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We hope that making available the relevant information on Pachyonychia Congenita will be a means of furthering research to find effective therapies and a cure for PC.
THREE CASES OF HEREDITARY HYPERKERATOSIS OF THE NAIL-BED.

By A. Garrick Wilson.

The first case I saw was an infant (Mrs. G—), for an affecting reason. On further inquiries I found that her mother (Mrs. G—) had the same condition, and that her two brothers had presented the condition from infancy. The nails grew opaque and thick, and the surface of the nails was rough, and towards the sides when examined up from its bed by a dark, pointed instrument, the free edge. The nail grew from under the skin, and the nail bed was not visible. The hair and skin were quiet, and there were no associated symptoms. The case was congenital or acquired, and not due to any other skin diseases. Examination of the bed after maceration in lye showed the condition of the spores.

The nails cause no pain in their normal condition. When knocked, the tissue around the nail and the nail bed is tender, but a gradual growth of horn raises the anterior part of the nail above the bed. The affection has been not only present throughout the whole of life, but is a condition of the nail bed.

The family history is as follows: Mrs. G— had a child, but she had thirteen children, only one now living. Of these children, the tenth and thirteenth were the only one now living. Of these children, the tenth and thirteenth were affected with the same condition. The nails grew opaque and thick, and the surface of the nails was rough. The nail grew from under the skin, and the nail bed was not visible. The hair and skin were quiet, and there were no associated symptoms. The case was congenital or acquired, and not due to any other skin diseases. Examination of the bed after maceration in lye showed the condition of the spores.

* Read before the Sheffield Medical Society.
THREE CASES OF HEREDITARY HYPERKERATOSIS OF 
THE NAIL-BED.*


The first case I saw was an infant, which was brought to me by its mother (Mrs. G—), for an affection of the nails, and whilst examining them I noticed that its mother's finger-nails were similarly affected. On further inquiries I found that the mother's toe-nails were in the same condition, and that her mother (Mrs. T—), one sister, and two brothers had presented the same peculiarity in the nails of both hands and feet.

The surface of the nails is smooth, and at the base normal in appearance, but towards the free extremity the nail becomes raised up from its bed by a dark, friable, horny mass which projects under the free edge. The nail grows much faster than the horny tissue underneath.

The hair and skin were quite normal in every case, as also were the teeth, and there were no associated congenital defects, such as sometimes occur in these cases. There was no evidence of syphilis, either congenital or acquired, and no signs of past or present ichthyosis or other skin diseases. Examination of the horny tissue from the nail-bed after maceration in liquor potassae showed the absence of any spores.

The nails cause no pain in themselves, but are very inconvenient. When knocked, the tissue around the nail readily becomes inflamed, and the nail comes off. The new nail at first appears to be normal, but a gradual growth of horny tissue takes place in the nail-bed and raises the anterior part of the nail from its bed.

The affection has been noticed in all the cases at birth and persists throughout the whole of life.

The family history is as follows: The grandmother was an only child, but she had thirteen children, the mother of the infant being the only one now living. Of these thirteen the first two had deformed nails, and the tenth and thirteenth; the first and second were boys and the tenth and thirteenth girls; the mother of our infant is the tenth.

* Read before the Sheffield Medico-Chirurgical Society, November 14th, 1904.
The infant, whose photo I reproduce, was a year old at the time and had a brother, a year and a half older, whose nails were quite normal. Since I first saw the family another male child has been born whose nails are exactly like those in the photograph.

The eldest and the youngest child are strong and healthy, but the subject of our photograph has always been weakly and anemic.

I have been unable to find any record of similar cases, but I believe that cases of hereditary affections of the nails are not very uncommon. I am unable to offer any suggestion as to the causation, and am inclined to put it down as simply an hereditary manifestation.

These cases are always interesting, but I regret that most of the interest lies in the rarity of the case and not in the possibility of treatment.

I have only been able to obtain photographs of the infant, its mother and grandmother; all the other affected members of the family are dead.

Mrs. T— (grandmother), affected.

1. Male, affected (dead).
2. Male, affected (dead).

10. Female, (Mrs. G—), affected (dead).
13. Female, affected (dead).

Male, aged 3 years, unaffected.
Male, aged 2 years, affected.
Male, aged 10 months, affected.

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