Case 8

Turner Syndrome with Aortic Coarctation and Thin Intraluminal Obstructive Ring: A Critical Stenosis Preserved by Iterative Reconstruction

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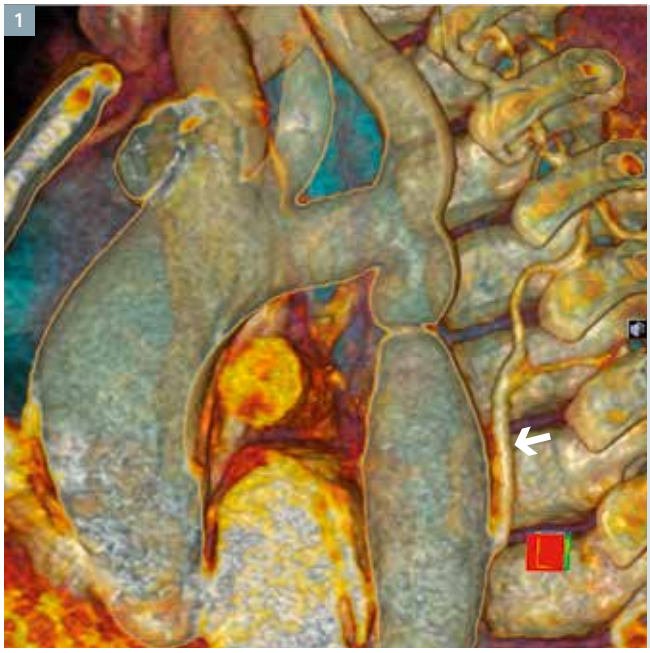
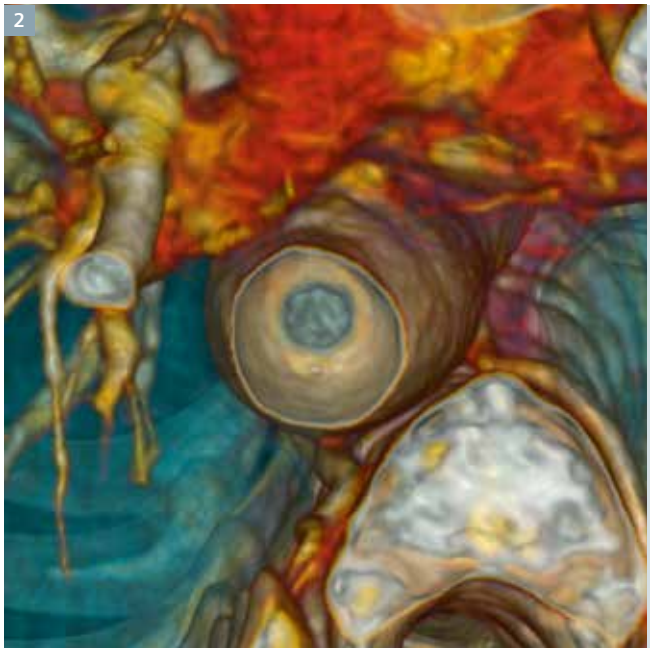


1

Volume-rendered view of the aortic arch: Note the hour-glass outer contour and the web-like ring within the flow lumen. A prominent intercostal artery drains into the distal aorta (arrow).



2

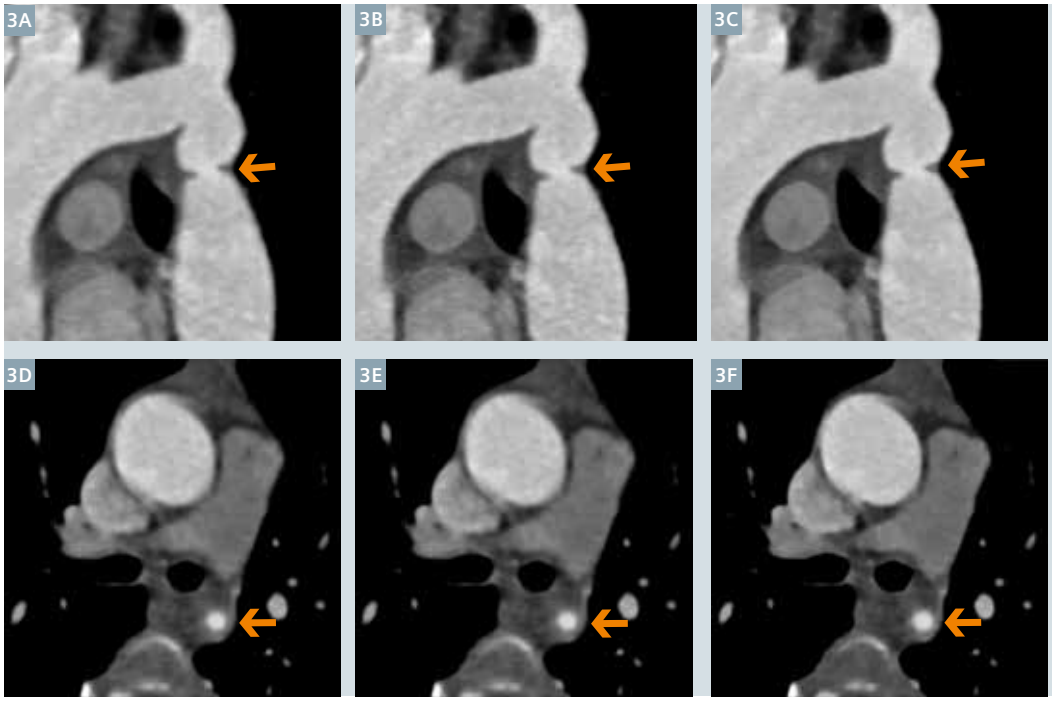
Volume-rendered view from the top of the aortic arch: It shows the opening of the intraluminal ring.

History

A 7-year-old girl was presented for an evaluation of her small stature when compared to her peers. She was evaluated by an endocrinologist and diagnosed with Turner syndrome. Due to the association of Turner syndrome with cardiovascular abnormalities, an echocardiography was performed. The echocardiography showed a discrete aortic narrowing, consistent with an aortic coarctation. The peak pressure gradient estimated by echocardiography was 50 mmHg. In order to decide between endovascular treatment with angioplasty and stenting versus surgical repair, a CT angiography of the chest was performed to define the length and narrowness of the coarctation, its distance to the cervical arteries, and any stenoses in these arteries. The coarctation was surgically resected and the aorta repaired with an end-to-side connection.

Diagnosis

The aortic arch was right-sided with a normal branching order of the cervical arteries. The coarctation was located distal to the left subclavian artery at the juxtaductal position (Fig. 1). All cervical arteries were patent. Prominent intercostal arteries (arrow) conducted retrograde collateral flow into the distal aorta, bypassing the obstruction. The outer contour of the coarctation had an “hour-glass” shape with a diameter of 9 mm at the waist, but inside the coarctation there was a web-like ring that further narrowed the flow lumen to a diameter of 4 mm (Fig. 2).



3

Multiplanar reformat of the web-like obstruction (arrows) in the sagittal plane (Figs. 3A–3C) and in the axial plane (Figs. 3D–3F) at different SAFIRE levels: Figs. 3A and 3D: none; Figs. 3B and 3E: SAFIRE level 2; Figs. 3C and 3F: SAFIRE level 4. The geometry of the obstruction is essentially unchanged.

Comments

Turner syndrome is a genetic disease in which the patient has only one copy of the sex-determining chromosome, an X-chromosome, without a second X- or Y-chromosome. Patients with Turner syndrome are phenotypically female. The disease is associated with a number of congenital cardiovascular abnormalities including coarctation of the aorta, bicuspid aortic valve, aortic valve stenosis, partial anomalous pulmonary venous return, and others. Coarctation, which occurs in 17% of these patients, is the most frequent and clinically important abnormality.

Management guidelines for Turner syndrome include noninvasive imaging, such as echocardiography and MR angiography, to detect coarctation.[1] CT angiography is an alternative examination if MR angiography is contraindicated or high-resolution imaging is needed for endovascular or surgical treatment planning. Surgical repair is preferred in young patients when their aortas have yet to grow to full size. An endovascular approach may be used in older patients with fully developed aortas.

The use of iterative image reconstruction, such as SAFIRE, enables CT scans with reduced radiation dose without associated increase in image noise. However, the image reconstruction must not obscure or alter diagnostically important features. This case demonstrated that an obstructive ring as thin as one millimeter was preserved by SAFIRE (Fig. 3). Accurate representation of a vascular obstruction was important not only for diagnostic assessment and treatment planning, it also enables the calculation of hemodynamic information, such as pressure gradients, shear stress, and ventricular loading by computational fluid dynamics.[2] ■

References

[1] Bondy CA and for The Turner Syndrome Consensus Study Group. Care of Girls and Women with Turner Synbdrome: A Guideline of the Turner Syndrome Study Group. J Clin Endocrinol Metab. 2007, 92(1): 10-25.

[2] Coogan FP, et al. Computational Fluid Dynamic Simulations for Determination of Ventricular Workload in Aortic Arch Obstructions. J Thorac Cardiovasc Surg. 2013, 145(2): 489-495.

In clinical practice, the use of SAFIRE may reduce CT patient dose depending on the clinical task, patient size, anatomical location, and clinical practice. A consultation with a radiologist and a physicist should be made to determine the appropriate dose to obtain diagnostic image quality for the particular clinical task. The following test method was used to determine a 54 to 60% dose reduction when using the SAFIRE reconstruction software. Noise, CT numbers, homogeneity, low-contrast resolution and high contrast resolution were assessed in a Gammex 438 phantom. Low-dose data reconstructed with SAFIRE showed the same image quality compared to full dose data based on this test. Data on file.

Examination Protocol

Scanner	SOMATOM Definition Flash
Scan area	Thorax
Scan length	160 mm
Scan direction	Cranio-caudal
Scan time	0.4 s
Tube voltage	80 kV
Tube current	60 eff. mAs
Dose modulation	CARE Dose4D
$CTDI_{vol}$	0.97 mGy
DLP	25 mGy cm
Effective dose	1 mSv[1]
Rotation time	0.28 s
Pitch	3
Slice collimation	128 × 0.6 mm
Slice width	0.6 mm
Reconstruction increment	0.6 mm
Reconstruction kernel	B26f/I26f (SAFIRE)
Contrast	350 mg/mL
Volume	30 mL
Flow rate	2 mL/s
Start delay	Bolus tracking

[1] Estimated by applying a conversion factor of 0.018, and an additional factor of 2.3 converting the reported DLP (32 cm) into the DLP (16 cm).