Childhood Solutions That Turn into Adult Problems: Long-term Implications of Congenital Aortic Coarctation Repair

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INTRODUCTION

Annually, congenital coarctation of the aorta (CoA) affects 1:1800 babies in the USA and treatment is early surgical repair. Medical advances are improving the life expectancy of CoA patients. Thus, timely recognition of long-term complications of post-surgical patients is crucial.

CASE REPORT

A 35-year-old female with bicuspid aortic valve, hypertension and a subclavian flap repair of CoA developed elevated supra-valvular gradients at the level of the sinotubular junction. Aortic valve (AV) area by planimetry was 2 cm ², however imaging at post-operative years 20, 24, and 33 showed worsening of AV gradients. Peak velocities were 4.4, 4.6, 4.9 m/sec; peak gradients were 78, 86, 83 mmHg and mean gradients were 42, 55, 58 mmHg, respectively. Multimodal imaging was used to investigate anatomic etiologies for the discrepancy (Figure 1). Echocardiogram revealed turbulent flow proximal and distal to a stenotic portion of the aortic arch, computed tomography angiography and cardiac MRI revealed a kink at the level of the ligamentum arteriosum. Three-dimensional (3-D) cardiac reconstruction confirmed a stable kink in the proximal descending aorta, 2 mm enlargement of aortic root, and a mean pressure gradient of 60.2 mmHg across the stenotic area.

IMAGING

Figure 1: Multimodal cardiac imaging obtained 33 years after a subclavian flap repair of congenital coarctation of the aorta.

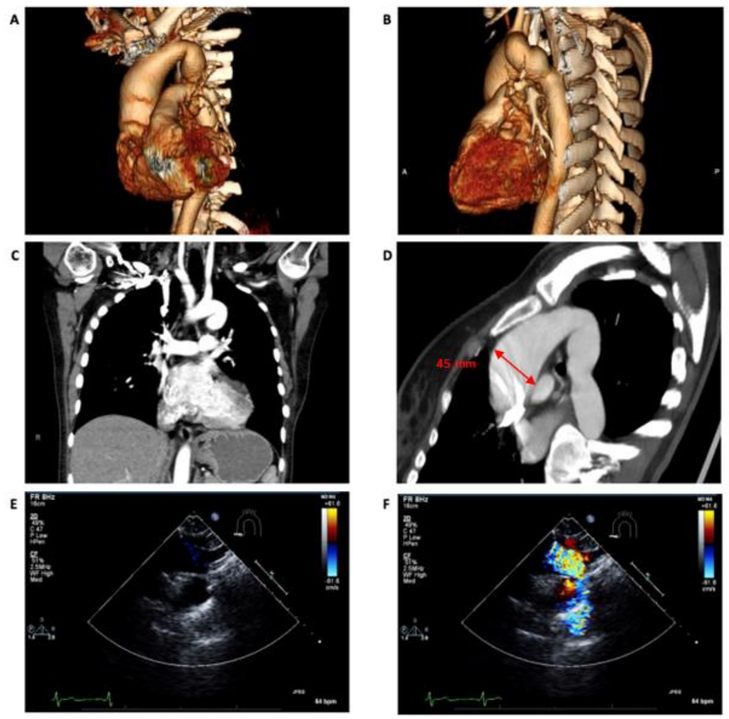

Figure 1A: Three-dimensional reconstruction of computed tomography angiography which showed a kink at the level of the ligamentum arteriosum, and surgical absence of the left subclavian artery.
Figure 1B: Three-dimensional reconstruction of computed tomography angiography showing lobulated contour of aortic arch. A proximal narrowing at the level of subclavian flap repair and a distal narrowing in the proximal descending thoracic aorta. Notably, there is absence of external structures causing pseudo-narrowing. This is supportive of intrinsic aortic wall disease. In CoA, the arterial medial layer has fragmented elastic fibers, high amounts of glycosaminoglycans, and few smooth muscle cells which degenerate over time. In surgical repairs, portions of abnormal media may be inadvertently left behind leading to aneurysmal dilatation or fibrous deposition resulting in aortic arch kinking or re-coarctation.
Figure 1C: Coronal view of computed tomography angiography with kinking at level of ligamentum arteriosum.
Figure 1D: Oblique view of computed tomography angiography demonstrating lobulated contour of aortic arch and proximal descending aorta.
Figure 1E: Suprasternal notch view of 2-D echocardiogram at the end of systole demonstrating the presence of a narrowing of the proximal descending aorta.
Figure 1F: Suprasternal notch view of 2-D echocardiogram at the end of diastole demonstrating turbulent flow proximal and distal to narrowing of the proximal descending aorta.

CONCLUSION AND DISCUSSION

Ongoing medical advances have improved the life expectancy of congenital coarctation of the aorta patients. Thus, the necessity to identify the long-term complications of surgical repairs and to recognize what increased risks these patients have--has become paramount for successful patient care. In CoA patients, early use of 3-D imaging can identify anatomical variations and guide the interpretation of traditional non-invasive imaging studies. CoA patients have an increased risk for aortic valve and arch pathologies, hence life-long monitoring and screening is key.

DISCLOSURES

The authors of this case report have no disclosures.

CITATIONS

Padalino MA, Frigo AC, Comisso M, et al. Early and late outcomes after surgical repair of congenital supravalvular aortic stenosis: A European congenital heart surgeons association multicentric study†. *European Journal of Cardio-Thoracic Surgery*. 2017;52(4):789-797. doi:10.1093/ejcts/ezx245

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