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Brief Report

Restenosis after aortic stenting

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Abstract Aortic coarctation is a recognized arteriopathy in patients with Williams' syndrome. We present an adolescent with Williams' syndrome who developed rapid restenosis after primary stenting of coarctation of the aorta. We believe such restenosis within the stent is due to a proliferative response of the abnormal aortic wall. Attention should be paid to the potential for restenosis during follow-up after stenting of aortic coarctation in patients with Williams' syndrome.

Keywords: William's syndrome; coarctation of aorta; arteriopathy

COARCTATION OF AORTA IS A RECOGNISED arteriopathy of Williams' syndrome.[1] Primary stenting of coarctation is an accepted intervention with results comparable to surgical repair.[2] As far as we are aware, there is only one reported case of failure of stenting in an infant with Williams' syndrome who had stenting of recoarctation following previous end-to-end repair.[3] We present an adolescent with Williams' syndrome who developed rapid restenosis within a stent.

Case report

BL is a 17-year-old boy who has Williams' syndrome with mild supravalvar aortic stenosis, and has been followed up since infancy. He was noted to be hypertensive when he was admitted to his local hospital with meningococcal meningitis at the age of 14 years. Further investigations were performed during follow-up, and showed widespread arteriopathy.

An echocardiogram, and subsequent angiography, revealed coarctation of thoracic aorta distal to origin of left subclavian artery. He also had bilateral intrarenal vascular disease, generalised mild cerebrovascular disease, and so-called "optic nerve drusen", extracellular deposits below the pigmented epithelium of the retina that are unrelated to Williams' syndrome.

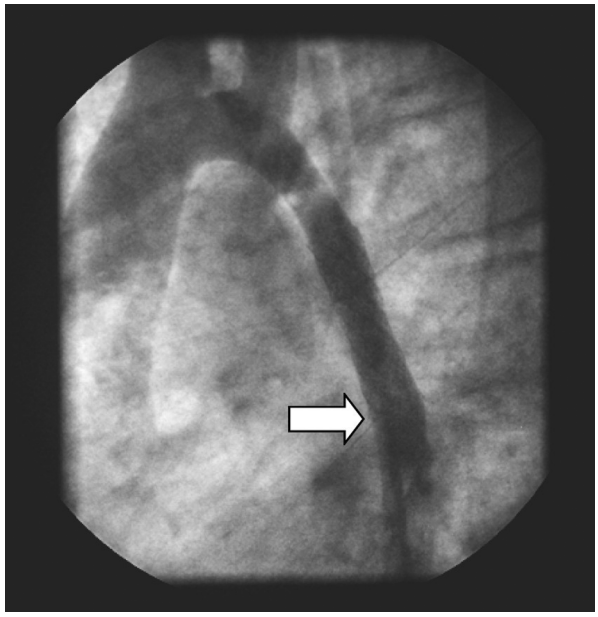

Figure 1.
The aortogram following stenting of the coarctation (arrow).

Primary stenting of the area of coarctation was performed in 2001 using a Cordis Palmaz P308 stent (Roden, The Netherlands), premounted on a 8 mm balloon, and dilated to 10 mm with a Cordis Power Flex Plus balloon (Fig. 1).

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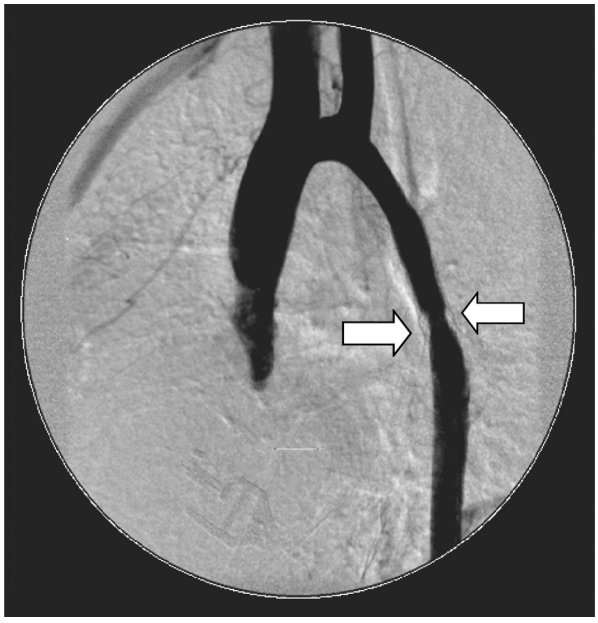

Figure 2.
The interval aortogram shows reduction of the calibre of the lumen within the stent (arrows).

Angiography was repeated after 18 months because of persistent hypertension. There was restenosis within the stent, with a gradient measured of 32 mmHg (Fig. 2). The descending aorta was also mildly hypoplastic. Intervention was thought to be unnecessary at this stage, and he is being followed up closely.

Discussion

Williams' syndrome is a contiguous gene syndrome caused by a deletion of multiple genes on the long arm of chromosome 7. Arteriopathy is caused by deficiency of elastin, and affects both small and large vessels. Arterial histology shows a haphazard arrangement of elastic fibres, excessive collagen, and hypertrophied smooth muscle. There is scant ground substance in the medial layer, with quantitative deficiency of elastic fibres.[4] There may also be significant neointimal proliferation resulting in acquired coarctation.[1]

Restenosis after primary stenting of coarctation of aorta is rare in the general population.[2] In the 2 published case reports of restenosis within a stent, one of the patients had Williams' syndrome.[3,5] Histopathologic examination of the resected aortic wall demonstrated extensive fibrosis with proliferation of intimal and smooth muscle cells.[3]

We believe such stenosis within a stent placed in the aorta of patients with Williams' syndrome is due to an abnormal proliferative response of the aortic wall. Heightened awareness of such stenosis should now predicate extra careful follow-up of patients with Williams' syndrome after aortic stenting.

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