

## EDITORIAL COMMENT

# Natural History of Discrete Subvalvar Aortic Stenosis: Management Implications\*

Welton M. Gersony, MD  
*New York, New York*

Discrete subvalvar aortic stenosis (DSVAS) represents a unique cardiac lesion. As compared to most other congenital heart defects, DSVAS is virtually never recognized in early infancy, but appears to be an “acquired” lesion, albeit with anatomic precursors (1–4). The stenosis is caused by a fibrous ridge in the left ventricular (LV) outflow tract just proximal to the aortic valve. An abnormal angle between the muscular and conal ventricular septum appears to be an important causative factor, but a definitive etiology has not been established (5–7). Some investigators have considered the disease to be a form of cardiomyopathy (8), and there are patients with DSVAS who develop an unusually profound LV myocardial hypertrophic response. However, more often there does not appear to be a global cardiomyopathic component. In the vast majority of cases, LV hypertrophy regresses after relief of outflow obstruction, as would be expected in a patient with valvar aortic stenosis.

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This condition occurs in patients with associated congenital heart disease, including ventricular septal defect, coarctation of the aorta, interrupted aortic arch, atrioventricular canal and others (2,9). The lesion may develop in unoperated patients with these defects, but also may appear and progress significantly after surgical correction of the associated defect. In addition, DSVAS presents as a primary defect.

Surgical management of DSVAS consists of removal of the circumferential fibrous obstructive ridge with or without septal myectomy. Early reports described complications of heart block and mitral valve injury as well as incomplete relief and/or recurrence of obstruction with reoperation rates of 20% or more (9–12). In recent years, circumferential enucleation of the fibrous ridge by blunt dissection (13), occasionally with septal myectomy, appears to provide the most efficient relief of obstruction. Nevertheless, recurrence still remains a significant problem, especially in the presence of a predisposing associated congenital heart defect.

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From the College of Physicians and Surgeons, Columbia University, New York, New York.

Most reports concerning DSVAS relate to the disease in infants and children. In the 1970s it was documented by a number of investigators that, once established, DSVAS progressed rapidly to severe LV outflow obstruction, with all of its hemodynamic consequences (14–17). These early cases defined management, which at most centers was to intervene surgically when gradients were 30 mm or more regardless of other aspects of the clinical and echocardiographic profile. Indications for DSVAS surgery became more aggressive than for valvar aortic stenosis, which is less likely to progress rapidly in children prior to puberty. However, it is important to be aware that in the pre-echocardiographic era, “mild” or “moderate” obstruction based on direct pressure measurements at cardiac catheterization often reflected rather significant obstruction. The average preoperative gradient was far higher than is usually reported in the modern era, when echocardiography identifies DSVAS at a much earlier stage.

The present-day management policy at most centers continues to require a significant LV-aortic gradient before surgery is recommended. However, operative indications have been expanded by some groups (10,18–20). Rapid surgical intervention is advocated in infants and children when a subaortic ridge is shown on echocardiographic study even when there is no significant predicted gradient. This rationale is based on the “inevitability” of rapid progression as well as concerns about eventual aortic valve injury and dysfunction secondary to unrelieved DSVAS, even in its mildest form. More recently, however, it has been demonstrated in children that the rate of progression of mild DSVAS is variable, and many patients remain stable for years (2,21). Prevention of aortic valve injury is also advanced as an indication for early surgery. This has been controversial as many cardiologists have observed more aortic insufficiency after DSVAS surgery than before. Centers that require at least a 30-mm gradient before advocating surgical intervention consider left ventricular outflow obstruction to be the key factor. Among patients with trivial or mild stenosis, progression of obstruction must be demonstrated before surgery is recommended.

Although considerations regarding surgical management and timing of surgical interventions are predicated on potential long-term effects, there has been little in the medical literature regarding the disease in adult patients (22–25), and early reports included small numbers of patients. In this issue of the *Journal*, Oliver et al. (26) present the most extensive data to date on the long-term aspects of DSVAS in adults and, thus, provide insights into some of the management issues and controversies. These investigators describe 134 adults with discrete subvalvar aortic stenosis, including 29 patients who were operated on in adult life, 41 operated on during childhood and 64 who had not had surgery. In the latter group of unoperated patients, LV outflow tract obstruction increased from

39.2 ± 28 mm Hg to 46.8 ± 34 mm Hg after a mean follow-up period of 4.8 ± 1.8 years in 25 patients. The investigators noted that in patients older than 50 years of age, severity increased significantly, but in young and middle-aged adults, there was no significant change during the follow-up period. It also can be noted from their data that the older patients had more severe gradients at the initial echocardiogram. In the overall group, 4 of 8 patients with gradients >50 mm Hg significantly increased the severity of obstruction, whereas only 3 of 18 patients with initial gradients under 50 mm Hg showed significant, albeit mild, progression of the subaortic gradients (19, 33 and 19 mm Hg, respectively). Those with gradients over 50 mm Hg would normally be expected to have surgical relief of obstruction at the time of the initial echocardiogram. Echocardiographic aortic regurgitation was noted in all patients who had had surgical intervention and in 75% of unoperated patients. Although not severe, the degree of regurgitation was significantly greater in the operated patients.

The conclusions from this study can be summarized as follows:

- Mild DSVAS most often remains stable or progresses slowly in adults.
- Aortic regurgitation is common, but is rarely hemodynamically significant.
- Aortic regurgitation is more prominent in patients *after* surgical intervention than in unoperated patients.

Although much remains to be learned about DSVAS, a number of management principles emerge from both the Oliver et al. (26) unique large experience in adults and the more recent pediatric reports. The major issue in determining whether surgery should be carried out is the severity of the LV outflow tract obstruction and the age of the patient. The disease is often rapidly progressive in infancy and early childhood, but it can also be stable for years; some patients reaching adult life with only mild obstruction. Aortic regurgitation associated with DSVAS is usually mild and is rarely progressive either in unoperated or postsurgical patients. It appears that adult patients who have had surgical repair of discrete LV outflow tract obstruction are more likely to have significant aortic regurgitation than unoperated patients with mild gradients.

At the present time, a reasonable approach to management of DSVAS might include the following guidelines:

1. Any patient with DSVAS who has severe obstruction requires surgical intervention.
2. An infant or young child with a well-defined 30-mm or more gradient should have removal of the subvalvar obstruction. Pediatric patients with <30-mm gradient and no significant LV hypertrophy must be followed closely for progression, especially in the first several years of life.
3. Older children with mild gradients (<30 mm) may be

followed until there is documented significant echocardiographic or catheterization progression of the obstruction, usually with supportive clinical and echocardiographic findings. The inexactness of predictions of gradient by color Doppler must always be considered in the decision-making process.

4. Unoperated adult patients with stable predicted gradients <50 mm without significant LV hypertrophy must also be followed closely, as some of these patients will eventually require surgery.
5. "Prevention" of aortic regurgitation is not a criteria for surgery in a patient with DSVAS in whom obstruction is insignificant.

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**Reprint requests and correspondence:** Dr. Welton M. Gersony, Children's Hospital of New York, 3959 Broadway 2-North, New York, New York 10032. E-mail: ds200@columbia.edu.

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