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Case Series

Radiology Section

Role of Multidetector Computed Tomography in Congenital Pulmonary Arterial Malformations: A Case Series

SHREYA P DESAI[1], BHADRA Y TRIVEDI[2], RADHIKA H PANDYA[3], SHREYA D PATEL[4], VIRAL B PATEL[5]

ABSTRACT

Congenital and developmental abnormalities of pulmonary vasculature encompass a wide-ranging spectrum of conditions, which exhibit myriad differences. Most of them present at birth; however, age of clinical presentation varies according to blood flow restriction and associated airway compromise. In this case series, we aim to describe embryological basis of development of pulmonary vasculature and normal anatomy. Authors intend to highlight case based imaging findings of pulmonary arterial congenital malformations both in isolation as well as in association with other congenital cardiac malformations referred at the rural tertiary healthcare hospital. Multidetector Computed Tomography (MDCT) scan not only proved to be an excellent non invasive modality in diagnosis of Pulmonary Artery (PA) malformations, associated cardiac and extra cardiac malformations, but is also an indispensable tool to provide adequate road map for intervention.

Keywords: Arterial sling, Pulmonary atresia, Tetralogy of fallot, Transposition, Vascular malformations

INTRODUCTION

Congenital abnormalities of the pulmonary vascular system are very heterogeneous, with variable presentation, management and outcome based on severity. Many of these malformations are associated with additional abnormalities of other organ systems or vascular structures [1].

Accurate and timely diagnosis of malformations of Pulmonary Arteries (PA) are essential as many of these are life threatening. The use of conventional modalities like X-ray and ultrasound has limitations in accurate detection and characterisation of pathology as well as evaluation of associated lung parenchymal involvement. Transthoracic Echocardiography (TTE) remains an effective tool in evaluation of these conditions; however, Multi Detector Computed Tomography (MDCT) proves as indispensable, operator independent tool with excellent resolution, faster scan time, and high quality reformatted images providing road map for intervention.

CASE SERIES

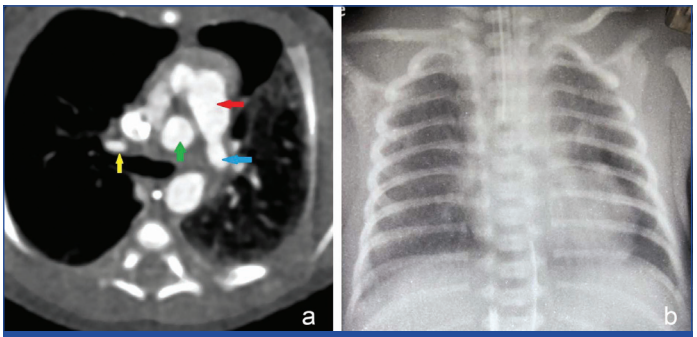
I) Unilateral Interruption of Pulmonary Artery (PA)

Case 1

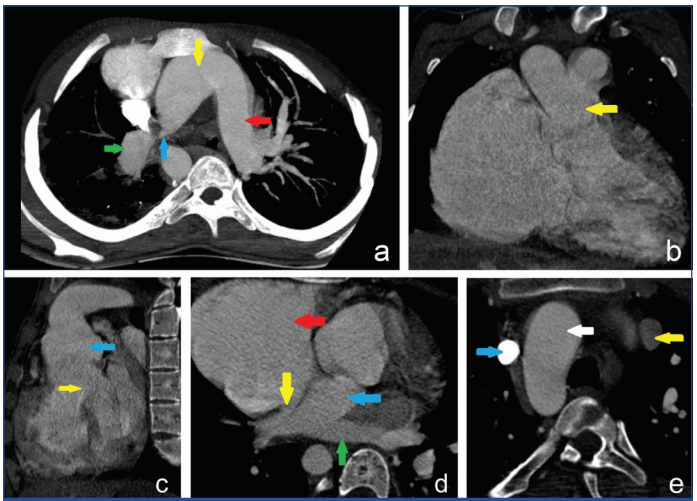
A newborn male was referred on day 7 of life for persistent pulmonary arterial hypertension. On MDCT, Right Pulmonary Artery (RPA) was not visualised in its usual location and a large collateral continuing from Patent Ductus Arteriosus (PDA) was supplying right intraparenchymal PA, suggesting diagnosis of unilateral interruption of PA [Table/Fig-1a]. Both lung fields appeared to have normal volume on chest X-ray [Table/Fig-1b]. The operative procedure considered was re-implantation of the RPA.

Case 2

An 18-year-old male patient presented with New York Heart Association (NYHA) grade III symptoms for past 12-13 years. His cardiac echocardiography was interpreted as double outlet Right Ventricle (RV) with malaligned great arteries, inlet Ventricular Septal Defect (VSD), dilated Right Atrium (RA) and RV, good biventricular function and moderate tricuspid regurgitation. The MDCT revealed conotruncal anomaly with absent RPA [Table/Fig-2a,b], perimembranous VSD [Table/Fig-2c], dilated RA [Table/Fig-2d], right sided aortic arch and bilateral superior vena cava [Table/Fig-2e] with oligemic right lung field.



[Table/Fig-1]: a) Absent Right Pulmonary Artery (RPA) and intact intrapulmonary part of PA (yellow arrow) indicating a separate embryonic origin. Main PA (red arrow) giving rise of Left Pulmonary Artery (LPA) (blue arrow) anterior to the right of ascending aorta (green arrow). b) Chest radiograph showing normal lung fields.



[Table/Fig-2]: a) Absent Right Pulmonary Artery (RPA), Truncus arteriosus (yellow arrow) with common origin of Pulmonary Artery (PA) and aorta, Left Pulmonary Artery (LPA) (red arrow), ductus arteriosus (blue arrow) arising from ascending aorta and supplying intra pulmonary RPA (green arrow); b) Single arterial trunk (circle and yellow arrow) giving origin to both aorta and LPA; c) Truncus arteriosus (blue arrow) overriding perimembranous VSD (yellow arrow); d) Dilated RA (red arrow), left atrium (blue arrow), patent foramen ovale (yellow arrow), common channel formed by left superior and inferior pulmonary veins draining into left atrium (green arrow); e) Right sided aortic arch (white arrow) right superior vena cava (blue arrow) and left superior vena cava (yellow arrow).

II) Pulmonary Arterial Sling

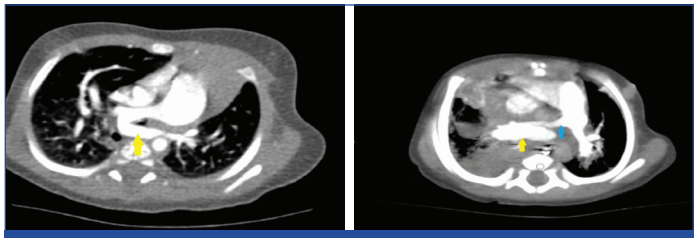
Case 3

A two-month-old female child with tachypnea underwent Transthoracic Echocardiography (TTE); which suggested ostium secundum Atrial Septal Defect (ASD), shunting left to right, with strong suspicion of anomalous origin of Left Pulmonary Artery (LPA) from RPA; dilated RA and RV with Pulmonary Arterial Hypertension (PAH). The MDCT confirmed the anomalous course of LPA, looping posterior to trachea between trachea and oesophagus. Mobilisation of LPA with reconnection to main pulmonary trunk was suggested as a part of surgical repair [Table/Fig-3].

III) Dilated main Pulmonary Artery (PA) with Right Pulmonary Artery (RPA) Aneurysm

Case 4

A 10-month-old female child was admitted with complaint of breathlessness and fever since 10 days. Clinical features and investigation were positive for sepsis. Pulmonary thromboembolism was suspected on TTE. However, MDCT revealed multiple varying sized outpouchings from the RPA and a large saccular aneurysm along its posterior wall. [Table/Fig-4]. The patient had normal pulmonary arterial pressure and no associated intra-cardiac shunts, chronic cardiac or pulmonary disease. The neonate was on high ventilator support despite of having normal lung parenchyma evident on CT. The surgery planned was plication of PA and release the pressure over airways.



[Table/Fig-3]: Anomalous left PA (yellow arrow) arising from posterior side of Right Pulmonary Artery (RPA) and looping posterior to trachea between trachea and oesophagus. **[Table/Fig-4]:** RPA aneurysm (yellow arrow), RPA (blue arrow). (Images from left to right)

IV) Pulmonary Atresia with Teratology Of Fallot (TOF)

A. Pulmonary atresia with confluent Pulmonary Arteries (PA)

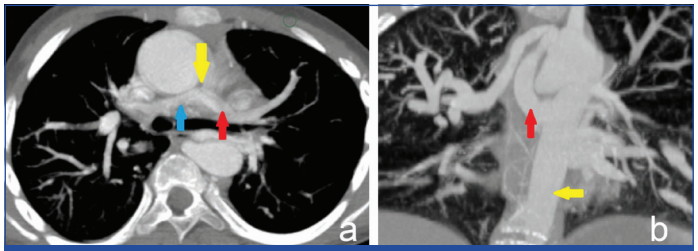
Case 5

A 13-year-old female child presented with breathlessness on exertion since one month. Examination, showed cyanosis, clubbing with SpO_2 of 70% on room air. The MDCT images showed pulmonary atresia with confluent hypoplastic RPA and LPA [Table/Fig-5a]. It was associated with big sub-aortic VSD, overriding of aorta and RV hypertrophy. Bilateral superior vena cava and three Major Aortopulmonary Collaterals (MAPCAs) were seen [Table/Fig-5b]. The McGoon ratio was 1.172 and Nakata index 192.5/Body Surface Area (BSA)=215.32 mm^2, thus patient was operated for Blalock Taussig shunt to increase pulmonary blood flow followed by second stage corrective surgery.

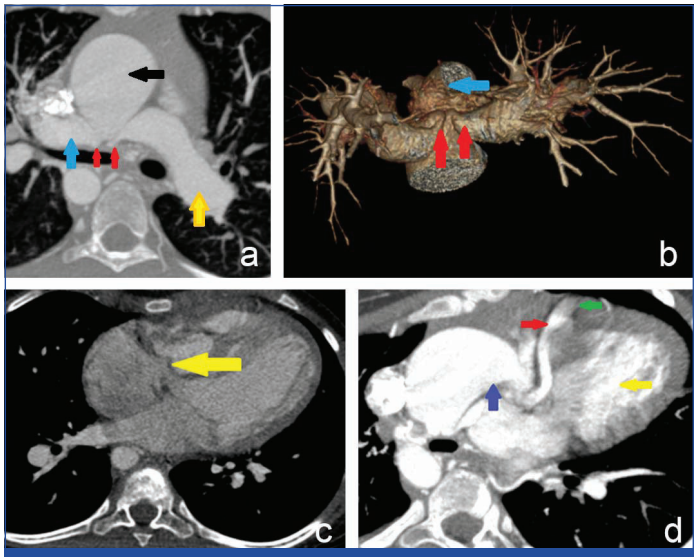
B. Pulmonary atresia with non confluent Pulmonary Arteries (PA)

Case 6

A 10-year-old female child presented with weakness, breathlessness on exertion and poor weight gain since birth. Cyanosis and clubbing was observed on examination. MDCT showed features of non confluent branch PA filled by bilateral PDA [Table/Fig-6a,b], TOF with MPA atresia as well as tricuspid atresia [Table/Fig-6c] and coronary cameral fistula with RV [Table/Fig-6d]. The patient underwent unifocalisation and confluence creation between RPA with LPA as well as bi-directional Glenn shunt, atrial septectomy, bilateral PDA division and suturing.



[Table/Fig-5]: a) Pulmonary trunk atresia (yellow arrow) with hypoplastic right and left Pulmonary Arteries (PA) (blue and red arrows); b) Large aortopulmonary collateral (red arrow) arising from descending thoracic aorta (yellow arrow).

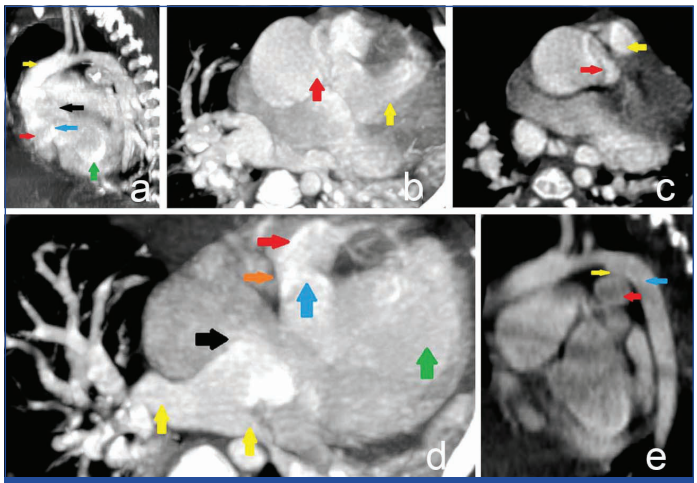


[Table/Fig-6]: a) Non confluent right pulmonary (blue arrow) and Left Pulmonary Artery (LPA) (yellow arrow) supplied by respective ductus arteriosus (red arrows) arising from ascending aorta (black arrow); b) Volume rendered image of Non confluent PA supplied by respective ductus arteriosus (red arrows) arising from ascending aorta (blue arrow); c) Tricuspid atresia; d) coronary cameral fistula (red arrow), arising from ascending aorta (blue arrow) and supplying right ventricular wall (green arrow), left ventricle (yellow arrow).

V) Transposition of Great Vessels (TGA)

Case 7

A four-month male child presented with fever and dyspnea for 15-20 days. TTE was suggestive of Complex Cyanotic Congenital Heart Disease with ASD, VSD, tricuspid valve atresia, hypoplastic RV, malposed great arteries and PDA and suspected coarctation of aorta. MDCT revealed transposition of great arteries and atrioventricular concordance [Table/Fig-7a,b] with antero-posterior relationship of aorta and PA at their origins [Table/Fig-7c], ASD, VSD [Table/Fig-7d] and wide short PDA [Table/Fig-7e]. Coarctation was ruled out on MDCT. The RV was hypoplastic with tricuspid atresia [Table/Fig-7d] thus the patient underwent PA banding to prepare for Glenn shunt in future.



[Table/Fig-7]: TGA a) Aorta (yellow arrow) arising from Right Ventricle (RV) (red arrow), Pulmonary Artery (PA) (black arrow) arising from left ventricle (green arrow), Ventricular Septal Defect (VSD) (blue); b) PA (red arrow) arising from left ventricle (yellow arrow); c) Aorta (yellow arrow) arising anterior to PA (red arrow); d) ASD (black arrow), VSD (blue arrow), RV (red arrow) with tricuspid atresia (orange arrow), left ventricle (green arrow) and pulmonary veins (yellow arrows) draining into left atrium; e) Short wide PDA (yellow) between aorta (blue arrow) and Left Pulmonary Artery (LPA) (red arrow)

DISCUSSION

Lee ML et al., refined the concept and classified bronchopulmonary malformations, taking into account isolated and concurrent abnormalities of airway, arteries, and veins [Table/Fig-8] [2]. The present case series shows type B group of these malformations in isolation as well as in association with other congenital cardiac malformations. Type B is a less common group consisting of isolated malinosculation of the PA with a normal pulmonary airway and venous connections as well as systemic arterial supply. It mainly includes unilateral absence of PA, PA sling and idiopathic dilatation of pulmonary trunk. Other malformations like pulmonary valvular stenosis, atresia, hypoplasia, segmental stenosis, and anomalous origin of PAs from aorta are more common than isolated ones and mostly associated with TOF and double outflow tract of RV.

Type	Congenital bronchopulmonary vascular malformations
A	Isolated bronchial
B	Isolated arterial
C	Isolated venous
D	Mixed bronchoarterial
E	Mixed bronchovenous
F	Mixed arteriovenous
G	Mixed bronchoarteriovenous

[Table/Fig-8]: Classification of Bronchopulmonary malformation [2].

Unilateral Absence of Pulmonary Artery (UAPA), is also known as proximal interruption of PA since distal portion inside the lung is intact as it has a different embryonic origin arising from lung buds [3,4]. It is uncommon and seen in one of every 200,000 individuals [5]. The UAPA commonly occurs in association with other cardiovascular anomalies like TOF or cardial septal defects as in our case where it was associated with ASD and rarely occurs in isolation, where the presentation can be in late adulthood due to non specific symptoms like chest pain, infections and or haemoptysis. Altered development of a sixth aortic arch segment is thought to result in a ductal origin to a PA that leads to the proximal interruption of that vessel when the ductal tissue regresses at the time of birth [6]. The present cases showed absence of RPA and visualisation of the intrapulmonary branch which suggest their separate embryonic origins.

The PA sling occurs due to an abnormality during involution of the left sixth proximal aortic arch [5,7]. Patients usually presents with respiratory distress or repeated pulmonary infections [8]. The aberrant disposition of the LPA is due to its abnormal origin from the RPA with an abnormal trajectory across the tracheoesophageal space leading to tracheal compression. Idiopathic dilatation of Pulmonary Artery (PA), is a rare condition; though occurs in isolation, but are also associated with other congenital heart anomalies [9]. Exact aetiology is still not established; though it is thought to be due to weakness of middle elastic layer of arterial wall [10]. The present case included saccular aneurysmal dilatation of RPA without associated intra-cardiac shunts, chronic cardiac or pulmonary disease and normal pulmonary pressures.

Pulmonary atresia with TOF are combined anomalies which are a result of alteration in anterocephalad deviation of the developing outlet ventricular septum, or its fibrous remnant. Not only the septum fails to muscularised but there is also associated abnormal morphology of the septoparietal trabeculation which encircle the subpulmonary outflow tract. The malaligned VSD and the over-riding aorta is a result of deviation of the muscular outlet septum [11].

Pulmonary atresia is typically described in relationship to the presence of an Intact (PA-IVS) or absent Inter-Ventricular Septum (PA-VSD). The presence of a VSD allows for blood from the right heart to flow across the septum, out the aorta through a PDA to the lungs, and thus relatively normal sized branch PA. In patients with PA/VSD, MAPCAs are typically present and native PA is generally hypoplastic or absent. In both lesions, pulmonary blood flow is via the systemic circulation either via the PDA or from MAPCAs. One of our cases had MAPCAS and the other had bilateral PDA.

In a study by Freedom RM et al., including analysis of 27 patients with bilateral ducti, 15 patients showed distal bilateral ductal origin of non confluent PA as was in our case and ectopic of distal ductal origin of 1 PA in 9 cases [12]. Similarly, Peirone A et al., in their retrospective study of 11 newborns with bilateral ducti showed pulmonary atresia with non confluent PA as a dominant lesion [13]. Urgent early intervention is required in order to ensure that the PDA remains patent when this is the primary source of pulmonary blood flow [14].

The PA-VSD with MAPCAs is an occasional anomaly occurring in about 10/100,000 live births and the most severe form of TOF [15]. TTE and cardiac catheterisation with direct injection into each collateral vessel with indirect imaging of the native PA were the main imaging modalities in these patients however; advances in cardiac CT imaging has led to a shift away from invasive imaging for delineation of collateral supply and demonstration of the native PA if present [16]. CT evaluation of MAPCAs should involve an exact explanation of location of origin, size, and presence of origin stenosis, lung segment supplied as well as the anatomic relationship to adjacent structures such as the carina, oesophagus and bronchi. Knowledge of the PA structure and the confluence of the left and right PA are imperious for surgical planning.

The TGA is a congenital cardiac defect occurring due to an embryological discordance between the pulmonary trunk and aorta, with a prevalence of 4.7 per 10,000 live births [17]. The normal cardiac development requires the conotruncal septum to spiral toward the aortic sac thus dividing the truncus into the pulmonary and aortic tubes subsequently forming PA and aorta, respectively. When the conotruncal septum fails to follow its spiral course and instead continues to have a linear orientation, the aorta arises from the RV and the pulmonary trunk arises from the left ventricle. Dextro-TGA (D-TGA) is the most common form which is characterised by RV being positioned to right of left ventricle and aorta arising anterior to PA, thus forming two parallel circuits as observed in our case.

The 'D' suggests that aorta is present in anterior and to the right of PA, also known as oblique relationship, which is the most common, however the term D TGA is controversial as studies have found other types including antero-posterior and side-by-side patterns [18]. Our case showed an antero-posterior relationship of aorta and PA at their origins.

Complete parallel circuits with no connections are incompatible with life and presence of a PDA or a ventriculoseptal defect will permit some mixing of oxygen-rich and oxygen-poor blood thus providing a window for survival [17]. The present case showed Dextro-TGA (D-TGA) pattern of TGA with ASD, VSD and wide short PDA. The RV was hypoplastic thus the patient underwent PA banding to prepare for Glen shunt in future. [Table/Fig-9] represents the summary of presented cases findings.

Case	Age/Sex	Radiological findings on MDCT	Diagnosis
1	7 Days/M	Right Pulmonary Artery (RPA) was not visualised in its usual location and a large collateral continuing from Patent Ductus Arteriosus (PDA) was supplying right intraparenchymal pulmonary artery.	Unilateral interruption of pulmonary artery
2	18 Years/M	Conotruncal anomaly with absent RPA perimembranous VSD, dilated RA, right sided aortic arch and bilateral superior vena cava with oligemic right lung field.	Conotruncal anomaly with interrupted RPA
3	2 Month/F	Anomalous course of LPA, looping posterior to trachea between trachea and oesophagus.	Pulmonary artery sling

4	10 Month/F	Multiple varying sized outpouchings from the RPA and a large saccular aneurysm along its posterior wall.	Dilated main pulmonary artery with Right Pulmonary Artery aneurysm.
5	13 Year/F	Pulmonary atresia with confluent hypoplastic RPA and LPA, associated with big sub-aortic VSD, over-riding of aorta and RV hypertrophy. Bilateral superior vena cava and three Major Aorto Pulmonary Collaterals (MAPCAs) were seen.	TOF along with Pulmonary atresia and confluent pulmonary arteries
6	10 Year/F	Non confluent branch Pulmonary Artery (PA) filled by bilateral PDA. TOF with MPA atresia as well as tricuspid atresia and coronary cameral fistula with Right Ventricle (RV).	TOF along with Pulmonary atresia and non confluent pulmonary arteries
7	4 Month/M	Transposition of great arteries and atrioventricular concordance with antero-posterior relationship of aorta and pulmonary artery at their origins, ASD, VSD and wide short PDA. The RV was hypoplastic with tricuspid atresia.	Transposition of Great vessels

[Table/Fig-9]: Summary of present cases.
VSD: Ventricular septal defect; RA: Right artium; LPA: Left pulmonary artery; TOF: Teratology of fallot; ASD: Atrial septal defect

CONCLUSION(S)

Computed tomography is an indispensable operator independent and helpful tool due to improved resolution, faster scan time, high quality reformatted images providing not only details of complex anomalous structures but also optimal sizing of various vasculature thus providing a road map for decision making in final surgical/non surgical interventions.

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PARTICULARS OF CONTRIBUTORS:
1. Assistant Professor, Department of Radiodiagnosis, PS Medical College and Shree Krishna Hospital, Bhaikaka University, Anand, Gujarat, India.
2. Consultant Paediatric Cardiologist, Department of Cardiology, PS Medical College and Shree Krishna Hospital, Bhaikaka University, Anand, Gujarat, India.
3. Senior Resident, Department of Radiodiagnosis, PS Medical College and Shree Krishna Hospital, Bhaikaka University, Anand, Gujarat, India.
4. Junior Resident, Department of Radiodiagnosis, PS Medical College and Shree Krishna Hospital, Bhaikaka University, Anand, Gujarat, India.
5. Professor, Department of Radiodiagnosis, PS Medical College and Shree Krishna Hospital, Bhaikaka University, Anand, Gujarat, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Viral B Patel,
Professor, Department of Radiodiagnosis, Shree Krishna Hospital, PSMC,
Karamsad, Anand-388325, Gujarat, India.
E-mail: viralvp@charutarhealth.org

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