Surgical Treatment of Discrete Subaortic Stenosis in an Adult

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We report on an adult patient with discrete-type subaortic stenosis. A 48-year-old man who had progressed asymptptomatically since childhood despite heart murmur was transferred to our hospital. The patient was diagnosed as having severe aortic stenosis with a pressure gradient of 100 mmHg across the aortic valve, associated with a grade II aortic regurgitation. A conventional aortic valve replacement was scheduled. During surgery, the aortic valve was found to be tricuspid but incompetent as a result of shrinking and thickening of the left coronary cusp. A circumferential fibromuscular ridge was observed under the cusps, which corresponded to Kelly's type II discrete subaortic stenosis. Because of the small subaortic area and deformity of the cusp, we performed aortic valve replacement after excision of all cusps and the fibromuscular ridge. Early corrective surgery is recommended for discrete subaortic stenosis to prevention regurgitation progression.

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Key Words: Discrete subaortic stenosis; Aortic stenosis and regurgitation; Aortic valve replacement

A 48-year-old man was operated on for aortic stenosis and regurgitation resulting from a localized subvalvular fibromuscular ridge. He had no symptoms but the pressure gradient (PG) across the aortic valve was 100 mmHg. Aortography revealed grade II aortic regurgitation according to Sellers' classification. He underwent resection of the fibromuscular ridge and aortic valve replacement with a CarboMedics prosthesis. His operative course was uneventful and the PG was decreased to 38 mmHg. Discrete subaortic stenosis accounts for 8–10% of all cases of congenital aortic stenosis and occurs twice as frequently in males as in females! Here we report a successful surgical case of a discrete subaortic stenosis in an adult.

Case Report

A 48-year-old man was transferred to our institution with a diagnosis of aortic stenosis. It was known that he had a heart murmur since childhood but had developed asymptptomatically. On admission, he was categorized as New York Heart Association (NYHA) class I under no medication. His blood pressure was 110/60 mmHg with a regular pulse rate of 76 per min. A grade 4/6 ejectional systolic murmur was heard in the aortic area. The cardiothoracic ratio was 0.57 with an enlarged left ventricle and ascending aorta observed on the chest radiograph (Fig 1). The electrocardiogram revealed left ventricular hypertrophy by voltage criteria with a normal sinus rhythm. Hematologic examination revealed no abnormalities. A severe PG (100 mmHg) across the aortic valve was apparent on cardiac catheterization, and enlargement of the proximal ascending aorta and grade II aortic regurgitation were detected by aortography. Left ventriculography revealed a filling defect beneath the aortic valve (Fig 2). Left ventricular wall motion was good.
Two-dimensional echocardiography revealed a marked thickening of the interventricular septum and left ventricular posterior wall. Doppler echocardiography revealed grade II aortic regurgitation and a PG of 100 mmHg across the aortic valve. There was no echocardiographic evidence of hypertrophic cardiomyopathy.

A diagnosis of aortic valve stenosis with left ventricular hypertrophy was made, and routine aortic valve replacement was scheduled. An operation was performed using a median sternotomy and employing a standard cardiopulmonary bypass with cold blood cardioplegia. The ascending aorta was markedly dilated (diameter 5.5 cm). The aortotomy was a J-shaped incision extending to the commissure between the non-coronary cusp and the left coronary cusp. The aortic valve was tricuspid but incompetent owing to shrinking and thickening of the left coronary cusp (Fig 3a). The aortic annulus was not observed to be...
small. When the aortic cusps were retracted, a sub-
valvular fibromuscular ridge was exposed (Fig 3b). It
was localized circumferentially and situated at the base
of the aortic leaflets. The ridge was particularly prom-
inent below the right aortic cusp. After the aortic
valve was excised, the fibromuscular ridge was
resected circumferentially until the mitral apparatus
was clearly visualized through the left ventricular out-
flow tract. The diameter of the left ventricular outflow
tract was enlarged from 19 mm to 23 mm. The aortic
valve was replaced with a CarboMedics prosthesis (size
23R) in the supra-annular position. The patient did
not need a blood transfusion. Postoperative recovery
was satisfactory. Doppler echocardiography showed
the PG across the aortic prosthesis to be 47 mmHg.
Histologic examination of the fibromuscular ridge
showed strong fibrous endocardial tissue.

At present, 3 months after surgery, the patient is in
NYHA class I and is being treated with 40 mg/day oral
furosemide and 0.375 mg/day digoxin. The PG across
the aortic prosthesis was reduced to 38 mmHg as deter-
mined by Doppler echocardiography.

Discussion

Discrete subaortic stenosis accounts for 8–10% of
all cases of congenital aortic stenosis and occurs
twice as frequently in males as in females! The lesion
consists of a membranous diaphragm (Kelly type I) or
fibromuscular ring encircling the left ventricular out-
flow tract just beneath the base of the aortic valve
(Kelly type II). The transitional type has also been
reported. This patient had a type II lesion.

Van Praagh et al stated that fibrous subaortic ste-
nosis results from malformation of the endocardial
cushion tissue of the atroventricular canal that usually
forms the anterior leaflet of the mitral valve. Somer-
ville et al proposed that the lesion is acquired rather
than congenital because an obstructive subaortic
membranous flap occurs rarely, if ever, in newborns.
They suggested that an abnormal flow pattern in the
left ventricle results in the deposition of fibroelastic
material below the aortic valve.

Mild degrees of aortic valvular regurgitation are
commonly observed in patients with discrete subaortic
stenosis and appear to be caused by thickening of the
valve and impaired mobility of the cusps secondary to
the trauma created by the high-velocity jet passing
through the subaortic lesion. Furthermore, infective
endocarditis is a serious and frequent complication
of subaortic stenosis that is not treated surgically.
Because of progressive obstruction and aortic regur-
gitation, the presence of even mild or moderate sub-
aortic stenosis warrants consideration of an elective
operation. Surgical correction is accomplished by
excising the membrane or fibrous ridge. If necessary,
the aortic valve is replaced. An operation is expected
to improve the hemodynamic state substantially but, in
a few patients, secondary muscular hypertrophy of the
outflow tract and PG may persist after the operation.

Conclusion

Resection of a fibromuscular ridge and aortic valve
replacement was performed in a patient with discrete
subaortic stenosis with aortic regurgitation; the out-
come was good. Immediate surgery is desirable in
patients with discrete subaortic stenosis to prevent
progression of aortic regurgitation.

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