Surgical Treatment of Ascending Aortic Complications in Marfan Syndrome: Early and Long-Term Outcomes

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Findings in 54 patients (mean age 39 years, range 18-66 years, 25% female) were analyzed. Of these patients, 21 had dissection of the ascending aorta (15 acute and 6 chronic) and 33 had aneurysm of the ascending aorta. Surgery was classified as emergency surgery in three cases, as urgent in 15, and as scheduled surgery in 36. The Bentall-De Bono procedure was performed in 39 patients, aortic valve reimplantation was carried out in nine, Cabrol's operation was performed in three, and a homograft was used in 3. The mean diameter of the ascending aorta was 66.6 mm. Overall, in-hospital mortality was 3.7% (33.3% for emergency surgery vs 2.8% for scheduled surgery; *P*<.001). During the mean follow-up period of 4 years (range, 2 months-14 years), 7 patients died, including four who died due to type-B aortic dissection. The actuarial survival rate at 2, 5, and 10 years was 94%, 83%, and 75%, respectively, with 88%, 67%, and 43% of patients, respectively, not requiring reoperation. Elective aortic root replacement was associated with a low risk and a good survival rate.

Key words: *Marfan Syndrome. Aortic Surgery. Results.*

Tratamiento quirúrgico de las complicaciones de la aorta ascendente en el síndrome de Marfan. Resultados inmediatos y a largo plazo

Se analizaron los resultados en 54 pacientes con una media de edad de 39 (18-66) años; el 25% eran mujeres; 21 pacientes presentaban disección (aguda en 15 y crónica en 6 pacientes) y 33, aneurisma. La operación fue de máxima urgencia en 3 pacientes, urgente en 15 y programada en 36. Utilizamos tubo valvulado en 39 pacientes, reimplante valvular en 9, técnica de Cabrol en 3 y homoinjerto en 3. El diámetro promedio aórtico fue de 66,6 mm. La mortalidad hospitalaria total fue del 3,7% (máxima urgencia, 33,3%; programada, 2,8%; p < 0,001). Durante el seguimiento (media, 4 años [2 meses-14 años]) fallecieron 7 pacientes, 4 por disección aórtica tipo B. A los 2, a los 5 y a los 10 años, la supervivencia actuarial fue del 94, el 83 y el 75% y la libertad de reoperación, del 88, el 67 y el 43%, respectivamente. El reemplazo electivo de la aorta ascendente tiene bajo riesgo y buena supervivencia.

Palabras clave: *Síndrome de Marfan. Cirugía aórtica. Resultados.*

INTRODUCTION

Aortic root dilatation in Marfan syndrome (MS) can cause acute dissection, aortic rupture, and aortic regurgitation[1] and is a common cause of premature death.[1,2] Prophylactic replacement has achieved a substantial improvement in life expectancy.[3,4] Most published studies are from Europe and the United States; no series in Latin America have been reported. In the present study, we describe the short-term results (in-hospital follow-up) and long-term outcome (complications) of surgery at a tertiary referral center in Argentina.

METHODS

Between July 1992 and December 2006, 54 patients with MS underwent surgery based on criteria subsequently confirmed at Ghent, Belgium.[5] The surgery was performed in accordance with routine procedures.[6] Surgery performed in a decompensated patient within 24 hours of diagnosis was considered as emergency, surgery performed during hospitalization but after 24 hours as urgent, and surgery undertaken while the patient was stable and asymptomatic as scheduled. The demographic

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TABLE 1. **Demographic, Clinical, and Echocardiographic Data of 54 Patients With Marfan Syndrome Who Underwent Ascending Aorta Replacement Surgery**[a]

	Acute Dissection (n=15)	Chronic Dissection (n=6)	Aneurysm Without Dissection (n=33)
Age, mean (range), y	37.26 (22-57)	42.8 (22-59)	37.12 (18-66)
Women, %	26.6	20	28
Heart rate, mean, bpm	62	64	62
Blood pressure, mean, mm Hg	124/75	128/65	125/95
Severe aortic regurgitation, n (%)	6 (40)	1 (17)	2 (6)
Aortic diameter, mean (range), mm	55.72 (43-63.6)	78 (67-81)	66.19 (40-104)
LVEF <40%, n (%)	1 (7)	0	2 (6)

[a]LVEF indicates left ventricular ejection fraction.

characteristics, in-hospital mortality, late mortality, and need for repeat surgery or interventional treatment on follow-up were analyzed retrospectively. The following events were considered: death, repeat surgery, infective endocarditis, thromboembolism, and major bleeding. Doppler transesophageal echocardiography (TEE), computed tomography (CT), or magnetic resonance imaging was used for the diagnosis. Doppler TEE was performed during surgery, and follow-up was based on TEE and CT. All patients received beta-blockers.

Statistical Analysis

Continuous variables are reported as the mean (SD) or range, and categorical variables, as percentages. The univariate analysis was performed using ANOVA for continuous variables with a normal distribution and the χ^2 test for qualitative variables. The Kaplan-Meier method was used to calculate patient survival. The survival rates and 95% confidence intervals (CI) for the different periods are shown.

RESULTS

Mean patient age was 39 years (range, 18-66); 25% were women. Fifteen patients had acute aortic dissection; 6, chronic aortic dissection; and 33, ascending aorta aneurysm. The average (SD) aortic diameter was 66.6 (3) mm. A comparison of the demographic, clinical, and echocardiographic data is presented in Table 1.

The surgery was emergency in 3 (5.6%) patients, urgent in 15 (27.8%), and scheduled in 36 (66.7%); 4 patients had previous aortic surgery. Aortic root replacement used a valved conduit (Bentall de Bono procedure) in 39 patients with a Cabrol modification in an additional 3, valve reimplantation (T. David) in 9, and a homograft in 3. Mitral valve replacement or plasty was performed in 6 patients.

Surgical mortality was 3.7% (2 patients) and was significantly higher in the emergency surgeries compared with elective surgery (33.3% vs 2.8%; *P*<.001). On follow-up (mean, 4 years [range, 2 months-14 years]), 3 (5.5%) patients were lost, and another 7 died; among them, 4 had type B redissection that had been reoperated and culminated in sepsis (3 from surgery and 1 from catheterization), 2 died of heart failure, and 1 of an unknown cause. Thirteen patients required a second operation, including 9 procedures to treat a distal redissection, 1 heart transplantation, and 3 homograft placement for infective endocarditis. Two patients had a third operation. Survival at 2, 5, and 10 years was 94% (95% CI, 88-100), 83% (95% CI, 70-97), and 75% (95% CI, 55-95), respectively (Figure 1). Reoperation at 2, 5, and 10 years was not necessary in 88% (95% CI, 78-98), 67% (95% CI, 50-85), and 43% (95% CI, 17-70), respectively (Figure 2).

DISCUSSION

Before the advent of prophylactic aortic root surgery, most patients died prematurely; life expectancy with the current treatment is 70 years.[3,4] There is a clear association between increased diameter and the risk of dissection and rupture in this condition: the risk of rupture of a 6-cm aneurysm is 4-fold.[7] The recommended aortic diameter for prophylactic surgery is 5 cm.[7,8] An accelerated increase (>1 cm/y), family history of premature death, or moderate-to-severe aortic regurgitation will require earlier surgery.[1] Some authors propose that surgery be done at diameters >3 cm because of the unpredictability of aortic rupture.[8] The efficacy, safety, and reproducibility of ascending aorta replacement with valved conduits were initially described by authors from a single institution.[9] Later, a report emerged with a series of 675 MS patients operated at 10 surgical centers.[10] In that study, 30% presented aortic dissection and 30-day mortality was 1.5% in elective surgery, 2.6% in urgent surgery, and 11.7% in emergency surgery. In 6.7 years of follow-up, there were 114 late deaths, most due to residual aortic dissection or rupture, and arrhythmias, mainly within the first 60 days after surgery. Later, mortality was low and constant: 93.5% of patients were alive at 5 years, 91% at 10 years, and 59% at 20 years. Subsequent experience confirmed these results.[11] Similar to the multicenter patient population, 38% of our patients presented aortic dissection. Overall mortality was somewhat higher at 3.7% (elective, 2.8%;

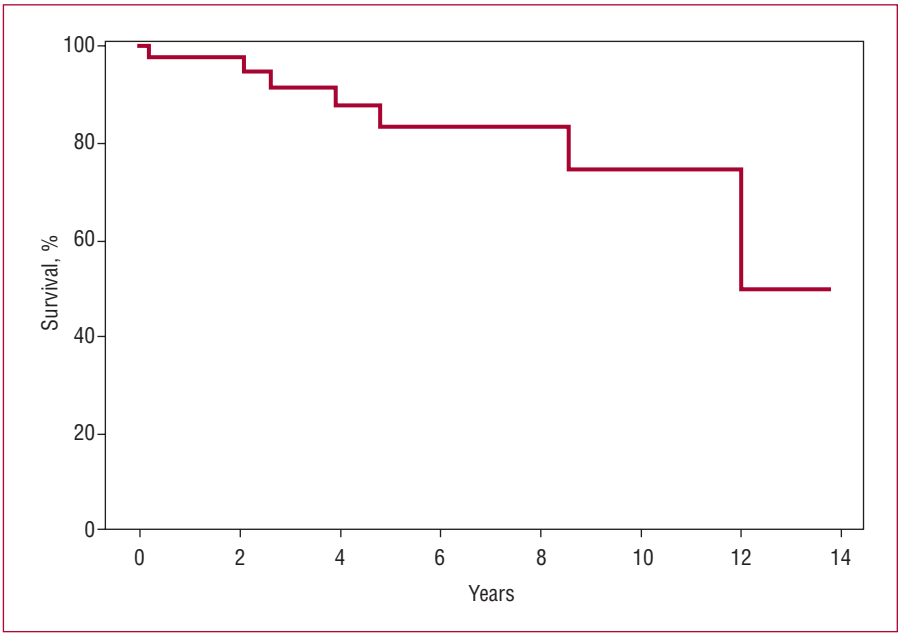

Figure 1. Survival curve of 54 patients with Marfan syndrome operated for complications of the ascending aorta.

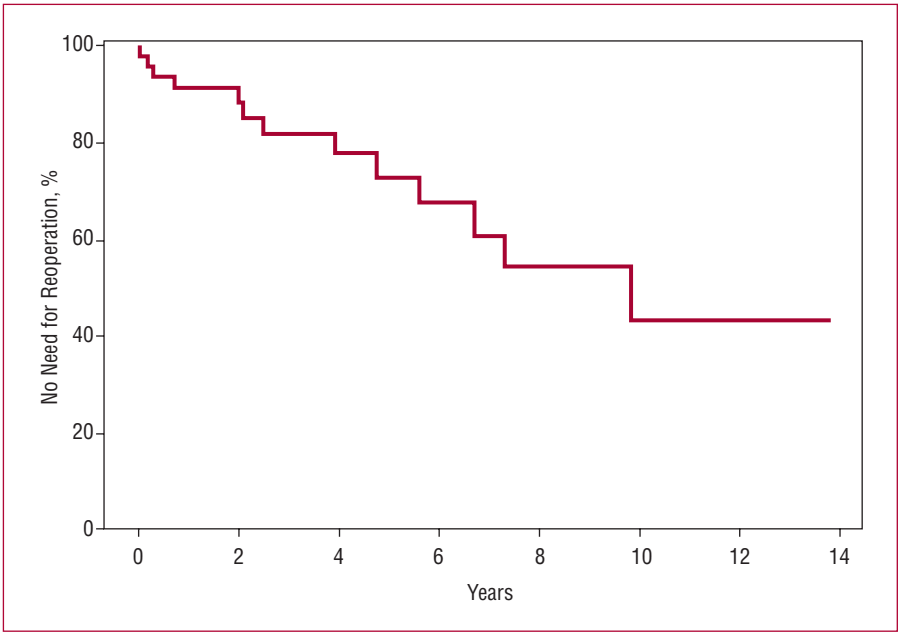

Figure 2. No need for reoperation in 54 patients with Marfan syndrome operated for complications of the ascending aorta.

emergency, 33%), survival rates were 94% at 2 years, 83% at 5 years, and 75% at 10 years, and there was no need for reoperation in 88%, 67%, and 43%.

Yacoub et al[12] described a valve-sparing "remodeling" technique for the aortic valve and David et al,[13] "reimplantation" in a tubular Dacron graft. The use of these techniques was restricted to patients with structurally normal aortic valves. David et al[14] retrospectively analyzed 220 valve-sparing surgeries (167 with reimplantation and 53 with remodeling), 40% in MS patients, with a follow-up of 5.3 years. There were 3 operative deaths and 13 late deaths. Overall survival at 10 years was 88% and freedom from significant aortic regurgitation was 85% (94% with reimplantation and 75% with remodeling). Reimplantation was used in 9 of our patients, with no surgical mortality or significant mid-term aortic regurgitation. However, there are no reports of long-term follow-up with this technique and the need for reoperation has not been established.[15,16]

A study of 78 MS patients who underwent surgery was recently conducted. During follow-up (mean, 5.4 years) 4 patients died and 46 "aortic events" (elective surgery

or dissection) occurred in 45 patients, with distal aortic involvement in 14 patients (31%).[17] Consistent with that series and with others,[9-12] 25% of our patients required repeat surgeries in the distal aorta. Hence, we believe that all MS patients who have undergone surgery should receive indefinite regular follow-up of the aorta.

We conclude that elective replacement of the ascending aorta with valved conduit is a low-risk, effective, and long-lasting procedure in patients with MS.

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