Case of Myotonia.

By REGINALD C. JEWESBURY, M.D.

Girl, aged 9 years. The child was quite well until two years ago, when wasting and weakness began simultaneously in both legs. This gradually increased, and she developed such a marked degree of talipes equino-varus that tenotomy of both Achilles tendons was done at a provincial hospital. She was able to walk better for a time after this, but gradually the legs became very stiff, and the arms, especially the left one, and the muscles of the neck also became affected.

On April 2 the child was admitted to the Victoria Hospital. There was no history of any previous illness. Both parents are healthy, and there are three other children quite normal. No history of any muscular or nervous disease in the family. No evidence of syphilis in this child or in the parents.

On examination, the patient is a healthy looking child and very intelligent. Motor system: There is slight wasting of the calf and thigh muscles on both sides. No loss of power, but on attempting to make any movement the legs become very stiff. Double talipes equino-varus, especially marked on left side. She is unable to stand with the left heel on the ground. On walking the legs become very stiff; there is no inco-ordination. No ataxy nor Rombergism. Arms: Some wasting of muscles but not marked, no loss of power but rigidity becomes very marked when she tries to make any definite action. The movements are performed jerkily and stiffly. The left arm is much more affected than the right. When she tries to grasp any object with the left hand all the voluntary muscles of the body seem to be thrown into a state of spasm. Muscles of neck: There is a perpetual right-sided torticollis owing to spasm of the sternomastoid and trapezius, which becomes very marked at times. Muscles of trunk and back are in a more or less constant state of spasm. Facial muscles not affected. Cranial nerves all normal. Optic disks normal. Tendon reflexes normal when muscular spasm is relaxed; superficial reflexes normal; plantar reflex flexor; sphincters not affected. Sensibility quite normal to touch, pain, heat and cold. No tremors, no athetosis, no evidence of any visceral disease. Wassermann test of blood negative.
The case appears to be a very unusual one of some form of myotonus. There is no evidence of any organic nervous lesion. The fact that the condition developed when the child was aged 7 years, up to which time she had been quite well, and the absence of any family history, makes the diagnosis of Thomsen's disease unlikely.

The child has been treated with massage and passive movements. She appears to be able to exercise a certain amount of voluntary control of the spasm of the muscles.

DISCUSSION.

Dr. F. Parkes Weber: I think the case is one of organic disease in the basal ganglia of the brain, and that it therefore belongs to a group which includes Wilson's family progressive lenticular degeneration. Apparently a number of various conditions have been suggested as belonging to that group.

Dr. Helen Ingleby: The stiffness which this child presents has some resemblance to that of certain cases of paralysis agitans in which there is rigidity but no tremor. But in paralysis agitans rigidity is never present without some motor weakness in the affected limbs. If the case belonged to this group, one would expect the lesion to begin on one side, to be for a time hemiplegic in distribution, or finally to cross to the other side. Most of the progressive degenerations involving basal ganglia follow this course. In this child, apparently, the lesion began as a paraplegia and was never hemiplegic in distribution. At present there seems to be no motor weakness at all. If there is any tremor or athetosis, it is so slight as to be doubtful and is not such as would suggest disease of basal ganglia. The absence of weakness is strongly against the latter diagnosis and in favour of this case belonging to the group of myotonias.

Dr. Porter Parkinson: Without suggesting what this condition is, I think that at any rate it is not Thomsen's disease. There is, here, an absence of hereditary history, and an absence of the chief symptom of Thomsen's disease, that of rigidity, which passes off very soon after the movements are started. In this child, apparently, movements do not lessen the rigidity at all. The third point against it being Thomsen's disease is the fact that it is not universal: the right arm has, apparently, escaped entirely at present, and the only parts affected are the neck, legs, and, to some extent, the left arm.

Dr. Helen Ingleby; I was only putting forward the instance of paralysis agitans because the work, showing the function of those motor cells, has been done on cases of that disease. Most diseases involving the pallidal system are at first unilateral and later become bilateral. I did not say the lesions were necessarily symmetrical.

Dr. Jewesbury (in reply): The condition is asymmetrical in distribution. It appeared to start in the legs, and now the leg and arm of the same side are involved. The left side is more affected than the right. On the other hand, the muscles on the right side of the neck are chiefly affected, so it is not a hemiplegic condition now, it is much more generalized. In answer to what Dr. Parkinson said, I did not want to suggest it was Thomsen's disease. It seemed to me to be some form of myotonus, and I am afraid I rather shut my eyes to the fact that it might possibly have some nervous origin, so I am very glad to have that suggestion to think about.