Cardiovascular Operations in Children With Marfan Syndrome

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Cardiovascular operations in children with Marfan syndrome are uncommonly performed and consequently, no physician, surgeon, or even institution has gained extensive experience. Age and size of the child are probably the most important factors in determining the outcomes of these operations. By the time children with Marfan syndrome reach the age of 10 or 12 years, the operations in the aortic root and mitral valve can be performed as safely as they are in young adults. However, the prognosis is usually poor if surgery is needed during infancy. The outcome of aortic surgery in neonates is likely worse than mitral valve surgery. There are case reports of successful mitral valve repair in neonates with survival up to a decade.[1,2] There is a very limited amount of information on surgery during childhood. One of the earliest reports on these operations was from The Johns Hopkins Hospital in 1997.[3] They described the results in 26 children with a mean age of 10 years (range 8 months to 17 years) who had aortic root replacement, mitral valve repair or replacement, and combined aortic root and mitral valve operations.[3] There was no operative death in this small series, and overall survival at 10 years was 79% but the freedom from reoperation was 41%, suggesting that most children need a reoperation within a decade of the initial procedure. There have been a few reports on the outcomes of aortic valve sparing operations in children with fairly good results.[4,5] Reimplantation of the aortic valve has provided better outcomes than remodeling of the aortic root or composite replacement of the aortic valve and ascending aorta, particularly in children older than 10 years of age.

This issue of *The Seminars* has a study by Knadler et al[6] on in-hospital outcomes of cardiovascular operations in 294 children with Marfan syndrome. The study was based entirely on the administrative database from the Pediatric Health Information System on children and young adults <25 years of age with the diagnosis of Marfan syndrome from 40 pediatric hospitals but contains only outcomes during the hospital stay as coded for administrative purposes. This is an important limitation of this study. The diagnosis of Marfan syndrome could not be verified because there were no records for review, and this diagnosis is more difficult to establish in children than in adults. There were 54 instances of mitral valve operations, 224 of aortic valve and aortic operations (not a single aortic valve sparing operation was reported in spite the fact that the mean age was 16 years) and 43 of combined mitral and aortic valves. The operative mortality rate was 2.2% (higher for mitral than for aortic operations) but complication rates were high. However, complications were listed as they were coded for administrative purpose. Although this is the largest series of children with the diagnosis of Marfan syndrome who had cardiovascular operations, I believe the study has more limitations than virtues. Hopefully, it will stimulate the participants of the Pediatric Heart Network Investigators to examine this issue in their database since it contains more detail on diagnosis, management, and outcomes.[7] In addition, a registry on children with the proven diagnosis of Marfan syndrome would improve our knowledge and skills to better serve this patient population.

Patients with Marfan syndrome and other genetic syndromes associated with cardiovascular diseases should be managed in

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Central Message

Cardiovascular operations are uncommon in children with Marfan syndrome. There is a need for a registry of these operations to increase our knowledge in this area.

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specialized centers able to care for all aspects of their cardiovascular and associated disorders. Hospitals where only 1 or 2 children with these inherited disorders are seen each year should refer their patients to centers with larger experience for the good of the patient.

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