made a full recovery when treatment was started 5 days after the onset of the spasms has also been reported (Oates and Stapleton, 1971). Because of delays in the diagnosis of hypsarrhythmia early treatment usually implies that it was started within 6 weeks of the onset of spasms (Chevrie, Aicardi, and Thieffrey, 1968).

A review of 150 cases (Jeavons, Bower, and Dimitrakoudi, 1973) showed no difference in long-term prognosis when steroids were used. This review showed that factors associated with a good prognosis were normal development before the onset of the spasms, a short period of spasms, and early steroid therapy in the cryptogenic group. 37% of children in the cryptogenic and immunization groups made a full recovery, the prognosis being much worse in the symptomatic group.

Summary

A child exposed to rubella at 16 weeks' gestation developed hypsarrhythmia at 5 months of age. Treatment with ACTH quickly improved the electroencephalogram and controlled the seizures, but at 2 years of age development was delayed. Hypsarrhythmia has not previously been described in association with congenital rubella infection.

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- R. K. OATES* and DAVID HARVEY
St. Charles's Hospital and Queen Charlotte's Hospital for Women, London.

*Correspondence to Dr. R. K. Oates, Dept. of Medicine, Royal Alexandra Hospital for Children, Camperdown, N.S.W. 2050, Australia.

Long-term results of surgical treatment for pulmonary valve stenosis

Since the introduction of surgical treatment for congenital pulmonary stenosis (Brock, 1948), the procedure has become well established with a low mortality (Nadas and Fyler, 1972). In reporting a series of 117 children who underwent pulmonary valvotomy or valve excision, we particularly refer to the progress of the 109 patients who survived.

Patients and indications for operation

All 117 patients had pulmonary valve stenosis with an intact ventricular septum, and underwent operation during the period 1956–1973. There were 60 boys and 57 girls, ranging in age from 1 to 14 years (mean 4.5 years). The indication for investigation was the clinical picture of substantial pulmonary stenosis, supported in most instances by characteristic x-ray signs and, in 41 patients (35%), by ECG evidence of right ventricular hypertrophy. 26 (22%) were investigated under the age of 4 years because of the onset of cardiac failure (4) or ECG evidence of severe right ventricular hypertrophy. Symptoms were present in a total of 12 patients (10%).

Investigations, which included cardiac catheterization and selective angiocardiography, were completed in most patients (58%) just before school entry. The systolic pressure gradient across the pulmonary valve ranged from 40–250 mmHg (mean 108).

Operation. Operation was performed in 102 patients with cardiopulmonary bypass support, but in 15 patients early in the series moderate hypothermia at 30°C was used. Using the transarterial approach, mobilization of the fused commissures from the pulmonary arterial wall followed by commissurotomy was sufficient to relieve the obstruction in 91, but valve excision was necessary in 26 either because of an underdeveloped valve ring or because the severity of the obstruction made commissurotomy inadequate. Furthermore, 12 patients required infundibulectomy. Even so, in 43 patients right ventricular hypertension had not been abolished at this stage. 8 patients died at operation or within 72 hours; 4 had undergone commissurotomy and 4 valve excision. 6 (75%) deaths were in patients with a valve gradient exceeding 150 mmHg.

Postoperative course. 109 patients were available for follow up and there were no late deaths. The review period ranged from 1–17 years (mean 6.5 years). All were active and symptom free. A systolic murmur was audible at the pulmonary area in 36 patients (33%) but was shorter and quieter than before operation. In 20 patients a short low-pitched pulmonary diastolic murmur was heard, occurring with equal frequency in the commissurotomy and valve excision groups. All x-ray and ECG abnormalities disappeared, though in some it took as long as 6 years for this to occur.

Cardiac catheterization with angiocardiography was repeated between 1 and 17 years after operation (mean 6.3 years) in the 43 patients with persisting right ventricular hypertension. The resting pressures in the right ventricle and pulmonary artery and the systolic pressure gradient across the valve were compared with the corresponding figures recorded before operation and in the operating theatre just before closure of the chest. The gradient recorded in theatre immediately after valve surgery had shown a significant fall in most patients; it had remained unaltered in 3, and substantially raised in 7 but fell subsequently to an acceptable level as shown by cardiac catheterization 2 to 5 years later. The gradient at review catheterization ranged from 4–40 (mean 18) mmHg.

Pulmonary regurgitation. Although after operation 20 patients had a short diastolic murmur at the upper left sternal border, pressure recording at repeat catheterization did not indicate substantial pulmonary regurgitation in any. Pulmonary artery angiography revealed regurgitation of contrast medium of mild degree in 12 patients, 6 after valve excision, and 6 after simple commissurotomy.

Discussion

Our ultimate criterion for selecting patients for operation was a systolic pressure gradient across the valve greater than 40 mmHg, as advocated by Campbell (1969). In most, however, the gradient was much higher (mean 108 mmHg). Few long-term studies have been reported. Braimbridge *et al.* (1966) reported the results of surgical treatment in 56 patients, most of whom were children. Postoperative catheterization in 18 showed a satisfactory result in 13 but right ventricular hypertension persisted in 5 due either to infundibular obstruction or a hypoplastic valve ring. This could have been attributable to the early use of the transventricular approach. In all our patients the transarterial was used, allowing when necessary the excision of any infundibular muscular obstruction at the time of valvotomy; this was done in 12 patients.

Danielson *et al.* (1971) gave an account of the operative results in 145 children. The mortality was 4%, and the results 1–10 years after operation were regarded by the authors as excellent, though

an analysis was not presented. Finnegan *et al.* (1974) made haemodynamic studies of 13 children 1 to 14 years after pulmonary valvotomy. The mean valve gradient before operation was 90 mmHg, and after operation was 25 mmHg at rest, with only a very slight rise in exercise. It can therefore be assumed that if the valve gradient at rest has fallen to a satisfactory level, the haemodynamic response to exercise will be normal.

A pulmonary diastolic murmur in 23 of their 56 patients (42%) was regarded by Braimbridge and his colleagues (1966) as evidence of pulmonary regurgitation when associated with a raised right ventricular end diastolic pressure. Although we suspected that a pulmonary diastolic murmur was a sign of pulmonary regurgitation in our patients, uncertainty arose from the fact that it was not uncommonly found after simple commissurotomy and that other clear signs of regurgitation were lacking. The right ventricular end diastolic pressure was not raised in any of our patients. Moreover, the regurgitation of contrast medium shown by angiocardiography was not only trivial in amount but present as often after commissurotomy as after valve excision, thus raising the possibility that it was merely an artifact of the technique. During the period of this review no evidence emerged of a deleterious effect of pulmonary valve excision in the 22 patients. They remained symptom free, and neither the ECG nor the chest x-ray showed any evidence of the development of right ventricular enlargement.

Certain conclusions on management were derived from the data obtained at postoperative catheterization. From the 11 patients catheterized more than 10 years after operation, it was evident that there is no tendency to recurrence of obstruction. From 18 patients catheterized at 5 years or earlier after operation, it was evident that persisting obstruction immediately after valvotomy quickly subsides thereafter, as it had done in these within the period of observation. We now believe that cardiac catheterization is necessary only when the valve gradient has remained significantly raised in theatre. If performed 3 years after operation it is likely to give a reliable assessment of the ultimate effect of the valvotomy and patients can then be dismissed from further review. Dismissal from supervision at the earliest possible opportunity is desirable not least on emotional grounds. We consider that because of the lack of long-term information on the subject those who have been subjected to valve excision require a longer period of review. No patient developed infective endocarditis during the study, but we have continued to advise prophylactic

antibiotic therapy to cover surgical operations likely to cause bacteraemia.

A feature of most patients was the lack of symptoms of exercise intolerance except for a few with very severe pulmonary stenosis. Although some patients after operation were said to have become more active and vigorous, most parents when questioned on the subject did not report any change.

Summary

109 children who survived surgical treatment for isolated pulmonary valve stenosis were followed for up to 17 years. In all the postoperative status was assessed as satisfactory. Cardiac catheterization repeated in 43 gave a resting valve gradient below 40 mmHg. The 22 children whose pulmonary valves had been excised were as healthy as the 87 who had undergone pulmonary valvotomy. Consideration was given to the desirable length of postoperative review. Except for the few children with symptoms before operation, a postoperative increase in exercise tolerance was not a feature.

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J. M. REID,* E. N. COLEMAN, J. G. STEVENSON,
J. A. INALL, and W. B. DOIG
University Department of Child Health and Department of Cardiology, Royal Hospital for Sick Children, Glasgow, and Cardio-Thoracic Unit, Mearnskirck Hospital, Glasgow.

*Correspondence to Dr. J. M. Reid, Cardio-Thoracic Unit, Mearnskirck Hospital, Newton Mearns, Glasgow G77 5RZ.