ABSTRACT

Fixed subaortic stenosis, commonly associated with other congenital cardiac defects, is the cause of 10 per cent of cases of congenital obstruction of the left ventricular outflow. Corrective surgery is frequently a successful treatment, recommendations being based on the transaortic gradient in Europe while in the USA the most prevalent opinion is surgical repair independently of the gradient. We present a case of adult clinical onset of a fixed subaortic stenosis during pregnancy, in which hemodynamic changes are significant, that was medically treated and followed in the outpatient clinic of our hospital, and review the state of the art of the management and surgical indications of this condition.

BACKGROUND

Fixed subaortic stenosis accounts for 8 to 10 per cent of all cases of congenital obstruction of the left ventricular outflow. Thirteen per cent have familial presentation, and between 25% and 45% of association with other congenital cardiac defects have been reported.

The presence of a tunnel or ridge type obstruction (ridge) is the cause of almost 10% of all cases of fixed subaortic stenosis, the membranous form being the most frequent. In this, a diaphragm or ring structure occludes the outflow tract near the aortic valve. In other cases, a fibromuscular narrowing is the cause of the obstruction (shelf).

Surgical treatment of this condition is usually curative, its morbidity and mortality being low, although dependent on the associated defects. A recurrence of the stenosis has been found in 27% of cases within five years of surgery. The moment at which surgical treatment should be performed is the subject of controversy, especially in cases of mild to moderate obstruction.

We present the case of a 23-year-old woman who was diagnosed with subaortic stenosis in the course of a twins pregnancy: A Controversial Management.

CASO CLÍNICO

Subaortic Stenosis Diagnosed in the Course of a Twins Pregnancy: A Controversial Management [33]

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Key words

Subaortic stenosis; Surgery indication; Pregnancy

RESUMO

Estenose Aórtica Diagnóstica no Decurso de uma Gravidez Gemelar: Abordagem Controversa

A estenose subaórtica fixa, frequentemente associada a outras cardiopatias congênitas, representa cerca de 10% dos casos de obstrução congênita do tracto de saída do ventrículo esquerdo. A cirurgia correctiva é frequentemente um tratamento eficaz, sendo as recomendações para a sua realização na Europa baseadas no gradiente transaórtico, enquanto dos EUA a opinião mais prevalente é a de intervir sempre, independentemente do gradiente. Apresentamos um caso clínico duma estenose subaórtica fixa com manifestação clínica inicial na fase adulta, durante a gravidez, altura em que as alterações hemodinâmicas são significativas, tendo a doente sido tratada medicamente e seguida na consulta de ambulatório do nosso hospital. Faz-se uma revisão sobre a abordagem e indicações cirúrgicas desta situação clínica.

Palavras-chave

Estenose subaórtica fixa; Correcção cirúrgica; Gravidez
membranous stenosis in the 24th week of a twin pregnancy and review the management and surgical indications for the treatment of this condition.

CLINICAL CASE

A 23-year-old woman, with a “cardiac murmur” diagnosed in childhood but not studied, mother of a 2-year-old son whose pregnancy had been uneventful, and with no physical limitations in her daily activity, having been diagnosed with a twin pregnancy, was admitted to the Gynecology Department in the 24th week for a threatened premature delivery that receded with ritodrine, and was discharged home after the clinical event was controlled.

Twenty-four hours after discharge, she returned to the Emergency Department with severe oppressive central chest pain irradiating to the left arm diagnosed as typically anginous by the clinician responsible for her admission, together with slight dyspnea. Her blood pressure at admission was 120/70 mmHg. In the physical examination, a IV/VI systolic ejection murmur with no signs of cardiac failure was found. Chest X-ray, biochemistry and hematological analysis showed no relevant abnormalities. The electrocardiogram revealed sinus tachycardia at 125 bpm, with no signs of hypertrophy of the chambers and a slight depression of the ST segment in V4-V6.

The pain was treated with magnesium metamizol, and beta-blockers were prescribed (propranolol 40 mg/8h) to slow the tachycardia. After her angina was treated, the patient remained asymptomatic and was examined by the Echocardiography and the Cardiology Outpatients services. Echocardiography showed a non-dilated left ventricle with signs of wall hypertrophy (wall thickness 13.7 mm, LV mass 265) (Fig. 1) and a thin subaortic shelf membrane located 13 mm from the aortic valve (Figs. 2 and 3). The septal veil of the mitral valve was preserved. The aortic valve had no structural abnormalities, an output tract peak systolic instantaneous gradient of 55 mmHg being measured by Doppler. No aortic valve regurgitation or other associated abnormalities were found.

The patient was followed by the Cardiology Department during the subsequent months of pregnancy, with no new episodes of thoracic pain or other symptoms reported. Beta-blockers were not withdrawn after the diagnosis of aortic stenosis, on the basis of the predominant tachycardia of the patient, the mild gradient of the obstruction and the safety of this product during pregnancy, the treatment being changed to atenolol 50 mg/24 h for its safer pharmacological profile and more convenient posology. Two other echocardiograms were done during this period, with no increase in the aortic subvalvular gradient detected.

In the 36th week of pregnancy a cesarean section with intradural anesthesia was performed because of metrorrhagia, no complications being reported during the procedure.

After the delivery, the woman was discharged with atenolol 50 mg/24h as the only cardiological medication. After the first year of follow-up she has not complained of any kind of symptoms and an echocardiogram showed a decrease in the gradient to 35 mmHg measur-

Fig. 1 A non dilated LV with signs of moderate hypertrophy is shown. Measures: Wall thickness 13.7 mm, LV mass 265.
ed by Doppler. Our management of the patient at present is limited to the performance of periodic echocardiographic checks, since the patient refused the option of surgical repair.

DISCUSSION

Congenital subaortic stenosis makes up 8 to 10% of all cases of aortic stenosis. Its prevalence is higher in males (2:1)\textsuperscript{9}. Obstruction is usually absent in the early stages of life and becomes more evident in late childhood and young adulthood. It is uncommon to detect the disease in people older than 30, which suggests that survival after this age without surgical treatment is exceptional\textsuperscript{10}. Several reports show that its progression is faster than aortic stenosis\textsuperscript{3}. The aortic regurgitation that is usually associated is probably caused by valve thickening due to subvalvular turbulence, which may also be the predisposing factor for endocarditis even after the excision of the membrane. On the basis of the progression of obstruction and aortic regurgitation, even a low grade of obstruction is an indication for elective surgery\textsuperscript{3, 4}.

The classical recommendations for the surgical repair of discrete subaortic stenosis depend on the angiographic visualization of a subvalvular membrane or diaphragm and a pressure gradient higher than 40 mmHg\textsuperscript{1, 3}. These recommendations are based on the potential progression of the condition (scarcely tested in these studies) and the intention of preserving the valve from damage caused by the high-pressure flow. The Guidelines of some cardiological associations, including the SEC (Sociedad Española de Cardiología), are for surgical repair in the above circumstances\textsuperscript{11}. Recent relevant studies also support this option\textsuperscript{12}, given that patients with a low gradient rarely have significant progression of the obstruction or develop aortic valvular regurgitation.

These standards, by contrast, go against the opinion of researchers at UCLA\textsuperscript{4}, who consider early surgery necessary independently of the gradient when diagnosed, reporting in a case-control study that surgical resection of fixed subaortic stenosis before the development of a significant (>40 mmHg) gradient might prevent recurrence, reoperation and secondary progressive aortic valve disease.

The case of our patient, aside from the singularity of onset, shows the need for a dynamic and thorough consideration of the individual circumstances, because in our case the hemodynamic changes induced by the twin pregnancy were the cause of the changes observed in the gradient. The absence of progression of gradient and symptoms during the third trimester of pregnancy supports our conservative attitude in the management of the condition. Besides, the application of the American guidelines for the management of other kinds of left ventricular output obstruction in the course of pregnancy supported our management approach\textsuperscript{13}.

Recently, an echocardiographic model to predict which individuals will develop progression of the stenosis has been proposed and validated. This model takes into account three
parameters: the gradient at the time of diagnosis, the distance from the membrane to the aortic valve, and whether there is involvement of the anterior leaflet of the mitral valve\(^{14}\). Using this standard, our patient probably has non-progressive discrete subaortic stenosis. Nevertheless, it will be the progression of her clinical and echocardiographical parameters, together with her personal decision, that will determine our attitude in the long term.

REFERENCES


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