An unusual murmur: The ‘Gothic’ aortic arch

Un souffle inhabituel : l’arche aortique « gothique »

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A 19-year-old man with a history of successful end-to-end anastomosis surgical repair of coarctation of the aorta was admitted to our institution for a murmur evaluation before applying for a sport licence. He was asymptomatic and was not taking any medication. A systolic murmur was heard in the aortic area and in the back. The patient was normotensive, with 120/60 mmHg on each arm. Echocardiography did not show valvular disease or intraventricular obstruction. The ascending aorta was normal and there was an acute angulation between the ascending and the descending aorta, without the horizontal part. This is characteristic of a ‘Gothic’ arch (Fig. 1). There was neither two-dimensional nor Doppler evidence of recoarctation. Cine cardiac magnetic resonance showed a hypointense jet in the angulation of the aorta (Fig. 2). Such an image is associated with acceleration of blood velocity in the angulation, which is responsible for the murmur. Unfortunately, no flow sensitive sequence was performed. Exercise testing identified an induced hypertension, reaching 250/90 mmHg, without any difference between arms.

Aortic arch growth after coarctation repair leads to three different shapes: Romanesque (normal), Crenel (rectangular form) and Gothic. The evolution mode leading to each shape is unknown. Resting and induced hypertension are long-term common complications after successful repair of coarctation of the aorta. An association between aortic arch deformation with ‘Gothic’ geometry and both resting and induced hypertension has been recently suggested. This hypothesis is still under discussion, but cardiologists should remember this anatomic form and its potential association with hypertension (see supplementary material).

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Figure 1. Two-dimensional suprasternal echocardiography view of the aortic arch showing a ‘Gothic’ shape characterized by an acute angulation between the ascending and the descending aorta. Left subclavian artery origin is in the descending part (arrow).

Figure 2. Cine cardiac magnetic resonance imaging confirms the successful repair of the aorta with no recoarctation, and visualizes a ‘Gothic’ shape (Panel A) and a hypointense jet just after the angulation (Panel B).

Conflict of interest statement

None.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.acvd.2010.05.008.