Interrupted Aortic Arch in the Adult

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Introduction:

- Interrupted aortic arch is a rare phenomena, occurring in approximately 3 of 1,000,000 cases. It was first recognized in 1778.¹
- Survival to adulthood is an even more rare occurrence with 30 cases reported.³
- Three types have been described which describe the relation of the interruption to the great vessels,¹³
  - Type A – distal to left subclavian artery (43%)
  - Type B – between left carotid and subclavian arteries (53%)
  - Type C – between the brachiocephalic and left carotid arteries (4%)
- Of note, other studies have reported Type A to be the most common

Case Presentation:

We describe a 53 year old male who presented for cardiac catheterization for pre-operative planning for severe aortic insufficiency (presumably from a history of rheumatic disease). He originally presented for several months of worsening dyspnea with exertion.

Physical Examination: Physical exam revealed >30 mmHg difference in systolic pressure between the upper extremities and lower extremities bilaterally without significantly different pressures in the left and right arms. (Figure 1)

Chest X-Ray: Demonstrated cardiomegaly. (Figure 2). Echocardiography revealed a tricuspid aortic valve with moderate to severe regurgitation and severe dilation of the ascending aorta.

Aortography: Aortography via a femoral approach revealed an interrupted arch distal to the left subclavian artery with large subcostal arteries serving as “bridging” collateral vessels. (Figure 3)

CT Angiography: CT Angiography demonstrated significant enlargement of the intercostal arteries as well as collateralization from the internal mammary arteries to the inferior epigastric arteries. A fibrous strand connected the ascending and descending aortas distal to the left subclavian artery. (Figures 4 & 5)

Discussion:

- Surgical intervention is high-risk in this patient due to the difficulty in achieving hemostasis due to massively enlarged intercostal arteries.
- However, he subsequently underwent unremarkable aortic valve replacement with plans for future aortic arch replacement.
- Previous surgical approaches have included 1 or 2 stage repairs¹, though two stage repair has been advocated in one study with repair of the aortic valve.²
- Consideration should also be given to extra-anatomic bypass³ of which ventral aortic repair has been shown to have good outcomes.⁴
- Interrupted aortic arch is a rare condition, however association with aortic arch dilation and aortic insufficiency seems to be even less frequent.

References: