A case report of cryoballoon-based pulmonary vein isolation in a patient with situs inversus abdominalis and levocardia

Omar Anwar, Tobias Espersen, Stephan Willems, and Christian Eickholt

Faculty of Medicine, Department of Cardiology, Asklepios Clinic St. Georg, Semmelweis University Campus Hamburg, Lohmühlenstrasse 5, 20099 Hamburg, Germany; DZHK (German Center for Cardiovascular Research), Partner Site Hamburg/Kiel/Lübeck, Berlin, Germany; Department of Cardiac Electrophysiology, Heart and Vascular Centre, University Hospital Hamburg Eppendorf, Hamburg, Germany; and Department of Cardiology, Klinikum Nordfriesland—Klinikum Husum, Husum, Germany

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Background
Cryoballoon-based pulmonary vein isolation (cbPVI) is a standardized treatment of atrial fibrillation. In complex anatomicies, radiofrequency ablation (rfPVI) is usually preferred. We describe the first cbPVI in a rare patient with SI and levocardia.

Case summary
A 41-year-old male patient with paroxysmal atrial fibrillation was referred to our clinic after a previous, unsuccessful cbPVI procedure. Observation of an atypical lead-wire position due to an abnormal anatomy of the inferior vena cava led to its initial termination. A subsequent thoraco-abdominal computed tomography revealed situs inversus abdominalis and levocardia and the procedure was re-attempted in our clinic. Transseptal puncture (TSP) was guided via transoesophageal echocardiography and fluoroscopy, using a SL0-Sheath and a standard BRK-needle. Advancement of the sheath initially failed but after additional dilatation with an Inoué dilator, transseptal passage of the sheath was successful. Due to the unusual antero-cranial TSP, the septal pulmonary veins (PV) contrasted poorly. After repeat TSP, a steerable FlexCath Advance sheath was introduced into the left atrium using an Amplatz Super Stiff guidewire. Subsequently, all PV were intubated with the Achieve catheter, over which a 2nd generation cryoballoon was introduced. Despite the practical challenges in this case, all PV were isolated.

Discussion
The main challenges include the achievement of transseptal access and manipulation of the cryoballoon to achieve a patent seal of the pulmonary veins. cbPVI eliminates the need for constant re-positioning of the ablation catheter and might facilitate the creation of durable lesions under such difficult anatomical conditions.

Keywords
Case report • Situs inversus • Atrial fibrillation • Cryoballoon • Pulmonary vein isolation

Learning points
• Situs inversus abdominalis with levocardia is a rare anatomical variation, this report is the first to demonstrate the use of cryoballoon based pulmonary vein isolation in this context.
• Pulmonary vein isolation using a cryoballoon can be safely and effectively performed in certain complex anatomicies.
• Use of single-shot devices such as the cryoballoon may reduce procedural complexity in these situations.

* Corresponding author. Tel: 040-1818-85-5411, Email: o.anwar@asklepios.com
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Introduction

Cryoballoon-based pulmonary vein isolation (cbPVI), alongside traditional radiofrequency ablation (rPVI), has become a standard in the interventional treatment of atrial fibrillation. Few case reports have been published describing the use of rPVI in patients with cardiac congenital abnormalities. While single reports of conventional rPVI in patients with situs inversus have been published,1–3 we describe the first procedure in a patient with situs inversus abdominalis coupled with levocardi. Situs inversus with levocardi alone is an extremely rare abnormality and is almost always associated with congenital heart disease. In this particular case, no associated congenital heart disease was found. This article will briefly discuss the underlying structural abnormality, as well as the successful application of cbPVI in this challenging scenario.

Timeline

| Date       | Event Description                  |
|------------|------------------------------------|
| October 2017 | Initial diagnosis of paroxysmal atrial fibrillation |
| February 2018 | Initial cryoballoon-based pulmonary vein isolation in the tertiary cardiac centre with subsequent computed tomography and initial diagnosis of situs inversus abdominalis with levocardi |
| March 2018   | Successful cryoballoon-based pulmonary vein isolation |
| September 2018 | 6-month follow-up                   |
| January 2021 | 2-year follow-up                    |

Case presentation

A 41-year-old patient was referred to our clinic through an outward cardiology centre after failure to perform a pulmonary vein isolation, due to a complex anatomy of the patient. In the external centre, the patient underwent pre-procedural screening procedures, including transoesophageal echocardiography (TOE), without remarkable results. During the procedure, after vascular access was established, an atypical positioning of the lead wire was noticed during fluoroscopy. Parallel TOE imaging was performed to confirm the venous position of the lead wire, as accidental arterial puncture was initially suspected. Transoesophageal echocardiography confirmed the correct placement of the lead-wire and normal cardiac anatomy, while an atypical entrance of the inferior vena cava (IVC) into the right atrium (RA) was noticed. The placement of a steerable diagnostic catheter in the coronary sinus (CS) was not possible from the IVC, and the procedure was finally aborted. Cursory abdominal sonography showed an atypical orientation of the upper abdominal organs, without prior history of any known anatomical or congenital abnormalities. A subsequent computed tomography of the thorax and abdomen showed an atypical situs inversus with levocardi. A normal anatomy of the thorax was verified; however, the inferior vena cava manifested an unusual course. It began distally on the left side as it coursed through the left-sided liver, crossing over to the right side to connect with the right atrium (Figure 1 and see Supplementary material online for cine-sequences).

Subsequently, the patient was referred for a repeat procedure in our tertiary electrophysiology centre. The patient history revealed symptomatic paroxysmal atrial fibrillation despite antiarrhythmic drug therapy with Flecainide. The patient did not have an additional past medical history and cardiovascular risk factors were minimal (Obese Class I, WHO-Criteria). There were no further salient physical exam findings. After an in-depth discussion with the patient regarding the possible therapeutic options (medical therapy with Amiodarone vs. pulmonary vein isolation), taking the patient history and current evidence-based guidelines into account, the shared team decision led to an interventional treatment approach, as it possibly offered the best therapeutic outcome. Additionally, due to the unusual anatomy and presumable difficulty in manipulating different sheaths and catheters, we chose cbPVI over rPVI in order to limit the procedure to a single-shot approach with one steerable sheath.

Vascular access was established successfully. Placement of the CS-Catheter (InquiryTM, 2-2-2 mm spacing, 6 Fr, St Jude, USA) posed a challenge once again due to the atypical pathway of the IVC and deviated axis of the heart and was aborted in order to limit mechanical manipulation. Successful transseptal puncture (Figure 2) was performed under TOE- and fluoroscopic-guidance using a SL0-Sheath (Fast-CathTM, St Jude, USA) and a standard BRK-Needle (BRK-1TM, St Jude, USA). Advancement of the SL0-Sheath initially failed due to a rigid septal wall. After dilatation with an Inoue mitral valvuloplasty dilator (Toray, USA), transseptal passage of the SL0-Sheath was successful (see Figure 4 for fluoroscopic comparison of SL0 Sheath placement before transseptal puncture in a patient with a normal anatomy). A normal anatomy of the left atrium was angiographically verified, however, because of the deviated axis of the heart and antero-cranial transseptal puncture, the right-sided pulmonary veins contrasted poorly. Selective angiography of the right pulmonary veins caused a displacement of the Sheath (Fast-CathTM) back into the right atrium. After a repeat transseptal puncture, a FlexCath Advance Sheath (Medtronic, USA) was placed using an extra-strength guidewire (Amplatz Super StiffTM, Boston Scientific, USA).

Using an Achieve-Catheter and a 2nd generation cryoballoon (Arctic Front AdvanceTM, Medtronic), the left superior pulmonary vein (LSPV) was occluded successfully. Despite multiple attempts (n = 2 freezes), a minimal temperature of only -26°C was achieved. Nevertheless, early isolation of the LSPV was attained after the first freeze. The left inferior pulmonary vein (LIPV), which was positioned in an atypical infero-posterior position with regard to the transseptal puncture site. In comparison to a normal anatomy, this pulmonary vein was situated more posterior than anticipated and in order to occlude this vein, an atypical posterior placement of the balloon was necessary, which resulted in the successful occlusion and isolation of the LIPV with a single freeze and a minimal temperature of -42°C. Prior to ablation of the right pulmonary veins, the CS-Catheter was repositioned to the superior vena cava and good electrical capture of the right phrenic nerve was confirmed. Due to prior displacement of the sheath into the right atrium, the right superior pulmonary vein was targeted first. Due to capture loss of the phrenic nerve caused by a displacement of the pacing catheter, the first freeze had to be terminated, and...
the catheter was repositioned. A second freeze was initiated, reaching a temperature of -49°C, without achieving complete isolation; an additional freeze however, lead to an early and continuous pulmonary vein isolation (minimal temperature -44°C). Finally, the right lower pulmonary vein, positioned more inferiorly than anticipated, was successfully occluded and isolated with a single freeze (minimal temperature -45°C) (see Figure 3 for cryoballoon occlusions of all pulmonary veins). Peri- and post-procedural complications were ruled out and the patient was discharged on the second post-interventional day. Fluoroscopy time was 30 min. The dose-area-product reached 1991.7 cGycm². The total procedure time was 110 min. At 2-year follow-up (via telephone), the patient remained in stable sinus rhythm.

Discussion

Catheter-based PVI using the cryoballoon-approach is a complex procedure in patients with situs inversus abdominalis and levocardia. However, cbPVI was successfully performed in this patient using standard fluoroscopy. The main procedural challenges were the safe achievement of transseptal access and the manipulation of the balloon catheter. Due to the atypical anatomical pathway and deviated axis of the heart, optimal occlusion of the targeted pulmonary veins posed a challenge. In contrast, rfPVI would have potentially allowed more freedom of movement of the ablation catheter, due to a diverse choice of ablation catheters and sheaths, leading to a more individualized 'shaping' of the lesion. Also, the utilization of real-time 3D-visualization with rfPVI may have provided a certain visual advantage, although this advantage might have been compromised by the anatomical complexity, which would have impeded the manipulation of the sheaths and catheters. Another concern was the increased risk of potential mechanical complications (e.g. perforation and cardiac tamponade) due to the atypical positioning and need for more vigorous manipulation of the ablation catheter. Should cbPVI not have sufficed in this patient, a hybrid approach with a switch to rfPVI would have been considered in order to achieve a durable and safe pulmonary vein isolation.

To the best of our knowledge, this is the first report demonstrating successful cbPVI in a patient with situs inversus abdominalis and levocardia. It is also one of the very few reports, if not the only report, illustrating this rare congenital abnormality as an initial diagnosis in a 41-year-old patient.

Previous cases describing pulmonary vein isolation in patients with situs inversus totalis with dextrocardia, used the radiofrequency approach, as well as cbPVI successfully.

As demonstrated by our case, the use of cbPVI in this even more complex anatomical condition is safely feasible, with good clinical
long-term results. Although the complex anatomy posed a challenge and an additional imaging modality (TOE) had to be used for transseptal puncture, the procedure time of 110 min and total fluoroscopy time of 30 min was within range compared to standard cbPVI procedures.

Traditional rfPVI, due to the use of 3D electroanatomical mapping is usually the primary choice for ablation in complex anatomies; however, there are certain advantages of cbPVI in this context. The manipulation and positioning of the cryoballoon requires more initial effort, yet—once in place—it facilitates the creation of continuous and durable lesions. The application of successive impulses, during the point-by-point RF ablation, requires constant re-positioning of the ablation catheter, increasing the chance of incomplete lesions and complications. This is the reason robotic or magnetic catheter manipulation, mostly abandoned in standard PVI, is utilized in complex cardiac anatomies.

Apart from the novelty of this clinical case, the successful application of cbPVI in this complex case is encouraging and might inspire its utilization in other challenging anatomies.

**Lead author biography**

Omar Anwar is a physician in the Department of Cardiology and Intensive Care Medicine at the Asklepios Clinic St. Georg in Hamburg, Germany. His research currently focuses on clinical electrophysiology under the lead of Prof. Dr. med. Willems.

**Supplementary material**

Supplementary material is available at European Heart Journal - Case Reports online.
Figure 3 (A) Fluoroscopic image showing occlusion of the right superior pulmonary vein with an expanded cryoballoon. Superiorly seen in the image is a decapolar HIS-Catheter placed in the superior vena for simultaneous phrenic nerve stimulation. (B) Occlusion of the left superior pulmonary vein. (C) Occlusion of the right inferior pulmonary vein with simultaneous pacing of the phrenic nerve via a decapolar HIS-Catheter. (D) Occlusion of the left inferior pulmonary vein. Images (A) and (C) were taken in RAO 30° viewing angles and images (B) and (D) were taken in anteroposterior-projection. Also see Supplementary material online for fluoroscopic video footage of pulmonary vein occlusion with the cryoballoon in this patient.

Figure 4 (A) Fluoroscopic image (anteroposterior-projection) showing the SL0-Sheath in the right atrium, ascending obliquely from the inferior vena cava after coursing through the left-sided liver. (B) Fluoroscopic image (anteroposterior-projection) showing an SL0-Sheath in the right atrium in regular position in a patient with a normal anatomy.
**Slide sets:** A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

**Consent:** The authors confirm that witnessed verbal consent for submission and publication of this case report including images and associated text has been obtained from the patients detailed in this case report. This has been discussed with the editors.

**Conflict of interest:** SW and CE have received speaker fees and scientific funding from Medtronic.

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### Table 1 Qualitative data of the cryoballoon-procedure

| Pulmonary vein | Application | Lowest temperature (°C) | Time to isolation (s) | Duration (s) | Cumulative time (s) | Real-time recordings | Occlusion quality (1–4*) |
|----------------|-------------|--------------------------|-----------------------|--------------|---------------------|----------------------|------------------------|
| LSPV           | 1           | -26                      | 77                    | 77           | 206                 | Yes                  | 3                      |
|                | 2           | -26                      | N/A                   | 129          |                     |                      |                        |
| LIPV           | 1           | -42                      | N/A                   | 240          | 240                 | No                   | 4                      |
|                | 2           | -47                      | N/A                   | 140          |                     |                      |                        |
| RSPV           | 1           | -27                      | N/A                   | 41           | 361                 | Yes                  | 3                      |
|                | 2           | -49                      | 90                    | 180          |                     |                      |                        |
| RIPV           | 1           | -45                      | 45                    | 240          | 240                 | No                   | 4                      |

*aPerfect circumferential occlusion of the PV was defined as 4 and ineffective position with a massive efflux of contrast medium was defined as 1.
LIPV, left inferior pulmonary vein; LSPV, left superior pulmonary vein; RIPV, right inferior pulmonary vein; RSPV, right superior pulmonary vein.

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