Section of Neurology

President—DOUGLAS MCALPINE, M.D., F.R.C.P.

[*February* 6, 1947]

CLINICAL MEETING HELD AT NATIONAL HOSPITAL, QUEEN SQUARE, LONDON

Dystonia Musculorum Deformans with Another Case in the Same Family.—DIANA BECK, F.R.C.S.

A. S., a schoolgirl, aged 8½ years, regarded as hysterical, but was referred to the Neurosurgical Unit from an orthopædic clinic for exclusion of organic disease.

The child of a normal, full-term first pregnancy terminated by a normal delivery without birth injury, the patient was perfectly well until a year ago when she began to kick up her left heel on walking. This was shortly followed by a similar uncontrollable disability on the right side; within six months she could only walk with support. Fine tremor of the hands had been observed throughout the illness.

The family history is of interest for the child's paternal uncle, L. S., now aged 39, began to have difficulty in walking at the age of 8 and with the years became progressively more unsteady. His disorder was diagnosed as chorea. He has never worked on account of his disability.

L. S. is an intelligent and otherwise healthy man. Spasmodic contractions of the masseters. Both sternomastoids contracted with periodic spasm; intermittent backward jerking of the head. Roving movements of the tongue. All limbs in continuous state of tremor and tension; rhythmic twitchings of biceps and triceps. Power in upper limbs good: impairment in lower limbs owing to severe spasm, especially marked in adductors and glutei. Great difficulty in walking: legs crossed; both feet in position of talipes equino-varus. No sensory or reflex disturbance.

The patient is a pale, auburn-haired little girl with a somewhat expressionless face. Very intelligent (I.Q. 148); emotionally mature and well balanced.

General examination normal. Cranial nerves normal. Roving movements of the tongue. Fine tremor of head and trunk, especially marked on standing up. Fine tremor of hands at rest; athetoid movements of outstretched arms; constant opening and closing movements of hands. There is diminished tone in the arms after initial difficulty in relaxation has been overcome; tone is increased in the lower limbs; both heels drawn up. No ataxia; no inco-ordination; no motor weakness. Fine movements somewhat clumsy. Stands and walks with hips and knees flexed; there is adduction and internal rotation at the hip-joints and the heels are drawn up whilst the feet are inverted. Gait very unsteady and only possible with support. Scoliosis with convexity to the right. Sensation normal. Tendon reflexes present and equal but difficult to elicit. Abdominal reflexes present and equal. Plantar reflexes flexor.

Investigations.—X-rays of skull and spine normal. Cerebrospinal fluid: clear, colourless: protein 25 mg.%; no cells; sugar 54 mg.%; chlorides 750 mg.%. Blood and cerebrospinal fluid: Wassermann negative. Liver function tests: Serum alkaline phosphatase 14 units; thymol turbidity 3 units; plasma proteins:—Total 5·9% = albumin 2·6%, globulin 3·02%, fibrinogen 0·28%, A/G ratio 1 : 1·2.

Electro-encephalogram.—Delta and theta discharge larger in postcentral regions and often focal on left side; almost completely inhibited on opening eyes. Frontal regions: strikingly little electrical activity. Electrical activity from central and postcentral structures closely resembles records from normal children of 1½ to 3 years. Tracings from precentral areas normal for child's age.

Electromyogram.—(*a*) Surface electrodes: motor unit discharge from both flexors and extensors. Relaxation slow. (*b*) Needle electrodes: some motor unit action potentials firing synchronously.

Comment.—In the absence of birth trauma, other injury or illness this condition must be ascribed to congenital defect. The points of interest are the family history, the changes in the plasma proteins and the electro-encephalograms. The patient has a diminished plasma protein content and reversal of the albumin-globulin ratio. It might be argued that there is some degree of liver dysfunction in this child; this suggests a link with Wilson's disease. The electro-encephalographic findings are important in view of recent American work in which precentral cortical excisions are undertaken in cases of dystonia musculorum deformans.

Post-encephalitic Parkinsonism with Marked Palilalia.—MACDONALD CRITCHLEY, M.D.

H. S., male, aged 40, sustained an attack of acute epidemic encephalitis in 1924. He made a good recovery until about 1934 when he began to develop drowsiness, and slowness and clumsiness of movements. Another disturbing symptom was a tendency for the eyes to turn up involuntarily (oculogyric crises). On this account he gave up work in 1941. In 1945 his speech showed the repetitive features of palilalia which are now so conspicuous.

Physical examination reveals a Parkinsonian syndrome of moderate severity, more marked on the right side, and with a mild and inconstant degree of tremor.

Dr. Critchley added that there were a number of interesting neuro-psychiatric features in this case. One was his tendency to compulsive thinking, especially during the oculogyric crises, but at other times also. He would be compelled to repeat to himself a number of rhymes or verses of poetry. He was also apt to identify himself in the cinema with the characters on the screen and this feeling would persist for a while even after leaving the theatre. A third feature of his case, which is now a thing of the past, was the occurrence of bouts of rapid noisy breathing. The outstanding characteristic at present is his palilalia whereby he repeats the last few words of a sentence over and over again—perhaps as many as 15 times. But if requested to narrate a preformed speech pattern (days of week, &c.) or to recite something he knew by heart, no palilalia would occur.

A final interesting feature is his tendency to continue unduly any repetitive act, such as hammering a nail, combing his hair, or brushing his teeth. Whether this movement-perseveration represents a real palipraxia, or whether his involuntary tremor in such circumstances takes control of his volitional movement, is uncertain. His tremor, however, was 5·5 per second and his movement repetitions were much slower and were perhaps on that account to be regarded as a true psychomotor disorder, or a palipraxia.

In reply to Sir Charles Symonds, who asked whether the patient still got his oculogyric crises, which on the whole carried a better prognosis than any of the other symptoms, Dr. Critchley said that though much less frequent than formerly he did not think they had entirely disappeared. His experience too was that these oculogyric crises, unlike the Parkinsonian syndrome, were features that improved.

The President asked about the psychological background of the patient, to which Dr. Critchley replied that the man's personality prior to his illness was not of the obsessional type.

Paraplegic Eunuch with Lues.—MACDONALD CRITCHLEY, M.D.

D. F., male, aged 46, entered hospital at the end of November 1946 on account of pains in the right side of the chest and in the right leg. Whilst under observation he developed numbness over the abdomen, and later, a total flaccid paraplegia with incontinence and loss of sensation up to the level Th. 6. Lumbar puncture: yellow fluid; Queckenstedt positive; 18 cells, protein 180 mg.%. W.R. and Kahn positive in fluid, negative in blood.

The patient was 6 ft. 2 in. in height, with long slender extremities, smooth hairless skin, small penis and testes, no beard, pubic or axillary hair, high-pitched voice.