

HEREDITARY OCCURRENCE OF HYPOTHYROIDISM WITH DYSTROPHIES OF THE NAILS AND HAIR *

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The study of individuals and families that may show various manifestations of mental and nervous degeneracy not infrequently discloses the existence of constitutional disorders that are direct or indirect causes of these.

These constitutional disorders appear sometimes as peculiar defects or abnormalities of body structures that tend to be transmitted through the family. Numerous instances of these are recorded in the literature dealing with problems of heredity. These are in most instances treated rather more from the point of view of being specific unit qualities that are transmitted than as the results of a more general inherent deficiency of organs whose healthy functioning is essential for determining normal development and successful adaptation to the requirements of life.

Recently we have had the opportunity of studying a family that for generations had shown among its members peculiar trophic disorders of the nails and hair, and various abnormalities of a mental or nervous character.

While the more obvious abnormalities of this group are of special dermatologic interest, the fact that these seem to be fundamentally related to some constitutional disorder of an endocrinopathic type that peculiarly affects the nervous and mental make-up of the members of this family, makes the entire subject one of much importance for neuropsychiatry.

Our acquaintance with this family began with a boy who, at the age of 18, had been brought into the Juvenile Court of Detroit because of several instances of petty thieving. The frequency of his delinquencies and certain mental traits suggested the existence of feeble-mindedness and led to his admission for observation to the Psychopathic Hospital at Ann Arbor.

REPORT OF A CASE

Examination.—The physical appearance of the boy was quite abnormal (Figs. 1 and 2). He was about 5 feet, 5 inches tall, and weighed 138 pounds. He appeared unusually well nourished. The skin of the face, trunk and

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extremities was plump and felt as if it were thickened in its deeper parts. Around the eyes the skin was puffy as if from increased subcutaneous fat. The hands were chubby and the fingers were short and clubbed. The hands and feet were cyanotic. The surface of the skin lacked moisture. The skin of the finger tips and soles of the feet was exfoliated in places.

A very striking feature in his appearance was the scant amount of hair on the head and body and peculiar abnormalities of the nails. While the head seemed fairly well covered by hair, this on close inspection was seen to be unusually thin, and there were patches in which the scalp was covered only by a fine lanugo. The eyebrows were reduced to a few scattered hairs and these were stiff and short. There was but little hair in the axillary and pubic regions and the latter conformed to the feminine type of arrangement.

The nails of the fingers and toes all lacked a quarter or a half inch of reaching the tips of the fingers or toes. There was no lunula. Their free margin was thickened and broken. Their surfaces were smooth and not ridged. The nail bed was exposed and in places showed suppuration (Fig. 3).

The pulse rate was 80; the blood pressure 130. There were no cardiac abnormalities. The body temperature was always a little below normal. On several occasions it was 96.8 F.; usually it was under 98. The Wassermann reaction test was negative on the blood. The urine at admission had a specific gravity of 1.010 and showed a few hyaline and granular casts. In other respects it was negative. The roentgen-ray examination showed a normal sella turcica.

There were a number of neurological abnormalities. There was a frequent tic-like movement of the face, and when he became emotionally excited there was a tic of the right shoulder. His speech was somewhat thick and nasal in quality. Bone conduction for sound was absent in the left ear. The left pupil was larger than the right. The fundi of the eyes were normal. Refraction tests showed a high degree of hyperopia. The form field of the left eye showed a retraction of the field for white. There were no paralyses or disturbances of sensibility. The fingers showed a fine tremor on extension.

His mental reactions were slow and he showed a definite intellectual defect. Psychometric tests gave him a mental age of 12 years and 6 months. His attitude toward his delinquencies was immature and unreasonable. In other respects he showed few definite peculiarities.

The general appearance of the boy, the peculiar condition of the skin and the dystrophies of the hair and nails suggested the existence of a myxedematous condition. This view was confirmed by a biopsy of the skin kindly made by Professor Wile of the department of dermatology.

The patient was tested for glucose tolerance and it was found that he could take 250 gm. of glucose in twenty-four hours before sugar appeared in the urine.

Treatment.—He was put on thyroid extract in 5 grain capsules three times a day. At the end of twenty-seven days there was increased perspiration. The exfoliation of the skin of the hands and feet ceased. The hair was less fragile and there was a very noticeable increase in the hair of the scalp and eyebrows. The nails appeared to be improved and they seemed to show some growth. Subjectively he appreciated an improvement in his general feelings. After a month of thyroid treatment he began to show toxic effects; the pulse rate was 110. Treatment was discontinued. The improvement of the skin condition at once ceased. The dryness and exfoliation again returned. On resuming treatment, improvement again occurred.

The clinical symptoms and their response to treatment in this case leave little doubt but that the disorder of the nails and hair were but parts of a constitutional disturbance due to hypothyroidism.

Family History.—The case became of increased interest through the fact that these same deficiencies had been present among members of the boy's family for at least six generations, and that in addition to the dystrophies of the hair and nails, both those affected in this way and many others without these defects showed various disorders of nervous functions (Fig. 4).

It has been possible personally to substantiate the accuracy of the data in this case in almost every instance. Our information regarding the family goes back to the great-grandmother of our patient, I—1 M. J. C. 1st. This woman lived in Canada and was of French extraction. It is known definitely to members of the present generations that she had the same characteristic abnormalities of nails and hair. A photograph in the family shows a marked facial similarity to her daughter, II—1 M. J. C. 2nd. It is probable that these abnormalities were present in still earlier generations. This is suggested in the reply that M. J. C. 1st gave to her daughter that these were the signs of an unhealthy generation. Information that we regard as trustworthy, but that has not yet been personally confirmed, is that a brother of II-1 has the abnormality and of his six children, three have the disorder, and one is insane.

II-1 married a man who was free from defect. From this marriage there were ten children. The oldest of these, a male III-2 had the characteristic defect. He married a woman without the defect, and from this marriage there were six children. The oldest, a female, L, IV-1, has normal nails and fairly abundant hair of the scalp, but few in her eyebrows. She is of short stature and has peculiar short, chubby fingers. She is of neurotic constitution, and from early childhood has had a severe facial tic. She has had no children. The second child, a female, A, IV-2, does not have the defect. She is either hysterical or epileptic. When a small child she would have attacks in which she was unclear. In these she would take hold of her mother and remark that the floor was sinking under her. Later on these ceased, but at the age of 16 she had attacks in which she was unconscious for a brief period. There were no convulsive movements. These have recurred from time to time during her adult life. It is of interest to note that although she has always had an abundance of hair on her head, in the spring of 1919 she had a miscarriage, and following this her hair fell out in large amounts. She is married to a man without the defect and has had two children. The first of these died in infancy from pneumonia, and the second during the first years of her life suffered from marasmus. P, IV-4, a male, was our patient. His physical and mental disorders have been noted previously. It is said by his family that he and his father closely resembled each other, not alone in the defects of nails and hair but in having a thick skin and in their general physical make-up. Since his discharge from our observation, this boy has been delinquent frequently, having been implicated in several instances of stealing, and at the present time his whereabouts are unknown.

Between the third and fourth child there were two miscarriages. The fourth child, a boy, G., IV-5, is now aged 19. He has thin eyebrows and a fair amount of head hair. His nails are normal. During an attack of typhoid fever he had a severe delirium and since then he has never been physically strong.

The sixth child, a boy, E., IV-6, has the characteristic defect of nails and hair (Figs. 1 and 2). He was first seen at the age of 7. His appearance

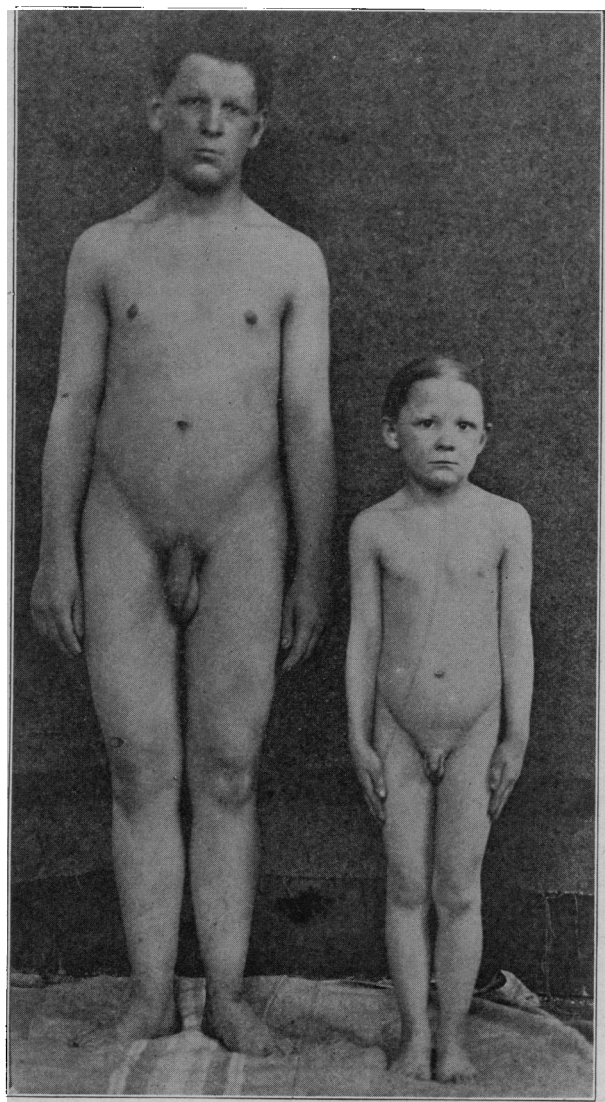


Fig. 1.—Patients IV-4 and IV-6.

at that time had a cretoid character. He looked like a little old man. His skin seemed myxedematous. There were patches of baldness on the top and back of the head and there were no eyebrows. Four years later his hair was uniformly scant over his head. There was a lateral scoliosis in the dorsal region. The thyroid gland was not palpable. There was a tic-like movement of the head and choreiform movements of the extremities. At the age of 13 he came into the courts because of malicious conduct and incorrigibility. He was sent to the State Training School for the Feebleminded. Psychometric examinations gave him a mental age of 9 years and 1 month. He responded very little to thyroid treatment.

The seventh child, R., IV-7, does not have the defect, but is mentally subnormal.

The second child of the third generation, W., III-3, a male, is said to have had hair at birth, but none after early childhood. At the present time, he is completely bald. He has no eyebrows or eyelashes and no body hair. His nails are defective in the characteristic way. Between the ages of 25 and 32 he had typical epileptic convulsions. At the present time he has peculiar feelings as if an attack were coming on, but no further development. He married a woman free from the defect and has had four children. The eldest, a girl, M., IV-8, has the defect. Her head hair is limited to a thin downy covering. Her nails are defective and she has thick skin and clubbed fingers. Mentally she is backward, and is now in a special room. Her thyroid is hard and abnormally small. Menstruation first appeared at the age of 16.

T., IV-9, a boy, aged 12, lacks the defect in the characteristic type, but his hair is unusually thin. He has nocturnal enuresis. Mentally he is feeble-minded.

H., IV-10, a boy, aged 10, does not have the defect, but is feeble-minded.

J., IV-11, a boy, aged 5, has thin hair, but normal nails. He has nocturnal enuresis.

The third child of the third generation, J., III-5, lacks the defect. He has had a bad criminal record in the courts. He has twice married, but has had no children.

The fourth child, E., III-7, seems to have been quite normal as to hair and nails. She married a normal man and all of her descendants, for two generations, have been free from the defect.

The fifth and sixth children of the third generation were twins and lacked the defect. One of these, III-9, died in infancy. The other, E., III-10, married a normal man and had two normal children. She, herself, died from acute tuberculosis.

The seventh child, L., III-2, had normal hair and nails. She married, and has had eleven children. All of these have remained free from the defect.

The eighth child, J., III-14, has no hair on her head and her nails are defective. She married a normal man, III-15, who is also a brother of III-4. She has had seven children. The first of these, T., IV-30, was free from the defect, but died in childhood from scarlet fever. The second and third children lacked the defect, but died in childhood from marasmus. The fourth child, W., IV-33, died in infancy from meningitis.

The fifth child, M., IV-34, a girl, aged 13, has normal nails and hair, but her teeth are notched along the cutting edge. Mentally she is feeble-minded.

The sixth child, C., IV-35, has the typical defect. He has nocturnal enuresis and is feeble-minded.

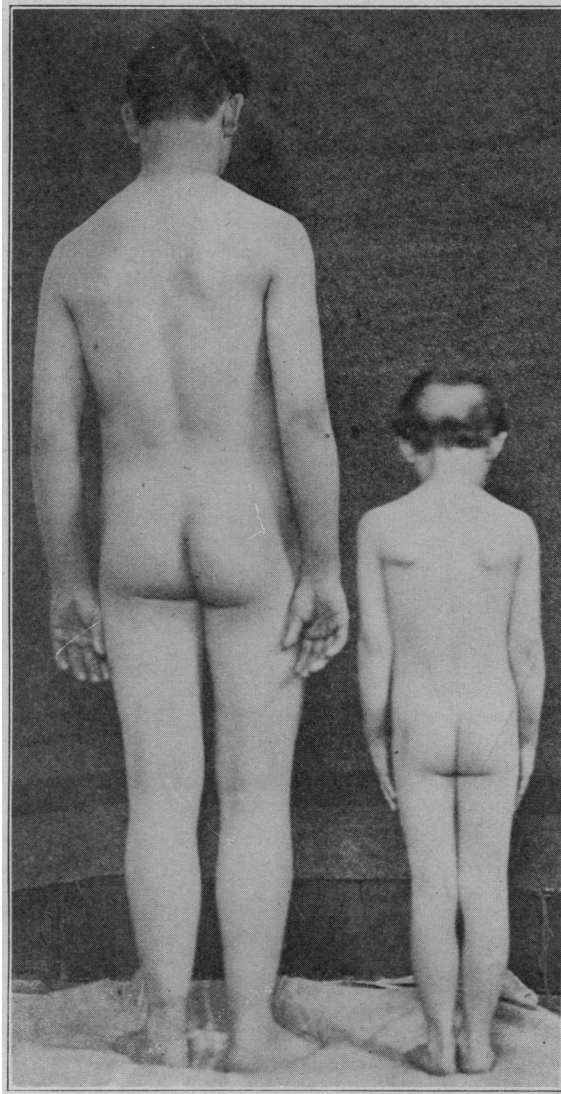


Fig. 2.—Patients IV-4 and IV-6.

The seventh child, M., IV-36, aged 7, has the family abnormality of nails and hair. She has nocturnal enuresis and is feeble-minded.

The ninth child of the third generation, M., III-16, lacks the defect. She married a normal man and has had nine children, all of whom are free from the defect and are of average mentality.

The tenth child, L., III-18, has the defect. Her teeth are worn away on the cutting edge. She married a normal man, and has had three children. The oldest of these, A., IV-46, has always had good hair, but his nails show the typical defect. He has some difficulty in hearing, but otherwise is healthy. The second child died in infancy and had abnormal nails. The third child, R., IV-48, has the defect. While the head seems fairly well covered by hair it is abnormally thin in amount. The teeth show irregularities of the cutting edge. Mentally he is subnormal.

Among sixty-one members of this family, belonging to six generations, the defect in its typical manifestations was present in fourteen instances. In these, both hair and nails were affected. The nail defect seemed to be about the same in all, but there was much variation in the

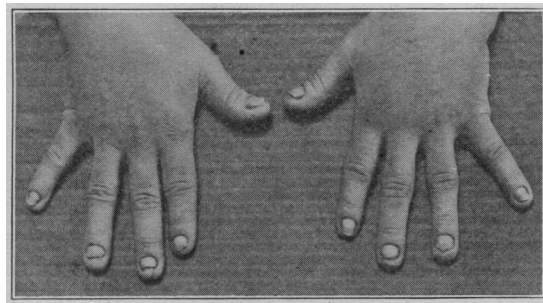


Fig. 3 (Patient IV-4).—Abnormalities of nails.

degrees of loss of head hair. In the greater number there was an extreme scantiness as to the amount of hair. In no instance was there a total loss of all hair of the head, the most extreme cases showing a fine lanugo-like covering of the scalp.

The defect tends to occur in a mendelian type of distribution, but the varied character of the abnormalities appearing in relation with the type defect of nails and hair is perhaps too complicated to be explained in a simple mendelian formula. The character of abnormal hair and defective nails behaves as a mendelian dominant. All persons that have the defect give a mixed progeny when crossed with normals. The defect never appears in a descendant who himself is free from the defect.

Aside from this particular abnormality, there are other features that show that in this family group there are conditions active in the production of a variety of disorders that are of much interest to neuropsychiatry. This is the high frequency of feeble-mindedness and

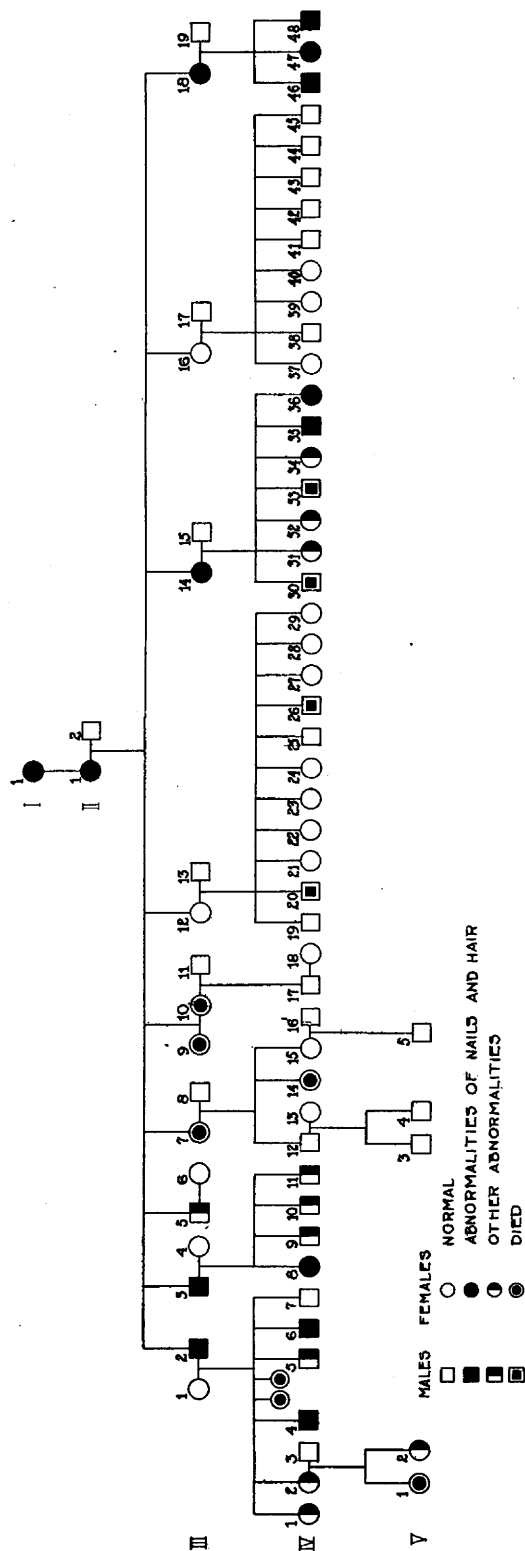


Fig. 4.

Heredity chart of family showing hypothyroidism and abnormalities of nails and hair.

neurological disorders of a degenerate type that are present among the family.

The members of the third generation who had dystrophies of nails and hair, and all of their descendants, numbered twenty-nine persons. Of these, twenty-two were definitely abnormal. Twelve of the latter had the characteristic family dystrophy, and ten others, who lacked this, showed other constitutional and nervous disorders. These included one case of epilepsy, one of hysteria, one of severe tic, four instances of feeble-mindedness, one of nocturnal enuresis and four died at an early age from marasmus. Even those who had the nail and hair dystrophy had other abnormalities. One of these was an epileptic; one had cancer; four were feeble-minded, one had nocturnal enuresis.

This multiplicity of characteristics of degeneracy bears out the view that we are concerned in this family with a more fundamental disorder than that of an isolated abnormality of nails and hair. It would seem that there was present some organic defect that had widely distributed constitutional influences.

The well-known association of abnormalities of hair and of nails in hypothyroidism and the two fairly well defined cases of juvenile myxedema, and the reaction of one case to thyroid feeding, seems to warrant the conclusion that the fundamental disorder in this family was of the thyroid gland.

Cases Reported in the Literature.—There are in the literature a few other observations on families showing similar dystrophies of nails and hair. The earliest one of these and the most extensive in its abnormalities was that reported by Nicolle and Hallipré.¹ In this family of fifty-five known persons, thirty-six showed the dystrophies. In 1896 White² reported the study of a family in which there were fourteen members in four generations. Of these, seven showed the characteristic abnormalities. Eisenstaedt,³ in 1913, reported observations on a family of thirteen members which showed the occurrence of the dystrophies in five generations. Among eight members of the fourth generation, the abnormality was present in three. It is also probable that a contribution by Hoffman⁴ should be included in this connection. This reports a line of thirteen persons, eight of whom through four generations had shown abnormally short and scant eyebrows and dystrophies of the nails of the hands and feet. In this family there were several instances of thyroid disease. The patient had a large struma, as had her mother. The appearance of the mother

1. Nicollé and Hallipré: *Maladie familiale caractérisée par des alterations des cheveux et des ongles*, *Ann. de Derm. et Syph.*, 3d Ser. **6**:804, 1895.

2. White, Charles J.: *Dystrophia Unguium et Pilorum*, *Hereditaria*, *J. Cut. and Gen. Urin. Dis.* **14**:220, 1896.

3. Eisenstaedt, J. S.: *Three Cases of Family Dystrophies of Hair and Nails*, *J. A. M. A.* **60**:27 (Jan. 4) 1913.

4. Hoffman: *Ueber Verkümmern der Augenbrauen und der Nägel bei Thyroidosen*, *Arch. Derm. u. Syph.* **89**:381, 1908.

suggested hypothyroidism and Hoffman comments that "perhaps one might believe that a dysthyroidism was the predisposing cause for the described malformations."

The frequency of disorders of the nails or of the hair in hypothyroidism is shown in several analyses of large groups of cases of myxedema. In an analysis of 150 cases, Hun and Prudden⁵ found malformations of nails in 75 per cent. of cases studied, and in a later series Howard⁶ found nails abnormal in 86 per cent. of cases, and hair in 93 per cent. of those studied.

As to the heredity of hypothyroid disorders, there is a good deal of confirmatory evidence. In the previously cited analyses of large groups of cases, Hun and Prudden found a direct inheritance in 8 per cent. of cases studied, and Howard in 6.6 per cent.

It would be of interest to know to what extent some of the so-called neurotic disorders of the hair are brought about through the reaction of the thyroid gland to disorder of the sympathetic nervous system coming primarily from affective disturbances such as the sudden loss of hair or change in color following fright and nervous stress. There may be persons whose thyroid functioning is just sufficient to maintain normal health, and under stress this balance may be so disturbed as to produce definite pathologic conditions. In this group we have reported there were two persons who lacked the characteristic family dystrophies, who, following slight constitutional disturbances suffered a loss of hair and later regained normal conditions.

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5. Hun and Prudden: Myxedema, *Am. J. M. Sc.* **96**: July and August, 1888.

6. Howard, C. P.: Myxedema: A Study, *J. A. M. A.* **48**:1226 (April 13) 1907; *ibid.* **48**:1403 (April 27) 1907.