

Abnormalities of aortic arch shape after successful repair of aortic coarctation and systemic arterial hypertension

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After successful repair of aortic coarctation (AoCo), many patients remain at risk for cardiovascular problems and major adverse prognostic factors have been identified¹⁻⁴. After AoCo repair, different pathogenic mechanisms leading to persistent systemic arterial hypertension have been identified, including mild residual arch stenosis, increased aortic wall stiffness, hyperactivity of renin-angiotensin system, impairment of the baroreflex system and peripheral vascular reactivity⁴.

An abnormal aortic arch shape has been associated with rest or exercise-induced arterial hypertension in the long-term follow-up of patients after repair of AoCo, without any residual arch obstruction^{3,5}. Native aortic arch morphology, age at repair and surgical approach were all identified as possible causes for a persistent postoperative abnormal arch shape; however all these findings are still debated. An abnormal arch shape with either gothic or crenel morphology after surgical repair leads to an abnormal central aortic flow dynamic and increased aortic wall stiffness⁶, which can both contribute to the development of systemic arterial hypertension in the long-term^{7,8} (see Figure 1 and Figure 2).



Figure 1. MR angiography of an aortic arch morphology in a patient following AoCo repair with normal arch shape.

Figure 1a. Dasc Ao: diameter of distal ascending aorta (before the brachiocephalic trunk); Dt₁: diameter of proximal transverse arch (between brachiocephalic trunk and left carotid artery); Dt₂: diameter of distal transverse arch (between left carotid artery and left subclavian artery); Dt₃: diameter of the isthmus (after the left subclavian artery); Dd: diaphragmatic aorta diameter.

Figure 1b. T: maximal distance between the midpoints of the ascending and descending aorta, close to the axial plane going through the right pulmonary artery; A: maximal vertical distance between T and the highest mid-point of the arch.

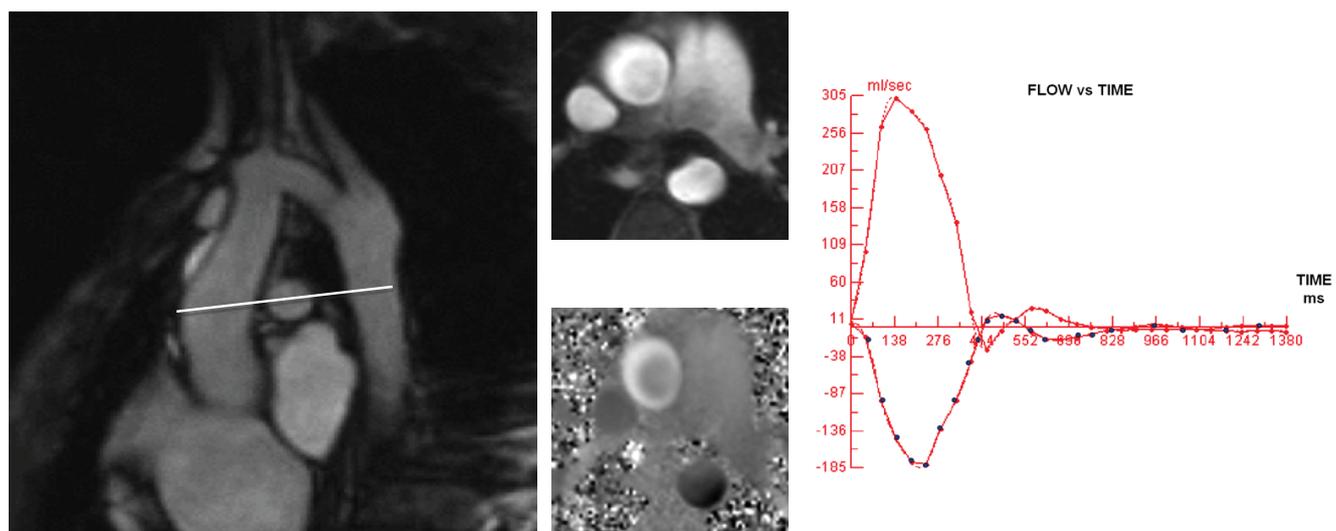


Figure 2. MR cine-sequence of an aortic arch morphology in a patient with gothic arch shape following AoCo repair. Flow time curves of ascending (upper curve) and descending aorta (lower curve) obtained at the level of the white line. From these pictures we can estimate the distensibility of the ascending aorta and the percentage (%) loss of systolic wave amplitude across the aortic arch (see Reference 13 for further details).

It has been widely shown²⁻⁵ that systemic hypertension is a major long-term complication after AoCo repair and moreover that patients with the gothic arch shape have a major risk to become hypertensive in the adult age. The etiology of gothic arch shape is still not clear, with many controversial explanations. The gothic shape after AoCo repair is characterized by a greater aortic arch growth in height rather than in width. Ou et al.³ reported that patients with gothic shape were of an older age, suggesting that this anomaly could be acquired with age. There are also evidences that the type of surgical intervention could influence the prevalence of hypertension in long term. In particular, patients undergoing end-to-end anastomosis have a lower systolic blood pressure at follow-up than those undergoing subclavian flap repair or patch aortoplasty^{2,9,10}. However other Authors report no significant differences in the prevalence of hypertension in relation to surgical repair techniques^{11,12}.

According to published data^{3,5,6,13}, a decreased ascending aorta distensibility coupled with an increased loss of systolic wave amplitude across the arch was reported in patients with a gothic shape after AoCo repair. The triangular-shaped morphology of the gothic arch could simply reflect the systolic wave inducing an early remodeling of the aortic wall. This is also confirmed by the evidence of morpho-functional alterations of the conduit arteries proximal to the AoCo repair site¹⁴⁻¹⁷, which are independent from time of intervention¹⁸⁻²⁰. An increased resistance to pulsed wave propagation across the arch has been documented in the gothic arch shape group⁶. It has been also demonstrated that this arch shape abnormality leads in the long term to structural changes of the aortic wall and is responsible for an increased wall stiffness

leading to exercise and rest hypertension together with left ventricular hypertrophy^{21,22}.

In summary, a vascular remodeling develops after AoCo repair and could be faster in patients with gothic aortic arch shape. Aortic stiffness and increased left ventricular mass are both independent predictors of late morbidity and mortality in hypertensive patients²³.

The gothic shape of the aortic arch, the reduced ascending aorta distensibility and the loss of systolic wave amplitude across the aortic arch, measured by MRI, correlate with systolic blood pressure and pulse pressure on ambulatory monitoring in a group of patients after successful AoCo repair¹³. An elevated pulse pressure is typical in elderly hypertensive patients and is due to a major vascular stiffness. As further confirmation of a pre-hypertensive state, subjects with a gothic arch shape show a higher day and night-time systolic blood pressure and a higher number of blood pressure measurements above the normal values for age¹³.

In conclusion, aortic wall remodeling is present in patients following AoCo repair. Those with a gothic arch shape may develop an earlier vascular stiffness and abnormal vascular hemodynamics, demonstrated also by a higher systolic blood pressure and pulse pressure on ambulatory monitoring and by exercise induced hypertension. These are all predisposing factors in developing arterial hypertension in the adult age.

We believe that all these observations highlight the need for a close follow-up of patients with gothic arch geometry after successful AoCo repair. The beneficial effects on outcome of pressure lowering treatment in subjects with increased central aortic stiffness are still under debate²⁴.

The identification of risk factors for hypertension associated with a gothic arch shape after AoCo repair might also help to improve surgical strategies towards techniques aimed to obtain a more regular and smooth arch geometry at the time of the surgical repair.

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