Case 1/2009 - 21-Month-Old Infant with Partial Atrioventricular Septal Defect, Consisting of Atrial Septal Defect and Tricuspid Insufficiency

Edmar Atik
Hospital Sírio-Libanês, São Paulo, SP - Brazil

Clinical data
Tiredness remained unaltered since birth, as well as failure to thrive. The child used anti-congestive drugs, such as digoxin, furosemide and spironolactone since 40 days of life. At the physical examination, in addition to Down’s syndrome signs, the infant was eupneic, exhibited normal skin color and peripheral pulses were normal. Weight was 8 Kg; BP: 95/60 mm Hg; HR: 130 bpm. The aorta was not palpable at the sternal notch. There were no precordial impulses and the ictus cordis was not palpable. The cardiac sounds were normal and the second heart sound was constant, with its two components of equal intensity. There was no heart murmur at the auscultation. The liver was not palpable.

The electrocardiogram (Fig.1) showed signs of right ventricular diastolic overload with polyphasic QRS complexes (rSR’s) in V1. AP:+60°, AQRS:+90°, AT:-90°.

Radiographic image
Showed a slightly increased cardiac area at the expense of the right atrium, corresponding to a more prominent right lower arch and slightly increased pulmonary vascular net. The middle arch was bulging (Fig. 1).

Diagnostic impression
This image is compatible with atrial septal defect (ASD) acyanogenic cardiopathy.

Differential diagnosis
Considering the unexpected increase in the right atrium, the possibility of tricuspid insufficiency (TI) in association with atrial septal defect must be recalled. In this situation, an atrioventricular septal defect (AVSD) is the most probable one.

Diagnostic confirmation
The clinical elements lead to the diagnosis of atrial septal defect (ASD) acyanogenic cardiopathy. However, in the presence of Down’s syndrome, and even without left anterior hemiblock, the diagnosis of atrioventricular septal defect cannot be ruled out. An echocardiogram showed the presence of an ostium primum ASD with 11 mm of diameter and moderate tricuspid insufficiency, in a partial atrioventricular septal defect.

Management
During a surgical intervention, the 12-mm diameter ostium primum atrial septal defect was closed with a bovine pericardium graft, together with the tricuspid valve plastic surgery, with approximation of the anterior and posterior valve at the groove, as well as suture of the anterior mitral valve sulcus. There was no interventricular communication and the small ductus arteriosus was ligated.

The immediate postoperative evolution was good, with the cardiac area returning to normal (Fig. 2), without audible murmurs and no specific medication.

Comments
The two defects that were repaired, the ASD and TI, are more rarely found within the context of atrioventricular septal defect. In general, the ASD is associated with mitral valve failure, as more common abnormalities in partial AVSD.

The diagnosis of AVSD becomes more difficult in the absence of left anterior hemiblock, even in the presence of Down’s syndrome. Hence, the warning for this diagnostic possibility is made in such situations.
Clinicoradiological Session

**Figure 1** - Preoperative chest X-ray and ECG show elements compatible with the diagnosis of atrial septal defect-type cardiopathy with an enlarged right atrium and LV diastolic overload.

**Figure 2** - Chest X-ray on the 3rd postoperative day shows a decrease in the right lower arch due to right atrium regression.