

A CASE OF PSEUDO-MUSCULAR HYPERTROPHY.¹

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CASES of pseudo-muscular hypertrophy are, on the one hand, not so rare as to be considered curiosities, nor, on the other hand, so common as to be without interest. Strictly speaking, the affection does not fall within the category of nervous diseases, being, so far as is known, a disorder of the muscles, pure and simple. In all likelihood, its occurrence depends upon some embryonic defect, some imperfection in the mesoblastic layer of the developing ovum, from which the muscular and fibrous tissues are derived. The designation pseudo-hypertrophic paralysis or pseudo-muscular paralysis is objectionable, inasmuch as the loss of motor power is but an incident, a secondary phenomenon in the symptom-complex.

This case is presented upon the suggestion of Dr. Weir Mitchell, in whose service it occurred, and to whom I owe the privilege of making this report.

J. R., ten and one-half years old, presented himself at the Infirmary for Nervous Diseases on January 4, 1895. He was born at term, without complication, and during infancy had been fed at the breast. At the age of nine months he had whooping-cough, and shortly afterward chicken-pox. He learned to speak at the usual time, and his teeth appeared normally, though with some difficulty of minor character. He has never had a convulsion. No abnormality was noted until the child should have walked but failed to do so. For a time, at the age of about two years, he was able to crawl and creep, but he never walked unaided. He has tried crutches and braces, but without noteworthy assistance. For five years he has been using a tricycle. He is unable to start himself, but once set going he can continue

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the propulsion. He has been at school for but a few months, and has received but little education. He appears, however, bright and intelligent. The muscular inability has grown steadily and progressively worse.



The boy is entirely unable to stand unsupported. Even when seated, the lumbar spine yields in a forward direction, leaving a marked concavity behind and the sacrum prominent. The legs and feet can be variously moved, though somewhat feebly; the thighs, also, but in somewhat less degree. Flexion of the thighs is particularly weak. The legs and feet are intensely cyanotic; in some places bluish, in some deep red, in some pink. They are also cold, and their cutaneous covering is



rough. The calves of the legs and the buttocks are distinctly full and firm, but not indurated. The musculature of the remainder of the body is rather spare. The child is unable to flex the trunk forward, and rotates the spine little and with difficulty. The movements of the upper extremities are well performed, although the muscles are small. The knee-jerks cannot be elicited, nor can the muscle-jerks in the lower extremities. The various reactions in the upper extremities are preserved. The several cutaneous reflexes are maintained. Cutaneous sensibility is likewise preserved.² The head ap-

² A separate study of the reflexes and of sensibility was made by Dr. F. S. Pearce, whose notes are appended: Superficial reflexes—Infraorbital present; pupils respond normally; epigastric, abdominal and

pears large; the ears are large; the complexion is pale. The appetite is excessive, and the child overeats at times. The bowels are regular, and the sphincters are continent. The hand-grasp is exceeding feeble, but it seems alike on both sides, although the parents express the opinion that the right side is the weaker. The disposition is good and the child is bright and cheerful. The rhythm of the heart and the character of its sounds suggest mitral obstruction, but no murmur can be detected. Dr. A. G. Thomson reports that the ocular media are clear; that the fundus and optic disc are normal, although pulsation is evident in the right eye; and that the muscular balance is normal. Nearly three years ago the feet were operated upon, but a tendency to valgus remains. In the family history the only noteworthy points are that a baby-brother died soon after birth in consequence of injuries received from dystocia due to unusual size; that a sister died at the age of four years, having had spinal curvature and finally meningitis; and that another sister died at the age of nine months during dentition. Three other brothers are well and present no obvious abnormality.

For the photographs I am indebted to Dr. H. P. Boyer.

cremasteric especially active; gluteal less so; erector spinæ cutaneous reflex good. Deep reflexes—Knee-jerks absent, not reinforcing; no ankle-clonus; no Achilles' tendon response; elbow-jerk preserved, but faint; no contra-lateral jerk; jaw-jerk present. There is fibrillary contraction generally in the chest-muscles and girdle-muscles on tapping; also in the arms and forearms, but wanting in the legs. The feet are cold and covered with cold perspiration. From the buttocks down there is marked pseudo-muscular hypertrophy. Sensation everywhere is preserved to touch, pain and temperature, but is the more acute in the upper extremities. There is undue mobility at many of the joints, including those of the spinal column. The child assumes an attitude of lordosis in the endeavor to maintain a position of equilibrium in sitting. There is a marked increase of the connective tissue over the lumbar vertebræ.

A study of the electric reactions was made by Dr. J. H. W. Rhein, who found only quantitative changes, particularly marked in the lower extremities. The left biceps and rectus femoris and the right rectus, as well as the triceps of the left arm, failed to yield any reaction to the strongest current that could be employed.