SHORT REPORTS

Pregnancy in women with Friedreich's ataxia

Friedreich's ataxia is an uncommon autosomal recessive disease of the nervous system affecting one in 50 000 people. In many cases its onset occurs in adolescence. As well as having severely disabling neurological symptoms 8-18% of patients have diabetes mellitus and 60-90% have electrocardiographic abnormalities, of whom one third have cardiac symptoms and signs.[1,2] Despite these severe complications affecting several organ systems no study of the effects of Friedreich's ataxia on pregnancy and its outcome has been reported previously.

Patients, methods, and results

I sent a questionnaire to women of childbearing age with Friedreich's ataxia on a list obtained from the Friedreich's Ataxia Association. A total of 18 women who had been pregnant were identified from this list, of whom three were excluded because the disease had been diagnosed after their pregnancies. In addition I myself investigated two cases. Of the 17 patients, three had identifiable cardiac signs: two had hypertrophic obstructive cardiomyopathy and one recurrent tachycardia and electrocardiographic abnormalities. One patient had insulin dependent diabetes mellitus, and one had yielded an abnormal result to a glucose tolerance test, although she was not taking any specific drugs.

The table shows the obstetric outcome of the 17 patients. There were no perinatal deaths. All women had been delivered of live babies in excess of 36 weeks' gestation. Five women had been offered termination of pregnancy because of their condition and four had refused; all four were delivered of a live baby with no obstetric complications. Two patients had hypertension induced by pregnancy but subsequent normal deliveries. Three women received epidural anaesthesia: two, one of whom had hypertrophic obstructive cardiomyopathy, had a normal delivery and one had a forceps delivery. One of the 16 women without diabetes mellitus developed gestational diabetes during her pregnancy.

Details of pregnancy in 17 patients with Friedreich's ataxia

Case No	Age at diagnosis of Friedreich's ataxia (years)	Parity	Delivery (all >36 weeks)			Associated conditions
			Spontaneous	Forceps	Epidural	
1	12	1+0	1	–	1	Hypertrophic obstructive cardiomyopathy
2	7	1+0	1	–	–	Insulin dependent diabetes
3	21	1+0	1	–	–	Supraventricular tachycardia
4	17	1+0	1	–	–	
5	23	2+0	2	–	–	
6	5	1+0	1	–	–	Hypertension induced by pregnancy
7	18	2+0	1	–	1	Hypertension induced by pregnancy
8	16	3+0	3	–	–	
9	20	1+0	–	1	–	Hypertrophic obstructive cardiomyopathy
10	15	2+0	2	–	–	Abnormal glucose tolerance test; gestational diabetes
11	15	1+0	1	–	–	
12	18	1+0	1	–	–	
13	18	2+0	2	–	–	
14	25	2+0	2	–	–	
15	16	2+0	2	–	–	
16	19	2+0	1	–	–	
17	29	1+1*	1	–	–	

*Termination of pregnancy and sterilisation at five months.

Contraceptive follow up for the 17 women varied. Two were sterilised, having completed their families; six used a combined oral contraceptive pill; three used an intrauterine contraceptive device; and the remainder used barrier methods of contraception.

Comment

Although Friedreich's ataxia is a serious disease with serious cardiovascular and metabolic problems, the reproductive performance of the 17 women was good. They did not seem to have been prone to antenatal complications, especially premature labour and hypertension induced by pregnancy. Thus a normal delivery should be expected in women with Friedreich's ataxia; when epidural anaesthesia is required it should not be withheld purely because of the condition. Friedreich's ataxia itself is not an indication for termination of pregnancy. Cardiovascular examination is mandatory in all patients, and early referral to a cardiologist is recommended. In those who have heart disease electrocardiographic monitoring throughout labour should be recommended.

Couples of whom one or both are affected with Friedreich's ataxia should seek full genetic counselling. The risk of a woman with Friedreich's ataxia and no family history having an affected child is about one in 220 (pamphlet obtainable from the Friedreich's Ataxia Association, Burleigh Lodge, Knowle Lane, Cranleigh, Surrey).

1 Harding AE, Hewer RL. The heart disease of Friedreich's ataxia: a clinical and electrocardiographic study of 115 patients with an analysis of serial electrocardiographic changes in 30 cases. *Q J Med* 1983;208:489-502.
2 Hewer RL. Study of fatal cases of Friedreich's ataxia. *Br Med J* 1968;iii:649-52.

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Ingestion of button batteries: hazards and management

The widespread use of small, button sized batteries in household objects has led to increased awareness of the special hazards caused by their ingestion. A few reports have been published in the United States and Great Britain, but most have been isolated case reports, mainly of serious complications including death. Most authors have recommended early surgical intervention in all cases. We report our experience with three patients and review the previous reports.

Case reports

Case 1—A 6 year old boy swallowed the battery from a digital watch. Radiographs showed the intact battery in his stomach and, 24 hours later, in his caecum. He remained asymptomatic and passed the battery six days later.

Case 2—A 20 month old girl swallowed the battery from a calculator. Its presence was confirmed by radiography. She passed the battery intact three days later.

Case 3—A 4 year old girl swallowed the battery from a toy calculator. This was confirmed by radiography; repeat radiography one week later did not show any foreign body.

Comment

Before 1983 only six cases of ingestion of button batteries had been reported. Two of the patients died, one required a prolonged stay in hospital, two had suspected mercury poisoning, and in the remaining patient the battery perforated a Meckel's diverticulum.[1] Accordingly, many workers recommended early intervention.[1,2] Litovitz, however, reviewed 56 cases in adults and children and recommended that surgical intervention should be withheld in the absence of specific clinical indicators.[3]

We reviewed the information available in reports on 58 children who ingested batteries. Forty eight were asymptomatic but, nevertheless, 16 underwent surgical intervention; seven of these showed evidence of minor mucosal damage. The remaining 32 asymptomatic patients were allowed to pass their batteries spontaneously. No late complications occurred in either group, which suggests that there is no benefit from surgical intervention in asymptomatic patients.

In five of the 10 patients with symptoms the battery lodged in the oesophagus; these five patients included the two who died and most of those with serious complications. The batteries ingested were 21-23 mm in diameter. These were the largest batteries in the entire series, and only one of the batteries that went beyond the oesophagus was of a similar size. The two patients who died had extensive liquefaction necrosis of the oesophagus, which led to severe mediastinitis in one and perforation of the aorta nine days after removal of the battery in the other. The three remaining patients suffered a second degree burn of the oesophagus (one) or tracheo-oesophageal fistula (one) or required a prolonged stay in hospital (one).

The battery went beyond the oesophagus in the five other patients with symptoms. Four of these batteries split, resulting in symptoms of mercury poisoning in two children. Two other patients with dark discoloration of