AORTOPULMONARY WINDOW: A RARE CONGENITAL HEART DISEASE

T.J. Costa[1], N. Damry[1], C. Jacquemart [2], C. Christophe[1]

We report the case of a 3,5 month-old infant presented at cardiology consultation of our institution for investigation of a heart murmur associated with failure to thrive and respiratory distress. Echocardiogram showed dilatation of left heart cavities, patent foramen ovale, small muscular ventricular septal defect and pulmonary hypertension. Angio-CT was performed on a 64-slice CT to better characterize the congenital heart disease. Aortopulmonary window was diagnosed and surgically corrected. Patient evolution was good with discharge 10 days after surgery. The authors review the literature and stress the importance of angio-CT in pre-operative evaluation.

Key-word: Infants, cardiovascular system.

Introduction

A 3.5-month-old infant presented at cardiology consultation for exploration of a heart murmur and failure to thrive. The patient was examined the first time when he was 3-month-old and presented with dyspnea and tachypnea with infra-costal retraction. Physical examination in our institution revealed a clinical picture that included poor general health, a systolic-diastolic murmur 4/6 and respiratory distress. ECG showed sinus rhythm and signs of biventricular hypertrophy. Echocardiogram detected dilatation of left heart cavities with left ventricle hyper-contractility, a patent foramen ovale, a small muscular ventricular septal defect and pulmonary hypertension.

An angio-CT was performed for pre-operative evaluation of congenital heart disease. The exam was realized in a 64-slice CT with a non ECG-synchronized spiral acquisition (tube voltage: 80 kV; tube current: 63 mAs), after manual non-ionic contrast injection of 2-ml/kg-body weight (IOMERON 400).

MIP reconstructions images were obtained in the axial (Fig. 1A, B, C) and coronal (D, E) planes, showing a large communication (window) between ascending aorta and pulmonary artery in proximity to the semilunar valves. VRT reconstruction image (F) shows the very anterior position of the defect between the arterial trunks. Based on the radiological findings, the diagnosis of an aortopulmonary window type I was made. The large defect was localized anteriorly in the proximal part of the aortopulmonary septum in the presence of two independent semilunar valves. Coronary arteries origin was
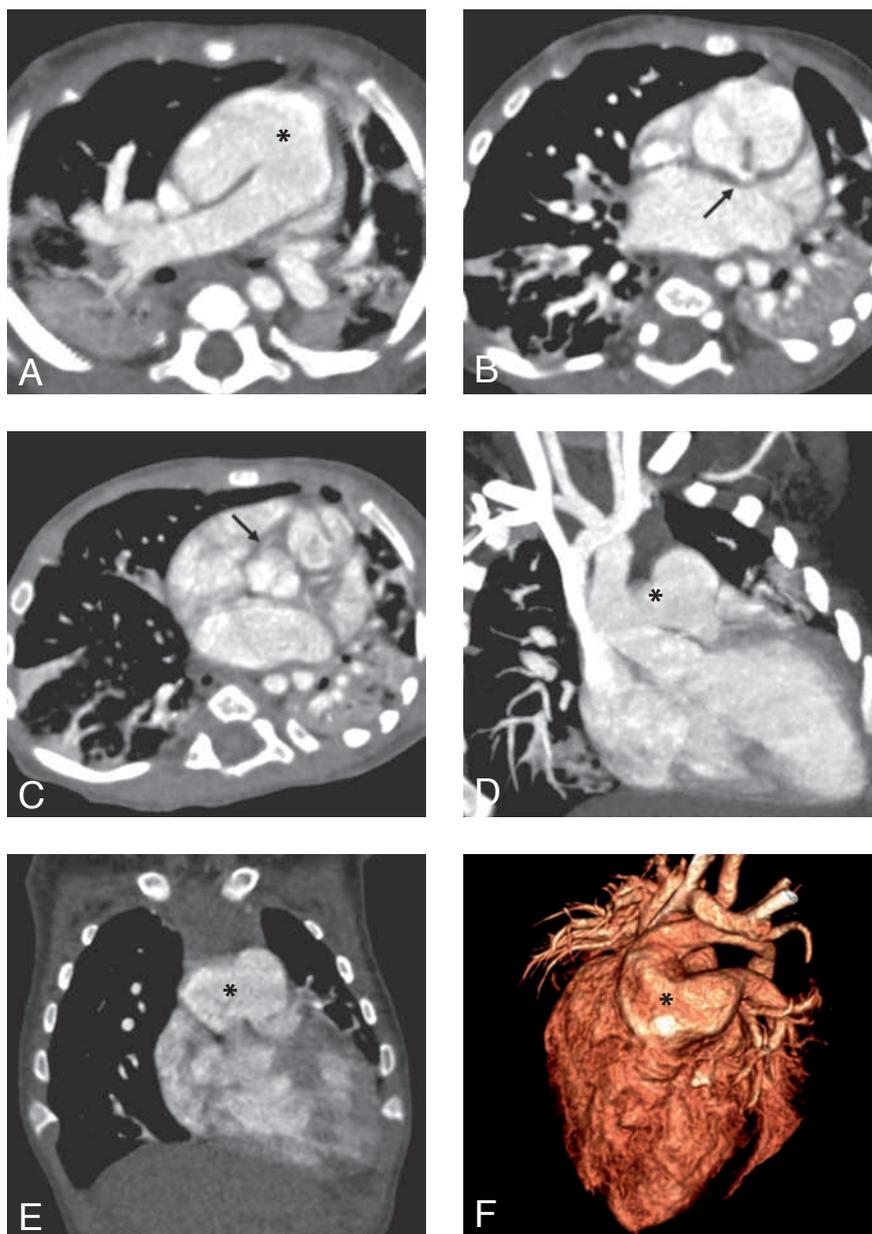
Fig. 1. — MIP reconstruction images in axial (A, B, C) and coronal planes (D, E) and VRT reconstruction image (F). In this case, the aortopulmonary window defect occurs in an anterior and proximal position (*). Coronary arteries origin is normal. B: left coronary artery (arrow), C: right coronary artery (arrow).

From: 1. Department of Radiology, 2. Department of Cardiology, Hôpital Universitaire Des Enfants Reine Fabiola, Brussels, Belgium.
Address for correspondence: Dr T.J. Costa, M.D., Avenue Ilya Prigogine, 15/10B, 1180 Brussels, Belgium. E-mail: joanaalvescosta@gmail.com

normal, as showed in images B and C. Vascular pulmonary accentuation (not show in images) was also present and interpreted as signs of pulmonary arterial hypertension.

Discussion

Aortopulmonary window was first described by Eliotson in 1830, as a communication between the ascending aorta and the pulmonary trunk in the presence of separated aortic and pulmonary valves. Robert E. Gross, MD, at Boston Children's Hospital, reported the first successful surgical repair in 1952 (1).

It is an uncommon entity, representing approximately 0.1% of all congenital heart diseases (2).

It may occur as an isolated lesion and/or associated with minor and simple repair anomalies (patent ductus arteriosus, atrial septal defect, patent foramen ovale) or as part of a larger complex of lesions such as tetralogy of Fallot, pulmonary atresia, aortic arch interruption, aortic arch hypoplasia with or without coarctation, and anomalous origin of coronary arteries. Fifty per cent of patients usually have no other heart defects.

The defect between the great vessels results from failure of the two embryonic conotruncal ridges to fuse. It is separate from *truncus arteriosus* because it occurs in the presence of essentially normal aortic and pulmonary valves. The "window" usually begins just above the sinuses of Valsalva and then extends into a variable distance distally into the arch (3).

Aortopulmonary window can be classified into 3 types, according to Mori's classification: type I (proximal) defects occur in the proximal part of aortopulmonary septum; type II (distal) defects occur in the distal part of the aortopulmonary

septum adjacent to the right pulmonary artery; type III (total) defect is a combination of types I and II , involving the entire length of the pulmonary trunk, from immediately above the semilunar valves to the level of pulmonary bifurcation and the proximal portion of the right pulmonary artery (4). Based on the recent utilization of transcatheter devices to close the defect, Ho et al. (5), added and additional type to this classification – type IV for intermediate defect with adequate superior and inferior rims.

Aortopulmonary window produces a large and usually unrestricted left-to-right shunt that worsens as pulmonary vascular resistance falls during the newborn period. Congestive heart failure and low cardiac output can rapidly follow. These patients are particularly susceptible to Eisenmenger's syndrome at an early age because of combined systolic and diastolic run-off into the pulmonary circulation.

Besides clinical suspicion of congenital heart disease and frequently ECG findings of biventricular hypertrophy, echocardiography can suspect or do the diagnosis by showing the "T sign" in the presence of two normal semilunar valves, downward flow through the right side of main pulmonary artery and diastolic flow reversal in both aortic arch and descending aorta.

However angio-CT gives additional anatomic information such as the precise location of the defect as well as associated coronary origin anomalies. In our case DLP was 24 mGy. cm, CTDI vol. 1.30 mGy and calculated effective dose was 2.0 mSv.

The patient was surgically treated and post-surgical evolution was good with just a transitory episode of myocardial dysfunction at Intensive Care Unit treated by dobutamine. Pulmonary hypertension episodes

were prevented by sildenafil administration during early hospitalisation.

Conclusion

This case shows that angio-CT is a valuable additional imaging method in the evaluation of this congenital heart disease, allowing a clear visualization of the defect location as well as coronary arteries origin. Indeed it enables a better planning of surgical approach.

The patient was surgically treated and could be discharged 10 days after surgery with no signs of pulmonary hypertension.

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