Transvenous pacing after the Mustard procedure: considering the complications

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Disturbances of rhythm and conduction in patients undergoing surgery for transposition of the great arteries have been widely reported. Some of these patients require implantation of a permanent pacemaker, especially those in whom symptomatic sick sinus syndrome is diagnosed. We present the case of a 29-year-old male corrected with a Mustard procedure, who received a pacemaker for progressive atrioventricular conduction disturbances and sinus node dysfunction, and we review the possible complications associated with transvenous pacemaker implantation in these patients. (*Neth Heart J* 2007;15:387-90.)

Keywords: transposition of the great arteries, pacemaker, complication, Mustard procedure

A 29-year-old male was born with a transposition of the great arteries, a large ventricular septal defect, valvular pulmonary stenosis and a patent ductus arteriosus. Because of progressive cyanosis a Rashkind septostomy was performed on the first day of life. At one year of age he underwent a Mustard operation including closure of the ventricular septal defect and ductus arteriosus as well as an open commissurotomy of the pulmonary valve. He was asymptomatic during regular follow-up at our outpatient clinic. However, recently, 28 years after his operation, he started complaining of progressive fatigue and dizziness. Echo-

cardiography revealed deterioration of his systemic right ventricular function. This was confirmed by magnetic resonance imaging (MRI), as a decrease in the ejection fraction from 48 to 37% within two years was noted. Subsequent Holter monitoring showed sinus rhythm with a prolonged PR interval up to 500 ms, periods of extreme sinus arrhythmia, sinus bradycardia and sinus arrests with frequent pauses of 2.6 sec. There were also episodes of atrioventricular junctional escape rhythm and high degree atrioventricular block with pauses of up to 5.4 sec. Because of the dizzy spells due to sinus node dysfunction in combination with atrioventricular conduction disturbances the patient underwent a transvenous double chamber pacemaker implantation. Through the left cephalic vein an active-fixation electrode was inserted and placed in the apex of the anatomic left (sub-pulmonary) ventricle. Subsequently, the atrial lead was inserted and screwed into the left atrial appendage. Sensing and pacing thresholds were excellent and a post-procedural X-ray showed adequate positioning of the leads (figure 1). The patient could be discharged uneventfully. At follow-up the patient had improved considerably and he did not have any complaints of fatigue or dizzy spells.

Discussion and literature review

Transposition of the great vessels accounts for 5 to 7% of all congenital cardiac disorders. Untreated, 90% of the infants born with this disorder will die within the first year of life. In 1959 Senning¹ described the first physiological surgical correction, the atrial switch procedure, which was modified in 1964 by Mustard.² Nowadays an anatomical correction is more preferred and this arterial switch procedure has largely replaced the atrial switch. However, the atrial switch procedure offered infants the possibility of reaching adulthood and at adult outpatient clinics we are now faced with late complications of this procedure, such as arrhythmias. After the Mustard operation, arrhythmia-free survival is 60 and 35% 10 and 20 years after repair, respectively.³ Most of these rhythm disturbances are identified as loss of sinus rhythm and atrial tachyarrhythmias and rarely as AV-node conduction disorder.^{4,5} The

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Figure 1. A) AP chest X-ray after pacemaker implantation to confirm the position of the pacemaker leads. The ventricular lead is situated in the anatomic left ventricle (LV), and the atrial lead in the left atrial appendage in the systemic venous baffle (SB). Ao=aorta, AP=pulmonary artery, RV=anatomic right ventricle. B) Lateral chest X-ray.

arrhythmias are probably related to the intra-atrial surgery that includes excision of the atrial septum and stitching of the intra-atrial baffle close to the atrioventricular node with damage to the sinus node and the artery to the sinus node.⁶ Pacemaker implantation is indicated for patients after Mustard repair who have a heart rate <30 beats/min or Stokes-Adams episodes, patients requiring pharmacological therapy for tachyarrhythmias, or patients with a poor systemic ventricular function and bradycardia.

The complexity of these patients warrants us to consider the possible procedural complications before transvenous pacemaker implantation (table 1). It is crucial to have exact knowledge about the anatomy. Old operation reports can give clues as to whether different adjunct operation techniques were used, whether the left atrial appendage is still in place, and whether there are any other structural variations. MRI can help in visualising the cardiac and vascular anatomy, which may guide the most suitable position for placing the leads (figure 2). Transvenous implantation is not possible if systemic venous baffle obstruction has occurred, which is currently in about 22% of patients after the Mustard operation.⁷ Venous obstruction, mostly in the left innominate vein, or the right or left subclavian vein, is present in 13.7% of patients scheduled for pacemaker implantation.⁸ It is therefore recommended to investigate the possibility of baffle or venous obstruction either by echocardiography, venography or intravenous digital subtraction angiography prior

to implantation. Intravascular stent placement may be applied before lead insertion, if systemic baffle or venous obstruction is demonstrated.⁹ There are no studies addressing the frequency of thrombosis within Mustard baffles after insertion of transvenous leads. The incidence of partial or total venous obstruction induced by transvenous leads is approximately 30 to 50% in adult patients early or late after pacemaker implantation, from which 1 to 3% of the patients are symptomatic.¹⁰ Figa et al. showed in children that the main risk factor for obstruction was found to be related to the total cross-sectional area of the lead as compared with the body surface area at implantation.¹¹ A ratio >6.6 mm²/m² was found to best predict venous obstruction. When selecting the pacemaker the leads,

Table 1. Key points for pacemaker implantation in a Mustard patient.

Abnormal anatomy, post surgery
Spontaneous systemic venous baffle and venous obstruction
Systemic venous baffle and venous obstruction after lead insertion
Lead positioning
Lead dislodgement
Ventricular rhythm disturbances
Adequate sensing and pacing thresholds
Endocarditis risk
Paradoxical thromboembolic events

Figure 2. MRI showing the pulmonary and systemic venous connection to the heart after Mustard procedure. LV=anatomic left ventricle, RV=anatomic right ventricle, SB=systemic venous baffle, PV=pulmonary venous baffle.

the use of an active-fixation lead seems preferable to a passive-fixation lead, given the abnormal positioning of the atrial lead in a systemic venous baffle and the placement in the nontrabeculated morphological left ventricle. However, the use of active-fixation pacing leads does not always prevent lead dislodgement. Especially in children, lead dislodgement from the left ventricle may occur secondary to traction due to the child's growth. In order to prevent this complication an exaggerated electrode loop is advised when growth is still suspected.¹² However, the systemic venous channel after Mustard operation is a tube-like channel which may not accommodate an electrode loop, and therefore this loop has to be formed in the anatomic left ventricle, creating a substrate for ventricular ectopic beats. Placement of the ventricular lead itself in the left ventricular apex may also produce ventricular tachycardia, making it necessary to position the lead in the left ventricular outflow tract. A second advantage of active-fixation leads is that they are more easily removed, even after being implanted for many years. This is especially useful in these young patients, where multiple pacemaker system replacements are often required. Positioning of the atrial lead for optimal sensing and pacing thresholds is difficult, as the systemic venous atrium consists partially of pericardial tissue. The superior part of the systemic venous atrium is the most optimal pacing site.¹³ If the left atrial appendage is still incorporated in the systemic venous atrium, placement of the lead in this appendage can also give good thresholds. Sensing capabilities can still be inadequate and poor sensing may lead to asynchronous atrial pacing during sinus rhythm which may induce tachyarrhythmias. Positioning the ventricular lead at the anatomic left ventricular apex puts it in close proximity to the diaphragm, and gives a high incidence of diaphragmatic pacing. To prevent this, the electrode can be placed on the midposterior free wall of the

anatomic left ventricle at the proximate level of the papillary muscles. This gives acceptable thresholds with a minimal risk for diaphragmatic pacing.¹³ As in regular implantations, after insertion of a pacemaker there is some risk of generator failure and erosion, lead fracture and pocket infections. Because of the abnormal anatomy, endocarditis prophylaxis during implantation is of utmost importance. Vegetations on the pacing leads are rarely described.¹⁴ The presence of an intracardiac shunt, including baffle leak, increases the risk of paradoxical thromboembolic events arising from transvenous leads >2-fold.¹⁵ Although there are few data, anticoagulation therapy should be considered in patients with intracardiac shunts receiving transvenous leads.

Conclusion

Reviewing the literature, several complications have been described in patients after a Mustard repair receiving a pacemaker. Most complications can be expected during venous access, positioning of the leads, and sensing and stimulation of the pacing system. The case of our patient illustrates that if care is taken, the procedure may be as simple and successfully performed as is routine in adults with normal cardiac anatomy. ■

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