Discrete subaortic stenosis 37 years after repair of a ventricular septal defect

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Abstract

Discrete subaortic stenosis (DSS) is uncommon in adults after surgical correction of congenital heart defects. There are only a few published reports on the occurrence of DSS in adults. We present an adult case with DSS after repair of a ventricular septal defect (VSD). The case was a 44-year old female patient who underwent VSD closure at 7 years of age. Thirty-seven years later, she presented with congestive heart failure associated with severe subaortic membranous stenosis and atrial fibrillation (AF) that required surgical repair. We report successful surgical treatment of this adult patient with DSS and AF 37 years after repair of a VSD.

Keywords: Discrete subaortic stenosis • Ventricular septal defect • Adult

INTRODUCTION

Discrete subaortic stenosis (DSS) is considered an acquired cardiac defect, and presumed to be secondary to altered flow patterns caused by morphological abnormalities in the left ventricular outflow tract (LVOT) [1]. Owing to its associated rapid clinical impairment during childhood, DSS has long been considered to only appear in childhood, and there are few published reports on the evolution of DSS during adult life [2, 3]. The incidence, characteristics and therapeutic options for DSS in adults are not well documented. This report documents the clinical features of DSS in an adult.

CASE REPORT

A 44-year old female had undergone surgery for a ventricular septal defect (VSD) at 7 years of age and had given safe, natural birth at 23 years of age. She remained asymptomatic for 37 years after the surgery, and then presented with dyspnoea. A chest X-ray showed cardiomegaly and pulmonary congestion. Brain natriuretic peptide was high at 773.2 pg/ml. An electrocardiogram showed atrial fibrillation (AF). Echocardiography showed atrial fibrillation (AF). Echocardiography showed no abnormal structure in the LVOT. A postoperative electrocardiogram showed sinus rhythm and echocardiography showed no abnormal structure in the LVOT.

Computed tomography confirmed the diagnosis of a discrete membrane at the LVOT level (Fig. 1B). The operation was performed on standard conventional cardiopulmonary bypass under moderate hypothermia. First, we performed pulmonary vein isolation for AF using a radiofrequency device. A transverse aortotomy was carried out and the aortic valve leaflets were retracted, revealing a discrete subaortic membrane at 8 mm from the aortic valve and a defined fibromembranous ring that completely encircled the subvalvular aortic region. Attachments to the anterior mitral leaflet were carefully defined and sharply divided. Complete resection of the circumferential obstructing discrete membrane, including the adjacent hypertrophied muscle, was performed (Fig. 2). The postoperative course was uncomplicated. A postoperative electrocardiogram showed sinus rhythm and echocardiography showed no abnormal structure in the LVOT.

DISCUSSION

The onset and progression of DSS are affected by rheologic abnormalities localized to the LVOT. A four-stage aetiology was proposed by Cape et al. [1], whereby different morphologic LVOT abnormalities, such as steeper aortoseptal angle and narrow LVOT, result in altered septal shear stress, which triggers a genetic predisposition leading to cellular proliferation and structures in the LVOT. Although progression of DSS obstruction can be very rapid in infants and small children, progression during adult life is slow [2]. DSS is not a congenital defect because it develops over an extended period after birth and is almost never encountered in infants. DSS was associated with another CHD in 33–44% of adult patients, and most lesions appeared after surgical repair of the associated CHD [2, 4]. Patients with associated CHD were younger and had less LVOT obstruction than those with isolated DSS [2]. The two most frequently associated cardiac malformations were VSDs and aortic coarctation, but DSS was also associated with atrioventricular septal defects, patent ductus arteriosus, bicuspid aortic valve or...
double-outlet right ventricle [5]. Our patient simultaneously had VSD and bicuspid aortic valve. There are no previous reports of cases resembling our patient. It is possible that an increase in the number of patients reaching adulthood after surgical correction of different CHDs may be causing an actual increase in the prevalence of DSS during adult life.

The natural history is unpredictable. Left untreated, progressive DSS can lead to important complications, including left ventricular hypertrophy, arrhythmias, AR and endocarditis. This may be explained morphologically by the degree of involvement of the anterior mitral leaflet, distance of the membrane from the aortic valve, steeper aortoseptal angle and narrower LVOT, which can serve as reliable predictors of progression [6]. AR is frequently found in patients with DSS [2]. The high-velocity jet may also damage the aortic valve leaflets, causing AR because of thick fibrous tissue on the left ventricular surface of the leaflets. However, significant progression of AR during adult life was not found, and there was no significant relationship between AR and age and no significant increase in AR over time [2]. Moreover, the severity of AR in DSS repaired during childhood was significantly greater than that in patients who survived naturally into adulthood.

Surgical decisions in adult patients with DSS should be based not only on the anatomic finding of subvalvular stenosis, but also on clinical evaluation, left ventricular hypertrophy, systolic function and aortic insufficiency. However, we think that early surgery may preserve the integrity of the aortic valve in a manner that avoids later valve replacement. Early intervention and complete resection can prevent the development of abnormal muscular hypertrophy and also prevent AR progression, and have therefore been recommended. The aortoseptal angle does not normalize when surgery is performed in older patients, suggesting that the geometry may be remodelled postoperatively in younger patients and making it advantageous to perform surgery earlier [7].

Despite adequate resection, there is a substantial recurrence rate 11.7–35% [8, 9]. Our surgical approach to DSS is transaortic enucleation of the fibromuscular membrane, combined with limited myectomy. Geva et al. [10] reported that independent predictors of reoperation for recurrent DSS available before surgery were peeling of the lesion from the aortic or mitral valve, distance between the lesion and aortic valve of <6 mm and peak gradient of ≥60 mmHg. In conclusion, we report successful surgical treatment of an adult patient with DSS and AF. This is a rare case, which was characterized by slow progression of growth after VSD repair. It is suggested that its prevalence in adults may be increasing because of the greater number of repaired CHDs. To prevent eventual haemodynamic collapse, early surgical resection is recommended for DSS. However, the long-term outcome of this treatment remains unknown, and meticulous follow-up of the LVOT and aortic valve function is mandatory.

Conflict of interest: none declared.

REFERENCES


