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Outcomes of Aortic Valve-Sparing Operations in Marfan Syndrome

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ABSTRACT

BACKGROUND In many cardiac units, aortic valve-sparing operations have become the preferred surgical procedure to treat aortic root aneurysm in patients with Marfan syndrome, based on relatively short-term outcomes.

OBJECTIVES This study examined the long-term outcomes of aortic valve-sparing operations in patients with Marfan syndrome.

METHODS All patients with Marfan syndrome operated on for aortic root aneurysm from 1988 through 2012 were followed prospectively for a median of 10 years. Follow-up was 100% complete. Time-to-event analyses were calculated using the Kaplan-Meier method with log-rank test for comparisons.

RESULTS A total of 146 patients with Marfan syndrome had aortic valve-sparing operations. Reimplantation of the aortic valve was performed in 121 and remodeling of the aortic root was performed in 25 patients. Mean age was 35.7 ± 11.4 years and two-thirds were men. Nine patients had acute, 2 had chronic type A, and 3 had chronic type B aortic dissections before surgery. There were 1 operative and 6 late deaths, 5 caused by complications of dissections. Mortality rate at 15 years was 6.8 ± 2.9%, higher than the general population matched for age and sex. Five patients required reoperation on the aortic valve: 2 for endocarditis and 3 for aortic insufficiency. Three patients developed severe, 4 moderate, and 3 mild-to-moderate aortic insufficiency. Rate of aortic insufficiency at 15 years was 7.9 ± 3.3%, lower after reimplantation than remodeling. Nine patients developed new distal aortic dissections during follow-up. Rate of dissection at 15 years was 16.5 ± 3.4%.

CONCLUSIONS Aortic valve-sparing operations in patients with Marfan syndrome were associated with low rates of valve-related complications in long-term follow-up. Residual and new aortic dissections were the leading cause of death. (J Am Coll Cardiol 2015;66:1445–53) © 2015 by the American College of Cardiology Foundation.

Aortic valve-sparing operations were developed to preserve the native aortic valve in patients with aortic root aneurysm (1,2). Their application in patients with Marfan syndrome was initially questioned because of the presence of abnormal fibrillin in the aortic valve (3). Early on in our experience, we treated only patients with aortic root aneurysms with normal aortic cusps and closely monitored post-operative valve function with periodic echocardiography. As our confidence in these operations increased, we expanded their use to patients with mild aortic cusp abnormalities, such as prolapse caused by elongation of free margins and stress fenestrations in the commissural areas caused by dilation of the aortic root. Elongation of the free margin of an aortic cusp was corrected by shortening its central portion along the nodule of Arantius, and large stress fenestrations by weaving a double layer of a fine expanded polytetrafluoroethylene suture along the free margin from commissure to commissure. These techniques of cusp repair proved to be durable; thus, the indications for aortic valve-sparing operations expanded accordingly (4,5).

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ABBREVIATIONS AND ACRONYMS

AI = aortic insufficiency

MV = mitral valve

Aortic valve-sparing operations have now become the preferred surgical procedure in our center to treat young patients with aortic root aneurysm (5) and approximately one-third of them had the diagnosis of Marfan syndrome according to Ghent criteria (6,7). This study examines the long-term outcomes of aortic valve-sparing operations in patients with Marfan syndrome.

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METHODS

STUDY POPULATION. From September 1988 through December 2012, a total of 149 patients with the diagnosis of Marfan syndrome by Ghent criteria (6,7) were operated on for aortic root aneurysm or Stanford type A aortic dissection at Toronto General Hospital by 1 surgeon (T.E.D.). The original Ghent criteria (6) were used retrospectively to identify patients operated on before 1997, then prospectively from 1997 through 2010; the revised criteria have been used since then (7). Three patients had aortic root replacement with a conduit containing a mechanical valve because of grossly abnormal aortic cusps and were excluded. The remaining 146 patients had aortic valve-sparing operations. This study was approved by the University Health Network Research Ethics Board and patients' consent was waived.

SURGICAL TECHNIQUES. Two types of aortic valve-sparing operations were performed in these patients (**Figure 1**): reimplantation of the aortic valve (the David operation [1]) and remodeling of the aortic root (the Yacoub operation [2]). Reimplantation of the aortic valve was performed by detaching the coronary arteries from the aneurysmal aortic sinuses, leaving a small rim of tissue around their orifices; a narrow rim of aortic sinus tissue was also left attached to the aortic annulus. Next, a tubular Dacron graft of appropriate diameter was sutured on the outside of the left ventricular outflow tract immediately below the level of the nadir of the aortic annulus. The remnants of aortic sinuses were sutured inside the graft, recreating the crescent shape of the aortic annulus. The cusps were examined to ensure that their coaptation level was well above the level of the nadir of the aortic annulus. If 1 or more aortic cusps were prolapsing, their free margin was shortened by plication along the nodule of Arantius. Additionally, if large fenestrations were present on the free margin along the commissural areas, a double layer of a fine expanded polytetrafluoroethylene suture was weaved along the free margin of the cusp from commissure to commissure. Finally, the coronary arteries were reimplanted on the Dacron graft. During the second decade of experience with this operation, neoaortic sinuses were created by using a slightly larger graft and reducing its diameter at the subannular and sinotubular junction levels (**Figure 1**).

Remodeling of the aortic root was performed by preparing the aortic root as described previously and tailoring a tubular Dacron in such a way as to recreate the aortic sinuses and suturing it to the remnants of aortic sinuses and aortic annulus. If the aortic annulus was deemed dilated at the time of surgery, an external annuloplasty with a Dacron band along the fibrous components of the left ventricular outflow tract was performed. The aortic cusps and coronary arteries were handled similarly as with the reimplantation procedure.

FOLLOW-UP. All patients have been followed prospectively with echocardiographic studies and periodic computed tomography or cardiac magnetic resonance imaging of the entire aorta every 3 to 5 years, unless the patient had a history of aortic dissection or another pre-existing aneurysm, in which case imaging of the aorta was performed more often. Given that aortic insufficiency (AI) was assessed by echocardiography, we used the date of the first post-operative study to reveal AI greater than mild as the time of event. Although the apparition of AI might predate the echocardiogram, using the first report of the abnormality is an adequate and only available surrogate.

STATISTICAL ANALYSIS. Data are described as mean ± SD (continuous variables) or frequencies (categorical variables) as appropriate. Rates of mortality, reoperations, AI, and aortic dissections were calculated using the Kaplan-Meier method with log-rank test for comparisons between groups. To account for the concomitant probability of death or other adverse event, competing risk analysis was used. In competing risk analysis, at time zero, all patients were free from adverse events; patients transiting to either death or other adverse event, whichever occurred first and at the same time, were removed from the proportion of patients who were alive and free from adverse events. Sex-specific life tables from the Province of Ontario from the 2000 to 2002 period (available from Statistics Canada) were used to estimate predicted survival of the patient cohort with yearly estimates fitted with an exponential regression model. All statistical analyses were performed using SAS statistical software version 9.3 (SAS Institute, Inc., Cary, North Carolina).

RESULTS

A total of 146 patients were included in this study. Clinical follow-up was 100% complete to a median of