elemental diet alone has been shown to be very effective in inducing remission in childhood Crohn's disease. In our child, however, the contribution of steroids to the bone disorder was likely to be minimal in that osteopenia was present in bone age radiographs and DEXA scans early in the illness after a very short duration of treatment. Sex steroids are known to be important mediators of ossification and of the pubertal growth spurt. Our child had evidence of both malnutrition and delayed puberty both of which are likely to have contributed to the poor bone mineralisation as BMD is known to increase with weight and pubertal Tanner stage and these have been found to be the best predictive indicators of bone mass and BMD. The marked improvement in our child's lumbar spine BMD was achieved by both improving his nutritional status by controlling the underlying Crohn's disease and by inducing puberty.

The lesson to be learned from this case is that gross osteopenia may be present at the time of diagnosis and it is therefore important to exclude it in all children with Crohn's disease. We suggest that the bone mineralisation status of all our children with inflammatory bowel disease should be determined at presentation and at regular intervals throughout their treatment.


Dopa responsive dystonia

K Patel, T Roskrow, J S Davis, J Z Heckmatt

Abstract

There may be insufficient awareness of dopa responsive dystonia (DRD), which has a characteristic diurnal variation of symptoms. Two children are reported in whom the diagnosis of DRD was missed. The first was thought to have hysteria and the second hereditary spastic paraparesis. A full history is vital for the diagnosis of this important treatable syndrome.

(Arch Dis Child 1995; 73: 256-257)

Keywords: dopa responsive dystonia.

Segawa et al in Japan reported a group of children with progressive idiopathic dystonia where the symptoms deteriorated through the day and improved after sleep. There was complete resolution with a small dose of levodopa. A gene for this disorder, inherited as a dominant, has been recently mapped to 14q.

The differential diagnosis includes other dystonias, juvenile onset parkinsonism, spastic and psychogenic disorders. We describe two patients to illustrate early recognition and differential diagnosis of dopa responsive dystonia (DRD). We particularly emphasise the need to consider the diagnosis in a child presenting with a progressive, apparently hysterical or spastic, gait disorder.

Case reports

PATIENT 1

This 6 year old boy was referred by an orthopaedic surgeon. He had presented at 5 years old with a three month history of walking on his toes, particularly on the left side. His symptoms had worsened by a plaster of Paris splint. After referral to the paediatric department the bizarre nature of his gait initially suggested a hysterical disorder.

His symptoms had worsened coincident with physical activity during the summer holidays. His difficulty walking was asymmetrical, with stiffness of the left leg. Running was easier than walking. We suggested the diagnosis of DRD when his parents reported that he had no symptoms in the morning, could walk to school without difficulty, but required physical support to walk home, and in the evening could not walk more than a few paces unaided.

On examination he walked, slightly stooped forward, with a stiff gait on his toes on the left with some stiffness of his left knee. He had associated movements of both upper limbs when walking, with abduction of the arms, flexion of the elbows, pronation and flexion of the wrists and metacarpophalangeal joints, and extension of the fingers. Lower limb tone was mildly increased, more marked on the left, with brisk reflexes, ill sustained bilateral ankle clonus, and normal plantar responses. Despite his difficulties with walking he could run, jump, and hop. Radiography of his hips, computed tomography of the head, and copper studies were all normal and there were no Kayser-Fleischer rings.

He responded excellently to a small dose of levodopa (25 mg) and carbipoda (6.25 mg) (Sinemet LS). Within a week, he had...
Dopa responsive dystonia

improved and within one month, abnormal neurological signs had resolved, apart from the hand posturing. He is now taking full part in school sports.

PATIENT 2
This 10 year old girl had already been extensively investigated for a progressive gait disorder and diagnosed with hereditary spastic paraparesis. The diagnosis of DRD was considered after a chance remark by her grandmother to the physiotherapist that she had no symptoms in the mornings. Further questioning revealed progressive worsening during the day, requiring assistance to walk home from school. This history had been recorded in the notes several times but its significance never appreciated. The family also reported facial grimacing and abnormal posturing of the hands occurring at the end of the day when she was tired.

Her symptoms had been slowly progressive from the age of 5 years. Investigations at age 9 years included a blood film (excluding acanthocytosis), amino and organic acid chromatography, liver function, ammonia, copper and zinc concentrations, white cell enzymes, computed tomography of the head, and magnetic resonance imaging of the spine; all gave normal results.

On examination at 10 years, she had a spastic gait with toe walking, a mild thoracic scoliosis with a right rib hump, expressionless face, markedly increased tone in her lower limbs with brisk reflexes, ankle clonus, and spontaneously up-going plantars. At the end of the day, walking induced fisting or claspers together of the hands (the latter possibly to increase truncal stability).

She responded in a similar fashion to levodopa (12.5 mg) and carbidopa (3.125 mg), and is now taking part in full normal sporting activities. The scoliosis has resolved.

Discussion
These two reports illustrate some of the difficulties in diagnosis of this treatable syndrome. Most accounts consider torsion dystonia and other extrapyramidal disorders as the main differential diagnosis, but DRD should be considered in any progressive gait disorder, in particular where there is no ‘glued to the floor sign’ despite significant disability. This sign has been considered pathognomonic of hysteria. Objective clinical signs of organic involvement of the central nervous system make a diagnosis of hysteria untenable but early in the course of DRD these may be ‘soft’.

Hereditary spastic paraplegia is a heterogeneous disorder characterised by a particular ‘dragging’ type of gait, occasionally asymmetrical, which our second patient did not show. An association with chorea has been reported and the need to consider DRD previously emphasised. Our second patient’s extrapyramidal signs were subtle and it was the grandmother’s chance remark that led to the diagnosis.

A history of diurnal variation should always be sought, as it is a vital diagnostic clue. Patients with neurological disorders may become easily fatigued but loss of function at the end of the day is probably more significant. Sequential video filming might be useful in objectifying progressive loss of function.


Co-trimoxazole induced aseptic meningitis

Dinesh Pashankar, Maureen McArule, Anthony Robinson

Abstract
A 15 year old boy presented with two episodes of aseptic meningitis-like reactions after ingestion of co-trimoxazole. The diagnosis of co-trimoxazole induced aseptic meningitis was made. This syndrome should be considered in the differential diagnosis of aseptic meningitis.

(Arch Dis Child 1995; 73: 257–258)

Keywords: aseptic meningitis, co-trimoxazole.

Aseptic meningitis is relatively common in childhood and has many causes besides viral infections. In the adult literature there are several case reports of drug induced aseptic meningitis. After an extensive search Maignen et al found 11 cases after ingestion of co-trimoxazole and eight cases after trimethoprim. So far we are aware of only one case report of aseptic meningitis after co-trimoxazole in childhood. This drug is currently widely used in paediatric practice and therefore to increase

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