

Ebstein's disease: the level of right ventricular dysfunction and the surgical approach

Doença de Ebstein: o grau de disfunção do ventrículo direito e a conduta cirúrgica

Alexandra Regina SISCAR¹, Marcelo Felipe KOZAK¹, Ulisses Alexandre CROTI¹, Domingo Marcolino BRAILE¹

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CLINICAL DATA

Full-term female child, 2.8 kg born by c-section. First-time mother denied use of medications during pregnancy. The hospital discharge occurred two days after birth, with the child breastfeeding well and without signs or symptoms of congenital heart disease.

At 6 months of age, the child was referred to our Service with a history of fatigue while breastfeeding, abundant sweating and tachycardia even at rest.

With the diagnosis confirmed, therapy was initiated with diuretic and six-month returns were scheduled. With 3 years of life the child began to complain of mild fatigue on efforts and cyanosis with peripheral saturation of 80% in ambient air.

On physical examination the patient was in good general condition, cyanotic

+/4+, normotensive and with appropriate weight gain. The *ictus cordis* was normopositioned, normal heart sound and no murmurs. Abdomen normal. Well perfused extremities, pulses present and symmetrical.

ELECTROCARDIOGRAM AND HOLTER

Sinus rhythm with first-degree AV block, heart rate of

115 beats/min. SÂQRS between +90° and +180°, PR 200 ms, QRS 80 ms. Right axis deviation with right atrial overload (Figure 1).

Holter ratified the electrocardiographic findings and showed presence of isolated supraventricular ectopy.

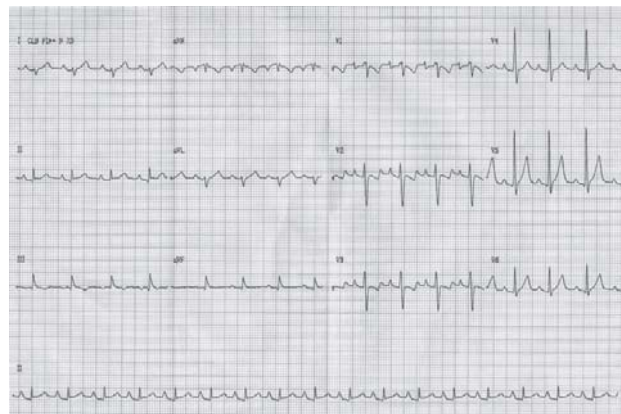


Fig. 1 – Electrocardiogram with prolonged PR interval and right atrial overload frequently common in Ebstein's disease

1. Pediatric Cardiovascular Surgery Service of São José do Rio Preto – Hospital de Base – São José do Rio Preto Medical School – São José do Rio Preto, SP, Brasil.

Correspondence address: Ulisses Alexandre Croti
Hospital de Base – FAMERP – Avenida Brigadeiro Faria Lima, 5544
– São José do Rio Preto, SP, Brazil – CEP 15090-000.
E-mail: uacroti@uol.com.br

RADIOGRAM

Visceral *situs solitus* in levocardia. Increased cardiac area with prevalence of right chambers and a cardiothoracic index of 0.60. Normal pulmonary vascular network (Figure 2).

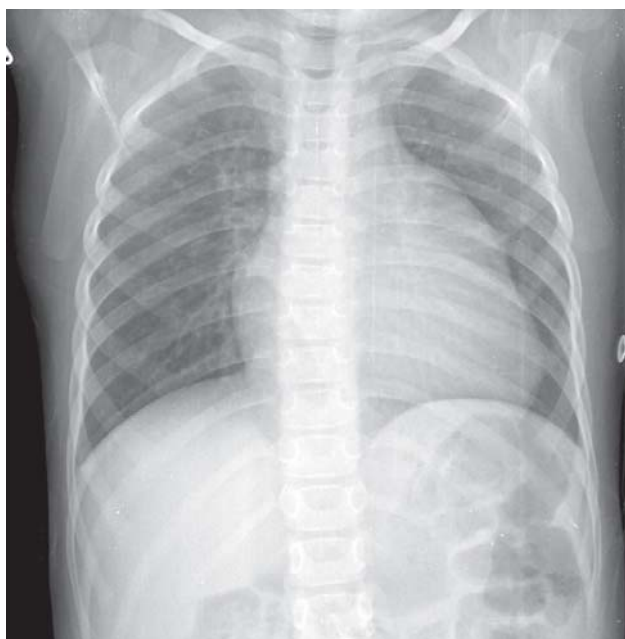


Fig. 2 – Preoperative chest radiography with cardiomegaly through the right cavities

ECHOCARDIOGRAM

Situs solitus in levocardia. Ebstein's disease with moderate-to-severe tricuspid valve insufficiency, large *ostium secundum* interatrial communication (IAC) with 11 mm and systolic dysfunction of the right ventricle (RV). The tricuspid valve ring measured 32 mm, there was significant adhesion of the septal leaflet and the RV presented with more than 50% of its cavity atrialized (Celemajer index of 1.5). At Doppler, the flow through IAC was bidirectional, the systolic pressure in RV was of 24 mmHg and the indexes of diastolic function were normal.

DIAGNOSIS

Since the beginning, echocardiography established the diagnosis of Ebstein's disease and detailed anatomical aspects. Then, it allowed the strict clinical monitoring. The balance of the variables: tricuspid valve insufficiency, size of interatrial communication and the RV contractility were responsible for the development and severity of the hypoxemic presentation [1].

With unsuccessful clinical treatment for decreasing the signs of hypoxemia and systemic venous congestion,

surgical treatment was indicated before the total deterioration of the RV function, since the patient was in functional class III (NYHA) and therefore at risk of polycythemia and events of paradoxical embolism [1,2].

The operation should primarily aim at preserving the tricuspid valve through valvuloplasty, avoiding prosthesis implantation. However, the greater concern was the significant degree of RV dysfunction, which might require an additional bidirectional cavopulmonary superior shunt (bidirectional Glenn procedure) [1,2].

OPERATION

Sternotomy, administration of heparin, introduction of cannulas in the aorta and vena cava, the aid of cardiopulmonary bypass (CPB) with moderate hypothermia at 28° C. Aortic clamping, antegrade blood cardioplegia, hypothermic at 4°C and intermittently every 20 minutes.

After opening of the right atrium, significant dysplasia of the tricuspid valve was found with large portion adhered to the RV, thickening and fusions compromising mainly the anterior leaflet.

Valvuloplasty using the Carpentier technique was initiated, with longitudinal plication of the RV wall, desinsertion of the anterior leaflet and part of the posterior leaflet, together with the tricuspid annulus, clockwise rotation and reinsertion [3], but without success in competence test with saline solution after several attempts of adjustment.

Thus, valve replacement was chosen by resecting the dysplastic valve and moving the points in the atrium so that the coronary sinus remained positioned below the ring of the prosthesis implanted, an unusual situation in a patient without Ebstein's disease and who requires tricuspid valve replacement.

The need for prosthesis raised the doubt about which one would be better, biological or mechanical, a controversial fact, since the degeneration of bioprostheses in tricuspid position seems to occur more slowly in relation to the prosthesis placed in left cavities. However, the calcification results in dysfunction and need for mid-term replacements, especially in children of lower age. Furthermore, the low pressure in the right cavities predispose to the formation of thrombi by mechanical prostheses [4]. Despite these considerations, it was chosen to implant a bivalvular, modern and low profile No. 31 mechanical prosthesis.

The *ostium secundum* IAC was closed with bovine pericardial patch, despite the great concern about the RV functionality, which might or not require the bidirectional Glenn procedure.

The disconnection of CPB was performed using inotropics, after 110 minutes of assistance from CPB and 86 minutes of myocardial ischemia. The patient stayed in

intensive care unit for seven days and, after this period, she evolved clinically well with mild pericardial effusion and moderate pleural effusion, which were reversed through the introduction of oral corticosteroids and respiratory therapy. She was discharged from hospital on the 13th

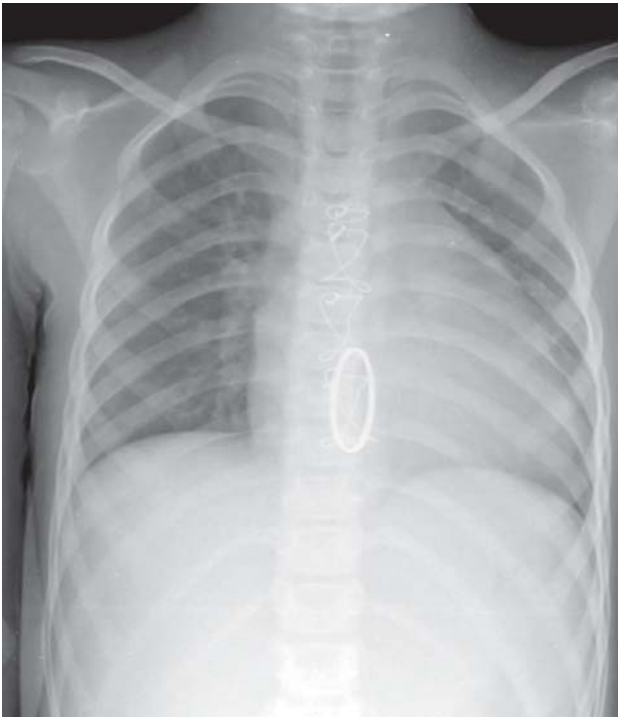


Fig. 3 – Postoperative chest radiography with decreased cardiac area and mechanical prosthesis in tricuspid position

postoperative day, under use of digital, diuretics and warfarin sodium.

The patient was followed-up in the outpatient clinics for 3 months and is currently acyanotic, with good weight gain, no complaint of fatigue, under control of INR, with decreased cardiac area on chest radiography (Figure 3) and the echocardiogram showed mild right ventricular dysfunction.

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