, 3B and 3C).

Discussion

A true tracheal bronchus is the upper lobe bronchus arising from the lateral wall of the trachea usually within 2 cm and up to 6 cm from the carina and it could be tiny, stenotic or blind-ended. The

incidence of right and left tracheal bronchus is 0.1%–2% and 0.3%–1% respectively.^{1,2} The tracheal bronchus is classified into displaced or supernumerary subtype. It is defined as displaced type if one branch of the upper lobe bronchus is missing and described as supernumerary subtype when the upper lobe bronchus has a normal and complete trifurcation. The displaced subtype is more common than supernumerary subtype.^{1,5} Anomalous tracheal bronchus is usually asymptomatic; however, recurrent pneumonia, bronchiectasis, focal emphysema, persistent right upper lobe atelectasis in intubated patients or respiratory distress can occur, which resulting from impaired drainage at the origin of tracheal bronchus.^{1,2,5} The angle between the tracheal bronchus and trachea can vary from 22° to 108°. Children with greater-angle tracheal bronchus or small-sized tracheal bronchus have less chance of accidental endotracheal intubation than the small-angle tracheal bronchus to occlude the lumen of tracheal bronchus leading to obstructed segmental airway and deficient ventilation of the remaining lung.² Associated anomalies include Down's syndrome, rib abnormalities, VATER syndrome, congenital heart diseases, abnormal pulmonary lobulation, and anomalous pulmonary arterial supply and venous drainage.^{4–6} Although true bilateral tracheal bronchi are extremely rare, it has been reported up to 24% of patients with asplenia syndrome.⁷ Patients with heterotaxy syndrome, asplenic subtype, have two “right sides” and absent spleen. The bronchial pattern in asplenia syndrome is bilateral right sided bronchi

or bilateral eparterial bronchi, thus the bilateral tracheal bronchi are believed to be bilateral right bronchi.^{2,7} MDCT is the non-invasive and reliable imaging technique to diagnose tracheal bronchus and also associated other anomalies, particularly for three dimensional (3D) assessment.^{7,8} The sensitivity of MDCT to diagnose tracheal bronchus is 100% and can be assistive to detect or exclude this airway abnormality.⁷ Furthermore, the MDCT can provide the distance between the tracheal bronchus and carina, the size of tracheal bronchus, the angle between the tracheal bronchus and the trachea, and particularly 3D-MDCT in particularly can dynamically demonstrate the special relationship between arterial and tracheal anomalies.^{7,8}



Figure 1A Axial enhanced MDCT shows tricuspid atresia (white arrow) seen as hypodense fatty tissue preventing connection between right atrium and hypoplastic right ventricle (*). Right sided aortic arch (gray arrow) is also demonstrated.

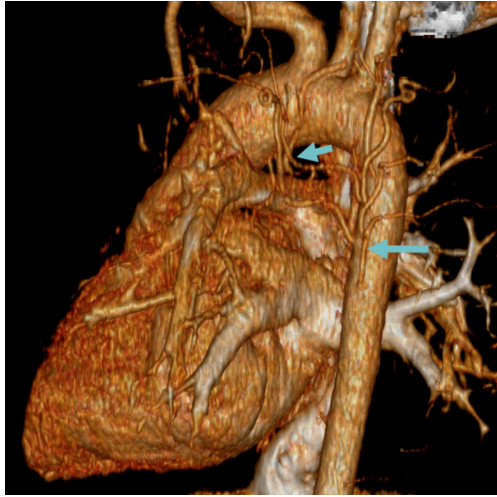


Figure 1B MDCT-3D volume rendering view of cardiac and great vessels showing multiple small major aortopulmonary collateral arteries (MAPCAs) (arrows) arising from the proximal descending thoracic aorta.

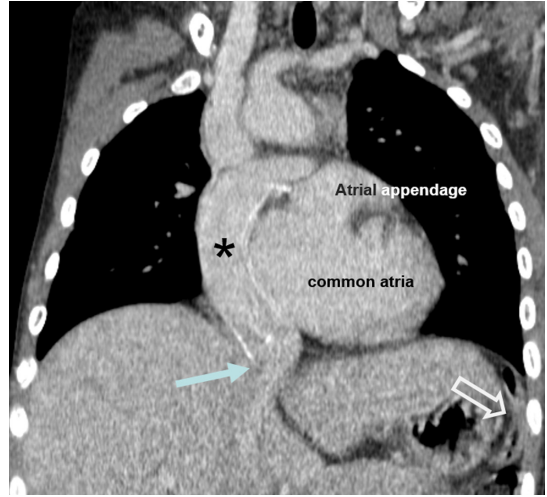


Figure 2B Coronal MPR enhanced MDCT showing lateral Fontan tunnel (*) with patent inferior vena cava (IVC) (arrow) pathway. The spleen is absent at left upper abdomen (open arrow) indicating asplenia or right isomerism.

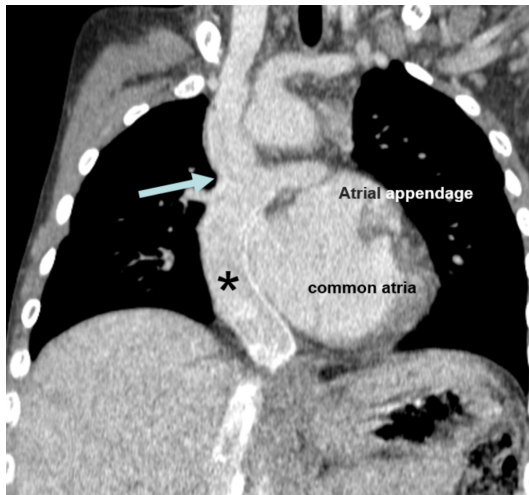


Figure 2A Coronal multi-planar reformation (MPR) enhanced MDCT showing lateral Fontan tunnel (*) with patent superior vena cava (SVC) (arrow) pathway.

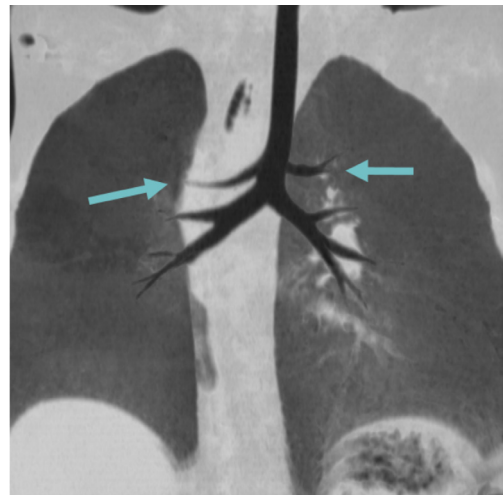


Figure 3A Coronal minimal-intensity projection (minIP) image reveals bilateral tracheal bronchi (displaced apical subtype) (arrows).



Figure 3B 3D-shaded surface display (SSD) opaque lung image displaying bilateral tracheal bronchi (arrows).



Figure 3C Coronal MPR MDCT (lung window) displaying bilateral minor fissures indicating bilateral trilobed lungs (arrows) in right isomerism.

Conclusion

The bilateral tracheal bronchi are more often found in patients with congenital heart disease and asplenia syndrome considered to be bilateral right bronchi. 3D-MDCT is the reliable, noninvasive and powerful technique to demonstrate associated arterial and tracheal anomalies.

References

1. Ghaye B, Szapiro D, Fanchamps JM, et al. Congenital bronchial abnormalities revisited. *Radiographics* 2001; 21: 105 - 19.
2. Ming Z, Lin Z. Evaluation of tracheal bronchus in Chinese children using multidetector CT. *Pediatr Radiol* 2007; 37: 1230 - 4.
3. Cope R, Campbell JR, Wall M. Bilateral tracheal bronchi. *J Pediatr Surg* 1986; 21: 443 - 4.
4. O'Sullivan BP, Frassica JJ, Rayder SM. Tracheal bronchus: a cause of prolonged atelectasis in intubated children. *Chest* 1998; 113: 537 - 40.
5. Kumagai Y, Jinguji M, Tanaka D, et al. An adult case of bilateral true tracheal bronchi associated with hemoptysis. *J Thorac Imaging* 2006; 21: 293 - 5.
6. Chau KW, Ng DK, Chong AS, et al. Tracheal bronchus. *Hong Kong Med J* 2003; 9: 71 - 2.
7. Ming Z, Hong S, Biao J. Images in cardiovascular medicine. Asplenia syndrome with bilateral tracheal bronchi. *Circulation* 2008; 118: 196 - 7.
8. Watabnabe K, Uese K, Higuchi O, et al. Three-dimensional computed tomographic findings of bilateral tracheal bronchus. *Pediatr Cardiol* 2009; 30: 87 - 8.