AORTIC DECISION MAKING IN LOEYS-DIEZT SYNDROME

To the Editor:

We read with great interest the report by Augoustides and colleagues¹ on aortic decision making in the Loeys-Dietz syndrome (LDS). They presented the surgical management of an adolescent with LDS. He was initially referred for aortic root aneurysm and underwent elective reconstruction of the aortic root and aortic arch during deep hypothermic arrest. The authors concluded that in patients with LDS, management should take into account not only the current aortic pathologic condition but also the likely future aortic syndromes based on the natural history of this vasculopathy.

This case provides yet another good example of the successful result of surgical intervention in patients with LDS. This characteristic distinguishes them from those with vascular Ehlers-Danlos syndrome.^{2,3} Patients with LDS generally undergo multiple cardiovascular operations, and they recover without sequela.²

We have treated a 32-year-old woman who arrived at the emergency room with new onset of chest discomfort. Echocardiography disclosed a huge aortic root aneurysm with severe aortic regurgitation and an ejection fraction of 45%. Multislice computed tomography showed an 8-cm aneurysm of the aortic root (Figure 1, A) with a dilated aortic annulus. She displayed distinctive physical stigmata of LDS, such as hypertelorism and

FIGURE 1. Preoperative and postoperative computed tomographic angiograms: A, At admission, an 8-cm aortic root aneurysm with a normal ascending aorta was seen. B, The ascending aorta was repaired with a Dacron graft without further dilation of the remaining aorta.

joint hyperlaxity. We replaced the aortic root with a graft conduit containing a mechanical valve. The aortic arch had normal diameter and was left intact. The patient was advised to avoid vigorous physical activities and to follow a regimen of beta-blockers. Genetic testing identified a transforming growth factor β -R1 mutation, which was also found in her daughter, confirming the diagnosis.

Two years after the Bentall procedure, she was free of symptoms and computed tomography showed normal dimension of the remaining thoracic aorta (Figure 1, B).

The strategy used by Augoustides and associates is not always feasible. The diagnosis of LDS is based on clinical and genetic testing,² which is not always available. Operating on a young patient with ascending aortic dissection or with a huge aortic aneurysm is standardized. In these particular cases, we know that we are facing a connective tissue disorder, but the natural history and the morbidity and mortality of the procedure in LDS differ from those of other inherited vasculopathy disorders. Importantly,

genetic confirmation is obtainable later after the surgical procedure. Ethically, we are amenable to replacing the aortic root and leaving the remaining apparently normal aorta.

We have a few questions for Augoustides and associates regarding their management. First, what is your opinion about leaving an elephant trunk during the arch replacement? What is your threshold of the aortic root size to operate on an adult with LDS? How often do you conduct follow-up in the patient with LDS after surgery? We have been concerned about the possibility of on aggressive aortopathy in our patient, but it seems that this disease is controlled by medical therapy. Additionally, if the patient with LDS undergoes serial computed tomographic analysis, then we will discover any significant increase in growth rate and we will act accordingly.

We greatly appreciate this report regarding the authors' management of this recently described syndrome and their extremely useful attitude. We congratulate them on the survival of their patient and the dissemination of important information.

The Editor welcomes submissions for possible publication in the Letters to the Editor section that consist of commentary on an article published in the Journal or other relevant issues. Authors should: • Include no more than 500 words of text, three authors, and five references. • Type with double-spacing. • See <http://jics.ctsnetjournals.org/misc/fora.shtml> for detailed submission instructions. • Submit the letter electronically via jtcvs.editorialmanager.com. Letters commenting on an article published in the JTCVS will be considered if they are received within 6 weeks of the time the article was published. Authors of the article being commented on will be given an opportunity of offer a timely response (2 weeks) to the letter. Authors of letters will be notified that the letter has been received. Unpublished letters cannot be returned.

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References

1. Augoustides J, Plappert T, Bavaria J. Aortic decision-making in the Loeys-Dietz syndrome: aortic root aneurysm and a normal-caliber ascending aorta and aortic arch. *J Thorac Cardiovasc Surg.* 2009; 138:502-3.
2. Loeys BL, Schwarze U, Holm T, Callewaert BL, Thomas GH, Pannu H, et al. Aneurysm syndromes caused by mutations in the TGF-beta receptor. *N Engl J Med.* 2006;355:788-98.
3. Melenovsky V, Adamira M, Kautznerova D, Voska L, Weichet J, Loeys B, et al. Aortic dissection in a young man with Loeys-Dietz syndrome. *J Thorac Cardiovasc Surg.* 2008;135:1174-5.

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Reply to the Editor:

We thank Drs Jayle, Hajj-Chahine, and Corbi for their report concerning an aortic root aneurysm in Loeys-Dietz syndrome (LDS) and their subsequent questions related to our clinical approach to this challenging aortopathy.¹ We congratulate this group on the successful surgical management of an aortic root aneurysm associated with LDS.

Their first question is whether to leave an elephant trunk during arch replacement in LDS. Although an elephant trunk is conceptually a possibility in this situation, it is not our standard approach at the University

of Pennsylvania because typically the descending aorta has a normal caliber until it dissects later in the natural history of this disease.²

Their second question relates to timing of surgical intervention for the aortic root in LDS. At the University of Pennsylvania, we recommend aortic root replacement in adults with LDS when the aortic root diameter is 40 to 45 mm. We typically also like to intervene before the development of aortic insufficiency so that, in the absence of aortic valve fenestrations, the native aortic valve can be preserved for optimal hemodynamics and long-term freedom from anticoagulation. In the setting of an aortic root replacement in LDS, we also favor hemiarch replacement under deep hypothermic circulatory arrest, not only to remove all the crossclamped aorta (which is likely at even higher risk for dissection) but also to protect further against future dissection of the distal ascending aorta and aortic arch.^{1,2-4}

Their third question concerned the ongoing surgical management of these patients with this aggressive aortopathy. At the University of Pennsylvania we recommend meticulous monitoring of the thoracic aorta for life in all patients with LDS. This clinical surveillance also includes regular thoracic aortic imaging to detect aneurysm and/or dissection to allow early surgical intervention to prevent death related to the aorta in this syndrome.

In summary, our clinical experience with LDS at the University of Pennsyl-

vania is that the surgical management of the thoracic aorta should be characterized by regular clinical examination, aortic imaging, and an aggressive surgical approach. The aortopathy associated with LDS merits meticulous management, given its life-threatening natural history.

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References

1. Augoustides J, Plappert T, Bavaria JE. Aortic decision-making in the Loeys-Dietz syndrome: aortic root aneurysm and a normal-caliber ascending aorta and aortic arch. *J Thorac Cardiovasc Surg.* 2009; 138:502-3.
2. Lee RS, Fazel S, Schwarze U, Fleischmann D, Berry GJ, Liang D, et al. Rapid aneurysmal degeneration of a Stanford type B aortic dissection in a patient with Loeys-Dietz syndrome. *J Thorac Cardiovasc Surg.* 2007;134:242-3.
3. Melenovsky V, Adamira M, Kautznerova D, Voska L, Weichet J, Loeys B, et al. Aortic dissection in a young man with Loeys-Dietz syndrome. *J Thorac Cardiovasc Surg.* 2008;135:1174-5.
4. Williams JA, Loeys BL, Nwakanma LU, Dietz HC, Spevak PJ, Patel ND, et al. Early surgical experience with Loeys-Dietz: a new syndrome of aggressive thoracic aortic aneurysm disease. *Ann Thorac Surg.* 2007;83:S757-63.

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Notice of Correction

Re: CCR3- and CXCR4-mediated interactions regulate migration of CD341 human bone marrow progenitors to ischemic myocardium and subsequent tissue repair. *J Thorac Cardiovasc Surg.* 2008;136:1044-53.

In the above-noted article, the spelling of Dr Sondermeijer's last name was incorrect. The corrected author list is printed below.

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