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BRIEF COMMUNICATION

Prenatal diagnosis of anomalous connection of the inferior caval vein to the left atrium associated with common arterial trunk

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Abstract

Anomalous connection of the inferior caval vein to the left atrium is exceedingly rare, and has even been considered by some authors an anatomic and embryologic impossibility. This study demonstrates for the first time the existence of this rare malformation, diagnosed on prenatal echo, and confirmed on post-mortem examination in a 24 WG fetus, in association with a common arterial trunk.

KEYWORDS
anomalous connection, common arterial trunk, inferior caval vein, left atrium, prenatal diagnosis

1 | INTRODUCTION

Anomalous connection of the inferior caval vein to the left atrium is exceedingly rare. The majority of cases reported in the literature actually correspond to an anomalous drainage of the inferior caval vein to the left atrium due to an abnormal Eustachian valve. The commonest associated anomaly is ostium secundum atrial septal defect, but associated anomalous pulmonary venous return (Lamb et al., 1987; Sanchez & Human, 1986), multiple pulmonary arteriovenous fistulas (Black et al., 1964), and tetralogy of Fallot (Sanchez & Human, 1986) have also been reported.

We describe here a fetus in whom anomalous connection of inferior caval vein to left atrium was diagnosed on antenatal echocardiography at 23 WG, in association with common arterial trunk, and confirmed at autopsy after termination of pregnancy at 24 WG. As far as we are aware, this combination has never been described.

2 | CASE REPORT

A 32-year-old woman was referred for expert fetal echocardiography at 23 WG because of fetal hydrops and complex cardiac defect diagnosed on routine fetal ultrasound. She had a history of dysplastic aortic valve with aortic insufficiency, mild pulmonary valvular stenosis, and spontaneously closed ventricular septal defect (VSD), associated with mild facial dysmorphic features. Fetal ultrasound confirmed fetal hydrops with ascites and right pleural effusion. The heart was in levocardia, with a small right superior caval vein draining into the right atrium and a normal pulmonary venous return to the left atrium. There was an inverted (left-to-right) shunt through the oval foramen (Figure 1). The inferior caval vein bordered by the Eustachian valve was dilated and directly connected to the left atrium, to the left of the atrial septum (Figure 1). In addition, both ventricles experienced severe systolic dysfunction. A common arterial trunk was located above a large outlet VSD with a

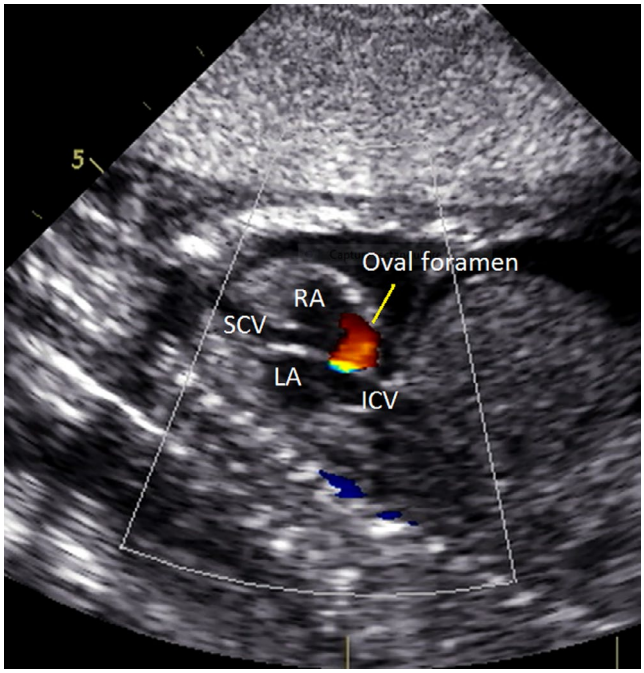

FIGURE 1 Fetal echocardiography, subcostal coronal view. The superior caval vein is connected to the right atrium, the inferior caval vein is connected to the left atrium. The oval foramen is shunting left to right. ICV, inferior caval vein; LA, left atrium; RA, right atrium; SCV, superior caval vein

dysplastic and stenotic truncal valve. Because of the complexity of the cardiac anomaly and the severe biventricular dysfunction, termination of pregnancy was chosen by the parents and performed at 24 WG.

At post-mortem examination, the disposition of intra-abdominal organs was normal. The inferior caval vein in all its segments was located to the right of the spine, and all the hepatic veins drained into the inferior caval vein. The portal vein was in normal position and the ductus venosus was patent. The lungs were normally lobated with normal bronchi, and the heart was in levocardia with normal segmental combinations. The atrial appendages were morphologically normal, with pectinate muscles extending to the crux in the right-sided atrium, and a smooth-walled left atrium. The right atrium received the right superior caval vein and the coronary sinus orifice bordered by a thebesian valve (Figure 2a). The orifice of the inferior caval vein was absent in the right atrium (Figure 2a). The oval foramen was patent with septum secundum above but there was no septum primum within the oval fossa (Figure 2a). The left atrium received the four pulmonary veins. The inferior caval vein drained directly into the left atrium to the left of the interatrial septum (Figure 2b). The inferior caval vein orifice was bordered by a well-developed Eustachian valve, located mostly to the left of the inferior caval vein with some attachments on the left interatrial septal surface (Figure 2b). The two ventricles were well developed. There was a common arterial trunk, type 1 of Collet-Edwards and Van Praagh classifications (aortic dominance with the orifices of the right and left pulmonary arteries originating from the left and posterior side of the common trunk, at the margins of the pericardial cavity) above a large outlet VSD (Figure 3) (Collett & Edwards, 1949; Van Praagh, 1987). The four-leaflet truncal valve was dysplastic. There was a short mitro-truncal discontinuity. The two coronary orifices were slit like and stenotic, the left one above a commissure and the right one below the sinotubular junction (Figure 3).

3 | DISCUSSION

Among the anomalies of the inferior systemic venous return, the rarest form is abnormal connection of the inferior caval vein to the morphologically left atrium. It is essential to make the distinction between connection and drainage, as true connection of the

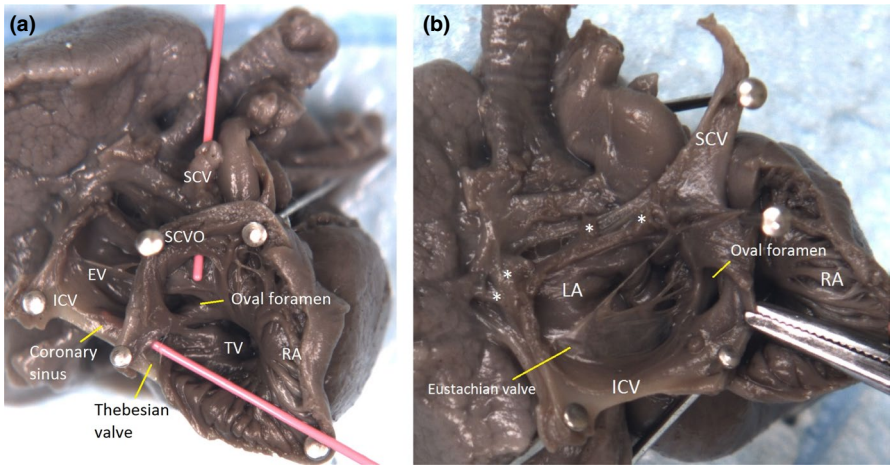

FIGURE 2 (a) Heart specimen, view of the right atrium. One probe passes through the superior caval vein, the other one is inserted in the coronary sinus. EV, Eustachian valve; ICV, inferior caval vein; RA, right atrium; SCV, superior caval vein; SCVO, superior caval vein orifice; TV, tricuspid valve. (b) Heart specimen, view of the left atrium. The four pulmonary veins are marked by asterisks. ICV, inferior caval vein; LA, left atrium; RA, right atrium; SCV, superior caval vein

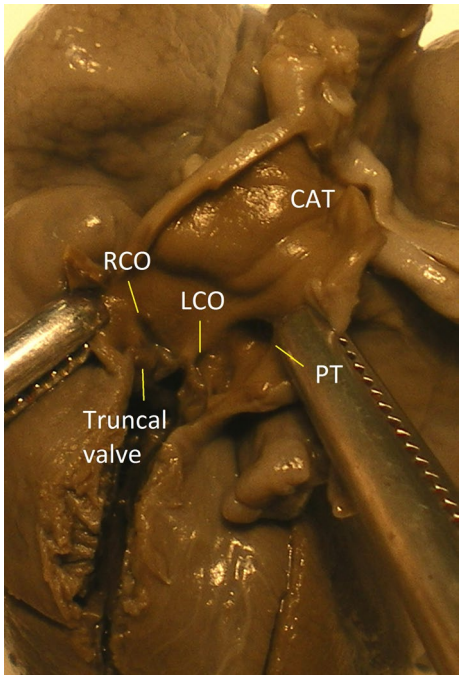

FIGURE 3 Heart specimen, view of the common arterial trunk. The two coronary orifices are slit-like and stenotic. CAT, common arterial trunk; LCO, left coronary orifice; PT, pulmonary trunk; RCO, right coronary orifice

inferior caval vein to left atrium seems to be exceedingly rare, and was considered impossible for some authors from an embryologic and anatomical standpoint (Van Praagh & Van Praagh, 1987). The embryologic development of the inferior caval vein is complex, each segment resulting from a different embryonic systemic vein. The supra-hepatic segment derives from the right vitelline vein (Oliveira & Martins, 2019). However, if most anomalies of the inferior caval vein can be explained by abnormal development of the different segments, there is no valuable explanation to date for abnormal connection of a right-sided inferior caval vein to the left-sided morphologically left atrium (Sierig et al., 2005).

Differential diagnoses include inferior sinus venosus defects, atrial situs inversus with inferior caval vein connected to a left sided but morphologically right atrium (Burri et al., 2003), heterotaxy with anomalous systemic vein connections (Miltner et al., 2017). Indeed, atrial situs inversus and heterotaxy might be underdiagnosed in some of the few cases reported, as the extension of the pectinate muscles was rarely, if never, described. Recently described totally anomalous systemic venous connection is a different entity, defined by the anomalous connection to the morphologically left atrium of all systemic veins, including the coronary sinus (Gupta et al. 2017, Zhang et al., 2009); this anomaly could be reinterpreted as total anomalous pulmonary venous connection in a left-sided right atrium because of the presence of the opening of the coronary sinus, and thus as situs inversus atria. However, if it was the case, the atrioventricular connections would be discordant, yet they were reported as concordant in the few cases published of this exceptional anomaly (Gupta et al. 2017, Zhang et al., 2009).

The most frequent differential diagnosis remains anomalous drainage of inferior caval vein to left atrium, and most of the reported cases could be indeed anomalous drainage rather than anomalous connection. Anomalous drainage is due to an abnormally developed Eustachian valve which fuses with the right edge of the superior interatrial fold (septum secundum) above the oval foramen, redirecting the venous flow from the inferior caval vein to the left atrium through the atrial septum, as demonstrated in some case reports (Genoni et al., 1999; Kogon et al., 2006; Sierig et al., 2005). In that case, the Eustachian valve remains to the right, and the atrial septum to the left, of the inferior caval vein. By contrast, according to Van Praagh, anomalous connection can be proven only if the inferior caval vein lies to the left of septum primum in atrial situs solitus (Van Praagh & Van Praagh, 1987). Very few cases in the literature could fit with this definition, as only the first case published in 1955 displays photos of an autopsied specimen; however, there is no view of the left atrium and the right atrial view shows only the atrial septal surface (Gardner & Cole, 1955). Figure 2a,b demonstrate that this was indeed the case in our specimen. The inferior caval vein is to the left of the atrial septum and the Eustachian valve is completely within the left-sided left atrium, although to the right of the mitral valve and the arrival of the pulmonary veins. One could argue that the structure that we describe as the Eustachian valve might be actually an abnormally located septum primum and if it was the case, the inferior caval vein would drain within, and not connect to, the morphologically left atrium. However, this fibrous structure borders the orifice of the inferior caval vein on half its circumference, is located to the left side of the atrial septum, and attaches on the left edge of the atrial septum. The authenticity of the atrial septum itself is ensured by three anatomic features: there is a superior interatrial fold below the orifice of the right superior caval vein; the orifice of the coronary sinus opens in usual position below the oval fossa and above the right atrioventricular junction; and pectinate muscles are present only to the right of this structure. If we admit that the inferior caval vein opens in the right atrium with the fibrous structure around its orifice being the septum primum, then there would be no superior interatrial fold and the orifice of the coronary sinus would open between the inferior caval vein orifice and the tricuspid annulus, but would not be separated from it by a muscular wall. In addition, the anterior wall of the coronary sinus is intact and its orifice is not enlarged, ruling out an abnormal connection of the inferior caval vein with the coronary sinus.

The association of abnormal connection or drainage of the inferior caval vein with common arterial trunk was never described in the literature. However, the association of common arterial trunk with abnormal pulmonary venous connections is known, pointing the possible involvement of the two parts of the second heart field in this rare phenotype (Bajolle et al., 2009). The outflow tract at the arterial pole of the heart is derived from the anterior second heart field (Waldo et al., 2005b; Waldo et al., 2005a). The venous

pole of the heart, including the atrial myocardium and the systemic, pulmonary, and cardinal veins, is derived from the posterior second heart field, with different genes expressed in systemic venous sinus compared to pulmonary veins (Sizarov et al., 2010). The connection of the sinus venosus to the right atrium results from asymmetrical growth of the right part of the common atrium, causing a rightward shift of the systemic venous compartment (Douglas et al., 2011; Jensen et al., 2017). The second heart field could therefore be involved in our case, together with a lack of rightward shift of the sinus venosus during atrial development.

4 | CONCLUSION

We demonstrate in a 24 WG fetus the existence of an abnormal connection of the inferior caval vein to the left atrium, both at fetal ultrasound and examination of the cardiac specimen, in association with common arterial trunk. The association between anomalies at both venous and arterial poles of the heart suggests the involvement of the second heart field in the morphogenesis of this rare cardiac phenotype.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

AUTHOR CONTRIBUTIONS

LH drafted the manuscript; LC, PB, and AH contributed to the acquisition of data; and DB reviewed the manuscript for intellectual content.

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