Genes, aortic diseases, and cardiovascular surgery

Tirone E. David, MD

From the Division of Cardiovascular Surgery of Toronto General Hospital and the University of Toronto, Toronto, Ontario, Canada.

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Address for reprints: Tirone E. David, MD, 200 Elizabeth St, 4N453 Toronto, Ontario M5G 2C4 Canada (E-mail: tirone.david@uhn.ca).

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Tirone E. David, MD

Central Message

It is becoming increasingly evident that aortic pathologies associated with genetic syndromes should be managed in aortic centers with dedicated professionals qualified to deliver comprehensive care.

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It is becoming increasingly more evident that most aortic pathologies are mediated by genes, and we are just beginning to understand the various phenotypes. For this reason alone, aortic surgeons can no longer practice in isolation and patients cannot expect the best care next door to their homes. An array of dedicated professionals is needed to deliver comprehensive care to patients with aortic pathologies, particularly the young ones. Cleveland Clinic leaders recognized the importance of developing an aortic program with the necessary infrastructure to care for these patients and have provided us with additional information on patients with Loeys-Dietz syndrome (LDS) in this issue of the *Journal*.¹ The article contains important information for aortic surgeons because there have been few reports on aortic surgery in patients with LDS.^{2,3} They identified 53 patients who fulfilled the criteria of LDS, and 33 of them required aortic surgery (11 for aortic dissections). During a median follow-up of 5.2 years, these 33 patients had 58 aortic and 81 cardiovascular operations. These numbers of operations in such a relatively small series underscore the importance of close surveillance of these patients.

Reimplantation of the aortic valve is the preferred operation to treat aortic root aneurysm associated with genetic syndromes in young patients.⁴ Reimplantation of the aortic valve was the procedure performed in 19 of the 33 patients in the Cleveland Clinic's series, and there was no aortic valve-related reoperation during the follow-up.¹ In the Johns Hopkins series of 79 surgically treated patients with LDS, 52 had reimplantation of the aortic valve using a modified technique whereby the Dacron graft was secured beneath the aorto-ventricular junction using only 3 sutures in the 3 nadirs of the aorto-ventricular junction.² Johns Hopkins investigators recently described an unusual complication of reimplantation of the aortic valve in 4 children who developed false aneurysms beneath the Dacron graft along the intervalvular fibrous body.⁵ This may be a complication of the technique of securing the graft in the left ventricular outflow tract with only 3 sutures

instead of a complete, circumferential subaortic annuloplasty with multiple sutures as originally described.⁶ Hopkins investigators also described false aneurysm in the coronary artery button after replacement of the aortic root.⁷ Their patients were much younger than the patients in the Cleveland Clinic's series, and it is possible that children with LDS have a more severe form of connective tissue degeneration that may predispose to false aneurysms.

These 2 recently published series^{1,2} and related publications^{5,7} of surgically treated patients with LDS demonstrated the importance of having these patients treated in centers of excellence in aortic disorders.

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