

Unusual Cause of Heart Failure in a Patient with Marfan Syndrome:

A Late Complication of Bioprosthetic Valved Graft Replacement

Perna B. Bansal, MBBS
Hari P. Chaliki, MD
Kantha R. Kolla, MBBS
Roger L. Click, MD, PhD
Alberto Pochettino, MD

A high-velocity gradient across the left ventricular outflow tract is most often caused by aortic valve stenosis. We describe the unusual case of a high-velocity gradient caused by a kinked ascending aortic graft in a 69-year-old man who had Marfan syndrome. The patient had a history of ascending aortic aneurysm and had previously undergone replacement of the aortic root and ascending aorta with use of a bioprosthetic valved graft. The kinking was caused by dilation of the native aortic arch. The patient underwent successful hemi-arch replacement and repair of the kinked graft. Late complications and reoperation after proximal aortic surgery in patients with Marfan syndrome are rare, and a high-velocity left ventricular outflow tract gradient caused by the kinking of the aorta is unusual. (Tex Heart Inst J 2020;47(1):38-40)

Increased echocardiographic Doppler gradients across the aortic valve (AV) and left ventricular outflow tract (LVOT) usually raise concern for valvular stenosis,¹ LVOT obstruction,² subvalvular stenosis,³ or supra-ventricular stenosis.⁴ We describe the case of a patient in whom a high-velocity Doppler gradient across the LVOT resulted from an unusual cause.

Key words: Aortic diseases/pathology; echocardiography, Doppler/methods; heart failure/etiology; Marfan syndrome/complications; reoperation; treatment outcome; ventricular outflow obstruction/diagnostic imaging/etiology/physiopathology

From: Division of Cardiovascular Diseases (Drs. Bansal, Chaliki, and Kolla), Mayo Clinic, Scottsdale, Arizona 85259; and Division of Cardiovascular Diseases (Dr. Click) and Department of Cardiovascular Surgery (Dr. Pochettino), Mayo Clinic, Rochester, Minnesota 55905

Address for reprints:
Hari P. Chaliki, MD,
Division of Cardiovascular
Diseases, Mayo Clinic,
13400 E. Shea Blvd.,
Scottsdale, AZ 85259

E-mail:
chaliki.hari@mayo.edu

© 2020 by the Texas Heart®
Institute, Houston

Case Report

In 2014, a 69-year-old man with Marfan syndrome was admitted to our hospital for exacerbation of heart failure. Eleven years earlier, he had undergone replacement of the aortic root and ascending aorta with use of a composite bioprosthetic valve graft to repair an ascending aortic aneurysm. One month before admission to our hospital, a 2-dimensional transthoracic echocardiogram was obtained at another institution when the patient presented with dyspnea; it showed thickened AV leaflets and an increased mean AV gradient (38 mmHg), which suggested prosthetic AV obstruction.

On admission, the patient had a harsh systolic murmur radiating throughout the precordium and a loud second heart sound. A chest radiograph showed pulmonary edema. The level of N-terminal fragment of the precursor to B-type natriuretic peptide was elevated (1,500 pg/mL). Echocardiograms showed a normal left ventricular ejection fraction of 0.60 to 0.65 and a 2-mmHg mean AV gradient from apical view. However, a high-velocity Doppler signal was obtained from the right parasternal position (>4 m/s), with a mean gradient of 48 mmHg (Fig. 1). This finding of a high-velocity mean gradient was not consistent with the normal visual appearance of the motion of the prosthetic AV leaflet (Fig. 2); therefore, we suspected aortic conduit obstruction. Further evaluation with use of magnetic resonance angiography showed severe kinking of the ascending aortic conduit 4 cm above the AV (Fig. 3).

The patient underwent hemi-arch replacement and repair of the kinked portion of the graft. The kink in the ascending aortic graft (Fig. 4) was attributed to dilation of the patient's native aortic arch, which displaced the anastomotic site at the distal ascending aorta caudad. The bioprosthetic AV was functioning normally. After graft replacement, an intraoperative transesophageal echocardiogram revealed no kinking in the ascending aorta and a normally functioning prosthetic AV. The patient was discharged from the hospital 8 days later.

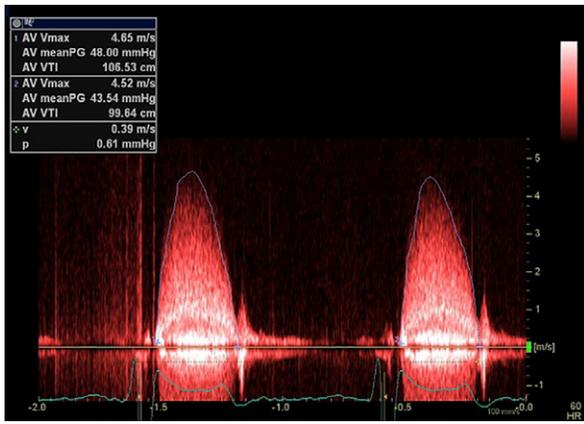


Fig. 1 Transthoracic Doppler ultrasonogram shows a high-velocity jet from the right parasternal window.

AV = aortic valve; PG = pressure gradient; Vmax = maximal velocity; VTI = velocity time integral

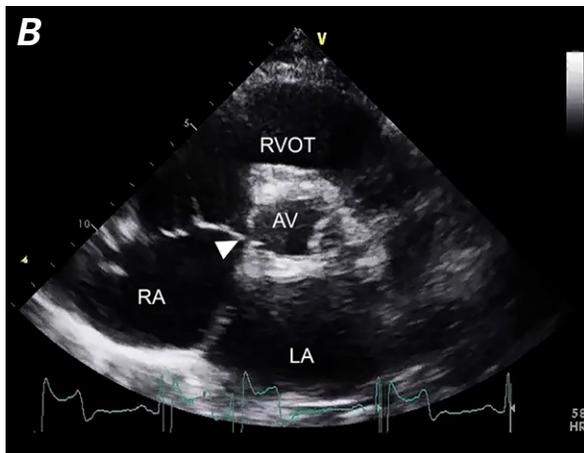
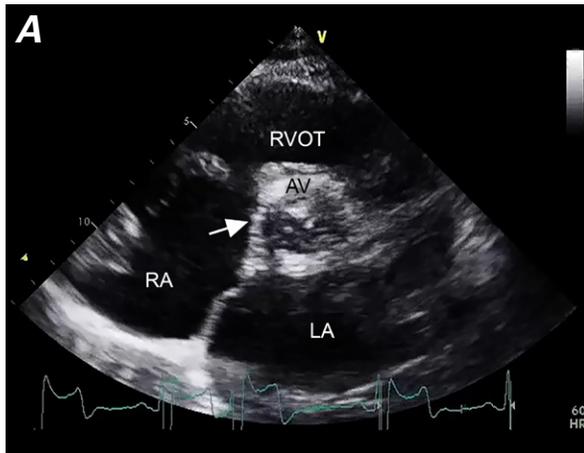


Fig. 2 Two-dimensional transthoracic echocardiograms. Views of the bioprosthetic aortic valve (AV) **A**) closed in diastole (arrow) and **B**) open in systole (arrowhead) show normal appearance without severe stenosis.

LA = left atrium; RA = right atrium; RVOT = right ventricular outflow tract

Supplemental motion image is available for [Figure 2](#).

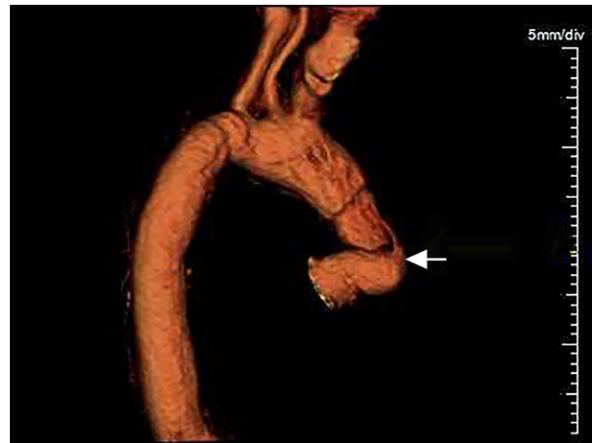


Fig. 3 Magnetic resonance angiogram shows kinking of the Dacron graft 4 cm above the aortic valve (arrow).

Supplemental motion image is available for [Figure 3](#).

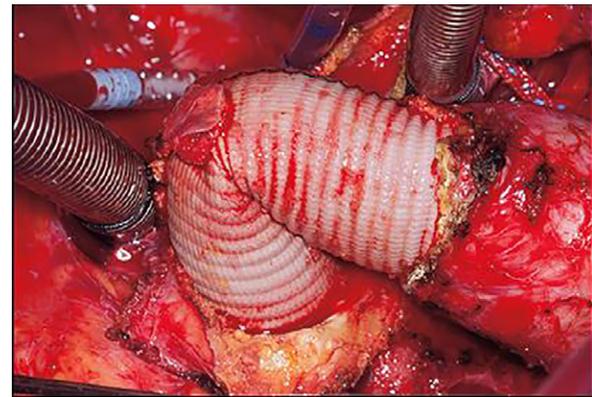


Fig. 4 Intraoperative photograph shows the severely kinked portion of the ascending aortic Dacron graft.

Discussion

Marfan syndrome, an autosomal dominant systemic disorder of the connective tissue, affects mostly the ocular, musculoskeletal, and cardiovascular organ systems. For patients who undergo proximal aortic surgery, 5% to 35% will need an additional aortic operation, depending on the extent of previous surgery and the quality of follow-up.⁵ Risk factors for late reoperations are acute or chronic dissections at the time of initial surgery, aneurysms involving major peripheral arteries, hypertension, and a history of smoking.⁶

Causes of increased echocardiographic Doppler gradients across the AV and LVOT include valvular stenosis, hypertrophic cardiomyopathy,¹ supra-aortic stenosis,⁴ subaortic obstruction,³ accessory mitral valve tissue attached to the ventricular side of the anterior mitral valve leaflet,² and mitral valve replacement with a high-profile mechanical or bioprosthetic valve.⁷⁻⁹ This case illustrates an unusual cause of a high-velocity gradient along the LVOT that was unrelated to prosthetic AV

dysfunction. Increased LVOT gradient and compromised hemodynamic status resulting from the kinking of an aortic graft is a rare occurrence.⁸

Echocardiography has a key role in diagnosing the cause of elevated pressure gradients across the LVOT. The nature of our patient's systolic murmur, however, would have made diagnosis difficult from the typical left parasternal or apical view. For all patients with suspected aortic disease, the European Association of Echocardiography recommends the use of right parasternal views.¹⁰ In our patient, the right parasternal view showed the highest Doppler velocity, which was disproportionately high for an aortic prosthesis that appeared to be normal. This was consistent with the physical examination findings, and it suggested obstruction of the aortic conduit. A magnetic resonance angiogram then showed kinking of the aortic graft due to dilation of the native aortic arch, warranting surgery. Our case demonstrates that high velocity across the LVOT can result from kinking of the aortic graft and that the aortic graft should be evaluated carefully in these patients.

References

1. Maron BJ, Maron MS, Wigle ED, Braunwald E. The 50-year history, controversy, and clinical implications of left ventricular outflow tract obstruction in hypertrophic cardiomyopathy: from idiopathic hypertrophic subaortic stenosis to hypertrophic cardiomyopathy. *J Am Coll Cardiol* 2009;54(3):191-200.
2. D'Aloia A, Vizzardi E, Chiari E, Fracassi F, Zanini G, Faggiano P, Dei Cas L. Dynamic mild subaortic left ventricular obstruction caused by an accessory mitral valve attached to the anterior mitral valve in a young pregnant woman. *Eur J Echocardiogr* 2008;9(1):160-1.
3. Ayoub C, Brieger D, Chard R, Yiannikas J. Fixed left ventricular outflow tract obstruction mimicking hypertrophic obstructive cardiomyopathy: pitfalls in diagnosis. *Echocardiography* 2016;33(11):1753-61.
4. Ghodduci KM, Antic M, Tanaka K, Verdries D, Kerkhove D. Sporadic supra-ventricular stenosis in a young man. *JBR-BTR* 2013;96(1):27-9.
5. Carrel T, Beyeler L, Schnyder A, Zurmuhle P, Berdat P, Schmidli J, Eckstein FS. Reoperations and late adverse outcome in Marfan patients following cardiovascular surgery. *Eur J Cardiothorac Surg* 2004;25(5):671-5.
6. Kawamoto S, Bluemke DA, Traill TA, Zerhouni EA. Thoracoabdominal aorta in Marfan syndrome: MR imaging findings of progression of vasculopathy after surgical repair. *Radiology* 1997;203(3):727-32.
7. Jett GK, Jett MD, Barnhart GR, van Rijk-Swikker GL, Jones M, Clark RE. Left ventricular outflow tract obstruction with mitral valve replacement in small ventricular cavities. *Ann Thorac Surg* 1986;41(1):70-4.
8. Al-Mohaisen MA, Skarsgard P, Khoo C, Virani SA, Munt B, Ignaszewski A. Expanding arch aneurysm causing a "kink" in a Bentall graft and heart failure. *J Thorac Imaging* 2012;27(4):W88-90.
9. Numaguchi K, Hatake S, Ueda T, Tomita Y. A kinked aortic tube graft after aortic root replacement causes heart failure: usefulness of the right parasternal approach on echocardiography. *J Med Ultrason* (2001) 2015;42(2):291-2.
10. Evangelista A, Flachskampf FA, Erbel R, Antonini-Canterin F, Vlachopoulos C, Rocchi G, et al. Echocardiography in aortic diseases: EAE recommendations for clinical practice [published erratum appears in *Eur J Echocardiogr* 2011;12(8):642]. *Eur J Echocardiogr* 2010;11(8):645-58.