

## Kinking of the aorta with calcified aortic valve stenosis: A case report



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### ABSTRACT

Congenital kinking of the aorta is an uncommon anomaly consisting of elongation of the aortic arch with kinking at the level of the ductal ligament. Herein we report a case of congenital kinking of the aorta with calcified aortic valve stenosis. The combination of a kinked aorta with severe calcified valve stenosis is very unusual.

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A 44-year-old woman was admitted to our hospital for aortic valve replacement. Her main complaints were cough for the past year, shortness of breath and headache. On admission, her blood pressure was 140/100 mm Hg in the right arm and 120/90 mm Hg in the left arm. Her pulse was regular with a rate of 82 bpm. She presented systolic ejection murmur, a grade of 5/6, with transmission to the interscapular area. She had no family history of heart disease. Echocardiography showed severe aortic valve stenosis with a peak gradient of 103 mm Hg, valve area of 1.1 cm<sup>2</sup>, aortic annulus diameter of 1.6 cm, moderate calcification of aortic valve, left ventricular ejection fraction of 69% and left ventricular hypertrophy (LVH). Electrocardiogram showed LVH. She was referred to catheterization laboratory for preoperative evaluation of the coronary arteries. During the procedure via right femoral artery it was impossible to pass through the isthmus and get into the aortic arch with a 0.035 inch guidewire. Contrast injection at the problematic site raised suspicion for aortic coarctation (Fig. 1). The angiographer did not switch to a radial access and preferred to send her to a CT-angio for evaluation of the isthmic site. Three-dimensional (3D) images obtained by means of contrasted CT-scan showed the following: the aortic arch was kinked and the diameter at the narrowest part, at the level of the isthmus just distal to the left subclavian artery, was 14.3 mm; furthermore, it was dilated just below the kinked site; the diameter of descending thoracic aorta was 34.6 mm and the diameter of ascending thoracic aorta measured 31.7 mm. No collateral circulation was detected; the aortic arch was also kinked between the LCCA and LSCA. The

brachiocephalic trunk was also dilated and had a diameter of 23 mm (Fig. 2).

We performed bronchoscopy to be sure that the kinked site of the aorta doesn't compress the trachea. Otherwise it could create serious problems with the tracheal tube in the postoperative period. She underwent AVR with a 17 mm metallic prosthetic valve. As the aortic root was hypoplastic, the valve was placed obliquely, because it didn't fit in the ordinary way. We didn't touch the kinked site. Postoperative

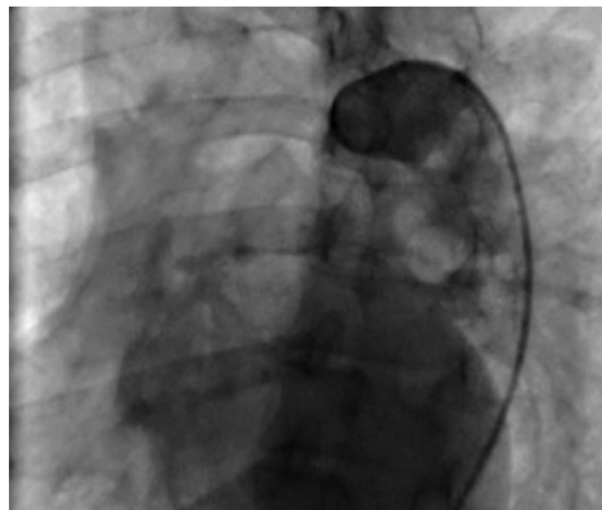


Fig. 1. Aortography at the level of isthmus showing blunt occlusion.

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Fig. 2. CT-scan image.

period went without any complication. She was discharged at the eighth postoperative day.

Two years after the surgery she feels good, with no serious complaints.

### 1. Discussion

Since the term “pseudocoarctation” was first introduced by Dotter and Steinberg in 1951, over 150 cases of the disease have been reported [1,2,7]. Associated congenital lesions reported in the literature have

been coarctation of the aorta, bicuspid aortic valve, congenital aortic stenosis, subaortic stenosis, corrected transposition, ventricular septal defect, patent ductus arteriosus, fibroelastosis and aneurysms of the aortic sinuses [3,4]. Steinberg and Hangstrom reported in 1962 the combination of congenital aortic valve stenosis with kinking of the aorta in four patients [4]. One case of kinking of the aorta with aortic stenosis was reported in a 5-year-old boy by Doris Kavanagh-Gray and Peter Chiu (1970).

The exact etiology of pseudocoarctation of the aorta is currently unknown. It has been proposed that the embryologic cause of pseudocoarctation is a failure of compression of the third through the seventh segments of the dorsal aortic roots and the fourth arch segments [2].

Compared with conventional aortography, 3D CT-angio is less invasive and is the most useful technique available for accurate evaluation of the morphologic orientation of the elongated aortic arch.

Hence, once the diagnosis of congenital kinking of the aorta has been established, a periodic follow-up is necessary to identify aneurismal dilatation [5]. Surgical treatment is recommended for all symptomatic patients and for those with associated aneurysm formation [6].

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