Cardiology in the Young

cambridge.org/cty

Premature closure of the arterial duct presenting with right heart failure of the fetus and ductal aneurysm postnatally

Mathias Michelitsch[1], Dominik Wiedemann[2], Elisabeth Schermer[3], Daniel Zimpfer[2] and Ina Michel-Behnke[1]

[1]Division of Pediatric Cardiology, Department of Pediatrics and Adolescent Medicine, Pediatric Heart Center, Medical University of Vienna, Vienna, Austria; [2]Department of Cardiac Surgery, Pediatric Heart Center, Medical University of Vienna, Vienna, Austria and [3]Department of Child and Adolescent Health, Pediatrics III (Pediatric Cardiology, Pulmonology, Allergology and Cystic Fibrosis), Medical University of Innsbruck, Innsbruck, TIR, Austria

Brief Report

Cite this article: Michelitsch M, Wiedemann D, Schermer E, Zimpfer D, and Michel-Behnke I (2023) Premature closure of the arterial duct presenting with right heart failure of the fetus and ductal aneurysm postnatally. *Cardiology in the Young* **33**: 2690–2692. doi: 10.1017/S1047951123003359

Received: 15 December 2022
Revised: 6 June 2023
Accepted: 3 September 2023
First published online: 31 October 2023

Keywords:
Premature; ductus; closure; ductal; aneurysm

Corresponding author:
M. Michelitsch;
Email: mathias.michelitsch@meduniwien.ac.at

Abstract

In utero idiopathic constriction of the arterial duct is a rare condition with only a handful reported cases. Ductal aneurysms with thrombus formations on the other hand are significantly more common. We report a case of a term infant who presented with right heart failure due to premature ductal closure and postnatal severe respiratory distress. Subsequent diagnostics revealed paresis of left laryngeal nerve and obstruction of the left pulmonary artery secondary to a ductal aneurysm. Consequently, surgical intervention was considered necessary. Post-operatively, right ventricular function and hoarseness resolved slowly.

Background

The arterial duct, in fetal circulation, allows relief of the right ventricle by bypassing the high-resistance lung. Depending on the gestational age, 50–75% of the right ventricular volume is pumped via the arterial duct into the systemic circuit.[1,2]

Consequently, premature ductus constriction or closure leads to a massive increase of right ventricular afterload with subsequent right heart failure. In rare cases, the excessive pulmonary blood flow leads to increased left ventricular preload and may result in left ventricular heart failure. Additionally, the fetal pulmonary volume overload can lead to endothelial damage, tunica media hypertrophy, and intima proliferation with irreversible structural pulmonary vascular disease and pulmonary hypertension. Either and/or both of these pathophysiological pathways contribute to a high morbidity and mortality.[3,4]

Idiopathic premature ductal closure is even more rare with only a handful known cases until now. Association with structural cardiac defects such as tetralogy of Fallot or truncus arteriosus communis had been observed in some of them.[5]

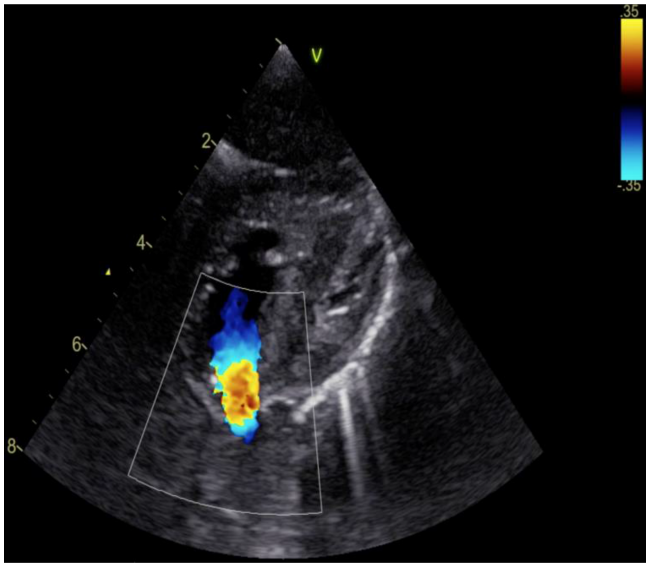

Figure 1. Subcostal view; right ventricular hypertrophy with deviation of the interventricular septum to the left, antegrade flow through the pulmonary valve could be demonstrated.

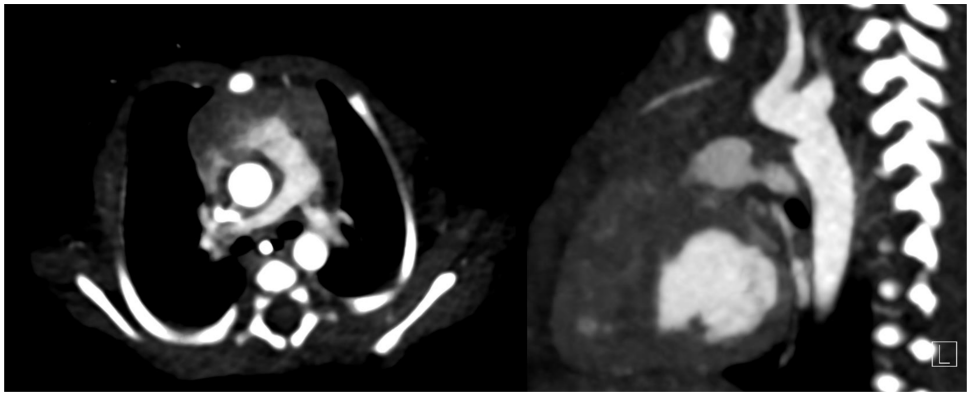

Figure 2. CT shows a 5,8 x 5,7 mm hypoechogenic formation with obstruction of the left pulmonary artery. Due to the proximity to the left laryngeal nerve and the presence of left vocal cord paresis, compression of the nerve was suspected.

In contrast, ductal arteriosus aneurysm has been observed more frequently with an incidence of 8.8%. Of these 98% regress spontaneously within the first 35 days of life. During this regression thrombus formation is frequently seen.[6]

Nonetheless, ductal arteriosus aneurysm is associated with a number of severe, potentially life-threatening sequelae. Spontaneous rupture, infections, thromboembolism, or compression of neighbouring structures (laryngeal nerve and bronchus) have been reported, in which surgical resection should be prompted.[7]

We report on a case of a 31-year old woman, gravida 3, partum 2 who was referred at gestational week 40 day 5 to our hospital. Routine fetal echocardiography had revealed right heart failure (as seen in Supplementary Material S1) and pseudoatresia of the pulmonary valve and absent flow through an arterial duct (Supplementary Material S2), which prompted an immediate caesarean section. A special diet and/or intake of drugs during any point of the pregnancy were negated. Previous check-ups, lastly performed 2 weeks before, had not shown any abnormalities.

Diagnostics and treatment

Postnatally the infant showed severe respiratory distress and respiratory acidosis. Echocardiography revealed massive hypertrophy and dysfunction of the right ventricle. Pulmonary valve had normal size, and opening with antegrade forward flow could be demonstrated as shown in Figure 1 and Supplementary Material S3.

However, we failed to show flow through the arterial duct, so premature ductal constriction was suspected. Exact estimation of pressure in the pulmonary circuit by echocardiography was not possible due to any regurgitation of the tricuspid or pulmonary valve (Supplementary Material S4). Nonetheless, Doppler flow profile with shortened acceleration time as well as R/L shunt via the foramen ovale indicated high pulmonary pressure/resistance.

Endotracheal intubation as well as inotropes were deemed necessary. Within the next days, heart function improved significantly, but a first attempt of extubation failed due to the presence of marked inspiratory stridor. Laryngoscopy revealed left-sided vocal cord paresis and a CT scan of the chest verified what has been suspected by echo: considerable stenosis of the left main pulmonary artery induced by a thrombus formation within an aneurysmatic arterial duct (Fig. 2). Consequently, heart surgery was planned to remove the thrombus, resect the aneurysm, and enlarge the left main pulmonary artery.

Results

Dissection of the arterial duct confirmed a 5,8 x 5,7 mm thrombus within the ductal arteriosus aneurysm. The compressed left main pulmonary artery at the level of insertion of the arterial duct was augmented with autologous pericardium. Post-operatively, unobstructed laminar blood flow in the left and right pulmonary artery could be demonstrated. Right ventricular function resolved slowly. However, hoarseness was ameliorated but enhanced breathing effort and stridor still present. It must be assumed that the ductal arteriosus aneurysm was causative for the paresis due to the proximity of the laryngeal nerve to the arterial duct.

Further follow-up was uneventful with no restenosis of the left pulmonary artery and restoration of cardiac function as well as normalisation of right ventricle hypertrophy.

Discussion

Despite repetitive fetal echocardiographic investigations, lastly performed 4 days before hospital admission, cardiac dysfunction could not be shown until gestational week 40 day 5. Therefore, it must be assumed that the arterial duct closed rapidly in-between those last two check-ups. Additionally, missing signs indicating heart failure like tricuspid and/or pulmonary regurgitation and right ventricular dilatation make a late and rapid process very likely.

Inspiratory stridor secondary to ductal arteriosus aneurysm although rare has been reported before. Walker et al. described a similar case with postnatal stridor and left as well as partial right vocal cord paresis.[8] Interestingly, they decided for tracheotomy and avoided surgery not to harm the laryngeal nerve even more by the procedure. The ductal arteriosus aneurysm as well as the paresis resolved spontaneously within 2 months. In our case, the additional obstruction of the left main pulmonary artery required enlargement, so surgery was deemed necessary. Ligation of the arterial duct and dissection of the ductal arteriosus aneurysm at the same time prevent secondary rupture of the ductal arteriosus aneurysm, a hazardous complication of ductal arteriosus aneurysm.

We did not include thrombophilia as a probable cause of thrombus formation in this specific child. Altered haemodynamics and blood rheology, presence of central venous lines, normal antithrombin III levels as well as a potentially physiological form of ductal closure were likely contributory factors so that specific diagnostics were not indicated.

Special attention deserves the coincidence of both pathologies. We would not have suspected ductal arteriosus aneurysm formation after premature ductus closure, and to the best of our knowledge, this is the first case which reports a combination of

https://doi.org/10.1017/S1047951123003359 Published online by Cambridge University Press

both. Right heart failure of the fetus can be induced by prenatal closure of the arterial duct and must be considered. Although rare, unexplained hoarseness in a newborn should include echocardiographic assessment of the arterial duct.

Conclusion

The arterial duct with its unique responsiveness to prostaglandins and oxygen impacts on haemodynamics both pre- and postnatally. Premature closure of the arterial duct can lead to impaired right ventricle function before birth, and postnatal sequelae might need surgical intervention. Timely delivery is of utmost importance, and regular postnatal check-ups are mandatory.

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S1047951123003359.

Acknowledgements. None.

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

Competing interests. None.

Ethical statement. Due to the nature of the article, a specific ethics approval was not required. However, written consent for publication was obtained from the parents.

References

1. Rasanen J, Wood DC, Weiner S, Ludomirski A, Huhta JC. Role of the pulmonary circulation in the distribution of human fetal cardiac output during the second half of pregnancy. Circulation 1996; 94: 1068–1073.
2. Okada S, Muneuchi J, Iwaya Y. Dilated cardiomyopathy due to premature ductus arteriosus constriction. Cardiol Young 2018; 28: p1172–1174.
3. Kondo T, Kitazawa R, Noda-Maeda N, Kitazawa S. Fetal hydrops associated with spontaneous premature closure of ductus arteriosus. Pathol Int 2006; 56: p554–7.
4. Soslow JH, Friedberg MK, Silverman NH. Idiopathic premature closure of the ductus arteriosus: an indication for early delivery. Echocardiography 2008; 25: 650–652.
5. Genovesea F, Marilli I, Benintende G, et al. Diagnosis and management of fetal ductus arteriosus constriction-closure. J Neonatal Perinatal Med 2015; 8: 57–62.
6. Jan S-L, Hwang B, Y-C Fu, Chai J-W, Chi C-S. Isolated neonatal ductus arteriosus aneurysm. J Am Coll Cardiol 2002; 39: 342–347.
7. Weichert J, Hartge DR, Axt-Fliedner R. The fetal ductus arteriosus and its abnormalities—a review. Congenit Heart Dis 2010; 5: 398–408.
8. Walker JC, Dikkers R, Halmos GB, Berger RMF, du Marchie Sarvaas GJ. Ductus arteriosus aneurysm and vocal cord paralysis. Circulation 2015; 12: 131–134.