Supravalvular aortic stenosis and total occlusion of the brachiocephalic trunk

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In a 4-year-old girl supravalvular aortic stenosis was diagnosed by echocardiography. Her younger brother was operated on for the same heart disease one year before. Since chromosomal disorder and Williams-Beuren syndrome were excluded we made the diagnosis of familial supravalvular aortic stenosis in these sibs.¹ Heart catheterisation confirmed the diagnosis and, surprisingly, a total occlusion of the brachiocephalic trunk (BT) was demonstrated (figure 1). Cerebral and thoracic/abdominal MR angiography showed retrograde filling of the right-

sided neck vessels with symmetrical jugular venous return (figure 2). No other systemic arteries were affected.

Total occlusion of the BT is not known in relation to supravalvular aortic stenosis.² Complete occlusion of the majority of aortic arch vessels is well known in the spectrum of Takayasu disease, but this patient has no signs of this disease.³ A 78-year-old woman with complete occlusion of the BT is reported without mention of a possible cause.⁴ ■

Figure 1. Aortogram. A) Normal left and right coronary artery, mild supravalvular stenosis (SVS). B) Mild SVS and occlusion of brachiocephalic trunk (BT). C) Occlusion of BT, no stenosis left carotid artery, no aortic coarctation.

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Figure 2. Thoracic/Cerebral MRA. A) Arterial phase with occlusion of brachiocephalic trunk and retrograde filling of the right internal/external carotid artery and right vertebral/subclavian artery. B) Symmetrical jugular venous return.

References

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- 2 Eronen M, Peippo M, Hiippala A, et al. Cardiovascular manifestations in 75 patients with Williams syndrome. *J Med Genet.* 2002;39:554-8.
- 3 Cheng C, Tsao T, Peng C Yang S. Severe arch vessel disease. *Neth J Med.* 2008;66:91-2.
- 4 Thalhammer C, Aschwanden M, Jager K. Occlusion of the brachiocephalic trunk. *Vasc Med.* 2004;9:313-4.

In this section a remarkable 'image' is presented and a short comment is given.

We invite you to send in images (in triplicate) with a short comment (one page at the most) to Bohn Stafleu van Loghum, PO Box 246, 3990 GA Houten, e-mail: l.meester@bsl.nl.

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