

from the disease. About the diagnosis in these three persons there can be, I think, no doubt ; that the father and mother were typical and undoubted cases no one of experience would question. Perhaps about the son some might be inclined to doubt, but it appears to me practically certain that he had the misfortune to die during his first congestive attack, and thus deprived us of the opportunity of further and minute observation of what must be regarded as a singularly unusual and interesting condition. No doubt some of the classical signs were wanting, but there were enough found during life and after death to make the diagnosis certain to my mind.

To have read a clinical account such as this upon general paralysis and to have omitted the word syphilis, must appear strange. We have no absolute proof unfortunately of specific infection in the case of father or mother.

The reason is that minute information could not be obtained which at the same time should be absolutely reliable regarding a person so long dead. The father was a loose liver—that is known. We cannot with certainty include syphilis, but at the same time it cannot be excluded.

The boy did not present any absolutely typical signs of congenital syphilis ; he had, however, a large head, with prominent parietal eminences, and he had irregular teeth. Many cases of juvenile general paralysis show no signs of syphilis on their body, though they are proved to be the offspring of syphilitic parents. The boy himself had never acquired syphilis. My own belief is that the boy was a juvenile general paralytic, the offspring of two people the subjects of general paralysis, who had in their earlier days acquired a specific infection.

(¹) A paper read at the Spring Meeting of the Northern and Midland Division at Storthes Hall, April 30th, 1908.

A Case of Cretinism. (¹) By GUY R. EAST, Assistant Medical Officer, Northumberland County Asylum.

S. B—, of no occupation, was born in co. Durham forty years ago and came under observation at the Northumberland County Asylum in March, 1908.

His certificate reads :

He states that his age is 45. He cannot read or tell the time, or

count beyond twelve, though he was taught. His whole conduct is not that of an adult, nor is it developed as that of a young child. His speech and appearance are those of a cretin.

His intellectual powers are undeveloped in proportion to his age. He is childish and is incapable of answering the simplest question in spelling and arithmetic. He is mischievous and subject to fits of temper, and is quite incapable of managing his own affairs.

The family history is negative. None of the patient's relatives suffered from goitre. The parents were in no way related to each other previous to marriage: the father died of "some internal complaint" at the age of 57, the mother died of senile decay aged 78. Seven children were born, of which four died in infancy, but inquiries elicit the fact that none of these presented signs of cretinism.

Regarding his personal history, the goitre appeared during his first year, and since then has gradually increased in size. At the age of two he was seized with convulsions, and from that date onwards did not develop mentally, though endeavours were made to teach him. All his life he has required attention like a child, and has been incapable of learning a trade or useful occupation. During childhood he was never treated with thyroid extract.

His condition on admission was as follows:

He stands 4 ft. 9 in. and weighs 8 st. 2 lb. The relatively normal size of the head contrasts with the dwarfish body. He has a degraded type of face—a receding forehead, eyebrows absent, eyes set rather widely apart, bridge of nose depressed with thickened *alæ nasi*, a prominent chin, a wide mouth with broad, thick lips, hair thin and brittle. The limbs and body are short and stunted, whilst there is some enlargement of epiphyses of knees and elbows. The long bones are generally shortened with the exception of the clavicle. There is heaviness in the limbs and movements are sluggish and clumsy. The hands are stunted and undeveloped. The thyroid gland is hypertrophied, the enlargement being chiefly confined to the right lobe, and is about the size of a man's fist; there is also some slight swelling of left lobe. The tumour is somewhat elastic to touch, is freely movable, quite painless, rises and falls with deglutition and in no way hampers respiration. The increased vascular supply to the gland is conspicuous on account of the dilated thyroid veins. There is a well-marked bruit on auscultation. The recurrent laryngeal nerve is evidently implicated, resulting in alteration of voice, which is harsh and croaking.

The temperature is subnormal, 96.4° F.

The tongue is hypertrophied and indented, teeth mostly carious, palate high-arched. He has a good appetite, but the bowels are obstinately constipated and require the frequent administration of aperients.

The heart-sounds are faint, but a tricuspid regurgitant murmur is audible at the lower end of sternum.

The pulse is weak but regular, the rate being 48 per minute. Examination of the blood reveals a reduction in the quantity of hæmoglobin, this diminution being about 30 *per cent.*, otherwise the blood exhibits little or no change. The liability to catching cold is one of the characteristics of cretinism, and this man is no exception, as he is acutely

sensitive to any change in temperature and at present has an obstinate cough.

It may be mentioned in passing that these patients usually succumb to pulmonary complications.

The total quantity of urine passed during twenty-four hours averages sixty-five ounces; specific gravity, 1025, acid in reaction contains no sugar or albumen, but there is an increase in the amount of urea excreted. This increase may be in part accounted for by the fact that lack of perspiration is a constant feature in cretinism, and in this way the excretion of urea is augmented.

The skin generally is dry, harsh, and thickened; It has a waxy, lemon tinge. The sweat and sebaceous secretions are absent. The nails are fragile and striated.

One of the most striking points in the nervous system has already been mentioned, namely, the abnormal sensitiveness to cold, the patient having to be supplied with extra under-clothing to promote warmth and comfort. The muscular nutrition is unimpaired whilst the motor functions, beyond clumsiness in movement, call for no further comment. Both the superficial and deep reflexes are diminished. The pupils react normally to light and accommodation.

The intellectual state is marked by apathetic enfeeblement, and cerebral torpor reveals itself in sluggish mentation, defective memory, slow speech, and lethargic movements. He is quite indifferent to his surroundings, never speaks to his fellow patients, nor attempts to work or amuse himself. The simplest questions are beyond his intelligence; he has no idea of time or place. He is childishly pleased with the slightest attention given to him. Is slovenly and untidy in dress, but has been educated to cleanly habits. He is indolent, both mentally and physically, having no muscular energy, and is promptly fatigued with the least continuous effort. He sleeps rather heavily and is with difficulty roused.

The patient has been under treatment with thyroid extract, this being administered in tabloid form. Starting with a dose of six grains daily this has been gradually increased until at the present time he is taking twenty grains *per diem*.

There has been a loss in body-weight amounting to 5 lb. The temperature has risen from 96.4 to 98.4° F. There is an increase in the pulse-rate, which on admission was 48 and is at present 80. The percentage of hæmoglobin present in the blood has increased 5 *per cent.*, whilst the amount of urea excreted has been slightly augmented. The skin remains inactive and the patient still complains of feeling cold. He has an excellent appetite, and the action of the bowels is regular.

As yet there is no alteration in his mental state, this condition being in all probability beyond improvement. Although not reported as epileptic, a few days ago patient had a typical seizure. This was evidently not his first attack, as just previous to it he informed an attendant that "he was going to have a fit."

During the whole time patient has been undergoing thyroid treatment he has been kept continuously in bed.

Regarding the manner in which defective thyroidation affects the trophic apparatus and induces mucoid deposits, two general points of

view depending on opposite physiological hypotheses are maintained. Many believe that the normal thyroid elaborates some substance indispensable to the proper action of the nervous system. Others maintain that the thyroid eliminates certain harmful elements in the blood, and in confirmation of this statement attention must be directed to two main points.

As previously stated, since the administration of thyroid extract to this patient, there has been a slight increase in the percentage of hæmoglobin present in the blood together with an increase in the output of urea excreted. This fact seems to prove the theory that the thyroid gland normally excretes a substance which, while aiding the oxygen-carrying power of the hæmoglobin, at the same time assists in the removal of toxic agents from the blood.

The conditions of origin of sporadic cretinism are obscure. Some authorities attribute it to parental consanguinity, others to alcoholism and syphilitic disease, but in the case under notice there is a point of importance. It has been observed that when a woman has frequent pregnancies this is often followed by an interval of sterility of some years' duration, and as she approaches the menopause fertility is again established. A child born at this time is usually smaller and not so well developed as the other members of the family. I am acquainted with two families in which the youngest child in both exhibits well-marked achondroplasia, the parental stock being healthy. I have a similar history in this case; the patient, a seventh child, was born when his mother was forty-three years of age, ten years having elapsed since her last pregnancy.

It is said that the offspring of two goitrous parents is invariably a cretin who may or may not be goitrous. In the goitrous cretins the thyroid disease may appear at any period of life, and acts, then, exactly as does spontaneous myxœdema or operative myxœdema, to stunt growth and stop mental development. The distribution of endemic cretinism is identical with that of endemic goitrous disease, and is probably due to some obscure telluric cause. A distinction between endemic cretins and other myxœdemic patients is the goitrous enlargement. This may be only a difference of degree, because the cystic degeneration and interstitial hypertrophy of a goitrous enlargement of the thyroid is destructive in character and effect, and in consequence myxœdema and mental disturbances will be developed proportionately to the functional inactivity of the thyroid. When the thyroid is entirely

wanting as in non-goitrous congenital cretins, or destroyed as in the present case, the myxœdema is correspondingly intense and the mental degeneration proportionately developed. Sporadic or congenital cretinism appears to be of two distinct kinds. The first in which a child at birth is found to present the most marked cretinous changes, being also invariably born dead ; the second, in which the child is born apparently quite normal, but at a variable period, from a few weeks to a few years after birth, begins to show symptoms of commencing cretinism which soon reach a maximum. The skull is full behind, contracted and narrow in front ; often the fontanelles remain unclosed. The features are flabby and thickened, nose snubbed, lips thick, eyelids swollen and drooping, mouth open and the tongue hypertrophied. Dentition is delayed and the teeth decay early, the neck is short and lipomatous, the abdomen swollen, the limbs dwarfed and crooked, the skin infiltrated and inactive. The thyroid is atrophied or absent ; the mental condition is usually one of idiocy. It is in this type of case that thyroid treatment is most beneficial.

Though cretins are comparatively rare in asylum populations, in certain mountainous regions throughout the world the disease is endemic. While on this point it may be advantageously remembered that the cause is distinctly hereditary in these districts where it largely prevails, and the birth of cretinous children can be avoided by emigration from affected districts. It is perfectly conceivable that insanitary or climatic conditions, which appear to determine the incidence of goitre or cretinism in these cases, do actually bring about one or more of these affections in certain instances. Professor Lebour, who has conducted the strictest inquiry into the geological distribution of this disorder, states that the greatest proportion of cretins are to be found in carboniferous limestone districts. The rarity of cretinism in asylums is in no small measure to be accounted for by the fact that the majority die in infancy or childhood, few—if untreated—reaching manhood.

(1) A paper read at Spring Meeting of the Northern and Midland Division at Starthes Hall, April 30th, 1908.