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Case Report and Literature Review: Berry Syndrome Diagnostic Approach by CT Angiography

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ABSTRACT

Berry syndrome is extremely rare multiple cardiac congenital anomalies that consist of; interrupted aortic arch, aortopulmonary window, but intact ventricular septum. To describe the advantages of CTA as one of the best modalities to diagnose berry syndrome which is able to display detailed morphology and anatomy of the heart, great vessels, and adjacent organs. A 4-month old baby boy has undergone CTA examination by giving iodinated contrast agent (370 mg/ml) 10 ml IV under sedation. The trigger scanning point was put on the cava superior vein. The findings are AP window of 1.9 cm, no aortic knob, PDA of 0.5 cm, and no defect on the ventricle or atrial septum. The findings were concluded as berry syndrome. CTA is non-invasive imaging that is believed as essential imaging of berry syndrome along with echocardiography. CTA with the radiation dose reduction technology is believed to have improved accuracy and reliability.

1. INTRODUCTION

Berry syndrome is a rare multiple cardiac congenital anomalies that was firstly reported by [3]. These cardiac congenital anomalies consist of; interrupted aortic arch (IAA), aortopulmonary window (AP Window), but an intact ventricular septum. The blood systemic circulation, in this case, raises from the pulmonary artery through AP Window and descendent aortic that is connected to the pulmonary artery like patent ductus arteriosus (PDA) [3], [4]. Berry syndrome is commonly diagnosed in neonatal and or childhood period by multimodality approach [15]. The initial diagnostic uses echocardiography with more than 60% accuracy rate reported. Echocardiography provides important information on several matters such as cardiac anatomy, functional and hemodynamic performance [4], [15]. As first described by [3], the diagnosis is provided by selective angiography [3]. However, angiography is the minimally invasive imaging procedure, since the non-invasive imaging will be most likely recommended. According to the surgical procedure of the berry syndrome, the radiologist needs to provide a detailed anatomic image of the cardiac and thoracic by Computed

Tomography Angiography (CTA) [6]. CTA is a non-invasive imaging modality that can provide high resolutions of an image with the detailed anatomic structure of cardiac and thoracic [5], [15]. However, the treatment management of these cases is a complex surgical procedure [5], [15].

Currently, CTA, as a sophisticated and non-invasive imaging modality, has improved and thus is more reliable [6]. CTA can provide high image detail and resolution. Moreover, if it is conducted by implementing new technology, which is possible to reduce the radiation, dose with optimum images quality. This is a breakthrough and a major advance in patients' diagnosis by CT scan. [6], [8] Hence, CTA will greatly support the surgical management procedure of these patients. In this paper, we report a 4-month-old boy with a diagnosis of berry syndrome.

2. Case Presentation

A 4-month-old boy has several complaints including cyanotic when crying, deep breathing, and recurring cold and cough. The patient is a second child who has a history of normal spontaneous birth, term, birth weight of 2500 grams, and spontaneous breathing. In addition, there is no history of any complaints during pregnancy. The problems began since the patient was two months old. The patient came to the Department of Radiology referred from Pediatric Department and request for CT Angiography examination. The patient was suspected of congenital cardiac anomaly. The vital signs were recorded and obtained a heart rate of 102 times/minute, blood pressure of 89/62 mmHg, breathing of 20 times/minute, and axillary temperature of 36.7°C. Furthermore, there is no cyanotic symptoms on the lips or distal extremities during the CTA examination. There was also no symptoms of the DiGeorge Syndrome. In this case, the laboratory examinations resulted blood urea nitrogen of 11 mg/dl and creatinine level of 6.28 mg/dl.

Based on the previous initial chest x-ray anterior-posterior projection, it reported the increase of pulmonary vascularization suggestive early pulmonary hypertension and inhomogeneous consolidation at the upper area of the right lung, which is concluded as pneumonia. Subsequently, previous echocardiography examination reported left atrial and ventricular dilatation with AV concordance, VA discordance, and atrial situs solitus, PDA of +/- 0.693 cm, AP Window of +/- 2 cm, and an ejection fraction of 85%.

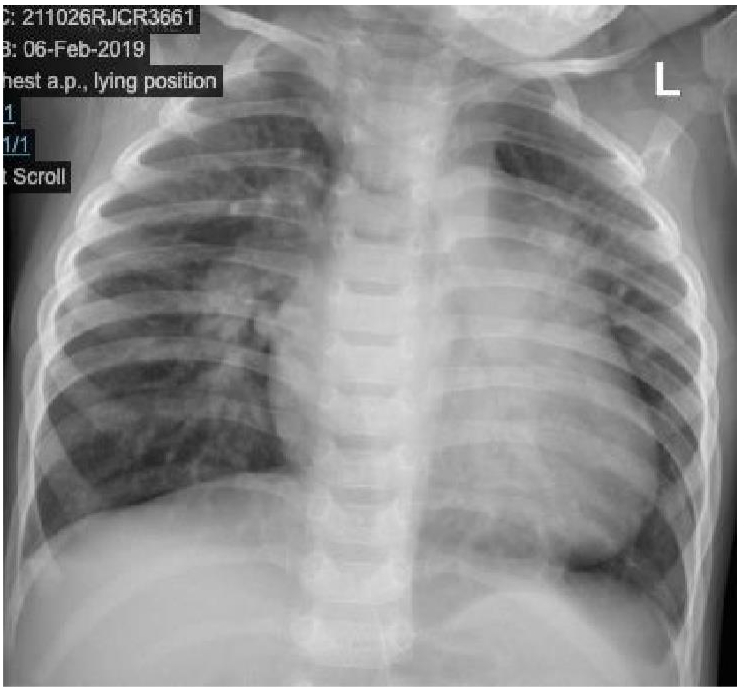

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Figure 1. A 4-month-old boy, Chest X-Ray AP projection shows increased pulmonary vascularity of both lungs and inhomogeneous consolidation in the upper area of the right lung (pneumonia and early pulmonary hypertension)

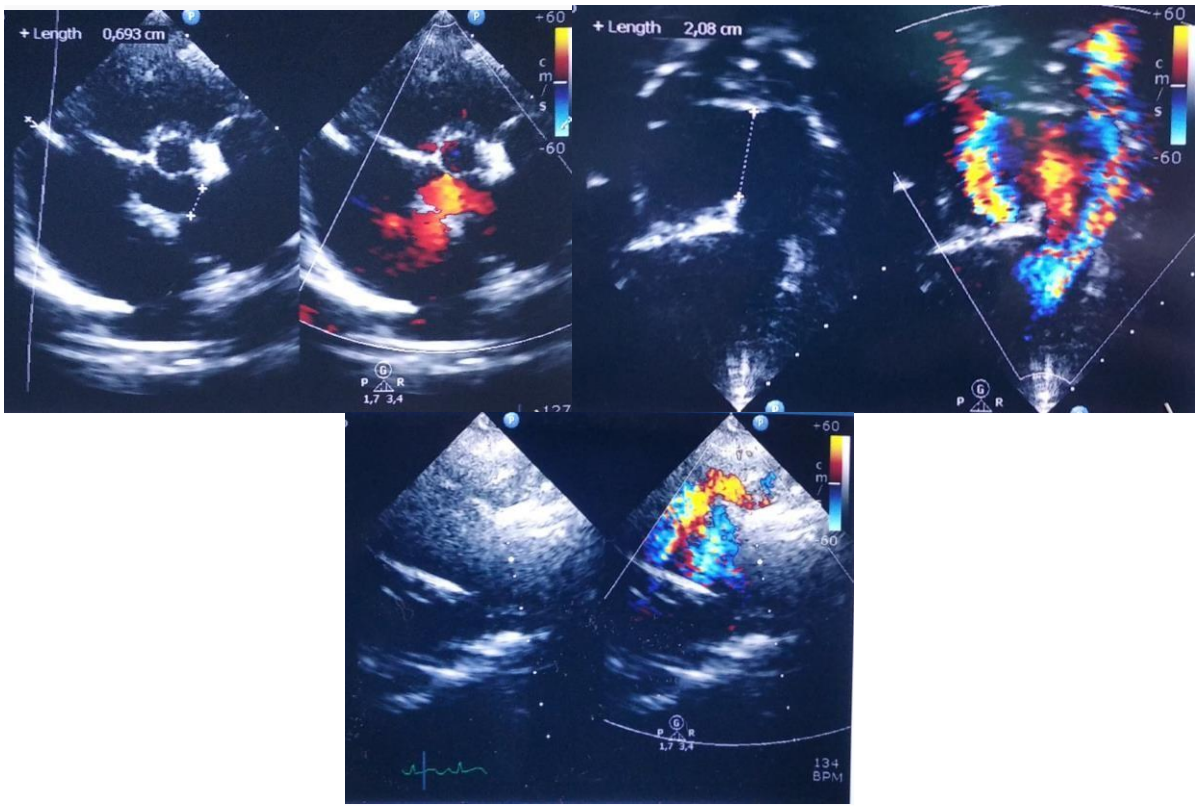

Figure 2. A 4-month-old boy's echocardiography examination shows that IAA associated with PDA (0.69 cm), the other findings is large AP Window (2 cm).

Based on the examinations above, the clinician required CTA. The CTA examination of this patient was conducted by using a CT Scan Philips 128 slices. The patient was at a supine position under propofol sedation and received iodinated contrast agent 10 ml (370 mg/ml) through the intravenous line. In addition, attached abbocath 24 gauge was attached at the right median cubital vein. Furthermore, right after the contrast was injected, 10 ml of saline flush was given. The scanning trigger point was put on the cava superior vein. We further provided the scanning for the pre-contrast, arterial phase, and vena portal phase. After the scanning was complete, the reformat image reconstruction was provided to be analysed and reported by an experienced radiologist.

The CTA findings are; the main pulmonary artery running out from the right ventricle with a diameter of 1.3 cm and there is an AP window fistulation at the aortic sinotubular junction with a diameter of 1.9 cm. The aorta runs out from the left ventricle with a diameter of 0.8 cm and the diameter of ascending aorta is 0.7 cm. In addition, there is no aortic knob formation in this patient. Other findings are the fistulation between the main pulmonary artery (MPA) and the descending aorta, which is defined as pattern ductus arteriosus (PDA) with a diameter of 0.5 cm. The diameter of the descending aorta is 0.7 cm at the level of diaphragm. Furthermore, there are no findings of septal atrial or ventricle defect reported. The CTA findings were concluded as berry syndrome.

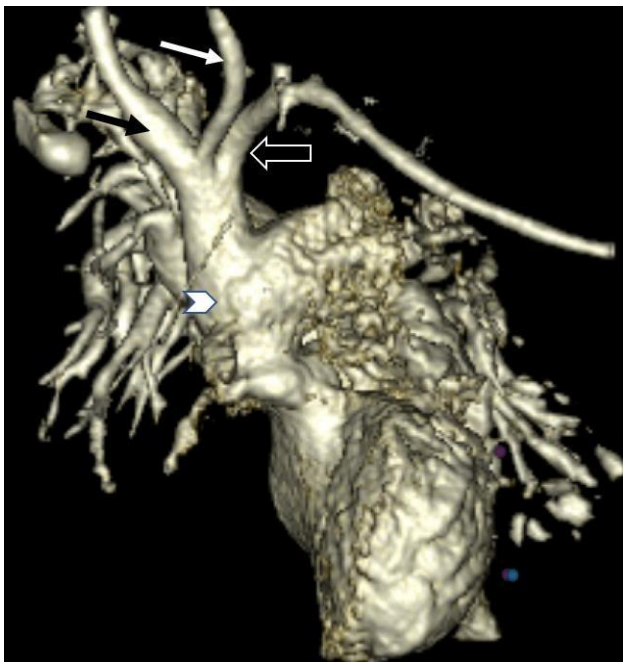

Figure 3. CT Angiography (CTA) of a 4-month-old boy patient at the 3D image of cardiac and great vascular showing the right brachiocephalic trunk (black arrow), left common carotid artery (white arrow), and left subclavian artery (empty arrow). An AP Window image at the sinotubular junction with MPA (head arrow)

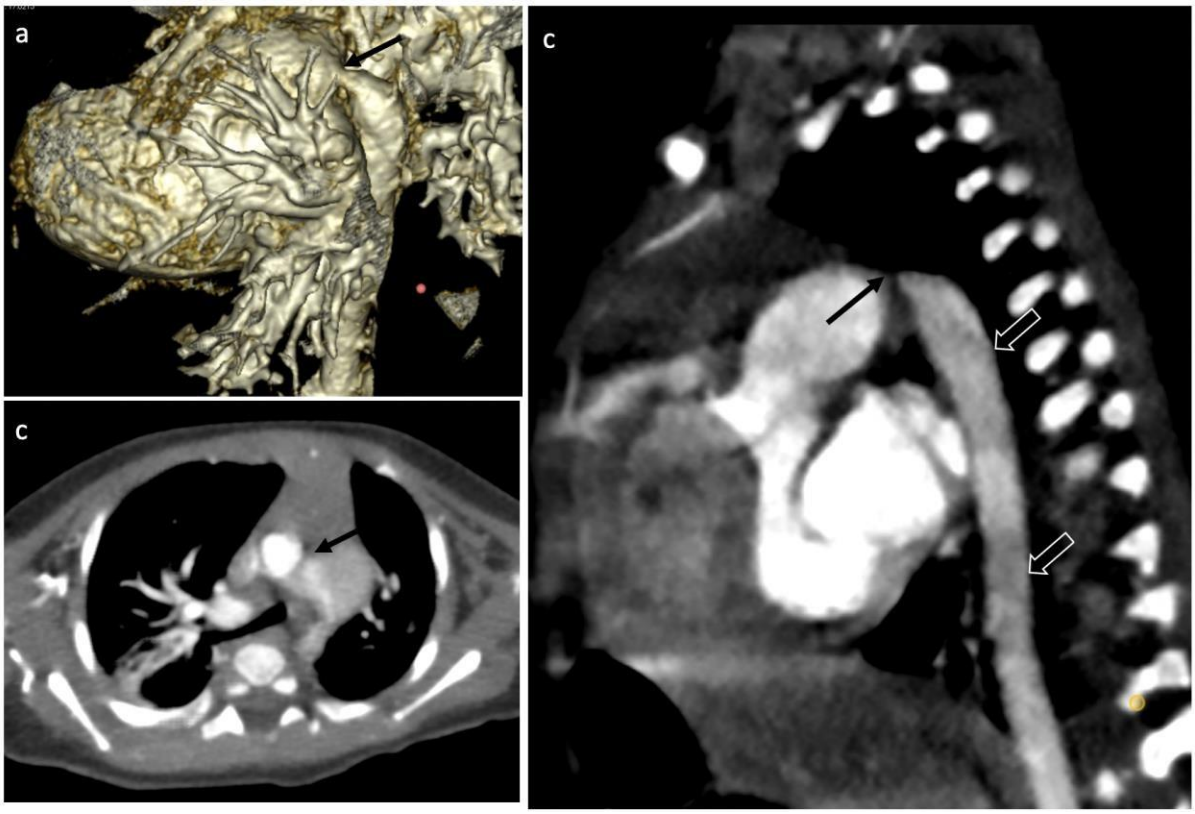

Figure 4. CTA of baby boy 4-month-old, (a) PDA, the connection between the descendent aorta and left pulmonary artery (calibre of 0.5 cm) at the 3D image, which is confirmed with the axial image (c), (b) coronal section, shows an interrupted aortic arch (IAA), no arch aortic finding, and the size and shape of the abdominal aorta is normal.

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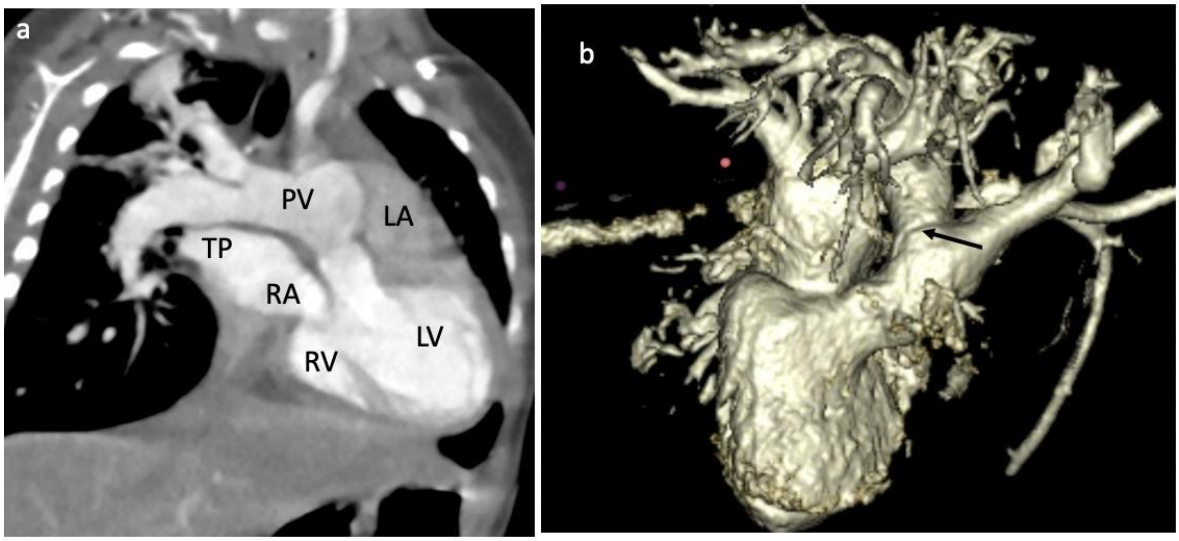

Figure 5. CTA of 4-month-old baby boy, (a) the septum ventricle is intact. (b) 3D image shows the connection of sinotubular junction of aortic and MPA (black arrow). The findings above are according to the Type A of IAA accompanied with the AP window and PDA from the MPA to the descending aorta without septal defect.

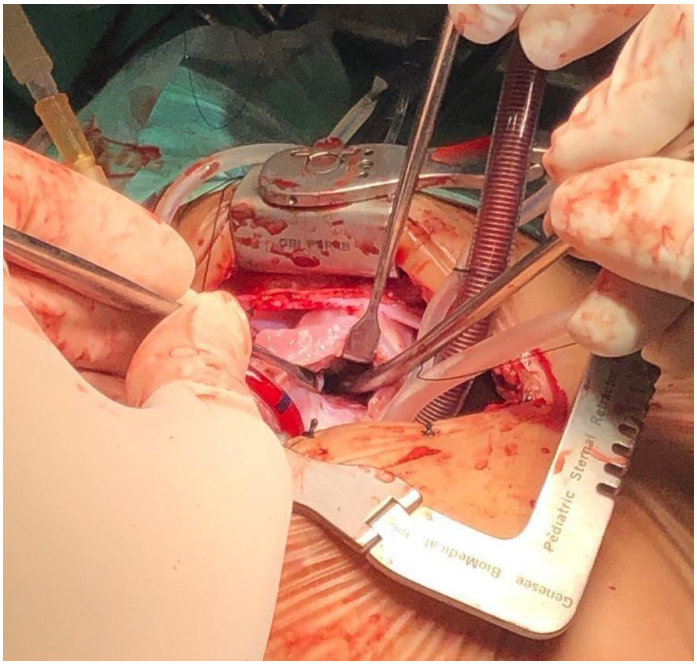

Figure 6. The surgical procedure of berry syndrome, a 4-month-old baby boy, the surgical findings confirm the previous echocardiography and CTA.

The findings above by diagnostic CTA and echocardiography are concluded as berry syndrome. The patient then underwent a surgical procedure and confirmed the berry syndrome.

The author has been approved to publish the case by the patient's parents.

3. Discussion

Berry syndrome is an extremely rare cardiac congenital anomaly [3], [4]. The incidence of berry syndrome is

only 0.046% of pediatric congenital heart anomalies. This case is slightly more common in the male gender (67.1%) [4]. The diagnostic of berry syndrome is provided by; clinical symptoms, echocardiography initial imaging, and advanced imaging by CTA.

The essential diagnostic imaging by echocardiography and CTA will provide better management of surgical procedures [4], [8], [18].

As described above, berry syndrome consists of; interrupted aortic arch (IAA), aortopulmonary window (AP Window), but intact ventricular septum defect [3], [4]. IAA is the congenital vascular malformation with discontinuity of the aortic arch. There are several classifications of IAA according to Celoria and Patton's classification in the Figure. 5 [9], [13]:

1. Type A: Aortic discontinuity is located between the left subclavian artery and descending aorta
2. Type B: Aortic discontinuity is located between the left subclavian artery and the left common carotid artery
3. Type C: Aortic discontinuity is located between the left common carotid artery and the innominate artery

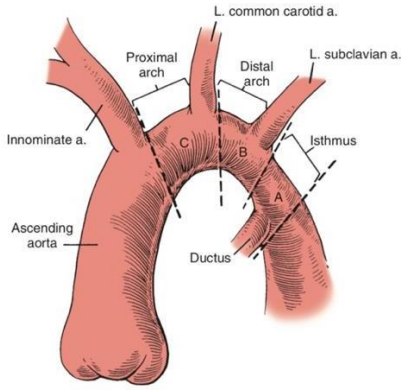

Figure 7. Overview of the anatomical location of IAA (adopted from Castañeda AR, Jonas RA, Mayer JE Jr, Hanley FL. Cardiac Surgery of the Neonate and Infant. Philadelphia, PA: WB Saunders; 1994) [9]

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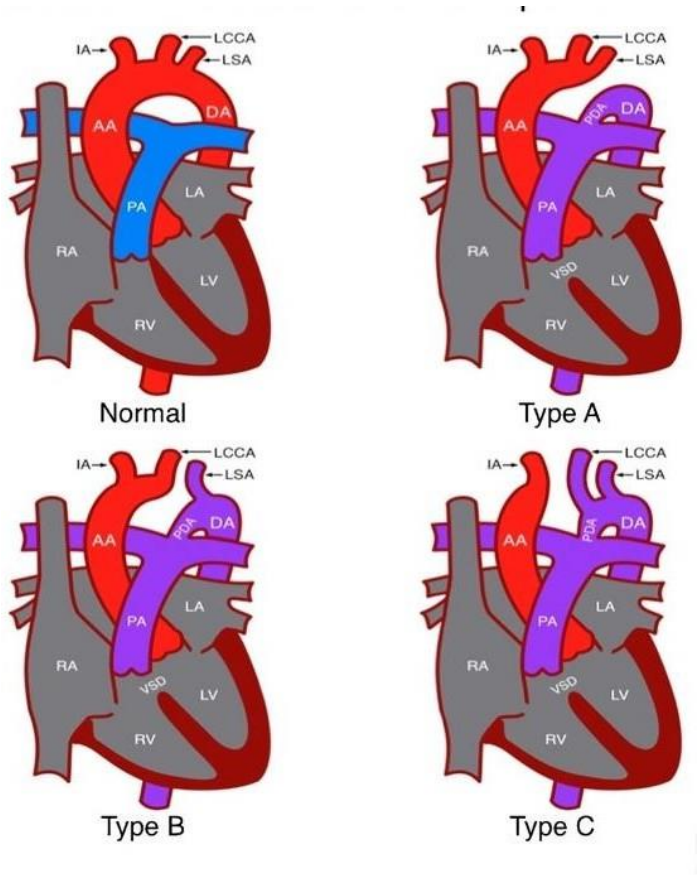

Figure 7. IAA classification by Celoria and Patton [11]

IAA is commonly associated with PDA, there is a disconnected tip of the descendent aorta that will receive the blood arising from MPA. As we were known, PDA is defined as the remaining opening of the connection between two great vessels arising from the cardiac, which are the aorta and pulmonary artery. The opening of the ductus arteriosus is normal on the baby's circulatory system [5]. However, the ductus is supposed to be close shortly after birth in 48 hours [10], [15]. The condition that remains open is defined as a patent ductus arteriosus (PDA). PDA is one of the common congenital heart diseases [1], [14], [16]. The risk factors of this abnormality are; prematurity, family history, and rubella infection during pregnancy. This is more commonly happen in Female than male [3], [4], [15].

The aortopulmonary window (AP Window) is a defect between two great vessels (aorta and pulmonary artery). This is caused by the failure of the conotruncal ridges to fuse during pregnancy [5], [16]. The defect usually begins just above the sinuses of Valsalva and then extends with various distances to distally heading to the aortic arch [9]. This abnormality causes the high pressure of arterial blood to mix with a lower pressure of venous blood circulation. Pulmonary hypertension commonly happens in this condition [9], [14]. AP Window was classified into 3 types; proximal defect, distal defect, and total defect. Furthermore, the Society of Thoracic Surgeons Congenital Heart Surgery Database Committee put the additional classification as an intermediate defect as described in Figure 8 [2].

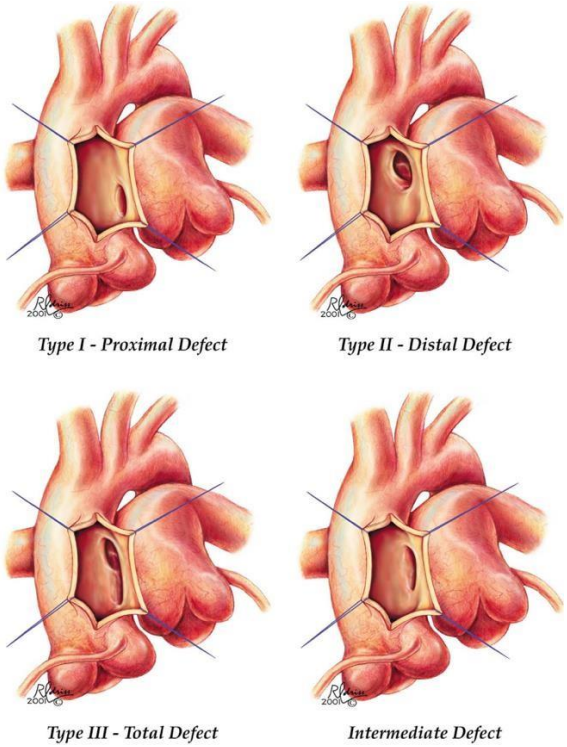

Figure 8. Types of AP windows based on the classification of the Society of Thoracic Surgeons Congenital Heart Surgery Database Committee, intermediate categories were not previously described Distal type of aortopulmonary window: report of four cases. Br Heart J 1978; 40:681– 689) [2]

Concerning the patient involved in this case report, his radiological diagnosis reported as berry syndrome, consisting of; IAA type A, AP window type 1, and PDA. In addition, there was no septal defect reported. The presented case is extremely rare and requires immediate treatment by surgical procedure. In case the treatment procedure is delayed, shunting of blood circulation through the AP window will increase the pulmonary pressure. Further, pulmonary hypertension will increase the complication during the surgical management and worst prognostic [2], [5].

Several imaging modalities are believed to have contribution in establishing the diagnosis of complex congenital cardiac anomalies. Many works of the literature suggest multimodality imaging to carry out the best imaging diagnostic result [4], [7], [17]. The chest x-ray is the common initial imaging in thoracic abnormality evaluation. In this case, the chest x-ray may evaluate cardiac contour, aortic knob, and pulmonary vascularity [16]. The diagnostic value of chest x-ray is less contributing to the diagnosis of berry syndrome. Echocardiography is the essential initial imaging of berry syndrome. Echocardiography can provide information related to hemodynamic, cardiac function, pressure, and also defining the defect of cardiac as well as the anatomy of cardiac [4], [15]. The strength points of this modality is that the absent of radiation exposure, non-invasive, and able to evaluate cardiac function [17]. Echocardiography is also able to evaluate the abnormality location of the aortic arch, the pattern of aortic branching (including evaluation for aberrant subclavian arteries) and site of obstruction, as well as other associated complex cardiac anomalies, including truncus arteriosus. The disadvantage of this modality is that it is quite operator-dependent and cannot provide a wide anatomical image of the adjacent of the cardiac [11], [17]. Furthermore, CTA is the modality that has many advantages in cardiac congenital anomaly diagnostic. Even though it cannot define functional and hemodynamic, CTA is significantly superior in providing detailed anatomy of the cardiac, great vascular, and thoracic organs as well as relative quick scanning time [17], [18]. However, detailed anatomy is needed to prepare the surgical procedures. The disadvantage of CTA is radiation exposure. The fact is that some

clinicians tend to be aware of unnecessary radiation exposure to pediatric patients [16]. Even more, CTA is superior compared to Magnetic Resonance Imaging (MRI), proven by the scanning duration, more sedation, and motion artifact due to child movement. Another advantage of CTA in the diagnosis of congenital cardiac anomaly is its ability to distinguish IAA and coarctation aorta [9].

CTA can provide high spatial images and resolution that may detect any vascular anomalies. Furthermore, CTA is able to provide a better assessment of the cardiac and vascular abnormalities which is also supported by the reformatting of coronal and sagittal images and 3D reconstruction [11], [17]. In practice, the CT Scan 16 slices are actually feasible to diagnose heart congenital anomalies, especially by providing the contrast agent. The CT Scan 16 slices are able to provide a thin slice 0.5 mm with a temporal resolution as low as 105–250 msec. However, the lack of 16 slices of CT scans will be affected by the heart movements [17]. This will then cause low image quality, reduce the anatomical morphological details of the heart organs obtained, as well as the possibility of the appearance of artifacts [18]. The CT Scan 64 or 128 slices is proven better to be able to reconstruct images at all phases of the cardiac cycle. There is a high temporal resolution for average rates of less than 70 beats per minute [12], [17]. The result of the higher slices level of CT Scan is better than the lowest slices. Furthermore, CTA also can simultaneously evaluate tracheal and oesophageal compression that may happen in cardiac problems [12], [16]. The new generation of CT Scans even has a sophisticated technology system that significantly reduces the dose of radiation exposure to the patient. This is really suitable for cardiac imaging evaluation especially on pediatric patients [12]. Scanning procedures are usually performed without gating, these are can further reduce radiation exposure as well. The high pitch spiral mode can be used without gating may significantly reduce artifacts due to heart rate movement. All of the improvements and developments above greatly increase the reliability and safety of the CTA for pediatric patients [6], [12], [17]. Therefore, CT Angiography is believed to be superior to diagnose congenital heart and blood vessel disorders of the pediatric patient [6]. Short scanning duration of the CTA may decrease the need for deep sedation duration. It will further increase the safety of the pediatric patient [17]. CTA is also able to describe the details of heart chambers, blood vessels, mediastinum, and chest wall very clearly. Hence, CTA plays a very promising role in diagnosing many types of congenital heart and vascular anomalies including berry syndrome.

4. Conclusion

Berry syndrome is an extremely rare case. The complex congenital cardiac anomaly in this syndrome includes IAA, AP window, and PDA without septal defect [3], [4]. The diagnostic procedure of berry syndrome should have high accuracy and be conducted immediately in order to reach a better prognostic [4], [6]. Echocardiography and CTA are both essential imaging in diagnostic procedures of berry syndrome, as well as other common congenital cardiac anomalies [4], [15]. CTA with radiation dose reduction technology has improved the safety as well as high accuracy and detailed anatomy image result of this modality. CTA is also a non-invasive imaging modality for evaluation of anatomical morphology, size details, and position of aortic interruption and other associated pathological abnormalities of cardiac [15].

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