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Case Reports

Computed tomography angiographic diagnosis of interrupted aortic arch in a neonate - a case report

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Abstract
Interrupted Aortic Arch (IAA) is a very rare congenital malformation and it is usually associated with other cardiovascular abnormalities. It is a cause of early neonatal morbidity and mortality if not diagnosed early and accurately. Computed tomographic angiography (CTA) and Magnetic Resonance Angiography (MRA) are the best imaging modalities for demonstrating the malformation and other associated anomalies. These also help the surgeons in the planning and approach during corrective surgery. Only very few cases of IAA have been reported in the literature. A case of a 13-day old neonate with IAA diagnosed with CTA, who had corrective surgery and was subsequently discharged home is report

Keywords: *Interrupted Aortic Arch, Computed Tomography Angiography, Neonat*e

Résumé
L'arc aortique interrompu (IAA) est une malformation congénitale très rare et elle est généralement associée à d'autres anomalies cardiovasculaires. Elle est une cause de morbidité et de mortalité néonatales précoces si elle n'est pas diagnostiquée précocement et avec précision. L'angiographie tomodensitométrique (CTA) et l'angiographie par résonance magnétique (ARM) sont les meilleures modalités d'imagerie pour mettre en évidence la malformation et les autres anomalies associées. Ceux-ci aident également les chirurgiens dans la planification et l'approche lors de la chirurgie corrective. Seuls très peu de cas d'IAA ont été rapportés dans la littérature. Je signale un cas d'un nouveau-né de 13 jours avec IAA diagnostiqué avec CTA, qui a subi une chirurgie corrective et a ensuite été renvoyé à la maison.

Mots clés: *arc aortique interrompu, angiographie par tomodensitométrie, nouveau-né*

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Introduction

Interrupted Aortic Arch (IAA) is a rare congenital malformation seen in 3 per million live births and accounts for less than 1% of all cases of congenital heart diseases [1,2]. It is defined as a complete loss of luminal continuity between the ascending and descending portions of the aorta [1]. Over 98% of IAA are associated with other cardiovascular abnormalities, such as persistent ductus arteriosus, ventricular septal defect, truncus arteriosus, or transposition of the great arteries [3]. IAA is a cause of early neonatal mortality if the ductus arteriosus closes and no correction by surgical intervention is made [1]. The diagnosis of IAA is confirmed by echocardiography and angiography. I present a 13-day old neonate with IAA with associated persistent ductus arteriosus and ventricular septal defect, who was diagnosed with echocardiography and CT angiography. He had a corrective surgery on the 15th day of life and was discharged home.

Case Report

A 13-day old male neonate with history of respiratory distress was referred to the Nelson Mandela Children hospital Johannesburg, South Africa for further management including ventilation. On physical examination, he was noted to be in cardiac failure with features of cardiogenic shock. The chest x-ray showed cardiomegaly with features of bronchopneumonia (Figure 1). An echocardiogram performed showed an interrupted aortic arch with an associated peri membranous ventricular septal defect. He was immediately placed on prostaglandin to keep the ductus arteriosus patent.

A CT angiography(Figures 2, 3 4 & 5) was performed in order to delineate the arch pathology better using multisliced spiral thin section scans plus maximum intensity projection (MIP), multiplanar reconstruction (MPR), and volume rendering reconstruction techniques.

CT angiography findings revealed normal origin of the aorta from the left ventricle with a common origin of the brachiocephalic trunk and the left common carotid artery. The brachiocephalic trunk divided normally into a right common carotid and right subclavian arteries. The aortic arch was not visualised

beyond the aforementioned branch in keeping with an interrupted aortic arch. The descending thoracic aorta was visualised from the level of T4/T5 intervertebral disc space. There was a patent ductus arteriosus, which communicated with the most

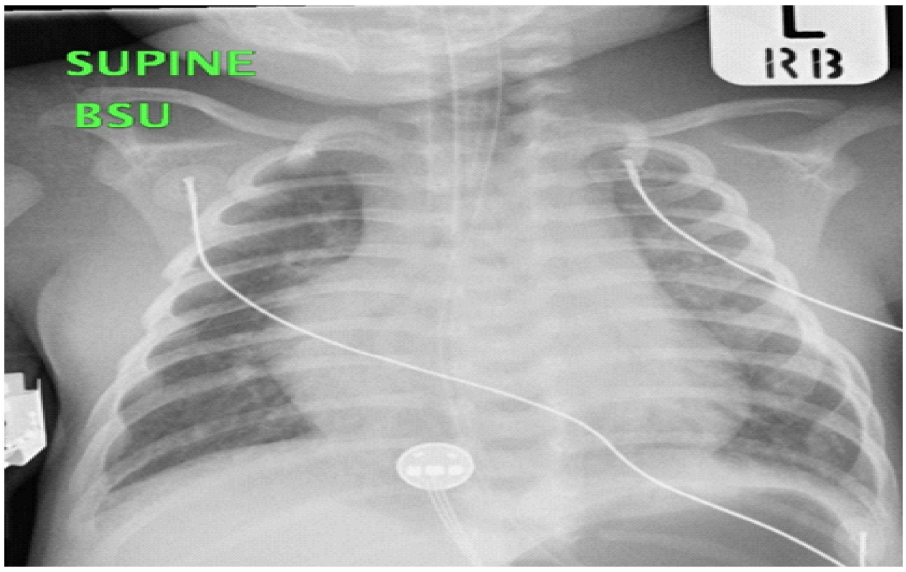

Fig. 1: *A supine anterior posterior (AP) chest radiograph of the neonate showing cardiomegaly (CTR- 0.62). There is associated hyperinflation of the lung fields with increased Broncho vascular markings. Endotracheal and nasogastric tubes are seen in-situ. Echocardiographic leads are also noticed.*

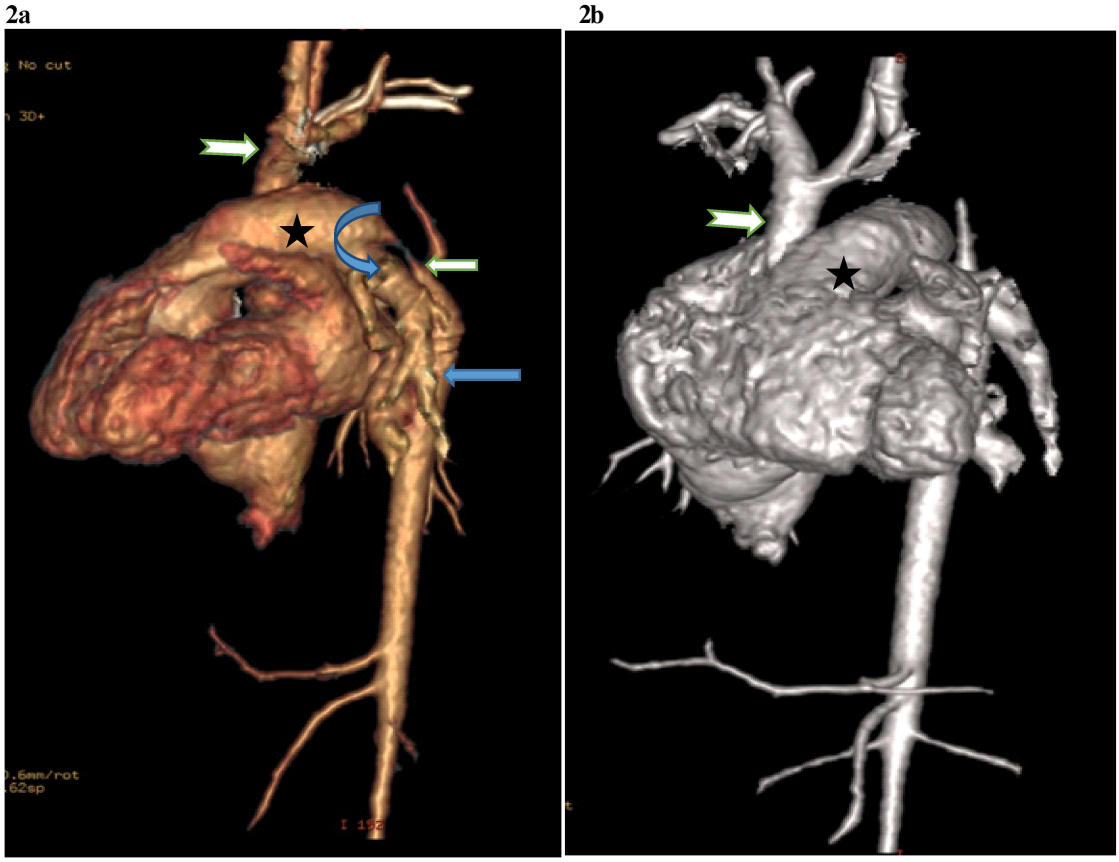

Fig. *2a and b: Volume rendering images of the heart and the great vessels showing normal origin of the aorta from the left ventricle (notched arrow) with a common origin of the brachiocephalic trunk and the left common carotid artery. The patent ductus arteriosus (curved arrow) is seen connecting it to the main pulmonary artery (star). A hypoplastic left subclavian artery (white arrow) originating off the proximal descending aorta (blue arrow)*

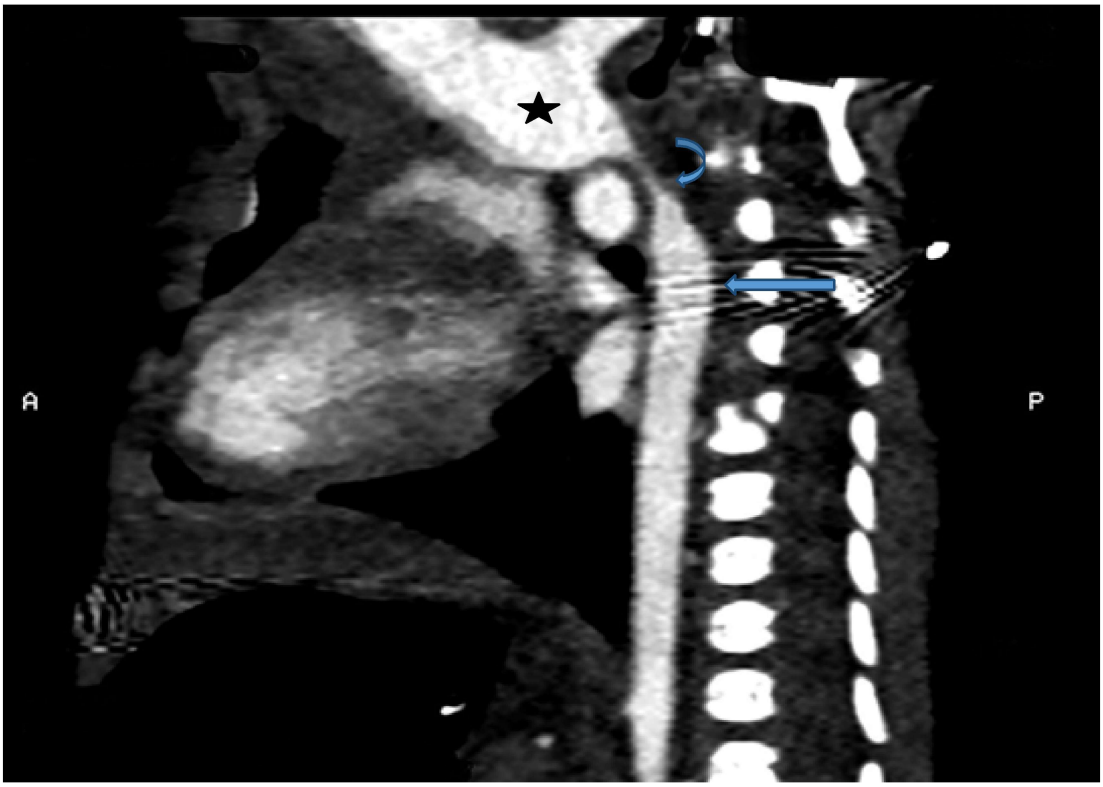

Fig. 3: *A multiplanar reconstructed (MPR) sagittal image showing the descending aorta (straight blue arrow) connecting with the main pulmonary artery (star) by a patent ductus arteriosus (curved blue arrow). The distal portion of the aortic arch is not visualized.*

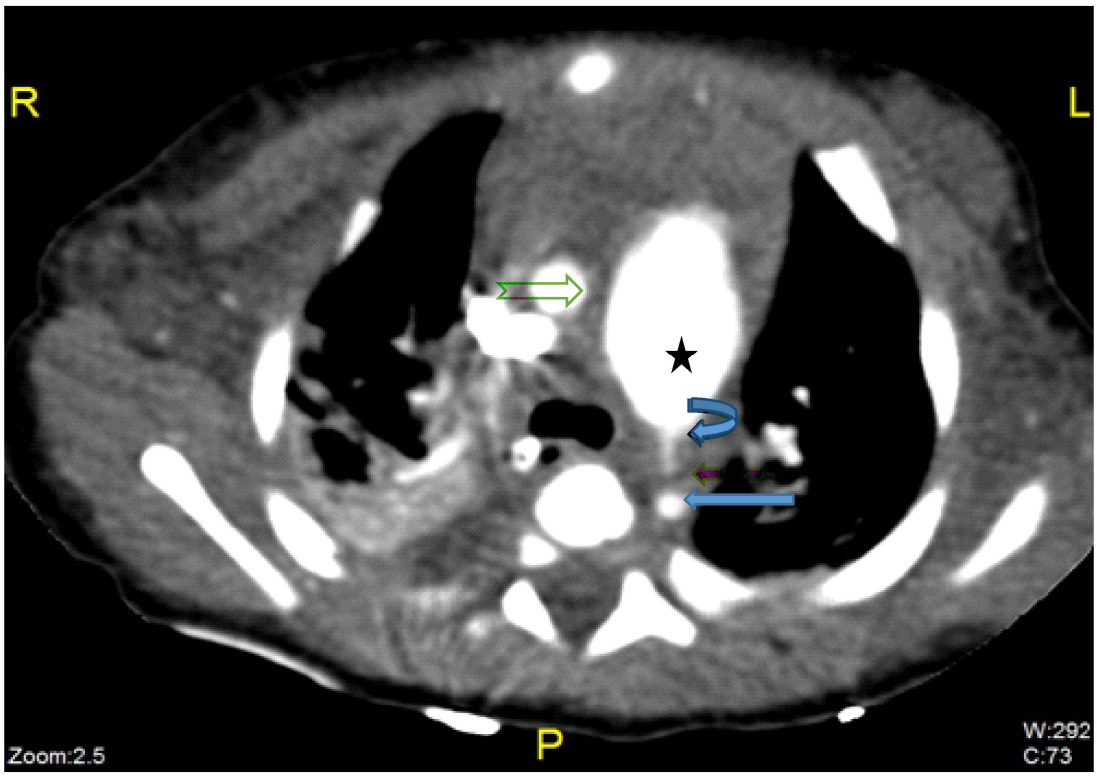

Fig. 4: *A contrasted CT, axial view at the level of the main pulmonary artery showing the patent ductus arteriosus (curved blue arrow) connecting the main pulmonary artery (star) to the small descending aorta (straight blue arrow). The ascending aorta is also noticed at this level (notched arrow).*

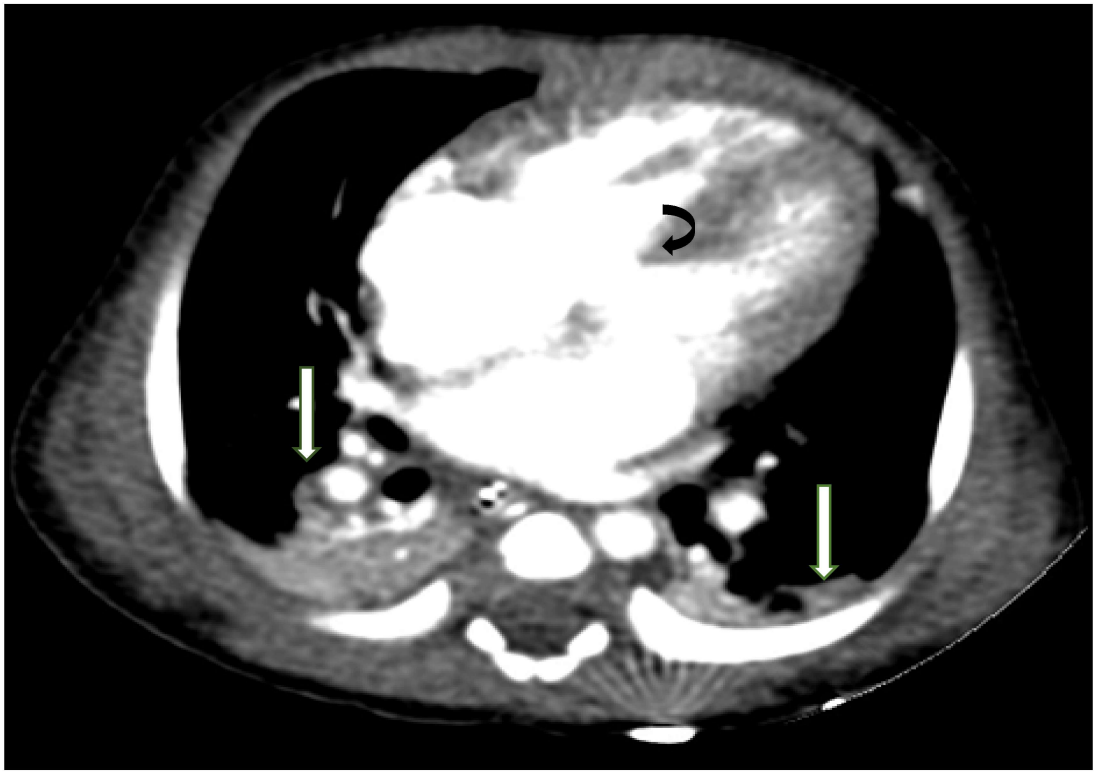

Fig. 5: *A contrasted axial CT at the level of the heart chambers showing a wide connection between the ventricles (curved arrow) consistent with a ventricular septal defect. Pneumonic changes are also noticed in both lungs posteriorly (white arrows).*

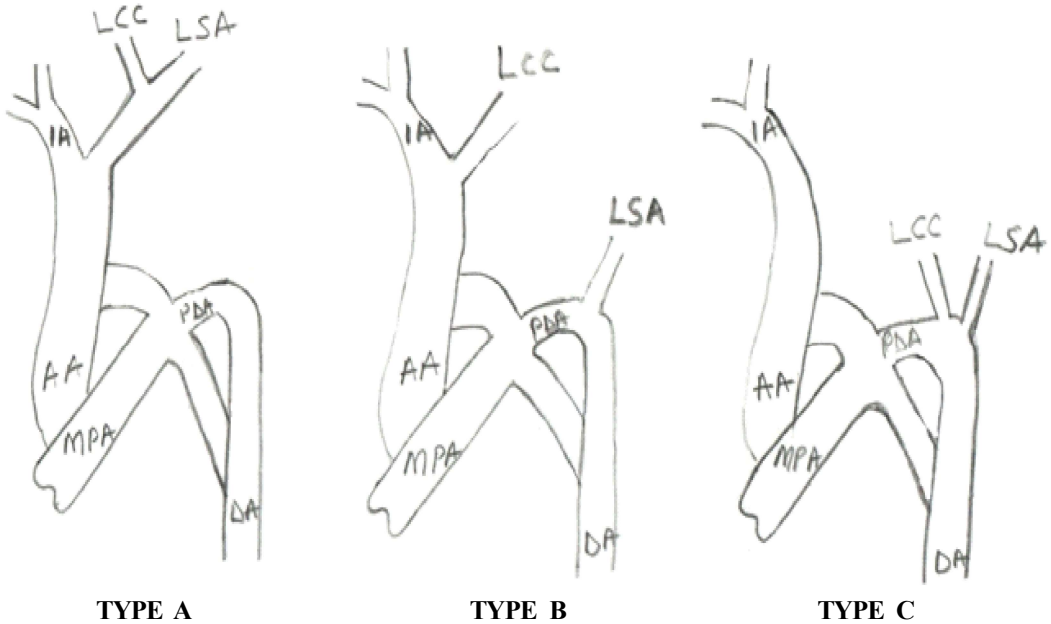

Fig. 6. *A sketch diagram showing the Celoria and Patton classification of IAA. Type A is defined as an interruption distal to the left subclavian artery (LSA). In type B, the absent segment is between the left common carotid artery (LCC) and left subclavian artery (LSA). Type C is defined as an interruption distal to the innominate artery (IA). Note that the descending aorta (DA) reconstitutes from the main pulmonary artery (MPA) through a patent ductus arteriosus (PDA). AA is the ascending aorta*

proximal aspect of the descending aorta (Figures 3 and 4). A narrow left subclavian artery arising from the descending thoracic aorta was noted.

There was a communication between both ventricles consistent with a ventricular septal defect (Figure 5). Examination of the lung fields showed consolidation in the lung fields posteriorly. (Figures 4 and 5).

In summary, a diagnosis was made of type B interrupted aortic arch with associated patent ductus arteriosus (PDA), ventricular septal defect (VSD) and underlying chest infection. Once stabilized, the patient was scheduled for corrective cardiac surgery. Findings from the surgery confirmed the diagnosis of IAA and VSD. The patient had a two-staged surgery. In the first stage surgery, the aortic arch was reconstructed by end-to-end anastomosis between the ascending and descending aorta. Thereafter, the VSD was repaired with a patch. After a few days in the critical care unit post-surgery, the patient recovered and was discharged home.

Discussion

Interrupted Aortic Arch (IAA) is a very rare anomaly of the aortic arch with an incidence of 3 in 1,000,000 births [1,2]. It was first described by Steidele [4] in 1778 while the first classification system was introduced in 1959 by Celoria and Patton [5], which is still being used**.** IAA is defined as a lack of luminal continuity between the ascending and descending thoracic aorta and it has been classified into three discrete types based on the location of the discontinuity [1,5] (figure 6). Type A is an interruption just beyond the left subclavian artery and this makes up about 30-40% of IAA cases. The type B defect is the commonest type and it occurs between the left common carotid and left subclavian arteries and is responsible for about 55% of the cases. Type C occurs in less than 5% of IAA cases making it the rarest type. It is the most proximal defect occurring between the innominate and left common carotid arteries [5]. The index patient had a type B IAA.

Congenital aortic arch anomalies result from errors in the embryologic development of the branchial arches, including errors of involution or migration [6]. The branchial arches or pharyngeal arch arteries are a series of six paired embryological vascular structures that give rise to the great arteries of the head and neck. These arches develop during the fourth and fifth week of fetal life. The third pair of arches constitutes common carotid arteries and the first part of internal carotids. The right 4th arch forms the right subclavian while the left 4th arch forms the arch of the aorta between the origin of the common carotid artery and the terminus of the ductus arteriosus [6].

Type A is likely the result of abnormal regression of the left fourth aortic arch after ascension of the left subclavian artery to its expected position. Type B occurs when the left fourth aortic arch regresses before normal ascension of the left subclavian artery to its expected position. Type C is seen when the ventral portion of the left third aortic arch and left fourth aortic arch involute, and there is a persistent ductus caroticus, a structure that normally regresses [3,6]. The patient presented had a type B IAA.

Over 98% of patients with IAA have other congenital anomalies such as sub aortic stenosis, bicuspid aortic valve, truncus arteriosus, and aortopulmonary window, VSD, aberrant innominate arteries, PDA and LVOT obstruction [2]. The presented case had associated PDA and VSD. Almost all cases present in the neonatal or early childhood period with symptoms of severe congestive heart failure with rapid clinical deterioration. There is poor prognosis with approximately 90% mortality at a median age of 4–10 days if untreated. This is usually because of the physiologic closure of patent ductus arteriosus [8]. The index patient presented on the 13th day of life with features of severe respiratory distress syndrome.

Prenatal diagnosis of IAA on echocardiogram can be challenging, however a number of anatomic features can facilitate the diagnosis. For example, a low aorta-pulmonary diameter ratio in the absence of a ventricular size discrepancy should prompt consideration of this diagnosis. Also, diagnosis of thymic hypoplasia or aplasia during fetal echocardiography can help in identifying a high-risk group for 22q11 deletion with possible associated IAA**.** In the index case, the diagnosis was not made prenatally [8].

Echocardiography has been the first-line imaging modality used in the diagnosis of IAA for a long time. However, because it is highly operator dependent and may not be sufficient for adequate evaluation of the great vessels due to acoustic window limitations, the current practice is to use additional 3D imaging in order to augment it. However, echocardiography remains the first line imaging modality to make the diagnosis [9]. The diagnosis of IAA was first made on echocardiography in this patient.

Computed Tomography (CT) or Magnetic Resonance Angiography can easily demonstrate the features of IAA and the associated cardiac anomalies because of their multiplanar capabilities. The

diagnosis is made when there is luminal discontinuity between ascending and descending aorta. The associated anomalies are better visualized on reformatted images. Multidetector CT (MDCT) is however said to have additional advantages over echocardiogram and magnetic resonance imaging [9,10]. These advantages include short scanning time which results in reduced sedation requirements, higher spatial resolution and the simultaneous evaluation of the airway and lungs.

CTA, in addition to visualization of the morphologic features of the IAA, could also help in the surgical plan and approach. It however has some disadvantages, especially in pediatric patients, which include the use of iodinated contrast medium as well as an increase in patient radiation exposure [10,11]. In the case presented MDCT was used in the diagnosis of IAA with associated PDA and VSD. The radiation dose to the patient was however lowered with the involvement of a physicist who reviewed the CTA protocol

In addition to the diagnosis of the cardiac anomalies, pneumonic consolidation was also detected. The main treatment for IAA is the reconstruction of the aortic continuity to enable appropriate blood ûow, usually by surgical means. The different surgical approaches include end-to-end anastomosis, graft interposition, or extra anastomotic bypass [12,13]. The surgical approach is usually determined by the age of the patient and the caliber of the aortic arch branches. Since this patient is a neonate, the most appropriate surgery is an end-to-end anastomosis of the ascending and descending aorta, which he had.

Conclusion

A 13-day old neonate with type B interrupted aortic arch with associated PDA and VSD has been presented. Imaging had an important role to play as there was prompt and accurate diagnosis of the anomalies with the aid of echocardiography and CTA, which assisted the surgeons in determining the surgical approach.

Acknowledgements

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