SPORADIC CRETINISM.¹

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THE two children whose portraits are here given afford good examples of this disease. The boys were brought to the Clinical Hospital last September, with the hope that something might be done for them, and perhaps, in the case of the younger patient it may still be possible for a proper course of education to ameliorate his mental condition; and with this object he is about to be sent to the Royal Albert Asylum, Lancaster. As regards the elder boy, however, there appears to be no prospect of any improvement.

Both patients are natives of Manchester, and have lived there since birth. Their family history, which is as follows, throws no light upon the etiology of the disease: their maternal grandfather was intemperate, but the other grandparents were temperate, and the parents are perfectly sober and healthy people, not related to one another except by marriage, coming, in fact, from different parts of England. No general causes of ill-health, no goitres in the family are known, and of five other children of the same parents none have shown the slightest symptom of cretinism or goitre.

The elder of the two boys, G. B., is 11 years old, 32½ inches in height, and weighs 34 lbs. He is the first child, and was born naturally. As not uncommonly happens in this disease, nothing roused the suspicion of his parents during his first year that he was not as other children. He was brought up at the breast, cut his first teeth at eight months, "noticed" tolerably early, and learned one or two of the usual childish words. When, however, he arrived at the age of twelve months, to use his mother's words, "he seemed to stop short;" his development (both mental and physical) ceased almost completely. He did not walk until he was 3½ years old.

At the present time he may be described in the following terms: His stature—and one might almost say, his intelligence—are those of a child of two years of age. He is very apathetic, and will sit for hours staring vacantly before him, or following with his eyes the other children in the ward, or poring over a picture book of which he has not the least com-

prehension. If he is compelled to walk he does so with a deliberate waddle, his legs wide apart, but it is patent that he is not only disinclined to exertion, but that, from his mental inertia, he would, if allowed, remain in one position (sitting or standing) for an indefinite time rather than take the initiative of changing. His expression is calm and disposition contented, but occasionally he has prolonged fits of sulkiness which are simply unconquerable. When pleased he has the smile common in cretins, slowly appearing and as slowly dying away; if highly delighted he laughs with a kind

of grunt. His voice is guttural, but he rarely utters a word, and has never spoken a connected sentence. All animals of which pictures are shown to him are either "dog" or "cat."

His head, though large in proportion to his height, is not so for his age (51 centimetres circumference), and is well shaped, with the exception of some flattening at the vertex. The anterior fontanelle is closed. His hair and eyelashes are normal. His features are broad and coarse, the eyes are set widely apart, the root of the nose being flattened, while the alæ and septum are thickened, and the nostrils are somewhat dilated. His thick everted lips are habitually open, with a large tongue protruding between them. There is no flow of saliva from the mouth. The face looks cedematous, the normal depression between the lower eyelid and the cheek being almost obliterated, but the skin is quite firm to the touch. There is a deep naso-labial line on either side of the mouth. His complexion is a dull yellowish white. He has only the blackened stumps of his milk teeth, worn down to a level with the gum in the front of the upper jaw, and in the lower his first teeth, set widely apart, are decaying, and some permanent incisors are making their appearance behind. A point of great interest lies in the fact that the thyroid gland is present and can be felt in his short neck as a small, firm, and

G. B., 11 years.

D. B., 6 years 3 months.

unsymmetrical body. A common feature in sporadic cretinism consists in the presence of lobulated masses of fat (pseudo-lipomata) in the supraclavicular regions or elsewhere, but in this case these swellings are entirely absent. The body is very bulky and the abdomen prominent. He has a small umbilical hernia. His chest is unsymmetrical, and he has a well-marked lateral curvature of the spine in the lower dorsal and lumbar regions. There is also a considerable lordosis, reminding one of pseudo-hypertrophic paralysis. The viscera of the chest and abdomen appear to be healthy, but his cir-

¹ Read before the Manchester Medical Society.

circulation is defective, his extremities being almost invariably blue and cold. He has a tendency to bronchitis in the winter months. His limbs, with their bony framework, are short and thick, but there is no absolute distortion except a bowing of the tibiae with the convexity forwards. The hands and feet are short and broad, with stumpy fingers and toes. The feet are flat, and the second toe on each foot is smaller than normal and overrides the rest. His skin is universally harsh, dry, and resistant, and his muscles are firm, but not larger than normal. There are no local swellings about the body. His senses of touch, sight, and hearing, so far as can be ascertained, are normal, though perception is distinctly retarded. He has occasionally suffered from slight hæmorrhages from the bowels, and once, when he had two teeth drawn, he bled considerably. His tendon reactions and superficial reflexes are active. He has never had convulsions.

D. B., aged 6 years and 3 months, is 33 inches in height, and weighs 32½ lbs. He is the fourth child (a healthy boy and girl intervening between him and G.). He cut his first teeth early, and is said to have been bright as an infant, but he did not walk until he was 2 years and 3 months old. Although still characteristic of cretinism, his expression appears to denote more alertness than that of his brother. He talks very little and usually wears an air of contentment, smiling very readily. He is subject to fits of sulkiness, exactly similar to those mentioned in the case of G. He waddles when he walks, but can move much faster than his brother. He can even run. His head is fairly well shaped, somewhat flattened at the vertex, with a depression in the region of the anterior fontanelle, which is closed. His hair is normal. The root of the nose is broad and flat and the tip is rather turned up, the alæ and septum are thickened and the nostrils are broad. He has rather a big mouth, which shows a tendency to remain open, but he only occasionally protrudes the tongue. The lips are not everted, but the lower lip is larger than usual. His cheeks are not puffy and there is the normal furrow beneath the lower eyelids. He has a pale yellow complexion, the cheeks being rather more yellow than the rest of the face. He has his first set of teeth, which are set widely apart. His neck is short and thick, and he has a small thyroid gland of very much the same character as that of his brother. There are no pseudo-lipomata about the neck or body. His abdomen is prominent and has a small umbilical hernia, and his chest is remarkable in the fact that it quite retains the circular form of infancy. There is a slight heaving of the ribs. A faint systolic murmur is audible at the apex of the heart, but not at the angle of the scapula, and there is no increase in the area of cardiac dullness. The rest of his viscera appear to be normal. His limbs show the same peculiarities as his brother's, including that of the second toe in each foot. His perceptive faculties are probably somewhat more acute than his brother's, but it is difficult to determine to what extent; there can be no doubt of his being at the present moment of extremely limited intellectual capacity. His skin is harsh and firm to the touch, and his circulation is sluggish. His tendon reactions are active. During his stay in the hospital he has passed blood from the bowels on one occasion. This however may have been due to the constipation from which he has constantly suffered.

A distinction has recently been drawn by Dr. Bourneville¹ between endemic and sporadic cretins, which appears to be somewhat unnecessary. He lays stress on the fact that endemic cretins are descended from goitrous parents, while sporadic cretins are not. Speaking generally, this statement may be correct, but Dr. Bourneville also proceeds to separate the two forms on the ground of their anatomical quite as much as of their etiological differences. He points out that endemic cretins have brachycephalic heads, are very frequently goitrous, and have no "pseudo-lipomata;" while, on the contrary, sporadic cretins have dolichocephalic heads, are invariably without thyroid glands, and present the "pseudo-lipomata." It may be that the etiological conditions of the two forms of disease are different; our knowledge at present is so limited that we are hardly in a position to discuss the question, but when we turn to the clinical features we scarcely find the difference between them sufficiently great to warrant our accepting them as two totally distinct diseases.

In the first place, it must be acknowledged that, although in the endemic form the disease appears able to go a step further in its effects, that is, the mental and bodily degeneration may be greater than is known to occur in sporadic cretinism, the two forms not only present the closest identity of symptoms, but have now been conclusively proved to depend upon the same fundamental pathological conditions—namely, loss of function of the thyroid gland. Then as to details—not more than two-thirds of endemic cretins are affected with goitre,² while cases have been described in which the thyroid gland has been entirely absent. On the other hand, as regards the statement that all sporadic cretins are destitute of thyroid gland, the patients described above are instances of its presence, and the late Dr. Hilton Fagge mentioned one case in which there was a goitre. Dolichocephalic heads, as seen in the above cases, are the rule in sporadic cretins, but brachycephalic heads have been described. Finally, the "pseudo-lipomata," though frequently met with in sporadic cretins, are not invariable, and when present are not always permanent, and as regards their presumed invariable absence in endemic cretins, it has been plausibly suggested that they have not been systematically looked for.

Minor points of difference between the two forms are mentioned by Dr. Bourneville, but as they are still more differences of degree rather than of kind than the above, they need not further be dwelt upon. For my part, I am satisfied that the distinction between the two forms is sufficiently marked by the terms endemic and sporadic cretinism.

EXPERIMENTAL RESEARCH AS TO THE PHYSIOLOGICAL ACTION OF PARA- CRESOTIC ACID.

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IN accordance with the promise made in a letter to the *BRITISH MEDICAL JOURNAL* of December 6th, 1890, I instituted a further series of experiments with regard to para-cresotic acid. This acid was forwarded to me by Messrs. Hopkins and Williams, London, and with it there was also sent at my request a sample of ortho-cresotic acid. Both these acids had been prepared from the corresponding cresols. I also obtained from another source samples of these acids, but, except for confirmatory purposes, they were not used, as their characters and melting points were identical with those obtained from Messrs. Hopkins and Williams.

Physical Characters.—*Para-cresotic acid*, melting point 150° C. to 151° C., a whitish powder, resembles the para-cresotic acid isolated from commercial salicylic acid by Professor Dunstan. The physiological action of this para-cresotic acid was stated at the meeting of the Pharmaceutical Society of London. *Ortho-cresotic acid*, melting point 163° C., distinct crystals. The only difference between it and the specimen of the ortho-cresotic acid received from Professor Dunstan was that in the latter the crystals were somewhat larger.

Physiological Experiments with Para-Cresotic Acid.—For this purpose two strong healthy rabbits were selected—(a) weighing 2½ lbs., (b) weighing 3½ lbs.

(a) To this animal an injection of 6 grains dissolved in rectified spirit was given. Fifteen minutes afterwards the fore-limbs slipped down, and the animal was unable to move easily. In forty-five minutes the hind limbs were stretched out, and a prone position was assumed. At the expiration of an hour and a half from receiving the injection the symptoms noted were "a prone position, great prostration, the head resting between the fore limbs, the breathing hurried and short." Three hours after the injection was given it died, the paralysis being complete half an hour before death.

(b) To this animal an injection of 8 grains similarly dissolved was given, and at the expiration of an hour other 4 grains. The result of the first injection was distinct disturbance, which deepened after the second injection into complete paralysis. Death occurred at the expiration of three hours. The same results followed upon injections of the two acids obtained

¹ *Progrès Médical*, Nos. 29, 30, 33, and 34, August, 1890.

² Baillarger and Krishaber, *Dict. Encycl. des Sci. Méd.*, Art. "Crétin."