In a prospective study made between 1 September 1996 and 31 December 2001, pediatric patients with a diagnosis of supravalvular aortic stenosis confirmed by a reduction in the aortic inner diameter and a gradient $\geq 50$ mmHg were detected.

Of 83 patients with aortic stenosis, only 7 (8.4%) had supravalvular aortic stenosis. All 7 patients underwent surgical treatment consisting of resection of fibrous tissue and reconstruction of the ascending aorta with a preclotted Dacron patch. One patient with severe, diffuse stenosis died and another had perioperative heart failure, cardiac arrest and reversible neurological sequelae. A significant decrease in the postoperative gradient was obtained ($p < 0.05$). At present all surviving patients are free of symptoms.

It was concluded that supravalvular aortic stenosis is infrequent in México. In our experience, surgical treatment produced good results and success depended on the magnitude and type of stenosis.

**Key words:** Williams syndrome. Supravalvular stenosis. Aortic surgery.

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**INTRODUCTION**

Supravalvular aortic stenosis is an obstruction caused by a narrowing in the aortic lumen immediately at the tip of the origin of the coronary vessels adjacent to the aortic valve. Mencarelli first described this condition in 1930, and its association with other facial de-
In the international literature, including that of Latin America, reveals a low occurrence of supravalvular aortic stenosis and in an extensive review of 653 cases of congenital cardiopathy by Vizcaíno and Ortega in Spain, the authors found 9 cases of idiopathic hypercalcemia, of which 5 were associated with aortic supravalvular stenosis.

**MATERIAL AND METHODS**

A prospective study was performed from September 1, 1996, through December 31, 2001, to identify and evaluate the results from pediatric patients (from newborns to 17 year-old individuals in accordance with the institutional guidelines) with aortic supravalvular stenosis who underwent surgical treatment.

Diagnosis was made using images indicative of a narrowing in the aortic lumen and a discrepancy in pressure between the left ventricle and aorta pre- and post-stenosis (≥50 mm Hg), on transthoracic M-mode and 2-dimensional echocardiography. We used cardiac catheterization, in patients whose clinical picture showed indications typical of aortic supravalvular stenosis associated with Williams syndrome such as: round forhead; depressed temporal bones; epicantus; depressed nasal bridge; anteverted nasal openings with prominent superior lip (elfin facies); and other characteristics. Correction of the defect was performed under general anesthesia, with cardiopulmonary shunt (CPS) implantation via mid sternotomy, induction of moderate hypothermia (28 °C to 32 °C), cardiac arrest induced with the administration of crystalloid cardioplegic solution at 4 °C, and the creation of a lateral longitudinal aortotomy over the defect and up to 2 mm over the noncoronary valve, resection of the redundant intraluminal tissue, and placement of a Dacron precoagulated patch to widen the diameter of the anastomosed aortic lumen with continuous 4-0 caliber polypropylene monofilament suture.

The patients were kept in a postsurgical therapy unit for immediate care, and during followup the supravalvular gradient values were measured by means of echocardiography immediately after surgery and at 6 months; the latter was compared to the initial preoperative value.

**Statistical analysis**

We used descriptive statistical analysis to characterize the group of patients and the Wilcoxon test to compare the preoperative and postoperative gradients, with a significant value considered to be \( P < 0.05 \).

**RESULTS**

During the evaluation period, 1390 pediatric patients of both sexes, with an average age of 6.9 years (range, 5 days in age to 17 years in age) who underwent surgery for congenital cardiopathy were studied. Aortic stenosis was present in 83 cases, and diaphragm-type subaortic stenosis predominated, followed by valvular stenosis; supravalvular stenosis was found in only 7 cases (8.4% of the 3 obstruction sites of the left ventricular outflow tract), which comprised 0.5% of the total number of pediatric patients (Table 1) studied.

In the patients with supravalvular stenosis, female sex predominated (4 cases). The youngest patient was 4 years of age and the oldest was 15 years of age. Five patients presented with the Williams syndrome. One patient with diffuse stenosis had undergone intervention 2 years earlier and due to a residual supravalvular pressure gradient required re-operation (Table 2). Another patient presented with congenital rubella.

The predominant type of lesion was the hourglass type of localized stenosis (5 of 7 cases), and diffuse stenosis in 2 cases, 1 of which affected the aortic arch and the descending aorta immediately below the emergence of the left subclavian artery (Table 2); this was the only case which required using the femoral approach to place the arterial CPS cannula. Ischemia time was 56.5 minutes±27 minutes and CPS time was 84.2 minutes±37.8 minutes; in the 2 cases with diffuse stenosis the myocardial ischemia time and CPS time were greater due to the complexity of the correction. Amount of time in postsurgery therapy was an average of 3 days, with a range of 20 hours to 5 days, and the total length of hospital stay was an average of 8.5 days, with a range of 7 to 10 days.

In the 7 cases surgical treatment consisted of resection of the intraluminal tissue and widening of the aortic diameter with a precoagulated Dacron graft, with which a significant decrease \( (P < 0.05) \) in the preoperative gradient (from 86.2 mm Hg±19.6 mm Hg to 12 mm Hg±8.7 mm Hg) was achieved at 3 and 6 months postsurgery.

The maximum followup period was 58 months and the minimum was 6 months at the time this manuscript was submitted.

**TABLE 1. Frequency of cases of supravalvular stenosis**

<table>
<thead>
<tr>
<th>Year</th>
<th>Total number of cases</th>
<th>Aortic stenosis</th>
<th>Supravalvular stenosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1996</td>
<td>81</td>
<td>7</td>
<td>-</td>
</tr>
<tr>
<td>1997</td>
<td>288</td>
<td>9</td>
<td>1</td>
</tr>
<tr>
<td>1998</td>
<td>216</td>
<td>13</td>
<td>1</td>
</tr>
<tr>
<td>1999</td>
<td>281</td>
<td>21</td>
<td>1</td>
</tr>
<tr>
<td>2000</td>
<td>275</td>
<td>21</td>
<td>2</td>
</tr>
<tr>
<td>2001</td>
<td>228</td>
<td>12</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>1390</td>
<td>83</td>
<td>7</td>
</tr>
</tbody>
</table>
TABLE 2. Patient characteristics

<table>
<thead>
<tr>
<th>Case</th>
<th>Age, years</th>
<th>Sex</th>
<th>Δpre mm Hg</th>
<th>Δ post mm Hg</th>
<th>Findings</th>
<th>CPS, min</th>
<th>AoC, min</th>
<th>Cirugía</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>10</td>
<td>F</td>
<td>90</td>
<td>20</td>
<td>Localized supravalvular stenosis</td>
<td>94</td>
<td>34</td>
<td>Tissue resection and graft</td>
</tr>
<tr>
<td>2</td>
<td>4</td>
<td>F</td>
<td>81</td>
<td>0</td>
<td>Aortic valve stenosis, subaortic stenosis and localized supravalvular stenosis</td>
<td>82</td>
<td>51</td>
<td>Tissue and graft resection and commissurotomy</td>
</tr>
<tr>
<td>3</td>
<td>11</td>
<td>M</td>
<td>85</td>
<td>11</td>
<td>Supravalvular ring and localized supravalvular stenosis</td>
<td>46</td>
<td>20</td>
<td>Ring and graft resection</td>
</tr>
<tr>
<td>4</td>
<td>11</td>
<td>F*</td>
<td>100</td>
<td>22</td>
<td>Diffuse supravalvular stenosis</td>
<td>104</td>
<td>65</td>
<td>Tissue and graft resection</td>
</tr>
<tr>
<td>5</td>
<td>15</td>
<td>F</td>
<td>56</td>
<td>15</td>
<td>Localized supravalvular stenosis and valvular aortic stenosis</td>
<td>67</td>
<td>45</td>
<td>Commissurotomy and graft</td>
</tr>
<tr>
<td>6</td>
<td>13</td>
<td>M</td>
<td>118</td>
<td>-</td>
<td>Diffuse supravalvular stenosis, aortic arch and descending aorta</td>
<td>153</td>
<td>88</td>
<td>Tissue and graft resection</td>
</tr>
<tr>
<td>7</td>
<td>11</td>
<td>M</td>
<td>74</td>
<td>4</td>
<td>Localized supravalvular stenosis</td>
<td>44</td>
<td>93</td>
<td>Ring and graft resection</td>
</tr>
</tbody>
</table>

Δpre indicates preoperative gradient; Δpost, post surgery gradient; CPS, cardiopulmonary shunt; AoC, aortic clamping; graft, widening with Dacron graft. *Case with a history of surgery due to supravalvular stenosis and residual gradient.

One patient with diffuse stenosis that involved the ascending aorta, the aortic arch, and the descending aorta immediately at the emergence of the left subclavian artery required 35 minutes of total circulatory arrest in deep hypothermia, and there were difficulties removing the CPS after performing the correction. After a postoperative period of 20 hours with hemodynamic instability, the patient died of refractory cardiogenic shock.

The remaining patients were alive and asymptomatic, with decreases in their postsurgery gradients, including the patient with diffuse stenosis.

DISCUSSION

Supravalvular aortic stenosis is an unusual form of obstruction of the left ventricle outflow tract which occurs in 3% to 6% of cases of various types of aortic obstruction; the occurrence of supravalvular aortic stenosis is surpassed by subaortic and valvular stenosis in studies that analyzed 4 to 20 years of cases; the anatomical variants of supravalvular aortic stenosis affect postsurgery results and survival rates. In our study, we found a frequency of 8.4% of the total cases had changes limiting blood flow in the left ventricle outflow tract and, as opposed to other studies, diaphragm subvalvular stenosis predominated in the surgical group studied, probably due to selection bias since many patients with stenosis of the aortic valve are treated with interventions that leave only cases of sub- or supravalvular obstruction remaining for surgery.

As reported in the international literature, the hourglass type of obstruction is the most common. It is important to remember that the diffuse type of stenosis has been described as the type that more commonly results in death. We also found this to be the case, and noted that it was associated with the persistence of symptoms and supravalvular pressure gradient in the patient who survived, who required re-intervention 2 years later, with a very favorable postoperative course after the second intervention.

The presence of hypercalcemia, which occurs during the first months of life in approximately 20% of cases, did not occur in our series of patients; although these patients came to us at a more advanced age, the number of cases studied is small. It is sometimes difficult to perform a complete study in patients, as reported by Soares et al in Brazil, who were only able to complete evaluation of 2 out of 3 cases and did not report the results of the cardiopathic correction if surgical intervention had been performed.

On the other hand, the repair technique used by our group is that described by McGoon et al. as the anatomical conditions of our patients allowed its use. Nevertheless, when the stenosis is very near the valves and the coronary ostia, the use of the Doty et al technique may be preferred, which involves an inverted Y patch. At present, there are several variations in the surgical technique for correcting this type of defect, although Hazekamp et al did not find any significant differences in change in valve function, and found the efficacy of reducing the pressure gradient was similar and acceptable with various techniques. Their findings are in contrast with those of Stamm et al in Boston, who analyzed cases occurring between 1957 and 1998 that included 75 patients, of whom 7 died perioperatively and the remainder of whom had a survival rate of 100% at 5 years and 77% at 20 years; at the end of the study the authors showed that diffuse stenosis had an influence on the outcome of this type of patient and that plasty of the 3 valves reduced the gradient more efficaciously than simple plasty of the noncoronary chest.

In the cases of recurrent serious stenosis, an alterna-
tive has even been to use the anastomosed valve graft of the of the free wall from the left ventricle to the descending aorta. Similarly, another option that must be mentioned not only for cases of recurring stenosis but also for complex cases of diffuse stenosis, is repair with an autologous arterial graft from the pulmonary artery, as described by Al-Halees et al in a 6-year-old patient.

In a series of 9 cases who underwent surgery between 1991 and 1998 stenosis of the left coronary trunk was found and the patients were treated immediately with widening of the aortic diameter; when the stenosis was localized in the ostium, resection of the redundant tissues was performed; on the other hand, if the coronary involvement was diffuse, an aortocoronary graft was used. On followup at 57 months, the patients had a favorable course, which points out the necessity of a thorough clinical evaluation that should include an angiographic study of the patient’s coronary anatomy in order to choose the most appropriate and effective surgical intervention. In contrast, McElhinney et al between 1992 and 1998 performed the Ross procedure in 4 of their 36 cases in which obstruction of the left ventricular outlet tract was considered complex. One death resulted and there were no reoperations. Their results included resection of the subaortic diaphragm in 11 patients.

In conclusion, supravalvular aortic stenosis is a complex and uncommon disease that requires careful evaluation and efficacious surgical treatment that provides anatomical improvement and improves quality of life. The followup period of our study is a bit too short to determine the influence of surgical treatment on the long-term survival of patients; they remain under the joint surveillance of the cardiology service that referred them for treatment and the Department of Congenital Cardiopathy in our hospital.

REFERENCES