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Brief Report

Anomalous connection of the right superior caval vein to the morphologically left atrium

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Abstract Anomalous drainage of the right superior caval vein into the morphologically left atrium as an isolated cardiac malformation is a rare anomaly. Most patients present with cyanosis. Thus far, about 20 cases have been reported in the literature. We report a case of cyanosis due to this malformation in a male neonate, which was complicated by the meconium aspiration syndrome. The malformation was diagnosed by echocardiography and cardiac catheterization. Surgery resulted in complete recovery.

Keywords: Anomalous systemic venous drainage; cyanosis; cardiac surgery

ANOMALOUS DRAINAGE OF THE RIGHT SUPERIOR caval vein into the morphologically left atrium is exceedingly rare as an isolated anomaly. Thus far, as far as we know, only about 20 cases have been reported in the literature.[1] Most patients with this lesion present with cyanosis at a young age. We report a neonate with this malformation who was diagnosed appropriately and corrected with surgical management.

Case report

A male neonate born at term presented with cyanosis and respiratory failure after delivery. The boy was intubated and transferred to a neonatal intensive care unit. The initial chest X-ray showed diffuse opacities consistent with the meconium aspiration syndrome. On initial transthoracic echocardiography, there were signs of right ventricular hypertrophy, and the pulmonary arteries seemed to be small in size. No specific congenital cardiac malformation could be detected. Analysis of blood gases disclosed a low arterial partial pressure of oxygen, but a normal arterial partial pressure of carbon dioxide. Since there was no improvement in his condition, he was referred to another clinic with the intention for treatment with extracorporeal membrane oxygenation. Contrast echocardiography showed filling of the left atrium with micro-bubbles after an injection into a cephalic vein. Cardiac catheterization was performed to rule out additional malformations. The catheter was advanced into the right superior caval vein after passage through the right inferior caval vein, the right atrium, a patent oval foramen, and the morphologically left atrium (Fig. 1a and b). Angiography into the right superior caval vein then demonstrated its origin from the morphologically left atrium. The inferior caval vein drained into the right atrium (Fig. 1c and d). Echocardiography confirmed presence of the antero superior part of the atrial septum and the terminal crest, and also confirmed the origin of the superior caval venous orifice from the left atrium, showing a clear separation from the right pulmonary veins (Fig. 2). There was no groove in the left atrial roof between the orifice of the superior caval vein and the right pulmonary veins. The right atrial appendage was anterior to the superior caval orifice, and clearly separated from it.

On surgery, the wall that partitioned the superior caval vein into the left atrium was felt to contain fibrous tissue. This structure, however, was not considered to represent a persistent and enlarged Eustachian valve. Surgical correction was performed by translocation of the superior caval vein into the right atrium, closure of the original site of drainage at the

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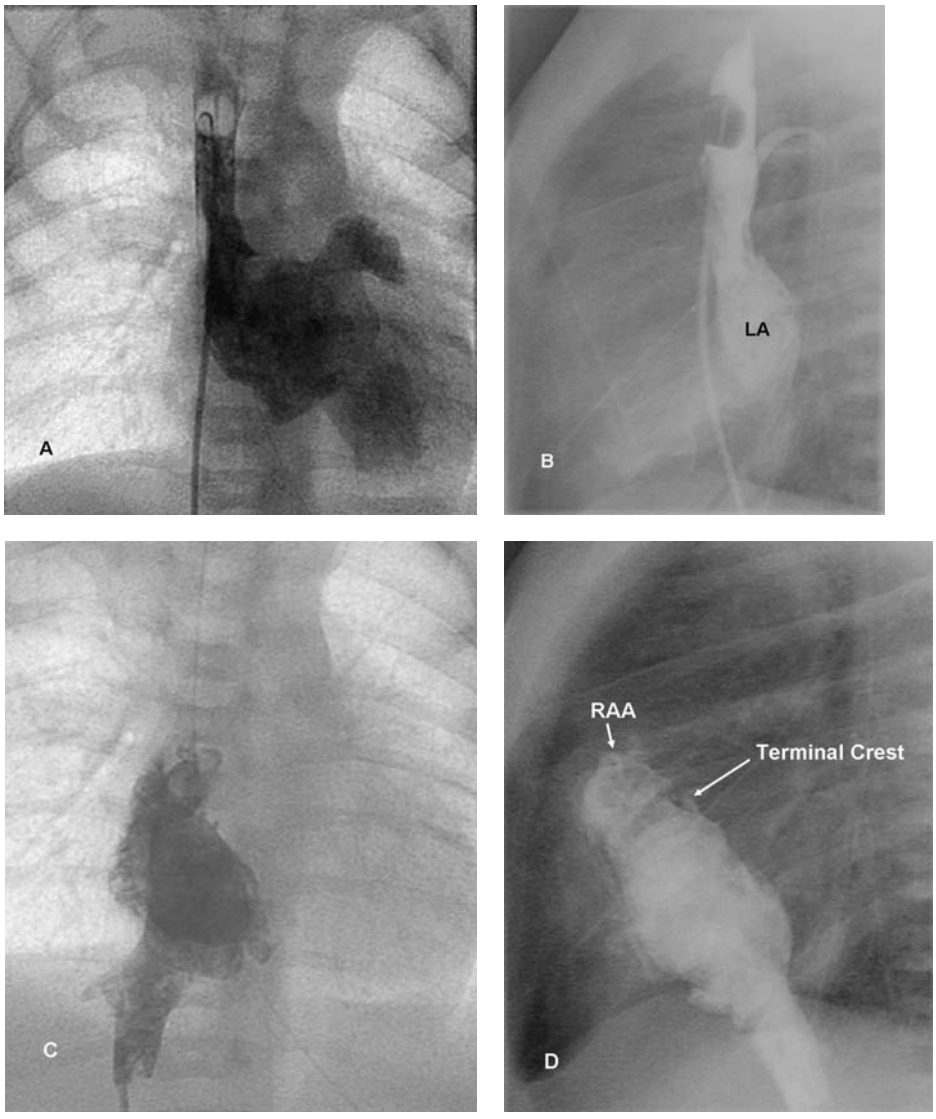

Figure 1.
*The right superior caval vein is connected unequivocally to the morphologically left atrium (LA), as shown in frontal (**a**) and lateral (**b**) projections. Injections in the right atrium (**c**, **d**) show the typical morphology of the right appendage (RAA), with the terminal crest, and show the inferior caval vein draining to this atrium.*

roof of the left atrium, and closure of the patent oval foramen. The further course and follow-up three months after discharge were uneventful.

Discussion

Most patients presenting with anomalous drainage of the right superior caval vein into the morphologically left atrium have been reported to be asymptomatic, presenting only with cyanosis.[2] In some cases published thus far, the diagnosis was made later in life,[3] but most patients were diagnosed as infants.[1,4,5] The precise aetiology of this malformation has not yet been clarified. Some authors hypothesized the leftward and cephalic malposition of the right horn of the systemic venous sinus[6] or, less likely, an abnormality in the evolution of the systemic venous sinus.[7] Alternatively, a persistence of the upper part of the Eustachian valve could plaster itself on the septum, leading to drainage of the superior caval vein into the left atrium. In our case, the surgeons were unable to find any part of the Eustachian valve within the left atrium, although the wall that partitioned the superior caval vein into the left atrium was felt to contain fibrous tissue.

Surgical correction is indicated in all cases to avoid the sequels of cyanosis and paradoxical embolisation to the brain. Contrast echocardiography usually allows non-invasive diagnosis. [2,5] Contrast echocardiography was not performed initially in our patient because of

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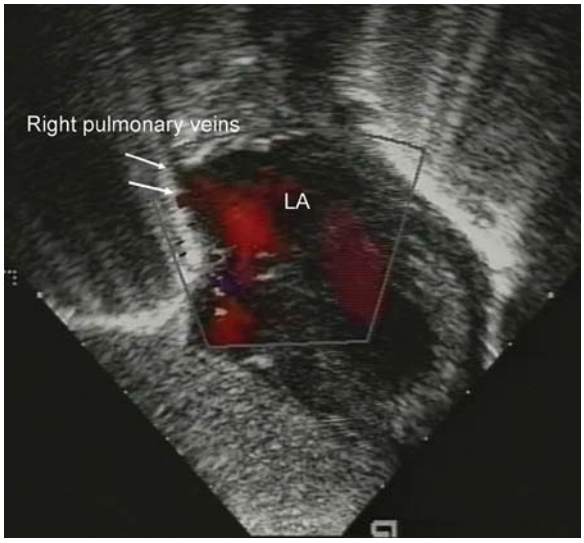

Figure 2.
The subcostal echocardiographic image shows the right pulmonary veins draining into the left atrium (LA), with a left-to-right shunt through the persistent oval foramen.

the presence of both clinical and radiographic signs of the meconium aspiration syndrome. After resolution of respiratory findings, nonetheless, the cyanosis persisted and necessitated further diagnostic procedures. Our case shows that contrast echocardiography should be performed in any neonate presenting with cyanosis that cannot be explained conclusively by other findings. Surgery led to complete recovery, and is the treatment of choice for this condition.[4]

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