

IMAGES

in
PAEDIATRIC
CARDIOLOGY

Images Paediatr Cardiol. 2010 Apr-Jun; 12(2): 5–9.

PMCID: PMC3228332

Diagnosis of aortic origin of a pulmonary artery by echocardiography

A Taksande, E Thomas, V Gautami, and KS Murthy

Department of Pediatric Cardiology, Innova Children Heart Hospital And Research Centre, White House, Tarnaka, Hyderabad, A.P. 500017

Corresponding author Amar Taksande, Innova Children Heart Hospital And Research Centre, White House, Tarnaka, Hyderabad, A.P. 500017 ; Email: amar_bharti2000@yahoo.co.uk

Copyright : © Images in Paediatric Cardiology

This is an open-access article distributed under the terms of the Creative Commons Attribution-Noncommercial-Share Alike 3.0 Unported, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Abstract

The aortic origin of right or left branch pulmonary artery is a rare cardiac anomaly in which the right pulmonary artery usually arises from the ascending aorta just above the aortic sinuses, whereas the main pulmonary artery and the other pulmonary branch arise in their normal position. We report a rare case of isolated origin of right pulmonary artery from the ascending aorta.

MeSH: Anomalous Pulmonary artery, Echocardiography

Introduction

The aortic origin of the right or left pulmonary artery is an infrequent cardiac anomaly whose incidence is <1% of all the congenital cardiac malformations. The right pulmonary artery (RPA) arises directly from the ascending aorta in 70-80% cases, and is associated with a patent ductus arteriosus (PDA) in 75% of cases.^{1,2} It is frequently associated with other cardiac malformations and rarely presents as an isolated anomaly as in our case.³ The clinical setting is characterized by increased pulmonary blood flow, congestive heart failure, and cyanosis when the pulmonary pressure and vascular resistance are too elevated. The early diagnosis should be made by echocardiography. Early surgical intervention is recommended in all patients because of the risk of rapid development of irreversible pulmonary vascular disease. Here, we report a rare case of an infant with aortic origin of a pulmonary artery which was diagnosed by echocardiography.

Case Report

A 45 day old female infant, weighing 3.2kg was referred for the assessment of a heart murmur. A history of tachypnoea was present. She was the offspring of non-consanguineous parentage and was born by normal vaginal delivery. This baby's general physical examination was unremarkable with no evidence of pallor, cyanosis or dysmorphism. Her heart rate and respiratory rates were 152/min & 52/min respectively. Blood pressure was recorded at 107/69 mmHg and oxygen saturation 90% in room air. There was no respiratory distress. All the peripheral pulses were well felt. Cardiac auscultation revealed a normal first heart sound and a loud pulmonary component of the second heart sound along with a grade 3 ejection systolic murmur audible best at the left upper parasternal area. Chest roentgenogram revealed moderate cardiomegaly and pulmonary plethora. Electrocardiography showed sinus rhythm with normal QRS axis and peaked p waves in the limb leads and right precordial leads. On echocardiography, parasternal short axis view (fig1) and suprasternal view (fig 2) clearly showed right pulmonary artery arising from the ascending aorta and left pulmonary artery from the main pulmonary artery. There was severe tricuspid regurgitation with a peak gradient of 120mmHg. There was in addition mild pulmonary regurgitation on colour flow mapping. The right atrium and ventricle were dilated with right ventricular dysfunction.

Figure 1 Parasternal shortaxis view showing the anomalous right pulmonary artery arising from the aorta and the normal left pulmonary branch from the main pulmonary artery.



Figure 2 Suprasternal view showing the anomalous right pulmonary artery arising from the ascending aorta.



Discussion

The aortic origin of a pulmonary artery is a rare congenital anomaly in which one pulmonary artery branch, usually the right, arises from the ascending aorta just above the aortic sinuses, whereas the main pulmonary artery and the other pulmonary branch arise in their normal position. Tetralogy of Fallot, patent ductus arteriosus, right aortic arch and aortopulmonary defect are common associations in anomalous origin of right pulmonary artery (AOPA).^{4,5} This type of cardiac malformation was described first by Fraentzel in 1868. Nakamura Y et al reported the two forms: a proximal one, the AOPA originates from the ascending aorta close to the valvar plane; and a distal one in which the AOPA arises via a patent ductus arteriosus.⁶ Tsutsumi Y et al on the other hand described one form in which the origin site is close to the innominate artery or in extreme cases, the right AOPA arising from the innominate artery.⁷ Congestive heart failure develops early in infancy, with respiratory distress and poor weight gain. Cyanosis may be present due to right to left shunting through the PDA or from the persistence of the foramen ovale in the presence of a high pulmonary resistance. Our child presented with history of tachypnoea. There was no history of cyanosis. The development of pulmonary vascular disease is attributed to several mechanisms such as increased pulmonary blood flow, circulating vasoconstrictor substances, neurogenic crossover from the unprotected lung, and left ventricular failure.³ A continuous murmur and bounding pulses may be present.

The echocardiography is usually sufficient for an accurate diagnosis of AOPA from aorta but the angiogram and 3-dimensional MR angiography may confirm it.^{4,5} The images obtained from parasternal short axis and apical 4-chamber views allow a

precise diagnosis of AOPA. Echocardiography demonstrates the presence of two concordant ventricular outflow tracts, the absence of the usual main pulmonary artery trunk (MPAT) bifurcation pattern and the right or left pulmonary artery arising directly from the aorta with the MPAT continuing with the contralateral pulmonary artery branch. All the echocardiographic characteristic features of AOPA were present in our case without any other cardiac malformations. Surgical intervention is recommended as early in life as possible.^{8,9} Different techniques have been successfully employed to reimplant the AOPA to the MPAT such as direct implantation, end-to-end anastomosis with a synthetic graft, homograft patch, and 'aortic-ring' flap. The banding of the anomalous pulmonary artery and PDA ligation have been described as a palliative alternative in special cases such as premature infants with very low weight.⁹⁻¹¹ In conclusion, isolated aortic origin of the right pulmonary artery without any other cardiac malformation is in itself a rare cardiac anomaly but can be diagnosed with transthoracic echocardiography by an alert echocardiographer.

References

1. Abu-Sulaiman RM, Hashmi A, McCrindle BW, Williams WG, Freedom RM. Anomalous origin of one pulmonary artery from the ascending aorta: 36 years' experience from one centre. *Cardiol Young.* 1998;8:449–54.[PubMed: 9855098]
2. Kirklin J, Barratt-Boyes B. New York: John Wiley & Sons; 1986. Origin of the right or left pulmonary artery from the ascending aorta. *En : Cardiac Surgery*; pp. 939–44.
3. Prifti E, Crucean A, Bonacchi M, Bernabei M, Leacche M, Murzi B, et al. Postoperative outcome in patients with anomalous origin of one pulmonary artery branch from the aorta. *Eur J Cardiothorac Surg.* 2003;24:21–27.[PubMed: 12853041]
4. Rivera IR, Moises VA, Silva CC, et al. Anomalous origin of the right pulmonary artery from the ascending aorta (Hemitruncus) *Arq Bras Cardiol.* 1998;70:341–4.[PubMed: 9687640]
5. Girona J, Casaldaliga J, Sánchez C, Yeste D, Miro L. Anomalous origin of the right pulmonary artery from the ascending aorta. Its echo-Doppler diagnosis. *Rev Esp Cardiol.* 1993;46:260–2.[PubMed: 8469813]
6. Nakamura Y, Yasui H, Kado H, Yonenaga K, Shiokawa Y, Tokunaga S. Anomalous origin of the right pulmonary artery from the ascending aorta. *Thorac Surg.* 1991;52:1285–1291.
7. Tsutsumi Y, Ohnaka M, Ohashi H, Murakami A, Takahashi M, Tanaka T. A case report of anomalous origin of right pulmonary artery from innominate artery associated with left sided unilateral pulmonary hypertension. *Nippon Kyobu Geka Gakkai Zasshi.* 1991;39:447–451.[PubMed: 2051111]
8. Fong LV, Anderson RH, Siewers RD, Trento A, Park SC. Anomalous origin of one pulmonary artery from the ascending aorta: a review of echocardiographic, catheter, and morphological features. *Br Heart J.* 1989;62:389–95. [PMCID: PMC1224839][PubMed: 2590592]
9. Kutsche LM, Van Mierop LH. Anomalous origin of a pulmonary artery from the ascending aorta: associated anomalies and pathogenesis. *Am J Cardiol.* 1988;61:850–6.[PubMed: 3354450]
10. Reyes de la Cruz L, Vizcaino Alarcon A, Arevalo Salas A, Espinosa Islas G, Bolio Cerdan A, Arteaga Martinez M. Echocardiographic diagnosis of anomalous origin of one pulmonary artery from the ascending aorta. *Arch Cardiol Mex.* 2003;73:115–23.[PubMed: 12894488]
11. Fontana GP, Spach MS, Effmann EL, Sabiston DC., Jr Origin of the right pulmonary artery from the ascending aorta. *Ann Surg.* 1987;206:102–13. [PMCID: PMC1492932][PubMed: 3606229]

© Images in
Paediatric Cardiology
(1999-2012)

