The pathogenesis of the cauda equina syndrome of ankylosing spondylitis is unknown. The long duration between the onset of ankylosing spondylitis and neurological symptoms (average 35 years in the Mayo Clinic series¹) argues against a shared inflammatory cause, as does the relative normality of CSF.^{1,2} Matthews suggested that arterial pulsations transmitted to the CSF might produce not only the bony erosion and arachnoid diverticula, but also contribute to sacral nerve damage.⁴ Atrophy of peridural tissues and adherence of dura to adjacent structures, as documented at operation^{1,6} and pathologically,⁴ might reduce elasticity and compliance of the caudal sac so impairing its ability to dampen CSF pressure fluctuations. Such excessive pulse pressure in CSF may, over the course of many years, produce the arachnoid diverticula and bony erosions and also have a deleterious effect on nerve roots.⁴ The impression that the cauda equina syndrome more often afflicts those with mild ankylosing spondylitis who remain ambulant may be a reflection of this pathogenetic mechanism. Intrathecal shunting could dampen such pathological pressure oscillations and hence might retard progression of the neuropathy.

A review of previous cases of cauda equina syndrome associated with ankylosing spondylitis has indicated that neither steroid treatment nor surgical exploration is of proved utility.² Moreover, instances of clinical deterioration after surgical intervention on the spine have been documented.^{1,6} Neurological improvement after L3-L5 laminectomy and marsupialisation of arachnoid cysts has been reported, but in this single case there was evidence of compression of the nerve roots, a distinctly uncommon finding in the idiopathic cauda equina syndrome of ankylosing spondylitis.⁷

The use of lumboperitoneal shunting is established for the treatment of idiopathic intracranial hypertension and cranial cerebrospinal fluid fistulae, but previous reports of its use in the cauda equina syndrome of ankylosing spondylitis have not been found. In view of our clinical findings, and the desirability of avoiding radical surgical intervention on the spine in ankylosing spondylitis, we suggest that lumboperitoneal shunting merits consideration in patients with ankylosing spondylitis presenting with an idiopathic cauda equina syndrome. If excessive CSF pressure fluctuations are important in pathogenesis, a case may be made for early surgical intervention by lumboperitoneal shunting in ambulant patients before the development of nerve damage.

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1 Bartleson JD, Cohen MD, Harrington TM, Goldstein NP, Ginsburg WW. Cauda equina syndrome secondary to long-standing ankylosing spondylitis. *Ann Neurol* 1983;14: 662-9.

2 Tullous MW, Skerhut HEI, Story JL, et al. Cauda equina syndrome of long-standing

ankylosing spondylitis. Case report and review of the literature. *J Neurosurg* 1990;73: 441-7.

3 Thomas DJ, Kendall MJ, Whitfield AGW. Nervous system involvement in ankylosing spondylitis. *BMJ* 1974;1:148-50.

4 Matthews WB. The neurological complications of ankylosing spondylitis. *J Neurol Sci* 1968; 6:561-73.

5 Kerslake RW, Mitchell LA, Worthington BS. Case report: CT and MRI of the cauda equina syndrome in ankylosing spondylitis. *Clin Radiol* 1992;45:134-6.

6 Auquier L, Siaud JR, Guiot G, Lhermitte F. Syndrome de la queue de cheval au cours de la spondylarthrite ankylosante: découverte opératoire d'une fusion entre vertèbre et dure-mère dans la cas rapporté. *Rev Rhum Mal Osteoartic* 1974;41:733-7.

7 Shaw PJ, Allcutt DA, Bates D, Crawford PJ. Cauda equina syndrome with multiple lumbar arachnoid cysts in ankylosing spondylitis: improvement following surgical therapy. *J Neurol Neurosurg Psychiatry* 1990;53:1076-9.

Diffuse neurofibrillary tangles with calcification in a non-demented woman

In 1994 we proposed the term "diffuse neurofibrillary tangles with calcification" (DNTC) for a new form of presenile dementia.¹ This disease is clinically characterised by progressive cortical dementia. Neuropathological features consist of temporal or temporofrontal atrophy with neuronal loss and astrogliosis, numerous neurofibrillary tangles spread throughout the cerebral cortex but lacking senile plaques, and Fahr's type calcification.

Recently, Langlois *et al* reported a case of DNTC without evidence of dementia, and pointed out that DNTC is not necessarily associated with dementia.² We have experienced a similar case.

A 64 year old woman was admitted to a mental hospital with anxiety attacks and hypochondrial complaints. Despite mild memory disturbance, dementia was not detected. She had hypochondria and delusions of persecution. She was dependent, and often displayed a negativistic attitude. Personality changes were considerable. At the age of 70 years, she fell down and soon died. At necropsy, fresh subdural bleeding with brain oedema (the cause of death) was found. The brain weighed 1265 g. Bilateral temporal atrophy was not so severe as in our previous patients with DNTC. Numerous neurofibrillary tangles were present in the hippocampus, entorhinal and transentorhinal cortex, and amygdala, but sparsely distributed in the neocortex. No senile plaques were found. Fahr's type calcification was present. Because of the lack of evidence of dementia, this case was not clinically diagnosed as having DNTC.

In this case neuronal loss and neurofibrillary tangles, which are thought to contribute to dementia, were much less obvious than those in our previous patients with profound dementia. Therefore, we diagnose this patient as having early stage DNTC.

Although Langlois *et al* did not describe the detailed distribution and degree of neurofibrillary tangles in their patient, it is possible that their case also exhibited early stage DNTC.

As we pointed out, all reported cases except one were Japanese. Recently, DNTC has received considerable attention, and more clinically diagnosed cases of DNTC have been reported in Japan.

The CT and MRI findings, consisting of localised temporal or temporofrontal atrophy and pronounced pallid and cerebellar calcification, are so characteristic of DNTC

that clinical diagnosis is not difficult. More cases of DNTC are expected to be reported, probably from other countries as well as Japan.

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1 Kosaka K. Diffuse neurofibrillary tangles with calcification: a new presenile dementia. *J Neurol Neurosurg Psychiatry* 1994;57:594-6.

2 Langlois NEI, Grieve JHK, Best PV. Changes of diffuse neurofibrillary tangles with calcification (DNTC) in a woman without evidence of dementia. *J Neurol Neurosurg Psychiatry* 1995;59:103.

Neurology of adult α -mannosidosis

Neurological findings in adult α -mannosidosis are cerebellar dysfunction, absent tendon reflexes, spasticity, and mental retardation.^{1,2} We present the most extensive longitudinal observation in one of the oldest known patients with adult α -mannosidosis.

In 1995, a 34 year old white man was seen for evaluation of progressive gait ataxia. Clinical data for this patient at the age of 4 have been reported.³ He had delayed developmental milestones from 4 months on; the patient did not sit without support until 2 years, and he first walked and spoke single words at 3 years of age. During early childhood he had recurrent respiratory infections. In 1967, mental retardation (IQ 60), hepatosplenomegaly, dysostosis multiplex, coarse facial features, severe deafness, but no ophthalmological abnormalities were noted. Cytoplasmic vacuoles were seen in 30% of the patient's peripheral lymphocytes and in a few peripheral lymphocytes from his parents. Many large foam cells were seen in a bone marrow aspiration from the patient. Further laboratory investigations including a urinary mucopolysaccharide screen gave normal results. Sural nerve biopsy showed neuropathy with myelin degeneration and metachromatic deposits. Based on these findings "lipomucopolysaccharidosis", subsequently called mucopolidosis I, was diagnosed.³ After a follow up investigation at 12 years, this diagnosis was abandoned; the patient was then classified as having α -mannosidosis.⁴

Since 1967, the patient has lived with his family and is now employed in a sheltered workshop. The parents were of Ukrainian origin and were first cousins. His three sisters, aged 35, 39, and 40 years are clinically healthy. The patient was mentally retarded but with an alert and pleasant personality, with brachycephalia and coarse facial features (prominent forehead, hypertelorism, wide spaced teeth, and a flattened nasal bridge). His height was 170.5 cm and his weight was 62 kg. He needed assistance to sit up and crutches for walking. He had pronounced kyphoscoliosis with gibbus deformity. Passive motion of both hips was limited in all ranges and painful. There was splenomegaly. No cardiac murmurs were heard; blood pressure and pulse were normal. Neurologically, there was no deficit on ocular and facial motor testing, pupils reacted normally on both light and convergence. There was bilateral deafness. The patient had slurred speech, clumsy tongue movements, and spoke sentences of only one to two words. Muscle power and tone were normal but the thigh muscles were wasted.