OBJECTIVES

We sought to determine the prevalence and rate of progression of left ventricular outflow tract obstruction (LVOTO) and aortic regurgitation (AR) in adults with discrete subaortic stenosis (DSS).

BACKGROUND

Discrete subaortic stenosis is an uncommon form of LVOTO, with rapid hemodynamic progression in children, but the prevalence and rate of progression in adults have not been studied so far.

METHODS

The prevalence of DSS was determined in 2,057 consecutive adults diagnosed with congenital heart disease (CHD). The relationship between LVOTO on Doppler echocardiography and patient age was analyzed. Sequential changes in LVOTO and AR were determined for patients with two or more Doppler echocardiograms obtained with at least a two-year interval.

RESULTS

A total of 134 adults (mean age 31 ± 17 years) were diagnosed with DSS. The prevalence was 6.5% for all adults with CHD. Sixty patients (44%) had other associated CHD. The mean age of 29 patients who had undergone an operation for DSS during their adult life (56 ± 15 years) was significantly higher than that of 64 patients (27 ± 13 years) who had not required a surgical intervention (p < 0.0001). A significant relationship between LVOTO and patient age (r = 0.61, p < 0.0001) was found: 21 ± 16 mm Hg in patients <25 years old, 51 ± 47 mm Hg for those between 25 and 50 years old, and 78 ± 36 mm Hg for those >50 years old. The LVOTO increased from 39.2 ± 28 to 46.8 ± 34 mm Hg (p = 0.01) during a mean follow-up of 4.8 ± 1.8 years in 25 patients. The slope of the change in LVOTO was 2.25 ± 4.7 mm Hg per year of follow-up. Aortic regurgitation was detected by color Doppler imaging in 109 patients (81%), but it was hemodynamically significant in <20%. An increase in the mean degree of AR over time was not significant (baseline: 1.3 ± 0.8; follow-up: 1.5 ± 0.9; p = 0.096).

CONCLUSIONS

The prevalence of DSS is increasing in adults due to the greater number of repaired CHDs that develop into evolutive DSS. In contrast to infants and children, adults with DSS show a slow rate of LVOTO progression. Aortic regurgitation is a common but usually mild and nonprogressive consequence. The current indications for surgical intervention should be revised. (J Am Coll Cardiol 2001;38:835–42) © 2001 by the American College of Cardiology

Discrete subaortic stenosis (DSS) is considered an “acquired” cardiac defect of postnatal development, as it does not appear during embryologic development of the heart and occurs very infrequently in the neonatal period (1). It usually appears after the first year of life and causes left ventricular outflow tract obstruction (LVOTO) of rapid hemodynamic progression (2–6). Due to its associated rapid clinical impairment during the first years of life, DSS has been considered, for a long time, to appear only in infancy and childhood (7–11), and there are few published reports on the evolution of DSS during adult life (12–15). Although rheologic factors affecting the onset and progression of DSS have been the subject of recent studies (16–21), no data have been published establishing whether the obstruction is also progressive during adult life. Aortic regurgitation (AR) is a common consequence of DSS, and the “prevention” of progressive AR by early operation has been advocated (22–24), but the rate of progression of AR in adults has not yet been established. Doppler echocardiography (DE) is a very useful tool for the evaluation and follow-up of LVOTO and AR in patients with valvular or subvalvular aortic stenosis (25). The main purposes of this study were to evaluate the prevalence of DSS in a large series of adult patients with CHD, as determined by DE, and to determine the rate of progression of LVOTO and AR in patients with DSS who did not have an operation during infancy or childhood.

METHODS

Study group. From the data base of the Adult Congenital Heart Diseases Unit in La Paz University Hospital, Madrid,
Abbreviations and Acronyms

AR = aortic regurgitation
CHD = congenital heart disease
CI = confidence interval
DE = Doppler echocardiography
DSS = discrete subaortic stenosis
LVOT = left ventricular outflow tract
LVOTO = left ventricular outflow tract obstruction

all patients diagnosed with DSS who were evaluated from January 1990 to December 1999 were identified. Only patients >15 years old and who had DSS diagnosed either by DE or during surgical intervention were included in the study. Discrete subaortic stenosis was diagnosed, by means of DE, when a fixed subvalvular obstruction causing subaortic flow acceleration detectable by color Doppler imaging was present. Patients having either the S arrangement of the ventricular septum protruding at the LVOT or a marked septal protrusion without a fixed subvalvular stenosis were not included. Patients with dynamic subaortic obstruction associated, or not, with hypertrophic cardiomyopathy and those with LVOTO caused by either accessory mitral valve tissue or the support system of mitral valve prosthesis were also excluded. Only patients fulfilling the anatomic criteria of Kelly's classification (26), with either short- or long-segment subaortic obstruction according to the morphologic classification of Choi and Sullivan (5), were included in the study.

All patients included had at least one follow-up visit and a DE study. To determine the prevalence of DSS among adults with CHD, we determined the number of adult patients for whom a DE had been obtained during the same period and who had been diagnosed with CHD. Patients with DSS were classified into three groups: group A included all patients who had undergone an operation for DSS for the first time during adult life; group B included all patients who survived naturally until adult age, who remained under clinical surveillance but without an operation for DSS; and group C included all patients who had an operation for DSS in the first 15 years of life. Age, gender distribution and frequency of other associated congenital or acquired heart diseases were determined for each of the three groups.

LVOTO. Comprehensive two-dimensional and Doppler echocardiographic assessments were performed using a Sonos 1000, 2500 or 5500 ultrasound system (Hewlett Packard, Andover, Massachusetts) fitted with a 2.5-MHz or multifrequency probe. The LVOTO was evaluated by continuous wave Doppler echocardiography obtained from the apical position, using a 2-MHz imageless probe. Peak pressure gradient was determined by Bernoulli’s simplified equation (27).

To determine the progression of LVOTO during adulthood, the relationship between age and LVOTO (presurgical LVOTO values in patients with a surgical interven-
(51%), the difference was not statistically significant. The mean age in group A (56 ± 15 years) was significantly higher than that in groups B (27 ± 13 years, p < 0.0001) and C (21 ± 4 years, p < 0.0001).

**Associated heart diseases.** Table 1 also shows the associated congenital or acquired heart diseases in the three groups. There were a total of 60 patients (44%) with another CHD. The percentage of patients with an associated CHD was significantly lower in group A (7%) than in groups B (61%, p < 0.0001) and C (42%, p < 0.0001). Nevertheless, there were three patients in group A (10%) with associated rheumatic mitral valve disease, and none in groups B and C.

The most frequently associated CHDs were ventricular septal defect (n = 20), aortic coarctation (n = 17), atrioventricular septal defect (n = 8), patent ductus arteriosus (n = 6), bicuspid aortic valve stenosis (n = 3) and double-outlet right ventricle (n = 3). In 49 patients (82%), the associated heart subaortic stenosis had been repaired during childhood, and DSS remained as noncorrected residua. Of the 93 patients in groups A and B whose DSS had not been surgically corrected during childhood, there were 43 cases with associated CHD and 50 cases with isolated DSS. Patients with associated CHD were younger (24 ± 8 vs. 46 ± 20 years, p < 0.001) and had less LVOTO (26 ± 28 vs. 63 ± 42 mm Hg, p < 0.001) than those with isolated DSS (Fig. 1).

**Progression of LVOTO.** Figure 2 shows a significant correlation (r = 0.61, p < 0.0001) between LVOTO and age in 93 patients with natural evolution of DSS into adult life (groups A and B). When patients were analyzed by age groups, a progressive increase in LVOTO was observed (p < 0.001). Patients <25 years old had LVOTO of 21 ± 16 mm Hg, those between 25 and 50 years old with 51 ± 47 mm Hg (p = 0.002) and those >50 years old with 78 ± 36 mm Hg (p < 0.02) with respect to the middle-age group (Fig. 3).

Table 2 shows the change in LVOTO in 25 patients who had at least two DE assessments separated by an average interval of 4.8 ± 1.8 years (range 2 to 8). During this interval, the variation of changes in LVOTO observed was wide, ranging from −10 to 41 mm Hg, but there was a significant increase in mean LVOTO, from 39.2 ± 28 to 46.8 ± 34 mm Hg (average increase 7.6 ± 14 mm Hg, 95% confidence interval [CI] 1.6 to 13.6, p = 0.01). The slope of change in LVOTO was 2.25 ± 4.7 mm Hg per year of follow-up. Changes in LVOTO over time correlated with age (r = 0.55, p = 0.004), but not with the baseline LVOTO (r = 0.22, p = 0.29). The mean increase in LVOTO in patients <50 years old was 19.5 ± 16 mm Hg (95% CI 5.7 to 33.2, p = 0.01), but there were no significant changes in LVOTO in patients <50 years old (95% CI −3.0 to 7.0, p = 0.4) (Fig. 4).

**AR.** This was detected by color Doppler imaging in 109 patients (81%). The grade of AR severity was 1 (trace to mild) in 36 patients (27%), 2 (mild to moderate) in 49 patients (37%) and 3 (moderate to severe) in 25 patients (19%). All patients in group C, but only 75% of patients in

**Table 1.** Mean Age, Gender Distribution and Associated Congenital or Acquired Heart Diseases in Patients With Discrete Subaortic Stenosis Who Had an Operation During Adulthood (Group A), No Operation (Group B) and an Operation During Childhood (Group C)

<table>
<thead>
<tr>
<th></th>
<th>Group A (n = 29)</th>
<th>Group B (n = 64)</th>
<th>Group C (n = 41)</th>
<th>Overall (n = 134)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender (% male)</td>
<td>41%</td>
<td>58%</td>
<td>51%</td>
<td>52%</td>
</tr>
<tr>
<td>Age (years)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>24 ± 8</td>
<td>27 ± 13</td>
<td>21 ± 4</td>
<td>31 ± 17</td>
<td></td>
</tr>
<tr>
<td>56 ± 15*</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Associated CHD</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>0</td>
<td>2</td>
<td>0</td>
<td>2 (7%)</td>
</tr>
<tr>
<td>Aortic coarctation</td>
<td>6</td>
<td>11</td>
<td>17</td>
<td></td>
</tr>
<tr>
<td>Atrioventricular septal defect</td>
<td>2</td>
<td>4</td>
<td>2</td>
<td>8</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>3</td>
<td>3</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>Aortic valve stenosis (bicuspid)</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Double-outlet right ventricle</td>
<td>2</td>
<td>1</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Other</td>
<td>3</td>
<td>0</td>
<td>3</td>
<td>3 (2%)</td>
</tr>
<tr>
<td>Rheumatic mitral valve disease</td>
<td>0</td>
<td>0</td>
<td>3</td>
<td></td>
</tr>
</tbody>
</table>

*p < 0.01. Data are presented as the number (%) of patients or mean value ± SD.

**CHD = congenital heart disease.**

**Figure 1.** Mean age and left ventricular outflow tract obstruction (LVOTO) in naturally surviving patients with isolated discrete subaortic stenosis (non-CHD) or discrete subaortic stenosis associated with other congenital heart disease (CHD).
groups A and B, had detectable AR (Fig. 5). The mean AR degree was 1.9 ± 0.7 in group C, compared with 1.4 ± 1.0 in groups A and B (p = 0.003). The AR degree was significantly related to LVOTO (p < 0.001), but not to age (p = 0.055). The increase in mean AR degree over time in patients with at least two DE assessments was not significant (baseline: 1.3 ± 0.8; follow-up: 1.5 ± 0.9; p = 0.096). Four of 17 patients in groups A and B with moderate-to-severe AR had aortic valve infective endocarditis.

**DISCUSSION**

**Prevalence of DSS in adult life.** Discrete subaortic stenosis has been considered an uncommon form of LVOTO, appearing almost exclusively in infancy or childhood and very infrequently in adulthood (7–10). In contrast, our data show that DSS is a relatively frequent heart disease in adult patients, with a prevalence of 6.5% of all adult CHDs studied at our institution during a 10-year period (29).

The increased prevalence of DSS in adults could be explained by an easier and earlier diagnosis of this lesion. Although the diagnosis of DSS has traditionally been difficult to differentiate from valvular aortic stenosis, DE has contributed to a precise identification of this cardiac abnormality and a correct assessment of the different anatomic patterns (30–34). Using DE, it is even possible to diagnose small subaortic membranes causing acceleration of the LVOT flow, but without a hemodynamically significant pressure gradient. Making a differential diagnosis of DSS versus obstructive hypertrophic cardiomyopathy can be more difficult (35), especially considering that patients with DSS can develop asymmetrical septal hypertrophy and secondary dynamic subaortic obstruction (36–38). Severe septal hypertrophy and dynamic obstruction of the LVOT can mask the existence of a subaortic membrane and cause a false diagnosis of obstructive hypertrophic cardiomyopathy. Although conventional DE may be inconclusive, transesophageal echocardiography is much more reliable for the determination of a subaortic membrane masked by the hypertrophied and prominent ventricular septum (39–41) (Fig. 6).

**Association with other CHDs.** It is also possible that an actual increase in the prevalence of this cardiac condition in adults is occurring. Our data show that DSS was associated with another CHD in 44% of patients. The two most frequently associated cardiac malformations were ventricular septal defect and aortic coarctation, but DSS was also associated with atrioventricular septal defect, patent ductus arteriosus, bicuspid aortic valve or double-outlet right ventricle. In most cases, DSS appeared as a secondary lesion years after the surgical repair of the associated cardiac malformation. The presence of DSS after surgical correction of aortic coarctation (42,43), ventricular septal defect (44), patent ductus arteriosus (45), ostium primum atrial septal defect (46), congenital valvular aortic stenosis (47), double-outlet right ventricle (48) and other diverse CHDs (3) has been described. It is possible that the 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.

**DSS after acquired heart diseases.** It has recently been shown that DSS can also develop in adulthood after...
acquired heart diseases. Khoshnevis et al. (49) reported three patients with rheumatic heart disease who developed severe stenosis by a subaortic membrane many years after the prosthetic replacement of the mitral valve. Our series also included three patients with rheumatic mitral valve disease and DSS. One of them had a previous mitral commissurotomy performed six years before the subaortic membrane was diagnosed. The other two patients were diagnosed with severe DSS and rheumatic mitral stenosis at 35 and 64 years of age, respectively. In the three patients, a subaortic membrane was confirmed and removed during the operation. In any case, the development of DSS caused by acquired mitral valve disease during adulthood must be exceptional.

Progression of DSS. The progressive nature of DSS obstruction has been well established in children (1–11). The stenosis progresses so rapidly that Brock (50) thought that it usually caused the death of patients who did not have an operation before they could reach adulthood. The present study shows that subaortic obstruction can also have a progressive nature during adult life, but in contrast to what happens in children, LVOTO progresses slowly along several decades. A number of findings support this statement: 1) the mean age of patients requiring an operation was almost 30 years older than that of patients remaining under clinical surveillance with no operation; 2) there was a significant correlation between patient age and LVOTO measured by DE; 3) the LVOTO value went from 21 ± 16 mm Hg in patients <25 years old to 51 ± 47 mm Hg in those between 25 and 50 years old and to 78 ± 36 mm Hg in those >50 years old; and 4) there was a small but significant increase in LVOTO over time in sequential studies covering intervals from two to eight years, with a slope of change in LVOTO of 2.25 ± 4.7 mm Hg per year of follow-up.

Table 2. Changes in Left Ventricular Outflow Tract Obstruction and Aortic Regurgitation Degree in 25 Patients Who Had Two Doppler Echocardiographic Assessments Separated by an Interval of ≥2 Years

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age (yrs)</th>
<th>Initial LVOTO (mm Hg)</th>
<th>Follow-Up LVOTO (mm Hg)</th>
<th>Initial AR Grade</th>
<th>Follow-Up AR Grade</th>
<th>Interval (years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>23</td>
<td>16</td>
<td>19</td>
<td>1</td>
<td>1</td>
<td>8</td>
</tr>
<tr>
<td>2</td>
<td>33</td>
<td>94</td>
<td>94</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>3</td>
<td>50</td>
<td>60</td>
<td>99</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>4</td>
<td>23</td>
<td>5</td>
<td>10</td>
<td>1</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>5</td>
<td>21</td>
<td>5</td>
<td>11</td>
<td>2</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>6</td>
<td>25</td>
<td>45</td>
<td>35</td>
<td>1</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>7</td>
<td>25</td>
<td>17</td>
<td>12</td>
<td>0</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>8</td>
<td>70</td>
<td>50</td>
<td>74</td>
<td>1</td>
<td>1</td>
<td>6</td>
</tr>
<tr>
<td>9</td>
<td>33</td>
<td>35</td>
<td>40</td>
<td>2</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>10</td>
<td>22</td>
<td>45</td>
<td>35</td>
<td>1</td>
<td>1</td>
<td>6</td>
</tr>
<tr>
<td>11</td>
<td>63</td>
<td>97</td>
<td>91</td>
<td>1</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>12</td>
<td>25</td>
<td>5</td>
<td>11</td>
<td>2</td>
<td>1</td>
<td>6</td>
</tr>
<tr>
<td>13</td>
<td>19</td>
<td>20</td>
<td>20</td>
<td>3</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>14</td>
<td>57</td>
<td>46</td>
<td>65</td>
<td>0</td>
<td>0</td>
<td>6</td>
</tr>
<tr>
<td>15</td>
<td>24</td>
<td>30</td>
<td>63</td>
<td>1</td>
<td>0</td>
<td>6</td>
</tr>
<tr>
<td>16</td>
<td>23</td>
<td>25</td>
<td>30</td>
<td>0</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>17</td>
<td>24</td>
<td>29</td>
<td>29</td>
<td>2</td>
<td>2</td>
<td>7</td>
</tr>
<tr>
<td>18</td>
<td>25</td>
<td>34</td>
<td>25</td>
<td>0</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>19</td>
<td>26</td>
<td>5</td>
<td>10</td>
<td>1</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>20</td>
<td>72</td>
<td>70</td>
<td>111</td>
<td>2</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>21</td>
<td>27</td>
<td>54</td>
<td>57</td>
<td>1</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>22</td>
<td>46</td>
<td>50</td>
<td>50</td>
<td>2</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>23</td>
<td>33</td>
<td>20</td>
<td>17</td>
<td>2</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>24</td>
<td>59</td>
<td>34</td>
<td>53</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>25</td>
<td>64</td>
<td>100</td>
<td>120</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

Mean ± SD 37.2 ± 17 39.2 ± 28 46.8 ± 34* 1.3 ± 0.8 1.5 ± 0.9† 4.8 ± 1.8

*p = 0.001 compared with initial LVOTO value; †p = NS compared with initial AR grade.

AR = aortic regurgitation; LVOTO = left ventricular outflow tract obstruction.

Figure 4. Comparison between baseline (initial) and follow-up left ventricular outflow tract obstruction (LVOTO) in patients ≥50 years old or <50 years old.
Mechanisms of DSS progression. The mechanisms of DSS development and progression have recently been studied in children (16–21). Most of the investigators accept that abnormal fluid dynamic forces at the LVOT level can cause septal shear stress. Different morphologic alterations can cause changes in fluid dynamic factors that increase septal shear stress. Both retrospective and prospective studies of children developing DSS have found an increased mitral-aortic fibrous distance, a steeper aortoseptal angle and a narrower LVOT. Minor morphologic changes in the LVOT can produce marked modifications in dynamic forces and septal shear stress. Cellular flow studies have shown that increased septal shear stress triggers a basic genetic predisposition to developing cellular growth factors. These morphologic changes act both in the onset and in the progression of the subaortic obstruction, but the cellular proliferation response in the child is probably very different from that in the adult. It is likely that the earlier in life the septal shear stress is increased, the more intense the response and the more rapid the progression of the LVOTO. Although progression of DSS obstruction can be very rapid in infants and small children at vulnerable periods of development (11), our data show that the obstruction progresses much more slowly during adult life.

AR. This occurs in >50% of patients with DSS (22). Damage to the aortic valve due to the subvalvular systolic jet appears to be the main cause, but direct extension of subvalvular fibrous tissue into the aortic valve has been advocated (51). Aortic regurgitation is usually mild in children diagnosed with DSS (52), but progression of the degree of AR in patients who have not had an operation has been reported (53). Our data show that AR, as detected by color Doppler imaging, is frequently found in adults with DSS (81%), but is hemodynamically significant (moderate to severe) in <20% of the patients. Previous reports have shown a relationship between the severity of stenosis and AR in children (52,53), and our results extend this relationship into adulthood. However, significant progression of AR during adult life was not found; there was no significant relationship between AR and age and there was no significant increase in AR over time. The risk of endocarditis in DSS is especially high (8,22), and it contributed to severe AR in four patients in this series.

Surgical implications. Early surgical repair of DSS has been advocated to prevent rapid progression of LVOTO and development of significant AR (22–24,54,55). Some investigators advise surgical repair, irrespective of the gradient, to reduce the damage to the aortic valve (56), but studies advocating early surgical repair are based on relatively small numbers of pediatric patients and short follow-up. No data on the benefits of early operation in adolescents and adults have been reported. The present study shows that LVOTO also has a progressive nature in adult patients, but the obstruction progresses very slowly along several decades, although it may be accelerated after the fifth decade of life. This pattern of progression makes the average age for surgical repair of DSS in adults to be >50 years old, with its
clinical manifestation in the 60s or even 70s is not infrequent. Aortic regurgitation is very common in adults, and it is related to the severity of LVOTO, but in most cases, AR is a mild and nonprogressive consequence of DSS. Moreover, the severity of AR in DSS repaired during childhood was significantly greater than that in patients who survived naturally into adulthood. Data from this study and others (57) show that the benefits of early surgical repair in adolescents and adults should be questioned. Surgical decisions in adult patients diagnosed with DSS should be based not only on the anatomic finding of subvalvular stenosis, but also on the clinical evaluation, left ventricular hypertrophy, systolic function and severity of LVOTO and aortic insufficiency.

Study limitations. The retrospective nature of this study may be considered an important limitation, and data should be confirmed by prospective studies following the evolution of young patients with mild DSS over several decades. The hemodynamic data of this study were based only on DE assessment, and data from cardiac catheterization were not analyzed. A fair correlation between DE and catheter-based LVOTO and AR has previously been reported (25,27), but Doppler flow peak velocity through the LVOT depends not only on the subaortic pressure gradient, but also on the severity of AR. Progression in the Doppler LVOT pressure gradient may be produced by an increase in the severity of either subaortic obstruction or aortic regurgitation, or both. Pressure recovery is another cause of increased Doppler flow peak velocity that is out of proportion to the catheter-measured LVOTO (58). In any case, this study shows that progression in the hemodynamic consequences of DSS during adult life is relatively slow.

Conclusions. Overall, the data from this study suggest that DSS is currently a relatively frequent heart disease in adults, and its prevalence may be increasing due to the greater number of repaired CHDs, which may enhance the evolution of DSS. In most cases, DSS is a primary lesion in patients with different cardiac conditions that have small morphologic or functional abnormalities in the LVOT in common, which increase septal shear stress and stimulate cellular proliferation in the interventricular septum and surroundings structures. In rare circumstances, DSS may also appear as a response to an acquired disease, such as rheumatic mitral valve disease. In adult patients, DSS progresses in severity during a lifetime, but in contrast to what happens in children, progression of the obstruction occurs slowly, over several decades. Aortic regurgitation is very common, but usually it is a mild and nonprogressive consequence of DSS, although severe AR may occur after infective endocarditis. Surgical repair of DSS in children does not prevent AR development in adults. The current indications for surgical intervention of DSS in adults need to be revised.

REFERENCES


