Ounsted[3] suggested that the gaze aversion of autistic children served to reduce arousal.

Very little is known about control of eye movement in patients with head injuries. Our patient's difficulty in disengaging from eye contact may be regarded as a form of the locking of fixation found in patients with Bálint's syndrome. Posner *et al*[4] have suggested that the act of shifting visual attention requires three operations; disengagement of attention from its current focus, followed by movement of attention to the target, and finally engagement of attention on the target. The parietal lobe is specifically involved in disengagement of attention. Our patient seems to have lost control of this disengagement system.

Two processes seem to have come together to result in our patient's symptom. On the one hand brain damage resulted in disinhibition of relatively primitive neurobehavioural responses to gaze in which the gaze of another on oneself is regarded as a threatening stimulus. This will have been exacerbated by neural mechanisms which result in selective fixation on eyes; babies show preferential fixation on eyes by seven weeks of age.[5] Thus the patient preferentially fixated on eyes, from which he had problems disengaging despite the profound arousal that he experienced on eye contact.

On the other hand his premorbid socially avoidant personality traits and aversion to disability will have facilitated the distress he felt when he was self consciously aware that others were aware of his disability. The most potent cue for such self consciousness is eye contact. The development of increasing self awareness on recovery from his head injury was therefore necessary for the fully developed behavioural syndrome and may partly explain why the symptom did not develop until three years after his head injury.

S FLEMINGER
Department of Psychological Medicine, St Bartholomew's and the Royal London School of Medicine and Dentistry, London E1 2AD, UK
L MURPHY
Royal Hospital for Neuro-disability, Putney, West Hill, London SW15 3SW, UK
WA LISHMAN
Institute of Psychiatry, De Crespigny Park, London SE5, UK

Correspondence to: Dr Simon Fleminger, Department of Psychological Medicine, St Bartholomew's and the Royal London School of Medicine and Dentistry, Turner Street, London E1 2AD, UK

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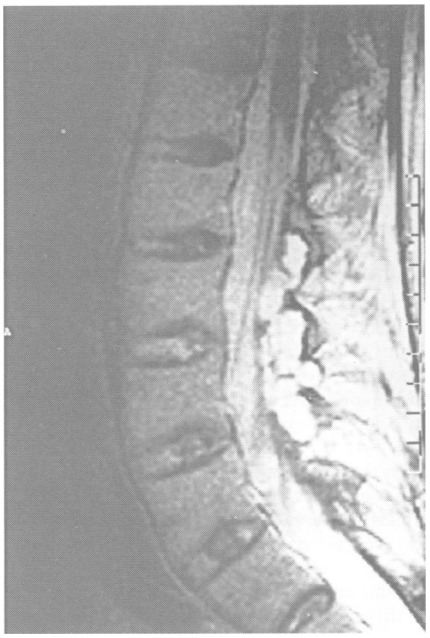
Arrested progression of the cauda equina syndrome of ankylosing spondylitis after lumboperitoneal shunting

An idiopathic cauda equina syndrome is a rare but well recognised complication of longstanding ankylosing spondylitis, usually developing many years after disease onset and after cessation of active joint pathology.[1 2] The frequency of this complication is unknown, but it is probably higher than the paucity of cases reported might suggest—for example, Thomas and colleagues found two cases among 45 patients with ankylosing spondylitis seen at this centre.[3] The characteristic pathological findings include erosion of the vertebral pedicles, laminae, and spinous processes ("vertebral scalloping"), widening of the thecal sac, and the presence of multiple dorsal arachnoid diverticula, but direct compression of the nerve roots is very uncommon. Affected nerve roots may show fibrosis and loss of myelin.[4] Clinically, the differential diagnosis includes compressive lesions of the cauda equina—for example, tumours (especially in patients with ankylosing spondylitis previously treated with spinal radiotherapy) but CT/MRI establishes the diagnosis.

Neither the natural history nor the pathogenesis of this condition are well defined. In the largest series reported to date (14 patients), the syndrome was slowly but relentlessly progressive without stabilisation or remission until complete sacral anaesthesia had developed, usually with double sphincter incontinence.[1] A review of 49 cases reported in the medical literature before 1990 concluded that 31 patients showed a progressive course, nine followed a stable course, and follow up was insufficient to permit comment in the remaining nine.[2] Of these 49, 47 eventually developed lumbosacral sensory disturbance, 44 sphincter disturbance, 27 motor deficits, and 23 pain.[2] Therapeutic interventions to try to arrest or reverse the progress of the syndrome, including the use of steroids, non-steroidal anti-inflammatory drugs, and surgical intervention, have produced disappointing results.[1 2]

A 56 year old man with a 32 year history of ankylosing spondylitis presented with sensory disturbance in the right leg. Aside from developing the typical bodily habitus of ankylosing spondylitis and having recurrent attacks of iritis in the left eye with subsequent cataract formation, his disease had caused him few problems. He took no regular medication and was able to walk a distance of several miles without difficulty. Radiographs of his dorsolumbar spine showed fusion of sacroiliac joints, squaring of the vertebrae and syndesmophyte formation, and ossification of interspinous ligaments and apophyseal joints, appearances typical of the "bamboo spine" of ankylosing spondylitis. Tissue typing was positive for HLA antigen B27.

Four years before presentation he noticed persistent numbness over the lateral border of the right foot, and seven weeks before presentation numbness over the right buttock. Clinically, all sensory modalities were impaired in the right L5, S1, S2, and S3 dermatomes, and the right ankle jerk was lost, but power was preserved. His left leg was normal. Electrophysiological studies failed to detect somatosensory responses from the right S1 dermatome and the right posterior tibial nerve. Some loss of motor fibres in the right lumbosacral distribution was indicated by delayed and abnormal F waves conducted via the right posterior tibial nerve in the foot and low amplitude evoked muscle action potentials in the abductor hallucis on stimulation of the posterior tibial nerve. Neurogenic abnormalities were found in the EMG of the right biceps femoris. Lumbar spine MRI showed erosion of the posterior lumbar arches with a wide and capacious spinal canal, in which multiple dorsal arachnoid diverticula were seen (figure). These electrophysiological and MRI findings are characteristic of the cauda equina syndrome of ankylosing spondylitis.[1 2 5]



T2 weighted MRI (sagittal section) of the lumbar spine, showing a capacious thecal sac and the roots of the cauda equina stretched over multiple dorsal arachnoid diverticula.

Further clinical deterioration occurred during follow up, with extension of the area of sensory impairment in the right leg to involve S4, sensory symptoms affecting the left buttock, and loss of vibration sense to the left ankle; the left plantar became extensor. These symptoms and signs were not influenced by an epidural steroid injection.

In view of the progressive nature of the patient's neurological deficit and the excessive spinal subarachnoid space, the relatively simple procedure of lumboperitoneal shunting was considered. This was carried out under general anaesthesia without complication. Cerebrospinal fluid taken at operation showed a normal cell count and protein concentration.

During 36 months of postoperative follow up, the patient's neurological symptoms and signs have remained unchanged. There has been no recovery of the function lost before operation, but no new neurological deficit has developed. Electrophysiological studies and lumbar spine MRI are also unchanged.

Although only limited information is available concerning the natural history of the cauda equina syndrome of ankylosing spondylitis, it seems to follow a slow but relentless progression in most cases, until complete sacral anaesthesia with impaired sphincter function is reached.[1 2] Intermittent and spontaneous periods of stabilisation do not seem to occur. Hence, a 36 month period of neurological stability, as seen in our patient, would seem exceptional, the more so in view of his continuing deterioration before operation. We therefore think that lumboperitoneal shunting has at worst stabilised his neurological deficit for a time and at best arrested its progression.

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 series[1]) 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 progession 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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ANDREW J LARNER
HARDEV S PALL
University Department and Regional Centre for Neurology
ANTHONY D HOCKLEY
Department of Neurosurgery, Queen Elizabeth Hospital, Edgbaston, Birmingham B15 2TH, UK

Correspondence to: Dr AJ Larner, National Hospital for Neurology and Neurosurgery, Queen Square, London WC1 3BG, UK.

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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 astrocytosis, 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 pallidal 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.

KENJI KOSAKA
Department of Psychiatry, Yokohama City University School of Medicine, Kanazawa-ku, Yokohama, Japan,
KENJI IKEDA
Division of Neuropathology, Tokyo Institute of Psychiatry, Tokyo, Japan

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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 mucolipidosis 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.