

Pathological Anatomy has as yet had the opportunity to take cognizance only of the condition of the muscles. These have been examined during life by M. Duchenne, by means of an instrument named "emporte piece."

I submit a brief summary of the results of M. Duchenne's observations:—

1. There is a large increase of connective tissue, and of interstitial fibroid tissue.
2. This interstitial deposit is mixed up with a greater or less number of fat vesicles or adipose tissue.
3. The muscular fibres are pale. Their transverse striæ are generally preserved, but have become extremely fine, and in some places they have even disappeared.

Dr. Lockhart Clarke, who had examined a portion of muscular fibre removed by the means just mentioned, from a patient of Duchenne's, confirms these conclusions.

Incomplete, however, as the history of this disease is, there are facts enough collected concerning it to call the attention of physicians and pathologists to its further investigation. Indeed, so much towards this end has already been accomplished by M. Duchenne de Boulogne, that inasmuch as we have the example before us of rendering honor to whom honor is due when we speak of "Bright's disease," "Addison's disease," &c., I believe we should herein only be doing justice if in future we shortened the lengthy title of "Pseudo-Hypertrophic Muscular Paralysis of Duchenne" into the brief, though not very scientific, form—"Duchenne's Paralysis." I would, therefore, with great deference, submit that this name should be adopted, and I have bestowed it accordingly upon the cases described in the present communication.

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*Case of Mental Feebleness associated with Pseudo-muscular Hypertrophy.* By LANGDON DOWN., M.D. LOND., F.R.C.P., Physician to the London Hospital.

(Read at the Quarterly Meeting of the Médico-Psychological Association January 27th, 1870.)

REVIEWING a large number of cases of idiocy and imbecility which have come under my observation, I am able to call prominently to mind three cases in which apparent hypertrophy of muscle, combined with motor paralysis, was present. As these occurred to me before Dr. Duchenne, of Bologne,

had pointed out their significance, I did not take steps to verify their condition. So marked, however, was the apparent muscular development, that the muscular feebleness puzzled me much. Faradization was tried in one case without any good result.

That it is not very common among the imbecile class may be gathered from my having noted this condition of the muscular system in only three instances among fifteen hundred cases of idiocy and imbecility that I had, prior to my meeting with the subject of the present paper, investigated.

Among my out-patients at the London Hospital, I recognised a boy whose condition forcibly reminded me of the three cases I had before observed, and which a careful examination led me to believe was a typical example of the *paralysie myosclérosique* described by Duchenne. Not having in existence any very accurate notes of my previous cases, I determined to make a careful examination of the present one, in which I have been ably aided by my clinical assistant, Dr. Woodman, and by my clinical clerk, Mr. Herman.

B.R., æt. eleven, admitted into the London Hospital, September 14th, 1869.

*History.*—His mother states that this disorder has been coming on five years. The first thing wrong that she noticed was that when he fell down he could not get up again. About two years ago he began to get unsteady in his gait; when running, if he tried to stop himself suddenly, he fell down, and even when walking, he had difficulty in stopping without tumbling over. Running was easier to him than walking, and walking than standing still. When he stood still he looked out for something to lean against, getting to the wall if he could. His mother noticed that as he ran he used to stick out his belly and throw back his shoulders. For the last twelve months he has been unable to walk, and for the last seven months has been gradually losing the use of his arms.

*Family History.*—His mother's health has always been good, and so has his father's. No history of any hereditary disease, except that his grandfather died from "consumption," and his father's grandfather was insane. All his brothers and sisters are healthy.

The only previous illness the boy has had was measles, seven years ago. Has never had thrush or any cutaneous eruption. Has had thread worms for six years. His mother suckled him for eighteen months. He was not any longer

teething or learning to walk than the other children. He has always been very fat; his belly has always been big, and the calves of his legs have always been large. During the last four years his mother thinks his ankles and thighs have wasted. His head has always been large. During the past two years he has not been well fed; the family have only had meat once a week, and then one and a half pounds divided among ten.

*Present Condition.*—He is utterly unable to stand upon his legs; they sprawl helplessly in every direction. He can sit up, but if he be gently pushed back, he falls quite passively upon the floor, and can by no efforts raise himself. As he sits up he likes to support himself by using his hands as props. When pushed back in bed against the pillows, so that he does not quite fall down, he can manage to raise himself by swinging his head laterally to the front. There is great loss of power over the arms. On being desired to raise his arm slowly, he can move it but a little distance from his side, and that by the aid of his scapular muscles, the shoulder being raised and thrown forward, and the lower angle of scapula tilted upwards and outwards. By a jerk he can raise it about sixty degrees, and by swinging his arm backwards and forwards, he can get it up nearly horizontally. If his arm be extended in the supine position, he can flex it at the elbow joint, but if a small book be placed in his hand, he cannot raise it; a weight of three or four ounces prevents his doing so. He can perform any movements with his fingers and thumb, but without much power. When he wishes to raise his hand, he does so by the aid of his fingers, making them climb, as it were, over his body and head, and drag his arm after them. He separates his fingers, and places his thumb on a higher point than the rest, he then approximates his fingers, and resting his little finger on the same level as his thumb, again separates his fingers, and plants his thumb on a higher level still; in this way he makes his arm travel to the desired place. He cannot flex the thigh upon the trunk at all in bed; out of bed he can do it a little. As he lies in bed he moves his lower extremities about by the movements of his feet, alternately using the heel and toe as a fixed point to advance the rest of the foot. He can rotate the limb at the hip joint. Out of bed, by swinging his legs, he can get them to an angle of about forty-five degrees from the perpendicular, but he cannot keep them in opposition to the force of gravity. He can also

swing his feet laterally. He can move his feet a little laterally at the ankle joint, but without much power. He flexes and extends his toes readily. He can adduct the limb slightly, but appears to do so by the action of his biceps.

*Muscular System—Lower Extremities.*—The muscles supplied by the gluteal and great sciatic nerves are immensely hypertrophied. On looking at the boy, the prominence of his gluteal region is very apparent; and on feeling the part the extreme hardness of the muscle is very striking. The lumbo-sacral curve is greatly exaggerated. The tensor vaginae femoris can be felt as a thick, hard, firm band under the skin. The flexors of the thigh are also much hypertrophied. The muscles of the calf are very hard and large. The extensor and peronei muscles on the outside of the tibia appear to share in this hypertrophy. The foot is in a state of talipes equino-varus. The adductor muscles on the inside of the thigh are very soft, small, and lax. The quadriceps extensor appear to be somewhat hypertrophied, but it is very difficult to estimate precisely its size. The sartorius is wasted.

*Measurements of Lower Extremities.*—

From one iliac spine to the other—8 inches.

Around pelvis, over most prominent part of glutæi—24 inches.

From anterior superior iliac spine to outer condyle—12½ inches.

From great trochanter to outer condyle—10½ inches.

Circumference at middle of thigh—12 inches.

Tibia, from inner tuberosity to inner malleolus—9½ inches.

Circumference of thickest part of leg—11 inches.

Circumference of leg, just above the ankle—6 inches.

*Trunk.*—As before said, the lumbo-sacral curve is greatly exaggerated. The erector spinæ is immensely hypertrophied, and can be felt as a thick, hard mass on each side of the spine. The spinous processes of the vertebræ lie in a groove between these masses of muscle. In the space between the crest of the ilium and the last rib, the thick hard edge of the erector spinæ can be felt distinctly. The belly is big, but the muscles forming its walls do not appear to be either wasted or hypertrophied. The cremasters act readily. The chest is broad and deep, and the costal angle exceedingly obtuse. Its conical shape can be very distinctly seen, owing to the wasting of the pectorals. The serratus magnus is large, and its serrations can be seen with great clearness through the skin; it is not hard, like the erector spinæ, but appears to act naturally.

The action of the diaphragm in respiration was carefully watched, and found to be the same as in a healthy person. The intercostals appear to act fairly.

*Measurement of Chest.*—

From acromion to acromion	...	...	...	10	inches.
Circumference just below axilla:—					
"    on expiration	...	...	...	24	"
"    on inspiration	...	...	...	25½	"
Circumference on a level with ensiform cartilage:—					
"    on expiration	...	...	...	26	"
"    on inspiration	...	...	...	27½	"

*Head and Neck.*—The sterno-mastoid is rather wasted. The depressors of the os hyoides are capable of fairly vigorous action. It is doubtful whether there is anything abnormal with the trapezius, and the other muscles of that group; if anything, hypertrophied. His temporal muscles are undoubtedly much hypertrophied, and seen from the front, form a small tumour, outside each orbit. His masseters seem somewhat increased in size, but this is difficult to ascertain. The facial muscles of expression appear sluggish in their action. His orbiculares are wasted; he can shut his eyes, but cannot screw them up tightly. He cannot frown, and can elevate his eyebrows but little. The muscles acting on the mouth do not seem to be defective at first sight, but on making him go through various grimaces, it is seen that he has not the command over the upper lip that he has over the lower. This, perhaps, is from a misapprehension of his directions. He puts out his tongue in a peculiar way, it always being concave, and he does not protrude it beyond his lips unless told to put it further out, when he makes an effort; but it is never convex from side to side. No affection of any ocular muscle.

*General Appearance, etc.*—He is a dull, heavy-looking boy, with a stolid, but somewhat cunning expression of countenance. His face is very fat, especially about the lower part; complexion sallow and pasty; skin thick; superficial veins cannot be distinctly seen through it. His head is very large (circumference where the hat fits, 21 inches). Its shape is broad rather than long; hair thick, brown and straight; forehead rather high; eyes brown; pupils large; eyelashes long; eyebrows thick at outer part, thick towards the median line; nose flat and broad, especially broad at the bridge; alenasi thin; nasal aperture large; ears small but well

shaped, lobules not ill-developed; lips thick, upper lip thicker than lower; angles of mouth somewhat inclined downwards; thyroid gland enlarged, but not greatly; voice natural, clear, low-pitched: bones of extremities well shaped; epiphyses not unduly large.

*Mental Condition.*—He is rather dull and stupid, somewhat cunning. He is more generally good-tempered, but is often passionate, and at times very sulky. He appears to possess a strong will. (He has probably been spoilt at home). He is not timid or bashful in any way. He smiles when spoken to, answers questions readily, and any experiments tried upon him he takes in very good part. He cannot read, but he knows his letters. He has an appreciation of music, and amuses the other patients by his singing, whistling, etc.

*Nervous System.*—He sleeps well; has no fits or cramps of any kind; sight good; ophthalmoscopic appearances normal; hearing, taste, and smell good; sensation appears to be very good all over the body, both painful, tactile, and thermic. It cannot be estimated with the compasses, because the boy, whether from laziness or any other motive, does not—or, perhaps, cannot—give consistent answers. The atrophied muscles do not respond at all to electricity. The gastrocnemii and hamstring muscles contract under it. The other hypertrophied muscles were not tested with it.

*Digestion* and appetite good. Bowels regular.

*Urine.*—Sp. gr. generally rather high; acid, no albumen, no sugar.

*Respirations.*—18 per minute, taken naturally; no cough or dyspnoea.

*Heart.*—Cardiac dulness reaches above to third rib; on the right, to an inch beyond the sternum; on the left to within half an inch of nipple; apex beat under fifth rib, one inch to right of nipple, 88 per minute; impulse normal; sounds sharp, and clearly defined, somewhat exaggerated; no murmur or thrill of any kind.

*Temperature*, taken at different times, always normal.

*Therapeutics.*—I determined to try the effect of Faradization, uncomplicated with any medication; but, although this has been persistently employed for nearly three months, I am unable to detect the slightest improvement.

*Histological Condition of the Muscle.*—There could be no doubt that the case was one of the kind described by Duchenne, but I was desirous of completing the diagnosis by a microscopic examination of his muscular system. By means

of a harpoon I removed specimens from both gastrocnemii. The microscope exhibited a vast increase of connective tissue elements, especially of the white fibrous tissue. The muscular fibres had, for the most part, lost their transverse striation. Here and there the transverse striæ were visible, but more generally they were indistinct, and still more frequently absent. There were numerous fat globules in the connective tissue and on the sarcous particles, but it was clearly made out that they were exterior to the sarcolemma, and that the muscular element had not undergone fatty degeneration.

I think it unwise to call this state of things Duchenne's Paralysis. Confusion is likely to result, in consequence of Locomotor Ataxy being frequently so designated. Better, I think, to use the term *Pseudo-hypertrophic Paralysis* as combining all we at present know of it, clinically and pathologically.

Since the above remarks were written, I have met with another less advanced case among my out-patients at the London hospital, and in which I have failed as yet in detecting any well-marked determining cause for the malady.

*Two Cases of Atheroma of the Blood Vessels at the Base of the Brain, with remarks upon the symptoms, diagnosis, prognosis, and pathological condition in that affection.* BY J. T. SABBEN, M.D.

IN publishing the following cases, recently under my charge, of mental derangement dependent upon atheromatous deposit in the coats of the larger cerebral arteries, without any apparent disease of the brain substance, I desire, if possible, to define the symptoms of that condition during life, so as to enable them to be distinguished from those of general paralysis, with which I believe them often to be confused.

#### CASE I.

*Atheroma of the blood vessels at the base, dilatation of and effusion into the lateral ventricles, thickening of the dura mater, with slight effusion into the arachnoid.*

A gentleman, æt. 50, a merchant, married, with several children, whom I first saw August 22nd, 1868.

*History.*—Till within a month of this date he had always enjoyed good health, with the exception of occasionally suffering from slight