



INTISARI SAINS MEDIS

Published by Intisari Sains Medis

Case report: interrupted aortic arch



CrossMark

Petrus Sewe Pajo^{1*}, Dewa Gde Mahiswara Suadiatmika²,
Elysanti Dwi Martadiani²

¹Radiology Resident Faculty of Medicine, Universitas Udayana, Prof. dr. I.G.N.G. Ngoerah General Hospital, Denpasar, Bali, Indonesia;

²Lecturer and Radiology Consultant, Department of Radiology, Faculty of Medicine, Universitas Udayana, Prof. dr. I.G.N.G. Ngoerah General Hospital, Denpasar, Bali, Indonesia.

*Corresponding author:

Petrus Sewe Pajo;
Radiology Resident Faculty of Medicine, Universitas Udayana, Prof. dr. I.G.N.G. Ngoerah General Hospital, Denpasar, Bali, Indonesia;
pajointeristi32@gmail.com

Received: 2023-10-05

Accepted: 2023-11-06

Published: 2023-12-28

ABSTRACT

Introduction: Interrupted aortic arch (IAA) is one of the congenital heart diseases characterized by a complete anatomical and luminal disruption between the ascending and descending aorta. It is frequently accompanied by other congenital heart defects, such as PDA and VSD. IAA in adults is exceptionally rare. The presence of collateral circulation and PDA are key factors for patient survival. Thorax CTA is an excellent modality for imaging and diagnosis, ensuring that appropriate treatment can be carried out.

Case Presentation: An 18-year-old male patient presented with complaints of worsening shortness of breath 10 days prior to the examination. The shortness of breath has been experienced since school age, often occurring during exercise and strenuous activity. Thorax CTA results indicated IAA type A subtype 1 according to

Celoria and Patton classification, PDA type A according to Krichenko classification, perimembranous VSD, cardiomegaly, and pulmonary hypertension.

Conclusion: IAA in adults is exceptionally rare. Adult IAA patients may exhibit no symptoms or may present with symptoms of CHF. Thorax CTA is one of the modalities employed to confirm the diagnosis of IAA. It can visualize the interruption and separation between the ascending and descending aorta, as well as the associated anomalies. The presence of a PDA and VSD ensures that the descending thoracic aorta distal to the defect continues to receive some oxygenated blood from the heart. Thorax CTA plays a pivotal role in establishing cases of IAA, as it helps clinicians to make a quick and accurate diagnosis.

Keywords: *Interrupted aortic arch, PDA, Thorax CTA, VSD.*

Cite This Article: Pajo, P.S., Suadiatmika, D.G.M., Martadiani, E.D. 2023. Case report: interrupted aortic arch. *Intisari Sains Medis* 14(3): 1240-1244. DOI: [10.15562/ism.v14i3.1888](https://doi.org/10.15562/ism.v14i3.1888)

INTRODUCTION

Interrupted aortic arch (IAA) is an exceptionally rare congenital heart disease, with approximately 1.5% incidence rate of all congenital heart diseases, which involves a complete anatomical and luminal disruption between the ascending and descending aorta.¹⁻³ The malformation of aortic arch may present without definite symptoms or may be accompanied by a variety of symptoms, including difficulty breathing or swallowing. Advances in imaging modalities and the increased use of cross-sectional imaging, including multidetector computed tomography (CT) and magnetic resonance imaging (MRI), have resulted in more incidental discoveries of such malformations.⁴

The incidence of IAA is estimated to be around 2 cases per 100,000 live births. Almost all patients with IAA present within the first 2 weeks of life when the

ductus arteriosus closes. In neonates, 53% of cases are type B, followed by type A (43%) and type C (4%). IAA in adults is very rare and is often overlooked in patients with secondary hypertension.^{3,5} Based on a report by Gordon EA et al., only 37 cases of adults with IAA have been reported over the past 40 years, with most of the cases being type A, and only 6 of them (16%) being type B.⁶

IAA patients have a very low survival rate, and the main treatment is early surgical intervention after diagnosis. Therefore, a rapid and precise diagnosis is crucial to prevent severe complications and potential fatalities. For diagnosis, in addition to anamnesis of patient complaints and physical examination results, it is required to combined with various examination modalities such as echocardiography, thorax X-ray, Thorax CT Angiography and Thorax MRI Angiography, where Thorax CTA being

considered the best modality for imaging and diagnosis.

CASE REPORT

An 18-year-old male patient presented with complaints of worsened shortness of breath 10 days prior to the examination, accompanied by complaints of chest palpitations without chest pain. This patient has experienced shortness of breath since school age, particularly during exercise and strenuous activity. Shortness of breath also often occurs when the patient sleeps on his back and improves when sitting or standing. The patient has a history of heart disease since the age of 3 and a history of hypertension.

Upon physical examination, the results were as follows: BP: 170/100 mmHg, pulse: 91 bpm, temperature: 36°C, RR: 20 breaths/min, SpO₂ 85%. Physical examination of the heart revealed a murmur (+), systolic



Figure 1. Cardiomegaly with left atrial enlargement and left ventricular hypertrophy configuration with suspicion of pulmonary hypertension

continuous at ICS IV PSL sinistra and ICS V anterior axillary line sinistra rumbling gr III/IV, with cyanosis on the superior and inferior extremities.

From thorax x-ray examination, the results were cardiomegaly with left atrial enlargement and left ventricular hypertrophy configuration with suspicion of pulmonary hypertension (Figure 1).

From Thorax CT Angiography examination, the results were Interrupted Aortic Arch type A subtype 1 (normal subclavian artery) according to Celoria and Patton classification accompanied by patent ductus arteriosus type A according to Krichenko classification and perimembranous ventricular septal defect, measured ascending aorta diameter +/- 2.14 cm and descending aorta pars thoracalis measured +/- 3.24 cm, cardiomegaly with pulmonary hypertension (Figure 2).

Considering the high risk of surgery, conservative therapy was chosen to treat the patient's complaints, with routine medications such as ramipril 5mg every 12 hours, adalat oros 60mg every 24 hours, bisoprolol 10mg every 24 hours, sildenafil 20mg every 8 hours, furosemide 40mg every 24 hours, spironolactone 25mg every 24 hours.

DISCUSSION

Interrupted aortic arch (IAA) is a type of congenital heart disease characterized

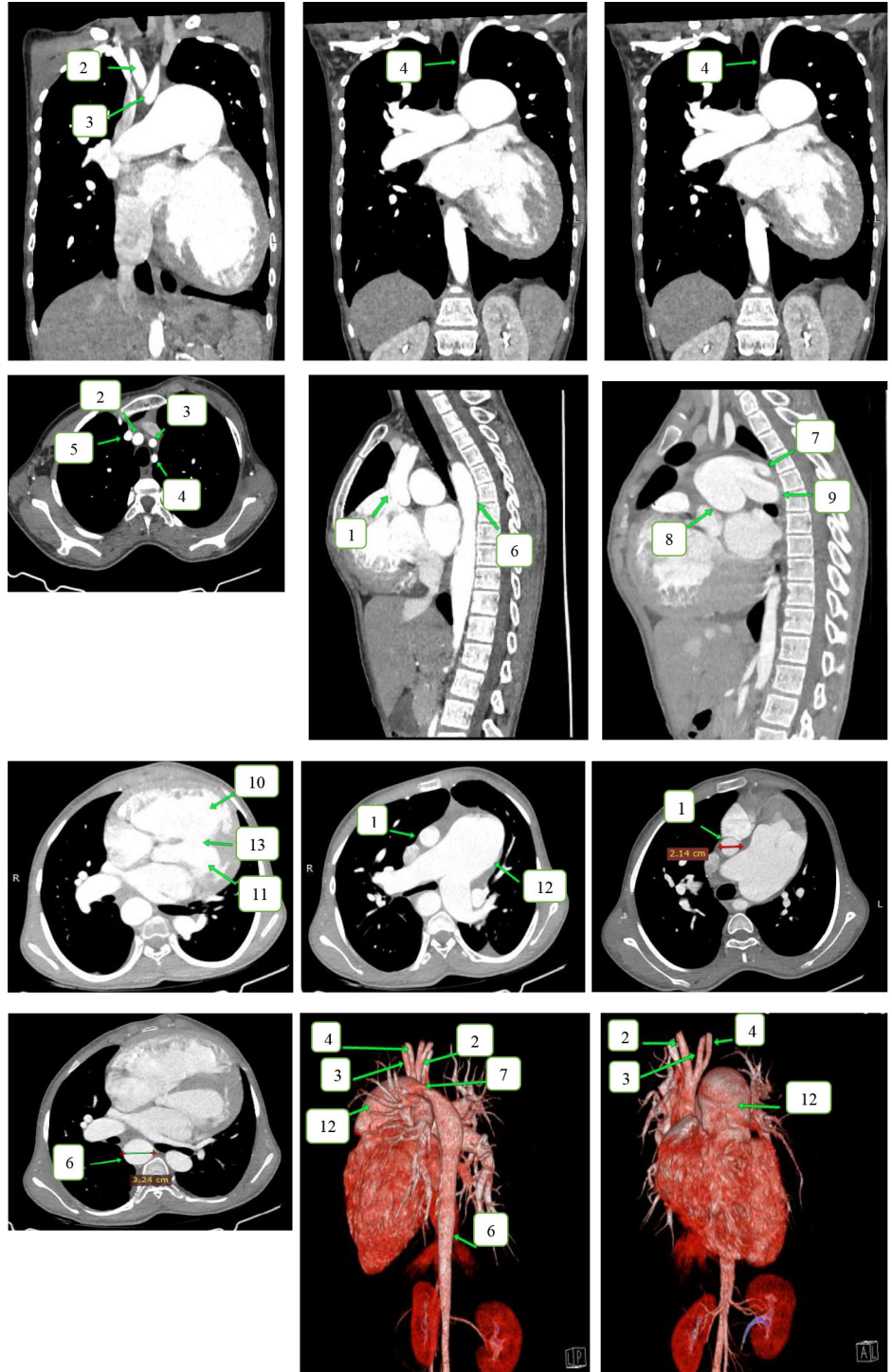


Figure 2. Interrupted Aortic Arch tipe A subtype 1 (normal subclavian artery), patent ductus arteriosus tipe A, perimembranous ventricle septal defect with features of pulmonary hypertension. Legend: 1: Ascending Aorta; 2: Brachiocephalic Trunk; 3: Left Carotis Communis Artery; 4: Left Subclavian Artery; 5: Superior Vena Cava; 6: Descending Aorta; 7: PDA; 8: Right Pulmonary Artery; 9: Left Pulmonary Artery; 10: Right Ventrikel; 11: Left Ventrikel; 12: Pulmonary Trunk; 13: VSD.

by complete anatomical and luminal disruption between the ascending and descending aorta, and it is considered the

most severe form of aortic coarctation. According to the literature, IAA is very rare with an incidence of approximately

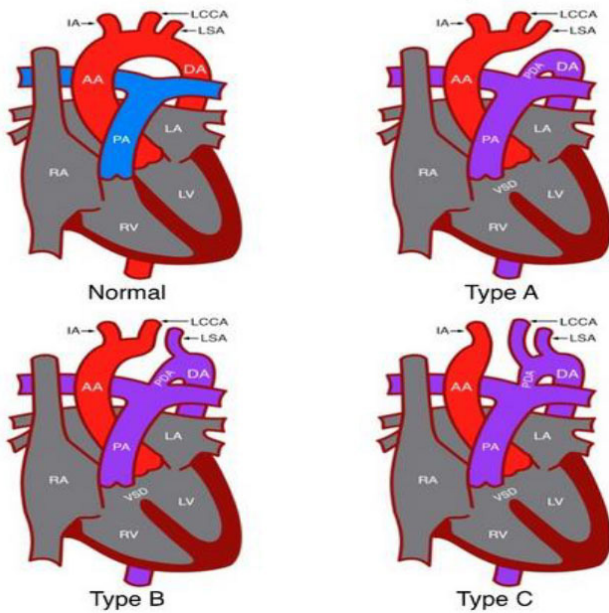


Figure 3. Celoria-Patton classification² (AA - ascending aorta; DA - descending aorta; PDA - patent ductus arteriosus; VSD - ventricular septal defect; PA - pulmonary artery; RA - right atrium; LA - left atrium; RV - right ventricle; LV - left ventricle).

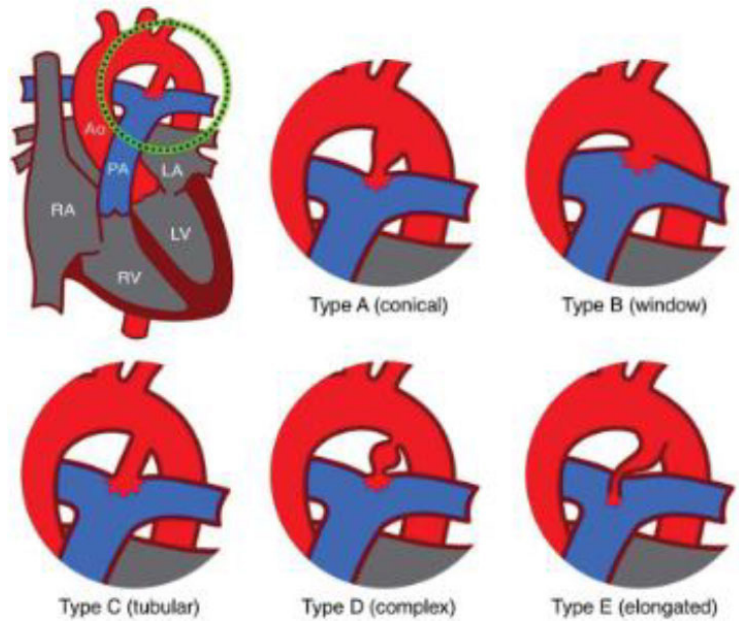


Figure 4. Variations in patent ductus arteriosus (PDA) configuration illustrated with the classification of Krichenko et al.¹⁰

1.5% among all congenital heart disease patients.¹⁻³

Infant patients with IAA may be asymptomatic until the ductus arteriosus closes resulting in signs of heart failure, such as tachycardia, tachypnea, and impaired growth, feeding difficulties, respiratory distress, cyanosis, and anuria which may eventually lead to shock and death.³ During fetal circulation, ductus arteriosus provides blood supply to the distal extremities of the fetus while the upper body of the fetus receives blood supply from the left ventricle to the aorta. After birth, pulmonary vascular resistance decreases leading to the closure of the ductus arteriosus. This leads to the inability of the heart to provide blood to the distal parts of the body. On physical examination of infants with IAA, there may be an absent pulse with a blood pressure difference between the right arm and inferior extremity. The incidence of IAA in adults is very rare. Although 90% of infants with IAA die of circulatory failure within the first year of life, there are some reports documenting survival of their patients to adulthood, which is very rare but possible. Patients may be asymptomatic or may present with complaints of headache, hypertension, claudication, differences in

upper and lower limb blood pressure, limb swelling, congestive heart failure, or aortic dissection.^{3,7,8}

A variety of examination modalities have been introduced to establish the diagnosis of IAA. While Chest X-rays are routinely performed, they do not provide specific images, lacking visibility of the aortic knob and cardiomegaly.² In contrast, a Thorax CTA examination of adult IAA patients may reveal interruption and separation of the ascending aorta and descending aorta, abnormal ratio of inner diameter of the ascending aorta to the main pulmonary artery, widening of the ascending aorta, and narrowing of the descending aorta. Thorax CTA allows for the visualization of the interrupted aortic arch and associated anomalies.⁸ According to the Celoria-Patton classification, IAA can be classified into three types according to the location of the anomaly (Figure 3).¹⁻⁴ Type A: The disruption is located in the distal left subclavian artery, which is the second most common disruption representing approximately 13% of cases. Type B: The disruption is located between the left carotid artery and the left subclavian artery, which is the most frequent disorder, representing approximately 84% of cases.

Type C: Disruption located between the innominate artery and the left carotid artery, is the rarest type, representing about 3% of all cases.

These three types of IAA can be further classified according to the origin of the subclavian artery.^{2,3} Type 1: Normal subclavian artery origin. Type 2: Aberrant right subclavian artery, found in the distal part of the left subclavian artery. Type 3: Isolated right subclavian artery, found to originate from the right patent ductus arteriosus.

IAA is often accompanied by cardiovascular anatomic defects in approximately 98% of cases. The most common cardiovascular defect is patent ductus arteriosus (PDA), which occurs in approximately 97% of patients with IAA. This vascular structure is required to provide blood supply to the descending thoracic aorta distal from the defect. Ventricular septal defects (VSD) are also commonly present, occurring in approximately 90% of individuals with IAA. VSD occurs as a result of posterior malalignment of the conal septum. The degree of collateral circulation and PDA are the key factors for survival into adulthood.^{3,8,9}

In fetal life, the arterial duct connects the descending thoracic aorta with the proximal left pulmonary artery. After birth the arterial duct usually closes functionally within 24-48 hours and anatomically within 1 month. Persistence of arterial duct for more than 3 months after birth is one of the congenital heart defects known as patent ductus arteriosus (PDA).¹⁰ According to Krichenko classification, PDA can be divided into five types (Figure 4).¹¹ Type A: conical ductus with prominent aortic ampulla and constriction near the pulmonary artery end. Type B: window-like (wide and very short) ductus. Type C: tubular ductus without constrictions. Type D: complex ductus with multiple constrictions Type E: elongated ductus with remote constriction

The results of the patient's Thorax CTA in this case are Interrupted Aortic Arch type A subtype 1 (normal subclavian artery) accompanied by patent ductus arteriosus type A, perimembranous ventricle septal defect with measured ascending aorta diameter +/- 2.14 cm and aorta descendens pars thoracalis measured +/- 3.24 cm accompanied by cardiomegaly and pulmonary hypertension. In addition to the Thorax CTA examination, previously the patient had also undergone a thorax X-ray examination with the results of the examination, namely cardiomegaly with left atrial enlargement and left ventricular hypertrophy configurations accompanied by suspicion of pulmonary hypertension. In this case, the patient's descending thoracic aorta originated from the pulmonary trunk without collateral circulation, leading to inadequate arterial oxygenation. However, the ventricular septal defect and PDA allowed some oxygenated blood to be pumped into the descending thoracic aorta distal from the defect, undoubtedly contributing to patient survival.⁸

The main treatment of IAA cases is early surgical intervention, typically in the form of bypass or percutaneous stent placement. In neonates and infants, there are many surgical methods, such as end-to-end or end-to-side anastomosis, where end-to-end anastomosis can be performed in older children and adults, while interposition transplantation can

be performed in the older population. A report revealed that most adult IAA patients (54%) underwent sternotomy or lateral thoracotomy for correction; 8% underwent percutaneous perforation with a stent placed between the ascending and descending aorta, and 10% were managed medically after refusing surgical treatment. Prostaglandin E1 should be started early to avoid sudden cardiac collapse and death by maintaining patency of the ductus arteriosus, thus ensuring perfusion of the lower body until surgical correction is performed.^{3,8} In this patient, conservative therapy was chosen due to the high risk of surgery, with routine medications such as ramipril 5mg every 12 hours, adalat oros 60mg every 24 hours, bisoprolol 10mg every 24 hours, sildenafil 20mg every 8 hours, furosemide 40mg every 24 hours, spironolactone 25mg every 24 hours.

CONCLUSION

Interrupted aortic arch (IAA) is one of the rarest types of congenital heart disease, with an incidence rate of approximately 1.5% among all congenital heart disease patients. The majority of patients with IAA present within the first 2 weeks of life when the ductus arteriosus closes. In contrast, the incidence of IAA in adults is exceptionally rare, with patients being asymptomatic or presenting with various complaints, ranging from mild to severe. Most of IAA patients have congenital heart defects, with PDA and VSD being the most common. The presence of PDA and VSD in these patients is a key factor for their survival to adulthood.

IAA patients have a very low survival rate, and the primary treatment is early surgical intervention after diagnosis. To establish the diagnosis of IAA, various examination modalities have been introduced, such as echocardiography, thorax X-ray, Thorax CT Angiography and Thorax MRI Angiography in combination with patient history and physical examination. Thorax CT Angiography is one of the main examinations to establish the diagnosis of IAA. This is because thorax CT Angiography allows visualization of interrupted aortic arch and related anomalies.

CONFLICT OF INTEREST

The authors report no conflict of interest in writing this case report.

FUNDING

In writing this case report, the author is responsible for funding without involving any funding source from any party

ETHICAL STATEMENT

The patient has given consent to participate and publish the data.

AUTHOR'S CONTRIBUTION

All authors have the same contribution in this case report, from the stage of case finding, reading of case radiology results, and clinical outcomes obtained, which are presented in scientific publications.

REFERENCES

1. Sato S, Akagi N, Uka M, Kato K, Okumura Y, Kanazawa S. Interruption of the aortic arch: diagnosis with multidetector computed tomography. *Jpn J Radiol.* 2011;29(1):46-50. Available from: <http://dx.doi.org/10.1007/s11604-010-0517-y>
2. Baba Y, Weerakkody Y. Interrupted aortic arch. In: *Radiopaedia.org.* Radiopaedia.org; 2009. Available from: <http://dx.doi.org/10.53347/rid-7805>
3. Ramirez Alcantara J, Mendez MD. Interrupted Aortic Arch Continuing Education Activity [Internet]. 2023. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK532902/>
4. Bae S Bin, Kang E-J, Choo KS, Lee J, Kim SH, Lim KJ, et al. Aortic Arch Variants and Anomalies: Embryology, Imaging Findings, and Clinical Considerations. *J Cardiovasc Imaging.* 2022;30(4):231-62. Available from: <https://pubmed.ncbi.nlm.nih.gov/36280266>
5. Chen G-W, Li H, Pu H. Isolated interrupted of aortic arch diagnosed using CT angiography: A case report and literature review. *Medicine.* 2018;97(21):e10569-e10569. Available from: <https://pubmed.ncbi.nlm.nih.gov/29794733>
6. Gordon EA, Person T, Kavarana M, Ikonomidis JS. Interrupted Aortic Arch in the Adult. *J Card Surg.* 2011;26(4):405-9. Available from: <http://dx.doi.org/10.1111/j.1540-8191.2011.01273.x>
7. Shirani S, Soleymanzadeh M. Diagnosis of aortic interruption by CT angiography. *Pol J Radiol.* 2013;78(1):72-4. Available from: <https://pubmed.ncbi.nlm.nih.gov/23493741>
8. Dong S-W, Di D-D, Cheng G-X. Isolated interrupted aortic arch in an adult: A case report. *World J Clin Cases.* 2021;9(4):992-8. Available from: <https://pubmed.ncbi.nlm.nih.gov/33585649>

9. Dillman JR, Yarram SG, D'Amico AR, Hernandez RJ. Interrupted Aortic Arch: Spectrum of MRI Findings. *American Journal of Roentgenology*. 2008;190(6):1467–74. Available from: <http://dx.doi.org/10.2214/ajr.07.3408>
10. Krupiński M, Irzyk M, Moczulski Z, Banyś R, Kuniewicz M, Urbańczyk-Zawadzka M. Detailed radiological study of the patent ductus arteriosus: a computed tomography study in the Polish population. *Folia Morphol (Warsz)*. 2020;79(3):462–8. Available from: <http://dx.doi.org/10.5603/fm.a2019.0116>
11. Krichenko A, Benson LN, Burrows P, Mões CAF, McLaughlin P, Freedom RM. Angiographic classification of the isolated, persistently patent ductus arteriosus and implications for percutaneous catheter occlusion. *Am J Cardiol*. 1989;63(12):877–80. Available from: [http://dx.doi.org/10.1016/0002-9149\(89\)90064-7](http://dx.doi.org/10.1016/0002-9149(89)90064-7)



This work is licensed under a Creative Commons Attribution