A 47-year-old woman with tricuspid atresia was referred for catheter ablation. After a palliation by Blalock–Taussig followed by an atriopulmonary Fontan surgery, the patient had a conversion to extra-cardiac total cavopulmonary connection in 1999 for Fontan obstruction. She then underwent a dual-chamber epicardial pacemaker implantation associated with Maze ablation in 2003 for sinus dysfunction and paroxysmal symptomatic atrial fibrillation. She presented with recurrent uncontrolled episodes of symptomatic atrial arrhythmia documented on 12-lead electrocardiogram. Cardiac echocardiogram revealed a unique ventricle with preserved function, and a dilated atrium. Pre-procedure intra-cardiac thrombus was ruled out by computed tomography scan.

A reference decapolar catheter was placed in the pulmonary artery. The clinical arrhythmia was easily inducible with a stable tachycardia cycle length of 260 ms. Trans-Gore-Tex tube puncture was performed as previously described. Multiple dilatations up to a 6-mm cutting balloon were required to advance a SL-0 sheath on the wire through the conduit into the atrium. The sheath and dilator were then exchanged for an Agilis 8.5-Fr steerable sheath (Agilis NxT™, Abbott) (Figure 1A). Bi-atrial activation mapping using a high-density mapping catheter (Pentaray, Biosense) revealed a macro intra-atrial re-entrant tachycardia around the atretic tricuspid annulus identified by a small scar area near to the His signal (Figure 1B, propagation map video is provided in Supplementary material online). Entrainment manoeuvres confirmed area between the atretic tricuspid annulus and the scar where the inferior vena cava was oversewn to be part of the circuit (post-pacing interval / tachycardia cycle length = 8 ms), and we decided to perform a linear ablation in this isthmus. The cycle length slowed and arrhythmia then terminated during the second radiofrequency application. At the end of the procedure, the conduction block was confirmed by activation mapping, and no further atrial arrhythmia was inducible. The patient had no recurrence after 3 months.

Compared with other congenital heart diseases, atrial arrhythmia circuits in patients with Fontan palliation more often involve non-cavooannular isthmus. However, in the specific population of patients with extra-cardiac conduits, a recent study has reported that the isthmus between the atrioventricular valve annulus and the oversewn inferior vena cava was involved in most cases. In this case, despite the
absence of tricuspid valve due to tricuspid atresia, the arrhythmia circuit rotated around the atretic annulus identified by a small scar area. This line may be targeted during Maze procedure in patients with tricuspid atresia.

Supplementary material is available at Europace online.

Conflict of interest: none declared.

References

