Major aortopulmonary collateral artery aneurysms- a rare cause of airway narrowing

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Text

To the Editor,

A 6-year-old cyanotic child with a diagnosis of ventricular septal defect- pulmonary atresia underwent computed tomography angiography (CTA) to look for the status of pulmonary arteries, aorto-pulmonary collaterals and the coronaries. CTA showed a perimembranous ventricular septal defect with pulmonary atresia. The pulmonary arteries were confluent, with the presence of multiple major aortopulmonary collateral arteries (MAPCAs). Three MAPCAs were seen supplying the right lung and one was supplying the left lung. The MAPCAs supplying the right lung showed aneurysmal dilatations along their course, with the largest collateral measuring 14 mm in calibre, arising from the descending thoracic aorta at D7 level. There was extrinsic compression of the bronchus intermedius between the dilated right descending pulmonary artery and one of the aneurysmally dilated MAPCAs **(Fig. 1-2)** . Aortic arch was right-sided, with four-vessel arch branching pattern, showing anomalous direct origin of right vertebral artery from the arch**(Fig. 2)** . The arch was dilated causing mild side-to-side tracheal flattening. Bilateral lungs showed mosaic attenuation, with presence of azygous fissure. Coronaries were normal with normal systemic and pulmonary venous drainage.

When the pulmonary arterial tree is underdeveloped, such as in pulmonary stenosis or atresia, aortopulmonary collaterals act as systemic to pulmonary shunts and maintain blood supply to the lungs (1,2). MAPCAs with calibre $>$ 3mm are usually characterized by high systemic pressure. Aneurysm formation and segmental pulmonary hypertension are known complications of hypertensive MAPCAs (1). Airway compression by MAPCAs is rare. Compression of central airways may present with choking episodes and respiratory distress (3). Moreover, airway compression can lead to bronchomalacia and patients may present with massive hemoptysis due to associated bronchial erosion (4,5). Variant arch anatomy is also associated with airway compression particularly when there is complete vascular ring. Moreover the right aortic arch can also compress the airway in cases with associated bronchial aberrations, such as tracheal bronchus (3). CTA nicely delineates the anatomy of aortopulmonary collaterals in addition to the native pulmonary circulation which helps in devising optimal management plan.

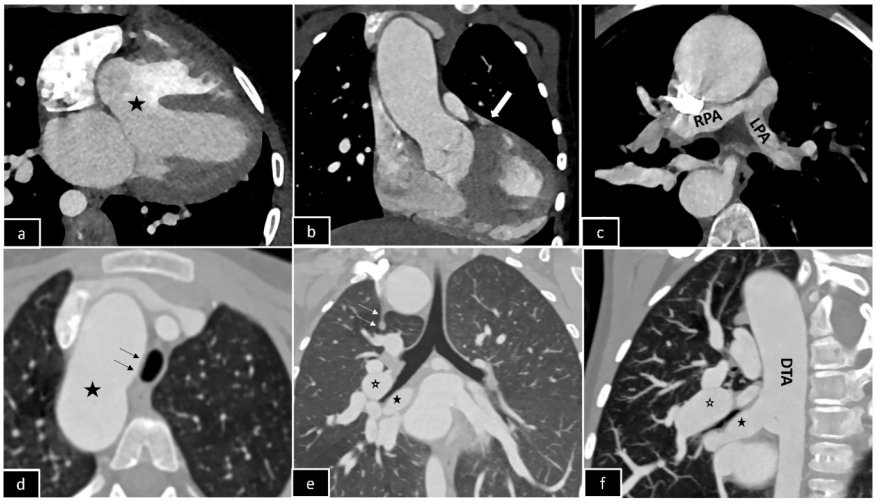

Figure 1: CTA images in a 6-year-old child with cyanosis. Axial images showing **a)** Perimembranous VSD (asterik), **b)**Pulmonary atresia (arrow) with confluent pulmonary arteries**(c)** . The right sided aortic arch (asterik) was dilated, causing side-to-side flattening of the trachea (arrows in **d**).**e)** Coronal maximum intensity projection image showing the extrinsic compression of bronchus intermedius between the right descending pulmonary artery (white asterik) and aneurysmal MAPCA (black asterik). Azygous fissure was present (white arrows). **f)**Oblique coronal maximum intensity projection image showing the extrinsic compression of bronchus intermedius by the right descending pulmonary artery (white asterik) and aneurysmal MAPCA (black asterik) arising from the descending thoracic aorta (DTA).

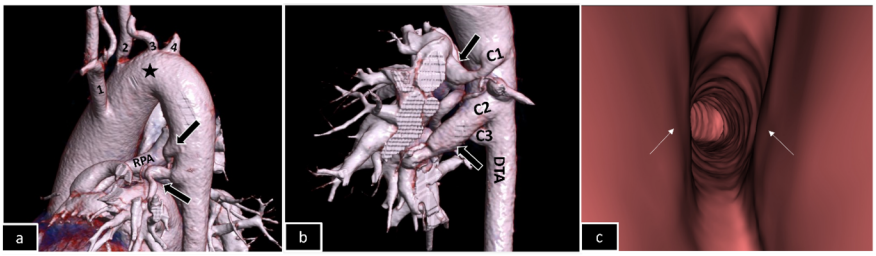

Figure 2: Three-dimensional virtual reconstructed images showing **a)** right sided aortic arch (asterik) with four-vessel arch branching pattern having separate origins of left brachiocephalic trunk (*1*), right common carotid artery (*2*), right vertebral artery (*3*) and right subclavian artery (*4*). The MAPCAs are seen arising from the DTA (arrows). **b)** Sagittal image of the descending thoracic aorta showing the right-sided MAPCAs (*C1, C2, C3*) arising from the DTA, with aneurysmal dilatations along their course (arrows). **c)** Virtual bronchoscopy image at the level of bronchus intermedius showing bilateral, smooth extrinsic compression (arrows).

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