


# Berry syndrome: a rare cause of cardiac failure in the early neonatal period

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## DESCRIPTION

The authors describe the case of a newborn who, at 24 hours of age, presented with respiratory distress, differential upper/lower limb oxygen saturations, cyanosis and filiform femoral pulses. He was started on prostaglandin E1 perfusion and was transferred to our centre.

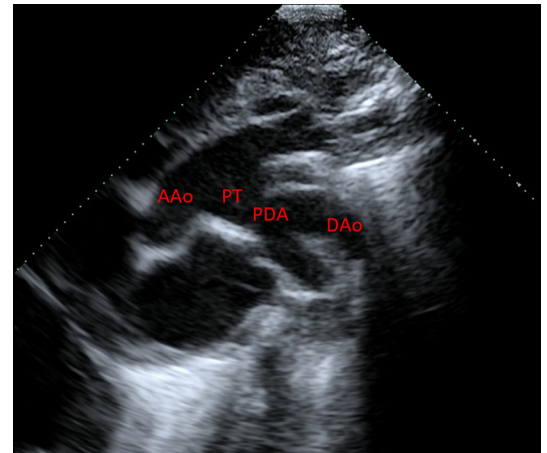
He had signs of lung congestion and required mechanical ventilation, improving clinically.

The chest X-ray showed asymmetrical bilateral pulmonary congestion, more pronounced on the right hemithorax (**figure 1**).

The more salient features on the echocardiogram included a type A interrupted aortic arch (IAA), a distal (type 2) aortopulmonary window (APW), the right pulmonary artery (RPA) arising from the aorta and the descending aorta continuous with the arterial duct (**figures 2 and 3**). The interventricular septum was intact. Angio-CT confirmed these findings (**figure 4**).

These features were compatible with Berry syndrome, a rare congenital duct dependent anomaly, with less than 100 cases described.<sup>1</sup> As shown in our case, it consists of an IAA, an APW, the RPA originating from the ascending aorta and an intact interventricular septum. The APW in often of the distal type (either type 2 or 3), which results in the pulmonary branches being widely separated and the IAA is often distal to the emergence of the left subclavian artery (type A).<sup>1</sup>

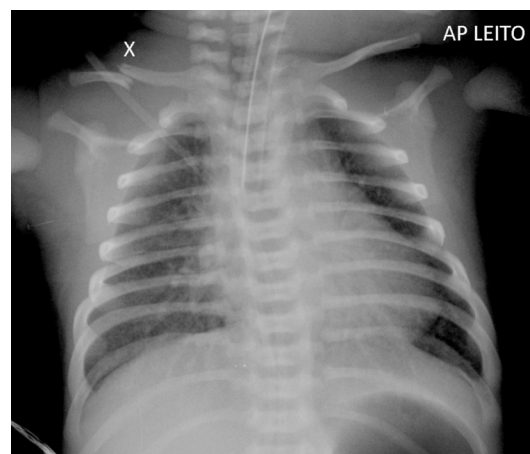
Newborns with Berry syndrome are usually critically ill. The aortic origin of the RPA results in early pulmonary overcirculation due to an obligatory left-to-right shunt. Simultaneously, the perfusion of the lower body organs is entirely dependent on a



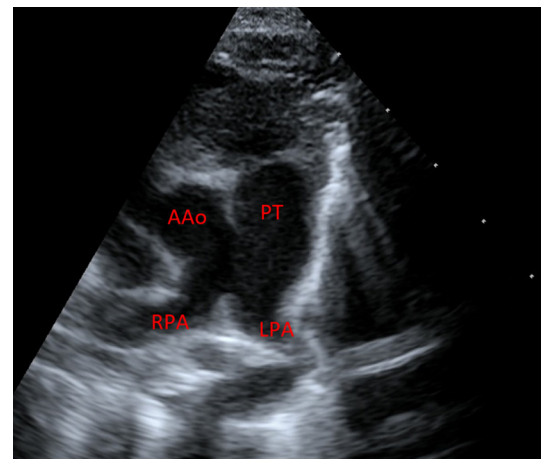
**Figure 2** Transthoracic echocardiogram (suprasternal view) showing a type A interrupted aortic arch and an aortopulmonary window. AAO, ascending aorta; DAA, descending aorta; PDA, patent ductus arteriosus; PT, pulmonary trunk.

patent ductus arteriosus due to an IAA.<sup>2</sup> Early clinical management is pivotal and includes diuretics and positive pressure ventilation to address the pulmonary congestion, as well as, when necessary, maintaining the arterial ductus patent with prostaglandin E1 infusion.

Despite optimal medical stabilisation, most require urgent surgery. Staged repair, consisting



**Figure 1** Anteroposterior (AP) chest X-ray showing asymmetrical pulmonary congestion.

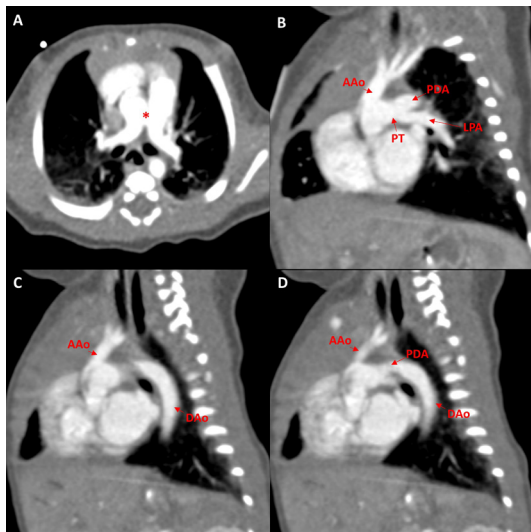


**Figure 3** Transthoracic echocardiogram (parasternal short-axis view) showing an aortopulmonary window with the right pulmonary artery (RPA) originating from the ascending aorta (AAO) and the left pulmonary artery (LPA) originating from the pulmonary trunk (PT), forming a 'butterfly sign'.



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**Figure 4** Angio-CT images. (A) Axial view reproducing the echo parasternal short axis view shown in figure 3, butterfly sign is also visible \*. (B, C and D) Oblique views, showing a type A interrupted aortic arch and aortopulmonary window. AAO, ascending aorta; DAAo, descending aorta; LPA, left pulmonary artery; PDA, patent ductus arteriosus; PT, pulmonary trunk.

of pulmonary banding followed by a later surgical correction, can be considered, particularly in premature or small-for-gestational-age infants.<sup>3</sup> However, the treatment of choice is single stage repair. Various surgical techniques are available, namely, APW closure and RPA connection with the main pulmonary artery can be done by either direct anastomosis or by several flap techniques.<sup>4–7</sup> Management of the IAA depends on its location and distance of both aortic ends. It can be carried out using various techniques such as end-to-end anastomosis or an extensive mobilisation and reconstruction of the aorta, requiring patch augmentation or conduit interposition.<sup>8</sup>

Berry syndrome, despite being rare, should be considered in the differential diagnosis of critically ill neonates presenting with severe coarctation presentation phenotype and signs of asymmetric pulmonary congestion.

## Learning points

- ▶ Berry syndrome is a potential cause of heart failure in neonates.
- ▶ Asymmetrical pulmonary congestion and the ‘butterfly sign’ should raise clinical suspicion about this entity.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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