Use of Cardiac Pacing After the Mustard Operation for Transposition of the Great Arteries

PAUL C. GILLETTE, MD, FACC, DEBORAH G. WAMPLER, CATHLEEN SHANNON, RN, DAVID OTT, MD
Charleston, South Carolina and Houston, Texas

The most frequent complication of the venous redirection (Mustard or Senning) operation for transposition of the great arteries is cardiac arrhythmia. Drug treatment of tachyarrhythmia often worsens bradyarrhythmia. Pacemakers can now treat both arrhythmias. The technique for implantation of pacemakers after redirection for transposition has changed over time from thoracotomy to subxiphoid to transvenous. Atrial pacing is almost always the mode of choice since the electrophysiologic abnormality is sinus node dysfunction with intact atrioventricular conduction.

Twenty-nine patients aged 3 to 19 years (mean 9.6) had implantation of a pacemaker a mean of 5.5 years (range 1 to 14) after undergoing the Mustard operation for transposition of the great arteries. Symptoms referable to bradycardia were eliminated in each case. Four patients who received an antitachycardia pacemaker no longer have symptomatic tachycardia. Four patients have required reoperation, three because of lead problems and one because of traumatic erosion of the pacemaker.

Pacemakers provide excellent relief of symptoms after the Mustard or Senning operation. Transvenous atrial automatic antitachycardia pacemakers offer the best combination of ease of implantation and symptomatic relief.

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Methods

Study patients. Records of the pacemaker clinic at the Medical University of South Carolina Hospital and the Texas Children’s Hospital were reviewed to determine which patients who received a pacemaker had previously undergone the Mustard operation for transposition of the great arteries. The patients’ clinical records were then evaluated from the files of the Divisions of Pediatric Cardiology. The patient’s age at implantation of the pacemaker, technique of implantation of the pacemaker, mode of pacing, duration of pacing, number of reoperations and effect of the pacemaker on the patient’s tachycardia spells were recorded. Symptomatic improvement was also evaluated.

Implantation techniques. The technique of implantation underwent a series of changes over the years. In the early 1970s, thoracotomy was performed for implantation of a pacemaker in a child. The thoracotomy was performed on the left side of the chest and either the base of the left atrial appendage or the left ventricle could be isolated for implantation of a permanent pacing lead. A screw-in lead was used for the ventricle and a plaque type sew-on lead maker. This report discusses our results in 29 patients who underwent permanent pacemaker implantation after undergoing the Mustard operation.
for the atrium. The left side was considered superior for atrial pacing in patients who had undergone the Mustard operation because there were fewer adhesions and less scarring of the atrial epicardium.

In the mid 1970s the techniques of subcostal and subxiphoid implantation of ventricular leads were developed. These had the advantage of not requiring a full thoracotomy and therefore shortened the postoperative recovery period considerably. The pulse generator was implanted beneath the rectus muscle so that the distance between the pulse generator and the tip of the lead was much smaller. Initially, atrial leads were not thought to be implantable by this technique. Therefore, most children had ventricular demand pacing even though it was believed to be more physiologic to use atrial pacing in patients with the sick sinus syndrome. Later, a technique of implantation of a stab-on lead on the atrium by the subxiphoid approach was developed for patients who had not had previous open heart surgery and subsequently for those who had.

Transvenous implantation (Fig. 1 to 3). In 1981, we began performing transvenous pacemaker implantation in children in the cardiac catheterization laboratory. This technique was used in patients who underwent the Mustard operation. It involved entry of the subclavian vein by an introducer technique, positioning of a screw-in lead in the roof of the new right atrium (the anatomic left atrium) (Fig. 1). Transvenous implantation was attempted in 18 patients in this series. In 2 of these 18 patients, initial catheterization studies demonstrated complete or nearly complete obstruction of the superior vena cava and right atrial junction, and no attempt was made to implant a transvenous lead. In a third patient, although there was total obstruction of blood flow between the superior vena cava and the new right atrium, a large portion of the right atrium, including most of the right atrial appendage, was left connected to the superior vena cava, and a bipolar J-shaped lead was positioned in the stump of the right atrial appendage and established excellent thresholds and sensing capabilities. In a
fourth patient, although a large portion of the atrium was accessible from the superior vena cava, adequate thresholds could not be obtained. In 14 patients, it was possible to pass a transvenous screw-in electrode through the superior vena cava-new right atrial junction and position it in the roof of the anatomic left atrium. In three patients, two screw-in leads were passed through the superior vena cava-new right atrial junction (Fig. 2 and 3). In two of these three, two leads were positioned in the atrium and used for a bipolar antitachycardia pacemaker (AAIMB). In the third patient, an automatic atrioventricular (AV) sequential pacemaker (DDD) was used.

Several different brands of standard single chamber pacemakers were used. Each had rate and output programmability in common. The same models of pulse generators were used for atrial and ventricular pacing. Standard AV sequential and automatic units were used. An automatic antitachycardia pacemaker was used in four patients (Cybertach in two and Intertach in two).

Follow-up. Follow-up study consisted of an outpatient evaluation at 6 weeks and yearly thereafter. Transthoracic follow-up study was performed every month for 1 year and every 3 months thereafter. Electrocardiograms and chest X-ray films were obtained at each visit. Twenty-four hour electrocardiographic monitoring and exercise testing were performed if any symptoms were reported. Antiarrhythmic medications were used as clinically indicated.

Results

Twenty-nine patients aged 3 to 19 years had implantation of a permanent cardiac pacemaker at an average age of 9.6 years (SD = 4.4). Three patients had their pacemaker implanted during their hospitalization for the Mustard operation. The difference in time between the Mustard operation and the pacemaker implantation for the remaining 26 patients ranged from 1 to 14 years (mean 5.5).

Indications. In 16 patients the indication for insertion of the pacemaker was the occurrence of symptoms: syncope, near syncope, dizziness or extreme exercise intolerance. Of the remaining 13 patients, 10 required drugs other than digitalis to control supraventricular arrhythmias and 3 had an extremely low heart rate although they were asymptomatic. In two of the three, the low heart rate (30 beats/min) occurred only at night; in the third, heart rates of less than 40 beats/min were recorded during awake activity.

Follow-up. The follow-up period ranged from 1 month to 9 years (mean 3 years). During follow-up, two patients died. Each had had a large ventricular septal defect closed in addition to undergoing the Mustard operation for transposition of the great arteries. Each patient had severe myocardial dysfunction and at recent follow-up before death was found to have a normally functioning pacemaker. One of these two patients had had a syncopal spell in the exercise laboratory due to hypotension with normal pacing by an AV sequential pacemaker.

Relief of symptoms. Symptoms referable to bradycardia were eliminated in each symptomatic patient, 16 of 16. That is, none of these patients had syncope or near syncope and in each the exercise tolerance improved. In 10 of the 12 patients with tachycardia the arrhythmia is no longer associated with symptoms. Four of these 10 patients were treated with an automatic atrial overdrive pacemaker (two with Cybertach and two with Intertach) (2). In three patients, the pacemaker has been shown to successfully overdrive episodes of atrial flutter. Two of these three experienced a brief palpitation with the flutter, whereas the third did not know when the flutter episodes were being overdriven. One patient had not had tachycardia. Of the remaining eight patients, six had an atrial pacemaker and two had a ventricular pacemaker implanted. Two patients continued to have tachycardia despite increased drug use, but it was made easier by the pacemaker. Both had an atrial demand pacemaker.

Reoperations. Twenty-five of 29 patients have not required reoperation. Two patients have required one reoperation and two required two reoperations. In the latter group, the first patient to receive a pacemaker after the Mustard operation had implantation of an epicardial atrial demand pacemaker. After 31/2 years, the atrial lead had slid, probably because of patient growth, so that it also paced the ventricle. At reoperation, the tip of the lead could not be found because of dense fibrous adhesions so a ventricular lead was placed. After 31/2 years, myocardial function had deteriorated and the heart size had increased; thus, a transvenous automatic dual chamber (DDD) pacemaker was inserted and the ventricular pacemaker was removed. The other patient who required two reoperations had pacing of the left phrenic nerve beginning 3 weeks after transvenous implantation. Pulse width and amplitude programming was unsuccessful in preventing this pacing so repositioning of the lead away from the left heart border was carried out. Six months later, the pacemaker eroded as a result of severe physical trauma and an epicardial ventricular pacemaker was implanted. One patient who had implantation of a ventricular epicardial pacemaker at the time of the original Mustard operation had a fractured lead 2 years after implantation; after careful ambulatory monitoring and electrophysiologic testing, it was decided that he no longer needed the pacemaker and it was removed. In the remaining patient the lead broke during a bicycle accident 2 years after implantation of an epicardial ventricular demand (VVI) pacemaker. The lead was replaced without incident.

Discussion

The Mustard operation continues to be a useful physiologic correction for transposition of the great arteries (1–6). Bradyarrhythmias and tachyarrhythmias are the most fre-
quent complications in the postoperative period. Use of an implanted cardiac pacemaker considerably eases the treatment of patients with both bradyarrhythmias and tachyarrhythmias. The usefulness of pacemakers has been limited in the past because of their large size in relation to that of the patient and the technical difficulty of implantation. Recent developments, including the use of microchips for circuitry and improvements in battery design, have reduced the single chamber cardiac pacemakers to a very acceptable size (7). Even a dual chamber pacemaker can now be considered for children of almost any age.

Transvenous implant technique. The advent of the transvenous technique has greatly decreased the morbidity of the implant procedure. Screw-in endocardial leads and the subclavian introducer technique have made transvenous implants possible in pediatric postoperative patients (8). In this study we found lead-related problems to be no more frequent in transvenous than in epicardial lead implants. The radiographic left heart border must be strictly avoided in transvenous implants to prevent phrenic nerve pacing. It is also imperative to determine that the superior vena cava-new right atrial junction is sufficiently wide to permit the pacemaker lead to pass. The long-term effect of a pacing lead on stenosis of this area remains to be determined but must be followed up carefully.

Antitachycardia pacemakers. Our results in four patients suggest that the greatest degree of tachycardia control will be achieved if an automatic antitachycardia pacemaker is used. In patients with known or potential 1:1 AV conduction of atrial flutter, consideration should be given to the use of digitalis to prevent death or severe symptoms from rapid ventricular response. A pacemaker, even an antitachycardia pacemaker, will not prevent all sudden deaths in post-Mustard patients. Attention must be paid to the possibility that supraventricular or ventricular tachyarrhythmias may be present, particularly in patients who have had additional surgery. Not only are these patients more prone to ventricular arrhythmias but they may be more prone to developing AV block. We have used atrial pacing in such patients if they had no evidence of AV block and if they maintained 1:1 AV conduction at atrial pacing rates of 120 beats/min or greater. None of our patients have developed AV block. With the present reliability and size of fully automatic (DDD) pacemakers, our threshold for their use is becoming increasingly low.

Patients who have undergone the Senning operation also suffer from bradyarrhythmias and tachyarrhythmias. Although we have no experience with these patients, we believe that they should be treated similarly to post-Mustard patients.

Indications. Our current indications for a pacemaker implant in a post-Mustard patient are: 1) syncope or near syncope, documented to be due to bradycardia; 2) the need to use antiarrhythmic drugs other than digitalis; 3) bradycardia of less than 40 beats/min while awake; and 4) bradycardia of less than 30 beats/min while asleep.

Conclusion. The results of this study indicate that pacing can be safely and effectively carried out in patients who have previously had the Mustard operation and who have sinus bradycardia or bradycardia-tachycardia syndrome. Symptoms can be relieved in most patients. The evolution of lead implant techniques indicates that in patients weighing more than 10 kg, the preferable technique is transvenous. Care must be taken to ensure that there is not total superior vena cava-right atrial junction obstruction preventing lead implantation before the incision is made for pacemaker implantation. The transvenous implant technique provides better thresholds and avoids the problem of the patient in whom an adequate threshold cannot be obtained by the epicardial technique.

References