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CASE REPORT

Prenatal Diagnosis of Cor Triatriatum Sinister in Association With Hypoplastic Left Heart Syndrome

J. Pruetz · J. Detterich · L. Trento · M. Sklansky

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Abstract Cor triatriatum sinister is a rare congenital heart defect related to incomplete common pulmonary vein resorption into the primitive heart. This lesion usually presents with pulmonary venous obstruction and can occur in association with left-sided obstructive lesions such as hypoplastic left heart syndrome (HLHS). In the context of HLHS, the presence of cor triatriatum sinister carries additional surgical and prognostic implications. Fetal diagnosis can enable appropriate counseling of the family and guide optimal peri- and postnatal management. The reported case represents the first prenatal description of cor triatriatum sinister in association with HLHS.

Keywords Common pulmonary vein resorption · Cor triatriatum sinister · Hypoplastic left heart syndrome · Pulmonary venous obstruction

Introduction

Cor triatriatum sinister is a rare congenital heart lesion related to incomplete common pulmonary vein resorption into the primitive heart. This early disturbance of embryologic development leads to the formation of two separate chambers in the left atrium: a posterior chamber that receives the pulmonary veins and an anterior chamber that communicates with the mitral valve and left atrial appendage [3]. The posterior chamber can have connections to the right atrium via an atrial communication, or it may have small fenestrations connecting it to the anterior chamber.

Typically, cor triatriatum sinister presents with pulmonary venous obstruction and commonly occurs in association with other left-sided heart lesions, including hypoplastic left heart syndrome (HLHS). In the context of HLHS, the presence of cor triatriatum sinister, much like the presence of a restrictive atrial communication, carries important surgical and prognostic implications [8]. Whereas the postnatal association of HLHS and cor triatriatum has been previously described, the prenatal diagnosis has not been reported to date [1, 2, 6, 7]. In this report, we describe the fetal diagnosis of cor triatriatum sinister in association with HLHS.

Case Report

A 34-year-old gravid 5, para 2 woman with a singleton fetus was referred at 22 3/7 weeks gestation for formal fetal echocardiography because of an abnormal-appearing four-chamber view on a routine second-trimester anatomic survey. Her family had no history of congenital heart disease, and the mother had no history of chronic illness or exposure to teratogens.

A detailed fetal echocardiogram (Philips IE-33 system, 5-MHz curvilinear probe, Philips Healthcare, Andover, MA) incorporating two-dimensional, spectral, and color flow Doppler methods demonstrated the diagnosis of HLHS with mitral atresia, aortic stenosis, moderate-size apical ventricular septal defect, and aortic arch hypoplasia.

J. Pruetz (✉) · J. Detterich · L. Trento
Division of Pediatric Cardiology, Children's Hospital Los Angeles, Keck School of Medicine, University of Southern California, 4650 Sunset Boulevard, Mailstop #34, Los Angeles, CA 90027, USA
e-mail: jpruetz@chla.usc.edu

M. Sklansky
Division of Pediatric Cardiology, Mattel Children's Hospital, David Geffen School of Medicine, University of California Los Angeles, Los Angeles, CA, USA

The patent foramen ovale was noted to have a mildly restrictive left-to-right shunting pattern, as seen by color flow Doppler. The four-chamber view suggested a second chamber in the left atrium (Fig. 1a), and spectral Doppler demonstrated very mild restriction of left-to-right flow across the atrial septum, with a mean gradient of 1 mmHg. However, the spectral Doppler waveform of the pulmonary veins suggested a more significant obstruction, with elevated peak systolic velocity (S), decreased peak diastolic velocity (D), and reversal of flow during late diastole/atrial systole (A).

An amniocentesis was declined, and the pregnancy continued without complication. A follow-up fetal echocardiogram at 36 2/7 weeks gestation again suggested a membrane dividing the left atrium. Repeat spectral Doppler interrogation of the pulmonary veins suggested worsening obstruction, with a decreased forward-to-reverse velocity time integral (VTI) ratio of 3 (Fig. 1b).

At 39 weeks gestation, the mother gave birth to a vigorous 3,256-g baby boy via vacuum-assisted vaginal delivery. The Apgar score was 8 at 1 min and 8 at 5 min. The baby was transferred to the neonatal intensive care unit receiving a prostaglandin E1 infusion. His initial chest radiograph demonstrated moderate cardiomegaly with diffuse interstitial pulmonary disease. Postnatal echocardiography (Philips IE-33 system, 8-MHz probe) confirmed the diagnosis of HLHS with mitral atresia, severe aortic stenosis, moderate-size muscular ventricular septal defect, and a restrictive patent foramen ovale. In addition, both two-dimensional (2D) and color flow Doppler imaging suggested cor triatriatum sinister (Fig. 2) but with limited visualization of a dividing membrane in the left atrium.

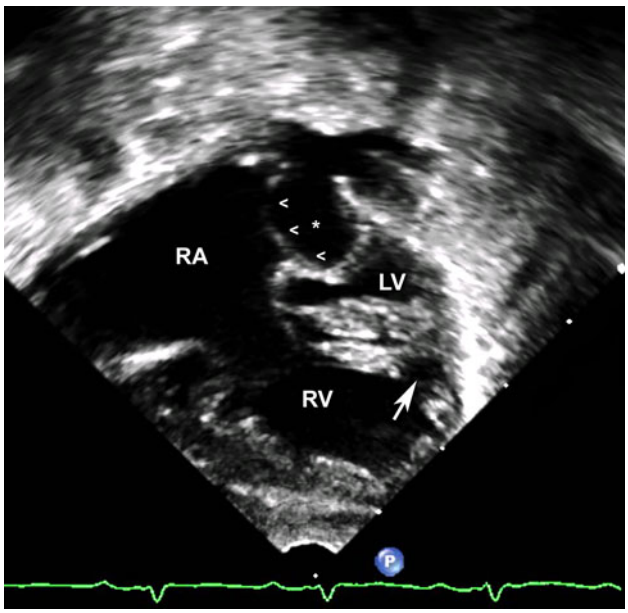

Fig. 2 Neonatal echocardiogram performed on the first day of life demonstrating an apical four-chamber view with cor triatriatum membrane dividing the left atrium into a posterior chamber (*asterisk*) and an anterior chamber bordered by the atrial septum and the dividing membrane, which are overlapping (*arrowheads*). The membrane dividing the posterior cor triatriatum chamber and the left atrium is thin and difficult to see. Dilation of the pulmonary veins is noted as well as mitral atresia with a moderate-size muscular ventricular septal defect (*arrow*) and hypoplastic left ventricle (LV). The right atrium (RA) and right ventricle (RV) both are dilated, and there is significant RV hypertrophy

Cardiac catheterization showed a small, left-sided posterior chamber that received the pulmonary venous return (Fig. 3). A gradient of 11 mmHg was noted between the

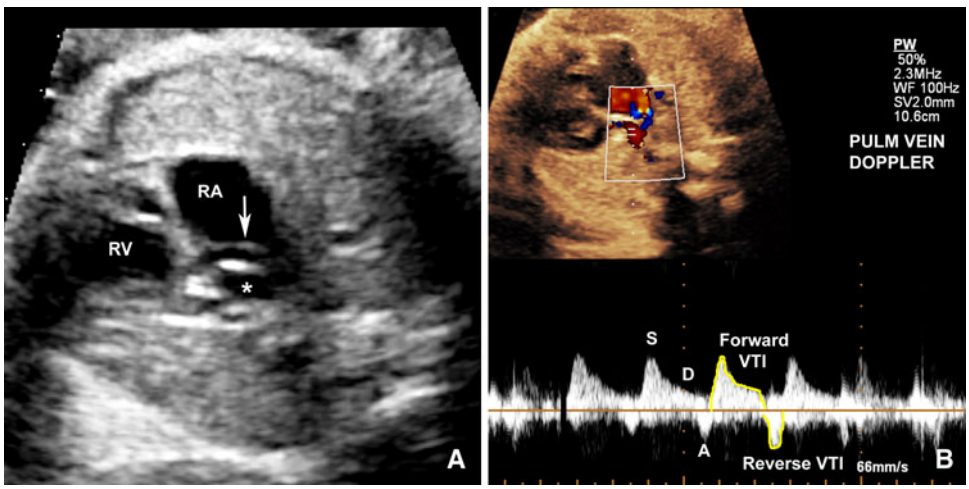

Fig. 1 **a** Fetal echocardiogram performed at 22 3/7 weeks gestation demonstrating a four-chamber view of the fetal heart with a separate chamber located in the left atrium (*asterisk*) that is distinctly separate from the atrial septum (*arrow*). Also noted are severe left ventricular hypoplasia and mitral atresia. **b** Spectral Doppler of the pulmonary veins showing a restrictive flow pattern with elevated systolic peak velocity (S), lower peak diastolic velocity (D), and reversal of flow during late diastole/atrial systole (A). Further evaluation of the pulmonary vein spectral Doppler shows abnormal A-wave duration and increased peak velocity with a decreased forward-to-reverse velocity time integral (VTI) ratio. *PFO* patent foramen ovale, *RA* right atrium, *RV* right ventricle

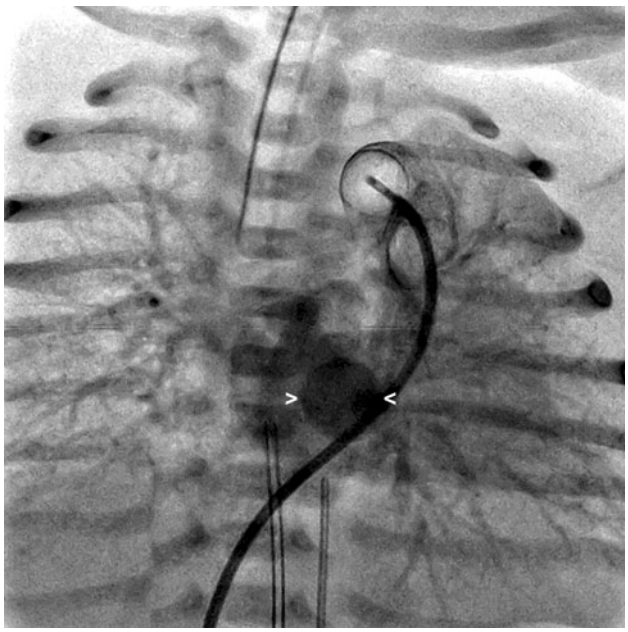

Fig. 3 Pulmonary artery angiogram from the second day of life. This angiogram is displayed in the straight anteroposterior projection. At levophase, the pulmonary venous flow drains to a small posterior chamber on the left side (*arrowheads*), which then drains into the left atrium via a restrictive communication

pulmonary capillary wedge pressure and the left atrial pressure, but only a 2-mmHg difference was noted across the atrial septum, suggesting significant obstruction proximal to the atrial septum. A preoperative transesophageal echocardiogram (Philips 7500 system, pediatric mini-multiplane TEE probe) demonstrated the posterior cor triatriatum chamber in the left atrium by both 2D and color flow Doppler (Fig. 4). Visual inspection by the surgeon confirmed the presence of a restrictive patent foramen ovale and a separate dividing membrane in the left atrium (cor triatriatum membrane).

The boy subsequently underwent a Norwood procedure with a right ventricle-to-pulmonary artery conduit (Sano shunt), atrial septectomy, and resection of the left atrial membrane. The postoperative period was complicated by difficulties weaning him from the ventilator, feeding issues, adrenal insufficiency, severe gastroesophageal reflux, and failure to thrive. The boy was discharged home at the age of 4 months receiving gastrostomy tube feedings and supplemental oxygen.

When the boy was 6 months old, a pre-stage 2 cardiac catheterization demonstrated proximal right pulmonary artery branch stenosis, elevated pulmonary vascular resistance of approximately 3 Wood units, and mild distal aortic arch coarctation, for which balloon angioplasty was performed. The patient underwent successful stage 2 palliation with a bidirectional cavopulmonary (Glenn) anastomosis and pulmonary arterioplasty.

Discussion

Whereas HLHS syndrome is one of the more common forms of cyanotic congenital heart disease (about 1 in 5,000 live births), cor triatriatum (in isolation or in association with other cardiac lesions) is a much rarer anomaly (about 1 in 30,000 live births) [9]. To our knowledge, this report is the first prenatal description of cor triatriatum sinister in association with HLHS. Eidem and Cetta [1] first reported this association observed in a 6-day-old neonate.

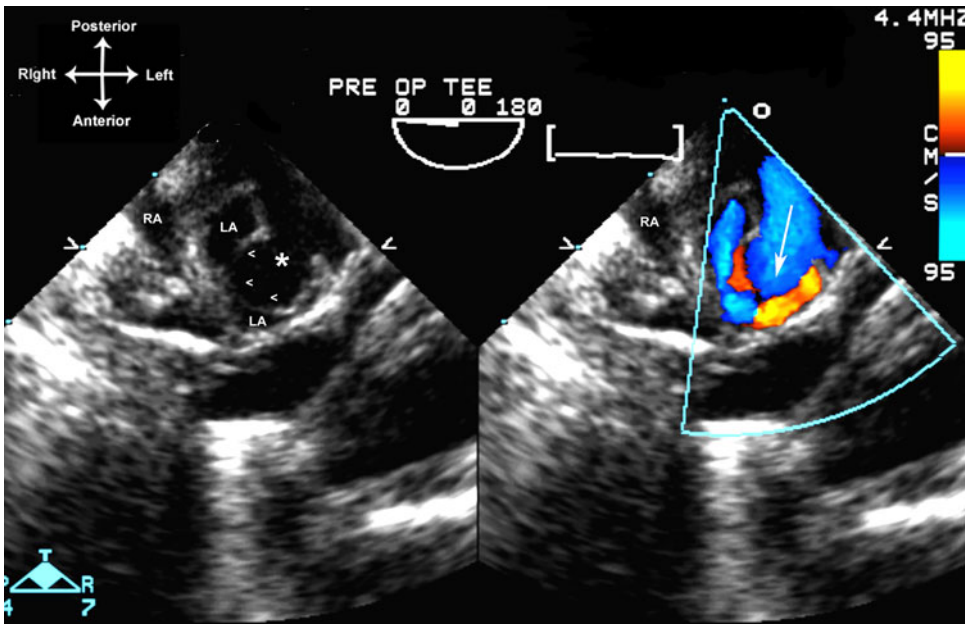

Fig. 4 Transesophageal echocardiogram performed preoperatively demonstrating a divided left atrium (LA) with a separate posterior chamber (*asterisk*) that receives the dilated pulmonary veins. A discrete membrane (*arrowheads*) separates the posterior chamber from the more anterior left atrium. There is flow into the posterior chamber (*arrow*) and then through a restrictive communication in the membrane into the left atrium before it crosses the patent foramen ovale (PFO). *RA* right atrium

Monaco et al. [6] described a neonate with a prenatal diagnosis of HLHS who ultimately was found also to have cor triatriatum sinister at postmortem examination.

Naito et al. [7] first described the surgical treatment of HLHS with a divided left atrium in 2007. However, in their reported case, the diagnosis of cor triatriatum sinister was not fully appreciated until postoperatively, when a cardiac magnetic resonance image (MRI) showed a muscular diaphragm completely dividing the left atrium. During the second-stage palliation for this patient, the surgeons successfully excised both the atrial septum and the diaphragm to create an unrestricted atrial communication.

In 2009, Taweevisit et al. [11] described a fetus with suspected Meckel-Gruber syndrome that had multiple congenital anomalies seen during fetal life. Postmortem autopsy confirmed multiple anomalies, including cor triatriatum sinister and HLHS. This appears to be the only description of both HLHS and cor triatriatum sinister reported in association with a specific genetic disorder.

Additional cardiac anomalies associated with HLHS include anomalous pulmonary venous return and atrial septal abnormalities. The detailed evaluation of atrial septal abnormalities can be critically important to appropriate counseling and surgical planning. The presence of an intact or restrictive atrial septum generally results in significant pulmonary venous obstruction with a pathophysiology similar to that of obstructed total anomalous pulmonary venous return. In both cases, abnormal prenatal development of the pulmonary vasculature can occur, leading to "arterialization" of the pulmonary veins and severe neonatal pulmonary hypertension with a very guarded prognosis [8].

Pulmonary venous Doppler flow patterns have been used to identify fetuses with HLHS who are at risk for severe left atrial hypertension at birth [10]. The ratios of pulmonary vein systolic velocity (S), diastolic velocity (D), and atrial systole velocity (A) as well as a forward-to-reverse VTI ratio all have been used to predict the need for emergent atrial septoplasty [5]. In our reported case, the assessment of the pulmonary venous Doppler flow patterns increased our clinical suspicion for cor triatriatum sinister because they suggested significant left atrial hypertension.

The association of HLHS with cor triatriatum sinister may carry prognostic and surgical implications similar to those for HLHS with restrictive atrial septum, particularly in cases with early prenatal obstruction at the level of the dividing membrane. Although attempts have been made to intervene during fetal life for cases of HLHS with restrictive or intact atrial septum [4], the presence of an additional dividing membrane may complicate the technical performance of this intervention and could limit the ability to relieve pulmonary venous congestion prenatally. Similarly, whereas emergent neonatal atrial septostomy/septectomy may be successful for cases of HLHS with restrictive atrial septum, the presence of cor triatriatum sinister likely would decrease the success of such transcatheter interventions. For these reasons, prenatal diagnosis of this lesion can have critical implications for prenatal and neonatal management.

In summary, the reported case represents the first published prenatal description of HLHS in association with cor triatriatum sinister. Prenatal diagnosis can improve risk stratification, enable appropriate counseling of the family, and guide optimal peri- and postnatal management. Thus, prenatal and neonatal evaluation of HLHS cases should always include a thorough evaluation of the left atrium for a dividing membrane.

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