Pediatr Cardiol 15:91–94, 1994

Pediatric Cardiology

Discrete Subaortic Stenosis after Successful Treatment of Congenital Aortic Valve Stenosis

M.A. Salim,[1] D.C. Watson,[2] B.S. Alpert,[1] and T.G. Di Sessa[1]

Departments of [1]Pediatric (Cardiology) and [2]Cardiovascular Surgery, University of Tennessee School of Medicine, Memphis, Tennessee, USA

SUMMARY. **Two cases of discrete subaortic obstruction which developed in a previously normal left ventricular outflow tract of patients with congenital valvar aortic stenosis are described. These examples emphasize the need for careful scrutiny of the etiology of recurrent postoperative left ventricular outflow tract obstruction.**

KEY WORDS: **Discrete subaortic stenosis — Congenital aortic valve stenosis — Cardiac surgery**

Discrete subaortic stenosis (DSAS) is an uncommon cause of left ventricular outflow tract (LVOT) obstruction [6]. Previous investigators have categorized fixed DSAS into three types: membranous, fibromuscular, and tunnel [5, 9, 11]. To our knowledge, DSAS has never been reported in newborn infants and appears to develop in the first few years of life [5, 9]. Furthermore, it can be an isolated lesion or can occur in combination with other forms of congenital heart disease [1, 2, 4, 5, 8–11]. We report the clinical, echocardiographic, hemodynamic, angiocardiographic, and surgical findings in two infants who developed DSAS after successful palliation of congenital valvar aortic stenosis.

Case 1

R.B. was the 3190 g male child of a normal gestation, labor, and delivery. On the first day of life, cardiac examination revealed a grade III/VI blowing holosystolic murmur which was heard best at the base of the heart and radiated to both the axilla and back. The amplitude of the pulses in the arms and legs was diminished. The chest radiograph demonstrated normal heart size with interstitial pulmonary edema. The electrocardiogram showed left atrial enlargement, biventricular hypertrophy, and nonspecific ST-T-wave changes in the right chest leads. An echocardiogram demonstrated a bicuspid aortic valve with an eccentric orifice and a normal LVOT (Fig. 1). Doppler interrogation of the LVOT predicted a maximal instantaneous pressure gradient of 80 mmHg. Color flow Doppler demonstrated laminar flow in the subaortic area.

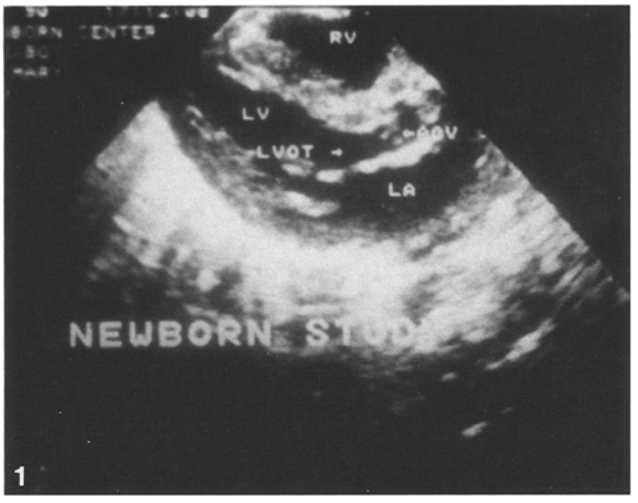
Address offprint requests to: Dr. Thomas Di Sessa, 777 Washington Avenue, Suite 215, Memphis, TN 38105, USA.

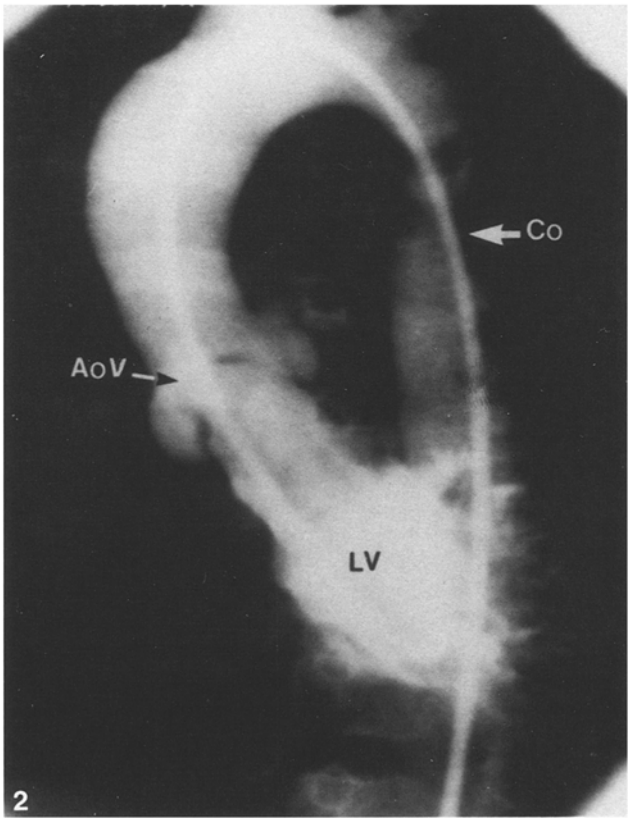
At 5 days of age, he underwent repair of duodenal atresia by means of resection and primary reanastomosis. Open aortic valvotomy of a bicuspid, thickened myxomatous valve was performed at 17 days of age. The commissure was opened completely. Postoperatively, he did well. Noninvasive evaluation at 7 months of age indicated recurrence of aortic stenosis. The patient, therefore, underwent cardiac catheterization and a successful aortic balloon valvuloplasty, using a balloon diameter to aortic annulus diameter ratio of 0.9–1.0:1 (Fig. 2, Table 1). There was angiographic evidence of a mild coarctation of the aorta. However, only a 10 mmHg pressure difference was measured from ascending to descending aorta. An echocardiogram after valvuloplasty demonstrated moderate aortic valve stenosis and aortic insufficiency with a normal left ventricular outflow tract.

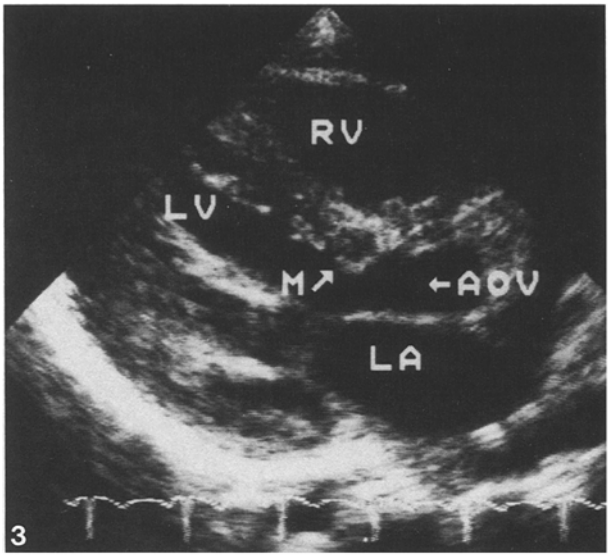
By 17 months of age, he developed left ventricular hypertrophy by electrocardiographic voltage criteria along with T-wave inversion in the left precordial leads. In addition to an abnormal aortic valve, echocardiogram demonstrated a fibromuscular ridge attached to the interventricular septum (Fig. 3). This structure narrowed the left ventricular outflow tract and had not been observed on previous echocardiograms. Cardiac catheterization and angiogram confirmed the presence of subaortic obstruction with a peak-to-peak gradient of 60 mmHg (Fig. 4, Table 1). At 19 months of age, the patient underwent successful resection of a fibromuscular subaortic obstruction.

Case 2

C.L. was admitted at 6 days of age in congestive heart failure with diminished pulses in all extremities and a short grade II/VI systolic murmur at the right upper sternal border. On chest radiograph, there was cardiomegaly with increased pulmonary vascularity. An electrocardiogram had voltage criteria for right ventricular hypertrophy along with nonspecific ST-T-wave changes. The echocardiogram revealed a thick dysplastic aortic valve that







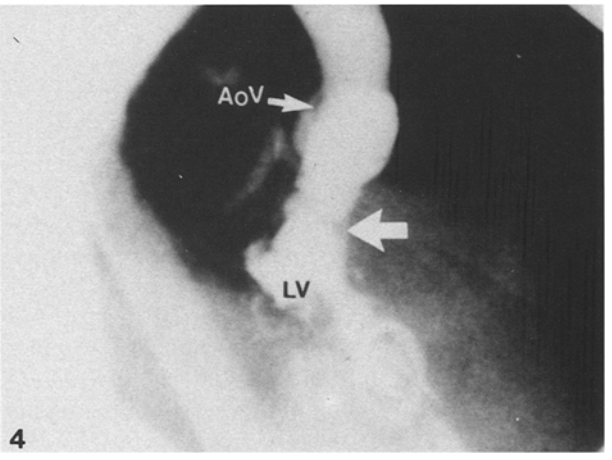


Fig. 1. Case 1, two-dimensional echocardiogram recorded in the parasternal long axis plane. In this diastolic frame, the left ventricular outflow tract (LVOT) is free of extraneous echoes. The aortic valve (AoV) is thickened. LA, left atrium; LV, left ventricle; RV, right ventricle.

Fig. 2. Case 1, left ventricular cineangiogram in the left anterior oblique projection performed at 7 months of age. This systolic frame demonstrated thickened domed aortic valve. The left ventricular outflow tract showed no evidence of subaortic obstruction. The aortic arch has a mild coarctation. However, there was only 10 mmHg pressure gradient across this narrowed area. AoV, aortic valve; CO, coarctation of the aorta; LV, left ventricle.

Fig. 3. Case 1, two-dimensional echocardiogram recorded in the apical long axis plane. In this diastolic frame, fibromuscular ridge (*M*) is imaged traversing the left ventricular outflow tract (*LVOT*) beneath the aortic valve (*AoV*). LV, left ventricle.

Fig. 4. Case 1, left ventricular cineangiogram in the long axial oblique projection at 17 months of age. This systolic frame demonstrated thickened domed aortic valve. Traversing the left ventricular outflow tract between the interventricular septum and the mitral valve is a radiolucent structure beneath the aortic valve (*arrow*). AoV, aortic valve; LV, left ventricle.

domed in systole. The LVOT was free of echogenic structures. Left ventricular function was diminished severely. Doppler demonstrated a peak LVOT velocity of 3.5 m/s across the aortic valve. A patent ductus arteriosus and an atrial septal defect were also present. At 8 days of age, cardiac catheterization and aortic balloon valvuloplasty were performed with subsequent improvement in left ventricular function (Table 1).

At 7 weeks of age, due to persistent ventilator dependence and evidence of a large left-to-right shunt, surgical closure of an atrial septal defect was performed. The aortic valve was inspected and found to be bicuspid, myxomatous, and thickened.

Table 1. Clinical and catheterizations data summary

Procedure	Age	Qp/Qs	RV	PA	LVapex	LVOT	Ao	Comment
Case I								
Ao valvotomy (surgical)	3 weeks							
Cath. no. 1	7 months	1:1	25/2	25/10	162/9		100/54	Pre-BV
					130/5		98/50	Post-BV
Cath. no. 2	18 months				160/14	100/10	80/40	
Resection of subaortic obstruction	19 months							
Case II								
Cath. no. 1	7 days	4.6:1	78/10	70/37	130/20		60/40	Radial artery
Cath. no. 2	8 days				130/15		60/35	Pre-BV
					105/20		75/45	Post-BV
Cath. no. 3	16 days	4:1	70/11	70/30	100/23		60/40	Cuff pressure
Surgical closure of ASD & opening of AoV	7 weeks							
Cath. no. 4	8 months				220/20	140/15	80/40	
Resection of subaortic obstruction	9 months							

Ao, aorta, aortic; AoV, aortic valve; ASD, atrial septal defect; BV, Balloon Valvuloplasty; LV apex, apex of left ventricle; LVOT, left ventricular outflow tract above the subaortic membrane; PA, pulmonary artery; pressures in mmHg; Qp : Qs, pulmonary to systemic flow; RV, right ventricle.

There were no intercommissural tears with almost complete opening of the commissure. Only 1–2 mm of residual fusion remained and it was incised.

At 8 months of age, repeat cardiac catheterization demonstrated a subaortic systolic pressure gradient of 80 mmHg (Table 1). Cineangiocardiogram demonstrated a narrowed LVOT with a radiolucent region between the interventricular septum and anterior mitral leaflet. At 9 months of age, subaortic fibromuscular obstruction was resected.

Discussion

These two cases are examples of DSAS that developed in patients who had a previously normal LVOT documented by angiogram and echocardiogram. The association of DSAS with ventricular septal defect [1, 5, 10], coarctation of the aorta [2], as well as other forms of congenital heart disease, is well described [4, 5, 10, 11]. However, the association of DSAS with aortic valve disease has been less well demonstrated. Newfeld and associates [5] reported a child who had aortic valvotomy at 2 years of age with "nonsignificant" DSAS and 1 year later died of severe congestive heart failure secondary to subaortic obstruction. Schneeweiss et al. [8] reported seven cases of coexisting subaortic stenosis and aortic valve disease. Similar data were reported by Wright et al. [11]. However, in all previous reports, patients had both lesions at the time of initial diagnosis.

To our knowledge there are no reports of the development of subaortic stenosis after relief of valvar disease. Both of these patients developed DSAS less than 1 year after angiogram and echocardiogram demonstrated a normal subaortic LVOT. In Case 2, it was less than 6 months after the surgery and 8 months after the last catheterization. Moreover, the diagnosis of DSAS in the first year of life is unusual. Freedom et al. [2] reported a case of fatal DSAS at 6 months of age. Of 83 patients reported by Wright et al. [11], only two patients were less than 1 year of age at diagnosis. Leichter et al. [4] reported only one out of 35 patients less than 1 year of age with DSAS. These reports emphasize the developmental nature of this lesion. However, the pathophysiology of this disease is elusive.

After closure of a ventricular septal defect, the development of subaortic stenosis most likely involves either malalignment or posterior displacement of the infundibular septum [10], or unrecognized subaortic obstruction preoperatively [1]. In the absence of ventricular septal defect, an explanation for the developmental nature of the obstruction may be deduced from the experience with the Newfoundland dog [7]. It has been shown that subvalvar aortic stenosis is developmental in this breed and appears only after 3 weeks of age. It is postulated that persistent embryonic endocardial tissue in the LVOT is capable of proliferating with age and causing obstruction. An alternate hypothesis invokes the presence of turbulent flow below the site of the subaortic obstruction as the causal factor in the pathogenesis of DSAS [3].

Both patients had balloon valvuloplasty several months prior to the development of subaortic stenosis. This raises the possibility that local trauma during balloon inflation may have triggered tissue proliferation in the LVOT. Long-term follow-up of the

new population of patients with a ballooned aortic valve will determine the validity of this speculation.

These two cases demonstrate the developmental nature of discrete subaortic stenosis in patients with congenital aortic stenosis. Moreover, they provide the foundation for speculation concerning the underlying pathophysiology.

References

1. Fisher DJ, Snider AR, Silverman NH (1982) Ventricular septal defect with silent discrete subaortic stenosis. *Pediatr Cardiol 2*:265–269
2. Freedom RM, Fowler RS, Duncan WJ (1981) Rapid evolution from "normal" left ventricular outflow tract to fatal subaortic stenosis in infancy. *Br Heart J 45*:605–609
3. Gewillig M, Daenen W, Dumoulin M, Van Der Hauwaert L (1992) Rheologic genesis of discrete subvalvar aortic stenosis: a Doppler study. *J Am Coll Cardiol 19*:818–824
4. Leichter DA, Sullivan I, Gersony WM (1989) "Acquired" discrete subvalvular aortic stenosis: normal history and hemodynamics. *J Am Coll Cardiol 14*:1539–1544
5. Newfeld EA, Muster AJ, Paul MH, Idriss FS, Riker WL (1976) Discrete subvalvular aortic stenosis: study of 51 patients. *Am J Cardiol 38*:53–61
6. Olley PM, Bloom KR, Row RD (1978) Aortic stenosis: valvular subaortic and supravalvular. In: Keith JD, Rowe RD, Vland P (eds) Heart disease in infancy and childhood, 3rd edn. Macmillan, New York, pp 698–710
7. Pyle RL, Patterson DF, Chacko S (1976) The genetics and pathology of discrete subaortic stenosis in the Newfoundland dog. *Am Heart J 92*:324–334
8. Schneeweiss A, Motro M, Shem-Tov A, Bleiden LC, Neufeld HN (1983) Discrete subaortic stenosis associated with congenital valvular aortic stenosis: a diagnostic challenge. *Am Heart J 106*:55–59
9. Shem-Tov A, Schneeweiss A, Motro M, Neufeld HN (1982) Clinical presentation and natural history of mild discrete subaortic stenosis: follow-up for 1–17 years. *Circulation 66*:509–512
10. Vogel M, Freedom RM, Brand A, Trusler GA, Williams WG, Rose RD (1983) Ventricular septal defect and subaortic stenosis: an analysis of 41 patients. *Am J Cardiol 52*:1258–1263
11. Wright GB, Keane JF, Nadas AS, Bernhard WF, Castaneda AR (1983) Fixed subaortic stenosis in the young: medical and surgical course in 83 patients. *Am J Cardiol 52*:830–835