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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.
The nomenclature relating to structures at the distal part of the nail is confusing. A distal yellow line traversing the nail, described by Pinkus, has been relatively ignored in the literature and remains unnamed. Clinical and histological studies presented in this article show that this band is present in more than 90% of normal adult fingernails and represents the most proximal point of attachment of the fingertip stratum corneum to the nail plate. Therefore, it should be referred to as the onychocorneal band or junction. This region has distinctive histological features and is the first major barrier to material passing proximally beneath the nail plate. It is possible that abnormalities of this structure may result in onycholysis, pachyonychia congenita, and pterygium inversum unguis.

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MORPHOLOGICAL descriptions of the distal region of the human nail are often incomplete, and the nomenclature is vague and confusing. Hyponychium is a common term that has been used variably to describe anything from the entire epidermis beneath the nail plate to a small area of epidermis in the region where the nail plate separates from the nail bed.

Sole-horn (sohlenhorn) is a term applied to an area of epidermis distally beneath the nail plate that is thought to bring horny substance to the nail but not to form an actual nail. This is not always present, particularly on the fingers. It has been suggested that this may be the only vestige of the deep stratum of an animal claw normally present in humans. Achten considered the terms to be synonymous, for he states, "The hyponychium (Sohlenhorn or terminal zone), the beginning of the nail bed and the finger tip consist of an epidermis which is comparable to the nail bed, with of course a thickened horny layer."

In 1955, Terry introduced the term onychodermal band to represent "a barely perceptible pale narrow band ... transversely across the distal portion of the nail bed, immediately proximal to the free edge of the nail." He also noted that it had a slightly amber tinge and a faintly translucent quality. However, Achten and Parent describe the onychodermal band as pink and corresponding to the site where the nail separates from the nail bed.

One distinctive morphological feature that has been largely ignored in the literature is the white band that runs transversely across the distal part of the nail. Pinkus referred to this in 1927 as the "zweite helle linie des Nagelbetts" (second light line of the nail bed), and it was mentioned by Terry as being present at the junction of his onychodermal band and the main pink zone.

The aim of our investigations was to review the structures and their relationships at the distal part of the human nail and, in particular, to determine the nature and significance of the transverse white band (Fig 1).

PATIENTS AND METHODS

The nails of the following groups of subjects were studied.

Cadavers

The morphological features of the finger and thumb nails of 10 cadavers were documented. The time of death before examination was between 1 day and 2 weeks (mean, 6 days).

Amputated Digits

Histological information was obtained by performing longitudinal nail biopsies on seven fingers from different subjects, that had been amputated after traffic accidents. The nails themselves had not been damaged by the accidents. Before biopsy, a nick was made on the surface of the nail plate overlying the position of the white band. Specimens were processed by a conventional alcohol-paraffin technique and stained with hematoxylin and eosin.

Normal Subjects

The fingernails and thumbnails of 100 normal subjects aged 16 to 65 years (mean age, 37 years) were examined for their morphological features, and 10 subjects were examined by transillumination from below. In 20 subjects, attempts were made to insert sharp and blunt probes proximally beneath the nail plate to assess the mechanical barrier properties of the distal nail structures.

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Fig 1. Diagrammatic representation of the morphological features of the normal nail.

Fig 5. (A) Seven-month fetal digits in longitudinal section. A thin layer of rudimentary nail plate covers the nail bed. (B) Just distal to the end of the growing nail, a compact mass of cells represents the hyponychium, or the future onychocorneal attachment zone.
Embryological Tissue

Sections were prepared from the hand of a 7-month fetus and stained with hematoxylin and eosin. Longitudinal sections were examined to obtain evidence of prenatal structures corresponding to the site of the transverse white band and/or solihorn.

RESULTS

Cadavers

A distal, thin (0.5 mm) transverse white band could be readily identified in at least seven of the fingernails and thumbnails of all cadavers.

Histology

All biopsy results showed the same basic features; the nick in the nail corresponded to the most proximal point of attachment of the digital stratum corneum to the nail plate (Fig 2). In this region there was, in every case, a distinct zone where the stratum corneum was paler staining and less compactly layered than in the adjacent fingertip stratum corneum. At the actual area of attachment of the stratum corneum to nail plate, the stratum corneum was more compact and darker staining, although ghosts of keratinocytes orientated in a distal direction could be seen. Similar cellular ghosts were orientated horizontally within the adjacent nail plate. In two of the sections, a small group of cells with well-preserved nuclei could be seen at the junction of the nail and stratum corneum.

The granular layer was particularly prominent where the stratum corneum joined the nail plate and also extended proximal to the junction.

In all specimens, the stratum corneum distal to the junction had become separated from the overlying nail plate, leaving only small remnants of stratum corneum still attached to the plate. However, in the two instances in which the proximal junction had been breached, the break was through the stratum corneum, leaving a column of stratum corneum still attached to the nail plate (Fig 3).

Normal Subjects

One thousand fingernails and thumbnails were examined in 100 subjects. A transverse distal white band could be seen in 909 (90.9%) (Fig 4). The width of the band varied from 0.1 mm to 1 mm. At its proximal border, there was a zone of increased redness compared with the remainder of the main pink zone extending proximally to the lunula. Immediately distal to the white band was a narrow pink zone (width, 0.5 mm to 2 mm) that culminated distally as a narrow (0.1 mm to 0.5 mm), rather translucent amber tinged band.

On transillumination, the white band did not transmit light as well as the pink zones on either side of it and therefore appeared as a dark line.

A sharp probe inserted proximally between the nail plate and the finger passed steadily and painlessly until the white band was reached. At this point, resistance occurred and pain was experienced. Using a blunt probe it was relatively easy to peel away the tissues in the region of the distal pink zone from the nail plate until the white band was reached. Resistance and pain then limited further separation.

Fetal Nail

In the 7-month fetus, a rudimentary nail plate was clearly visible covering the nail bed (Fig 5A and B) to the end of the nail plate, and a pad of dark staining cells is seen distally.

DISCUSSION

This investigation has shown that a thin, distal, transverse white band is visible in more than 90%
of normal adult fingernails and thumbnails. It has been suggested that this band is a manifestation of the local vasculature and that it is a vital process present only in life. However, its presence in the majority of the nails of the cadavers shows that it is not dependant on a dynamic circulation. Its relative opacity upon transillumination indicates a solid structure rather than a vascular phenomenon.

Histological studies showed that the white band corresponds to the most proximal point of attachment of the fingertip stratum corneum to the nail plate. Therefore, it is appropriate to refer to this band as the onychocorneal band or onychocorneal junction.

Although the stratum corneum is loosely adherent to the nail plate at its most distal point, it can be easily peeled away from the nail plate by using a probe. However, it is only at the site of the white band that substantial resistance to objects passing proximally beneath the nail plate is met and pain is first experienced. This band, therefore represents the most distal point of firm attachment of nail to nail bed and is the major barrier to exogenous agents' passing proximally beneath the nail plate.

The efficiency of this junction is well illustrated in the two sections showing cleavage of nail plate from nail bed occurring through the stratum corneum rather than through the attachment zone itself (Fig 3).

The difference in the pattern of keratinization and staining properties of the stratum corneum in this area compared with adjacent fingertip stratum corneum suggests a modification of keratin production in this region. However, the significance of this finding and of the groups of cells observed within the stratum corneum in two specimens is
uncertain. One possibility may be that modification of the stratum corneum is necessary in this region for it to adhere efficiently to the nail plate as well as to serve its normal barrier functions.

It is essential that this seal remain intact while the nail plate continues to grow distally. Therefore, it was interesting to find ghosts of cells oriented distally beneath the junction and horizontally within the adjacent nail plate. Therefore, the junction is a site of cellular activity, with the attached stratum corneum moving distally with the nail but then shearing off, with continual replacement occurring proximally.

We believe the structure of the nail apparatus at this point to be a dynamic and essential feature maintaining the integrity of the nail attachment. The pad of darkly staining cells seen at the tip of the 7-month fetal specimen may represent the sohlenhorn or hyponychium. We do not regard it as a vestigial remnant of the animal claw, although its ontogenetic relationship may be accepted. It may develop into the important attachment zone seen in cadaver specimens and in normal adults as the onychocorneal band.

An abnormality of the onychocorneal junction may be responsible for some observed conditions. Defective adhesion of the stratum corneum to the nail plate may permit distal separation of nail plate from nail bed and allow material to pass proximally beneath the nail plate to give the clinical picture of onycholysis. The parakeratotic stratum corneum of psoriasis is generally considered to be less adherent than normal orthokeratosis, and the consequent reduction in adhesion at the onychocorneal junction may result in psoriatic onycholysis. Similarly, infection of the stratum corneum at this point by dermatophytes may allow onycholysis to develop, and interruption of the junction may be an important pathogenetic factor in idiopathic onycholysis.

A primary defect in the mechanism by which the stratum corneum remains adherent at this junction could result in pterygium inversum unguis, a condition in which the distal part of the nail bed remains adherent to the ventral surface of the nail plate through a hyperkeratotic layer. Caputo and Prandi also suggest that this condition results from an abnormality at the point at which nail plate separates from the nail bed and that in their patient this "sole horn was more extensive than normal, which indicates the possibility of an acquired reversion, without any apparent cause, towards a more primitive condition."

Hyperplasia in this region would result in substantial subungual hyperkeratosis with lifting of the nail plate. A primary hyperplastic abnormality of the junction may be responsible for conditions such as pachyonychia congenita.

We suggest that the nomenclature relating to the distal nail apparatus could be simplified by discarding the term onychodermal band. This is an inaccurate description because the nail plate does not directly come into contact with the dermis. The term hyponychium should be retained to describe the area of epithelium extending proximally beneath the nail plate to where the granular layer ceases. The term onychocorneal band or junction should be added because this appears to be a distinct and important clinical and functional structure.

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REFERENCES