

**Case report**

**INCIDENTAL FINDING OF INTERRUPTED AORTIC ARCH IN AN ADULT PATIENT UNDERGOING URGENT PERCUTANEOUS CORONARY INTERVENTION**

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**ABSTRACT**

Interrupted Aortic Arch (IAA) is a rare congenital abnormality characterized by a complete discontinuity of the aortic lumen, usually located after the origin of the left subclavian artery. IAA is mainly diagnosed during childhood and has an extremely high mortality rate if left untreated. Therefore, only a few cases have been diagnosed in adulthood. We report the case of a patient with Non-ST Segment Elevation Myocardial Infarction (NSTEMI) and unknown IAA abnormality, who underwent urgent percutaneous coronary angioplasty (PCI). It was not possible to reach ascending aorta from the right radial artery because of the presence of tangled arteries connecting the pre-vertebral subclavian segment to the descending aorta. PCI was completed successfully through the left radial artery. A post-procedural Angio-CT scan confirmed the Aortic Arch interruption. The presented case highlights the crucial role of a multi-imaging modality approach for those patients with such congenital abnormalities before undergoing PCI.

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**1. Background**

Interrupted Aortic Arch (IAA) is an extremely rare congenital abnormality characterized by a complete discontinuity of the aortic lumen between ascending and descending aorta (1). This condition affects 3 per million live births; if left untreated, it is associated with a mortality rate of more than 90% at 1 year (2). IAA is classified into three types: Type A, where the interruption of the aortic lumen takes place distal to the origin of the left subclavian artery; Type B, characterized by interruption between the left carotid and left subclavian arteries; and Type C, involving the interruption between the carotid arteries (3). Although this condition is usually considered a pediatric disorder, more than 30 cases have been reported in the adult population (4). The majority of patients reported hypertension unresponsive to medical therapy (70%). Among symptoms reported at the time of diagnosis, claudicatio intermittens, congestive heart failure, and aortic insufficiency have been observed in 13%, 6%, and 10% of the cases, respectively (4).

Occasionally, exertional angina has been reported as one of the initial symptoms associated with IAA (5–8). Performing percutaneous coronary intervention (PCI) in patients with this particular condition poses notable challenges, especially in emergency care scenarios and when the underlying abnormality is unknown. To our best knowledge, only one case report of IAA in the context of Acute Coronary Syndrome has been reported. (9)

**2. Case Report**

A 58-year-old woman, with a history of hyperlipidemia and hypertension since adolescence, was referred to the emergency department, complaining of constrictive chest pain. The electrocardiogram showed ST segment depression and inverted T waves in the infero-lateral leads and high sensitivity cardiac troponin T level was high. The patient continued to complain of chest pain despite medical therapy with aspirin, low molecular weight heparin, and intravenous nitrates and for this reason, she underwent urgent coronary angiography.

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It was not possible to reach ascending aorta via the right radial artery, because of the presence of multiple tangled arteries connecting the pre-vertebral subclavian segment to the descending aorta. A digital subtraction angiography (DSA) of the aorta, performed through left radial access, showed the interruption of the aortic arch, distal to the origin of the left subclavian artery (Fig 1. A-B). The coronary angiography showed a tight stenosis of the right coronary artery, and right coronary artery PCI was performed successfully through left radial access. (Fig 1. C-D). The computed tomography angiography (CTA) confirmed the Aortic Arch interruption and showed the tangled collateral vessels connecting both subclavian arteries to the descending aorta (Fig 1. E-G-H). The cardiac surgery consultation judged conservative management of the aortic arch congenital abnormality to be appropriate. The patient was discharged five days later without further complications.



**Figure 1 (A-H).** A-B: Digital Subtraction Angiography. A. Tangle vessel connection from Right Subclavian Artery to the Descending Aorta. B. Short occlusion of the Descending Aorta distal to the left subclavian artery origin. C: Coronary Angiography showed Proximal right coronary artery critical stenosis (White Arrow). D: Final Result after successful deployment of Coroflex Isar Neo 3.0/24 mm new generation DES (B. Braun, Germany). E: Computed Tomography Angiography (CTA) MPR reconstruction. See the aortic occlusion after the left subclavian artery origin. F: Computed Tomography Angiography (CTA) MPR reconstruction showing left ventricular hypertrophy. G-H: Angio CT Volume-rendered images. G: Lateral view projection showing the interruption of the Aortic Arch, distally to the left Subclavian Artery. H: antero-posterior projection showing the tangled vessel connections, running very close to the back spine, from both Subclavian Arteries to the Descending Aorta.

This condition avoids the onset of severe symptoms (4,10) Second, it is suggested that a subset of Type A interruptions may represent the final stage of a progressive aortic coarctation narrowing. (4)

In this latter condition, when an acute myocardial infarction occurs, the presence of an unknown Interrupted Aortic Arch represents a challenge for the following urgent percutaneous coronary revascularization. Finally, the presented case highlights the essential role of a multi-imaging modality approach in managing patients with such anatomical complexity.

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## 3. Discussion

Interrupted Aortic Arch is a rare congenital malformation, accounting for less than 1% of all reported cases of congenital heart disease. (3) Despite being highly uncommon, IAA should be suspected next to aortic coarctation, in those adult patients with hypertension unresponsive to medical management and weak arterial pulses in the lower extremities. (4) Among adults, Type A is observed with significantly higher frequency, accounting for 79% of cases. The higher incidence of Type A IAA in adults can be attributed to main two factors. First, Type A interruptions are more likely associated with well-developed collateral vessels, that can guarantee a sufficient blood distal to the interruption.