

“Corkscrew” aortic arch branching pattern

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A 10-month-old infant was referred for cardiac evaluation because of a mild inspiratory stridor during feeding and crying. At rest, no significant cardiac or respiratory finding was evident, except for a soft 2/6 systolic murmur at the cardiac base, widely irradiating to the vessels of the neck. Neither chest X-ray or electrocardiography revealed any significant anomaly. At echocardiography, an anomalous splitting of the ascending aorta prompted us to suspect a double aortic arch malformation as a cause of the “noisy” breathing. A confirmatory bronchoscopy showed a faint pulsatile extrinsic compression on the anterior tracheal wall. Thus, in order to better visualize the anatomic pattern of the aortic arch, a cardiac catheterization was planned. At angiography, an anomalous aortic shape, course and branching pattern were found

(Fig. 1A). The aortic arch started as if it was going to be a right arch and then turned to the left across the mediastinum, to cross over the left main stem bronchus. In addition, the left carotid artery arose very proximally from the ascending aorta and paralleled the transverse aortic arch, accounting for the echocardiographic misdiagnosis of a double aortic arch. Moreover, there was a “corkscrew” appearance of the epiaortic vessels that were coiled from their origin up to the periphery (Fig. 1B). The anomalous course of the aortic arch and its branches presumably caused the mild extrinsic compression of the upper airway imaged at bronchoscopy.

The pathogenetic mechanism of these anomalous anatomic findings remains unknown. The unusual shape of the aortic arch and the airway obstruction might be

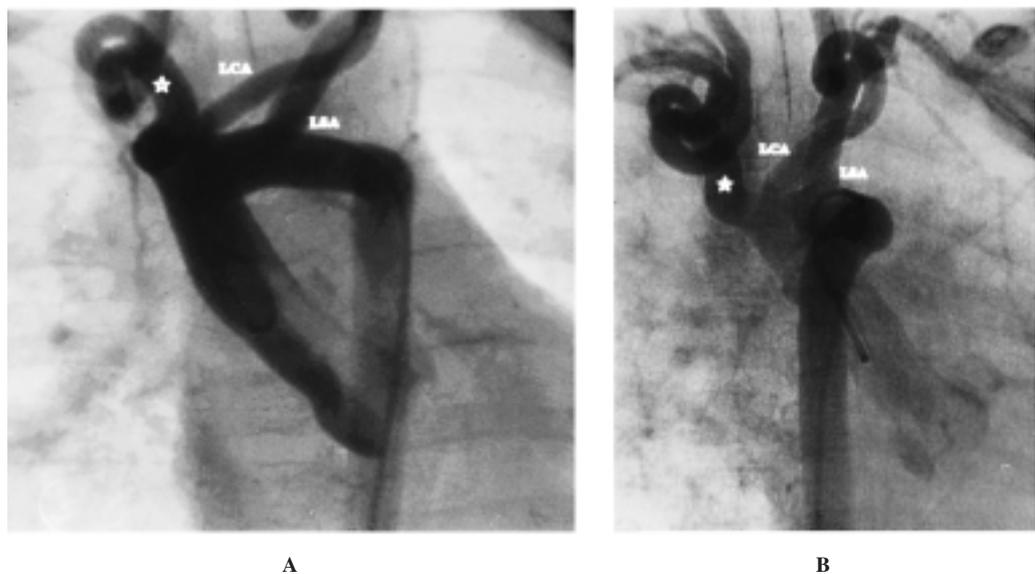


Figure 1. Postero-anterior (A) and right oblique (B) views of the aortic arch branching pattern. Note the “corkscrew” appearance of the epiaortic vessels that spiral downstream from their origin from the ascending aorta. The left carotid artery (LCA) arises very close to the right innominate artery (*) and its horizontal course presumably causes the mild tracheal compression found at bronchoscopy. LSA = left subclavian artery.

due to some tethering down of the right epiaortic vessels by a potentially atretic right arch or right ligamentum arteriosum, so figuring a very unusual form of vascular ring. Another possibility could be that the tortuosity was due to an increased elongation of the aortic arch and of its branches (Fig. 2) resulting from some arterial vasculopathy of unknown origin. This latter hypothesis seems to be supported by the extensive coiling of the peripheral arterial branches, but disproved by the lack of any systemic inflammatory sign at both clinical evaluation as well as at blood analysis.

Due to the mildness of the respiratory symptoms, the lack of any significant obstruction across the aortic arch and its epicardial vessels, as well as the perceived unfavorable cost-effectiveness of any surgical option, it was decided to submit the patient to close clinical follow-up and to perform surgery in case of significant worsening of the respiratory symptoms.



Figure 2. Selective injection into the right common carotid artery that shows the coiled peripheral branches of both the internal and external carotid arteries (arrows). The faint opacification of the coiled origin of the right subclavian artery is also evident.