

# Journal of Atrial Fibrillation



www. jafib.com

# **Atrial Fibrillation Ablation in a Patient with Absent Pericardium**

Vineet Kumar, MD, Takumi Yamada, MD, and G. Neal Kay, MD.

Division of Cardiovascular Disease, University of Alabama at Birmingham, Birmingham, AL, USA

#### Abstract

A 45-year-old woman with drug-refractory paroxysmal atrial fibrillation (AF) underwent AF ablation. She had a history of abnormal chest radiograph, which on review was consistent with congenital absence of pericardium and this was later confirmed on cardiac magnetic resonance imaging. She had extreme leftward and posterior rotation of the heart, resulting in abnormal fluoroscopic appearance of the electrophysiological catheters and orientation of the interatrial septum. This along with exaggerated beat to beat motion of the heart posed significant technical challenges for the electrophysiologist during left atrial access and pulmonary vein isolation.

## Introduction

Atrial fibrillation (AF) ablation usually requires transseptal access to the left atrium. Safe transseptal catheterization requires appreciation of the anatomical orientation of the inter-atrial septum and its relationship to the aorta, right atrial posterior wall, and the left atrium. Congenital complete absence of the pericardium is a rare cardiac anomaly and may be complicated with a markedly rotated heart. We describe the technical challenges this anatomical anomaly presents for left atrial catheterization by illustrating a case that underwent pulmonary vein isolation (PVI) for drug-refractory symptomatic AF.

#### Case

A 45-year-old woman with a history of atrial and ventricular septal defects repaired at the age of six was referred for catheter ablation of paroxysmal AF and atrial flutter. Prior ineffective medical therapy included flecainide and amiodarone. Written, informed consent was obtained, and an electrophysiological study was performed. Multipolar mapping catheters were positioned in the coronary sinus (CS), and His bundle (HB) region. Given the abnormal orientation of the heart on the chest radiograph (Figure 1A), a left ventriculogram was performed (Figure 2A and 2B) demonstrating

# Key Words:

Absent pericardium; atrial fibrillation; pulmonary vein isolation

Disclosures: None.

Corresponding Author:

Takumi Yamada, MD, PhD,

marked leftward and posterior rotation of the cardiac silhouette. This was later confirmed with cardiac magnetic resonance imaging (MRI) (Figure 1B). Rapid pacing from the CS catheter easily induced counterclockwise cavo-tricuspid isthmus dependent atrial flutter confirmed by electroanatomical mapping and transient entrainment. A cavo-tricuspid isthmus ablation line was created and bi-directional conduction block was confirmed by differential pacing. After the ablation of atrial flutter, incessant atrial fibrillation was observed and transseptal catheterization was performed using biplane fluoroscopy without the use of intracardiac echocardiography. The inter-atrial septum was oriented abnormally in the thoracic cavity and intracardiac landmarks such as the HB catheter were used to guide transseptal puncture (Figures 2C and 2D).

A narrow superior vena cava and exaggerated cardiac motion within the thoracic cavity made transseptal puncture under biplane fluoroscopy challenging. After transseptal access was obtained, intravenous heparin was administered to maintain activated clotting time of greater than 400 sec. After pulmonary venograms were obtained, a 20-mm, 20-pole circular mapping catheter was positioned selectively at the ostium of each of the pulmonary veins and successful PVI was achieved using irrigated radiofrequency current (ThermoCool, Bio-Sense Webster, Diamond Bar, Ca, USA). Atrial fibrillation converted to sinus rhythm during the ablation around the left-sided common pulmonary vein. Following PVI, no further atrial arrhythmias could be induced with rapid atrial pacing or during isoproterenol infusion. The total fluoroscopic and procedural times were 46 and 170 minutes, respectively. No complications occurred.

### Discussion

Congenital absence of the pericardium is a very rare and usually asymptomatic cardiac anomaly. This congenital anomaly is frequently

Division of Cardiovascular Disease, University of Alabama at Birmingham, FOT 930E, 510 20th Street South, 1530 3rd AVE S, Birmingham, AL 35294-0019 USA.



Figure 1:

A; Chest radiograph showing classical "snoopy dog" sign due to extreme levorotation and increased radiolucency between the aortic and pulmonary arteries (arrowhead). B; Cardiac magnetic resonance image exhibiting marked levorotation of the heart with the apex rotated posteriorly. Notice the inter-atrial septum is shifted (white dotted line) as compared with normal positioning (red dotted line)Ant=anterior, Post=posterior, Lt=left, Rt=right.

discovered incidentally either during surgery or during imaging for unrelated indications.<sup>1</sup> The characteristic findings on chest imaging include a "snoopy dog" appearance of the cardiac silhouette, and a teardrop appearance with a bulbous ventricle on the chest radiograph, and elongated atria on the apical four chamber view of transthoracic echocardiography. In addition, there is usually exaggerated motion of the heart and interposition of the lung in the usually lung-free space between heart and diaphragm as observed in both the coronal and sagittal axis on cardiac MRI.<sup>2</sup> Although patients with isolated complete absence of the pericardium are asymptomatic, those with partial defects can have symptoms ranging from atypical chest pain to more serious presentations as a result of incarceration of cardiac tissue.<sup>3,4</sup>

Performing invasive cardiac procedures on patients with these pericardial defects can be technically challenging given the marked displacement of the heart and its pendulum-like motion within the thoracic cavity.<sup>5</sup> A few surgical case reports have emphasized the procedural difficulties and associated complications during coronary artery bypass grafting and aortic dissection repair in these patients.<sup>6,7</sup> To our knowledge this is the first case report describing an invasive electrophysiological procedure in a patient with this congenital anomaly. The orientation of the CS catheter on RAO projection appeared twisted and the HB catheter was displaced leftward in this case (Figures 2C and 2D) as compared with a normal heart (Figures 2E and 2F). As demonstrated by the cardiac MRI, the inter-atrial septum was oriented more leftward and posteriorly making transseptal puncture challenging. Therefore, intracardiac echocardiography was likely to enhance the safety of

transseptal puncture in this case although it might not be necessary if a well-experienced operator was aware of the anatomy on the preprocedural imaging and was comfortable with a procedure using only the HB landmark and biplane fluoroscopy. Catheter ablation was also made more challenging because stability of the ablation catheter was compromised by exaggerated beat-to-beat cardiac motion. Electrophysiologists should also be vigilant for the presence of coexistent congenital cardiac defects and valvular abnormalities associated with absence of the pericardium.<sup>8,9</sup> There have been case reports of paroxysmal AF resulting from herniation of the left atrial appendage in patients with partial absence of pericardium requiring pericardial defect repair.<sup>10</sup> However, the prevalence of AF in patients with absent pericardium is not clear.

#### **Conclusions:**

Congenital absence of the pericardium presents challenges for catheter ablation because of the unusual orientation of the heart within the thorax and an exaggerated, pendulum-like cardiac motion. The use of pre-procedural imaging with cardiac MRI and intracardiac echocardiography may assist in the performance of these procedures.

#### **References:**

- Gatzoulis MA, Munk MD, Merchant N, Van Arsdell GS, McCrindle BW, Webb GD. Isolated Congenital Absence of the Pericardium: Clinical Presentation, Diagnosis, and Management. Ann Thorac Surg 2000;69:1209-1215.
- Morgan JR, Rogers AK, Forker AD. Congenital Absence of the Left Pericardium. Clinical Findings. Ann Intern Med 1971;74:370-376.
- 3. Ellis K, Leeds NE, Himmelstein A. Congenital Deficiencies in the Parietal Pericardium: A Review with 2 New Cases Including Successful Diagnosis by



Figure 2: A and B: Left ventriculogram in the RAO and LAO projections. Notice the leftward and posteriorly tilted left ventricle. Fluoroscopic images in the RAO and LAO projections exhibiting catheter positions during the trans-septal puncture in a patient with congenital absence of pericardium (C and D) as compared with normal heart (E and F). In LAO projection, the His bundle (HB) catheter was shifted leftward as compared with almost en face position in normal hearts. In RAO projection, the CS catheter appears markedly rotated.

CS=coronary sinus, LAO=left anterior oblique, RAO=right anterior oblique, TS=transseptal sheath

Plain Roentgenography. Am J Roentgenol Radium Ther Nucl Med 1959;82:125-137.

- Saito R, Hotta F. Congenital Pericardial Defect Associated with Cardiac Incarceration: Case Report. Am Heart J 1980;100:866-870.
- Topilsky Y, Tabatabaei N, Freeman WK, Saleh HK, Villarraga HR, Mulvagh SL. Images in Cardiovascular Medicine. Pendulum Heart in Congenital Absence of the Pericardium. Circulation 2010;121:1272-1274.
- Amiri A, Weber C, Schlosser V, Meinertz TH. Coronary Artery Disease in a Patient with a Congenital Pericardial Defect. Thorac Cardiovasc Surg 1989;37:379-381.
- Gersbach P, Kaufmann U, Althaus U. Congenital Absence of the Left Pericardium with Extreme Intrathoracic Displacement of the Heart. Case Report and Review of the Literature. Eur J Cardiothorac Surg 1996;10:473-476.

- Lu C, Ridker PM. Echocardiographic Diagnosis of Congenital Absence of the Pericardium in a Patient with Vater Association Defects. Clin Cardiol 1994;17:503-504.
- Rashid A, Ahluwalia G, Griselli M, Scheuermann-Freestone M, Neubauer S, Gaztoulis M, Kilner P, Shore DF. Congenital Partial Absence of the Left Pericardium Associated with Tricuspid Regurgitation. Ann Thorac Surg 2008;85:645-647.
- Misthos P, Neofotistos K, Drosos P, Kokotsakis J, Lioulias A. Paroxysmal Atrial Fibrillation Due to Left Atrial Appendage Herniation and Review of the Literature. Int J Cardiol 2009;133:e122-124.