Percutaneous Balloon Aortic Valvuloplasty in a Pregnant Adolescent

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We report the case of a 16-year-old pregnant patient with severe aortic stenosis and pulmonary congestion clinically uncontrolled, in whom percutaneous balloon aortic valvuloplasty was used as the first choice of treatment in an emergency procedure. The clinical findings, pathophysiology, diagnostic features, and indications for percutaneous treatment are reported.

Severe congenital aortic stenosis is rare in children and young individuals. Bicuspid aortic valve occurs in 3% to 6% of patients with congenital heart disease; when associated with commissural fusion, significant stenosis may be present in childhood. The association of severe congenital aortic stenosis and pregnancy is difficult to control clinically, carrying a high risk of maternal and fetal mortality, mainly when manifested with symptoms of pulmonary congestion [1,2].

Case Report

The patient is a 16-year-old pregnant white woman in the 27th gestational week, who was admitted with severe dyspnea and precordial pain. Her personal antecedent was a cardiac murmur without diagnosis or follow-up. When starting the prenatal follow-up 2 weeks before and complaining of dyspnea on exertion, the patient was referred for cardiac assessment. She evolved with symptoms at rest, being then hospitalized. She reported smoking and no familial history of heart disease, diabetes mellitus, hypertension, or dyslipidemia.

On physical examination, the patient was in regular general condition, tachypneic, acyanotic, and her facies was not characteristic. Her weight and height were, respectively, 40 kg and 1.50 m. Her blood pressure was 90/60 mmHg, and, on heart auscultation, she had a regular rhythm, tachycardia (heart rate=120 bpm), and an ejection systolic murmur (5+/6+) in the aortic region with irradiation to the carotid arteries. The pulmonary auscultation showed crepitant rales in both bases.

The electrocardiography revealed sinus rhythm, hypertrophy of the left chambers, and ventricular repolarization changes secondary to overload (fig. 1).

The chest radiography showed a normal cardiac area, cranial diversion of the pulmonary circulation and Kerley B lines compatible with pulmonary congestion.

On admission, the echocardiogram showed a bicuspid aortic valve with commissural fusion of the right and left coronary leaflets, moderately reduced systolic mobility, systolic dynamics in cupule, and no calcification. In addition, left ventricular (LV) concentric hypertrophy, left ventricle/aorta (LV/AO) gradient = 100 mmHg, and an aortic ring of 1.48 cm^2 were observed.

The patient remained at the coronary unit under clinical treatment with digitalis, a diuretic, nitroglycerine, and oxygen therapy with a mask.

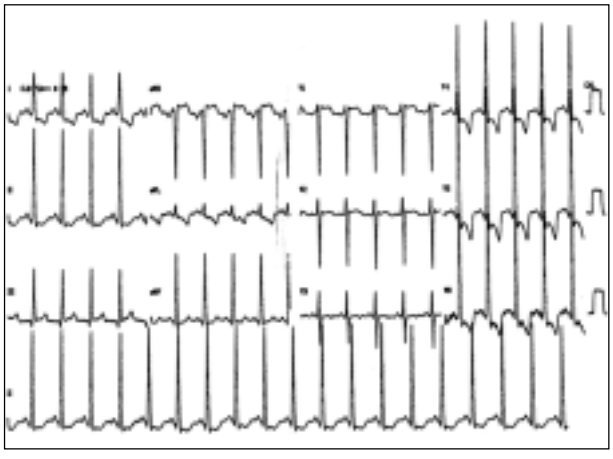

Fig. 1 - ECG at rest on hospital admission showing hypertrophy of the left chambers.

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On admission, the obstetric ultrasound showed the presence of heart beats and fetal movements, as well as a Grannun zero-degree placenta with normal thickness. The amniotic fluid volume was normal. The fetus weighed approximately 920 g and had no morphological peculiarities. The fetal biometry for the echographic gestational age of 27 weeks was average. A new ultrasound was performed every 48 hours for fetal follow-up.

In the 30th gestational week, the patient experienced progressive pulmonary congestion, which did not respond properly to clinical treatment. Percutaneous aortic valvuloplasty was then indicated.

On cardiac catheterization, the findings were as follows: bicuspid aortic valve with an LV/AO gradient = 105 mmHg (fig. 2), ascending aorta with normal angiographic appearance, coronary circulation with normal origin and trajectory, and hypertrophic left ventricle.

Procedure – The patient was sedated and the cardiofetal beats were monitored. Catheterization of the left ventricle was performed through puncture of the right femoral artery with an 8F Sones catheter. A 0.35 guidewire (Amplatz "extra-stiff" 260 cm – Boston Scientific) was positioned in the left ventricle. Aortography was performed in left anterior oblique projection and left ventriculography was performed in right anterior oblique projection (Multi-Track 5F angiographic catheter, NuMed Inc) with radiographic nonionic low-osmolarity contrast medium. An 18x4-cm balloon (Z-MED – NuMed) was used (fig. 3). At the end of the procedure, the peak-to-peak ejection gradient LV/AO was 20 mmHg, and mild-to-moderate aortic valve insufficiency was observed (fig. 4).

After the procedure, the electrocardiogram showed a reduction in the ventricular rate and maintenance of the signs of hypertrophy in the left chambers.

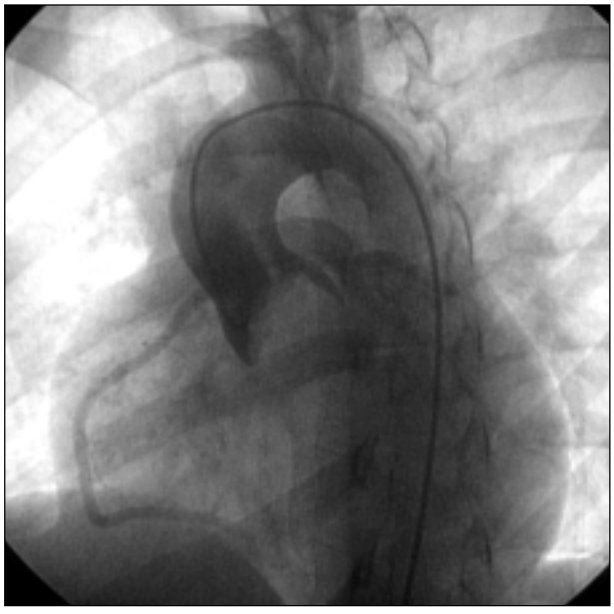

Fig. 2 - Aortogram in left anterior oblique projection. A bicuspid aortic valve with severe stenosis is seen (the decrease in opacity indicates flow through the stenotic valve).

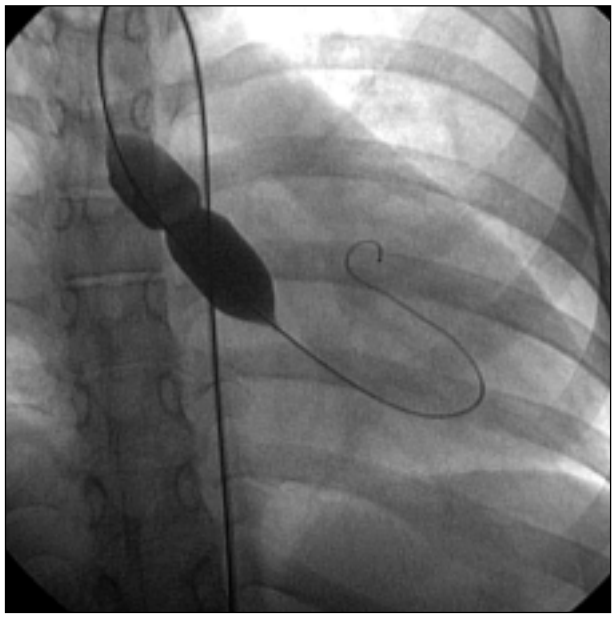

Fig. 3 - Aortogram in right anterior oblique projection. Balloon dilation of the aortic valve. A constriction corresponding to valvular stenosis may be seen.

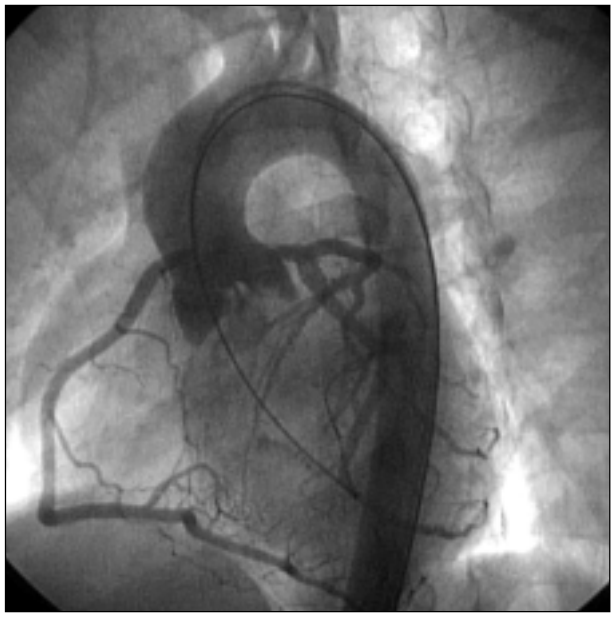

Fig. 4 - Aortogram in left anterior oblique projection. Final result showing mild aortic regurgitation and disappearance of the central jet.

The chest radiography showed a reduction in the signs of pulmonary congestion, and the echocardiogram showed a mild residual aortic stenosis (LV/AO gradient = 29 mmHg) and moderate aortic valvular insufficiency. The other parameters remained unchanged.

After the procedure, fetal monitoring with cardiotocography evidenced a reactive tracing. The obstetric echography and Doppler color velocimetry revealed good fetal vitality. On the 4th day after percutaneous aortic valvuloplasty, a new obstetric echography revealed severe oligohydramnios with an amniotic fluid index (AFI) = 2.5 cm (normal

AFI=8-18 cm). Due to the association of oligohydramnios and placental insufficiency, interruption of the pregnancy with a Cesarean delivery was indicated.

The male newborn infant weighed 1.425 g, had an Apgar score of 8 and 9, and a Capurro index of 32 weeks.

The mother was discharged from the hospital on the 4th day after the Cesarean delivery, in good general condition and without dyspnea. The diuretic and angiotensin-converting-enzyme inhibitor were maintained for the clinical control of heart failure.

The premature newborn had hyaline membrane disease, extensive bronchopneumonia, septicemia, disseminated intravascular coagulation, and anemia. In addition, as his ductus arteriosus was patent, indomethacin was used until its closure on the 10th day of life. The infant was discharged from the hospital on the 46th day of life weighing 2.125 g, in good general condition and stable.

On the clinical 9-month follow-up, the patient remained asymptomatic using an angiotensin-converting-enzyme inhibitor and a diuretic. Due to the good clinical evolution, the surgical valvular correction was postponed.

Discussion

Severe congenital aortic valve stenosis is rare in children and young individuals [1]. Bicuspid aortic valve accounts for 3% to 6% of congenital heart diseases [1,2]. It is more frequently found among males (male to female proportion of 4:1) and may be associated with cardiovascular anomalies in up to 20% of the cases [2]. The trauma produced by the turbulence of the blood flow leads to thickening, fibrosis, calcification, and stiffness of the valve, and the clinical manifestations occur from the 3rd decade of life onwards, mainly in the male sex [2,3]. In some cases, when the bicuspid valvular stenosis is associated with commissural fusion, it may already present itself as severe during childhood [3]. Some authors have considered aortic valve stenosis the most frequent congenital heart disease, because it is not usually detected during the first years of life. With the routine use of echocardiographic techniques, the recognition of aortic valve stenosis has been facilitated [2].

The physiological changes induced by gestation cause the following 3 major hemodynamic changes in the heart: an increase in the cardiac output (30-40%); an increase in heart rate (10-20 bpm); and expansion of the blood volume (20-100%) [4,5]. The association of these factors with obstruction of the left ventricular outflow tract, which limits the variations in cardiac output, may lead to hemodynamic decompensation, which is frequently portrayed as symptoms and signs of pulmonary congestion, syncope, and sudden death.

According to Arias and Pineda [6], in a series of 38 gestations with 23 severe aortic stenoses, the natural history of this disease encompasses a maternal mortality rate during pregnancy of 17.4% for nontreated patients and a fetal mortality rate of 34%. An invasive cardiac intervention may be necessary in patients whose clinical condition has deteriorated during pregnancy, aiming at reducing the peak-to-peak ejection gradient by 60% to 70% [7,8]. Surgical treatment for severe bicuspid aortic stenosis has been recommended by the Brazilian Consensus on Heart Disease and Pregnancy at any time during gestation when the LV/AO gradient is greater than 70 mmHg [9]. Cheitlin [10] recommends surgical balloon valvuloplasty or even surgical valve replacement in the presence of symptoms, immediately if evidence of pulmonary congestion exists, and when the valvular area is 0.7 cm^2 or lower, measured on the echocardiogram or during cardiac catheterization. Balloon valvuloplasty successfully used in severe mitral stenosis during pregnancy and in aortic stenoses in nonpregnant, but at high risk for aortic valve replacement [11,12], patients was used in 2 pregnant patients with aortic stenosis [13,14]. Both cases had favorable maternal and fetal outcomes. A series of valvuloplasties performed in the 30th gestational week [15] has been associated with preterm birth of healthy babies 2 weeks later. In 2 studies [16,17] with 11 patients undergoing aortic valve replacement, no maternal death was reported although the overall maternal surgical mortality rate with cardiopulmonary bypass was 1.5. On the other hand, the overall fetal surgical mortality ranged from 16% [17] to 20% [18]. Aortic valve replacement, in particular, seems to be associated with an exceptionally high fetal mortality rate of 40% in the entire group and 57% in patients with aortic stenosis [17]. Therefore, open surgery for aortic valve stenosis during pregnancy should be considered as the last choice.

Percutaneous balloon aortic valvuloplasty began to be used in the mid 1980s. In 1984, based on the initial experience with pulmonary valvuloplasty, Lababidi [19] and Lababidi and Wu [20] extended the method for the treatment of aortic stenosis. Currently, the refinement of materials provided the use of that less invasive method in situations in which the surgical risk is unacceptable.

Balloon valvuloplasty and surgical valvuloplasty have been limited by their late results, which turn these procedures into palliative treatment [21]. The immediate appearance of aortic regurgitation or its progression, and the later appearance of restenosis are the major complications of valvuloplasty. Other complications during the procedure include bleeding, arrhythmias, stroke, iliac-femoral arterial complications, injury to mitral valve, and death [7].

Exposure to X-rays during the procedure is another issue that deserves special attention, and for a short time it may be minimized by using radiological protection for the abdomen and pelvis of the pregnant female. This fully reduces the risks of congenital malformations [22].

The experience of percutaneous aortic valvuloplasty in pregnant women has been limited to case reports, because it has been performed in clinically unstable patients who are under high surgical maternal-fetal risk and require intervention. Percutaneous aortic valvuloplasty during gestation is a safe and effective procedure that reduces morbidity and mortality and provides opportunity for an elective surgical valvular correction.

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