

Pulmonary vein isolation in a patient with congenital pulmonary atresia: a case report

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Background

Tetralogy of Fallot is a congenital heart defect characterized by pulmonary valve stenosis, ventricular septal defect (VSD), overriding aorta, and right ventricular hypertrophy. In its' extreme form, the pulmonary valve orifice does not develop during organogenesis, resulting in pulmonary atresia. We report a case of catheter ablation of symptomatic atrial fibrillation (AF) in a 37-year-old patient with congenital pulmonary atresia.

Case summary

The young man described paroxysmal tachycardia correlating to AF episodes in the previously implanted event recorder. Computed tomography scan described the complex anatomy with congenital pulmonary atresia, VSD, and major aortopulmonary collateral arteries. Electroanatomical mapping revealed typical pulmonary vein electrograms in a hypotrophic left atrium. Modified pulmonary vein isolation was successfully performed and non-excitability of the ablation line was reached. The patient recovered uneventfully and event recorder interrogation showed no AF recurrence after 3 months.

Discussion

Incidence of pulmonary atresia is low. Untreated survival rate is 50% after 1 year and 8% after 10 years. Tachycardia is a major cause of increased morbidity and mortality in patients with cyanotic congenital heart defects and pulmonary vein foci are described as driver for AF. Considerations preceding catheter ablation included pathophysiological mechanism, complex anatomy, atypical left atrium access, and reduced pulmonary perfusion resulting in a hypotrophic left atrium. Pulmonary veins showed typical electrograms, and isolation of pulmonary veins was feasible without adverse events.

Keywords

Congenital heart defect • Pulmonary atresia • Atrial fibrillation • Pulmonary vein isolation • Case report

Learning points

- Pulmonary atresia is a complex heart defect and the extreme form of Fallot's syndrome.
- Atrial fibrillation and atrial re-entry tachycardia are frequent in patients with congenital heart defects.
- Pulmonary vein isolation was feasible without adverse events in this patient with pulmonary atresia.

Introduction

Tetralogy of Fallot is a congenital heart defect characterized by pulmonary valve stenosis, ventricular septal defect (VSD), overriding aorta, and right ventricular hypertrophy. In its' extreme form, the pulmonary valve orifice does not develop during organogenesis, resulting in a pulmonary atresia. We report a case of catheter ablation of symptomatic atrial fibrillation (AF) in a 37-year-old patient with congenital pulmonary atresia (Figure 1).

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Timeline

Patients history	<ul style="list-style-type: none"> • Congenital heart defect • Symptomatic tachycardia • Implantation of an event recorder • 12 months electrocardiogram monitoring
Intervention	<ul style="list-style-type: none"> • Correlation of episodes to symptoms • Magnetic resonance imaging • Computed tomography scan • Cardiac electrophysiology study • Electroanatomical mapping • Image integration and catheter ablation
Outcome and follow-up	<ul style="list-style-type: none"> • Isolation of pulmonary veins • Non-excitability of ablation line • No adverse events • No atrial fibrillation recurrence • Follow-up 3 months and ongoing

Case presentation

The young man described paroxysmal tachycardia according to EHRA Class III. A previously implanted event recorder showed episodes of AF, which could be correlated to the patients' symptoms. The device revealed 22 symptomatic AF episodes in 12 months. During the last 6 months prior to catheter ablation, the patient experienced six sustained AF episodes between 20 min and 2.5 h. Four episodes lasted longer than 1 h (1.5–2.5 h) and led to the hospitalization of the patient. Another 16 episodes with non-sustained AF were marked by the patient but were not detected by the event recorder. In total, AF induced severe symptoms but episodes were rather short, resulting in an AF burden of 0.1%.

Besides pulmonary atresia with nail clubbing and compensatory polycythaemia (Hb 23.1 g/dL, haematocrit 66%, SpO₂ 81%, blood pressure 115/70 mmHg, heart rate 85 b.p.m., respiratory rate 13/min, and body mass index 18.9 kg/m²), patients' history revealed resection of a cerebral abscess (22 years ago) with residual epilepsy. Medical treatment consisted of anticonvulsants (lamotrigine 100 mg/day, levetiracetam 2500 mg/day, and gabapentin 2800 mg/day) and Losartan (aortic dilation; beta-blocker not tolerated). Class I antiarrhythmics were not well tolerated (headache and dizziness) and therefore, discontinued. No anticoagulation was prescribed (CHADS-VASc Score 0).

Pre-procedural computed tomography scan (Figure 2) described the pre-known pulmonary atresia with a malalignment VSD, an overriding aorta shifted to the right with an aneurysmal dilatation up to 45 mm. Ascending aorta was dilated to 57 mm. Aortic arch and descending aorta were also shifted to the right side of the spine. Lung perfusion was enabled with multiple major aortopulmonary collateral arteries (MAPCA) and vasa privata were arising from peribronchial vessels. Ejection fraction of the hypertrophic right ventricle was



Figure 1 Magnetic resonance imaging scan of our patient with pulmonary atresia. Aorta, overriding aortic valve and dilated ascending aorta; LA, hypotrophic left atrium; LV, left ventricle; RV, hypertrophic right ventricle; VSD, ventricular septal defect.

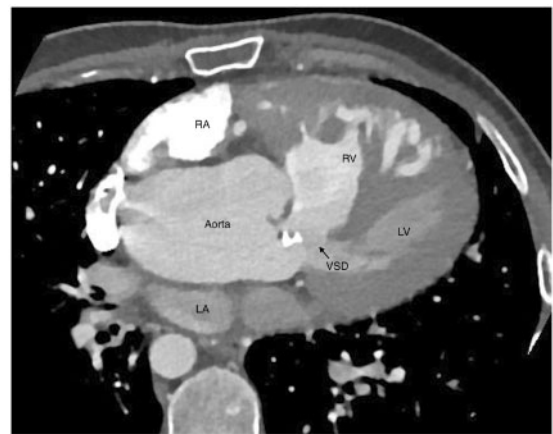


Figure 2 Computed tomography scan of our patient with pulmonary atresia. Aorta, overriding aortic valve and dilated ascending aorta; LA, hypotrophic left atrium; LV, left ventricle; RA, right atrium; RV, hypertrophic right ventricle; VSD, ventricular septal defect.

preserved. Vena cava inferior drains into right atrium forming a dilated venous convolute of 65 × 56 mm (Figure 3). Combination of pulmonary atresia with an atrophic pulmonary artery truncus resulted in multiple MAPCA. Although considered, the patient was ineligible for surgical repair.

Available images were integrated into a three-dimensional mapping system and transfemoral access to the venous system was obtained in analgosedation. Cardiac electrophysiology studies, including pharmacological challenge, did not trigger any non-pulmonary focal activity. In a tailored approach, we considered pulmonary vein isolation as reasonable.

Fluoroscopy-guided transseptal puncture was successfully performed without affecting the shifted and dilated aorta. Electroanatomical mapping revealed a hypoplastic left atrium with a narrow antero-posterior dimension (18 mm). Left pulmonary veins were configured as an

anterior and posterior pulmonary vein with a common ostium. Right pulmonary veins showed small diameters (15 mm) in concordance with the hypoplastic left atrium (Figures 3 and 4).

The multipolar mapping catheter showed typical pulmonary vein potentials in all pulmonary veins. During ablation, AF was induced mechanically. Modified antral pulmonary vein isolation was performed. Encircling of ipsilateral veins led to isolation of all veins and after electrocardioversion, endpoint of non-excitability of the ablation line was reached with a common ostium of the left superior vein (Figures 5–8). The patient recovered uneventfully and event-recorder interrogation showed no recurrence of AF during three months follow-up.

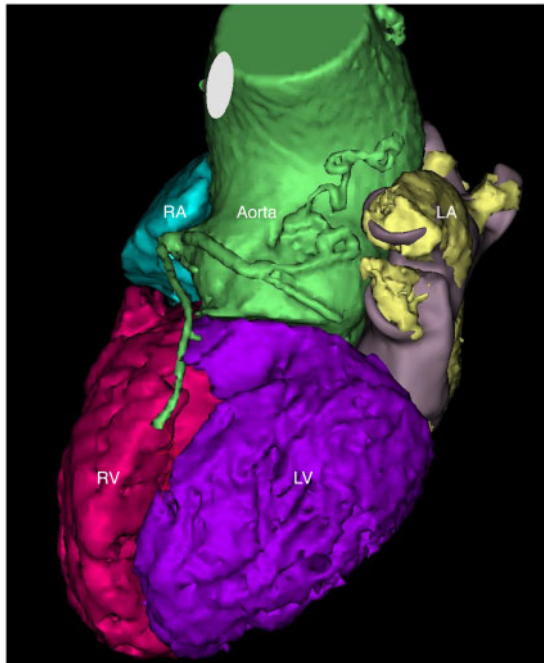


Figure 3 Electroanatomical map merged with three-dimensional computed tomography data (left-anterior-oblique/cranial view). Aorta, dilated ascending aorta; LA, hypoplastic left atrium; LV, left ventricle; RA, right atrium; RV, hypertrophic right ventricle.

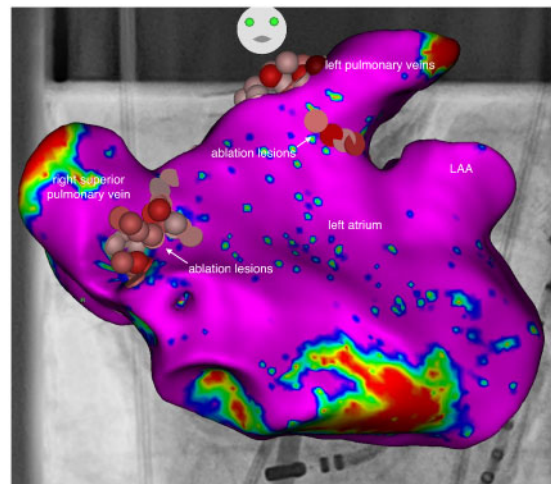


Figure 5 Electroanatomical map showing the left atrium (anterior-posterior view) and ablation marker encircling pulmonary vein ostia (red dots). LAA, left atrial appendage.

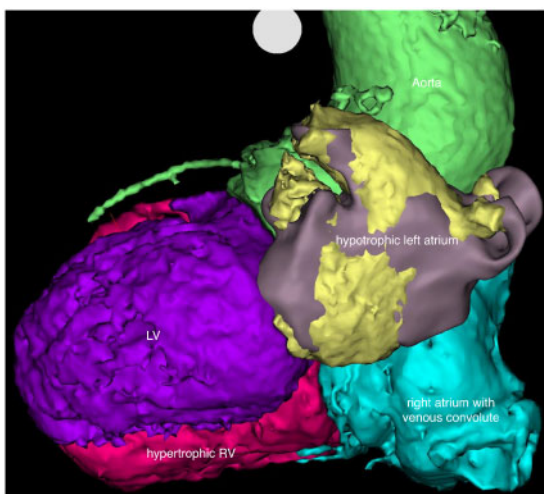


Figure 4 Electroanatomical map merged with three-dimensional computed tomography data (posterior-anterior view). Aorta, dilated ascending aorta; LA, hypoplastic left atrium; LV, left ventricle; RA, right atrium; RV, hypertrophic right ventricle.

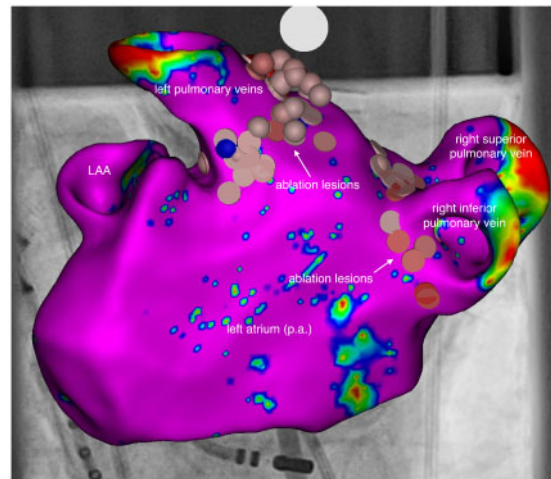


Figure 6 Electroanatomical map showing the left atrium (posterior-anterior view) and ablation marker encircling pulmonary vein ostia (red dots). LAA, left atrial appendage.

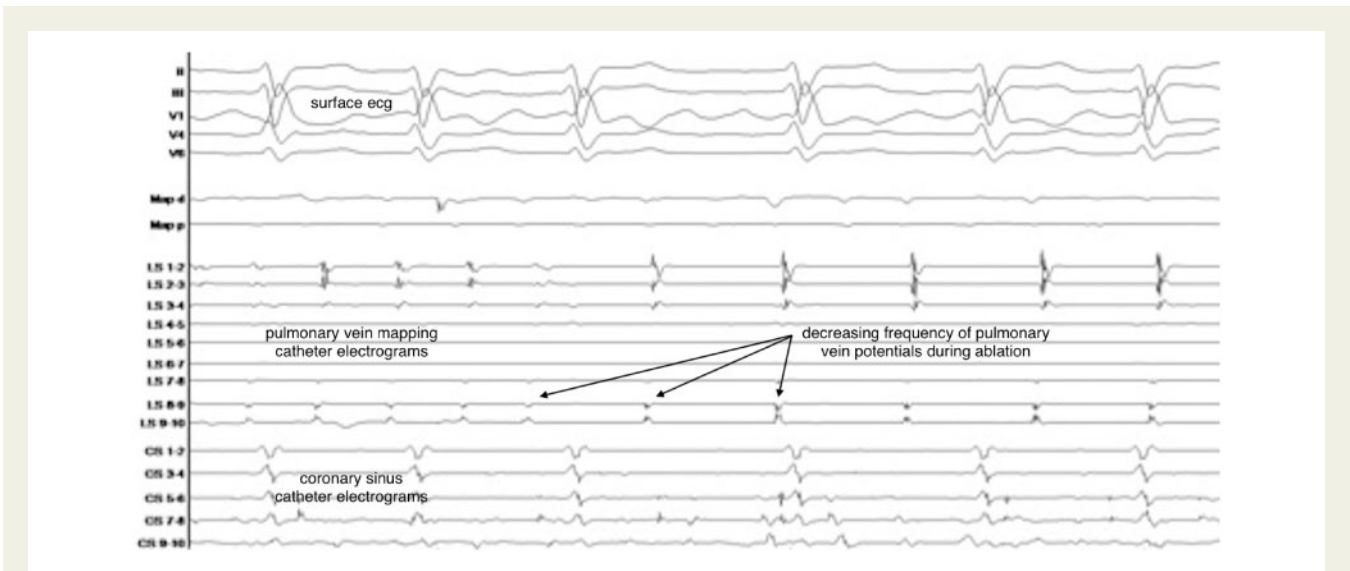


Figure 7 Peri-procedural recording of surface electrocardiogram (leads II, III, V1, V4, and V6) and intracardiac electrograms. Frequency of pulmonary vein signals decreases during ablation (black arrows) showing initial reduction of conduction capacity. CS 1–10, coronary sinus electrograms as reference; LS 1–10, circular mapping catheter in left pulmonary vein.

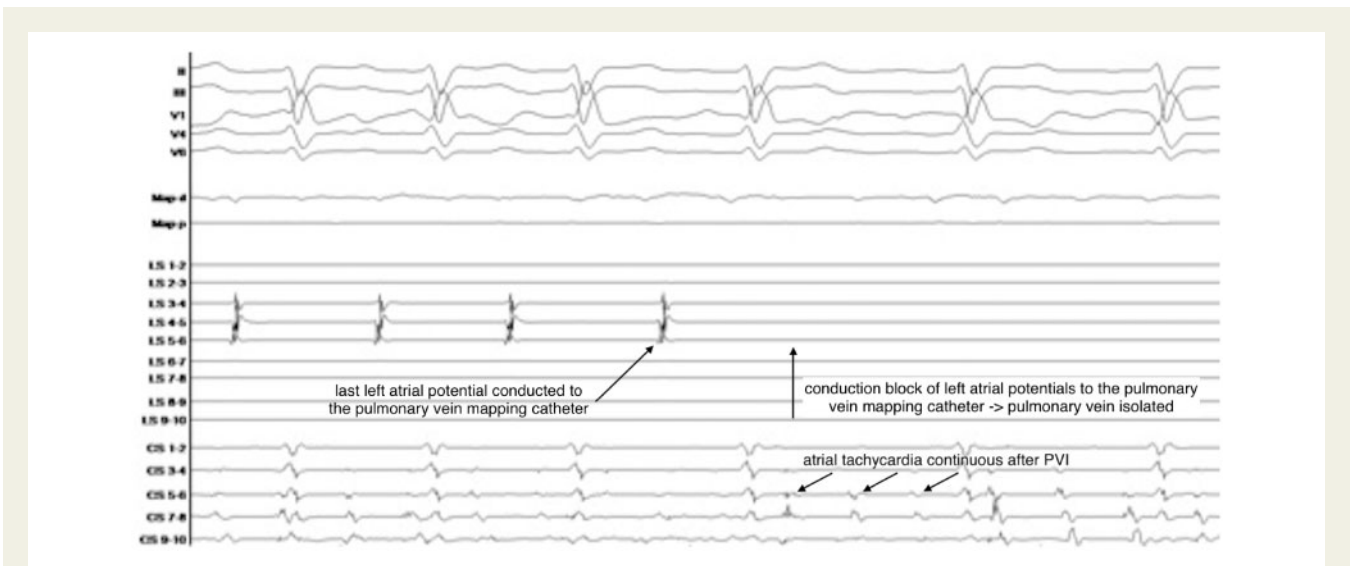


Figure 8 Peri-procedural recording of surface electrocardiogram (leads II, III, V1, V4, and V6) and intracardiac electrograms. Pulmonary vein signals vanish during ablation (perpendicular black arrow) indicating isolation of pulmonary vein. CS 1–10, coronary sinus electrograms as reference; LS 1–10, circular mapping catheter electrograms of left pulmonary vein; Map d and Map p, ablation catheter electrograms.

Discussion

Pulmonary atresia can present with or without VSD. In absence of a VSD, lung perfusion is realized via a persistent foramen ovale and ductus arteriosus. Our patient presented with the more frequent type of pulmonary atresia with VSD (1% vs. 0.3% of congenital heart defects).¹ Incidence of both types is low: 7/100 000 live birth with pulmonary atresia and VSD and 4.5/100 000 live birth for pulmonary

atresia with intact ventricular septum.² Untreated survival rate is 50% after 1 year and 8% after 10 years.³ Surgical repair of pulmonary atresia with VSD is challenging and to a large extent dependent on anatomical variations of MAPCA and diminutive central pulmonary artery truncus.^{4,5} Algorithms intend either multi- or single-stage unifocalization. Single-stage unifocalization can be performed with a total repair (of VSD) or a central shunt preceding total repair. Widely preferred surgical algorithm is a single-stage unifocalization at a young age (4–7 months).⁶

In cyanotic congenital heart defects, tachycardia is a major cause of increased morbidity and mortality.⁷ About 11% of all patients with congenital heart defect develop atrial tachycardia with intra-atrial re-entry tachycardia being more frequent than AF.^{7,8} Moe *et al.*⁷ suggest typical pulmonary vein foci as driver for AF in patients with congenital heart defects.

Data on pulmonary vein isolation in pulmonary atresia patients is sparse. Considerations preceding catheter ablation of AF included anatomical changes and access to venous system plus right atrium, atypical left atrium access, and reduced pulmonary perfusion resulting in a hypoplastic left atrium. Ablation catheter impedance was high (170–200 Ω) during pulmonary vein isolation. This may be due to the narrow left atrial chamber (18 mm) and potentially hyperdense pulmonary tissue secondary to reduced lung perfusion. No perforation, pericardial effusion or steam pop occurred.

In conclusion, reduced lung perfusion resulted in a hypotrophic left atrium in this patient with pulmonary atresia. Pulmonary veins showed typical potentials and isolation of pulmonary veins was feasible without adverse events. Event recorder interrogation showed no AF recurrence during three months follow-up.

Lead author biography



Tobias Plenge is an interventional cardiac Electrophysiologist at the University Hospital in Cologne, Germany. He has worked in the field of Cardiology since 2009 and his interests include catheter ablation of atrial and ventricular arrhythmias as well as cardiac implantable devices. Research activities focus on catheter ablation in a large animal model and clinical AF investigations.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none to declare.

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