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Case Report

# A Rare Case of Survival to Adulthood with an Interrupted Aortic Arch

**Satya Rashmit Durugu[1]* and Chaitanya Rojulpote[2]**

[1]*Intern, Department of Medicine, Kasturba Medical College, Manipal, India*

[2]*Research Fellow, Department of Diagnostic Radiology, University of Pennsylvania, Philadelphia, USA*

***Corresponding Author:** Satya Rashmit Durugu, Intern, Department of Medicine, Kasturba Medical College, Manipal, India.

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## Abstract

Transthoracic echocardiography has been considered the gold standard for cardiac evaluation of young patients with hypertension. This however, may not reveal conditions such as interrupted aortic arch which can be found in young adults. This article seeks to establish the need for CT angiogram of the heart to establish the diagnosis. Further numerous cases have been analyzed to establish single stage Dacron graft repair as the most successful method to treat such patients.

***Keywords:*** *Interrupted Aortic Arch; CT Angiogram*

## Introduction

Hypertension is an increasingly important medical and public health issue with a prevalence of 30 - 45% of the general population. Among the hypertensive population, there exists a small subgroup called the young adult hypertensive. This population is at a significantly higher risk of increased morbidity and mortality and therefore must be timely diagnosed and appropriately managed. A thorough physical examination in these young hypertensive patients is crucial in the diagnosis by providing clues for rare secondary causes such as interrupted aortic arch and coarctation of aorta. Interrupted aortic arch presents at an infantile age with a poor prognosis without surgical intervention. That said, some patients, as evidenced in this case, may survive beyond the expected mean age. Therefore, these conditions must always be kept in mind as a routine transthoracic echocardiography may not detect them. In regard to such a presentation, conventional angiography and other imaging modalities play an important role in the background of clinically evident physical signs. In this case, we focus on the importance of clinical signs, various imaging modalities, and conventional angiography for diagnosing a rare but potentially treatable causes of hypertension in a young population.

## Case Presentation

In January 2015, a 33-year-old female patient presented to the cardiology OPD with a history of palpitations over the past 2 months. She had previously been detected to have a high blood pressure reading at another hospital and was started on a combination of beta blocker- metoprolol 50mg and a calcium channel blocker- amlodipine 5mg OD. There was no significant medical history, family history or a history of drug abuse. Her obstetric history showed two uncomplicated pregnancies which she tolerated well.

On physical examination, the patient was alert and oriented. No cyanosis or clubbing was present. Pulse was 86 beats per minute, regular rate with normal rhythm and volume. Blood pressure was 170/9 6mmhg in both upper limbs. However, when BP was measured in the lower limbs there was approximately a 40-mmHg difference from the value obtained in the upper limb. Examination of the cardiovascular system revealed no significant findings.

## Investigations

Routine laboratory investigations were normal. Ultrasound of the abdomen did not reveal any abnormality. Electrocardiogram was found to be normal.

Renal artery doppler revealed increased acceleration time values and slow rising wave forms on both sides, suggestive of possible bilateral renal artery stenosis. However, a renal angiography done later showed no stenosis in either of the renal arteries.

Cardiac catheterization was tried via left radial artery. Aortography showed a complete occlusion of aortic arch distal to the origin of left subclavian artery. To delineate the anatomy of the abnormality, an aortography via the right femoral artery was performed. The peak systolic pressure gradient above the interrupted segment was 170 mmHg and the pressure below was 130 mmHg.

An Echocardiography did not reveal any abnormality.

This was followed up with a multi-slice computed tomography which revealed a 7 mm aortic interruption distal to the left subclavian artery with multiple collaterals arising from the internal mammary artery, superior epigastric artery, inferior epigastric artery and descending scapular arteries. Furthermore, multiple intercostal collaterals were seen arising from 3$^{rd}$ to 10$^{th}$ intercostal spaces from the vertebrobasilar system, extending to the posterior chest wall and spine.

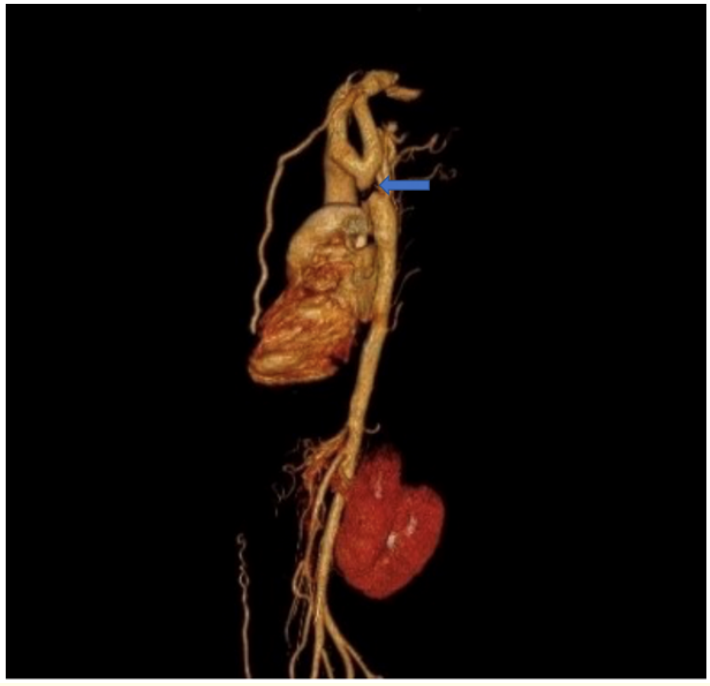

***Figure 1:*** *The interruption in the aortic arch is shown by the blue arrow.*

## Diagnosis

On the basis of the clinical findings and the CT study result, the patient was diagnosed with an interrupted aortic arch type A, with the break in continuity seen after the left subclavian artery.

### Treatment

Patient in view of IAA type A was advised a single stage repair by extra anatomical approach. However, the patient refused the surgical correction and was therefore put on a medical management primarily aimed at treating the high blood pressure. She was put on tablet Metoprolol 50 mg OD and tablet Amlodipine 5 mg BD. On follow up, her blood pressure had come down to 144/92 mmhg when measured as a 24-hour ambulatory blood pressure.

## Discussion

Interrupted aortic arch is a rare congenital malformation of the heart that occurs at a frequency of 3 cases in a million live births and accounts for 1% of all congenital heart diseases [1]. This anomaly is defined as a loss of luminal continuity between the ascending and descending portions of the aorta [2]. A classification system introduced by Celoria and Patton [3] in 1959 divides IAA into three types: type A-interruption distal to the left subclavian artery (LSA); type B- interruption between the left common carotid artery and the LSA; and type C- interruption between the brachiocephalic trunk and the left common carotid arteries. In infants, type B is more common (53%) and is associated with DiGeorge syndrome and chromosome 22q11.2 deletion [4], whereas in adults type A has been reported more (79%) [5] as this type has better chances of collateral formation as seen in this patient.

As compared to neonates and infants, the diagnosis of interrupted aortic arch is very rare in adults and only a limited number of cases have been reported in literature so far worldwide.

Adult Patients with Surgical Repair of Isolated Interrupted Aortic Arch.

| Patient | Age/Sex | Type of Lesion | Treatment | Reference |
|---|---|---|---|---|
| 1 | 42/F | B | Single stage extra anatomic repair with Dacron graft | Messner G., *et al*. 2002 |
| 2 | 18/M | A | Single stage extra anatomic repair with Dacron graft | Shyamadeep B., *et al.* 2010 |
| 3 | 19/M | B | Single stage extra anatomic repair with Dacron graft | Morgan., *et al.* 1970 |
| 4 | 19/M | A | Single stage extra anatomic repair with Dacron graft | Todoric., *et al.* 1985 |
| 5 | 24/M | A | Single stage extra anatomic repair with Dacron graft | Burton BJ., *et al*. 1995 |
| 6 | 32/M | A | Single stage extra anatomic repair with Dacron graft | Ogino Hj., *et al*. 1995 |
| 7 | 36/M | B | Single stage extra anatomic repair with Dacron graft | Kauff MK., *et al*. 1973 |
| 8 | 49/M | A | Single stage extra anatomic repair with Dacron graft | Kauff MK., *et al*. 1973 |
| 9 | 72/F | A | Single stage extra anatomic repair with Dacron graft | Canova CR., *et al.* 1995 |

In all these cases, it has been noticed that extensive collateral network formation to maintain distal blood flow plays a crucial role in survival to adulthood.

Multiple theories have been proposed which attempt to explain how a patient can survive the initial years of infancy when the collateral circulation has yet to develop. The most plausible explanation states that in cases of survival to adulthood, the initial defect may have been a coarctation of aorta [6], which later progresses to a complete obstruction of the vessel lumen and forms an interruption. This is further substantiated by the fact that IAA type A and coarctation of aorta show clinical and anatomical similarities.

A careful physical examination is of utmost importance in dealing with a young patient with hypertension. A radio-femoral delay and a discrepancy in upper and lower limb blood pressure may be present in both coarctation of aorta and an IAA. However, in coarctation of aorta, there is an audible systolic murmur during blood flow across the constriction which is absent in an IAA.

As seen in this case, it is important to realize that a routine transthoracic echocardiography may miss a significant IAA or its close differential, a coarctation of aorta. Therefore, when clinical signs show evidence of the condition, it should be confirmed by a computed tomography or an MR angiography.

The definitive treatment of IAA is surgical correction, either by single-stage repair (extra- anatomic approach) or a two-stage repair (in the presence of other heart abnormalities). In regards to our case, the patient was unwilling to undergo a surgery. Therefore, she was managed conservatively with the aim of controlling blood pressure and preventing heart failure.

## Conclusion

IAA is seldom seen in patients beyond childhood. However, some individuals may present with refractory hypertension and manage to reach adulthood with an IAA. To note, a thorough clinical examination plays a pivotal role in detecting IAA. When the clinical signs are suggestive of IAA, CT or a MR angiography is recommended as a transthoracic echocardiography may not detect the lesion and the primary diagnosis may be missed. A single stage repair with a Dacron graft method is the gold standard to repair an IAA that has survived to adulthood and has consistently been shown to be curative for both type A and B lesions.

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