An exceedingly rare cause of neonatal shock: persistent fifth aortic arch with critical coarctation

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Abstract

We present a case of a 13-day-old neonate with shock who was diagnosed with persistent fifth aortic with critical coarctation by cardiac point-of-care ultrasound and surgery confirmed the anomalies. This case demonstrates the value of cardiac point-of-care ultrasound in detecting congenital anomalies of great vessels in neonates and may be beneficial in clinical management.

An exceedingly rare cause of neonatal shock:

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Keywords

Neonatal shock; Aorta; Persistent fifth aortic arch; Ultrasound

Introduction

Neonatal shock is a life-threatening condition. Timely diagnosis of the cause of the shock and management of the hemodynamic disturbance can be life-saving in the emergency department (ED)[1,2].We present a case of a 13-day-old neonate with shock who was diagnosed with persistent fifth aortic arch (PFAA) type B with critical coarctation in the ED by cardiac point-of-care ultrasound (POCUS).

Case presentation

A 13-day-old female neonate was referred to the ED with symptoms and signs of shock, including drowsiness, tachypnea, and cold, wet, and cyanotic extremities. Physical examination revealed a pulse rate of 161/min, respiratory rate of 51/min, and blood pressure of 82/47 mmHg. The neonate had a history of assisted reproductive technology. POCUS was performed immediately. Notably, generalized cardiac enlargement was observed (Fig. 1A, videos 1 and 2), with a left ventricular ejection fraction of 24.6% and a right ventricular fractional area change of 29.8%. Severe pulmonary hypertension was revealed, with significant valve insufficiency. In addition, an atrial septal defect with bidirectional shunting (Fig. 1B) and patent ductus arteriosus almost closed were found (Fig. 1C). Furthermore, two aortic arches were detected in the high-parasternal and suprasternal notch view: the upper arch was interrupted distal to the origin of the subclavian artery, while the lower arch was connected to the descending aorta with coarctation (Fig. 2). The neonate was diagnosed with PFAA type B with critical coarctation. Computed tomography angiography and three-dimensional reconstruction confirmed the congenital abnormalities of the aortic arch (Fig. 3) with the narrowest diameter of the fifth aortic arch measuring 0.9 mm. After emergency surgical repairs, cardiac POCUS showed normal aortic flow velocity (Fig. 4). The neonate was discharged 24 days postoperatively with normal cardiac function.

Discussion

The symptoms and treatment of neonatal shock vary significantly with the causes of the condition. The most common etiological factors underlying neonatal shock include hypovolemia, myocardial dysfunction, vasodilation, and acute mechanical blockage of blood flow, such as aortic coarctation (CoA).[1,2] However, PFAA is a rare congenital great artery variant that can cause neonatal shock. It is generated from failed degeneration of embryonic fifth pharyngeal arches[3] and is frequently associated with CoA or IAA, which could cause critical hemodynamic disorder [4-6]. Without prompt medical intervention, neonates are likely to succumb to multiple organ failure and death[7]. Therefore, early recognition and appropriate treatment are crucial[1].

PFAA can be classified as type A, B, and C [4-6]. The severity of PFAA type B is dependent on the extent of CoA. Critical coarctation in our case has not only decreased blood flow to the lower extremities, but also increased the afterload of neonatal immature myocardium and further led to biventricular dysfunction. However, there are many other causes, such as various cardiomyopathies, incessant arrhythmias, and viral myocarditis, that can result in poor myocardial function and thus have been mistaken as the etiology of neonatal shock.[2,8] Therefore, PFAA with significant hemodynamic disturbance needs to be identified early because its prognosis depends heavily on the correct treatment before severe outcome[9].

Routine physical examinations are limited to seeking the causal factors underlying neonatal shock while POCUS can be deemed as an adjunct[10]. POCUS appears to play a more important role among neonates and children, where other monitoring techniques may not be available, but POCUS imaging can be technically easier by reducing the need for radiation and monitoring progress repeatedly in a cost-effective manner. In addition, neonates' and children's lower lung volumes, fewer chronic diseases and thinner chest walls guarantee better image quality for interpretation[11]. Furthermore, the high-parasternal and suprasternal notch view can serve to rapidly evaluate large vessel diseases that will cause hemodynamic instability, such as PFAA, IAA and CoA[12]. Scanning from the two views helped us demonstrate the spatial relationship between these two almost parallel aortic arches, clarify the scheme of vascular branching and identify the PFAA with CoA. Therefore, integrating the high-parasternal and suprasternal notch view into routine cardiac POCUS can significantly benefit vulnerable neonates and children.

Except for POCUS as a typical screening and monitoring tool, computed tomography angiography and magnetic resonance angiography can be used to confirm the abnormalities and assist with surgical planning, because they "lights up" blood vessels, and three-dimensional reconstruction can directly display the branching patterns and entire course of the aorta.[13-15] Prompt causal treatment of the obstruction could achieve good outcomes.

Conclusion

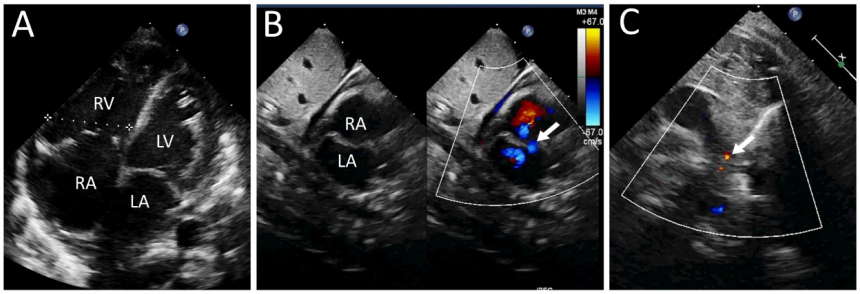
This case highlights the application of POCUS in the swift diagnosis of congenital anomalies underlying neonatal shock for the first time and emphasizes the role of the high-parasternal and suprasternal notch view. Cardiac POCUS used in critical patients can lead to rapid and well-aimed therapy. For the pediatric population, congenital anomalies are a concern. When performing cardiac POCUS, great vessels should also be evaluated in the high-parasternal and suprasternal notch view.

Fig.1 A: Apical 4-chamber view of the enlarged heart. B: Subcostal view focused interatrial septum showing atrial septal defect, white arrow indicted right-to-left shunt. C: Parasternal short-axis focus on pulmonary artery showed patent ductus arteriosus nearly closed.

Fig. 2 A: Cardiac point-of-care ultrasound parasternal long-axis view of aortic arch in suprasternal fossa depicting persistent fifth aortic arch type B. The upper arch was the (fourth) aortic arch (white asterisks) interrupted, the lower arch was persistent fifth aortic (yellow asterisks) with critical coarctation at the origin of the ductus arteriosus (white arrow). B: Continuous wave Doppler echocardiography demonstrated that the peak flow velocity through the narrowest point of the descending aorta was 3.1m/s and pressure was 37 mmHg.

Fig. 3 Computed tomography angiography and three-dimensional reconstruction showing the upper (fourth) aortic arch (white asterisks) interrupted and the lower persistent fifth aortic arch (yellow asterisks) with critical coarctation at the origin of the ductus arteriosus (white arrow).

Fig. 4 A: Postoperative cardiac point-of-care ultrasound showed that the lower persistent fifth aortic arch was broadened, with the narrowest measuring 6.5mm. Color Doppler echocardiography showed patent arterial flow. B: Postoperative continuous wave Doppler echocardiography showed normal descending aorta flow velocity.



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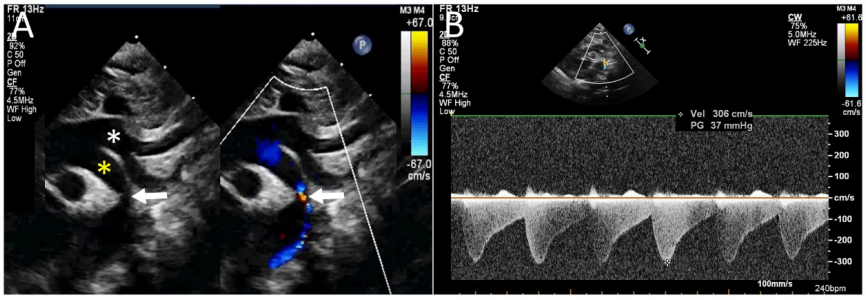

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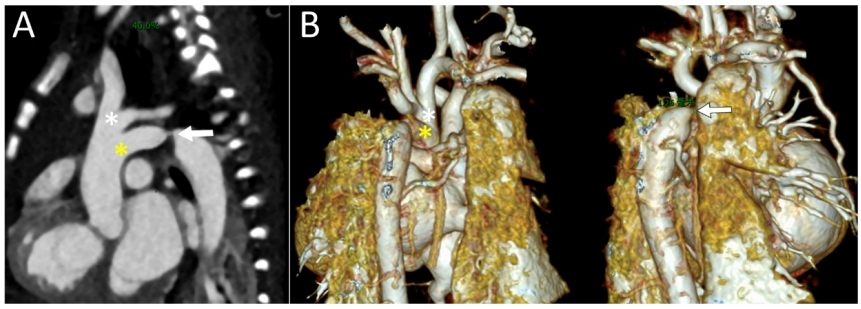

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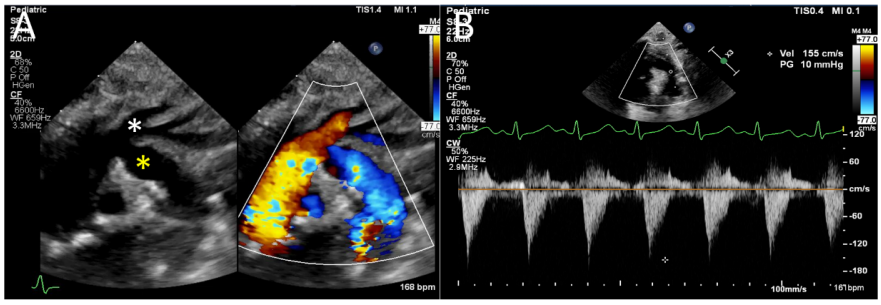

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References

1. Bachur RG, Shaw KN, Chamberlain J, Lavelle J, Nagler J, Shook J.*Fleisher & Ludwig's Textbook of Pediatric Emergency Medicine*Wolters Kluwer Health; 2020.

2. Charles S. Kleinman, Seri I. *Hemodynamics and cardiology : neonatology questions and controversies.* 2 ed2012.

3. Liu Y, Zhang H, Ren J, et al. Persistent fifth aortic arch: a single-center experience, case series. *Transl Pediatr.*2021;10(6):1566-1572.

4. Weinberg PM. Aortic arch anomalies. *J Cardiovasc Magn Reson.*2006;8(4):633-643.

5. Kusano N, Marutani S, Masumi H, et al. A Case of Persistent Fifth Aortic Arch with an Interrupted Fourth Aortic Arch. *Ann Thorac Surg.* 2021.

6. Zhao YH, Su ZK, Liu JF, Cao DF, Ding WX. Surgical treatment of persistent fifth aortic arch associated with interrupted aortic arch. *Ann Thorac Surg.* 2007;84(3):1016-1019.

7. Standl T, Annecke T, Cascorbi I, Heller AR, Sabashnikov A, Teske W. The Nomenclature, Definition and Distinction of Types of Shock. *Dtsch Arztebl Int.* 2018;115(45):757-768.

8. Rowland DG, Gutgesell HP. Noninvasive assessment of myocardial contractility, preload, and afterload in healthy newborn infants. *Am J Cardiol.* 1995;75(12):818-821.

9. Zotzmann V, Rottmann FA, Müller-Pelzer K, Bode C, Wengenmayer T, Staudacher DL. Obstructive Shock, from Diagnosis to Treatment. *RCM.* 2022;23(7).

10. Perera P, Mailhot T, Riley D, Mandavia D. The RUSH exam: Rapid Ultrasound in SHock in the evaluation of the critically lll. *Emerg Med Clin North Am.* 2010;28(1):29-56, vii.

11. Singh Y, Tissot C, Fraga MV, et al. International evidence-based guidelines on Point of Care Ultrasound (POCUS) for critically ill neonates and children issued by the POCUS Working Group of the European Society of Paediatric and Neonatal Intensive Care (ESPNIC). *Crit Care.* 2020;24(1):65.

12. Wang RN, Escobar M, North S, Lin J. Point-of-care Ultrasound Diagnosis of Type B Aortic Dissection on the Suprasternal Notch View. *Cureus.* 2019;11(10):e6005.

13. Goldstein SA, Evangelista A, Abbara S, et al. Multimodality imaging of diseases of the thoracic aorta in adults: from the American Society of Echocardiography and the European Association of Cardiovascular Imaging: endorsed by the Society of Cardiovascular Computed Tomography and Society for Cardiovascular Magnetic Resonance. *J Am Soc Echocardiogr.* 2015;28(2):119-182.

14. Doshi AR, Chikkabyrappa S. Coarctation of Aorta in Children. *Cureus.* 2018;10(12):e3690.

15. Moes CA, Freedom RM. Rare types of aortic arch anomalies. *Pediatr Cardiol.* 1993;14(2):93-101.