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.

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
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 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 micro-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).

### 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
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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