Incomplete Shone's complex with cor triatriatum sinister and interrupted aortic arch: Prenatal diagnosis of a rare association

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ABSTRACT

Shone's complex is a rare constellation of multilevel left cardiac obstructive lesions involving both the inflow and outflow. Four primary lesions were included in the initial lesion description. However, other obstruction-causing structural lesions could be infrequently associated. The current report describes a fetal diagnosis of an unusual association of incomplete Shone's complex with cor triatriatum sinister and interrupted aortic arch.

Keywords: Fetal echocardiography, left-sided obstructive lesions, parachute-like mitral valve, supramitral ring

INTRODUCTION

Shone's complex is a rare multilevel left heart obstructive combination first described by Shone *et al.* in 1963. Four primary lesions were described for complete form, including supramitral ring, parachute mitral valve, subaortic stenosis, and coarctation of the aorta.[1] Although incomplete forms have been described to include two or three, suggestions were made to include other lesions such as cor triatriatum sinister, bicuspid aortic valve (AV) and small AV annulus, hypoplastic stiff left ventricle, and small aortic arch.[2] The current work describes a prenatally diagnosed case with an unusual association of incomplete Shone's complex with cor triatriatum sinister and interrupted aortic arch.

CASE REPORT

A 30-year-old, sixth gravida, a first para woman, had a history of four miscarriages and one living child. Unfortunately, she was from a remote rural area with limited prenatal care, and she was reluctant to seek medical care in a tertiary center until the late third trimester. She was referred for fetal echocardiography at 35 weeks of gestation due to a suspected abnormal cardiac obstetric screen, possibly hypoplastic left heart syndrome.

Transabdominal fetal echocardiography showed situs solitus and levocardia with concordant atrioventricular and ventriculoarterial connections. The right ventricle was dilated with a smaller yet apex-forming left ventricle. On careful assessment of the left side of the heart, pulmonary veins were seen draining into a separate chamber with an incomplete cor triatriatum membrane, causing no obstruction. However, turbulent obstructed inflow was seen starting at a small incomplete supramitral ring and through a suspected parachute mitral valve with an E: A ratio of 0.7. Nevertheless, the velocities of both waves were increased with E wave 72 cm/s (z score + 5.08) and A wave 106 cm/s (*z* score + 3.9) with a mean pressure gradient of 2 mmHg [Figure 1a-d and Video 1]. The tricuspid valve annulus to mitral valve annulus ratio was 1.6. The left ventricular subaortic area showed no

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obstruction; however, a mildly stenotic bicuspid AV was seen with mild turbulence across the AV at a velocity of 1.3 m/s. Diagnosis of the incomplete form of Shone's complex was made in addition to a small midmuscular ventricular septal defect with a right-to-left flow across the atrial septal defect; the foramen ovale flap was not properly seen antenatally. In the three-vessel view, four vessels were seen, indicating bilateral superior venae cavae with a dilated pulmonary artery and a smaller aorta failing to connect to the ductal arch with an aberrant right subclavian artery seen from the ductal arch [Figure 1e and f]. The aortic arch interruption type B was evident in the arch views [Figure 1g and h].

The baby was male, born at 37 weeks of gestation, with a low birth weight of 2110 g for gestational age. On postnatal transthoracic echocardiography,

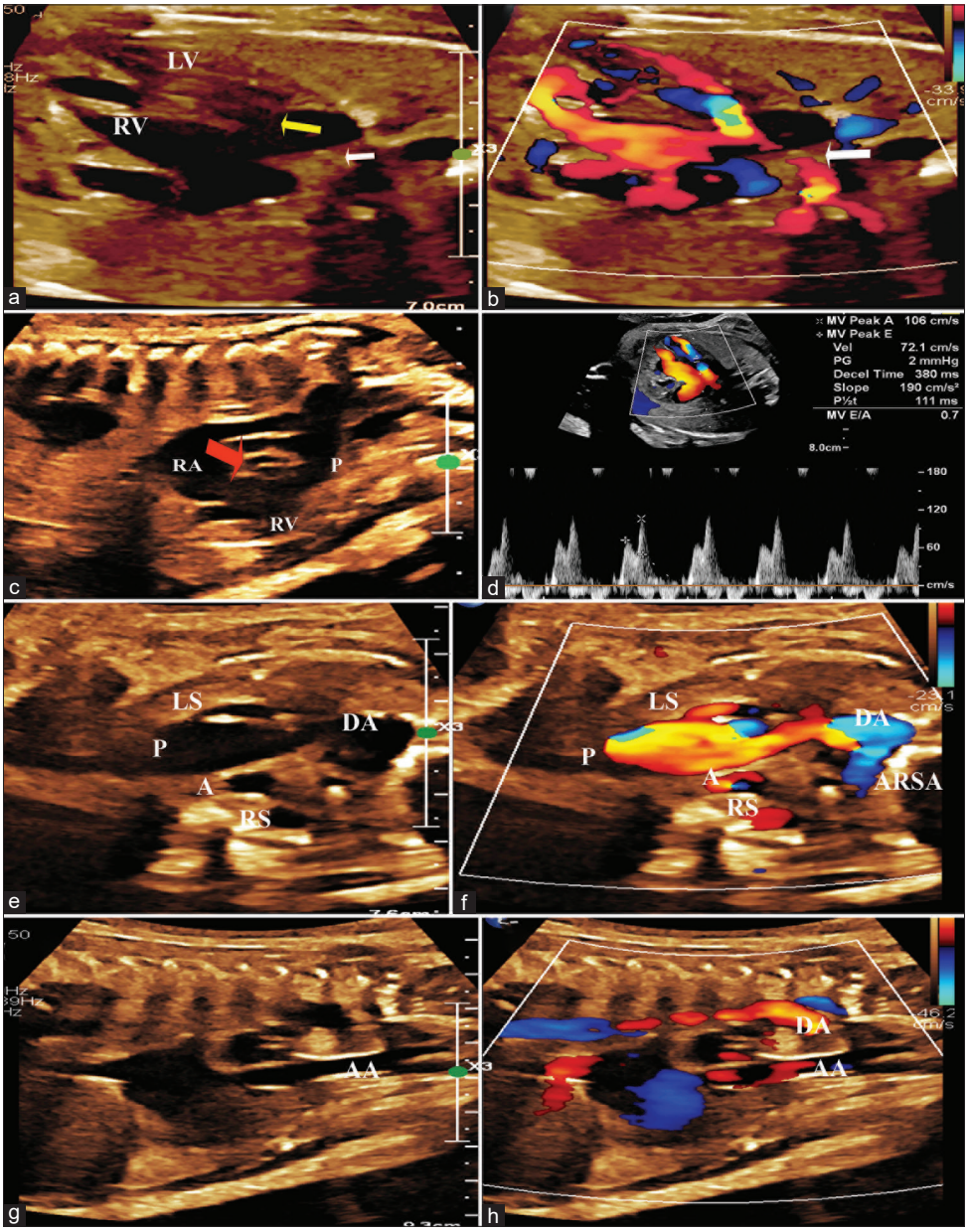

Figure 1: Fetal echocardiography: (a and b) Color-compare of four-chamber view demonstrating pulmonary veins draining to an accessory chamber with a membrane dividing left atrium (cor triatriatum), yellow arrow pointing to small supramitral ring with turbulent flow across and dilated right ventricle with smaller left ventricle with the white arrow is pointing to the cor triatriatum membrane. (c) Short-axis view demonstrating the aortic valve as bicuspid with red arrow pointing to it. (d) Pulsed wave Doppler across mitral valve demonstrating high velocities of E and A waves. (e and f) Color-compare of three-vessel ductal arch view demonstrating bilateral superior venae cavae with dilated pulmonary artery, smaller aorta, and aberrant right subclavian artery. (g and h) Color-compare of aortic arch view showing interrupted aortic arch type B with ascending aorta bifurcating into the right and left common carotids, then failing to arch to join the descending part. A: aorta, AA: ascending aorta, AC: accessory chamber, ARSA: aberrant right subclavian artery, DA: ductal arch, LA: left atrium, SVC: Superior vena cava, LS: left SVC, LV: left ventricle, P: main pulmonary artery, RA: right atrium, RS: right SVC, RV: right ventricle

the diagnosis of incomplete Shone's syndrome was confirmed with a parachute-like mitral valve including hypoplastic anterolateral papillary muscle and incomplete supramitral ring with a mean pressure gradient of 5 mmHg in addition to small secundum atrial septal defect in communication with the true distal left atrium (LA) and a moderate midmuscular ventricular septal defect 3.6 mm in diameter shunting left to right. The cor triatriatum sinister did not cause a significant obstruction, with a tiny patent foramen ovale in the atrial septum of the proximal accessory chamber [Figure 2 and Video 2]. The interrupted aortic arch was seen with bilateral superior venae cavae without a bridging vein. The left superior venae cavae drained to a dilated coronary sinus. Prostaglandin E1 infusion was commenced once the baby was delivered, and then, he was admitted to the neonatal intensive care unit. A multislice cardiac computed tomography angiogram with three-dimensional-reconstructed images [Figure 3] accurately demonstrated the anatomy of aortic arch interruption, confirming that it was type B with an aberrant right subclavian artery. Karyotyping was normal; however, no other genetic testing was performed due to financial constraints. Unfortunately, the baby required mechanical ventilation and died at the age of 1 week of extensive pneumonic consolidation and respiratory failure before scheduled arch repair.

DISCUSSION

Shone's complex is a phenotypically variable constellation of left-sided obstructive lesions. Pathogenesis of the complex is not well delineated; it has been suggested that disruption of left ventricular inflow leads to the underdevelopment of downstream structures.[3] The embryology of both Shone's complex and cor triatriatum is still poorly understood. For cor triatriatum, several theories were suggested. The mal-incorporation theory where it is suggested that the primary common pulmonary vein fails to incorporate normally into LA,

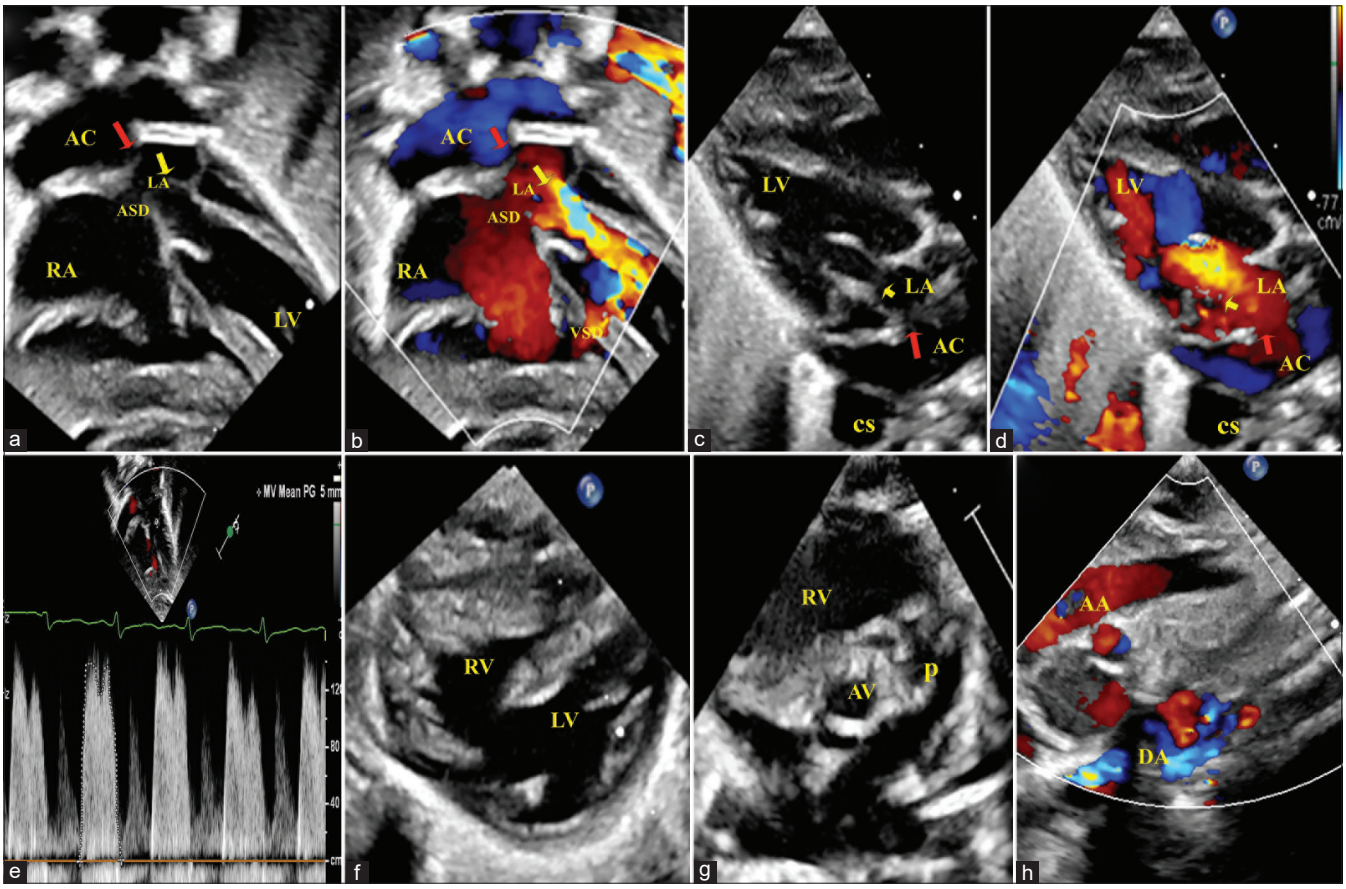

Figure 2: Neonatal transthoracic echocardiography. (a and b) Color-compare of subcostal view and (c and d) Color-compare of long parasternal view, showing the red arrow pointing to the cor triatriatum membrane, the yellow arrow pointing to the supramitral ring with turbulent flow across and parachute-like mitral valve in addition to secundum atrial septal defect and midmuscular ventricular septal defect both draining from left to right. Mitral z score: -3.8, tricuspid z score: -0.53, aortic z score: -1.6 , and ascending aorta z score: -1.5. (e) Doppler showing a mean of 5 mmHg across the ring, (f) short-axis view at papillary muscle level showing hypoplastic anterolateral papillary muscle with prominent posteromedial indicating parachute-like mitral valve, (g) short-axis view showing bicuspid aortic valve, (h) Suprasternal view showing the ascending aorta and interrupted arch. AA: aortic arch, AC: accessory chamber, AV: aortic valve, cs: coronary sinus, DA: descending aorta, LA: left atrium, LV: left ventricle, p: main pulmonary artery, RA: right atrium, RV: right ventricle, ASD: Atrial septal defect, VSD: Ventricular septal defect

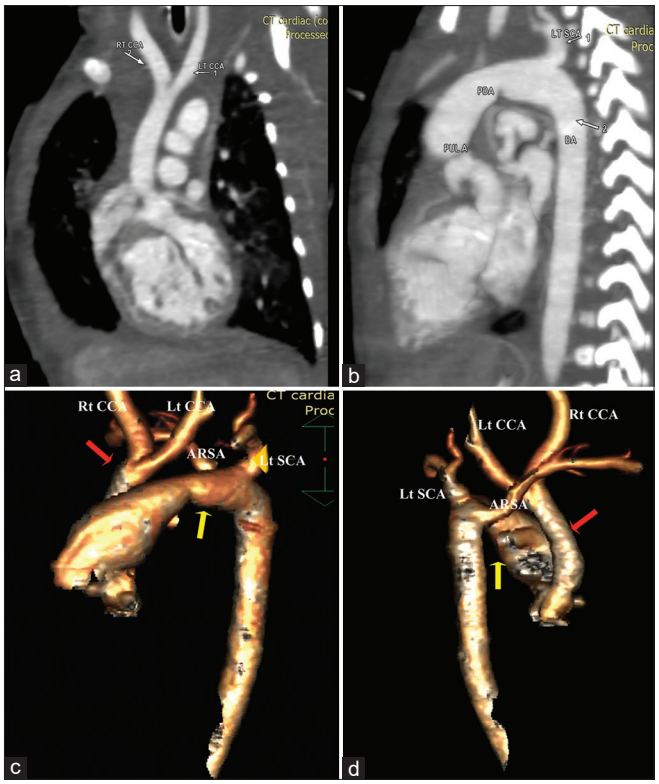

Figure 3: Cardiac computed tomography (CT): (a and b) Contrast-enhanced CT sagittal view showing aortic arch interruption type B. (c and d) Three-dimensional reconstruction of multidetector CT showing the aortic arch interruption type B with ascending aorta (red arrow) supplying left and right carotid arteries, then ductal arch (yellow arrow) supplying left subclavian artery and aberrant right subclavian artery. ARSA: aberrant right subclavian artery, Lt SCA: left subclavian artery, Lt CCA: left common carotid artery, Rt CCA: right common carotid artery, CT: Computed tomography, DA: descending aorta, PDA: patent ductus arteriosus, PuL A: pulmonary artery

resulting in forming two chambers separated by a narrow opening. Another theory is the malseptation, a membrane bisecting the atria due to an abnormal growth of the septum primum. Finally, the entrapment theory, where the left horn of the embryonic sinus venosus entraps the common pulmonary vein, preventing its incorporation into LA.[4] For the supramitral ring, it was suggested that it could be a developmental anomaly of the AV canal, with defective subdivision of the endocardial cushions resulting in a fibrous band that forms the ring; however, with the absence of other cushion defects and the association with other left ventricular outflow tract defects, this theory would not fully support Shone's complex genesis.[5] Moreover, the parachute mitral develops when the connection between the anterior and posterior parts of the trabecular ridge condenses to form a single papillary muscle.[6] Hence, further studies are required to explore the developmental background of these associations, as there could be interpolated elements, not the mere disruption of flow at the left ventricle inflow initially suggested by Shone *et al.*[1]

In cor triatriatum sinister, the LA is divided by a fibromuscular septum into a proximal or superior chamber, which mainly receives the pulmonary venous drainage, and a distal or inferior chamber (the true LA), which is in contact with the mitral valve and contains the atrial appendage. Several classifications were proposed for this rare cardiac anomaly in the literature.[7,8] Mashadi *et al.* proposed a new classification that considers the presence of other cardiac anomalies and obstruction of the cor triatriatum in addition to atrial communication and venous drainage.[9] The current case has cor triatriatum type 3 Loeffler classification, and according to Mashadi classification, it is type II B nonobstructive with a combined level of atrial communication and normal pulmonary venous drainage.

Few cases of Shone's complex diagnosed in intrauterine life have been reported in the literature. Only four cases were diagnosed among 12,520 fetal echocardiographic examinations in a study by Zucker *et al.*[10] Another study included 17 cases of fetal Shone's complex, showed ventricular disproportion and a lower left ventricular global longitudinal strain compared to aortic coarctation and control fetuses. They found that mitral valve to tricuspid valve ratio and global longitudinal strain could predict fetal Shone's complex.[11] Another report documented fetal partial Shone's complex with coarctation and parachute mitral valve. Diagnosis of parachute mitral in fetal life could be challenging. In a four-chamber view, the limited opening of the mitral valve leaflet is seen with chordae that converge into a single papillary muscle. Doming of elongated chords in diastole or enlarged LA could aid the diagnosis. Furthermore, a single papillary muscle could be detected in the short-axis at mid-papillary level.[12]

A case report documented the association of Shone's complex, including aortic coarctation, parachute mitral valve, and supramitral stenosis with cor triatriatum sinister in a 15-month-old male patient.[13] However, to the best of our knowledge, the currently described case is the first to report the association of partial Shone's complex with cor triatriatum and aortic arch interruption in fetal life.

Patients with Shone's complex often require multiple surgical and transcatheter interventions at an early age with favorable survival odds.[14] However, the prognosis remains uncertain in some cases. The variability in obstruction levels of the complex causes difficulty in predicting neonatal outcomes. Cases such as our patient with multi-major obstructive lesions might have a poor prognosis, and this should be clarified on counseling the parents prenatally.

Ethical standard

The authors assert that all procedures contributing

to this work comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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