Fetal Diagnosis in a Unique Case of Vascular and Cardiac Interdependence in Omphaloischiopagus Conjoined Twins

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INTRODUCTION

A woman was referred to our fetal cardiac center for omphaloischiopagus conjoined twins, each with a separate thorax, heart, head, and upper extremities. Fetal echocardiography in twin A showed complete atrioventricular canal defect with aortic and pulmonary atresia and severe atrioventricular valve regurgitation. Twin B had a structurally normal heart. An arterial vessel from twin B’s descending aorta entered twin A and provided retrograde filling of twin A’s aorta. A venous vessel from twin A’s common atrium drained into twin B’s portal venous system. Postnatal computed tomography (CT) angiography confirmed the lack of connection between the cardiac mass and aorta of twin A. There appeared to be complete dependence of twin A on twin B’s cardiac output. Fetal echocardiography was critical in delineating the vascular anatomy of the conjoined twins prenatally, allowing for delivery planning and parental counseling. This constellation of findings is the first of its kind described in the literature, being significantly more complex and more developed than reported cases of parasitic conjoined twins. These findings may provide insight for management strategies of future conjoined twins with complex cardiac interdependence.

CASE PRESENTATION

Clinical Presentation

A 34-year-old gravida 2 para 1 woman was referred to our fetal cardiac center at 31 weeks’ gestation due to the finding of conjoined twins. The twins were suspected to be conjoined at the liver with diaphragm interdependence. These findings may provide insight for management strategies of future conjoined twins with complex cardiac interdependence.

Imaging Findings

Fetal echocardiography demonstrated severe complex congenital heart disease in twin A consisting of a complete atrioventricular canal defect with both aortic atresia and pulmonary atresia. There was no demonstrable pulmonary or aortic outflow from either ventricle. There was severe atrioventricular valve regurgitation and an extremely dilated common atrial chamber (Figures 1A and 2, Video 1). Twin A was severely hydropic with severely diminutive lungs compressed by a massive pericardial effusion occupying the entire chest and extending into twin B (Figure 1C). Twin B was found to have a structurally normal heart without signs of high-output heart failure prenatally.

A large arterial vessel was seen emerging from the abdominal descending aorta of twin B coursing toward twin A’s abdomen and providing retrograde filling of twin A’s descending aorta, aortic arch, and aortic root (Figures 1B and 2, Videos 2 and 3). The aorta of twin A did not appear to connect to the ventricles. Additionally, a tortuous venous structure from twin A’s common atrium was found to drain inferiorly toward twin B and was suspected prenatally to connect into twin B’s portal venous system in the liver (Figure 3, Video 4).

Postnatally, these findings were confirmed by transthoracic echocardiography. Contrast-enhanced CT angiography of the chest, abdomen, and pelvis demonstrated that twin B provided arterial supply for twin A through an arterial vessel, which connected to the thoracic descending aorta of twin A, filling its descending aorta, aortic arch, and aortic root (Figure 4). No significant large arterial branching vessels to the lower body of twin A were seen below the level of this connection. Interestingly, the cardiac mass of twin A did not fill with contrast, confirming the lack of connection between its cardiac mass and aorta.

Role of Imaging in Patient Care

Fetal echocardiography was critical in delineating the venous and arterial anatomy of the conjoined twins, which helped provide appropriate counseling to the parents and a delivery plan for the perinatologist and birth hospital. A multidisciplinary discussion was held prenatally to guide the delivery team and neonatal intensive care unit about the safest course of action. The prenatal multidisciplinary discussion included the perinatologist, fetal cardiologist, pediatric cardiac intensivist, neonatal intensivist, and pediatric surgeon. Given the lack of functional lung tissue and nonviable congenital heart disease in twin A, it was determined that twin B alone would be intubated after birth and monitored for signs of high-output heart failure, as twin B was determined to serve as the sole cardiopulmonary support for the twin pair.
The twins were delivered at 37 weeks of gestation with a pediatric cardiologist present at the delivery. Twin B was intubated at birth, and twin A remained on room air. No inotropes were required after birth, and twin A exhibited independent purposeful movement. The twins were transferred to the neonatal intensive care unit for further imaging. Postnatal transthoracic echocardiography confirmed that the venous vessel from twin A did drain into the portal venous system of twin B, providing some protection as a vascular resistor from high-output heart failure in the immediate postnatal period. In the days after birth, feasibility of separation was deemed too difficult in the early neonatal period due to the complexity of skeletal reconstruction, single gastrointestinal tract and stomach, and their small size. Pediatric surgery consultation reassessed operative candidacy for separation regularly. Twin B was extubated at 2 weeks of age but required reintubation the following day for inefficient ventilation. Twin B underwent weekly serial echocardiograms for surveillance of development of high-output heart failure or pulmonary hypertension. Mild pulmonary hypertension was evident on twin B’s echocardiogram on day 24 of life. Ultimately, twin B developed a severe pulmonary hypertension crisis on day 32 of life, leading to the demise of both twins.

DISCUSSION

This constellation of findings in conjoined twins with cardiac interdependence is the first of its kind described in the literature, being significantly more complex—and in the case of

Figure 1  Echocardiographic findings. (A) Postnatal echocardiogram of twin A demonstrated a severe complete atrioventricular canal defect with severely dilated atria. (B) Postnatal transthoracic echocardiogram of twin A showed retrograde filling of the entire aortic arch with color Doppler, suggesting aortic atresia. (C) Fetal echocardiography demonstrated a severely hydropic twin A with significant absence of lung tissue in the chest cavity and a large pericardial effusion extending into twin B.
Figure 2 Severe atrioventricular valve regurgitation in twin A. Fetal echocardiography showed the presence of severe atrioventricular valve regurgitation in twin A.

Figure 3 Arterial and venous connections in twin A. (A) Fetal echocardiography showed retrograde filling of the entire aorta (dashed arrow) of twin A without connection to the ventricles. (B) A large venous vessel (solid arrow) seen posterior to the cardiac mass appeared to carry venous drainage of twin A into twin B.

Figure 4 CT angiographic reconstruction of conjoined twins. Whole body CT angiography reconstruction of the conjoined twins demonstrated large arterial vessel (arrow) arising from the abdominal descending aorta of twin B to supply the entire aortic arch of twin A without filling of contrast into twin A’s cardiac mass (region encompassed by circle), confirming aortic atresia. As this large arterial vessel coursed superiorly into twin A, there were no major inferiorly directed arterial branches to supply the lower extremities of twin A. Additionally, the absence of lung tissue in twin A and a single gastrointestinal tract and pelvis shared by both twins were noted.
twin A, more developed—than reported cases of parasitic conjoined twins or twin reversed arterial perfusion sequence. Fetal echocardiographic imaging was critical in delineating the complex cardiopulmonary physiology in this unique case of conjoined twins and allowed for accurate counseling and perinatal care planning. The unusual vascular connections were defined accurately by fetal echocardiography in the hands of experienced cardiac sonographers. The rate of congenital heart defects in conjoined twins is high and appears to be an important prognostic factor for survival. As expected, the risk of high-output heart failure and pulmonary hypertension in the donor twin was high, although the timing of postnatal decompensation was difficult to predict prenatally, and even postnatally, given the rarity of these findings. If reconstruction is feasible, early separation of twins in the setting of complex cardiac anomalies seems improve the chance for survival of the twin without congenital heart disease.

**CONCLUSION**

Although the ultimate outcome could not be altered, the findings can provide insight for prenatal management strategies of conjoined twins with complex cardiac interdependence. Conjoined twins should be referred to centers that can provide detailed fetal cardiac assessments to guide counseling regarding prognosis and management.

**SUPPLEMENTARY DATA**

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2021.01.007.

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