Radiofrequency catheter septal ablation for hypertrophic obstructive cardiomyopathy in children

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Background. The definitive therapeutic options for symptomatic obstructive cardiomyopathy in childhood are restricted. At present, extensive surgical myectomy is the only procedure that is of proven benefit.
Patients and Methods. Three patients, aged 5, 11 and 17 years, respectively, with progressive hypertrophic obstructive cardiomyopathy and increasing symptoms were considered for radiofrequency catheter septal ablation. The peak Doppler gradient recorded on several occasions ranged between 50 to 90mmHg. Via a femoral arterial approach, the His bundle was initially plotted and marked using the LocaLisa navigation system. Subsequently, using a cooled tip catheter a series of lesions were placed in the hypertrophied septum, taking care to stay away from the His bundle. A total of 17, 50 and 45 lesions were applied in the three patients. In one case, the procedure was complicated by two episodes of ventricular fibrillation requiring DC cardioversion but without any neurological sequelae.
Results. The preablation peak-to-peak gradient between left ventricle and aorta was 50 mmHg, 60 mmHg and 60 mmHg, respectively, and remained unchanged immediately after the procedure. All patients were discharged from hospital 48 hours later. Serial measurement of serum troponin T and CK-MB isoenzyme confirmed significant myocardial necrosis. Follow-up echocardiography both at seven days and at six weeks postablation confirmed a beneficial haemodynamic result, with reduction of left ventricular outflow obstruction and relief of symptoms.
Conclusion. In young children, in whom alcohol-induced septal ablation is not an option, radiofrequency catheter ablation offers an alternative to surgery, with the benefits of repeatability and a lower risk of procedure-related permanent AV block. (*Neth Heart J* 2005;13:448-51.)

Keywords: hypertrophic obstructive cardiomyopathy, radiofrequency catheter ablation, childhood

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Hypertrophic obstructive cardiomyopathy (HOCM) is a primary myocardial disorder characterised by inappropriate myocardial hypertrophy. The clinical course is highly variable.[1-3] In children, the prognosis is determined by the degree and rate of myocardial hypertrophy and corresponding obstruction to blood flow.[4] Sudden death as a result of severe outflow obstruction, myocardial ischaemia and malignant ventricular tachycardias is well recognised.[4-6] Possible definitive approaches to therapy include surgical resection of the obstructive outlet septum and transcoronary septal reduction by selective alcohol injection into the septal branches of the left anterior descending coronary artery. Surgery carries a risk of AV block requiring permanent pacemaker implantation. This risk is approximately 5% in very experienced hands. Selective alcohol ablation is technically unfeasible in young children.[5,7,8] We report transcatheter ablation of the left ventricular outlet septum using radiofrequency current in three children.

Patients and methods

The three children were aged 5, 11 and 17 years, respectively. In all of them, the diagnosis of HOCM had been made in infancy. All patients had remained under serial follow-up for several years, and had received a variety of pharmacological agents including β-blockers and verapamil in appropriate dosages. The obstruction was noted to be progressive in all cases, and was associated with a variety of symptoms including progressive exercise intolerance (n=3), unexplained chest and abdominal pain suggestive of angina (n=1), and decreased spontaneous activity (n=2). The peak

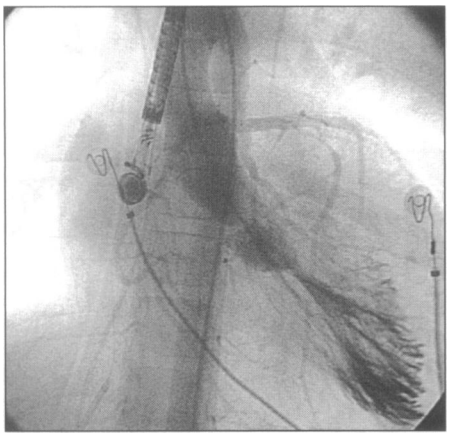

Figure 1. Left ventricular angiogram demonstrating the location and extent of septal hypertrophy, producing outflow obstruction in patient 2.

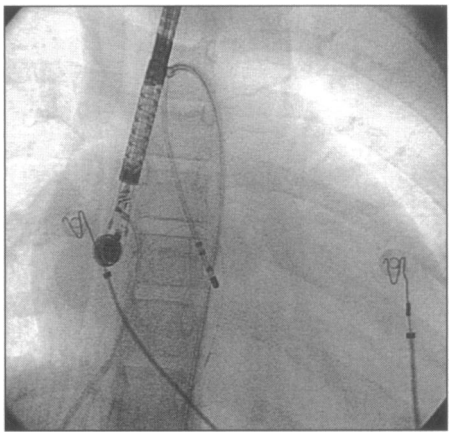

Figure 2. Fluoroscopic delineation of the location of the ablation catheter prior to applying the initial radiofrequency current lesions in the same patient. Two additional catheters, in the right atrium and right ventricle, respectively, for AV sequential pacing, are seen.

Doppler echocardiographic gradient in the left ventricular outflow tract at rest, measured on several occasions prior to considering definitive therapy, ranged between 50 and 80 mmHg (patient 1), 70 and 90 mmHg (patient 2), and 64 and 90 mmHg (patient 3). In addition to the pronounced septal hypertrophy, all three patients exhibited generalised left ventricular hypertrophy (left ventricular free wall thickness of >30 mm). Two of the three patients (aged 5 and 17 years, respectively) also had associated abnormalities of the mitral subvalvular apparatus, consisting of septal attachment of the mitral valve chordae and an anomalous apical mitral valve papillary muscle. In both of these patients, surgical excision of the obstructive septum was also deemed to carry a significant risk of the patient requiring concomitant mitral valve replacement. In view of the young age of the patients, selective transcoronary alcohol ablation of the septal hypertrophy was deemed to be technically unfeasible or risky. Transluminal radiofrequency catheter ablation of the hypertrophied septum was considered to be an alternative to surgery, and informed consent for the procedure was obtained from the patients and their parents, as appropriate. This procedure has hitherto been described in a single adult patient, in whom it appeared to produce adequate relief of septal hypertrophy.[9]

Cardiac catheterisation and ablation procedure

Cardiac catheterisation was performed under general anaesthesia. Left ventricular angiography was performed to demonstrate the extent of septal obstruction (figure 1). A direct pullback gradient across the left ventricular outflow tract was also measured using a 5F Tracker catheter, which allows serial measurements to be made by withdrawal of the catheter over a 0.035 inch guidewire prepositioned in the left ventricle. This has the added advantage that the aortic valve does not require to be recrossed every time a gradient needs to be measured. The peak withdrawal gradients across the left ventricular outflow tract were 50 mmHg, 60 mmHg and 60 mmHg, respectively.

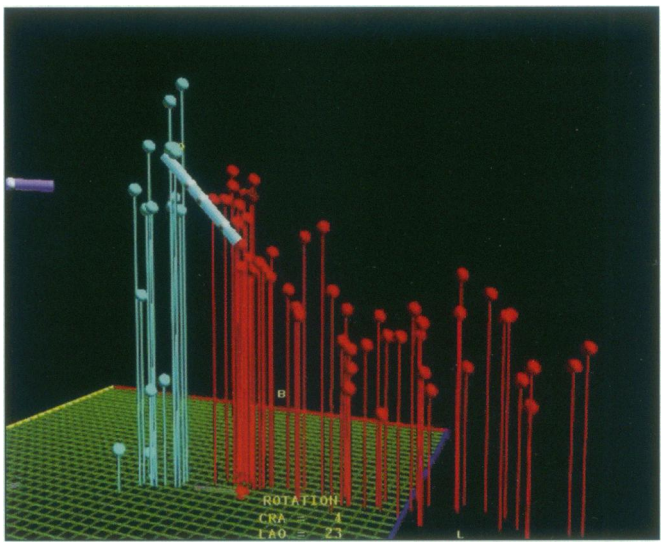

Figure 3. The LocaLisa map showing the location of the His bundle (in blue), and the location of the individual lesions (red) in patient 2.

Sequential AV pacing at progressively shorter AV coupling intervals (ranging between 120 and 80 ms) was performed from the right atrium and right ventricle, and the outflow tract catheter withdrawal gradient was remeasured during each pacing manoeuvre, to assess any possible haemodynamic benefit from permanent right ventricular apical pacing or from induced left bundle branch block. It was demonstrated, however, that sequential pacing did not influence the left ventricular gradient. Using a 7F steerable cooled tip ablation catheter (Sprinklr, Medtronic) introduced via the femoral artery and through the aortic valve, the location of the His bundle and proximal left bundle branch was mapped out using the LocaLisa mapping system (Medtronic) (figures 2 and 3).

The 7F cooled tip ablation catheter was then used for ablation of the hypertrophic septum. This catheter was chosen in order to increase the depth of the lesions. Sequential lesions were made on the left ventricular

septum, commencing distally within the ventricle, and proceeding more proximally with subsequent lesions to just under the aortic valve. Three such linear lesions were made between the left ventricular apical septum and the aortic valve. Care was taken to stay away from the His bundle, by monitoring catheter location continuously using the LocaLisa system. The local electrogram was scrutinised at each potential ablation site for the presence of a His bundle potential. If this was seen the catheter was moved away from that site and reengaged on the septum at an adjacent site which did not demonstrate a discrete His bundle potential. At locations where frequent junctional ectopic beats were observed during RF energy application, atrial overdrive pacing was performed from the right atrium to confirm that AV conduction was intact. During lesion application the catheter tip was continuously irrigated with normal saline at a flow rate of 300 ml/hour. Each lesion was applied for a period of 60 seconds. In addition to local electrical signals, catheter position was also monitored using angiographic roadmapping, and by transoesophageal echocardiography. Transoesophageal echocardiography demonstrated echodense lesions in the left ventricular septum, at the sites of RF current application. It was also useful in identifying the insertions of the mitral valve chordal apparatus to the anomalous papillary muscle in the left ventricle, thereby avoiding damage to the papillary muscle and creating iatrogenic mitral valve dysfunction. When three linear lesions had been completed as described, the catheter withdrawal gradient was remeasured, and the procedure terminated regardless of the final measured gradient, which remained unaltered in all three patients.

The total fluoroscopic times were 15, 21 and 25 minutes, respectively. A total of 17, 50 and 45 RF lesions, respectively, were applied in the three patients, to achieve three linear lesions extending from the most apical location of septal hypertrophy to just under the aortic valve.

The youngest child, aged 5 years (patient 1), developed two episodes of ventricular fibrillation during RF energy application, necessitating DC cardioversion (2 Joules/kg). Apart from this, there were no other procedure-related complications.

Results

Clinical course

All patients were discharged from hospital 48 hours after ablation. All were in sinus rhythm at discharge. The ventricular fibrillation in patient 1 was felt to be the result of catheter manipulation in the narrow left ventricular outflow tract, and was thought to be unlikely to recur. Telemetric Holter monitoring during the 48 hours of in-hospital observation did not demonstrate any ventricular ectopy. As is our policy for standard catheter ablation procedures in children with tachyarrhythmias, oral aspirin (5 mg/kg/day administered as a single dose, up to a maximum of 300 mg) was commenced on the afternoon of the procedure, and continued for six weeks postprocedure.

Cardiac enzymes

Serial blood samples were taken at time 0 (preprocedure), and at six hours and 24 hours postablation, for measurement of serum troponin T and CK-MB isoenzyme, to attempt to quantify the extent of myocardial necrosis produced by the procedure. The maximum values, measured postablation, were >5 µg/l (range 5.1 to 8.0; troponin T baseline value <0.1 µg/l) and >45 units/l (range 45 to 74; CK-MB baseline value <20 u/l) respectively.

Follow-up

Follow-up echocardiography performed five days later revealed a residual left ventricular outflow gradient of 25 mmHg, 30 mmHg and 25 mmHg, respectively, which had diminished further to <20 mmHg at six weeks' follow-up in all three patients. Serial echodense spots could be seen in the left ventricular outflow tract (figure 4), corresponding to the sites of RF energy application.

Mitral valve and aortic valve function was unchanged. There was no new aortic valve insufficiency; the pre-existent mitral valve insufficiency present in two of the three patients was also present at follow-up. None of the patients have had symptoms at follow-up. All continue to take their preprocedure medications (β-blocker) at the same dosage.

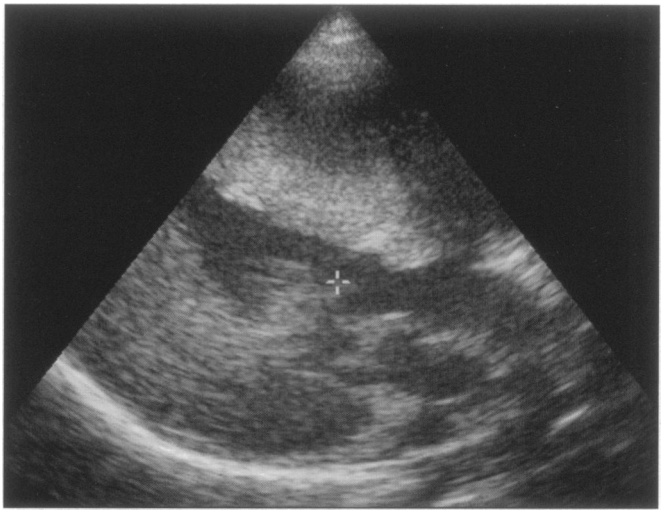

Figure 4. Echocardiographic appearance of the left ventricle following ablation in patient 2. This frame, in peak systole, demonstrates that the outflow tract remains patent. The sites of application of radiofrequency current are seen as echodense spots on the septal aspect of the outflow tract. This echocardiographic appearance was associated with a marked decrease in the Doppler-derived gradient.

Discussion

The accepted 'gold standard' for therapy of HOCM in children with persistent symptoms and a significant outflow tract gradient of >40mmHg is surgical myectomy using cardiopulmonary bypass.[1,6] Surgical myectomy is associated with a good functional outcome, as has most recently been demonstrated in the study of Woo et al., although no children were included in their series,[7] but it carries a significant risk of permanent AV block requiring pacemaker therapy. In patients with abnormal mitral valve anatomy, as was the case in patients 1 and 3 described above, mitral valve function may be further compromised by surgical myectomy, necessitating concomitant mitral valve replacement.[3,6] This is in contrast to the functional mitral insufficiency sometimes observed after myectomy, which is in most instances nonprogressive. In some patients, AV sequential pacing with a short AV delay may reduce the left ventricular outflow tract gradient, and be haemodynamically beneficial. This was excluded in our patients, prior to radiofrequency current application. However, it is probable that the beneficial effects of sequential pacing are only manifest after a period of follow-up. An alternative to surgery in older patients is percutaneous transluminal septal myocardial ablation by alcohol-induced obstruction of a septal branch of the left anterior descending coronary artery. In recent studies, the results of alcohol ablation of the septum have been shown to be at least comparable to surgical resection, with high success rates and a lower morbidity.[8,10,11] This approach is technically unfeasible in small children, in whom selective cannulation of the target septal artery and controlled myocardial necrosis without permanent sequelae of AV block have not previously been reported. An alternative to alcohol ablation is intracavity ablation of the obstructing interventricular septum using radiofrequency current. This approach has been reported in a single adult, in whom the ablation was performed from the right ventricular side of the interventricular septum.[9] In contrast to this report, we performed direct septal ablation from within the left ventricle, using continuous monitoring of catheter tip location after having previously mapped out the location of the His bundle. Initial sequential atrioventricular pacing at different AV intervals confirmed that permanent pacing would not be beneficial. The number of lesions applied was decided upon empirically; the aim was to ablate the segment of hypertrophied left ventricular septum commencing apically and moving progressively towards the aortic valve. Lesions were applied continuously, to avoid the theoretical risk of creating narrow corridors of intact myocardium which might result in reentrant ventricular tachycardia circuits in the future.

The follow-up data on our patients suggest that this technique is feasible, and produces a beneficial reduction in outflow tract gradient. An additional potential advantage of the approach described here includes the possibility of performing repeat procedures in the future, when clinically indicated. Clearly further data on a larger series of patients are required to confirm the beneficial effect reported here, and at the present time the procedure should be considered experimental. The procedure is not without risk, and one patient developed two episodes of ventricular fibrillation requiring cardioversion. It is important therefore that the procedure is performed by paediatric cardiologists with extensive experience of catheter ablation techniques. ■

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