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CASE REPORT
Electrophysiology

Ablation of atrial flutter in a patient after Fontan procedure: a case report of challenging access to the common atrium

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Background In patients with a total cavopulmonary connection in Fontan circulation, the access to the common atrium (CA) during a catheter ablation can be challenging, even in the presence of fenestration in an intra-atrial lateral tunnel (IALT). In our department, the fenestration is typically marked with metal clips (MCs). To the best of our knowledge, there is no previous report of balloonoplasty of clipped fenestration.

Case summary A 19-year-old male with hypoplastic left heart syndrome (HLHS) was scheduled for catheter ablation of recurrent atrial tachycardia. He was diagnosed with HLHS prenatally and underwent a stepwise surgical palliation. Fontan circulation was completed with the creation of a fenestrated IALT. The fenestration was marked by four MCs. During the ablation procedure, the passage of the steerable sheath with mapping catheter to the CA was prevented by a small fenestration size and rigidness of the edges of the fenestration caused by the MCs. Multiple attempts to dilate the fenestration using a peripheric angioplasty balloon failed. Only angioplasty with the 'balloon-against-dilator' technique was finally successful. Activation map showed a counterclockwise atrial flutter in the CA; successful ablation was performed.

Discussion We present a case of challenging access to the CA through a clipped fenestration in a polytetrafluoroethylene baffle for atrial tachycardia ablation. Even though a tunnel fenestration in Fontan patients facilitates access to the CA, the passage of a steerable introducer with a mapping catheter may be challenging due to diameter mismatch and the rigidity of its edges caused by MCs. The balloon-against-dilator technique might be helpful when conventional balloon angioplasty fails.

Keywords Ablation • Angioplasty • Atrial tachycardia • Case report • Hypoplastic left heart syndrome

ESC curriculum 5.4 Atrial flutter • 5.5 Supraventricular tachycardia

Learning points

- In patients with hypoplastic left heart syndrome after Fontan procedure, access to the common atrium could be challenging, despite the presence of fenestration in the intra-atrial lateral tunnel. Metal clips around the fenestration could make the dilation of fenestration even more complex.
- Balloon-against-dilator technique for dilation of the fenestration in polytetrafluoroethylene baffle might be helpful when conventional balloon angioplasty fails.

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Introduction

Hypoplastic left heart syndrome (HLHS) is a rare congenital heart defect, in which the left side of the heart and the aorta are underdeveloped, resulting in a functional single right ventricle. The treatment of choice is a three-stage surgical palliation with the final stage being the completion of a total cavopulmonary connection.[1] Different types of Fontan procedures exist; one common way is to create an intra-atrial lateral tunnel (IALT) with fenestration between the IALT and the atria.[2] In our department, the fenestration is typically marked with metal clips (MCs) to create easy access to the common atrium (CA) for standard diagnostic catheters, but those can make the passage of a long sheath challenging due to the rigidness of the edges caused by the MC. Patients with Fontan circulation have a high risk of developing atrial tachycardias (ATs) due to surgical scarring and due to pressure and volume overload.[3] Atrial arrhythmias in Fontan patients may lead to haemodynamic deterioration and are associated with substantial morbidity and mortality.[4] Most ATs after the Fontan procedure are drug refractory; therefore, catheter ablation is an alternative therapy option.

Here, we report a successful radio-frequency catheter ablation (RFA) of re-entry AT in an adolescent with HLHS after Fontan completion with challenging access through the clip-marked fenestration of the IALT.

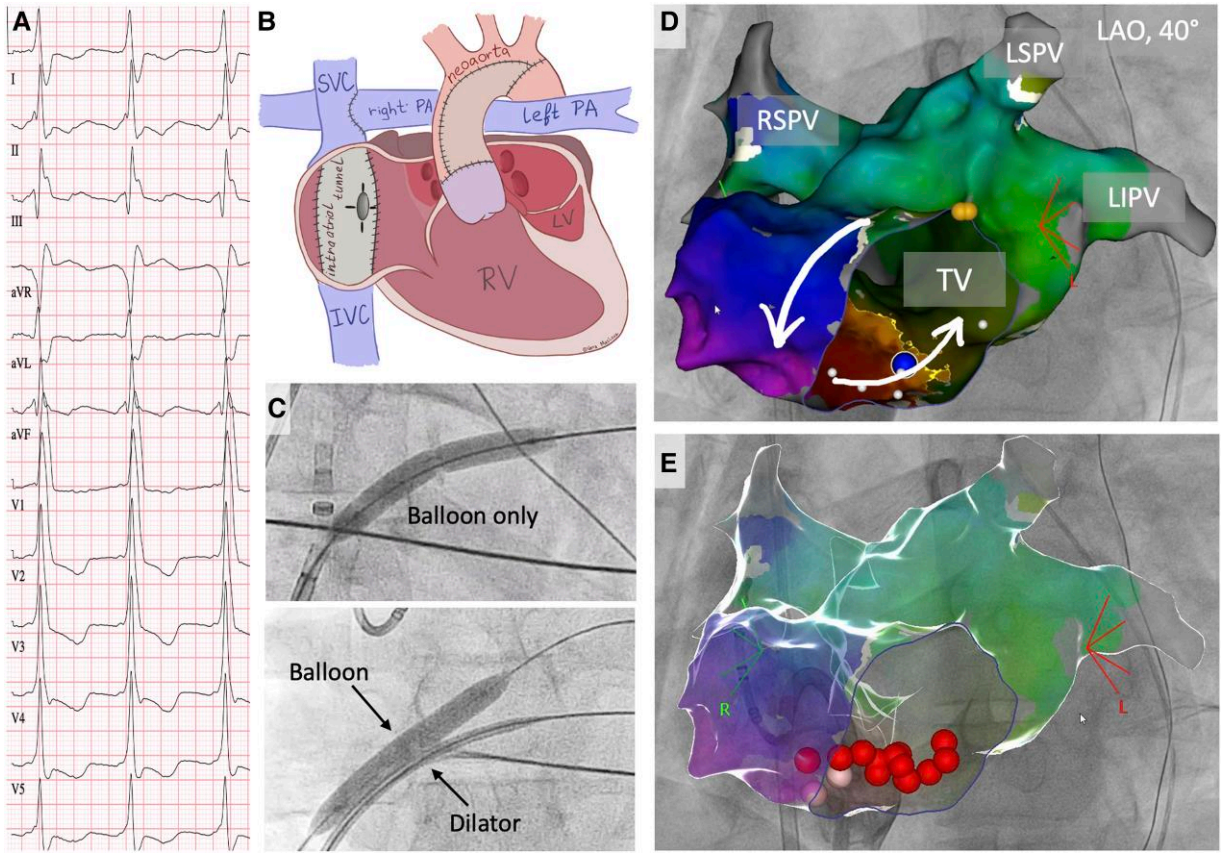
Summary figure

Case presentation

A 19-year-old male was scheduled for RFA of symptomatic and recurrent AT (*Figure 1*), resistant to anti-arrhythmic therapy with a beta-blocker and flecainide. He was prenatally diagnosed with HLHS and underwent a stepwise surgical palliation with modified Norwood procedure, hemi-Fontan operation, and, finally, Fontan completion with the creation of a fenestrated IALT using a polytetrafluoroethylene (PTFE) patch. The fenestration had a size of 4 mm. It was marked by four MCs, normally used for vessel closure (*Figure 2*).

The ablation procedure was performed under deep sedation with fentanyl and propofol. At the beginning of the procedure, the patient presented with ongoing clinical AT. The cannulation of the coronary sinus (CS) was not possible, because the CS orifice was located in the CA. Therefore, the 10-pole diagnostic catheter (DECANAV®, Biosense Webster) was placed at the lateral wall of the IALT to obtain the referent signal.

Due to the small diameter of the fenestration, the ablation catheter (THERMOCOOL SMARTTOUCH™ SF, Biosense Webster) could only be inserted into the CA. However, the position of the fenestration did not allow to reach reliable contact, especially in the areas around the tricuspid valve (TV). Insertion of the multi-polar mapping catheter (PENTARAY®, Biosense Webster) and steerable sheath (Agilis™, Abbott) was also not possible. The tip of a steerable introducer was repeatedly jammed by the edges of fenestration. Multiple attempts to dilate the fenestration using a 6 mm peripheric angioplasty balloon (Charger™, Boston Scientific) failed due to the rigidness of the edges of the fenestration caused by the MC. The inflated balloon remained in the characteristic



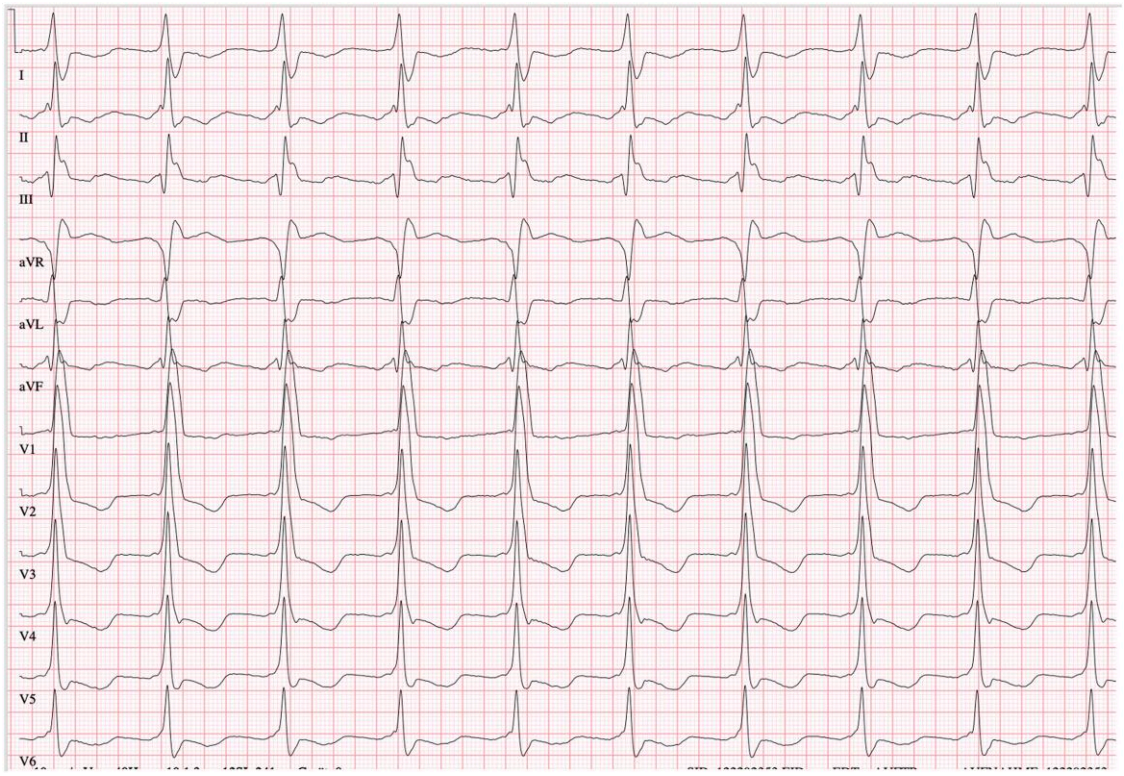


Figure 1 The baseline electrocardiogram of atrial tachycardia with heart rate 115 b.p.m., 2:1 atrioventricular conduction. Speed 50 mm/s.

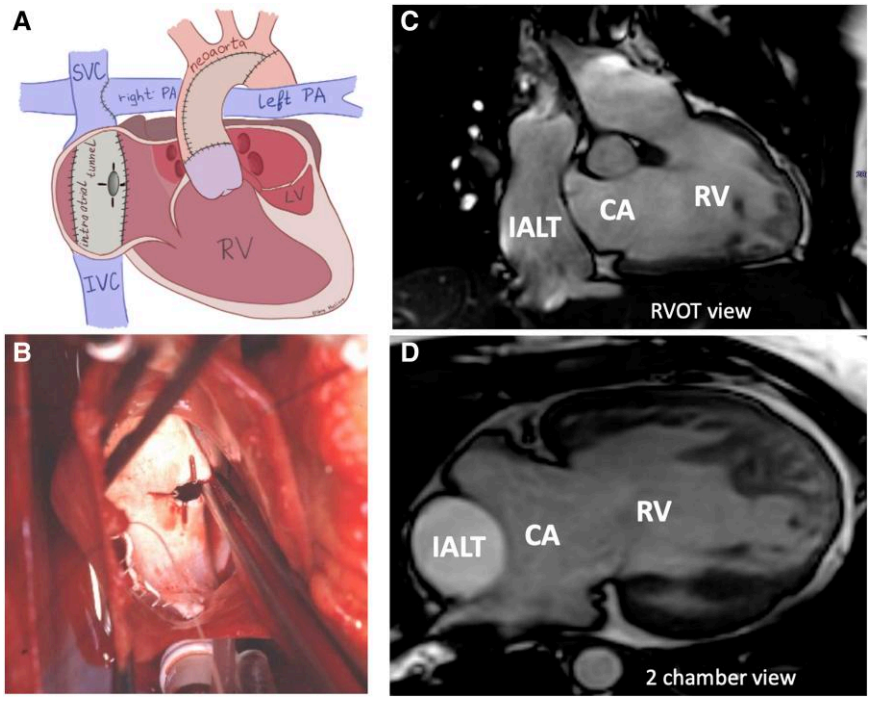


Figure 2 (*A*) Surgical correction, performed in our patient. (*B*) Intra-atrial lateral tunnel with fenestration, marked by four metal clips. (*C*, *D*) Cardiac magnetic resonance imaging, showing the Fontan circulation. CA, common atrium; IALT, intra-atrial lateral tunnel; IVC, inferior vena cava; LV, left ventricle; PA, pulmonary artery; RV, right ventricle; RVOT, right ventricular outflow tract; SVC, superior vena cava.

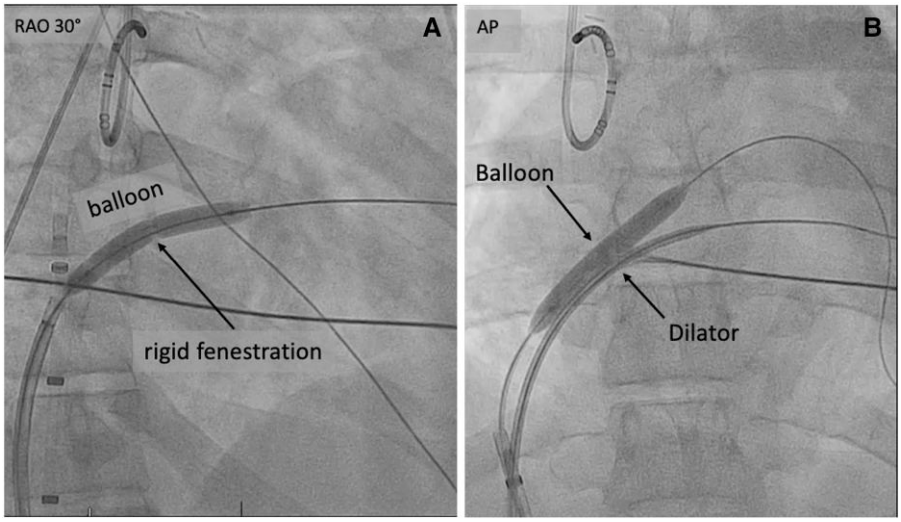


Figure 3 Angioplasty of the fenestration. (*A*) Incomplete inflation of the balloon. (*B*) Complete inflation of the balloon, using the balloon against a dilator technique. AP, anterior–posterior projection; RAO, right anterior oblique projection.

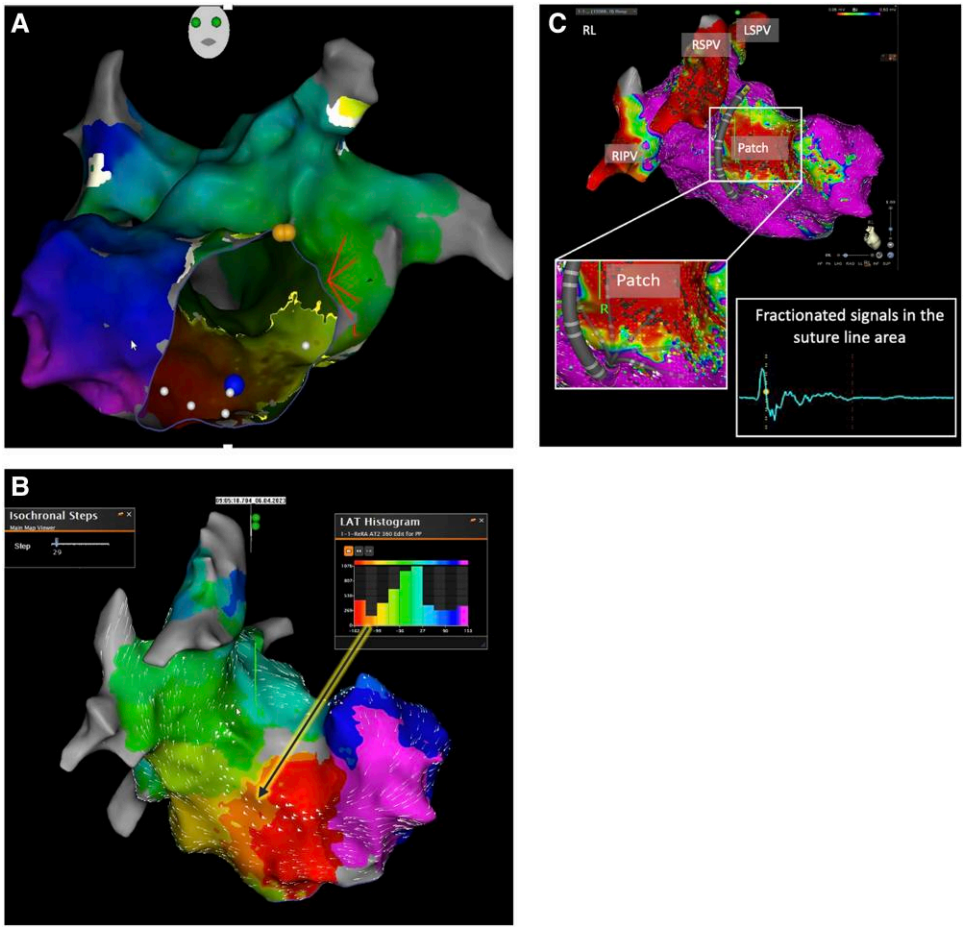


Figure 4 (*A*) Activation map of the atrial tachycardia, showing re-entry in the common atrium. (*B*) Isochronal map and local activation time histogram, showing the slowest conduction in the orange area (arrow). (*C*) Voltage map of the common atrium in right lateral projection, showing the fractionated signals in the suture line area of the patch. LSPV, left superior pulmonary vein; LAT, local activation time; PA, pulmonary artery; RIPV, right inferior pulmonary vein; RSPV, right superior pulmonary vein.

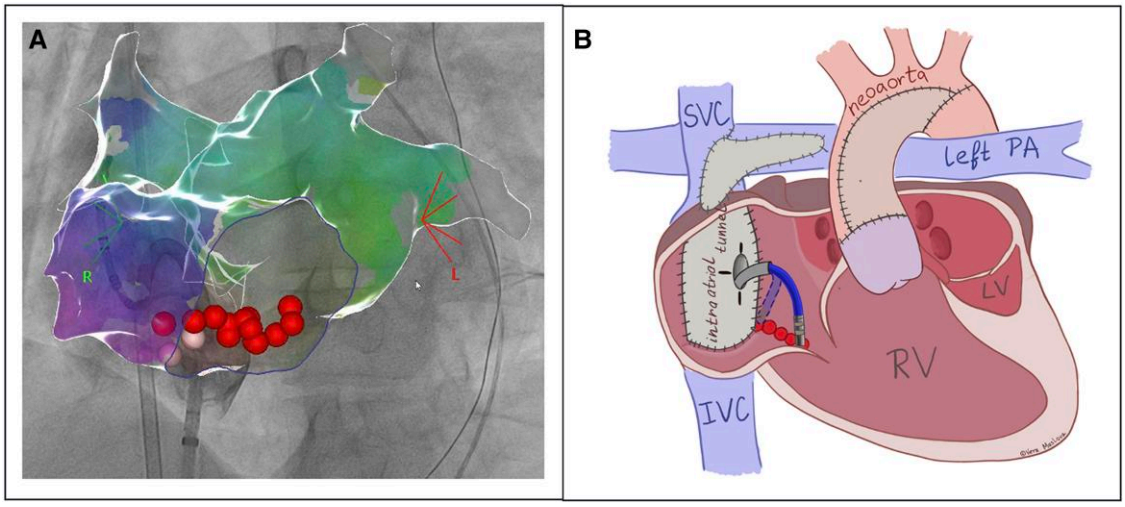


Figure 5 (*A*) Ablation points (red dots) in cavotricuspid isthmus area. (*B*) Schematic picture of the ablation line between the intra-atrial tunnel and tricuspid valve.

dumbbell-shape despite the use of high pressures (up to 16 atm) (*Figure 3A*). Only angioplasty with the 'balloon-against-dilator' technique was finally successful and allowed complete inflation of the balloon, dilation of the fenestration, and insertion of the steerable introducer into the atrium (*Figure 3B*). For this, two J wires were inserted to the CA through the fenestration. The dilator of the steerable sheath was then positioned over the first J wire, ensuring its placement within the fenestration. Once in position, the balloon was inflated with moderate pressure, not exceeding a maximum of 12 atmospheres exerting pressure against the dilator. This resulted in a localized, asymmetrical rise in radial pressure and dilation of the orifice despite the mechanical resistance of the MCs.

Activation mapping of the AT (CARTO3 V7; Biosense Webster, Diamond Bar, USA) revealed a counterclockwise re-entry around the TV. In our case, not only the right atrium (as typical in atrial flutter) but also the whole CA was included in the re-entry circuit. The zone of the slowest activation during the AT was located between the PTFE patch and the tricuspid annulus, corresponding anatomically to the cavotricuspid isthmus (CTI) (*Figure 4*; Supplementary material online, *Video S1*). After linear radio-frequency application at the isthmus abolished the AT, no other AT was inducible (*Figure 5*). A CTI block was confirmed. One week later, the patient experienced a different AT (longer cycle length = 405 ms with 1 to 1 AV conduction and different P-wave morphology), which was successfully terminated by electrical cardioversion. Over the following 9 months, the patient remained free of arrhythmias.

Discussion

Here, we present a case of challenging access to a CA through a clipped fenestration in a PTFE baffle for AT ablation.

Although we could insert the ablation catheter through the fenestration into the CA before dilation, good tissue contact could not be reached. The insertion of the multi-polar mapping catheter was, however, necessary for performing the complete map and understanding the atrial anatomy and mechanism of the AT in a patient after complex cardiac operations. Therefore, the decision to dilate the fenestration was made.

Tunnel fenestrations in Fontan patients typically have a size of 4 mm. They allow shunting from the tunnel to the CA and decompression of the Fontan pathway but can also be very useful in patients with AT. However, there are no recommendations regarding the balloon size or pressure for dilation of the fenestration in cases where it is too small. In one experimental *in vitro* study that assessed the dilatation of PTFE baffle fenestrations this size, usually 6–7 atm pressure was enough for complete balloon inflation,[5] but this study was performed in non-clipped fenestrations.

In our case, dilation of the fenestration with a 6 mm incompliant angioplasty balloon was unsuccessful due to the rigidness of the clips despite the use of the maximum recommended pressure (16 bar) for the applied balloon.

The successful dilation of the balloon was reached, when at first the dilator from the steerable sheath was inserted through the fenestration, and then, over a second wire the balloon. Using the balloon-against-dilator technique, the balloon could be leaned on the hard dilator. This resulted in an increased focal instead of radial pressure against the baffle and was followed by successful dilation.

Prediction of the structural damage to the fenestration in our case was not possible. Imaging did not reveal whether the orifice maintained a circular configuration after dilation, or it was torn. Previously mentioned *in vitro* experiments showed that baffles can be torn using balloons of larger size (8–9 mm), but no excessive tears of baffle or excessive enlargements of fenestration were reported. There is one case report about an uncontrolled tear of a baffle, created from native atrial tissue using a 16 mm balloon.[6] This caused the patient's death after the procedure.[6] The tear was explained due to easy stretchability and subsequent recoil of the native atrium. To the best of our knowledge, there are no reports about uncontrolled tearing of synthetic baffles. In our patient, dilation (or tear) of fenestration had no negative consequences for the patient. Transthoracic echocardiogram 3 months after the procedure revealed unchanged flow through the fenestration; there was no deterioration of New York Hear Association Class, decrease of oxygen saturation, and quality of life.

Conclusion

Even though the tunnel fenestration during the Fontan completion facilitates access to the CA, the passage of a steerable introducer with a mapping catheter may be challenging due to diameter mismatch and the rigidity of its edges caused by MC markers. The balloon-against-dilator

technique might be helpful when conventional balloonoplasty fails, but potential risks have to be taken into account.

Lead author biography

Vera Maslova is a cardiology resident at University Hospital Kiel, Germany. After graduating from the First Moscow State Medical University, Russia, she continued her career in Germany. Here, she follows her fascination for electrophysiology and strives to specialize her career in this field.

Supplementary material

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

Consent: The authors declare that written consent was acquired from the patient for the submission and publishing of this case report, including images and textual content, in accordance with COPE guidelines.

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Data availability

The data used to support the findings of this case report are included in this manuscript.

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