Transvenous pacing in complex post-operative congenital heart disease guided by angiography: A case report

Jayaprakash Shenthar a,*, Maneesh K. Rai a, Tammo Delhaas b

a Electrophysiology Unit, Department of Cardiology, Sri Jayadeva Institute of Cardiovascular Sciences and Research, Bengaluru, India
b Department of Biomedical Engineering, Maastricht UMC+, Universiteitskliniek 50, Room 3366, 6229 ER, Maastricht, the Netherlands

ARTICLE INFO

Article history:
Received 27 August 2018
Received in revised form 11 November 2018
Accepted 12 November 2018
Available online 16 November 2018

Keywords:
Congenital heart disease
Transvenous pacing
Angiography
Complex congenital heart disease
Post-operative complex congenital heart disease

ABSTRACT

Transvenous pacing in patients with postoperative complex congenital heart disease (CHD) can be challenging and pose technical challenges to lead placement because of the complex anatomy, distortions produced by the surgical procedures, and the altered relationship of cardiac chambers. We describe the utility of angiography for transvenous dual chamber pacemaker implantation in a post-operative complex congenital heart disease.

Copyright © 2018, Indian Heart Rhythm Society. Production and hosting by Elsevier B.V. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

1. Introduction

Permanent pacemaker implantation is indicated in patients with congenital heart disease (CHD) with associated sinus node dysfunction or atrioventricular block that is either de novo or if it occurs following surgical correction. Pacing therapy in CHD reduces morbidity and mortality [1]. Epicardial pacing which is usually preferred in pediatric patients tends to fail earlier, and more often than transvenous leads regardless of steroid-eluting characteristics [2]. Transvenous pacing is an important option in patients with CHD because of its better long-term performance compared to epicardial pacing [3]. Before implanting a permanent transvenous pacemaker in complex CHD, the implanting physician needs to have a complete understanding of the abnormal cardiovascular anatomy using an imaging technique [4]. Though pre-procedure computed tomography (CT) is an option, there are concerns of cancer risk in children with congenital heart disease, especially females, which require repetitive high-exposure imaging (catheterization and computed tomography) due to radiation exposure [5]. We illustrate the utility of angiography instead of CT as a pre-procedure aid in guiding transvenous dual chamber pacemaker implantation in a complex post-operative CHD patient with failed surgically implanted epicardial lead.

2. Case report

A 13-year-old female weighing 24 kg, with abdominal situs inversus, levocardia, total anomalous pulmonary venous connection to the coronary sinus, atrioventricular discordance and double outlet right ventricle and surgically corrected subpulmonic ventricular septal defect, implanted with a permanent pacemaker for postsurgical AV block, presented with five episodes of syncope within one month. At the age of 15 months, she had undergone arterial switch, atrial septectomy, Senning procedure to reroute pulmonary venous drainage through the mitral valve, and patch closure of the ventricular septal defect as per the detailed operative records available with the patient. A single chamber (VVI) pacemaker with an epicardial lead was implanted for postoperative AV block, and she had a pulse generator replacement at the age of 11 years. At presentation, pacemaker interrogation revealed a threshold of 5 V at 1.5-ms pulse width, and lead impedance of 1256 Ω. Holter monitoring demonstrated intermittent loss of capture during episodes of
syncope. She underwent an evaluation by the pediatric cardiologist with a detailed echocardiographic evaluation that revealed normal biventricular function, and confirmed the diagnosis. With a decision to implant a transvenous dual chamber pacemaker, bilateral upper limb venogram performed with 5 ml non-ionic (Iohexol) contrast revealed patent upper limb veins with a single left-sided superior vena cava. Through a left infraclavicular incision, two separate venous accesses were obtained by extrathoracic axillary vein punctures. Attempt to negotiate the leads into the venous chamber was unsuccessful because of the inability to visualize the complex venous

Fig. 1. A) SVC angiography demonstrating the IVC to the left of the spine and also the major cardiac silhouette to the left of the spine indicating abdominal situs inversus and levocardia. The venous blood drains to the morphologic left atrium (MLA). B) Angiography reveals the acute angulation of the baffle crossing the spine from left to right to enter the morphological left atrium with the absence of atrial appendage as depicted in the cartoon E. C) morphological left atrium is connected to the morphological right ventricle (MRV) which drains into the pulmonary artery as depicted in cartoon E. D) shows levophase of the angiogram demonstrating the morphological right atrium (MRA) draining into smooth-walled morphologic left ventricle (MLV) as depicted in the cartoon F.

Fig. 2. A) and B) the course of the renal catheter through the complicated route in to the right ventricle in AP view, C) and D) shows LAO and shallow RAO views demonstrating the ventricular lead in the mid-septal position in the right ventricle and the atrial lead in the venous atrium. The tortuosity and the complex course taken by the leads can be appreciated.
anatomy. A 6 F pigtail catheter was positioned through one of the venous accesses and positioned in the superior vena cava. Thirty ml of non-ionic intravenous contrast (Iohexol) was injected at 15 ml/sec using a flow injector, and cine films were acquired in anteroposterior view at 30 frames/second. Review of the acquired films revealed an acute rightward course of venous drainage from the superior vena cava to the morphologic left atrium across the spine and then a leftward course to the morphologic right ventricle resulting in a double bend (Fig. 1). The tortuous course was negotiated using two 6 F angiographic catheters with a renal curve (Medtronic Inc. Minneapolis, MN, USA) and a Terumo 0.035” Glide wire® (Terumo interventional systems) to gain access to the venous atrium and ventricle. The Terumo 0.035” Glide wire was exchanged for 0.035” extra support Teflon guidewires and two 7F peel away introducers (Medtronic Inc. Minneapolis, MN, USA), were advanced into the respective chambers and 7F active fixation lead (model 4076, Medtronic Inc., Minneapolis, MN, USA) was deployed successfully into the venous chambers with minimal manipulation (Fig. 2). The measured parameters were R wave of 12 mV, a threshold of 0.6 V at 0.5 ms pulse width, and an impedance of 780 Ohms in the ventricle, and a P wave of 3.4 mV, a threshold of 0.5V at 0.5 ms pulse width and an impedance of 550 Ohms in the atrium. A dual chamber pulse generator (Model Relia DDD, Medtronic Inc. Minneapolis, MN, USA) was connected to the leads and placed in a subpectoral pocket, and the abdominal pulse generator was explanted. Post-procedure 12-lead ECG revealed atrial sensed and ventricular paced complexes with inverted P-waves in lead I (red arrow heads) indicating atrial situs inversus. The QRS complex in lead I is upright with left bundle and left axis configuration with normal QRS progression in the chest leads preceded by a pacing spike (black arrows) in lead II indicating right ventricular apical pacing. The QRS shows normal progression across the chest leads indicating levocardia. At one year follow up patient is doing well and asymptomatic with preserved biventricular function (see Fig. 3).

3. Discussion

This case highlights the utility of angiography in defining the complex anatomy that is encountered by an implanter attempting to perform transvenous permanent pacemaker implantation in...
surgically corrected congenital heart disease. Angiography reduces the procedure complexity and fluoroscopic exposure, and assists simplified transvenous lead placement in complex anatomy.

Transvenous pacemaker implantation is an option when epicardial leads implanted at the time of surgery fails over the long-term [3,6]. Repaired congenital heart disease is not synonymous with a "normal heart" because the postoperative state introduces many technical challenges to the implanter attempting a pacemaker or ICD implantation [7]. Device implanters mostly rely on fluoroscopy to guide the positioning of the atrial and ventricular leads during the implant procedure. Fluoroscopy shows only the cardiac silhouette without details of the complex intracardiac anatomy and the altered relationship of the various chambers in patients with complex anatomical abnormalities. Fluoroscopy alone may not be helpful to the implanter in patients with complex anatomy. In a patient with a complex anatomy CT or MRI is usually done before pacemaker implantation as the venous and ventricular anatomy can be complex and variable as is shown in the case. Angiogram at the time of implantation is considered a bail out procedure. Various other imaging modalities such as echocardiography and CT scan have been used to aid the implanters in patients with CHD. The use of intraprocedural echocardiography for guidance has the disadvantage of the need for extra personnel, the concern of compromising asepsis near the field of the procedure, and the inability to demonstrate the tortuosity and angulations accurately in the venous anatomy. Though CT scan and MRI can define the complex anatomy, it needs to be done in advance and adds additional radiation to young patients who may require repeated or additional procedures. The advantages of angiography are that it does not require additional equipment or personal, can be performed at the time of implantation with the available equipment, is available for immediate review, displays complex anatomy for repeated review, and adds very little to the fluoroscopy time. However, there are two caveats to this method: (a) there should be no contraindication to angiography, and (b) the operator should be familiar with the angiography anatomy of the altered cardiac morphology.

In conclusion, we demonstrate the utility of angiography during dual chamber pacemaker implantation in complex CHD as an aid to facilitate the implantation in a patient with complex anatomy.

**Funding sources**

None.

**Conflicts of interest**

None.

**References**

[1] Batra AS, Balaji S. Pacing in adults with congenital heart disease. Expert Rev Cardiovasc Ther 2006;4:663–70. https://doi.org/10.1586/14779072.4.5.663.

[2] Fortescue EB, Berul CI, Cecchin F, Walsh EP, Triedman JK, Alexander ME. Patient, procedural, and hardware factors associated with pacemaker lead failures in pediatrics and congenital heart disease. Heart Rhythm 2004;1:150–9. https://doi.org/10.1016/j.hrthm.2004.02.020.

[3] Silvetti MS, Drago F, Di Carlo D, Placidi S, Brancaccio G, Carotti A. Cardiac pacing in pediatric patients with congenital heart defects: transvenous or epicardial? Europace 2013;15:1280–6. https://doi.org/10.1093/europace/eut029.

[4] Khairy P, Van Hare GF, Balaji S, Berul CI, Cecchin F, Cohen MF, et al. PACES/HRS expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease. Heart Rhythm 2014;11:e102–65. https://doi.org/10.1016/j.hrthm.2014.05.009.

[5] Johnson JN, Hornik CP, Li JS, Benjamin DK, Yoshizumi TT, Reiman RE, et al. Cumulative radiation exposure and cancer risk estimation in children with heart disease. Circulation 2014;130:161–7. https://doi.org/10.1161/CIRCULATIONAHA.113.005425.

[6] Sachweh JS, Vazquez-Jimenez JF, Schönürbe Fa, Daebritz SH, Dörg H, Mühler EG, et al. Twenty years experience with pediatric pacing: epicardial and transvenous stimulation. Eur J CardioThorac Surg 2000;17:455–61.

[7] Mond HGKP. Tricks of the trade. In: Mond HG, Karpawich PP, editors. Pacing options adult patient with congenit. Hear. Dis. 2007th. Malden, Massachusetts, USA: Blackwell Publishing; 2007. https://doi.org/10.1002/9780470750940.part1.1–1.