

Congenital Aortic Supravalvular Stenosis. Frequency and Results of Surgical Treatment in a Third Level Hospital

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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 ≥ 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 the 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-

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Estenosis supravalvular aórtica congénita. Frecuencia y resultados del tratamiento quirúrgico en un hospital de tercer nivel

En un estudio prospectivo realizado entre el 1 de septiembre de 1996 y el 31 de diciembre de 2001, se identificó a los pacientes pediátricos con diagnóstico confirmado de estenosis supravalvular aórtica. Se consideró estenosis supravalvular cuando se detectó la disminución del calibre aórtico y un gradiente ≥ 50 mmHg.

De 83 pacientes con estenosis aórtica, sólo en 7 (8,4%) fue de tipo supravalvular y fue tratada con resección del tejido fibroso y ampliación de la luz aórtica con injerto de dacrón precoagulado, sin accidentes ni complicaciones. Hubo disminución significativa del gradiente preoperatorio ($p < 0,05$). Un paciente falleció y otro presentó choque cardiogénico y paro cardiorrespiratorio reversible con secuelas neurológicas en remisión, y en la actualidad los pacientes se encuentran asintomáticos.

Concluimos que la estenosis supravalvular aórtica es poco frecuente en México y los resultados del tratamiento quirúrgico son satisfactorios en función del tipo de lesión y de acuerdo con ello.

Palabras clave: *Síndrome de Williams. Estenosis supravalvular. Cirugía aórtica.*

fects and psychomotor retardation was established in 1961.¹ In the same year, McGoon et al,² from the Mayo Clinic, proposed widening the aorta diameter with a synthetic graft as a treatment option.

Although clinical presentation of supravalvular aortic stenosis is similar to that in valvular and subvalvular stenosis, supravalvular aortic stenosis has been found in association with hypercalcemia, elfin facies, and mental retardation; there is also a familial form that is not associated with elfin facies.³⁻⁶

Aortic supravalvular stenosis is the least common form of aortic obstruction and affects both sexes equally.

Aortic supravalvular stenosis may be due to the presence of: a discrete fibrous membrane, an hour-glass narrowing, or a diffuse narrowing.⁶⁻⁸

In the international literature, including that of Latin America, reveals a low occurrence of supravalvular aortic stenosis^{1,6,7,9,10} and in an extensive review of 653 cases of congenital cardiopathy by Vizcaíno and Ortega¹¹ 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 forehead; depressed temporal bones; epicanthus; depressed nasal bridge; anteverted nasal openings with prominent superior lip (elfin facies); and other characteristics.⁹ 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 $P < .05$.

RESULTS

During the evaluation period, 1390 pediatric patients of both sexes, with an average age of 6.9 years

TABLE 1. Frequency of cases of supravalvular stenosis

Year	Total number of cases	Aortic stenosis	Supravalvular stenosis
1996	81	7	-
1997	288	9	1
1998	216	13	1
1999	281	21	1
2000	275	21	2
2001	228	12	2
Total	1390	83	7

(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 rubeola.

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 \pm 27 minutes and CPS time was 84.2 minutes \pm 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 ($P < .05$) in the preoperative gradient (from 86.2 mm Hg \pm 19.6 mm Hg to 12 mm Hg \pm 8.7 mm Hg) was achieved at 3 and 6 months post-surgery.

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

TABLE 2. Patient characteristics

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

Δ pre indicates preoperative gradient; Δ post, 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 valvular⁶ stenosis in studies that analyzed 4 to 20 years of cases; the anatomical variants of supravalvular aortic stenosis affect postsurgery results and survival rates.⁷ 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.^{1,6,7} 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 life¹¹ in approximately 20% of cases,⁶ 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 al⁹ 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,² 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 al¹² 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 al¹³ 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 al¹⁴ 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.⁸ 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 al¹⁵ 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.¹⁶ 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 al¹⁷ 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.

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