Indian Pacing and Electrophysiology Journal 16 (2016) 201–204

Contents lists available at ScienceDirect

Indian Pacing and Electrophysiology Journal

journal homepage: www.elsevier.com/locate/IPEJ

Leadless pacemaker implantation in a patient with complex congenital heart disease and limited vascular access

Paolo Ferrero [a, b, *], Michael Yeong [a], Emilia D'Elia [b], Edward Duncan [a], Alan Graham Stuart [a]

[a] Bristol Heart Institute, Adult Congenital Heart Disease Department, University Hospital of Bristol, Bristol, United Kingdom
[b] Hospital Papa Giovanni XXIII, Cardiovascular Department, Bergamo, Italy

ARTICLE INFO

Article history:
Received 21 August 2016
Accepted 21 October 2016
Available online 26 October 2016

Keywords:
Adult congenital heart disease
Leadless pacemaker
Univentricular physiology

ABSTRACT

Management of rhythm related issues might be particularly challenging in patients with congenital heart disease due to complex anatomy and restricted vascular access. The leadless technology appears a suitable and attractive alternative for this population. We describe a patient with single ventricle physiology who successfully underwent implantation of a leadless pacemaker.

1. Introduction

Rhythm related issues are common in adults with congenital heart disease (ACHD).

Endocardial pacemaker lead implantation in congenital patients with complex anatomies may be particularly challenging, owing to restricted or abnormal venous access to the heart. In addition to the epicardial approach, which may be problematic after multiple sternotomies, many innovative strategies have been devised to offset difficulties with venous access to the heart, including hybrid approaches with endocardial trans-atrial lead deployment [1].

The introduction of the leadless intracardiac pacemaker has enabled the implantation of an active-fixation leadless device in patients who require permanent single-chamber ventricular pacing. This technology has been proven to be effective and safe in initial clinical trials and it has many theoretical attractions in ACHD patients with complex anatomy [2–5].

We describe a patient with a single ventricle, complex congenital heart disease and paroxysmal atrioventricular conduction block who had no superior caval vein access to the heart and was unsuitable for epicardial pacing, but who underwent successful implantation of a leadless pacemaker (Medtronic Micra TM) using the left femoral vein.

2. Case report

The patient was a 47 years old lady who presented with cyanosis in infancy. Her underlying cardiac diagnosis was atrial situs solitus with atrioventricular concordance, a superior-inferior ventricular arrangement, uncommitted ventricular septal defect, arterial malposition with the anterior aorta arising from the right ventricle, and pulmonary atresia. She underwent a left modified Blalock Taussig (BT) shunt aged 6 years, a Waterston shunt aged 7 years and right BT shunt aged 28 years. In 1995, the Waterston and right BT shunts were taken down and a classical Glenn anastomosis was performed. The post surgical anatomy is demonstrated in Fig. 1 and Fig. 2.

During follow up, she developed atrial tachyarrhythmia and symptomatic paroxysmal AV block. In view of multiple previous sternotomies, she was felt unsuitable for epicardial pacing. The right femoral vein was occluded and she had no superior caval venous access to the heart, so a dual chamber pacemaker was implanted using the left femoral vein (Advisa Medtronic DDDR®: Select Secure 85 cm atrial lead and Capsure Fix 85 cm ventricular lead Medtronic®). The entire system was removed two months later due to wound infection. She then underwent implantation of a second endocardial DDD pacemaker system by direct double transatrial puncture after access was obtained by sternotomy with X ray screening to position the leads. The surgical exposure was complicated by multiple adhesions and calcification and it took 2 h to find a suitable entry point to the systemic venous atrium. The

* Corresponding author. Adult Congenital Heart Disease, Bristol Heart Institute, Upper Maudlin St, Bristol BS2 8HW, United Kingdom.
E-mail address: pferrero@asst-pg23.it (P. Ferrero).

http://dx.doi.org/10.1016/j.ipej.2016.10.007

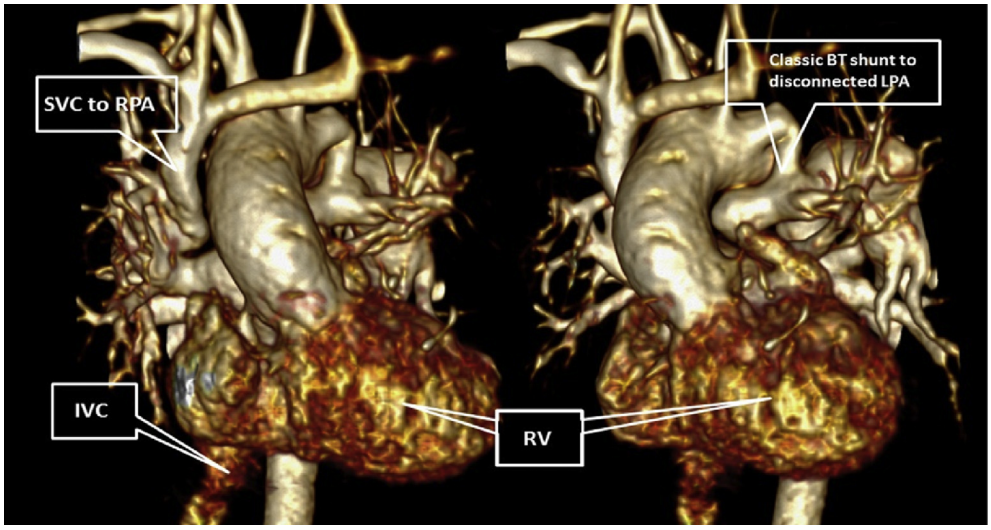

Fig. 1. Magnetic resonance imaging (MRI) 3D reconstruction showing the anatomic details.

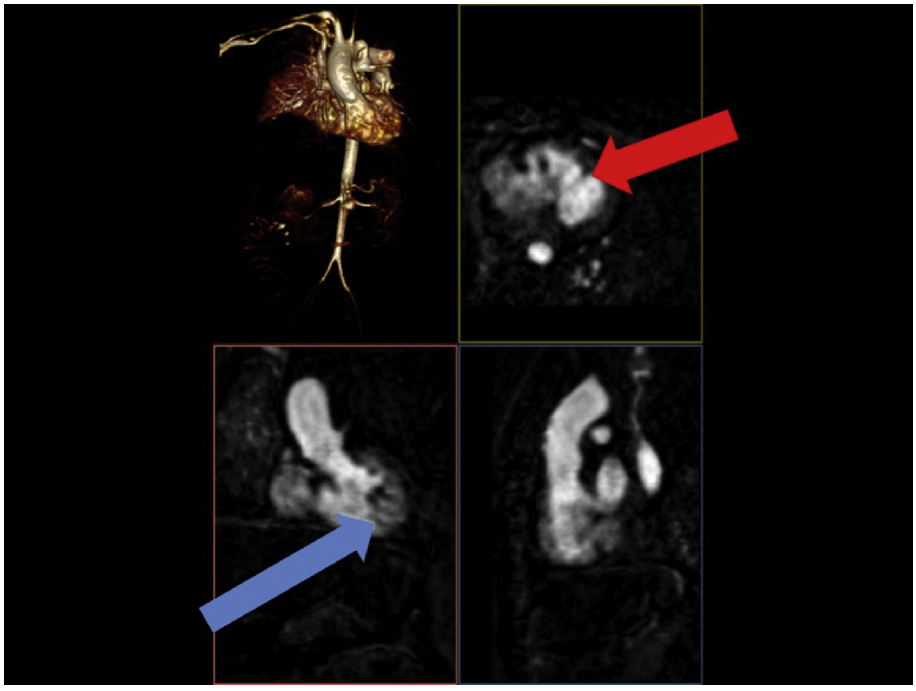

Fig. 2. MRI 3D reconstruction and cross-sectional images showing the ventricular mass arrangement, ventricular septal defect (red arrow) and RV apex (blue arrow).

pacing leads (Select Secure 74 cm Medtronic) were tunnelled to the abdomen and attached to a dual chamber generator (Advisa Medtronic DDDR®). However, after 3 months she developed muscle twitching and loss of atrial capture. A chest X-ray demonstrated that the atrial lead had retracted through the tunnelled access and was lying underneath the abdominal generator. The device was reprogrammed to VVIR mode pacing, but six months later the patient developed further dizzy spells. Chest X-ray and device interrogation unveiled complete ventricular lead dislocation. Owning to recurrent symptoms a leadless device implantation procedure was planned. The procedure was carried out under general anaesthesia and under fluoroscopic and transoesophageal (TOE) guidance. Access to the left femoral vein was achieved and dilators of increasing size were passed over a stiff wire to 24 F, allowing final advancement of the 27 F Micra sheath (Medtronic®) to the inferior caval vein-systemic venous atrium junction (Fig. 3A and C). After systemic anticoagulation with unfractionated heparin achieving an ACT of around 300 s, the Micra device mounted on the dedicated steerable delivery catheter was advanced through the right atrioventricular valve to the peri-apical portion of the right ventricle and gently pushed obtaining the classical gooseneck shape. After confirming the contact of the device with the ventricular trabeculae by contrast angiography, the delivery system was partially withdrawn to check the device stability by gently pulling (Fig. 3B and C). The sensing was around 7 mV, the threshold was repeatedly below 0.8 V/1 ms, hence the device was successfully deployed in the final position. No procedural complications occurred. The chest X-ray and transthoracic echo were repeated the following day showing a

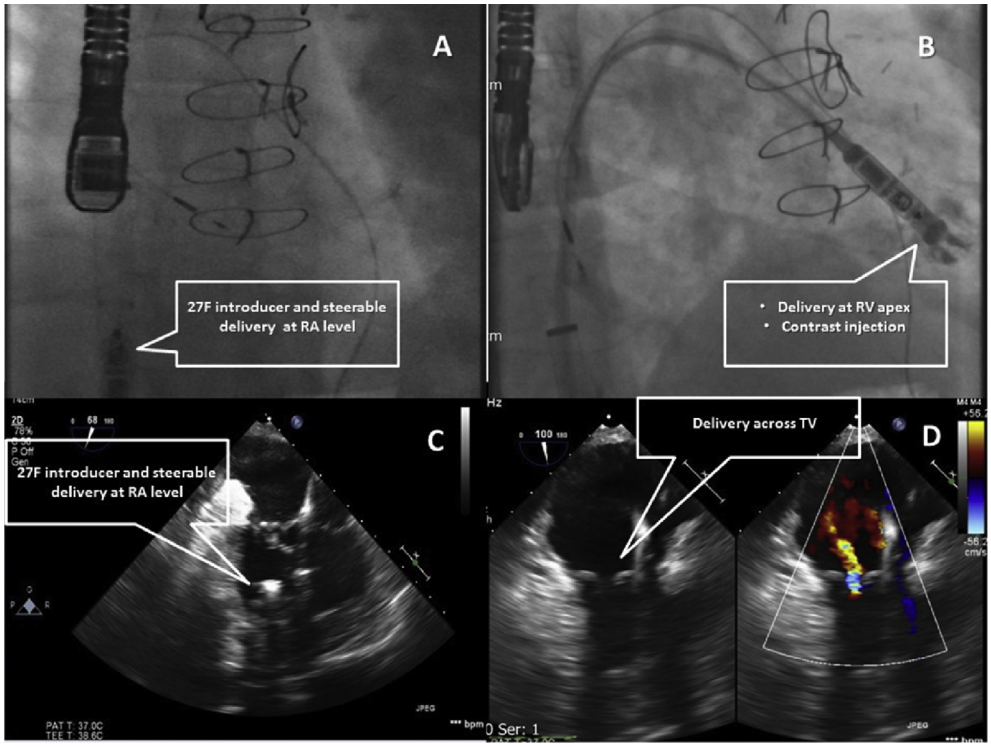

Fig. 3. Panel A: Fluoroscopy image showing the Micra dedicated steerable delivery at the inferior vena cava-right atrium junction. Panel B: Fluoroscopy image showing the Micra dedicated steerable delivery at the target deployment site. Panel C: Corresponding cross-sectional transoesophageal echo image. Panel D: Corresponding cross-sectional trans-oesophageal echo image, depicting the delivery shaft across the tricuspid valve (TV).

stable position of the device with sensing of 7.5 mV and threshold of 0.6 V@1 ms.

The pacing parameter were stable at three months follow up with a sensed R wave amplitude around 8 mV and a threshold of 0.5 V @ 1 ms. No infective or embolic complications occurred.

3. Discussion

Management of symptomatic bradycardias in patients with complex congenital heart disease is particularly challenging due to several factors including complex anatomy, lack of venous access to the heart due to venous occlusion or previous surgery and an increased risk of infection and right to left emboli.

Miniaturization of pacemaker batteries has made it possible to contemplate deployment of a device with direct contact to the endocardium of the right ventricle allowing direct cardiac electrical capture. Percutaneous implantation of an entirely intracardiac leadless pacemaker has been recently demonstrated to be feasible and safe in humans, assuring stable pacing performance in a large cohort of patients at one year follow-up [2–5].

Although the leadless pacing technique appears to be a potential option for patients not suitable for conventional endocardial lead implantation or at prohibitive risk for epicardial pacing, there are few reports of its use in the ACHD population and no previous reports of leadless pacemaker use in patients with a single ventricle and complex atrioventricular valve anatomy [3]. The possibility to deploy the device from the femoral vein, without the need to create a pocket and tunnelling the pacing leads, is a major theoretical benefit in ACHD patients who may have limited vascular access, a bleeding diathesis associated with chronic cyanosis, and the potential for right to left emboli from leads. The avoidance of venous occlusion by pacing leads is an additional potential advantage in allowing future percutaneous therapy and optimised cardiac output.

The use of leadless pacemaker technology has led to concern regarding the risk of cardiac perforation due to erosion. However in the Leadless trial the rate of cardiac perforation was around 1.5%, which was not significantly higher than the rate observed with trans-venous leads [2,5]. Our patient displayed several peculiarities which made conventional pacing throughout both the endocardial and epicardial route particularly challenging. Firstly, our patient underwent univentricular palliation by Glenn cavo-pulmonary derivation, making femoral venous access the only available trans-venous route to achieve endocardial pacing. Secondly, she had undergone five previous sternotomies or thoracotomies, making the risk of a new surgical dissection particularly high. Furthermore, the pericardial adhesions might have reduced the likelihood of finding an epicardial pacing site with acceptable thresholds. Finally, a previous hybrid approach of trans-atrial endocardial lead implantation had failed, discouraging further attempts.

In this patient the complex ventricular and atrioventricular valve anatomy was clearly delineated during leadless pacemaker implantation using a combination of fluoroscopy and TOE echocardiography. In this setting, we would recommend using multimodality imaging.

In conclusion, to our knowledge this is the first report of a leadless pacing in a patient with complex ACHD and univentricular physiology. We believe that this may prove an extremely useful technique in patients with complex congenital heart disease who require pacemaker implantation.

References

[1] Sherwin ED, Triedman JK, Walsh EP. Update on interventional electrophysiology in congenital heart disease: evolving solutions for complex hearts. Circ Arrhythm Electrophysiol 2013;6:1032–40.
[2] Reddy V, Exner DV, Cantillon DJ, Doshi R, Bunch TJ, Tomassoni GF, et al.

LEADLESS II study investigators. Percutaneous implantation of an entirely intracardiac leadless pacemaker. N Engl J Med 2015;373:1125–35.
[3] Wilson DG, Yue A, Roberts PR, Morgan JM. Leadless pacing: the old with the new. Int J Cardiol 2016;203:407–8.
[4] Khairy P, Landzberg MJ, Gatzoulis MA, Mercier LA, Fernandes SM, Côté JM, et al. Transvenous pacing leads and systemic thromboemboli in patients with intracardiac shunts. A multicenter study. Circulation 2006;113:2391–7.
[5] Reynolds D, Gabor Z, Duray GZ, Omar R, Soejima K, Neuzil P, et al., Micra Transcatheter Pacing Study Group. Leadless intracardiac transcatheter pacing system. N Engl J Med 2016;374:533–41.