Cor Triatriatum Dexter: Two-Dimensional Echocardiographic Diagnosis

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Cor triatriatum dexter is a malformation resulting from lack of normal regression of the embryonic right valve of the sinus venosus. In this situation, the right atrium is divided by a membrane into two chambers. Two-dimensional echocardiography was used in the antemortem diagnosis of this rare cardiac anomaly in a neonate. Associated cardiac lesions were also documented. The patient died, and findings were verified at autopsy.

Case Report

A 2.61 kg white girl was born to a 30 year old mother who had a bicornuate uterus and three previous spontaneous abortions. Because of spontaneous membrane rupture and fetal bradycardia, the baby was delivered at 37 weeks' gestation by cesarean section.

Apgar scores were 7 at 1 minute and 5 minutes. Heart rate during sleep was 55 beats/min. There was a grade 2/6 systolic ejection murmur at the left sternal border. There was no diastolic murmur. The liver was not enlarged. The electrocardiographic findings were consistent with congenital heart block.

Cor triatriatum dexter is a very rare cardiac anomaly in which a membrane divides the right atrium into two chambers (1). Antemortem diagnosis of this and other variations of pathologic persistence of the embryonic right valve of the sinus venosus can be difficult. Real-time two-dimensional echocardiography allows for an accurate identification of such malformations in the right atrium. We describe a neonate who had complex congenital heart disease and associated cor triatriatum dexter diagnosed antemortem by two-dimensional echocardiography and confirmed at autopsy.

Two-dimensional echocardiogram. There was situs solitus of the atria and viscera, with atrioventricular and ventriculoarterial concordance. A large membrane within the right atrium divided the atrial cavity into two chambers (Fig. 1). The membrane extended from an inferior attachment just above the tricuspid anulus, at the level of the rim of the inferior vena cava orifice, to a superior attachment at the roof of the right atrium, along the crista terminalis and the lateral rim of the superior vena cava. The inferior portion of the membrane was shaped like a wind sock, with its convexity toward the tricuspid orifice.

During ventricular systole (Fig. 1A), the portion of the right atrium medial to the membrane appeared to be distended, probably secondary to the returning systemic venous blood. During ventricular diastole (Fig. 1B), the superior portion of the membrane was pulled toward the atrial septum. This pulling was probably due to the shunting of the accumulated blood across the foramen ovale during the cardiac cycle. Also, during diastole, the inferior wind sock portion of the membrane was carried toward the orifice of the tricuspid valve and the right ventricle.

Injection of contrast solution (0.5 ml of saline solution) into the inferior vena cava through an umbilical venous catheter showed opacification of that portion of the right atrium medial to the membrane. The entire contrast flow pattern was from the right atrium to the left atrium through a patent foramen ovale. Subsequent flow was coursed to the left ventricle, with eventual opacification of the right ventricle through a ventricular septal defect.

Additional echocardiographic findings included an 8 mm membranous ventricular septal defect with an aneurysm of the membranous septal remnant. The myocardium appeared to be hyperrefractile and thickened and had a "spongy,"
highly trabeculated appearance and decreased contractility. The pulmonary valve was thickened, domed during systole and had a greatly reduced orifice. There was a patent but narrow ductus arteriosus.

**Hospital course.** Cardiac catheterization was performed when the infant was 2 days old and findings were similar to those of echocardiography. Total tricuspid obstruction by the right atrial membrane could not be documented. The catheter could be advanced easily from the right atrium to the right ventricle. The left ventricular end-diastolic pressure was 23 mm Hg, the end-diastolic volume was 224% of normal and the ejection fraction was 46%. The peak systolic gradient across the pulmonary valve was 12 mm Hg. The infant developed severe metabolic acidosis toward the end of cardiac catheterization. There was progressive deterioration despite the use of alkali, inotropic agents and transvenous cardiac pacing. She died a day later.

**Postmortem examination.** Congenital heart disease was associated with thoracoabdominal situs solitus and a unilobed left lung. A large septating membrane was present in the right atrium, consistent with cor triatriatum dexter (Fig. 2). Except for a slitlike perforation along its posterosuperior aspect, the membrane separated the venae cavae and coronary sinus from the right atrial appendage and tricuspid orifice. The lower half of the membrane was redundant and was shaped like a wind sock, with the convexity toward the tricuspid orifice. The foramen ovale was patent, and the tricuspid annular size was normal. There was an 8 mm di-
Figure 2. Postmortem specimen bisected to simulate the subcostal four chamber view (compare with Fig. 1A). The membrane (white arrows) divides the right atrial cavity in two. The smooth-walled portion of the right atrium, which receives the venae cavae, is medial to the membrane, whereas the pectinate muscles, atrial appendage (not seen) and tricuspid valve orifice are lateral. The lower portion of the membrane is shaped like a wind sock and overlies the tricuspid valve orifice. Both ventricles have trabecular (spongy) apices. FO = foramen ovale (black arrows); IVC = inferior vena cava; other abbreviations as in Figure 1.

ameter membranous ventricular septal defect partially obstructed by the septal tricuspid leaflet and by a small aneurysm of the anterior tricuspid leaflet. In addition, there were four to 2 mm muscular ventricular septal defects of the anteroapical trabecular septum. The myocardium had a spongy characteristic with extensive fine intracavitary trabeculations. There was pulmonary stenosis due to a thickened three cusped pulmonary valve without commissural fusion. The ductus arteriosus was patent but narrow. A nonobstructive shallow supravalvular mitral ridge was also observed.

**Discussion**

**Normal embryogenesis.** During early cardiogenesis, the right horn of the sinus venosus is guarded by two valves, the right and the left venous valves (2). The smaller left valve becomes incorporated in the septum secundum, but the right valve almost completely divides the right atrium into two chambers. This structure normally regresses between the 9th and the 15th week of gestation, as the cecal portion forms the crista terminalis and the caudal portion develops into the eustachian valve of the inferior vena cava and the thebesian valve of the coronary sinus. Extensive fenestrations of the right venous valve may result in a web-like Chiari’s network (3).

**Persistence of right sinus venous valve.** Lack of normal regression of the right valve of the sinus venosus may rarely occur. This can result in abnormalities ranging from partial septation of the right atrium by a prominent eustachian valve to complete division of the right atrium (cor triatriatum dexter) (4). In other cases, the right valve of the sinus venosus may form a pendulous wind-sock-like structure (5). Depending on the length of the “stalk” and where it is carried by blood flow, the wind sock may obstruct the tricuspid orifice (6), right ventricular outflow tract (5), inferior vena cava (7) or atrial septal defect (8). Antemortem diagnosis of this malformation is difficult and, in most cases, can be appreciated only by visual inspection at operation or autopsy.

**Role of echocardiography.** High resolution two-dimensional echocardiography is a valuable tool in identifying the location, size and site of attachment of anomalous remnants of the right sinus venosus valve. To our knowledge, very few echocardiographic reports on this malformation exist in the English literature. M-mode echocardiographic features have been described (5,9). Using two-dimensional echocardiography, Gussenhoven et al. (9) described a pedunculated structure extending from the right atrium, just above the tricuspid valve, into the right ventricle and across the pulmonary valve. In another report (6), a Chiari’s network coexisted with a wind-sock-like structure that arose near the inferior vena cava orifice. However, we could not find any report describing the two-dimensional echocardiographic findings of cor triatriatum dexter.

In the present case, the diagnosis of cor triatriatum dexter is warranted because of the extensive superoinferior attachment of the septating membrane. In this condition, the amount of blood that is shunted across a patent foramen ovale is determined by the extent of septation of the right atrium, the presence of fenestrations in the membrane and the mechanical obstruction of the tricuspid orifice by the redundant portion of the septating membrane. An echocardiographic contrast study enabled us to determine the degree of right to left shunting of the systemic venous blood across the patent foramen ovale. Although complete right to left shunting occurred at the atrial level, the presence of ventricular septal defects and a patent ductus arteriosus allowed pulmonary blood flow to be maintained. The fenestration in the superoposterior aspect of the membrane may have permitted passage of the catheter from the right atrium to the right ventricle during cardiac catheterization.

**Associated cardiac lesions.** There is a high frequency of serious congenital cardiac malformations associated with pathologic persistence of the right valve of sinus venosus. The most commonly observed anomalies include pulmonary atresia or stenosis, hypoplastic right ventricle and tricuspid valve atresia or hypoplasia (1,4). A substantial contributing
factor to the hypoplasia of the right ventricle and pulmonary arterial trunk has been believed to be the direction of blood flow into the left atrium and away from the right ventricle (4). The hypoplastic left heart syndrome also has been reported (6).

Implications. It is important to look for malformations in the right atrium, particularly when congenital right heart lesions are present. Two-dimensional echocardiography is a valuable tool for the evaluation of the subdivided right atrium that, if not identified, could complicate surgical intervention for the more serious associated cardiac malformations.

References