ACTA RADIOLÓGICA PORTUGUESA
January-April 2019 | Vol 31 nº 3 | 13-14

ARP Case Report Nº 15: Pulmonary Atresia with Ventricular Septal Defect and Major Aortopulmonary Collateral Arteries

Caso Clínico ARP Nº15: Atrésia da Válvula Pulmonar com Defeito do Septo Interventricular e Macroartérias Colaterais Aortopulmonares

Nuno Pereira da Silva, Paulo Donato

Imaging Department, University Centre Hospitals of Coimbra. Faculty of Medicine, University of Coimbra, Portugal

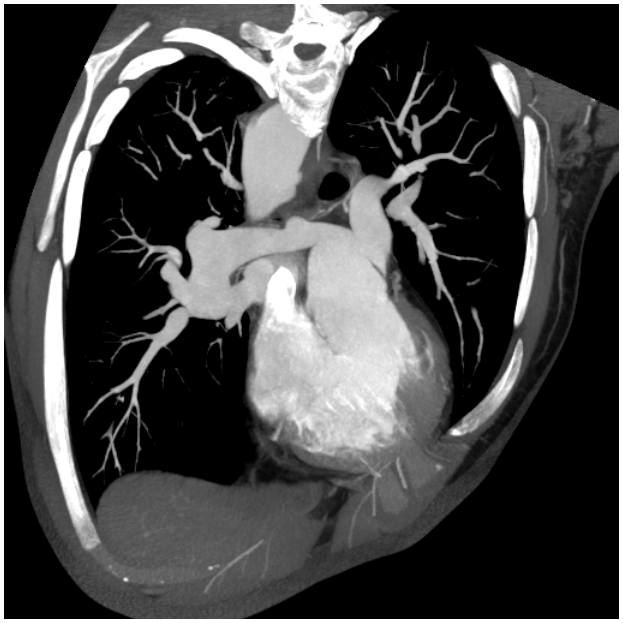
Presentation of the case

A 21-year-old male is admitted to our hospital's emergency ward with cyanosis. Laboratory routines revealed severe hypoxia. The patient had an history of congenital heart defect. Computer tomography angiography with ECG gating was performed.

The exam revealed right aortic arch, an interventricular communication associated with an "overriding" aorta and increased trabeculation of the right ventricular cavity.

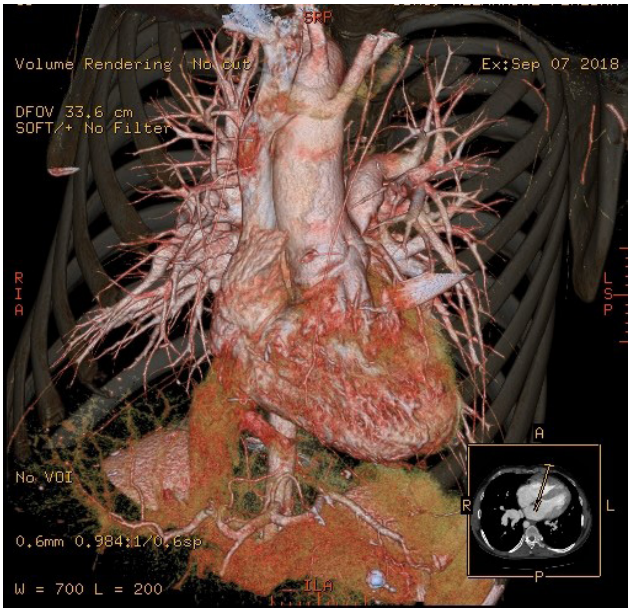
Three major aortopulmonary collateral arteries (MAPCA) can be seen branching from the descending aorta. The pulmonary arteries are hypoplastic and have no visible connection to the right ventricle, arising from the largest MAPCA which completely supplies them.

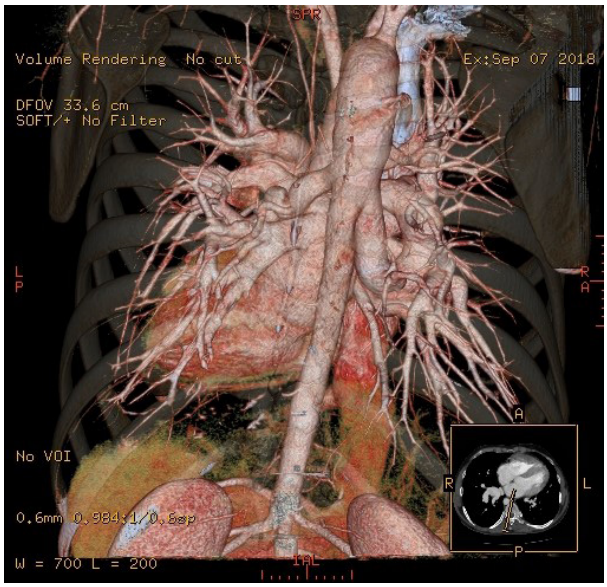
The findings are consistent with Pulmonary Atresia with Ventricular Septal Defect and Major Aortopulmonary Collateral Arteries (PA-VSD-MAPCAs).



Discussion

PA-VSD-MAPCAs is a rare complex congenital heart defect, the most severe form of tetralogy of Fallot. In this condition, the normal connection between the right ventricle and the pulmonary arteries is absent and a ventricular sept defect is apparent. Central pulmonary arteries may be hypoplastic or even non-existing and, if present, tend to be confluent. Consequently, patients develop aortopulmonary collaterals in utero to compensate for a lack normal pulmonary blood flow. The anatomical layout and dimension of the collateral vessels is extremely diverse and is the reason for the variety in clinical progressions and post-surgical outcomes. Aneurysmal dilatation of MAPCAs may be visible, especially in cases of long duration of the disease.





Cardiac catheterization remains the diagnostic gold standard thanks to the vast and accurate hemodynamic data provided in the procedure. However, MR and CT angiography can accurately identify the pulmonary blood flow with similar results. CT is especially elucidative in the characterization of MAPCA's, of paramount importance as the surgical goal is often to eliminate the extracardiac sources of pulmonary arterial blood flow by combining the MAPCAs into a confluent pulmonary artery, thus creating a complete pulmonary arterial system. This process is called "unifocalization".

Mortality rates are high without surgical intervention, up to 60% in the first year, less than 50% survival in 10 years, and less than 20% in 20 years.

Bibliography

Presnell LB, et al. 2015. An overview of pulmonary atresia and major aortopulmonary collateral arteries. World Journal for Pediatric and Congenital Heart Surgery. 2015;6:630-9.

Rajeshkannan R, et al. Role of 64-MDCT in evaluation of pulmonary atresia with ventricular septal defect. American Journal of Roentgenology. 2010;194:110-8.