Atrioventricular Septal Defect with Cor Triatriatum Sinister

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Cor triatriatum (CT) is a rare cardiac anomaly, characterized by a membrane in the left atrium which separates the atrium into the proximal and distal chambers. Association of CT with atrioventricular septal defects (AVSD) is extremely rare; only three cases with complete AVSD and 10 with partial AVSD have been reported. In this study, we present an 11-month-old female infant with complete AVSD and cor triatriatum. The patient recovered uneventfully after surgery and normal cardiac performance was achieved 2 years later. *(Chang Gung Med J 2007;30:270-3)*

Key words: atrio-ventricular septal defect, cor triatriatum

CASE REPORT

An 11-month-old female infant was admitted to the Chang Gung Children Hospital because of lip cyanosis after feeding and crying. History was notable for Down's syndrome and the birth body weight was registered as 2900 g. Physical examination identified that the child was mildly cyanotic but no evidence of congestive heart failure. Retarded development was found with body weight of 6 kg. A systolic murmur over the left middle sternal border was heard.

On her chest plain film, we identified pulmonary artery engorgement and increased bilateral lung markings. Electrocardiography showed sinus rhythm with complete right bundle branch block. Transthoracic echocardiographic study results demonstrated atrial septal defect, ventricular septal defect (atrioventricular canal type), severe mitral regurgitation and severe tricuspid regurgitation. Pulmonary artery pressure was 66/34 mm Hg. The patient's great vessels were in the normal relationship and the ductus arteriosus was patent. Intra-operative trans-esophageal echocardiography was arranged for an equivocal membranous structure in the left atrium.

Two-dimensional trans-esophageal echocardiography revealed an abnormal membranous structure in the left atrium, separating the pulmonary venous drainage from the mitral valve. Based on these findings, the patient was clinically diagnosed with atrioventricular septal defect (AVSD) with cor triatriatum sinister (Fig. 1).

Cardiac surgery was initiated from the median sternotomy. A cardiopulmonary bypass was established by arterial cannulation of the ascending aorta and bi-caval venous cannulation. Inspection of the atrioventricular defect and pulmonary veins was performed through right atriotomy. All pulmonary veins were drained into the proximal chamber, communicating with the distal chamber through a pin-point hole in the left atrial membrane. The hole in the left atrial membrane was enlarged toward the atrial appendage, followed by excision of the remainder of the membrane. The anterior bridging leaflet was tethered to the ventricular septum crest, which was classified as Rastelli's type A. The AVSD was reconstructed using a two-patch method for septal defects

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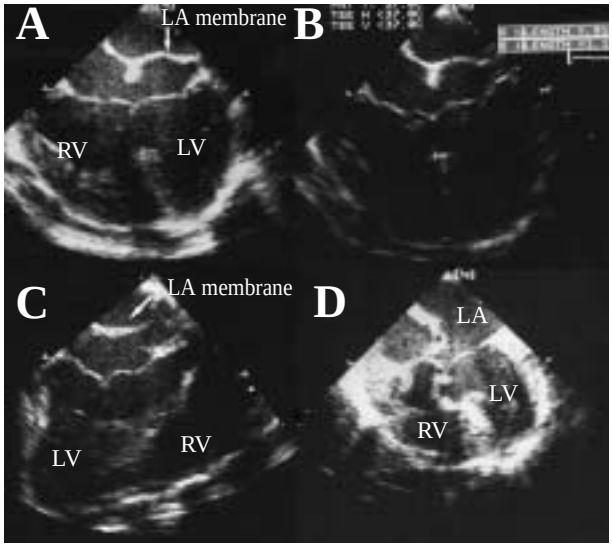

Fig. 1 Intra-operative trans-esophageal echocardiography: (A) Preoperative transesophageal echocardiography demonstrated a membranous structure in the left atrium in the four chambers view. (B) Atrio-ventricular defect was also documented by preoperative transesophageal echocardiography. (C) The membranous structure was also evident in the longitudinal two chamber view. (D) Post-operative transesophageal echocardiography proved no residual lesion in the left atrium, atrial septum and ventricular septum (*Asterisks indicate Dacron patch for atrial and ventricular septal reconstruction). White arrows indicate left atrium membrane; LA: left atrium; RA: right atrium; LV: left ventricle; RV: right ventricle.

and mitral valve was repaired. Aortic clamping time was 126 minutes, and total cardiopulmonary bypass time was 172 minutes.

Postoperative course was smooth and nitric oxide inhalation therapy was employed for 3 days. The patient was weaned off ventilation on the fourth post-operative day, and discharged 3 weeks after the operation. This patient recovered well at the 20th month follow-up examination.

DISCUSSION

Cor triatriatum (CT), first described by Church in 1868, occurs in approximately 0.1% of children with congenital heart disease.[1-3] In its classic form, CT is characterized by the presence of a fibromuscular diaphragm subdividing the left atrium into a proximal accessory chamber and a true distal chamber.

Association of CT with AVSD is rare; only three cases with complete AVSD and 10 with partial AVSD have been reported to the best of our knowledge.[4] Patients with AVSD are prone to develop pulmonary hypertension, and often lead to inoperable pulmonary vascular occlusive disease. Pulmonary venous obstruction with CT, which is also rare, is a correctable cause of pulmonary arterial hypertension. In AVSD, CT presents a diagnostic pitfall, which can lead to inadequate surgical intervention or misdiagnosis as inoperable. As first noted by Gahagan and Ziegler, CT has been missed during surgery, which led to postoperative death and subsequent diagnosis at autopsy.[5] Definite diagnoses rely on surgical exploration or autopsy prior to the development of echocardiography. Starc et al. made preoperative diagnoses using 2-D echocardiography in 1987.[6] In trans-thoracic echocardiography, the left atrium is foreshortened and, thus, the associated CT is often not visible in an apical 4-chamber view. Transesophageal echocardiography provides an alternative diagnostic modality under this scenario. In our case, CT and AVSD were reevaluated preoperatively using trans-esophageal echocardiography and confirmed by surgery.

Upgraded surgical outcomes for AVSD patients with CT has been recently reported, likely resulting from improved diagnostic procedures (i.e., echocardiography), enhanced knowledge of the anomaly, and enhanced postoperative intensive care (e.g., nitric oxide inhalation therapy).[2-4] Cardiac catheterization is usually performed for diagnostic confirmation and to access associated cardiac anomalies. Generally, patients with AVSD and CT have high right ventricular pressure, pulmonary artery pressure and pulmonary wedge pressure. Cardiac magnetic resonance imaging can be a highly effective means of diagnosing anomalies with a pulmonary connection and drainage in CT.[7,8] However, this modality is not feasible for neonates and infants and it is very expensive.

In conclusion, in this report, we presented a rare case of CT with complete AVSD. Precise preoperative echocardiography and intra-operative evaluation are essential to achieving promising surgical results for CT with AVSD. Cor triatriatum, as a correctable cause of pulmonary hypertension, should be considered in cases of complete atrioventricular septal defects.

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心室心房中隔缺損合併三心房畸形

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三心房 (cor triatriatum) 是一種少見的先天性心臟畸形，主要是心房內有一層膜將心房分成前後腔，前腔收集所有肺靜脈回流的血液，並將血液經由膜上的孔引流過後腔中。合併三心房的畸形與心房－心室中膈缺損 (Atrioventricular septal defect) 更是少見，文獻上僅有三個完全心房－心室中膈缺損和十個部分心房－心室中膈缺損的病例。本病例是一個十一月大的小女孩，合併完全心房－心室中膈缺損與三心房的畸形，經手術矯正，在術後兩年仍保有正常的心臟功能及發育。(長庚醫誌 2007;30:270-3)

關鍵詞：心房－心室中膈缺損，三心房畸形

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