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IMAGE

A rare case of aortic stenosis in adulthood

Un cas rare de sténose aortique chez l'adulte

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MOTS CLÉS

Sténose supravalvulaire aortique ;
Syndrome de Williams ;
Évaluation scanographique ;
Traitement chirurgical

A 52-year-old woman was referred for surgical treatment of a symptomatic aortic stenosis. Her medical history included unexplored mild mental retardation. Physical examination revealed high blood pressure (140/87 mmHg) with a systolic aortic 3/6 murmur and a distinctive facial appearance (spaced teeth, long philtrum and sunken nasal bridge), suggestive of Williams's syndrome. Transthoracic echocardiography showed a calcified bicuspid aortic valve with mild insufficiency, a supravalvular aortic stenosis (SVAS) with a mean gradient of 78 mmHg and normal left ventricular systolic function (Fig. 1). Pulmonary stenosis, mitral involvement and ventricular septal defect were not found. A computed tomography scan confirmed SVAS (SVAS area, 1.35 cm²) (Fig. 2). There was no aortic coarctation. An angiogram showed a slight right coronary artery and blood analysis revealed normal calcaemia. The deletion of an elastin allele was subsequently found by fluorescence in situ hybridization and confirmed the 7q11.23 chromosomal microdeletion. A tight SVAS was found at the sinotubular junction after median sternotomy. The aortic valve was bicuspid type I with bulky calcifications. The non-coronary sinus was fully split then the sinotubular junction was enlarged using a Dacron patch to allow aortic valve replacement with a mechanical prosthesis (On-X 19 mm) (Fig. 3). The postoperative course was uneventful and the patient was discharged 11 days postoperatively.

Abbreviation: SVAS, supravalvular aortic stenosis.

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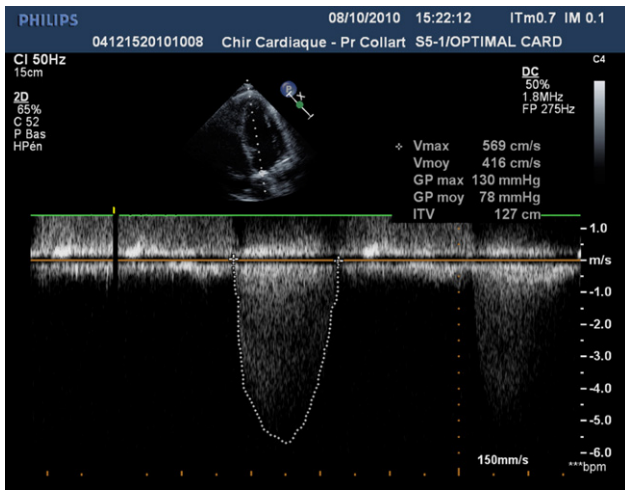


Figure 1. Assessment of mean supravalvular stenosis gradient with transaortic continuous Doppler using Bernoulli's law. Mean gradient, 78 mmHg; Vmax, 5.69 m/s.

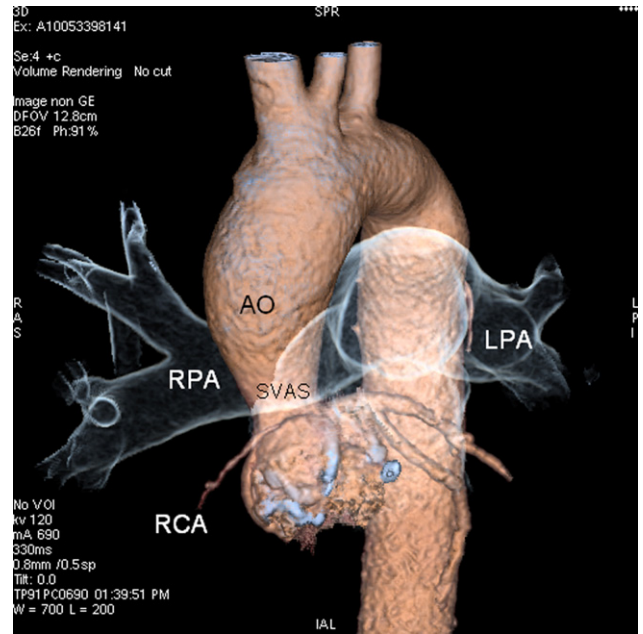


Figure 2. Three-dimensional computed tomography scan reconstruction: supravalvular aortic stenosis (SVAS). Note the absence of pulmonary tract stenosis (in transparency). AO: aorta; LPA: left pulmonary artery; RPA: right pulmonary artery.

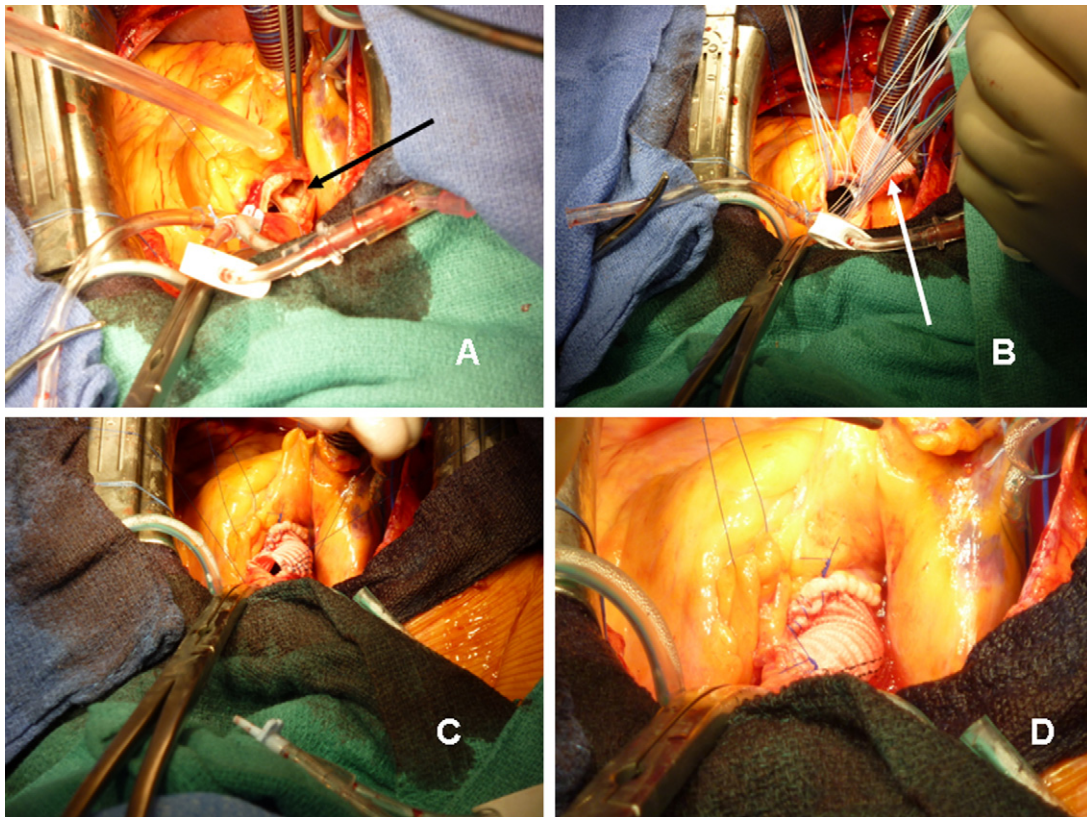


Figure 3. Surgical views after median sternotomy. (A) Bicuspid aortic valve with tight aortic outflow (black arrow). (B) Enlargement of the sinotubular junction with a Dacron patch (white arrow). (C and D) Final result after aortic valve replacement with mechanical prosthesis.

This case is one of the longest natural courses of SVAS, which is the most common cardiac abnormality in Williams's syndrome. Spontaneous improvement may occur in mild SVAS but severe SVAS always needs surgical correction. Coronary involvement increased the mortality risk.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.