Rapidly fatal encephalopathy with shigella infection

Convulsions in children with shigella infection are well recognised (see Archivist 1991, p203) but probably most paediatricians are less familiar with shigella-associated fulminating encephalopathy. It may even have slipped your mind that three cases from South Africa were described in this journal in 1983.¹ Further data from Israel (Avner Goren and colleagues, *Pediatrics* 1992;89:1189-93) put the incidence of this complication in children under 10 years of age with shigellosis at one in 4600, whereas convulsions have been estimated to occur in between 12 and 45% of such children. My one in 4600 occurred not long ago so I'm sensitised to the subject.

In the 1980s in Israel there were 15 deaths of children with shigella found on stool culture. They were eight boys and seven girls and their ages ranged from 5 months to 11.5 years. All died from an acute fulminating encephalopathy with brain swelling and brain death usually within 48 hours. The 15 fatal cases were compared with 30 age and sex matched controls with non-fatal shigellosis but the two groups were very similar. Those who died did not have more severe gastrointestinal symptoms nor significantly more frequent convulsions. There was no evidence that the deaths were due to complications such as septicaemia, dehydration, fluid overload, disseminated intravascular coagulation, acute surgical abdomen, haemolytic uraemic syndrome, pneumonia, or meningitis. The only clinical differences between the two groups were an increase in mild hyponatraemia and in headache in the fatal cases but in no case was biochemical disturbance thought to be an important factor. The organism in the stools in the 15 study cases was *Shigella flexneri* in eight, *S sonnei* in four, and *S dysenteriae* in one. In two cases the type of shigella was not identified. In the 30 controls *S sonnei* was found in 21, *S flexneri* in eight, and *S boydii* in one.

The mode of death suggests a 'toxic' encephalopathy. Four patterns of neurological disturbance have been described with shigellosis: seizures or transient encephalopathy both of which appear to be benign, peripheral neuropathy described rarely and in adults, and the acute fulminating encephalopathy which has been invariably fatal in reported cases. The cause is unknown. It seems that shiga toxin and shiga-like toxins I and II are not the cause and a different neurotoxic protein has been implicated.

What can be done to prevent these deaths? Antibiotic treatment is unlikely to be the answer as the encephalopathy is so rapidly progressive that it seems extremely doubtful that giving an antibiotic after its onset would be effective and it seems unjustified to treat all cases of shigella infections especially as antibiotic resistance is common. The authors suggest that we must look to immunisation as the most effective means of reducing mortality in endemic areas.

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¹ Sandyck R, Prenon M J W. Fulminating encephalopathy associated with *Shigella flexneri* infection. *Arch Dis Child* 1983;58:70-1.