JISS Journal of Indonesian Social Science

Vol. 6, No. 5, May 2025
E-ISSN:2723 – 6692
P-ISSN:2723– 6595
http://jiss.publikasiindonesia.id/

Early Recognition of Anomalous Origin of Right Pulmonary Artery From The Ascending Aorta with Minimal Limited Resources

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KEYWORDS
anomalous, right pulmonary artery, ascending aorta, pediatric, echocardiography.

ABSTRACT
The anomalous origin of the right pulmonary artery (AORPA) is a rare congenital heart defect with a poor prognosis if not surgically corrected. Atypical symptoms often lead to underdiagnosis and delayed treatment, necessitating timely and accurate examination. The utility of diagnostic tools, especially in resource-limited settings, remains a subject of debate. This research aims to assess the effectiveness of echocardiography and cardiac CT in detecting AORPA in children suspected of having congenital heart disease (CHD), compared to catheter angiography and/or surgery. A case report of a male term newborn presenting with respiratory distress and heart failure was presented. Initial diagnosis was made via echocardiography, which was later confirmed by computed tomography angiography (CTA). A literature search was conducted using PubMed, Google Scholar, and ScienceDirect with keywords such as "anomalous origin of right pulmonary artery," "ascending aorta," "children," "echocardiography," and "cardiac CT". Two relevant articles were critically appraised. The case demonstrated that transthoracic echocardiography (TTE) provided a timely diagnosis of AORPA at 11 days old, even when MDCT was delayed due to the patient's critical condition. While MDCT is superior for detailed anatomical features, TTE proved valuable as an initial diagnostic modality in a resource-limited area. Literature review showed TTE diagnostic accuracy at 88.9%, and MDCT with high sensitivity and specificity for pulmonary arterial anomalies. Despite diagnosis, the patient deteriorated and died without surgical repair. Early diagnosis and prompt surgical correction are essential for improved outcomes.

INTRODUCTION

The anomalous origin of right pulmonary artery (AORPA) is a rare cardiopulmonary defect that constitutes less than 0.5% of all congenital heart diseases (CHD) (Molossi et al., 2024; Pumacayo-Cárdenas et al., 2020). The artery abnormally originates from the ascending aorta

instead of main pulmonary artery, causes a significant left-to-right shunt, directing blood from the aorta into the pulmonary circulation (Hartmann & Screaton, 2020). This imbalance can lead to lung overload, pulmonary hypertension and congestive heart failure, therefore the common symptom is clinical heart failure. Continuous murmur and bounding pulses may be found in physical examination. Electrocardiogram can show biventricular hypertrophy, and chest imaging reveals cardiomegaly with increasing increased pulmonary vascular markings (Liu et al., 2015; Park & Salamat, 2021; Tsoutsinos et al., 2023).

Due to unspecified clinical symptoms, AORPA is easily underdiagnosed and has high mortality. The mortality rates are around 30% at three months and 70% at six months after diagnosis without surgical correction. High suspicion for AORPA needs to be applied promptly to patients with heart failure progressively after birth. Early surgical correction results excellent outcomes, therefore prompt diagnosis is essential (Nathan et al., 2007; Prifti et al., 2003).

Cardiac catheterization with angiocardiography is still considered as the gold standard for diagnostic and is useful for assessing vascular resistance. Cardiac computed tomography (CT) has been considered as standard non-invasive diagnostic in recent years for identifying AORPA as it provides clear three-dimensional images of anatomical structures from various angles and less radiation exposure. However, not all health centres have both facilities especially in remote areas with limited resources. Despite it may be insufficient for assessing extracardiac structures because of limitations in the acoustic window, echocardiography can be initial modalities for confirming timely diagnosis of AORPA due to its accessibility across all health centre.

The current research highlights the novelty of transthoracic echocardiography (TTE) as a timely and accessible diagnostic tool for anomalous origin of the right pulmonary artery (AORPA) in resource-limited settings, where advanced imaging like cardiac CT or catheter angiography may be delayed or unavailable. While prior studies emphasize MDCT's superiority for anatomical detail (Liu et al., 2015; Harraz et al., 2019) or surgical outcomes (Nathan et al., 2007), this study demonstrates TTE's 88.9% diagnostic accuracy (Wang et al., 2015) in detecting AORPA early (at 11 days old), even when MDCT was deferred due to patient instability. By validating TTE's role in critical, low-resource scenarios—complementing but not replacing advanced imaging—the research addresses a gap in the literature, proposing practical diagnostic pathways for early intervention where access to gold-standard tools is limited (Park & Salamat, 2021; Taylor, 2008). This focus on real-world applicability distinguishes it from studies that primarily advocate for MDCT or angiography without considering resource constraints.

RESEARCH METHOD

A male term newborn was born spontaneously in 38 week gestation on the way to hospital with clear amniotic fluid. He did not immediately cry and had moderate asphyxia so he got continuous positive of airway pressure (CPAP) as oxygen support. He was suspected with neonatal

pneumonia. No congenital abnormalities were detected when the mother had routine pregnancy control at the obstetrician.

At two-day-old, he experienced increasingly deeper chest wall retraction accompanied by rapid breathing despite of CPAP, therefore the patient was referred for further investigation. He was referred to our hospital then at four-day-old, presented main complaint with shortness of breath since birth. When the patient was arrived in our triage, the chest retraction still remained. On physical examination, heart rate was 162 times per minute, oxygen saturation was 99% with bubble CPAP and all peripheral pulses were equal and well felt. The capillary refill time was less than two seconds. The respiratory rate was 70 times per minute with features of moderate respiratory distress. The Downes score was seven. Breath sound was normal. The patient was treated with non-invasive positive pressure ventilation (NIPPV) and first line antibiotics.

Initial complete blood count showed hemoglobin of 16.1 g/dL, MCV 50.90 fL, MCH 16.70 pg, MCHC 32.90 g/dL, normal amount of leucocyte and thrombocyte, and without any vacuolization and toxic granulation found. differential count lymphopenia, atypical lymphocyte. Neither CRP nor IT ratio elevated. The symptoms were initially improved but on the seventh day the shortness of breath returned with ROSS score was six. The murmur was heard at the level of intercostal space IV of the left parasternal line grade II/6. Chest radiography evaluation revealed cardiomegaly with cardiothoracic ratio 64% and pulmonary plethora **(Figure 1)**. Echocardiography showed atrial situs solitus, normal systemic and pulmonal veins drain, AV-VA concordant, anomalous origin of right pulmonary artery that originates from ascending aorta, prominent eustachian valves in right atrial, patent foramen ovale (PFO) left to right shunt with 3.4 mm diameter, patent ductus arteriosus (PDA) bidirectional shunt with size of isthmus was 2 (The Ohio State University, College of Nursing, 2017). mm and ampulla was 5 (Bu & Gong, 2020). mm, no ventricular septal defect (VSD), left Ao arch, discrete CoA, mild mitral regurgitation (MR VC 2.3 mm), mild tricuspid regurgitation (TR), normal semilunar valves, no pericardial effusion, normal left ventricle (LV) and right ventricle (RV) systolic function, normal LV diastolic function. There was anomalous origin of right pulmonary artery (RPA) from ascending aorta (AAo), patent foramen ovale, patent ductus arteriosus, Discrete Coarctation of the Aorta, mild mitral regurgitation and mild tricuspid regurgitation **(Figure 2)**. He got anti failure treatment (Furosemide 3 mg, Captopril 1.5 mg, spironolactone 3.125 mg, and Digoxin 12.5 mcg) per oral every 12 hours.

Cardiac computed tomography angiography (CTA) examination was carried out with the aim of obtaining detailed anatomical structures of blood vessels for determining the 3D reconstruction to plan AORPA corrective surgery and CoA repair. It was performed with Siemens SOMATOM® Flash CT scanner that confirmed that the origin of RPA comes from the AAo as far as +/- 1.0 cm from the aortic valve with a branch diameter of +/- 0.48 cm, without visible connection from the RPA to the pulmonary trunk (PT) or left pulmonary artery (LPA), discrete coarctation at descending aorta (DAo) at Thoracic 3 level, accompanied by the presence of PDA **(Figure 3)**.

Weaning the patient off the ventilator was challenging due to the presence of pneumonia. The patient passed away two days after the CT examination. No additional comorbidities were identified, and there was no family history of heart disease. Even if the diagnosis can be established immediately, the mortality rate remains high if surgical correction is not performed soon.

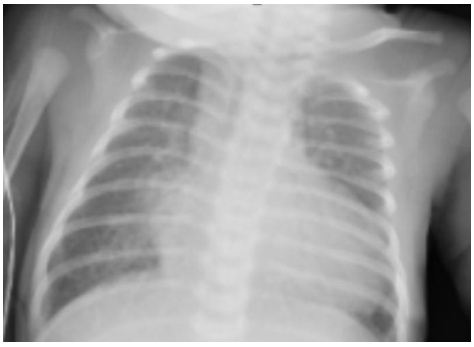

Figure 1. Rontgen Thorax AP (antero-posterior) (5 May 2023, at 9-day-old): Cardiomegaly and pulmonary plethora

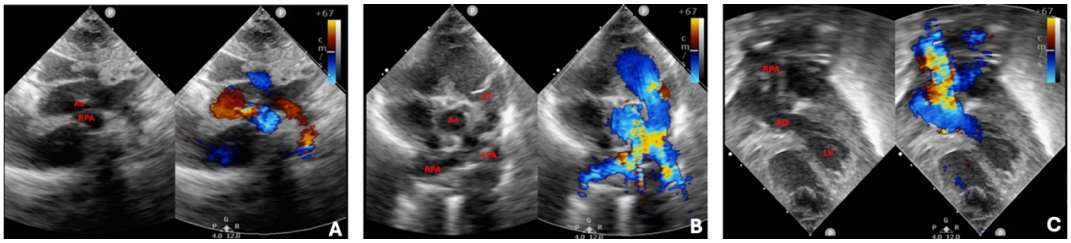

Figure 2. Transthoracic echocardiography Examination result (7 May 2023, at 11-day-old). **A**, Apical 5 chamber view shows that right pulmonary artery (RPA) arises from aorta (Ao). **B**, Suprasternal aortic arch in long-axis view showed the RPA originating from the ascending Ao with discrete CoA. **C,** Parasternal short-axis view showed that the pulmonary trunk bifurcation was absent. RPA originates from ascending Ao.

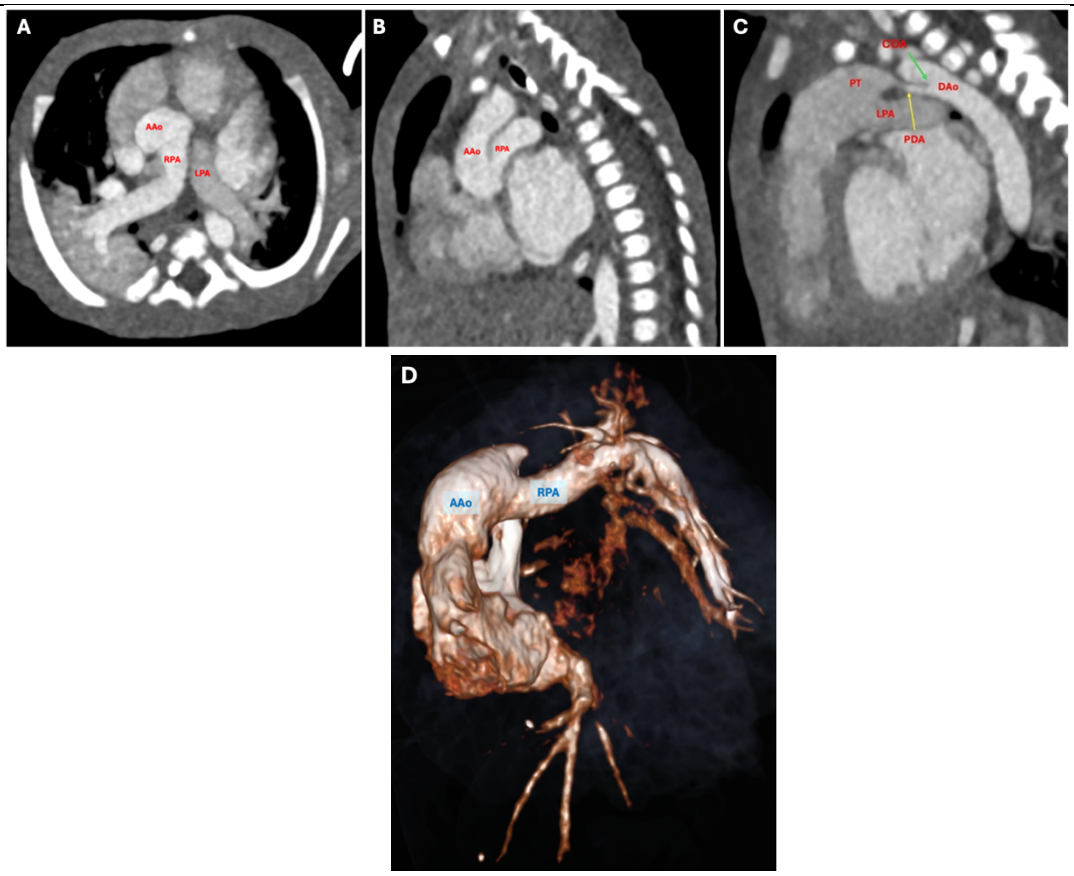

Figure 3. Cardiac CTA result (24/06/2023, at 59-day-old). **A,** Axial view of MDCT angiography reveals right pulmonary artery (RPA) originating from ascending aorta (AAo) with left pulmonary artery (LPA) arising normally from main pulmonary artery. **B,** Sagittal view shows RPA branches from the AAo and cardiomegaly. **C,** The anomaly was accompanied by the presence of PDA (yellow arrow) and discrete coarctation at DAo at Thoracic 3 level (green arrow). **D,** Three-dimensional volume rendering technique (3D VRT) image shows RPA arises from AAo, around 1 cm away from aortic valve, representing proximal subtype.

Clinical Problem

The case raises clinical question as following: in children suspected of having CHD, how effective is echocardiography and cardiac CT in detecting the presence of AORPA compared to the catheter angiography and/or surgery?

Methode

Clinical question (PICO)

Patient (P)	: children suspected of having CHD
Intervention (I)	: echocardiography or cardiac CT
Comparison (C)	: Surgery and/or catheter angiography
Outcome (O)	: AORPA diagnosis

Search Strategy

Literature search conducted to answer problem by browsing the library online using PubMed, Google Scholar, and ScienceDirect search instrument.

Selection Criteria

The keyword used is "anomalous origin of right pulmonary artery", "ascending aorta", "children", "echocardiography" and "cardiac CT" with some similar words or have the same meaning. Literature that appears during searching then adjusted to the inclusion criteria include: 1) published in English; 2) relevant to the clinical question. Exclusion criteria included: 1) book chapters and abstract conferences; 2) not available in full form text or open access; and 3) not done on pediatric patients. Based on the search method with with the above criteria, 69 articles were obtained after being reviewed **(table 1)**. Those three articles are then double filtered, abstract screening, study type selection, and full text so that it is known that there are 2 articles can be used in evidence-based case reviews with this topic. The two articles that have been selected undergo selection with the following flow **(Figure 4)**.

Table 1. Literature search results (conducted on June 30th, 2024)

Database	Search methode	Findings	Relevant articles
PubMed	(pediatric OR child OR children) AND (anomalous origin of right pulmonary artery) AND (cardiac CT OR MDCT) AND echocardiography	30	1
Google Scholar		38	0
ScienceDirect		11	1

Critical Appraisal

Two relevant articles were then reviewed critical based on the criteria of the Oxford Center for Evidence-based Medicine (Oxford Centre of Evidence-Based Medicine, 2011) for diagnostic studies and rapid critical appraisal of a descriptive study for The Helene Fuld Health Trust National Institute for Evidence-based Practice in Nursing and Healthcare (The Ohio State University, College of Nursing, 2017) for a descriptive study. The result of the critical review can be seen in Attachment 1 and 2.

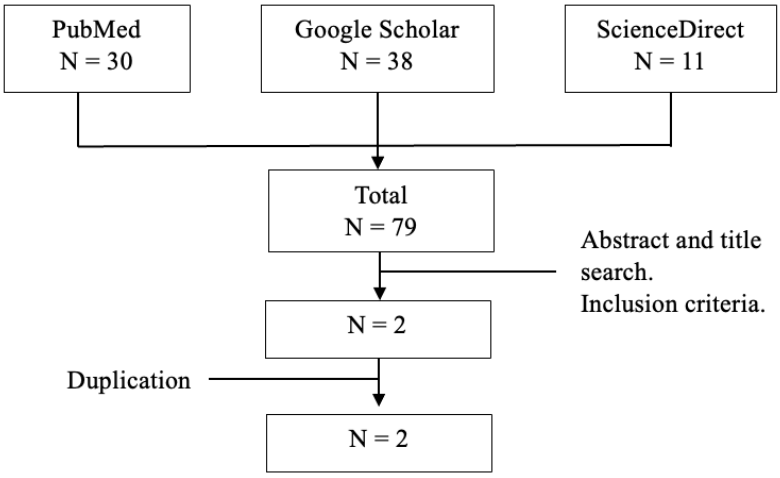

Figure 4. Literature selection flow

RESULTS AND DISCUSSIONS

A retrospective analysis performed in 9 cases with AORPA that underwent surgical correction. Transthoracic echocardiography (TTE) was able to identify eight cases, performed prior to surgery. Only one case mistakenly identified as AOLPA from the ascending aorta, leading to a conclusion that diagnostic accuracy rate between TTE and surgical findings was 88.9% (Wang et al., 2015).

A diagnostic study conducted to 52 patients aged 1 day to 4 years who had CHD and possible pulmonary arteries anomalies either clinically or by echo. All children with contraindication to contrast injection either due to allergy or impaired renal functions but not dialysis, and patients missed follow-up were excluded. All children were subjected to have MDCTA (Multi Detector Computed Tomography Angiography) that was performed by a 128-section CT scanner (Siemens SOMATOM® Definition AS) and with non-ionic iodinated contrast material. The scan was started while the contrast was seen in the upper part of descending aorta. CTA findings were compared to echocardiography (all cases), surgery (26 cases), conventional angiography (preoperative diagnosis) 19 cases and 7 cases underwent therapeutic angiography. CT pulmonary angiography was superior to echocardiography in the visualization of the morphology of the pulmonary artery congenital anomaly MDCT could diagnose all cases of pulmonary arterial anomalies with 96% sensitivity, 100% specificity, 100% positive, and 94% negative predictive values. The anomalous origin of pulmonary artery from ascending aorta was categorized into aberrant of right pulmonary artery found in 3 patients; and for the specific anomaly, MDCT can detect it with 92% sensitivity, 100% specificity, 100% positive, and 93.8% negative predictive values (Harraz et al., 2019).

Though AORPA is a lethal condition, physical examinations typically do not reveal distinctive signs. Some prior studies used different imaging modalities for diagnosis AOPA, most commonly echocardiography and angiography. Diagnosis is generally made through 2-dimensional echocardiography, but imaging of the pulmonary artery branches can be challenging, and catheter angiography would be the gold standard (Mitropoulos et al., 2010).

Cardiovascular MR angiography (MRA) risks less radiation to the patient but it is high-cost and needs longer time to image and to analyse. This means more general anesthesia will be required and the neonatal patient is at greater risk of hypothermia. Catheter angiography, which is the gold standard for AORPA diagnosis, is an invasive procedure using radiological contrast and exposing patients to radiation. The problem with both diagnostic tools is not all health centers have access to perform catheter angiography, specifically in remote areas where resources are limited (Taylor, 2008).

Multi-detector computed tomography (MDCT) is a two-dimensional array of detector elements replaces the linear detector array used in conventional and helical CT scanners. It allows the CT scanner to capture multiple slices or portions at the same time, significantly increasing the speed of CT image acquisition. Moreover, MDCT captures volume data rather than individual slice

data. These factors, combined with the use of thin section slices, allow the new technique to produce nearly isotropic data without losing the spatial resolution of the original axial images (Burrill et al., 2007). In this case, we used 128-MDCT scanners from Siemens SOMATOM® Flash CT scanner.

Compared to echocardiography, MDCT is superior due to its ability to facilitate rapid and accurate depiction of the cardiovascular anatomic features and extracardiac structures However, the severity of the clinical condition in this patient caused the patient could not be transferred to the radiology department, resulting MDCT examination to be delayed until the age of two months, whereas AORPA had been previously diagnosed by bedside echocardiography while he was 11 day-old.

Prior study revealed that the value of TTE in diagnosing AORPA was fairly high, 88,9% (Wang et al., 2015). It was confirmed by another study within 19 patients, that revealed a total of four patients (21%) with misdiagnosis by TTE compared to MDCT, including three patients with underdiagnosis of anomalous origin of any pulmonary arteries and one patient with misdiagnosis as transposition of great arteries (TGA).3 In this case, the initial TTE examination had been able to diagnose AORPA.

The definitive management for AORPA is surgical correction. Various other modifications to aortic and pulmonary arterial flap techniques have also been documented (Armer et al., 1961; Kirkpatrick et al., 1967; Prifti et al., 2002; van Son & Hanley, 1996). It is widely acknowledged that early diagnosis and prompt surgical treatment of AORPA lead to excellent short-term and long-term outcomes, 94% probability of survival at 20 years (Nathan et al., 2007).

CONCLUSION

The anomalous origin of the right pulmonary artery (AORPA) is a rare and often fatal congenital cardiovascular anomaly characterized by atypical symptoms, making early and accurate diagnosis crucial for improved prognosis. This case demonstrates that transthoracic echocardiography (TTE) can provide a timely diagnosis with a high level of accuracy, particularly in minimal resource-limited areas where access to more advanced imaging techniques like cardiac computed tomography (CT) or catheter angiography may be restricted or delayed due to patient instability. While MDCT offers superior anatomical detail, the accessibility and promptness of TTE make it an invaluable initial diagnostic modality. Despite early diagnosis, the high mortality rate without prompt surgical correction underscores the urgent need for timely intervention. Future research should focus on developing standardized protocols for early AORPA detection using readily available imaging modalities like TTE in diverse clinical settings, especially in regions with limited resources, and explore strategies to facilitate rapid transfer and surgical management for these critically ill neonates.

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