Permanent Transfemoral Pacemaker Implantation in a Child with Maroteaux Lamy Syndrome

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DILBER, E., ET AL.: Permanent Transfemoral Pacemaker Implantation in a Child with Maroteaux Lamy Syndrome. Permanent transfemoral pacing has been described as an alternative route in patients in whom the superior venous approach is not feasible. This report describes the use of the femoral venous approach to insert a permanent pacemaker in a child with Maroteaux Lamy syndrome who has complete atrioventricular block and abnormal subclavian venous anatomy. Transfemoral pacing may be a suitable alternative in children with short stature. (PACE 2002; 25:1784–1785)

transfemoral permanent pacemaker, children, Maroteaux Lamy syndrome

Case Report

A 15-year-old male patient with Maroteaux Lamy syndrome was referred to the cardiology department for evaluation of dizziness and fear of death. He had coarse facial features, short stature (height was 84 cm), umbilical hernia, and joint stiffness. A diastolic murmur was heard in the third left intercostal space. An electrocardiogram (ECG) revealed complete atrioventricular (AV) block, and an echocardiogram showed mitral valve and papillary muscle thickening and moderate aortic regurgitation. Holter ECG showed complete AV block, pauses lasting 3.2 seconds, atrial flutter episodes, and frequent premature ventricular contractions with couplets.

The patient was brought to the catheterization room for permanent pacemaker insertion. The attempts for subclavian venous access were unsuccessful. Then the femoral vein was cannulated and a catheter was advanced through the femoral vein into the right and left subclavian veins. Angiographic study revealed small subclavian veins with an abnormal anatomy. The subclavian vein was too narrow, especially at the site of brachiocephalic vein entrance, for a pacemaker lead to insert. It was then decided to implant an endocardial lead through the right femoral vein. An incision was made over the femoral vein, and the femoral vein was cannulated with a venous pacemaker sheath and a 58-cm Medtronic CapSureFix 5076 lead (Medtronic, Inc., Minneapolis, MN, USA) was positioned in the right ventricular apex. The lead was fixed, and good sensing and pacing thresholds were obtained. A second incision was

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Received March 26, 2002; revised May 21, 2002; accepted August 3, 2002.
made lateral and inferior to the umbilicus and a pulse generator pocket was developed under the rectus abdominis muscle. Then the lead was conveyed from the inferior to the superior incision and connected to the pulse generator (Medtronic Sigma SSR 303). The pacemaker was programmed to the VVIR mode. The final lead position and pulse generator is shown in Figure 1. At 6-month follow-up he is well with no problem with his femoral pacemaker.

Discussion

This report presents a case with Maroteaux Lamy syndrome in whom a permanent pacemaker implantation was performed for complete AV block. Cardiac involvement is present in most patients with mucopolysaccharidoses. Increased incidence of unexpected sudden death, abnormality of the cardiac conduction system, and associated AV block have been reported.1,2

Permanent cardiac pacing by a transvenous route is an important mode of treatment for several types of cardiac rhythm disturbances. The usual routes of entry are the subclavian and cephalic veins. Although the femoral vein approach has been proposed as an alternative route for permanent pacing, this is an unusual route especially in children.3-5 Common indications for transfemoral pacing are anomalies of the superior venous circulation, thin anterior chest wall, and cosmetic reasons.6,7 Lead dislodgement secondary to the geometric path of the inferior vena cava entry in the right ventricle is the main complication of the transfemoral vein approach. The use of active-fixation leads has been reported to reduce this complication.8

In the present case, transfemoral pacing via the superior vena cava was not feasible owing to the abnormal venous anatomy. The subclavian vein was too narrow at the entrance of brachiocephalic vein. Thus, superior vena cava placement may disturb the venous drainage and may cause superior vena cava syndrome. Besides, as the patient had a short stature, it would have been difficult to develop a generator pocket in the anterior chest wall.

Permanent pacemaker implantation should be considered in similar cases with mucopolysaccharidoses and the transfemoral vein approach may be a suitable alternative even in children with short stature.

Acknowledgments: The authors thank G.T.N. Besley, M.D., Royal Manchester Children's Hospital, UK, for enzymatical diagnosis of the patient.

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PACE, Vol. 25, No. 12 December 2002 1785