

NEUROMUSCULAR DISORDERS

GUILLAIN-BARRE SYNDROME AND CAMPYLOBACTER JEJUNI

A case control study of patients with Guillain-Barre syndrome (GBS) in South East England, reported from Guy's Hospital, London, has uncovered a strong association between *C jejuni* infection and a pure motor GBS, characterized by axonal degeneration either alone or combined with demyelination. *C jejuni* were isolated from the stools of 4 of 36 (11%) patients compared to 1 of 49 (2%) controls. A strong serological evidence of recent infection was found in an additional 5 patients. Of the total of 9 (25%) infected patients, 8 had a recent history of diarrhea, and 7 (78%) had one or more antibodies to glycoconjugates. (Rees JH, Hughes RAC. *Campylobacter jejuni* and Guillain-Barre syndrome. Anna Neurol Feb 1994;35:248-249). (Respond: Dr JH Rees, Department of Neurology, UMDS, Guy's Hospital, London SE1 9RT, UK).

COMMENT. The combination of recent *C jejuni* infection and positive anti-ganglioside GM1 antibodies heralds a poor prognosis in patients with GBS. The association of antecedent infection with *C jejuni* and GBS is also reported from the University of Texas Health Science Center, Houston (Vriesendorp FJ et al) and Julius-Maximilians-Universitat, Wurzburg, Germany (Enders U et al) (Ann Neurol Feb 1994;35:249).

A role for *C jejuni* infection in the etiology of a Chinese paralytic syndrome (acute motor axonal neuropathy) which resembles GBS has been proposed. (Gordon N. Arch Dis Childhood 1994;70:64-65). This disease shares clinical and CSF findings with the demyelinating GBS, but electrophysiological tests indicate an axonal neuropathy. A febrile illness preceded muscle weakness in 30% of patients, and some had diarrhea. Outbreaks of the Chinese paralytic syndrome associated with diarrhea and *C jejuni* infection have also occurred in Japan and Bangladesh. Environmental waterborne infections may have serious neurological complications. (Environmental Food Poisons, PNB Publ).

DISTAL VACUOLAR MYOPATHY IN CYSTINOSIS

Distal vacuolar myopathy in 13 post-renal-transplant cystinosis patients, ages 17 to 27 years, and studied at multiple centers, is reported from the National Institute of Child Health and Human Development, Bethesda, MD. Among 54 untreated patients with cystinosis, 13 (24%) developed hand weakness and wasting, sometimes accompanied by facial weakness and dysphagia, and becoming progressively more generalized. Tendon reflexes were preserved, and sensory testing and nerve conduction velocities were normal. EMG of affected distal muscles showed reduced amplitude and brief duration voluntary motor unit potentials. Muscle biopsy revealed fiber size variability, acid phosphatase-positive vacuoles, and absent fiber grouping or inflammation. Muscle cystine content of clinically affected muscles was markedly elevated. The cause of the distal myopathy was unclear. Systemic complications of nephropathic cystinosis or its treatment were excluded. (Charnas LR et al. Distal vacuolar myopathy in nephropathic cystinosis. Ann Neurol Feb 1994;35:181-188). (Respond: Dr Charnas, NICHD, NIH, Bethesda, MD 20892).

COMMENT. Distal myopathy is a relatively common late complication of nephropathic cystinosis. Cysteamine therapy may prove effective.