# A NEUROLOGICAL SYNDROME IN CHILDREN RECOVERING FROM MALNUTRITION*

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This article discusses some of the features of a neurological syndrome which has been observed in Johannesburg in 16 children recovering from malnutrition (kwashiorkor).

Since publication of my first account of this condition[1] I have received communications from other centres indicating that this disorder is not merely of local interest. Thus one case has been noticed recently in Durban,[2] others in Central Africa,[3, 4] and Gerbasi has described the same disorder in malnourished, anaemic children in Sicily.[5] The syndrome is not mentioned in contemporary medical literature, but there is evidence that similar, if not identical, cases were seen in the USA, France, Italy and Austria when malnutrition in infants was still common there some 50 years ago. In 1909, 18 cases were published by Zappert[6] under the title 'Acute cerebral tremors'. He attributed them to toxic-infective causes.

*Symptoms and Signs*

The syndrome is characterized by coarse tremors, which affect most frequently the upper extremities and somewhat less commonly the legs, the muscles of the face, the tongue, the neck and the abdomen. The tremors may occur unilaterally or bilaterally or they may affect both sides of the body with different intensity. They cease only during deep sleep. Cog-wheel rigidity may be associated with the tremors.

Postural abnormalities can usually be detected in the upper extremities of these children. The arms are abducted at the shoulders and flexed at the elbows. The hands are held in ulnar deviation, the fingers are flexed at the metacarpo-phalangeal joints and extended at the interphalangeal joints, and the thumb is flexed into the palm.

Tremors of the tongue are associated with drooping of the lower jaw, but excessive salivation has not been observed.

The tendon reflexes are nearly always markedly exaggerated. A few cases have shown myoclonic jerks at a rate of 1 per second. Other inconstant features of the syndrome are muscular weakness, insomnia, epistho-tonus and irritability. There is no clouding of consciousness.

*Prognosis*

Recovery from the syndrome has been complete within weeks or months of onset, except in one child who died of intercurrent gastro-enteritis and in another who, nearly 3 years after cessation of tremors, was found to have markedly exaggerated tendon reflexes and who also suffered from mild mental retardation and grand-mal epilepsy.

*The Cause of the Syndrome*

None of the numerous investigations carried out on these children has yielded a clue to the pathogenesis of the neurological disorder.

Similar disturbances have been noticed in adult pellagrins[7] in whom a favourable therapeutic response was obtained with injections of pyridoxin hydrochloride. This vitamin was used in some of our cases with equivocal results.

It is also doubtful whether the syndrome is related to the portal-systemic encephalopathy of liver failure, which has aroused interest in recent years.[8] The two conditions seem to differ in that our cases have not shown any clouding of consciousness or a favourable response to a low-protein diet. Furthermore, histological examinations of the livers of our patients have revealed only mild fatty changes, and biochemical tests have indicated varying degrees of impaired hepatic function, findings which are almost the rule in children recovering from malnutrition.

At this stage, there is no explanation why the syndrome should occur in only about one of 300 severe cases of malnutrition in childhood.

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