Successful catheter ablation using real-time ultrasound-assisted 3-D electroanatomical mapping system for atrioventricular accessory pathway in a 1-year-old girl with criss-cross heart

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Introduction

Criss-cross heart, defined as crossing of the long axis of the atrioventricular (AV) valves, is an extremely rare congenital heart defect. The unusual arrangement of the cardiac inlets observed in criss-cross heart was first described by Lev and Rowlatt[1] in 1961. The term "criss-cross heart" was initially coined by Anderson et al[2] in 1972. The reported incidence of criss-cross heart is no greater than 8 per 1,000,000 and accounts for $<0.1\%$ of congenital heart defects.[3] According to the relationship of the AV connection, criss-cross heart is divided into 2 types: criss-cross heart with AV concordance is known as concordant criss-cross heart, whereas criss-cross heart with AV discordance is termed discordant criss-cross heart. Catheter ablation of complex congenital heart defects is clinically challenging because the embryology of the cardiac conduction system, particularly the AV node, is complex and the associated anatomical structures are difficult to identify accurately.

The CARTOSOUND system (Biosense Webster Inc, Diamond Bar, CA), an ultrasound-based 3-D imaging modality for catheter navigation, was introduced by Forleo et al[4] in 2011. Because this technique provides anatomically accurate volumes from real-time 2-D intracardiac echocardiographic images, it might have particular utility in accessing challenging anatomical structures, as observed in congenital heart disease.

Herein, we present a very rare case of successful catheter ablation of concealed accessory pathway using the CARTOSOUND system in a 1-year-old girl with criss-cross heart and AV discordance following bidirectional Glenn procedure.

KEYWORDS Catheter ablation; CARTOSOUND; Accessory pathway; Criss-cross heart; His potential; Complex congenital heart disease
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Case report

A 1-year-old girl with criss-cross heart, AV discordance, double-outlet right ventricle, pulmonary atresia, and ventricular septal defect following surgical palliation of bidirectional Glenn procedure and atrial septal defect enlargement was referred to our hospital. She had a history of recurrent and drug-resistant supraventricular tachycardia from the age of 9 months. On admission, her body weight was 8.9 kg. Her electrocardiogram at rest and during tachycardia revealed a short RP tachycardia, with orthodromic reciprocating tachycardia being high in the differential diagnosis (Figure 1).

We planned deliberate electrophysiology study and catheter ablation under general anesthesia preceding the Fontan operation. Enhanced cardiac computed tomography (CT) was performed prior to catheter ablation and revealed the precise cardiac anatomical structures. The right atrium drained through the mitral valve to the posterior morphologic left ventricle. The left atrium was observed lying posterosuperior and left of the right atrium and drained through the tricuspid valve to the anterior morphologic right ventricle, from which the aorta originated anteriorly. The tricuspid valve opened anteriorly, and the mitral valve opened leftward. The ventricular septum was oriented vertically (Figure 2A).

We ensured vascular access via the right femoral vein and the left femoral artery. Baseline electrophysiology study was performed using an antegrade 5 French steerable 10-polar electrode through both atriums via the atrial septal defect and a retrograde 2 French steerable 4-polar electrode through the left ventricle via the ventricular septal defect. Her ventriculoatrial conduction without decremental property indicated a concealed AV accessory pathway. A 10 French SOUNDSTAR catheter (Biosense Webster Inc, Diamond Bar, CA), typically used for intracardiac echocardiography (ICE), was

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KEY TEACHING POINTS

- CARTOSOUND system facilitate the acquisition of more accurate geometry around AV annuli of complex congenital heart defects than electroanatomical mapping alone.
- The identification of the His bundle potential is one of the most important points for safe ablation related to accessory pathways in cases with congenital heart disease.
- Esophageal SOUNDSTAR placement for small patients is "off-label" use, but provides us with images of adequate quality and no esophageal injuries.

placed in the esophagus because of her small body size and difficulty in obtaining vascular access. Using SOUNDSTAR, we traced the endocardial contours of the aortic root, right ventricle, left ventricle, and left atrium. We were able to obtain cross-sectional images, including those of the AV annulus. Furthermore, CT images were integrated into the CARTOSOUND system (Figure 2B). Integrated images on CARTOSOUND revealed wired anatomical information regarding the AV annulus. We then replaced the 10-polar electrode with a 7 French NAVISTAR nonirrigated mapping and ablation catheter (Biosense Webster Inc, Diamond Bar, CA). We observed the AV annulus using intracardiac electrography with an atrial-to-ventricular voltage ratio of approximately 1:1. Her His bundle potential was identified at the inferior "right-sided" mitral annulus at the junction of the mitral and tricuspid annuli (Figure 3A and B). We were then able to induce clinical tachycardia. Sustained tachycardia was confirmed as orthodromic AV reentrant tachycardia (AVRT). During tachycardia, we investigated the earliest atrial excitation site and found it to be located at the superior "left-sided" tricuspid annulus, 20 mm from the site at which the His bundle potential was recorded. Echocardiography provided by SOUNDSTAR revealed that the target point was located at the tricuspid annulus (Figure 2C). A single application of radiofrequency current (limit, 40 W 55°C; total duration, 60 s) immediately terminated tachycardia (Figure 3A and C). After ablation, ventriculoatrial conduction disappeared and tachycardia was no longer inducible. No recurrences of AVRT or late complications have been observed over a 6-month follow-up period. The patient remains scheduled to undergo the Fontan operation.

Parental consent for this case report to be published was provided.

Discussion

We report a very rare case report of catheter ablation in criss-cross heart. To the best of our knowledge, only 1 case of catheter ablation in criss-cross heart has previously been reported: the case of a 13-year-old boy with remote surgical palliation for cyanotic criss-cross heart with AVRT, in which a bidirectional AV accessory pathway was successfully ablated guided by nonfluoroscopic mapping and the use of 3-D magnetic resonance imaging integrated into the Nav-X system.[5] There are only 3 case reports of Wolff-Parkinson-White syndrome in criss-cross heart. The first case was an 8-month old girl with intermittent ventricular pre-excitation in criss-cross heart with concordant AV connections and a small ventricular septal defect.[6] The second case was a newborn with cyanosis, dyspnea, and supraventricular tachycardia because of reentry who died suddenly after 6 days of follow-up. Necropsy confirmed the diagnosis of criss-cross heart with AV discordance, straddling right AV valve, double-outlet right ventricle, and ventricular pre-excitation.[7] The third case was a 13-year-old boy who received catheter ablation, as previously described.[5]

The identification of the His bundle potential is one of the most important points for safe ablation related to accessory pathways in cases with congenital heart disease. AV conduction block should be prevented. To date, there have been no review articles concerning the AV conduction system in criss-cross heart. The AV conduction system in cases of discordant criss-cross heart is speculated to be similar to that of corrected transposition of the great arteries (AV discordant and ventriculoarterial discordant heart). The location of the AV node in cases of corrected transposition of the great arteries is more anterior in the right atrium at the lateral junction of pulmonary and mitral valves. Further, an anteriorly situated bundle is seen to descend into the morphologic left ventricle and encircle the anterolateral quadrant of the pulmonary outflow tract before descending into the anterior septum and bifurcating with inversion of the bundle branches.[8] In the present case, we predicted that the location of the His bundle would be anterior to the right atrium at the junction of both AV annuli. We identified the dull potential at the predicted site. Although we meticulously sought around both AV annuli, we could not obtain high frequency potential. So we concluded that dull potential as the far field His potential. We could not perform atrial pacing confirming AH prolongation by limitation of the number of electrode catheters.

The recognition of the anatomical heart structures around the twisted AV annulus was essential in mapping the earliest atrial excitation site during tachycardia. The CARTOSOUND system has a substantial advantage in allowing modeling of anatomical shapes. During tachycardia, the CARTOSOUND system provides the most accurate real-time geometry and reveals the catheter position on real-time echo images.[9] The CARTOSOUND system enables the depiction of the ultrasonic contours in an arbitrary phase of the heart cycle, which results in more accurate merging with electrocardiography-gated angiographic images, such as coronary CT angiograms. ICE also provides direct visual control of lesion formation during ablation.[10] The CARTOSOUND system has utility in image integration with 3-D CT. Matching both respiratory phase and cardiac cycle between CARTOSOUND and CT image acquisition has been shown to improve image integration accuracy.[11]