Multidetector CT angiographic evaluation of intralobar pulmonary sequestration with congenital cystic adenomatoid malformation in a patient of tetralogy of Fallot (TOF).

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Learning objectives

- The aim of this poster is to show the role of 64- multidetector computed tomography (MDCT) angiography in the comprehensive evaluation of anatomic variations in pulmonary arterial circulation and other associated lung parenchymal anomalies in patients of tetralogy of fallot (TOF).
- Complex pulmonary vascular blood supply is common in patients of TOF with pulmonary atresia, major systemic to pulmonary collateral arteries and hypoplastic or deficient central pulmonary arteries.
- Arterial and venous vessels were analyzed using three-dimensional vascular exploration tools.
- This report presents an unique case of intralobar pulmonary sequestration and congenital cystic adenomatoid malformation (CCAM) combined with complex congenital heart disease.
Background

A 20-year old female presented in cardiology department with clinical features of cyanotic heart disease. On echocardiography pulmonary atresia with VSD was found. Lung parenchymal anomalies were also suspected on basis of chest x-ray (Fig.1) which showed a large hyperlucent area in left lower zone and an ill-defined oval opacity in right lower zone. So, patient was referred to radiology department for MDCT angiography for the evaluation of anatomic variations in pulmonary arterial circulation, major aorto-pulmonary collateral arteries (MAPCAs) and other associated lung parenchymal abnormalities.

- In a subset of TOF with marked infundibular stenosis, there may be complete obstruction of pulmonary blood flow. This subset has been described as TOF with pulmonary atresia and forms a specific type of pulmonary atresia with ventricular septal defect (PA-VSD).
- The pulmonary circulation in PA-VSD is extremely variable and depends on the presence or absence of native pulmonary arteries, PDA, and MAPCAs.¹
- On the basis of the characterization of the pulmonary circulation, PA-VSD is classified into three types.² In type A, the native pulmonary arteries are present and are supplied by the PDA. In type B, pulmonary blood flow is provided by both native pulmonary arteries and by MAPCAs. In type C, native pulmonary arteries are absent and the blood supply is only through MAPCAs.
- Precise characterization of the condition of pulmonary arteries and MAPCAs is of paramount importance in managing patients with PA-VSD. Using 3D imaging software, a complex pulmonary blood supply can be noninvasively and accurately imaged with high-resolution MDCT.³⁵

In addition to complex pulmonary vascular blood supply, pulmonary sequestration of the right lower lung and CCAM of left lower lobe were also detected (Fig.4).

Pulmonary sequestration is a rare congenital malformation defined by dysplastic and nonfunctioning pulmonary tissue which receives its blood supply from an anomalous systemic artery. Extralobar and intralobar sequestrations are distinguished by their venous drainage pattern: in intralobar sequestration, the venous drainage is through the pulmonary veins, whereas in extralobar sequestration, the veins drain into the systemic veins.⁶⁷

CCAM is a congenital lung mass of unknown cause resulting from disorganized hamartomatous and adenomatoid proliferation of primary bronchioles, which are in communication with the bronchial tree. There are three types of CCAM. The most
common subtype is a type 1CCAM, in which there is at least one dominant cyst that is larger than 2 cm in size.\textsuperscript{8}
Imaging findings OR Procedure details

MDCTA scanning protocol

MDCT angiographic evaluation of the thorax and upper abdomen was performed on 64-slice scanner (Philips Brilliance 64, Netherland) using a single injection of iohexol 350mgI/ml (Omnipaque, GE Healthcare) at a dose of 1.5 mL/kg. Contrast medium was injected with dual-head power injector at a rate of 3.0 mL/s followed by 20 ml of saline chase. Other technical parameters are as follows: detector collimation, 64 × 0.6 mm; pitch, 1.2; slice thickness, 0.75 mm; reconstruction interval, 0.4 mm; and gantry rotation time, 0.33 second. All image data are evaluated using a 3D post processing workstation with Syngo software (Siemens Healthcare). Various image reformatting techniques including curved planar reconstruction, maximum intensity projection (MIP), and volume-rendering technique (VRT) are used to get all the clinically relevant information. Earlier reports have shown that echocardiography can accurately delineate the anatomy of the central pulmonary arteries in infants with PA-VSD, but echocardiography is often of limited value in older children because of poor acoustic windows. Furthermore, echocardiography does not provide the surgeon with a precise anatomic road map.9-10

Final Report - Tetralogy of Fallot (TOF) with marked pulmonary infundibular stenosis, subaortic ventricular septal defect (VSD), patent ductous arteriosus (PDA) and partial overriding of aorta were found (Fig.2). PDA was joining with left pulmonary artery (LPA) (Fig.3). A large collateral was arising from the PDA and supplying to the right lower medial basal segment of lung (Fig.3). An intralobar sequestration with Type-1 congenital cystic adenomatoid malformation (Fig.4), which has not been described previously in these patients, were imaged. The arterial blood supply to sequestrated lung from a branch of celiac trunk and venous drainage through the pulmonary veins are shown on the volume rendered images (Fig.4E)
Fig. 1: Chest X-ray PA view of a 20-year old female shows a large hyperlucent area in left lower zone and a faint oval opacity in right lower zone.

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**Fig. 2:** Transverse and oblique axial maximum-intensity-projection images A & B in a 20-year-old female show subaortic ventricular septal defect and partial overriding of aorta. Oblique maximum-intensity-projection (C) and volume rendered MDCT (D) images show marked pulmonary infundibular stenosis and hypoplastic main pulmonary artery.

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Fig. 3: Sagittal oblique maximum-intensity-projection (A) and oblique volume-rendered MDCT (B-D) images show patent ductus arteriosus (arrow) to left pulmonary artery. A large collateral is arising from the PDA and supplying to the right lower medial basal segment of lung (double arrow in C).

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**Fig. 4:** Coronal and axial CT lung window images (A&C) show type 1 CCAM manifesting as a large cystic lung lesion in the left lower lobe and sequestrated lung (S) as a complex thick walled multiseptated cystic lesion with air-fluid levels in posterior basal segment of right lower lobe (A,C&D). Coronal oblique contrast-enhanced maximum intensity projection MDCT image shows an anomalous artery (arrow) arising from coeliac trunk into sequestered lung (S). Two anomalous veins (double arrows) draining into the left atrium are also noted preoperatively.

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Conclusion

- This poster illustrates the use of 64-MDCT in comprehensive evaluation of different anatomic structures, including the heart, pulmonary and systemic thoracic vasculature, lungs, and abdomen, when evaluating patients with PA-VSD.
- Multidetector CT angiography is the investigation of choice and allows the noninvasive and accurate delineation of all of these abnormalities.
- Multiplanar and 3D CT reconstruction images can improve communication of anatomic details to clinicians.
- MDCT has proved to be an invaluable diagnostic and decision-making tool as a compliment to echocardiography and increasingly as a substitute for invasive angiography in the management of PA-VSD.
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