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"ACQUIRED" SUBVALVULAR AORTIC STENOSIS AFTER REPAIR OF SEVERAL CONGENITAL CARDIAC DEFECTS*

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Discrete subvalvular aortic stenosis is a progressive lesion. In this report we presented nine patients who had no significant left ventricular-aortic obstruction at initial cardiac catheterization or echocardiographic examination, but later developed significant sbvalvular aortic stenosis. Associated lesions included ventricular septal defect in three, patent ductus arteriosus in two, aorticopulmonary window in one, tetralogy of Fallot in one, supramitral membrane in one, and ventricular septal defect and patent ductus arteriosus in one case.

Nine patients were diagnosed with subvalvular aortic stenosis 18 months to eight years after surgical correction. Eight of the patients required surgery for subvalvular obstruction.

In conclusion, discrete subaortic stenosis is a rare, late complication of the surgical repair of several congenital heart defects. It is a progressive lesion after surgery; therefore these patients require careful follow-up. *Key words: subaortic stenosis, surgery, follow-up.*

Discrete subaortic stenosis accounts for approximately 10 percent all cases of congenital aortic stenosis[1]. It is frequently associated with congenital heart defects, including ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot and coarctation of the aorta[2-4]. Although the development of "acquired" subaortic stenosis (SAS) after surgical repair of ventricular septal defect, tetralogy of Fallot, double outlet right ventricle and common atrioventricular canal defects has been described previously, there is little data about the relative incidence of post-surgical subaortic stenosis.

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The purpose of this report was to present the development of subaortic stenosis after surgical treatment in patients who had no subaortic obstruction at initial echocardiography or cardiac catheterization.

Material and Methods

Nine patients who had undergone cardiovascular surgery for congenital heart defects between 1982 and 1989, and who later developed subaortic stenosis as shown by two-dimensional echocardiography during follow-up, were reviewed retrospectively. Patients who had subaortic stenosis preoperatively were excluded.

Two-dimensional and Doppler echocardiographic examination was performed with Toshiba SSH 60 equipment with 2.5, 3.75 and 5.0 MHz transducers pre- and postoperatively. The standard subcostal, parasternal, apical and supra-sternal views were used.

Seven patients underwent left cardiac catheterization. Left ventricular angiography was performed by using long-axis (60° left anterior oblique with 30° cranial angulation) and four-chamber (45° left anterior oblique with 30° cranial angulation) wiews. The diagnosis of acquired subaortic stenosis was made in all patients by two-dimensional and Doppler echocardiography. The last two patients underwent surgery without cardiac catheterization.

The indications for surgery included a subaortic gradient > 25 mmHg or a subaortic gradient < 25 mmHg with aortic regurgitation on echocardiography or cardiac catheterization.

Results

We studied nine patients (six boys and three girls). The median age on admission was 10 months (range six months to five years). The mean age at first operation was 3.6 ± 1.4 years (range 2-6.5 years). The mean age at diagnosis of subvalvular aortic stenosis was 7.8 ± 1.8 years (range 4.9-11 years). The diagnosis of SAS was made at a mean interval of 4.26 ± 1.7 years (range 1.5-8 years) after the operation.

Table I shows the age at operation, type of surgical repair and follow-up of the nine patients. Before surgery we did not demonstrate a subaortic membrane in any patient on echocardiographic examination. In three patients there was no pressure gradient between the left ventricle and the aorta. We diagnosed a ventricular septal defect in three patients, patent ductus arteriosus in two patients, aorticopulmonary window, supramitral membrane, tetralogy of Fallot, ventricular septal defect and patent ductus arteriosus in one patient each.

Two patients underwent surgery for subaortic stenosis after echocardiographic examination only. Table II shows the results of the cardiac catheterization in seven patients together with Doppler gradients. The subaortic gradient on Doppler

examination was 3.5 ± 9.5 mmHg. The mean gradient between the left ventricle and the aorta was much higher (mean 58.8 ± 32 mmHg, range 0-90 mmHg). The difference between data related to gradients gathered by Doppler versus cardiac catheterization was statistically significant. This difference may be related to the time between Doppler examination and catheterization. (i.e., in one patient admitted with the complaint of chest pain four years after Doppler examination, an 82 mmHg left ventricle to aortic gradient was detected on cardiac catheterization).

The subaortic membrane was resected in five patients. Three patients were awaiting surgery as of this writing.

Table I: Age at Operation, Type of Surgical Repair and Follow-up in 9 Patients

Age on Admission	Age at Operation and Type of Surgical Repair	Procedure Before Operation	Period Before Subaortic Stenosis Occurence	Follow-up
7 moths	2 Y PDA ligation 3 Y VSD closure	E + C	8 years	Operation
5 years	5 Y BT Shunt 6.5 Y Total correction	E	1.5 years	Membrane resection
2.5 years	3 Y APW closure	C + E	3 years	Clinical follow-up
7 months	10 months Pulmonary banding 22 months Depanding + VSD closure + patch for RVOT	E	4 years	Operation
7 months	3 Y Supramitral memb. resection	E + C	3.5 years	Operation
6 months	4 Y VSD closure	E + C	4.5 years	Membrane resection
5 years	5 Y PDA ligation	E	5 years	Membrane resection
16 months	2 Y PDA ligation	E + C	4 years	Membrane resection
10 months	4 Y VSD closure	E + C	4 years + 9 month	

Y : Years.
E : Echocardiography.
C : Catheterization.
PDA : Patent ductus arteriosus.
VSD : Ventricular septal defect.
APW : Aorticopulmonary window.
BT : Blalock-Taussing.
RVOT : Right ventricular outflow tract.

Table II: Catheterization Results and Doppler Echocardiography Gradients

Number	Echo Gradient*	LV Pressure*	Subaortic Pressure*	Aortic Pressure*	Subaortic Gradient*
1	28	177	95	95/71 Mean: 81	82
2	24	195	115	100/62 Mean: 79	80
3	20	120	120	120/66 Mean: 90	0
4	31	Underwent Operation with Echocardiography			
5	40	165	120	115/78 Mean: 98	45
6	40	180	140	120/80 Mean: 100	40
7	45	200	110	95/60 Mean: 71	90
8	40	Underwent Operation with Echocardiography			
9	47	200	125	125/75 Mean: 92	75
Mean	35 + 9.5**				58.8 + 32

* mmHg.
** Mean Doppler gradient in patients who have a catheterization subaortic gradient.
LV: Left ventricle.

Discussion

The development and incidence of subaortic stenosis after surgical repair of several congenital heart defects have been documented previously[4-7]. Although the number of patients in the series was small, Ciccini et al.[8] reported that the incidences of subaortic stenosis in ventricular septal defect, tetralogy of Fallot and double outlet right ventricle were 3.2, 2.1 and 21.4 percent, respectively. In the presence of a ventricular septal defect, hemodynamic diagnosis of a subaortic obstruction may be difficult to make using Doppler echocardiography or peak-to-peak catheterization gradient. A ventricular septal defect decompresses the left ventricle and minimizes the left ventricular outflow tract pressure gradient until it is closed. The other factor masking the presence of the membrane is shunting of the contrast material through the ventricular septal defect at angiocardiography[8-9]. Therefore, the diagnosis of subaortic obstruction is usually made on the basis of morphologic data obtained by two-dimensional echocardiography rather than on the basis of the hemodynamic gradient[8]. In our cases, there was no evidence of obstruction on angiocardiographic (in seven patients) and two-dimensional echocardiographic (in nine patients) examination obtained before surgical correction of the cardiac defects.

The occurrence of a subaortic membrane post-operatively has been reported in cases of tetralogy of Fallot, ventricular septal defect, common atrioventricular canal, double outlet right ventricle and patent ductus arteriosus. The etiology and pathogenesis of subaortic stenosis are still not completely understood.

Possible mechanisms have been proposed. Pathogenesis as suggested by Keith[10] may be caused by malamalgamation of the subaortic conus with the ventricular septum, followed by incomplete absorption of the aortic infundibulum. Van Praagh et al.[11] suggested that maldevelopment of the atrioventricular canal and subsequent constriction of the left ventricular outflow tract by abnormal migration of the endocardial cushion tissue into the subaortic region could be responsible. Pyle et al.[12] hypothesized that the fibrocartilagenous ring of subaortic stenosis is derived from persistent embryonal endocardial cushion tissue that retains its proliferative capacity. They also suggested that subaortic stenosis was not a true congenital defect but developed postnatally. Recently Gewillig et al.[13] suggested that the turbulent blood flow that may be created by the patch sewn on the right ventricular side of the interventricular septum may damage the endothelium and thereby initiate the formation of a subaortic ridge. Sommerville[14] postulated the idea of turbulence which stimulates the formation of discrete subaortic stenosis.

In one of our cases, the cause of the discrete subaortic membrane may have been a malalignment-type ventricular septal defect. Zielinsky et al.[15] demonstrated that a malalignment-type ventricular septal defect is a constant feature in patients who develop preoperative or postoperative discrete subaortic stenosis with ventricular septal defect[14]. In the other cases with ventricular septal defect, turbulence originating from an abnormally contracting myocardium and patch may have been the reason for a discrete subaortic membrane.

In two patients with patent ductus arteriosus and one patient with an aorticopulmonary window, it is impossible to explain the mechanism of formation of the subaortic membrane after surgery. However, there are reports in the literature about the formation of a membrane after patent ductus arteriosus ligation. Leichter et al.[6] reported 12 cases with patent ductus arteriosus either alone or in combination with other cardiac defects, such as ventricular septal defect, aorticopulmonary window, atrioventricular canal defect and pulmonary stenosis. Two of our cases underwent surgery after only echocardiographic examination. In one patient with aorticopulmonary window we did not find any gradient between the left ventricle and the aorta in the pullback of the left ventricle to aorta before surgery. However, three years after the operation we demonstrated a subaortic membrane with 31 mmHg instantaneous peak gradient by two-dimensional and Doppler echocardiographic examination.

In previous reports the time lapse between surgery and development of the subaortic membrane was reported as three months to 18 years, with a mean of two years and nine months[5, 6, 8]. In our study this period averaged 4.2 ± 1.7 years (range 18 months to eight years). Five patients were operated on for resection of membranes. Three patients were awaiting surgery as of this writing.

In conclusion we believe that discrete subaortic stenosis is a rare, late complication of several congenital heart defects and that it is an acquired condition. Because of its progressive nature after surgery, these patients must be followed carefully. If the patient has a significant heart murmur or electrocardiographic findings, two-dimensional echocardiography and catheterization must be performed. Because two-dimensional echocardiography is useful in demonstrating the subaortic membrane, we recommend echocardiography for patients with subaortic stenosis to assure accurate diagnosis and proper management.

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