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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.^{1,3}
 - 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 studies³ 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)

References:

- 1 Messner et. al. Interrupted Aortic Arch in the Adult. Texas Heart Institute J. 2002; 29(2): 118-121.
- 2 Ogino et. al. Two stage repair for aortic regurgitation with interrupted aortic arch. Ann Thor Surg 1998 Apr; 65(4): 1151-3.
- 3 Borgohain et. al. Isolated Interrupted Aortic Arch in an 18-Year Old Man. Texas Heart Institute J. 2013; 40(1): 79-81.
- 4 Sai Krishna et. al. Interruption of Aortic Arch in Adults. Texas Heart Institute J. 2005; 32(2): 147-150.



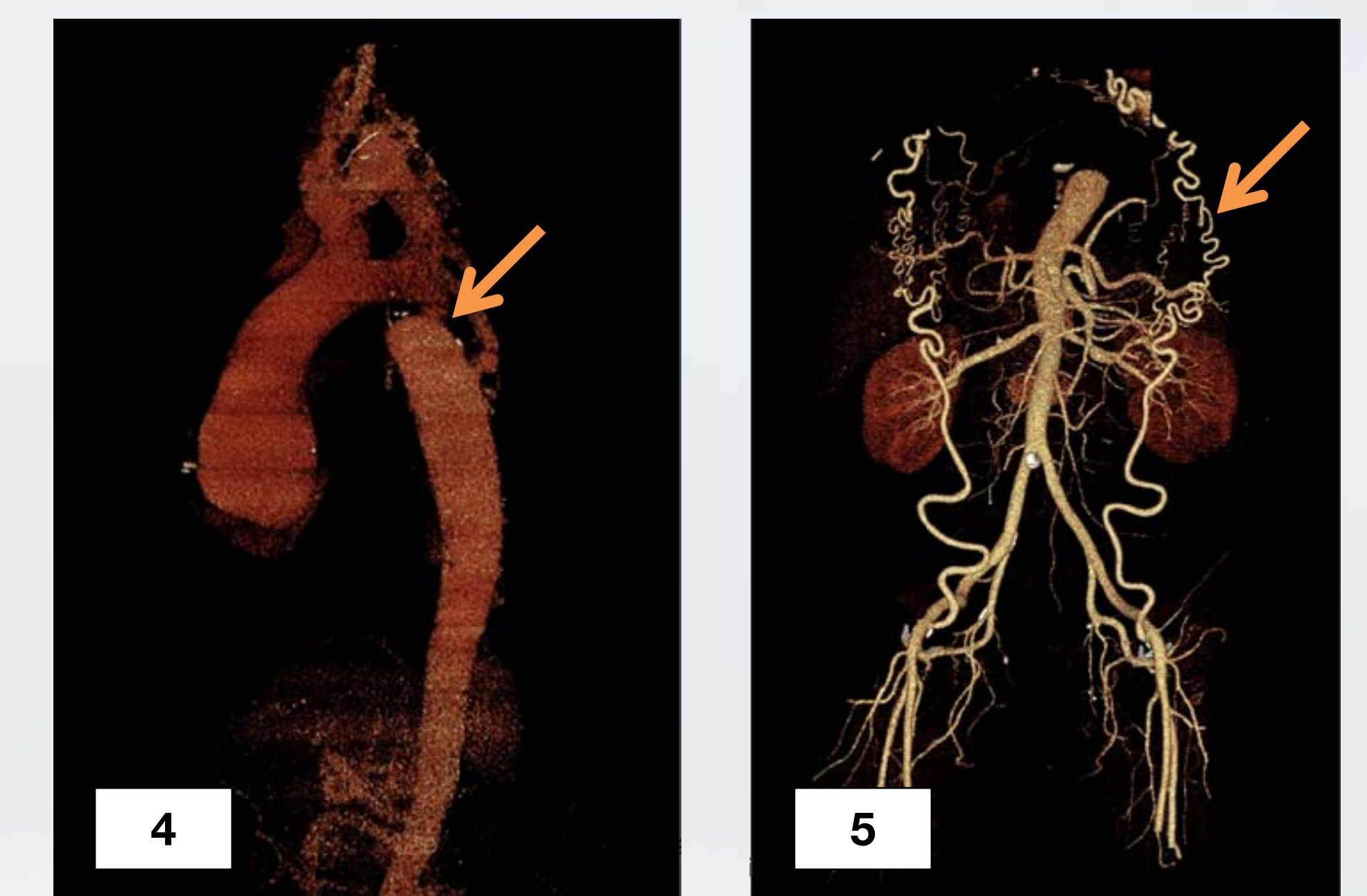
Figure 1: Schematic of Blood Pressure and Pulse Recordings.

Figure 2: Chest x-ray demonstrating cardiomegaly.

Figure 3: Aortic angiogram demonstrating interruption and collateral vessels (arrows).

Figure 4: Interruption in the aortic arch (arrow) is noted distal to the left subclavian artery.

Figure 5: Significant collateralization is noted from the internal mammary to the inferior epigastric arteries (arrow). Enlarged intercostal arteries were also noted.



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.

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