CASE REPORT

A rare cause of early neonatal cyanosis: absent right pulmonary artery

Avadhesh Joshi,¹ Manish Kumar,² Sumitha Arun,¹ Mane Manisha Sheshrao³

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¹Department of Neonatology, Christian Medical College, Vellore, Tamil Nadu, India

²Department of Neonatology, Christian Medical College, Vellore, Tamilnadu, India

³Department of Radiology, Christian Medical College, Vellore, Tamilnadu, India

Correspondence to

Dr Avadhesh Joshi,
joshi.kmc@gmail.com

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SUMMARY

Unilateral absent right pulmonary artery is a rare developmental anomaly that usually presents in late childhood and adolescence as recurrent respiratory tract infections, dyspnoea and haemoptysis. We report a case of a 2-day-old baby with respiratory distress and differential cyanosis. Echocardiogram showed pulmonary hypertension with absent right pulmonary artery. The findings were confirmed by CT angiogram. The baby improved with pulmonary vasodilators and antifailure medications.

BACKGROUND

Cyanosis in newborn may herald serious underlying illness. Cyanosis results from a wide range of pulmonary, cardiac, neurological and metabolic disorders. Prompt and early diagnosis of underlying pathology helps in the appropriate management of the condition. We present a 2-day-old baby with a rare cause of cyanosis who presented with pulmonary hypertension due to absent right pulmonary artery.

CASE PRESENTATION

A term male baby, with birth weight 2.82 kg, was born to a 29-year-old mother with an obstetric score of G2P1L1 by normal vaginal delivery with Apgar score of 9 and 10 at 1 and 5 min of life. The mother did not have gestational diabetes mellitus and group B streptococcus colonisation status was not known. The baby was of non-consanguineous parentage. Amniotic fluid was clear and there was no history of aspiration pneumonia. He was active, alert and feeding well in the postnatal ward. At 26 hours of life, the patient was found to have central cyanosis and was admitted to neonatal intensive care unit for further evaluation. Examination revealed a respiratory rate of 68/min and blood pressure was 65/52 (57) mm Hg. Preductal and postductal saturation (SpO₂) difference of more than 10% was seen (preductal, 93%; postductal, 80%). Heart sounds were normal. Capillary refill time was normal. SpO₂ improved after starting oxygen (preductal, 94%; postductal, 91%).

INVESTIGATIONS

Blood counts were normal. Blood culture was sterile and sepsis screen (total leucocyte count, absolute neutrophil count and C reactive protein (CRP)) was negative. Chest radiograph was unremarkable (figure 1). Echocardiography showed absent right

pulmonary artery (online supplementary video 1 and video 2) and features suggestive of pulmonary hypertension in the form of dilated right atrium and right ventricle, patent ductus arteriosus (PDA) and patent foramen ovale (PFO) with predominant right to left shunt. CT angiogram (figures 2 and 3) showed absence of right pulmonary artery and its branches with oligoemic right lung and normal venous drainage. Tiny peripheral pulmonary arterial branches were noted on the right side with small collaterals originating from intercostal arteries and bronchial arteries. The main pulmonary artery was dilated with narrowing at the origin of the left pulmonary artery. A 2-mm PDA was noted.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses considered were neonatal sepsis, pneumonia, air leaks, congenital pulmonary malformations, PPHN (Persistent Pulmonary Hypertension of Newborn) and congenital heart disease. Sepsis workup was normal. Arterial blood gas was done for inborn error of metabolism and was normal. Chest radiograph did not show features of pneumonia or congenital pulmonary malformations. Echocardiography established the diagnosis of absent right pulmonary artery with pulmonary hypertension, which was confirmed by CT angiography.

TREATMENT

The baby was started on headbox oxygen at 8 L/min with FiO₂ 100%. He was given one normal saline bolus and started on antibiotics. On day 3 of life, tachypnoea decreased. Oxygen was tapered off and stopped when he was maintaining SpO₂ in room air. The preductal and postductal SpO₂ difference also improved. On day 4 of life, the baby worsened and had tachypnoea, increased work of breathing and oxygen requirement with hepatomegaly. Furosemide was given considering the possibility of congestive cardiac failure (CCF). The respiratory distress diminished with two doses of furosemide. Antibiotics were stopped after 3 days as culture was sterile and CRP was negative. On day 5 of life, the baby needed more oxygen. He again developed pronounced differential cyanosis. Repeat echocardiography was suggestive of pulmonary hypertension with dilated right atrium, right ventricle and predominant right to left shunt at PDA and PFO level. Oral sildenafil was started at 2 mg/kg/dose Q6H. The baby required oxygen via nasal catheter for another 48 hours, which was tapered off and stopped on day 7 of life. The baby

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Figure 1 Frontal radiograph of chest: diminished pulmonary vascular markings.

was discharged on sildenafil. There was no hypotension related to use of sildenafil. At discharge, SpO₂ was >95% in room air with no respiratory distress.

OUTCOME AND FOLLOW-UP

The patient was discharged home on exclusive breast feeding and oral sildenafil, which was stopped after 2 weeks of follow-up as

Figure 2 Axial contrast-enhanced CT scan showing absent right pulmonary artery and oligoemic right lung field.

Figure 3 CT angiogram reconstruction showing main and left pulmonary artery and absent right pulmonary artery.

opined by a paediatric cardiologist. The baby had no respiratory distress and central cyanosis on follow-up at 1 month of life. Paediatric cardiology review, repeat CT angiogram and echocardiography were planned at the age of 3 months to know the suitability for corrective surgery at a later time.

DISCUSSION

Unilateral absence of pulmonary artery (UAPA) is a rare anomaly first reported in 1868 by Frentzel.¹ The prevalence is estimated to be 1 in 200 000 in young adults with no sex predilection.² However, true prevalence in neonates is not known. The sixth aortic arch gives rise to extrapulmonary part of pulmonary arteries. Distal intrapulmonary branches arise from the lung buds. Origin of pulmonary artery from ductus arteriosus due to altered development of the sixth aortic arch is thought to be responsible for this defect. After birth, as the duct closes, the pulmonary artery segment also disappears. Almost all cases of absent right pulmonary artery have bilateral ductus arteriosus.³ However, in this baby, only left-sided ductus arteriosus was seen. Distal intrapulmonary branches usually are supplied by collateral vessels from bronchial, intercostal, internal mammary, subclavian, subdiaphragmatic or even coronary arteries.⁴ In this baby, collaterals were seen arising from intercostal arteries and bronchial arteries.

Pulmonary hypertension in a case of UAPA is due to persistence of thickened media and decreased lumen-to-wall ratio of the unaffected pulmonary artery and its branches.^{5 6} Other theories include reduced elasticity of pulmonary vasculature of the unaffected side receiving full cardiac output, shear stress on endothelium leading to endothelin release and abnormal response to vasoconstrictors.^{7 8} Two types of presentation of UAPA have been described. In infants, it presents as dyspnoea, failure to thrive, CCF and pulmonary hypertension, whereas in older patients, it presents as exercise intolerance, haemoptysis, progressive scoliosis or incidental finding on chest radiography.^{9 10} Median age of presentation in a review was 14 years (range 0.1 to 58 years).¹⁰ Singhi *et al* in their case series had five patients who were infants between 40 and 120 days (mean 86 days) at presentation and weighing 3.1–5.5 kg (mean weight 3.65 kg).⁸ This case was diagnosed on day 2 of life, which is the earliest age at diagnosis according to literature. Common cardiovascular abnormalities associated with UAPA include tetralogy of Fallot, PDA and septal defects. This baby had right-sided UAPA with PDA.

Chest radiograph may show an absent hilar shadow, absence of the left or right pulmonary artery, oligoemic lung field on the affected side, small haemothorax and intercostal space, shrunk lung and mediastinal shift to the affected side with hyperinflation of the contralateral lung.^{10 11} In our baby, chest radiograph

appeared normal initially. CT showed an oligoemic lung field on the right side. Chest radiograph was reviewed following CT scan and appeared to have diminished pulmonary vascular markings on the right side.

Revascularisation of branches of pulmonary artery to hilum on the affected side can be done. If intrapulmonary arteries are well developed, primary anastomosis with central pulmonary artery can be made. Modified Blalock-Taussig shunt can be used if intrapulmonary arteries are small. This leads to better growth of affected intrapulmonary arteries.^{3 12} A two-stage procedure described by Krammoh *et al* includes PDA stenting followed by surgical anastomosis in the second stage.⁹

In this baby, only tiny peripheral pulmonary arterial branches were noted on the right side. Hence, surgical correction was not possible. Our baby was managed conservatively as the left lung supply was receiving the whole right ventricular output. The blood supply to the right lung, which was supplied by small collaterals, was significantly reduced. This can cause left lung volume overload due to excessive flow, while the right lung is oligoemic. Corrective surgery is possible only if the right lung supply can be clearly identified and effort is done to unifocalise the supply to become a right pulmonary artery if possible. Corrective surgery should not be postponed too long as the left lung can be damaged by prolonged severe pulmonary oedema.

Diagnostic catheterisation will help to identify hilar arteries better and measure differential pressures in each segment. This will help to plan for corrective surgery.¹⁰ Diagnostic catheterisation has been planned for this patient as preoperative workup.

The management of pulmonary hypertension includes the use of vasodilator therapy (nitric oxide, magnesium sulfate, phosphodiesterase 5 inhibitors like sildenafil and endothelin inhibitors like bosentan). Sildenafil is a pulmonary vasodilator that can lead to excessive pulmonary artery flow if used in conditions with low pulmonary vascular resistance (PVR) and high Qp/Qs. In a case of UAPA, this will lead to increased flow to lung with normal vasculature and volume overload, deteriorating the clinical condition. Sildenafil was started in this patient in view of pulmonary arterial hypertension on echocardiography with normal left lung field on CT scan. In our case, there was clinical improvement with sildenafil, which implies that the left pulmonary artery has developed pulmonary arterial hypertension and has elevated PVR while blood flow to the right lung is severely reduced.

In babies with UAPA, increased pulmonary blood flow on the normal side can be managed by restricting flow with a pulmonary artery band to prevent pulmonary arterial hypertension. Unifocalisation and connection to main pulmonary artery can be planned later.

Isolated unilateral absence of pulmonary artery can be a rare cause of unexplained cyanosis in newborn period. Early diagnosis will help in better planning of management.

Learning points

- ▶ Absent right pulmonary artery is a rare cause of neonatal cyanosis.
- ▶ It can present in neonatal period with cyanosis, pulmonary hypertension and cardiac failure.
- ▶ Echocardiography is diagnostic.
- ▶ CT angiogram can describe associated cardiac defects and collaterals.
- ▶ Conservative treatment with oral sildenafil may be beneficial.

Contributors MK and SA were involved in diagnosis. AJ collected clinical details. MK and AJ edited the manuscript. MMS helped with CT reconstruction images.

Competing interests None declared.

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