Common pulmonary vein atresia

Thomas Glenn¹, Jose Honold², Beth F. Printz¹ and Dana Mueller¹

¹Division of Cardiology, Department of Pediatrics, University of California San Diego, Rady Children’s Hospital San Diego, San Diego, CA, USA and ²Division of Neonatology, Department of Pediatrics, University of California San Diego, Rady Children’s Hospital San Diego, San Diego, CA, USA

Abstract

A 4-hour-old infant with profound cyanosis on an alprostadil infusion was urgently transferred to Rady Children’s Hospital with suspected CHD. Upon arrival, urgent echocardiography was performed but could not confirm the presence of discrete pulmonary veins or pulmonary venous drainage. Given the difficulty in delineating the anatomy, a cardiac CT scan was performed and demonstrated a nearly atretic common pulmonary vein with multiple small collaterals that drained to systemic veins. Due to the high risk of mortality associated with operative repair, the decision was made to proceed with compassionate withdrawal of care. The described anatomy of common pulmonary vein atresia remains rare, and to our knowledge, fewer than 40 cases have been reported in the literature. Albeit rare, common pulmonary vein atresia should be considered in the differential diagnosis of a severely cyanotic neonate.

Case report

A newborn female was born at 39 3/7 weeks gestational age at an outside hospital to a 32-year-old G2P0 Hawaiian female with adequate prenatal care. The mother had no history of any chronic health conditions and the pregnancy was reportedly uncomplicated, with normal prenatal ultrasounds and laboratory results. Apgar scores were 7 and 8 at 1 and 5 minutes, respectively. Shortly following delivery, the patient developed significant tachypnoea and cyanosis with oxygen desaturations to the 50’s on room air. The patient was started on non-invasive continuous positive airway pressure and an alprostadil infusion. Initial echocardiogram was concerning for hypoplastic left heart syndrome-variant versus Shone’s complex, and the patient was transferred to the cardiothoracic ICU at our institution for further care.

On arrival, the patient remained on an alprostadil infusion at 0.1 mcg/kg/minute, with oxygen saturations persistently in the 50’s despite ongoing respiratory support with nasal continuous positive airway pressure with FiO2 100%. The patient was immediately intubated but remained significantly desaturated. Repeat echocardiogram was obtained (Fig 1), which revealed the absence of pulmonary veins entering the atrium and absence of dilated pulmonary veins or large vertical vein. There was all right to left shunting across an unrestricted atrial communication, and there was a large patent ductus arteriosus with predominantly right to left flow. A trivial pulmonary venous confluence was seen posterior to the left atrium. The right ventricle was dilated and the left ventricle was apex forming but appeared underfilled. Given that discrete pulmonary veins or venous drainage could not be delineated by echocardiography, cardiac CT scan was emergently obtained. Cardiac CT revealed a trivial pulmonary venous confluence with small left and right veins posterior to the left atrium; however, no discrete connection to the heart was identified (Fig 2). Due to the near atretic nature of this patient’s pulmonary veins and common pulmonary vein and lack of identified collaterals, extensive multidisciplinary discussions were held between Cardiology, Cardiac Intensive Care, Radiology, and Cardiothoracic Surgery teams. Ultimately, it was determined that there was no viable operative repair, and the decision was made between the family and the medical team to proceed with withdrawal of care. The patient was transferred back to the cardiothoracic ICU, compassionately extubated, and died shortly thereafter. An autopsy was performed which confirmed the absence of any pulmonary venous connection to the left atrium. There was a trivial pulmonary venous confluence superior to the left atrium as noted on the echocardiogram and CT, with a trivial vein that appeared to drain from the confluence to the innominate vein. No dilated pulmonary veins were demonstrated by autopsy. On gross examination, the lungs had a diffusely nodular appearance and histological examination revealed findings consistent with cystic lymphangiectasia.

Discussion

Common pulmonary vein atresia is a rare, but serious, form of total anomalous pulmonary venous return, which can lead to cyanosis and respiratory distress in the early postnatal period. As Lucas et al first described in 1962, the common pulmonary vein is an embryonic structure...
that develops as an outgrowth of the heart that makes connection
to the venous plexus which drains the pulmonary primordia. As
development continues, the common pulmonary vein is eventually
incorporated into the left atrial wall. The group suggests that in
total anomalous pulmonary venous return with anomalous drain-
age, this connection between the left atrium and common pulmo-
nary vein is obliterated early in development, such that collateral
venous drainage develops and anomalous venous return persists.
In cases of common pulmonary vein atresia, the common vein
is thought to obliterate late in development, leading to the inability
of collaterals to form and as a result, the presentation described
above. Various institutions have attempted to determine the exact
incidence of this condition based on retrospective autopsy reviews,
with reported results varying from 0.03% to 0.22% of patients with
suspected CHD. In 2008, Vaideeswar et al reported that the total
number of documented cases of common pulmonary vein atresia
in the literature to be around 25, a number which had grown to 35
by 2015, as reported by Perez et al. This number likely underre-
presents the number of affected patients, given the difficulties asso-
ciated with accurately establishing this diagnosis.

Cases of common pulmonary vein atresia present a unique sur-
gical challenge, which is made more difficult because of the lack of
prenatal diagnosis. In some instances, prenatal diagnosis of total
anomalous pulmonary venous return is incorrectly made, and only
after postnatal echocardiogram is performed, the diagnosis of
common pulmonary vein atresia is made. Unfortunately, even with prompt diagnosis, survival is suboptimal even if operative
repair is attempted and commonly depends on the presence of a
sizeable venous confluence, which the patient in our case was lack-
ing. In 2015, Perez et al described a series of three patients with
similar anatomic findings to the patient in our institution, none
of whom were offered surgical repair. Of the less than 40 reported
cases of common pulmonary vein atresia, there have been only 5 successful surgical repairs,\(^6\)\(^8\) highlighting the typical poor prognosis. Additionally, long-term data on these few survivors are not available.

Unfortunately, without surgical repair, common pulmonary vein atresia is universally fatal. Our report of a patient with common pulmonary vein atresia demonstrates the need for prompt diagnosis and evaluation by echocardiography, CT, and/or cardiac catheterisation to determine the presence and/or size of a pulmonary venous confluence. It also emphasises the importance of a cohesive, multidisciplinary approach between multiple subspecialty teams in evaluating and determining surgical candidacy and best course of action. As more patients with common pulmonary vein atresia are identified, hopefully necessary, continued reporting and expanded, ongoing research into this rare population will continue.

Acknowledgements. The authors would like to acknowledge the cardiothoracic surgical team at our institution for their contributions to medical discussions about this case.

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

Conflicts of interest. None.

References
1. Lucas RV, Woolfrey BF, Anderson RC, et al. Atresia of the common pulmonary vein. Pediatrics 1962; 29: 729–739.
2. Vaideeswar P, Tullu MS, Sathe PA, et al. Atresia of common pulmonary vein: a rare congenital anomaly. Congenit Heart Dis 2008; 3: 431–434.
3. Deshpande JR, Kinare SG. Atresia of the common pulmonary vein. Int J Cardiol 1991; 30: 221–226.
4. Perez M, Susheel Kumar TK, Briceno-Medina M, et al. Common pulmonary vein atresia: report of three cases and review of the literature. Cardiol Young 2015; 26: 636–637. DOI 10.1017/S1047951115002450.
5. Nakamura Y, et al. An extremely rare variant of pulmonary venous atresia. Ann Thoracic Surg 2016; 101: 2382–2384. DOI 10.1016/j.athoracsur.2015.08.085.
6. Dudell GG, Evans ML, Krous HF, et al. Common pulmonary vein atresia: the role of extracorporeal membrane oxygenation. Pediatrics 1993; 91: 403–410.
7. Mas C, Cochrane A, Menahem S, et al. Common pulmonary vein atresia: a diagnostic and therapeutic challenge. Pediatr Cardiol 2000; 21: 490–492.
8. Suzuki T, Sato M, Murai T, et al. Successful surgical repair of common pulmonary vein atresia in a newborn. Pediatr Cardiol 2001; 22: 255–257.