Ross procedure for severe congenital aortic regurgitation in a three-month-old patient

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We here describe the case of an infant with severe aortic regurgitation due to a congenital dysplastic aortic valve. Surgical treatment (autograft replacement of the aortic valve) was carried out when the patient was 3 months old. The technical aspects and the results of instrumental examination during follow-up are discussed.

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Isolated congenital aortic regurgitation due to a malformation of the valve leaflets is a rare condition. The timing of surgical treatment depends on the severity of the defect and on its hemodynamic and clinical consequences; furthermore, other implications influencing the surgical decision need to be considered when adopting a valve replacement procedure.

We here present a case in which 3-month monitoring of severe aortic regurgitation revealed progressive dilation and dysfunction of the left ventricle despite full medical treatment (digoxin, frusemide and captopril). Surgical replacement of the aortic root with a pulmonary autograft was followed by a prompt improvement in the patient's clinical conditions and left ventricular (LV) function.

Case report

A 7-day-old female patient was first referred to our Center with a diagnosis of suspected aortic insufficiency. She looked pink and healthy (weight 3.8 kg), and showed no clinical signs of heart failure. A grade 3/6 diastolic murmur was audible along the left sternal border, whereas the peripheral pulses were strong. Echocardiography revealed severe aortic regurgitation due to aplasia of the right coronary cusp, and dysplasia of the other two cusps (Fig. 1); the left ventricle was dilated but with a normal contractility. The patient was then scheduled for fortnightly clinical and echocardiographic control examinations. After 1 month, digoxin, frusemide and captopril were started because of the appearance of tachypnea and hepatomegaly, and the progressive LV dilation was found to be associated with mild-to-moderate mitral valve incompetence. After an initial positive response to medical treatment, the infant stopped growing and was hospitalized for further investigations. At the time of admission, she was 3 months old (weight 5.4 kg). Clinical examination showed polypnea, hepatomegaly, bounding pulses, an apical thrust over the precordium, and a grade 3/6 systolic and a grade 4/6 diastolic murmur along the left sternal border. The cardiothoracic ratio calculated on a chest X-ray was 0.7. Echocardiography revealed massive aortic regurgitation, marked dilation of the left ventricle (an end-diastolic diameter of 31 mm and an end-diastolic volume of 37 ml, corresponding to 122 ml/m^2), an impaired systolic function (LV ejection fraction 45%) and grade 2 mitral valve insufficiency. The morphology of the aortic valve was determined and was suggestive of a slight tendency of the dysplastic non-coronary cusp to prolapse; the coronary ostia were normally positioned; analysis of the pulmonary trunk showed a normally functioning tricuspid valve whose annular dimensions matched those of the aorta. Urgent surgical correction was therefore indicated.

After median sternotomy, the ascending aorta and both venae cavae were cannulated, and a cardiopulmonary bypass was instituted. During systemic cooling to

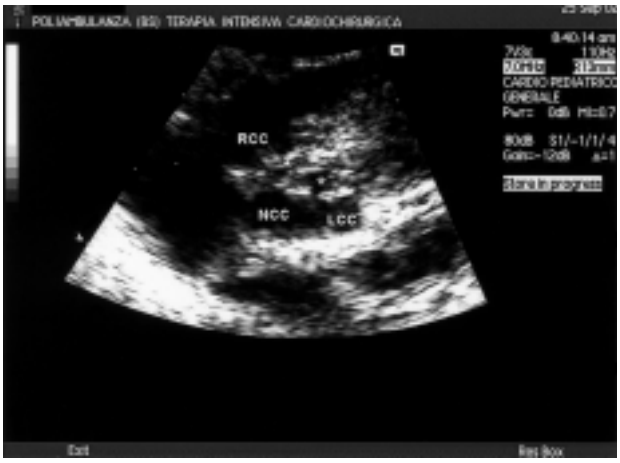

Figure 1. *Parasternal short-axis view of the aortic valve during diastole. Note the wide area of non-coaptation (*) due to the absence of the right coronary cusp (RCC)* (arrow). *LCC = left coronary cusp; NCC = non-coronary cusp.*

24°C, the left ventricle was vented through the atrial septum and a retrograde cardioplegia catheter was inserted into the coronary sinus. The aorta was cross-clamped, cold (4°C) retrograde blood cardioplegia was administered every 20 min, and the ascending aorta was transected just distally to the sino-tubular junction. Inspection of the aortic valve revealed thick and prolapsing left and non-coronary cusps, and the absence of the right coronary cusp. A very thin rim of myxoid tissue was found at the proto-site of the insertion of the right cusp. The pulmonary autograft was then harvested and implanted as a standard root replacement. The right ventricular-to-pulmonary connection was re-established by means of a posterior direct anastomosis and an anterior outflow patch of bovine pericardium containing a 0.1 mm polytetrafluoroethylene (PTFE) monocusp valve. After 159 min of cross-clamping and 202 min of cardiopulmonary bypass, the child was easily weaned from the bypass without the need of inotropic support. Transesophageal echocardiography showed the absence of autograft regurgitation, negligible turbulence along the right ventricular outflow tract (RVOT) without any significant gradient, and a well functioning monocusp valve.

The postoperative course was uneventful; the child was extubated on day 5 and discharged home on day 14. The discharge echocardiogram showed a significantly reduced LV volume (an end-diastolic diameter of 27 mm and an end-diastolic volume of 27 ml, corresponding to 77 ml/m^2) with a LV ejection fraction of 56%. Frusemide and captopril were stopped 2 months following surgery. At the time of the last follow-up examination (12 months postoperatively), the patient was in good clinical conditions, well nourished and growing normally without medications. Despite the fact that she had doubled in weight, echocardiography confirmed that there was no autograft regurgitation and that the LV end-diastolic diameter and LV ejection fraction were 28 mm and 58% respectively. The right ventricle appeared to be normal in size and function, with no significant gradient across the RVOT. Echocardiography also revealed that the monocusp valve was still functioning well and the presence of mild pulmonary regurgitation.

Discussion

Dysplasia of the aortic valve, with the total absence of one cusp causing congenital aortic insufficiency, is a very rare pathological entity. The most frequently described morphology is the absence of the right coronary cusp[1-3] but a rudimentary non-coronary cusp has also been reported[4]. Despite early recognition of the disease, most of the patients described in the literature underwent surgical treatment in pediatric age, depending on the severity of the aortic regurgitation and the degree of LV impairment. The presence of two well-functioning leaflets may sometimes allow a conservative approach to the valve[4], but the myxomatous degeneration involving all the cusps described in almost all cases makes aortic valve replacement the only surgical option. This condition supports an attempt to delay surgery as long as possible, but sudden infant death related to congenital aortic insufficiency has also been described[5]. We have found only one case report in which aortic valve replacement associated with annular enlargement was carried out in a neonate because of the severity of the defect[6].

In our patient, the diffuse malformation of the aortic valve was clearly recognizable at the time of the first echocardiographic evaluation, thus suggesting the impossibility of surgical repair. A close follow-up was therefore started in order to monitor the evolution of the LV dilation and systolic dysfunction. The accelerated progression of the ventricular dysfunction despite medical treatment was the major determinant of early surgical correction. When dealing with a small baby, a pulmonary autograft is the preferred form of aortic valve replacement because of its growth potential, optimal hemodynamic performance, and freedom from anticoagulation and hemolysis[7]. Some concerns regard the higher risk of autograft failure in patients with a primary diagnosis of aortic insufficiency, which is probably related to a significant incidence of annular dilation[8]. However, this has not been clearly proved for patients with a normal-sized aortic annulus undergoing surgery in the first months of life.

There is still no convincing alternative to cryopreserved pulmonary homografts when addressing the problem of RVOT reconstruction, although a younger age has been found to be a significant risk factor for homograft reoperation[9]. This is especially true for small sized patients. In order to avoid the ear-

ly obstruction of the right ventricular-pulmonary conduit, we performed a direct posterior anastomosis between the pulmonary trunk and the infundibulum using an anterior patch containing a PTFE monocusp valve. This technical choice was based on experience with the use of a monocusp patch to correct RVOT obstruction, mainly in tetralogy of Fallot patients[10]. The rationale behind this choice for our patient was to allow a certain growth of the pulmonary-right ventricular junction, at least in the posterior aspect, with the further advantage of the presence of a competent pulmonary valve. An intraoperative transesophageal echocardiogram revealed an unobstructed RVOT and a competent monocusp valve. The hemodynamic recovery after cardiopulmonary bypass and the postoperative course were uneventful. The same echocardiographic results were found 12 months postoperatively, together with a normalized LV volume and ejection fraction. On the basis of published reports concerning the good short- and mid-term functions of PTFE monocusp valves for RVOT reconstruction[10], we are quite optimistic that the reoperation-free interval of our patient will be long. A longer follow-up is required before any definite conclusions can be drawn about the effectiveness of this approach.

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