We present the case of a 39-year-old physically active male amateur hockey player with a history of preductal coarctation of the aorta repaired at the age of 10 years with an ascending-to-descending aortic conduit. He presented with neurological symptoms, which were present at rest but paradoxically resolved with physical exertion, including left upper extremity weakness, dizziness, visual changes, and facial tingling.

A 2008 echocardiogram demonstrated a bicuspid aortic valve with moderate regurgitation and a mildly to moderately dilated and hypertrophied left ventricle with ejection fraction of 50%. A 2009 carotid Doppler ultrasound examination demonstrated high-velocity retrograde flow in the left vertebral artery, which reduced on exertion. Stenosis of the subclavian artery, and thus subclavian steal, which has been shown to cause neurological symptoms, was ruled out by a 2010 arteriogram.1 A recent magnetic resonance imaging (MRI) evaluation (in 2011) demonstrated worsening (moderate to severe) aortic insufficiency with a clearly visible asymmetrical diastolic flow jet (Figure 1, white arrows), mild decrease of left ventricular ejection fraction, and a patent aortic bypass conduit.

To further interrogate the paradoxical reversal of neurological symptoms on exertion, we performed flow-sensitive 4D MRI.2 The aim was to evaluate time-resolved 3D blood flow during rest and exertion with full volumetric coverage of the entire aorta, bypass conduit, and supra-aortic vessels (1.5-Tesla MRI; Siemens, Erlangen, Germany; velocity sensitivity=150 cm/s, spatial resolution 2.6×2.6×3.2 mm³, temporal resolution=38 ms, ECG gating, respiratory navigator gating, scan time=20 minutes). Flow-sensitive 4D MRI was performed after administration of intravascular gadolinium contrast agent (Ablavar; Lantheus Medical Imaging, North Billerica, MA; dose=0.03 mmol/kg) at rest. Imaging was repeated after administration of 1.5 mg of atropine intravenously in an attempt to evaluate the effect of exercise on aortic hemodynamics. A moderate increase in heart rate was achieved (from 45 bpm to 67–71 bpm). The study was approved by the institutional review board, and informed consent was obtained from the patient.

To visualize blood flow in 3D, we calculated time-resolved path lines depicting the direction and temporal evolution of flow through the entire thoracic aorta, conduit, and 3 supra-aortic branches.3 4D flow data were also used to retrospectively quantify retrograde fraction (retrograde flow divided by antegrade flow) at several locations in the vessels.

Figure 1. Cardiac magnetic resonance angiography demonstrating aortic valve insufficiency and a clearly visible asymmetrical flow jet (white arrows, regions with signal void in the left ventricle). The individual images represent sagittal (left) and coronal (right) reformats of a 3D respiratory and ECG-gated magnetic resonance angiography acquisition. AAo indicates ascending aorta; LV, left ventricle; and RV, right ventricle.
3D flow visualization demonstrated high flow in both the native aorta and bypass graft (Figure 2), with substantial diastolic retrograde flow throughout the system (Figure 3). The complex dynamics of 3D blood flow are best appreciated in the online-only Data Supplement Movie. Specifically, the retrograde fractions in the conduit, innominate, left common carotid, and left subclavian arteries were consistently reduced after administration of atropine, as summarized in Figure 2.

Our patient’s symptoms closely mimicked those of subclavian steal, which occurs when stenosis of the left subclavian artery induces retrograde flow in the left vertebral artery and causes symptoms of vertebrobasilar insufficiency. Although ultrasound examination demonstrated retrograde flow in the left vertebral artery, angiography ruled out subclavian stenosis and thus subclavian steal.

4D flow analysis demonstrated high flow velocities through the bypass conduit and native hypoplastic descending aorta and, most noticeably, enhanced diastolic retrograde flow throughout the system, likely exacerbated by the severe aortic regurgitation. The worsening of aortic insufficiency, demonstrated on serial imaging examinations, correlates well with the progression of symptoms and provides an explanation for the absence of neurological symptoms during the first 25 years after repair.

Interestingly, the patient’s symptoms resolved with exertion. We attempted to recreate this effect during 4D flow analysis with the administration of atropine. Although the patient only achieved a modest increase in heart rate, the retrograde flow fraction in the system noticeably decreased, especially in the left common carotid and subclavian arteries.

As evident in Figure 3, retrograde flow in the supra-aortic vessels was present even in late diastole. As a result, the low resting heart rate (45–50 bpm) of this physically fit patient (a hockey player) may have contributed to the neurological impact of the retrograde flow. Shorting of diastole associated with an increase in heart rate may thus offer a potential explanation for the reduction of retrograde flow and neurological improvement during exercise.

Flow-sensitive 4D MRI analysis helped to elucidate the complex flow dynamics and offered an explanation for the paradoxical resolution of symptoms with exertion and helped guide management. The patient is scheduled for aortic valve replacement.

Disclosures
None.

References
Figure 3. Temporal evolution of 3D path lines over the cardiac cycle (before atropine). The individual images represent selected systolic (top row) and diastolic (bottom row) cardiac time frames. High systolic velocities and thus earlier wave reflection at the periphery in combination with aortic valve insufficiency resulted in substantial retrograde flow throughout the entire thoracic aorta, even reaching the distal descending aorta (DAo). Retrograde flow clearly extended into the supra-aortic branches, which offers a potential explanation for neurological symptoms due to insufficient delivery of blood to the cranial circulation. See also online-only Data Supplement Movie. AAo indicates ascending aorta.
Four-Dimensional Magnetic Resonance Flow Analysis Clarifies Paradoxical Symptoms in a Patient With Aortic Bypass and Retrograde Flow Mimicking Subclavian Steal

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