quality of the child's life and the lack of any other possible cure for his severe cardiomyopathy; on the other hand his underlying malignant disease was a major obstacle. Now, more than three years later, that choice seems to have been correct. The child has regained a completely normal life: he attends school, has normal heart function, and is able to take part in normal physical activities.

Because of the improvement in cure rate provided by combined treatment of some childhood cancers, the limits for indication of organ transplantation may well be revised. We are aware that a few children with cancer have recently undergone heart transplantation (J Le Bidois, Paris, personal communication⁴), and this treatment is also being given for some non-malignant chronic diseases of children.⁵

The mechanisms of development of distant tumour metastases are not yet completely clarified. A possible role for immune surveillance has been postulated but is still under investigation. In the present case long term immunosuppressive drugs, given to prevent graft rejection, could cause some depression of the immune system, and possibly enhance the risk of tumour recurrence. To assess the real degree of immune depression we had induced in this patient, we tested his immune function. Humoral immunity, assessed by serum immunoglobulin concentrations, was normal. Cellular immunity was also substantially normal.

Only the delayed type hypersensitivity skin test reactions, reliable markers of overall immune function, were persistently depressed, as is also seen in children undergoing chemotherapy for leukaemia.

To conclude, heart transplantation successfully cured doxorubicin induced cardiomyopathy despite the underlying tumour. Long term administration of steroids, azathioprine, and cyclosporin caused only mild impairment of immune function; he had no recurrent infection, local recurrence of tumour, or distant metastases.

With continuing advances in our understanding of tumour biology and the techniques of organ transplantation, eligibility criteria for transplantation should continually be re-evaluated. Nevertheless further experience is necessary before this becomes the accepted treatment.

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Life threatening 'epilepsy'

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Abstract

A 5 year old girl presenting with episodes of sudden loss of consciousness was found to have intermittent ventricular tachycardia and, on one occasion, self limiting fibrillation. Corrected QT interval was normal. After several therapeutic measures clinical and electrocardiographic improvement was achieved by administration of sotalol.

We present the case of a girl with a sudden loss of consciousness that was thought to be caused by epilepsy.

Case history

The patient, a 5 year old girl, was admitted to hospital having collapsed at home. She was found by her mother unconscious, limp, and cyanosed. She had been incontinent of urine and faeces. She recovered after four minutes and appeared confused. She had been born after

42 weeks' gestation by caesarean section for fetal distress evidenced by meconium staining. She had been referred to a paediatric neurologist at 3 years of age with a history of never running. The only abnormalities at that time were mild generalised hypotonia and a slightly broad based gait suggestive of a mild degree of ataxia. Her physical state was thought to be in keeping with a benign congenital hypotonia. At nursery she was noted to have poor fine hand movements and was referred for occupational therapy. Fifteen days before this admission she had tripped and banged her head on the floor. She was then unconscious for four minutes. Skull radiography had shown no fracture and observations overnight in hospital had been normal. A cousin had been investigated for recurrent hypoglycaemia when aged 2 years and ketotic hypoglycaemia diagnosed.

On admission she was fully conscious and alert. Her height was on the 97th centile and weight on the 75th. Cardiovascular, respiratory, and abdominal examinations were normal. She

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was a clumsy girl with mildly ataxic gait but no other neurological abnormality. Blood biochemistry including fasting blood glucose was normal. Computed tomography of her brain, a 12 lead electrocardiogram (ECG), and an electroencephalogram also gave normal results.

A month later she was again found unconscious. As before there were no witnesses of her behaviour or activity level before her collapse. On admission the only abnormal finding was an irregular pulse. Electrocardiography showed sinus rhythm with intermittent junctional beats. A 24 hour ECG showed episodes of sustained polymorphic ventricular ectopics and ventricular tachycardia. Exercise testing, with intensive care cover, did not produce any arrhythmia; in particular the QT interval did not alter. Cardiac ultrasound scan was normal. Disopyramide treatment was commenced and therapeutic concentrations attained. A CardioMemo system was supplied to the family and its use demonstrated. This small instrument is held against the bare chest during any abnormal episodes and a button pressed to record a 32 second electrocardiographic strip. This recording can then be transmitted by telephone to a monitoring service at the hospital. Several days later this method demonstrated for the first time that collapse of the patient, during normal quiet activity, coincided with an episode of ventricular tachycardia. Flecainide was commenced after readmission to hospital and commencement of continuous electrocardiographic monitoring. Later the same week an episode of self limiting ventricular fibrillation was accompanied clinically by a complaint of sore knees but no loss of consciousness. The blood flecainide concentration was found to be within the therapeutic range. Electrophysiological studies failed to initiate any arrhythmia. Sotalol treatment was commenced. The ECG showed an appreciable improvement with only occasional ventricular ectopics. There have been no further episodes of collapse.

Discussion

Before concluding that acute loss of consciousness is due to epilepsy a number of investigations are generally considered mandatory to rule out anatomical, electrolyte, infective, metabolic, or neoplastic disturbances. In this case the history was misleading in two respects. Firstly, the possibility of asphyxial damage at birth was raised by the presence of meconium and subsequent poor gross and fine motor coordination; this was excluded by review of the neonatal record and normal brain tomography. Secondly, there was a second degree relative who had suffered intermittent hypoglycaemia, but a prolonged fast in this patient failed to produce hypoglycaemia.

Twelve lead electrocardiography, routine as an investigation for interruption of normal consciousness in this unit, was unrewarding.

Garson reports the separate sudden deaths of two siblings whose mother was subsequently found to have a prolonged QT interval.¹ Garson therefore recommends the calculation of the corrected value QTc (divided by the square root of the length of each cardiac cycle in seconds), although this has been disputed.^{2 3} In this case QTc before commencement of treatment was not prolonged (0.37 seconds) at rest or during exercise. Previous reports of patients with normal QT and QTc values and yet suffering recurrent syncope due to ventricular fibrillation suggest a forme fruste of the long QT syndrome.⁴

The nature of the episodes (sudden falls or 'drop attacks') together with the finding on one occasion of an irregular pulse nevertheless strongly suggested the possible involvement of an arrhythmia. Although a 24 hour ECG did reveal intermittent ventricular tachycardia, it was not until some time later with the aid of a CardioMemo system that one of the patient's attacks was found to coincide with the arrhythmia. The lesson learnt is that during the investigation of such symptoms in the presence of a normal electroencephalogram, efforts to exclude a causal cardiac arrhythmia should proceed via 12 lead ECG, 24 hour monitoring, and CardioMemo systems without delay in order to prevent further life threatening episodes.

Once the diagnosis was reached it proved difficult to find a suitable treatment to decrease the frequency and severity of ventricular arrhythmia. As a class Ic antiarrhythmic agent flecainide acts as a membrane stabiliser to reduce myocardial excitability with no effect on action potential duration. Unfortunately proarrhythmia may occur in patients with ventricular arrhythmia. This treatment was therefore initiated under continuous electrocardiographic monitoring and terminated after one episode of self limiting ventricular fibrillation. Sotalol was used next as it is a drug with a wide range of action by reason of its combined β blocking activity (class II antiarrhythmic action) and its property of prolonging the action potential duration in myocardial tissues (class III action). The case described here supports available data which indicate that sotalol is an effective antiarrhythmic compound for a significant number of patients with life threatening ventricular tachycardia/ventricular fibrillation refractory to conventional antiarrhythmic drugs.⁵

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