Cardiology in the Young

cambridge.org/cty

Images in Congenital Cardiac Disease

Cite this article: Pandey NN, Sinha M, and Kumar S (2020) Unilateral pulmonary aplasia and congenital diaphragmatic hernia associated with tetralogy of Fallot: a rare trifecta. *Cardiology in the Young* **30**: 121–122. doi: 10.1017/S1047951119003251

Received: 30 August 2019
Revised: 9 December 2019
Accepted: 16 December 2019
First published online: 9 January 2020

Keywords:
Tetralogy of Fallot; pulmonary aplasia; congenital diaphragmatic hernia

Author for Correspondence: Dr S. Kumar, Department of Cardiovascular Radiology & Endovascular Interventions, All India Institute of Medical Sciences, New Delhi 110029, India. Tel.: +91-9968968918;
E-mail: sanjeevradio@gmail.com

CAMBRIDGE UNIVERSITY PRESS

Unilateral pulmonary aplasia and congenital diaphragmatic hernia associated with tetralogy of Fallot: a rare trifecta

Niraj N. Pandey, Mumun Sinha and Sanjeev Kumar

Department of Cardiovascular Radiology & Endovascular Interventions, All India Institute of Medical Sciences, New Delhi, India

Abstract

We present a case of a 7-month-old boy with tetralogy of Fallot associated with unilateral pulmonary aplasia and herniation of the liver and small bowel loops in the right hemithorax.

Case report

A 7-month-old boy presented with respiratory distress, cyanosis, and feeding difficulties. Oxygen saturation on room air was 78%, which improved to 83% on oxygen supplementation. Chest radiograph revealed near-complete opacification of the right hemithorax with herniation of liver and small bowel loops; however, no apparent tracheo-mediastinal shift was noted (Fig 1a). Transthoracic echocardiography established a diagnosis of tetralogy of Fallot with non-visualisation of right pulmonary artery. Patient was subsequently referred for computed tomography angiography for evaluation of the pulmonary vasculature and airways. Computed tomography angiography confirmed the diagnosis of tetralogy of Fallot with complete absence of right lung, right pulmonary artery, and right pulmonary veins. A rudimentary right-sided

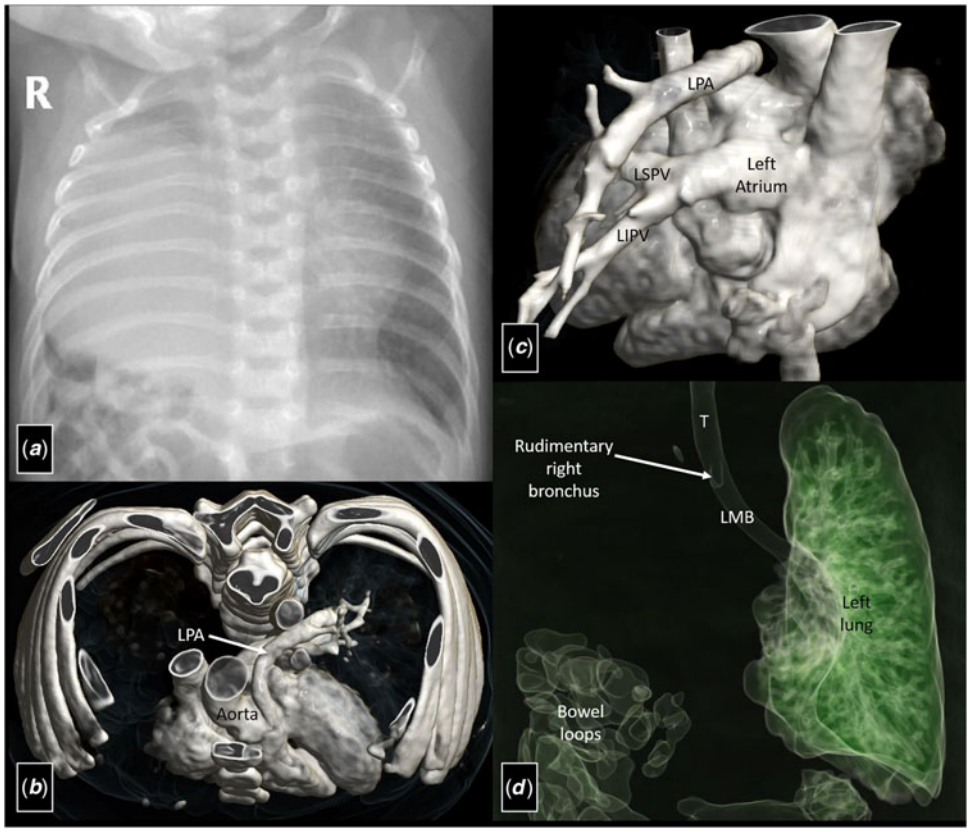

Figure 1. Chest radiograph (***a***) reveals near-complete opacification of the right hemithorax with herniation of the liver and small bowel loops without any apparent tracheo-mediastinal shift. Volume-rendered images (***b***–***d***) of computed tomography angiography show the absence of the right pulmonary artery and the right pulmonary veins with a rudimentary right-sided bronchus. LIPV = left inferior pulmonary vein; LMB = left main bronchus; LPA = left pulmonary artery; LSPV = left superior pulmonary vein; T = trachea.

bronchus was, however, present (Fig 1b–d). Associated herniation of liver and small bowel loops in the right hemithorax was noted.

Unilateral pulmonary agenesis is an exceedingly rare anomaly and is associated with other congenital anomalies in ~50% cases. Pulmonary agenesis is classified into three types depending on the degree of arrest of development. Type I (pulmonary agenesis) is characterised by complete absence of a lung along with its bronchus and vessels. Type II (pulmonary aplasia) shows complete absence of a lung albeit with a rudimentary bronchus present. In type III (pulmonary hypoplasia), there is partial presence of the bronchial tree along with parts of unilateral lung and its vasculature.[1] As development of the lung occurs in the 4th–5th week of gestation around the same time the migration of heart occurs, pulmonary agenesis is frequently associated with CHDs. To the best of our knowledge, this unique combination of anomalies in association with tetralogy of Fallot has been reported only once previously.[2]

Acknowledgements. None.

Financial Support. This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Conflicts of Interest. None.

References

1. Boyden EA. Developmental anomalies of the lungs. Am J Surg 1955; 89: 79–89.
2. Labovsky K, Hoffman G, Scott J. Management of a patient with tetralogy of Fallot, congenital diaphragmatic hernia, and complete left lung agenesis. A A Case Rep 2016; 7: 16–20.

https://doi.org/10.1017/S1047951119003251 Published online by Cambridge University Press