

Clinical-Medical Image

## MPA Aneurysm

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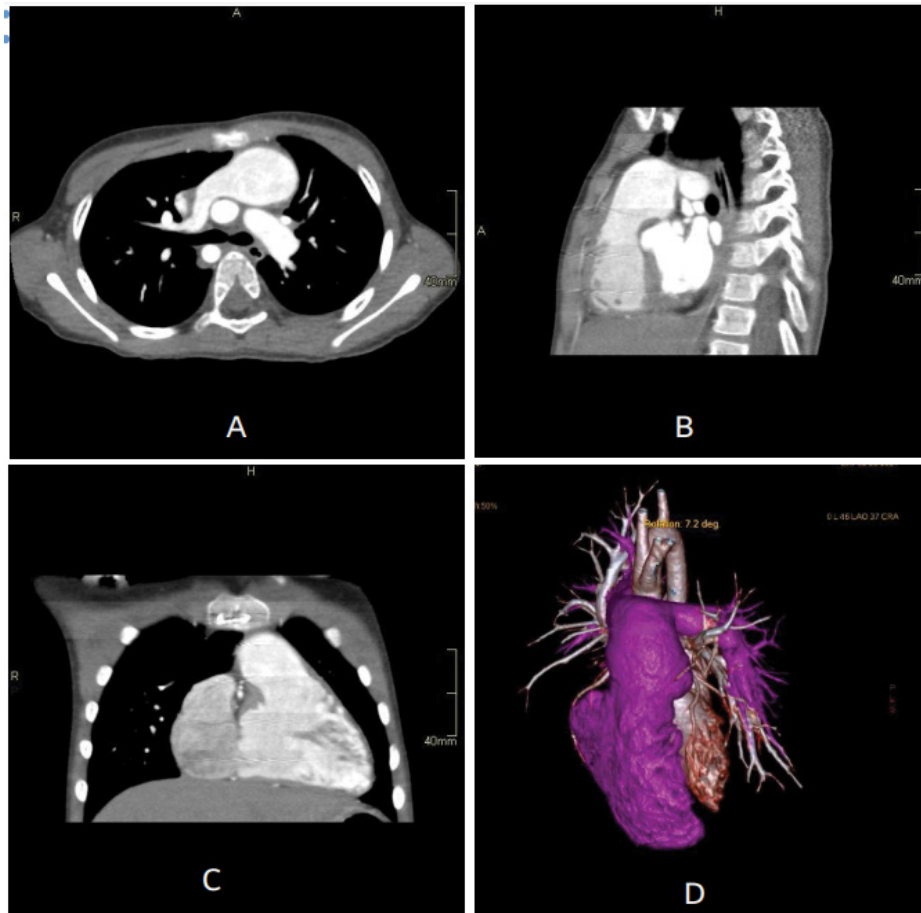


Figure 1: 4 years old child chest CT angiogram images (A) axial (C) coronal, (B) sagittal and (D) 3D reconstruction.

### Clinical Image

Congenital causes have been recognized as the major reason for PAA formation. More than 50% of all cases were associated with congenital heart disease (Figure 1). Congenital causes of pulmonary artery aneurysms and pseudoaneurysms include deficiency of the vessel wall, valvular and post valvular stenosis, and increased flow due to left to right shunts. Increased haemodynamic shear stresses and increased flow due to congenital heart disease can result in giant pulmonary artery aneurysms. The 3 most frequent congenital heart defects associated with a PAA are in decreasing order, persistent ductus arteriosus, ventricular septal defects, and atrial septal defects.

The aortic valve has also been identified as a major congenital cause of PAA formation. In fact, the fourth and fifth most frequent causes of PAA formation are a hypoplastic aortic valve and a bicuspid aortic valve, respectively. Pulmonary valve stenosis, including postvalvular stenosis, has frequently been described as an isolated cause of PAA formation. In fact, early pulmonary valve commissurotomy in the patient's history may precipitate aneurysm formation because of an eccentric right ventricular outflow jet, which may lead to weakening of the vascular wall. Furthermore, 1 case report described PAA formation in a patient presenting with the Noonan syndrome, a relatively common autosomal-dominant congenital disorder that is also associated with pulmonary stenosis [1,2].

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Many patients with PAA also present with pulmonary valve regurgitation, and even though it is more plausible that it is a consequence of annulus dilatation by the PAA, it may also be an independent etiologic factor in the formation of a PAA. Patients with the congenital malformation of an absent pulmonary valve syndrome represent a PAA subset that might link pulmonary valve regurgitation to PAA formation. Absent pulmonary valve syndrome has been described as a rare variant of tetralogy of fallow but has also been associated with ventricular septal defects and Uhl anomaly and very seldom occurs as an isolated congenital heart defect. In fact, early mortality in patients with absent pulmonary valve syndrome is high as a result of significant PAA formation and bronchi compression.

### Diagnosis

During a consultation, a systolic heart sound is generally present and might be combined with a diastolic murmur.

- The ECG shows signs of right ventricular or right atrial hypertrophy.
- In a standard x-ray PAA may appear as a hilar enlargement, a lung nodule, or a pulmonary mass. Some x-rays illustrate an aneurysmal main PA segment or dilatation of the PA.
- Transthoracic or transesophageal echocardiography is an important tool to evaluate heart function and valvular function to reveal shunts and may show the presence of a PAA.
- A bronchoscopy may show compression of the bronchus.
- Angiography allows delineation of the PAA within the pulmonary vasculature, involvement of the vascular structure, and assessment of the right-side hemodynamic pressure. However, an angiography can visualize only the patent lumen of the PAA, and it is invasive.
- In general, contrast-enhanced computed tomography confirms the diagnosis and provides useful information on size, number, location, and extent of the PAA. Furthermore, magnetic resonance imaging or 4-dimensional magnetic resonance imaging may show arterial wall thickening, provide information on blood flow, and characterize aortic and pulmonary hemodynamics without any radiation exposure.

### Declaration of Interests

The authors declare that they have no competing interests.

### References

1. Kreibich M, Siepe M, Kroll J, et al. Aneurysms of the pulmonary artery. *CIRC J*. 2015;131: 310-316.
2. <https://radiopaedia.org/cases/main-pulmonary-artery-aneurysm>