Study Backs Prophylactic Aortic Root Repair

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NEW YORK — Rupture and tear from an aortic root aneurysm is a major cause of death in Marfan syndrome and Loeys-Dietz syndrome, but preventative aortic root repair in these diseases has advanced over the decades to prolong lives and avoid the need for future surgery.

"A prophylactic aortic root replacement in both Marfan syndrome and Loeys-Dietz syndrome has very low operative risks and excellent long -term results," Dr. Duke Cameron of Johns Hopkins in Baltimore said at the symposium, which was sponsored by the American Association for Thoracic Surgery.

Dr. Cameron reported on results of 417 patients with Marfan syndrome who were treated at Johns Hopkins since 1976 and 31 patients with Loeys-Dietz syndrome treated over an 8-year period.

In Marfan syndrome, the threshold aortic root diameter indicated for surgery declined over the years to 5 cm, Dr. Cameron said.

"If there's been a family history of aneurysm or dissection, we've lowered the threshold by about 0.5 cm," he said, adding that in younger patients, enlargement of the aortic root diameter of more than 1 cm a year is also a reason to operate.

The Hopkins approach for aortic root repair in Marfan syndrome is straightforward, he said: full root replacement with full-thickness, end-to-end anastomosis, mobilization of the three coronary arteries, and root reimplantation. The Hopkins group used a remodeling technique exclusively until 2002, "but became disappointed with annual dilatation and aortic regurgitation seen in some of these patients," Dr. Cameron said. Now, they perform reimplantation with a Valsalva graft.

The average age of Marfan syndrome patients in the Hopkins' treatment group was 32; ages ranged from 1.5 to 73 years. Three patients died, including 2 of the 45 patients who had urgent or emergency surgery. None of the patients who had elective operations died within 30 days of the operation. At 20 years after surgery, 75% of patients survived.

Aortic root dissection present at the time of the operation reduced a patient's late-term survival time by 50%, Dr. Cameron said, "underscoring again the importance of root replacement before dissection occurs."

A history of mitral valve surgery, seen mostly in younger patients, was also a predictor of late death. The two most prevalent late-term causes of death were dissection or rupture in the distal aortic or ileac arteries and arrhythmia, he said.

However, freedom from thromboembolism in Bentall procedures with mechanical valves surpassed 90% at 25 years, as did freedom from endocarditis, Dr. Cameron said. "This is better than what one would expect to see with an isolated mechanical aortic valve replacement," he said. "It makes the point that the Bentall procedure is still one of the very best operations we have in cardiac surgery."

A previous Hopkins' study showed the Bentall procedure had higher rates of thromboembolism than did valve-sparing aortic root replacement in Marfan syndrome, but valve-sparing surgery had higher rates of reoperation (Ann. Thorac. Surg. 2008;85:2003-10).

Preventative operations in Loeys-Dietz syndrome are similar to those for Marfan syndrome, but the threshold aortic root diameter for intervention is lower: 3 cm in children and 4 cm in adults, Dr. Cameron said.

Overall, the 31 patients with Loeys-Dietz syndrome who had aortic root repair were half the age of Marfan syndrome patients (15 years on average) with an average sinus diameter of 3.9 cm. All had a valve-sparing procedure, and none died during the operation or at follow-up at an average of 3.6 years, Dr. Cameron said.

"When comparing Loeys-Dietz to Marfan syndrome at early to mid-term follow-up, they have very similar operative outcomes," Dr. Cameron said. ■

Disclosures: *Dr. Cameron had no disclosures relevant to his presentation.*