

A case of sporadic cretinism with appearance of myxoedema / by Amand Routh.

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A CASE OF

15

SPORADIC CRETINISM,

WITH

APPEARANCE OF MYXŒDEMA.

BY

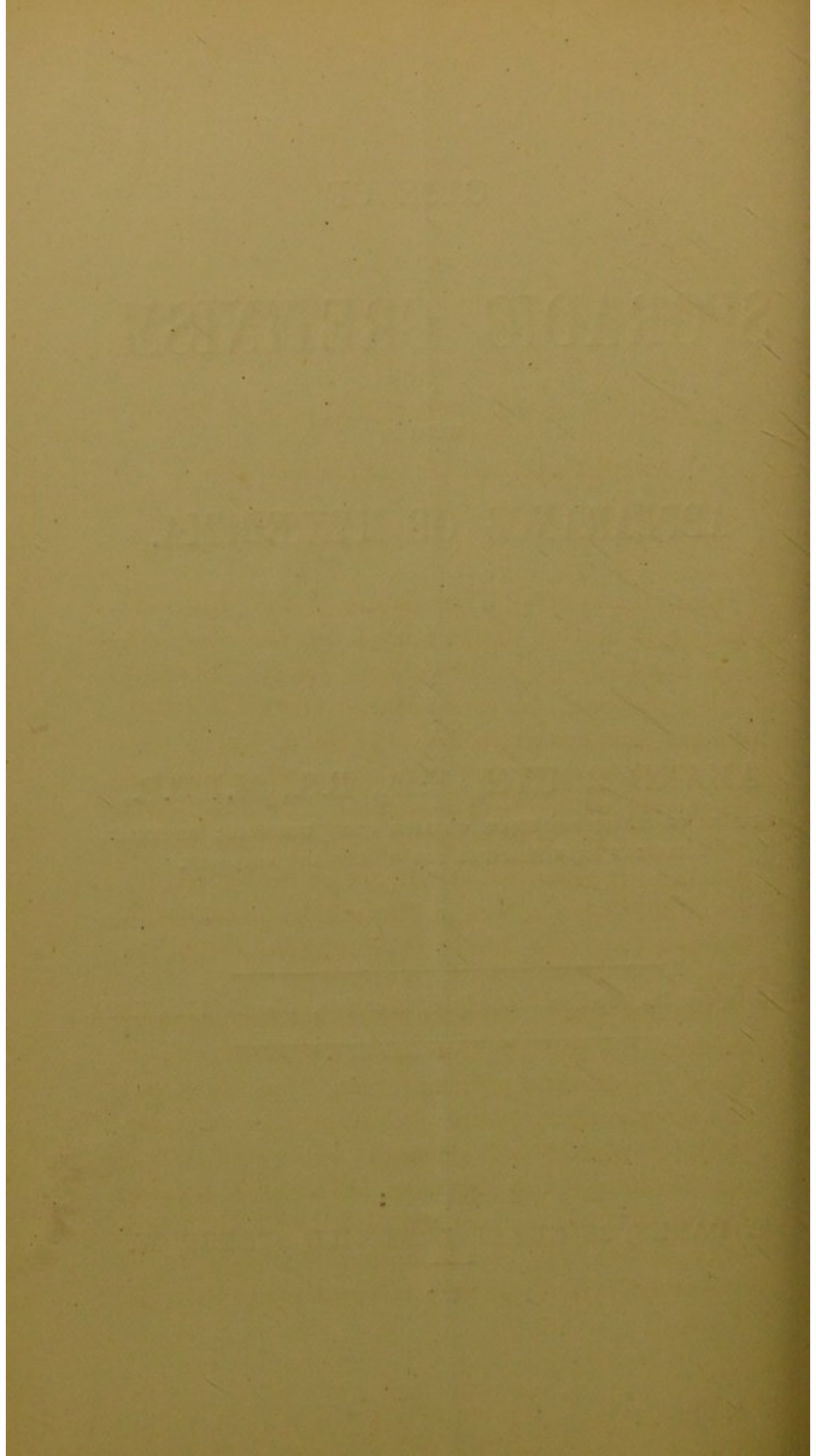
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A CASE OF SPORADIC CRETINISM WITH
APPEARANCE OF MYXEDEMA.

ELIZ. H., æt. 25 $\frac{3}{4}$, is an inmate of the Home for Female Incurables, Marylebone Road, and was born near Southampton, in Hampshire. She was the eldest of four children, both parents being quite healthy when she was born, being respectively 24 and 22 years of age. The father, however, died twelve years afterwards, and the mother sixteen years afterwards, both of consumption—a disease which also caused the death of the patient's own sister, æt. 17, and her mother's sister, æt. 27. The father was born in Hampshire, the mother in Berkshire, and there was no blood-relationship between them.

Up to the age of 7 years the patient was just like other children, but plumper than usual, and always looked like a "large white pudding," being "fat, flabby, and of a doughy-white colour." Whilst pregnant with this patient the mother was frightened by a bull. At 7 years of age the girl ceased growing taller, but continued growing stouter and flabbier, becoming slow in all her movements, and so stupid that all attempts at teaching her to read or write were fruitless. At 8 years of age the child

came to London, and beyond getting less able to help herself and more silly-looking, the friends say there has been little or no change, with the exception that latterly she has become thinner.

Family History.—Beyond the apparent heredity of consumption, there is no family history of any disease, such as syphilis or Bright's disease, and no history of alcoholism, neither is there any history of insanity or cretinism in the family. A sister of the patient, *æt.* 23, is a fairly healthy-looking girl, well-developed, and regular in all her functions, though somewhat anæmic. She has, however, an enlarged thyroid gland, now somewhat diminished under treatment, and there is a systolic murmur to be heard at the heart's apex and left axilla, dating from an attack of acute rheumatism three years ago. The patient also has a healthy brother, *æt.* 19, now in the army. All the children were born in Hampshire.

Present State.—Height, 3ft. 5in., the average height of a child of 5 years of age. Weight, 4st. 2lbs. 12ozs. She is much stunted in growth, and her bony skeleton is very considerably altered by rickety changes. Thus, there is a lateral curvature of the spine in the dorsal region, with the ribs thereby secondarily distorted. Lordosis is well marked. The tibiæ are curved. The epiphyses of the bones of the lower limbs are somewhat enlarged, and the ribs beaded at their junction with their cartilages. The head is very dolicho-cephalic—a condition of the skull which Dr. Gee has stated is often due to rickets. Some of these bony changes are not unlike those described by Dr. Barlow and Mr. Bowlby as occurring in foetal rickets (sporadic cretinism as it occurs in the foetus?), and it is possible that one reason

why they do not coincide more exactly is that in this case these changes may not have occurred till 7 years of age, when the sporadic cretinism first became apparent. The face is absolutely expressionless, and the lines obliterated. The features are swollen; the lips thickened, high-coloured, and shapeless; the mouth usually open; the nose is flattened and broadened between the eyes, and the nostrils dilated; the ears large and flat. The head does not hang forward, but the neck appears shortened, being lost in the supra-clavicular tumefaction.

The Skin is everywhere waxy-looking and pallid, with the exception of the face, where there is a slight flush, absent at times, over the flabby, pendulous cheeks, and on the feet and legs, where the circulation is so poor that a state of chronic chillblain exists. The skin is dry and rough in parts, but the palms of the hands are occasionally moist. The hands are spade-like in form. There is no true œdema.

The Hair is jet black, and fairly plentiful, but very harsh.

The temperature of the body averages 98.8° F. in rectum, but the surface temperature is always sub-normal, being usually between 95° and 96° in the axilla or groin. Once only was it as high as 97° , and that was after the patient had been covered up in bed for some hours.

The Thyroid Body cannot be felt. Above the clavicle on either side, and to a less extent immediately below that bone, are well-marked *tumefactions*, which slip away when grasped, and though somewhat resembling lipomatous or myxomatous tissue to the fingers, also remind one of the sensation conveyed by grasping a varicocele.

A clear respiratory murmur can be heard over the swellings, but no venous bruit. Similar subcutaneous swellings can be felt over the ribs in posterior axillary line.

The Abdomen is tumid and pendulous, owing partly to the shallow pelvis.

The Umbilicus is protruded, and the recti are non-adherent in the middle line.

The patient can walk slowly and totteringly for short distances, but often falls suddenly, so never walks without assistance being at hand. The legs seem unable to bear the weight of the body, and give way suddenly at the ankle or knees. She has free movement of her arms, but does everything very slowly and methodically, yet quite unhesitatingly. She does not dress herself, because she would be all the morning doing so, but can put on or off her clothes if allowed to take her own time.

The Speech is slow and monotonous, and pitched in a high key. She answers questions correctly enough, but her answers are delayed. She appears to take a longer time than usual to understand a question, and then to require a still longer time to frame the reply.

Nervous System.—Patient can neither read nor write, and every attempt to teach her anything has failed ; in fact, it is impossible to keep her attention concentrated. She cannot remember recent events, but can recall events which occurred before she left Hampshire at 7 years of age, the growth of both body and mind seeming to have been then arrested. She sleeps well and heavily. She is almost absolutely indifferent to pain, yet often complains of feeling cold. The “knee reflexes” are exaggerated ; and Dr. Angel Money drew my attention to Gower’s “front tap contraction,” which is also well

marked. Nothing appears to interest her for more than a few seconds, but when cheerily spoken to a momentary gleam of pleasure flits across her face. Otherwise, her expression is always motionless, callous, and somewhat sad.

The Eyesight is good, and ophthalmoscopic appearances normal. Pupils are somewhat dilated, and rather sluggish.

Urinary Organs.—Patient passes on an average 25 ozs. of urine in 24 hours. Sp. g., 1015. There is no sugar, albumen, or casts. The urea is deficient, being only about one-third of the normal quantity, viz., $10\frac{1}{2}$ grammes, or 172 grains per diem (percentage 1·3). This result was obtained by the method devised by Drs. Russell and West.

Digestion, Organs of.—The tongue is not notably enlarged, nor indented by the teeth; the faucial arch is broad; the teeth are badly formed, and several have been removed, being decayed. The alveolar processes are unusually prominent, and the mucous membrane of the gums much thickened. Patient's appetite is good; her breath is usually foul; her bowels constipated. She constantly complains of a nasty bitter taste in her mouth.

Organs of Circulation.—Pulse 72. It is very difficult to feel the pulse at the wrist owing to its distance from the surface, and its weakness. The heart-sounds are normal but weak.

The Blood.—The blood was very kindly examined for me by Dr. Montagu Murray by means of Dr. Gower's hæmocyto-meter and hæmodynamometer, as well as microscopically. Dr. Murray states that the proportion of white to red corpuscles is about 1 to 250; that the

percentage of red blood discs to the normal standard is 86 ; whilst the percentage of hæmoglobin is 77. There was no obvious change in the appearance of the blood corpuscles, red or white.

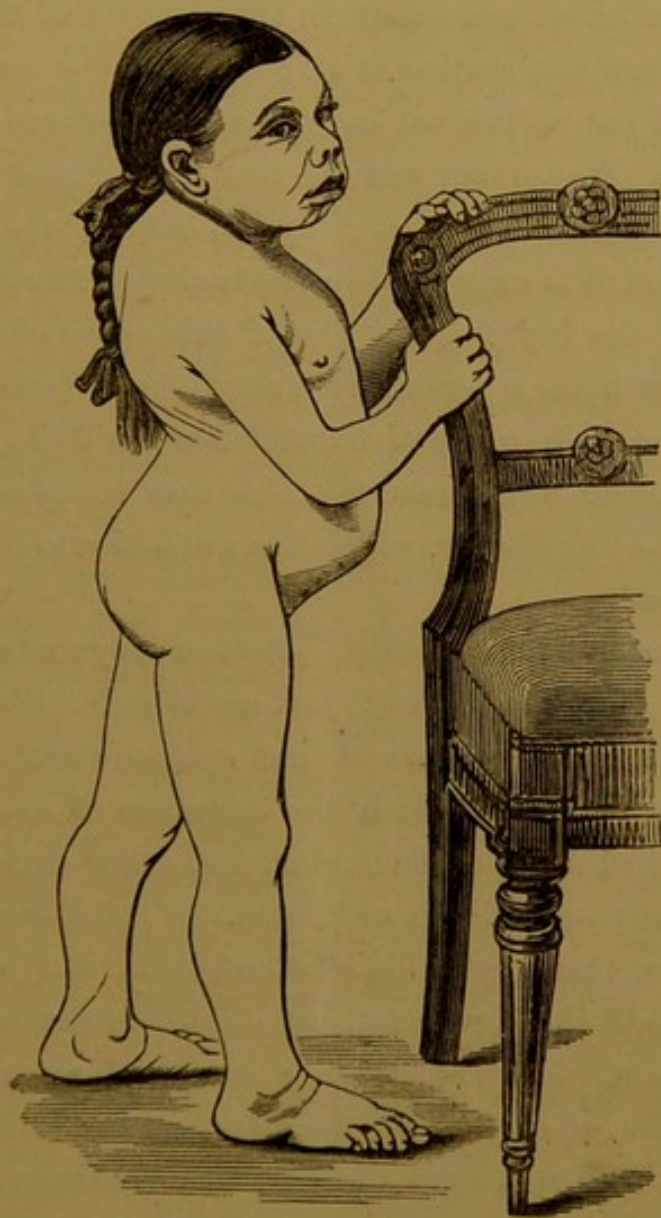
Organs of Respiration.—Quite normal in every respect.

Liver and Spleen.—Nothing abnormal is to be made out in these organs. They are not perceptibly enlarged.

Generative Organs.—Patient has never menstruated. There is no leucorrhœa. The external organs are quite undeveloped, and destitute of hair. The labia minora are elongated and thickened, protruding some distance from the vulva. The mammæ are quite infantile. The vagina and os uteri are normal. Per rectum, the pelvis is found to be infantile in shape and size, and the uterus is half its normal adult size, but normal in shape and position. The ovaries lie against the sides of the pelvis, lower and more posterior than usual, and are, at least, twice their normal bulk ; and are softer than usual, and cause no sense of pain when grasped between the finger in the rectum and the hand on the abdomen. It is probable that the ovaries have undergone a change analogous to myxœdema, which would account for the persistent amenorrhœa, and the infantile condition of both external and internal organs of generation.

Remarks.—I first saw this patient on account of her chilblains, and because the other inmates of the "Home for Incurables," where she is staying, complained of her bad breath. I believed the case to be sporadic cretinism, with myxœdema, and this diagnosis was kindly confirmed subsequently by Drs. Gowers, Ord, Barlow, and my father. Dr. Ord pointed out that the bony deformities appeared to be of rickety nature. The case agrees in most of its features with the description given by Sir

William Gull and the late Dr. Hilton Fagge, in their papers on sporadic cretinism—especially as possessing the subcutaneous symmetrical swellings, as first pointed out by Mr. Curling, and considered by many, and by Dr. Ord in particular, to be similar to myxœdema, a disease named



and described by that gentleman in 1878. In his paper read before the Medico Chir. Soc. in 1871, the late Dr. Hilton Fagge laid stress upon the fact that sporadic cre-

tinism was probably due to atrophy of the thyroid, and he further stated that he believed that a healthy, and still more an enlarged, thyroid was to be regarded as a safeguard against cretinism—in other words, that a healthy thyroid exercised a protective influence over the production of cretinism. He showed that in countries where cretinism was endemic, those who had the largest goîtres were not cretins at all; and that a large goître protected the individual from the more severe effects of cretinism. He further showed that in England the cause of cretinism, whatever it may be, only acts with a low power, which is expended in producing goïtrous enlargements of the thyroid, but that if this cause of cretinism should act upon a person with an undeveloped or absent thyroid no such protective influence could be afforded, and cretinism would result. This case lends some little support to this theory, which has not hitherto been much entertained.

The exact relationship between myxœdema and sporadic cretinism is not yet made out. Sir William Gull and Dr. Fagge showed that sporadic cretinism was invariably associated with the presence of symmetrical tumours in the supra-clavicular, and other regions; but their relative priority is unknown. In Dr. Hilton Fagge's four cases, those in which the cretinism had lasted the longest, had these symmetrical swellings the least well-marked, and *vice versa*. Hence it would seem that the myxœdema, if such it be, decreased as the cretinism increased. The case I am showing this evening also seems to indicate this, as the girl has latterly been getting thinner and losing weight. The cretinoid change does not appear to have commenced in this case till 7 years of age, up to which time the patient was much as other

children, and this is in accordance with Dr. Fagge's remark that sporadic cretinism may arise as late as the 8th year. It would, however, seem, from the aunt's description of the case, that in this patient the myxœdema had commenced much earlier; for she says the patient, as a baby, always looked "fat and flabby," and like a "doughy white pudding," a very fair popular description, I think, of a myxœdematous baby. This description is also in accord with that given before the Sardinian Commission on Cretinism, by Foederé, who states that those infants destined to become cretins have a body extraordinarily voluminous, and are mostly œdematous. There is reason, therefore, for believing that myxœdema precedes, or, at all events, may precede the development of sporadic cretinism. It seems now also clearly made out that non-development or atrophy of the thyroid gland precedes both myxœdema and sporadic cretinism. In his paper on myxœdema read before the Med. Chir. Soc. in 1878, Dr. Ord suggested that the mucous œdema might precede the atrophy of the thyroid, but the cases of Mr. Curling, the papers of Sir William Gull and Dr. Fagge, together with Kocher's remarkable results following complete extirpation of the thyroid gland, as narrated by Dr. Felix Semon, have led Dr. Ord lately to admit that evidence is now tending to show that atrophy of the thyroid gland precedes the myxœdema.

Resumé.—No date can be fixed in this case for the atrophy of the thyroid gland, but it seems likely that it is a case of intra-uterine non-development of the gland, tending first to myxœdema, and secondarily to cretinism, for we have a clear history that the mother, whilst pregnant with this child, was frightened and chased by a bull, and maternal fright during pregnancy was given by

Dr. Hilton Fagge as having occurred in one of his four cases. After birth, up to 7 years of age, the child's general appearance was that of a myxœdematous baby. At that age sporadic cretinism supervened ; whilst quite latterly the mucous œdema has been showing signs of subsidence. The girl continues to be under medical supervision at the above-named establishment.

