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PART 12 MISCELLANEOUS CONDITIONS

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Nail anatomy and physiology

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The nail is an important skin appendage. It is useful, not only for the aesthetic appearance, but also for its role as a diagnostic clue to various cutaneous and systemic disorders. The nail also protects the underlying rich neurovascular supply, which is used for thermoregulation and sensory purposes. Knowledge of nail development, anatomy, and physiology helps us in better understanding of nail disorders and it also guides us in therapeutic interventions and innovations.

ANATOMY OF NAIL UNIT

Embryology and development

The fingernail primordium develops during the 8th embryonic week from the epidermis slightly earlier than the initiation of hair follicle development, as a transverse ridge on the distal dorsal surface of the digit. Development of toenails starts 4 weeks later in similar pattern. The nail fold is delineated by a continuous groove. A group of cells from the proximal part of the nail fold then grows and extends downwards and proximally into the dermis of the digit, stopping approximately 1 mm from the phalanx and giving rise to the matrix primordium. This site will in turn form the epithelium of the proximal nail fold, the distal and intermediate matrix epithelium. The presumptive nail matrix cells that are present on the ventral side of the proximal invagination differentiate and keratinize to become the nail plate. The distal part of the nail fold forms the distal ridge, which is a visible group of cells, on the dorsum of the distal tip of each digit (Figure 2.1).[1]

At 11 weeks, the dorsal nail bed surface begins to keratinize. At 13 weeks' gestation, the proximal nail fold is formed and the first signs of nail plate growth are observed from the lunula. Moreover, nail fold epithelium starts to keratinize with formation of the stratum granulosum, beginning distally and advancing towards the proximal nail fold. The nail matrix is completely developed by 15 weeks and it starts to produce the nail plate, which will continue to grow until death.[2,3]

The granular layer recedes at 18 weeks' gestation and the nail bed epithelium takes on a postnatal appearance. The process of cellular differentiation and maturation within the matrix is similar to that seen in adult nails at 20 weeks' gestation. By 32 weeks' gestation, the nail plate reaches the tip of the fingers and all the components of the nail are recognizable. Nail plate in toenails reaches the tip later at 36 weeks only and absent nail plates at the tips of digits are another indicator of prematurity.[2,3]

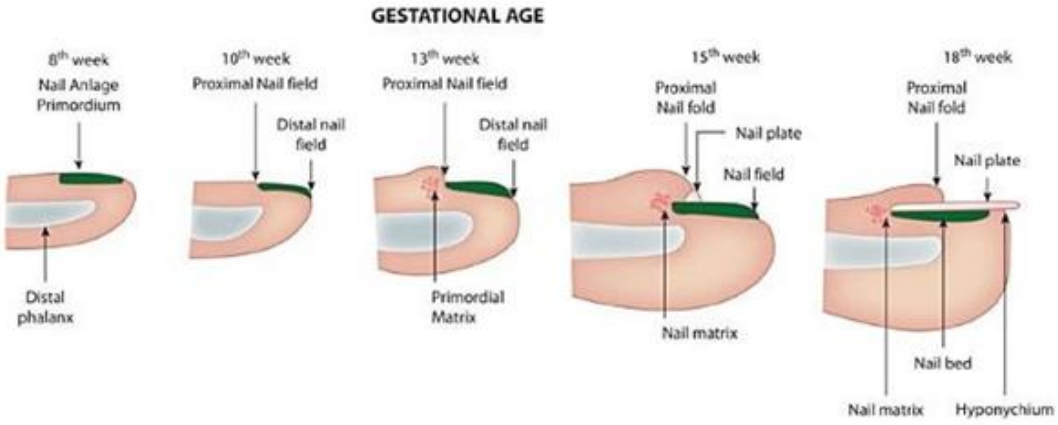

Figure 2.1 Embryologic development of the nail unit.

Transcription factor R-spondin 4 initiates nail development, and its mutation leads to congenital anonychia. Functional *p63* is required for the formation and maintenance of the apical ectodermal ridge, which is an embryonic signaling center required for limb outgrowth and hand plate formation. So, mutations in *p63* affect nail development in syndromes like ankyloblepharon, ectodermal dysplasia, and cleft lip/palate syndrome as well as ectrodactyly, ectodermal dysplasia, and cleft lip/palate syndrome. Wnt7a is also important for dorsal limb patterning and, hence, nail formation. Primary signaling abnormalities in Wnt7a are also associated with inherited nail dysplasias such as Schöpf–Schulz–Passarge syndrome (*Wnt10a*). *LMX1b* and *MSX1* are important for nail differentiation. *LMX1b is* mutated in nail-patella syndrome and *MSX1* in Witkop syndrome. However, in contrast to follicular development, the Shh gene is not required for nail plate formation. In murine models, *Hoxc13 is* also an important homeodomain-containing gene for both follicular and nail development.[2–4]

ANATOMY

The nail unit lies immediately above the distal phalanx and the ligaments, and tendons and ligaments around the distal interphalangeal joints not only help anchoring the nail apparatus to underlying bone, but also are essential for nails' mechanical functions. Moreover, fibers from extensor tendons attach to proximal nail fold, thus further strengthening the attachment of nail unit to bone. Thus, the entire nail apparatus is kept in place over underlying distal phalanx, the size and shape of which greatly determine those of the nail plate.[1,2] Close proximity of nail apparatus to periosteum and relative lack of dermis and subcutaneous fat necessitate strict asepsis and careful handling of tissues during the procedures on nail.

The nail unit consists of a nail plate, four specialized epithelia (the proximal nail fold, the nail matrix, the nail bed, and the hyponychium), and lateral nail folds (Figures 2.2 and 2.3).[1]

Nail plate

The nail plate is a hard, semi-transparent, slightly convex "dead" keratinized structure. It is comprised of tightly packed onychocytes that contain abundant hard hair-type keratins embedded in a matrix of sulfur-rich high-cysteine and high-glycine/tyrosine proteins. The nail plate, analogous to stratum corneum of skin, arises from the lower surface of the proximal nail fold. It is surrounded and partially covered by the proximal and lateral nail folds. Nail plate is a translucent structure but appears pink due to underlying blood vessels except for the free distal margin that appears white. Lunula, a white semi-circular area, may be present in the proximal nail plate. This corresponds to the visible portions of the distal matrix and loose attachment of proximal nail plate to the underlying epithelium. Lunula is most easily visible in thumbnails and great toenails. The natural shape of the free margin of the nail is the same as the contour of the distal border of the lunula. The hyponychium is the gap beneath the free edge of the nail plate distally. Onychocorneal band, a thin distal transverse white band, present in more than 90% of fingernails, represents the last point of firm attachment of the nail plate to the nail bed. Another zone, called onychodermal band, lies just distal to onychocorneal band and is seen as a 1.0–1.5-mm pink (Caucasian) or brown (Afro-Caribbean) colored band.[1,4]

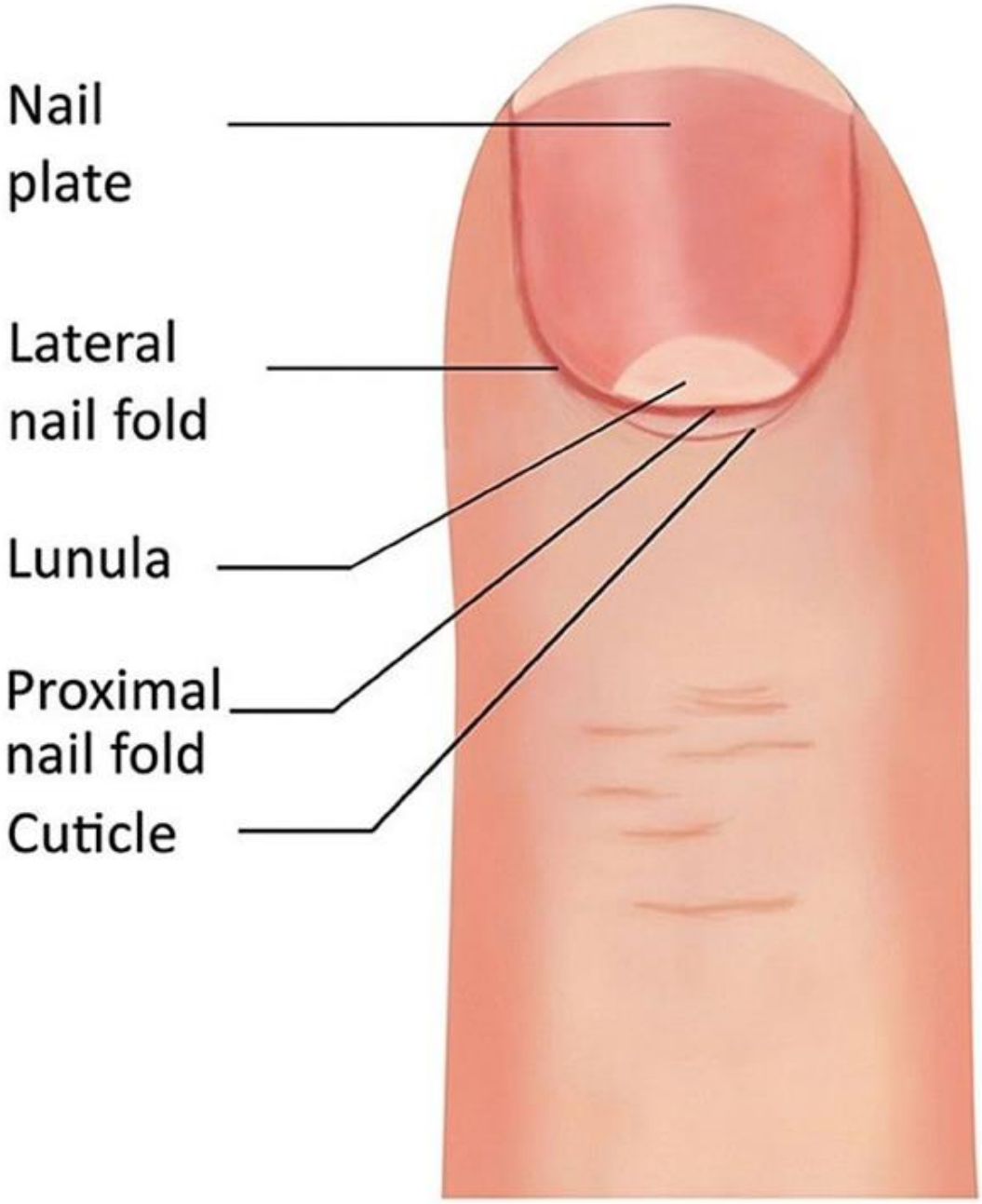

Figure 2.2 The nail unit (surface view). (Courtesy of Dr. Sunil Kothiwala.)

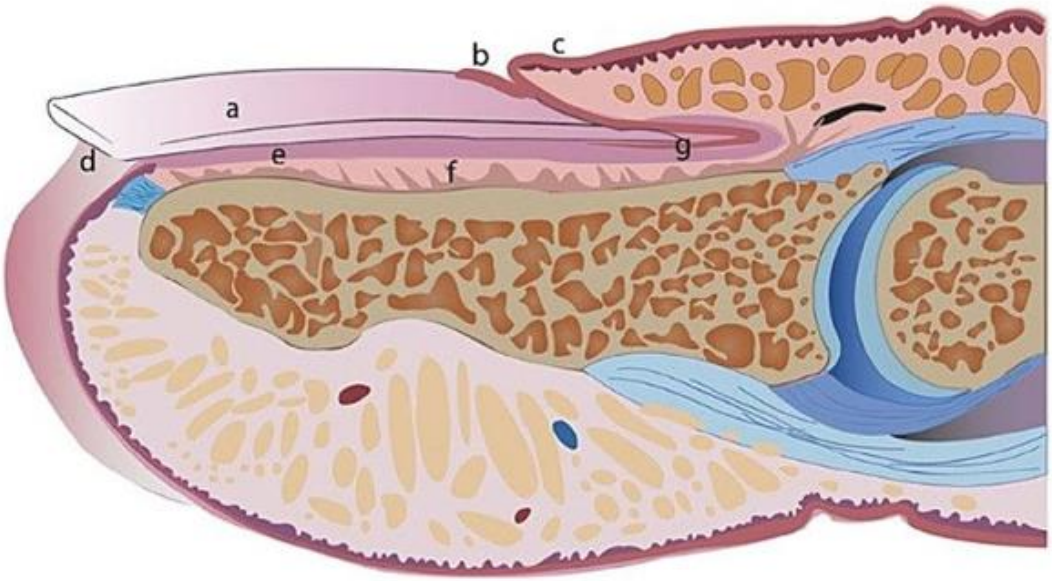

Figure 2.3 The nail unit (longitudinal section) – **(a)** nail plate, **(b)** cuticle, **(c)** proximal nail fold, **(d)** hyponychium, **(e)** nail bed, **(f)** terminal phalanx bone, **(g)** nail matrix. (Courtesy of Dr. Sunil Kothiwala.)

The nail plate is curved in both the longitudinal and transverse axes. This curvature allows nail plates to be embedded in nail folds, providing stronger attachment. The surface of nail plate is not completely smooth, but shows fine longitudinal ridges that correspond to complementary ridges on the underlying nail bed to which it is attached. The pattern of ridges in childhood is different; short partial oblique ridges are noted in herringbone or chevron pattern. The longitudinal ridges appear as children grow and increase with advancing age. These ridges are considered to be specific to the individual, allowing the distinction between identical twins and, hence, may be used for forensic identification.

Sagittal section of the nail plate shows three portions: (1) dorsal nail plate, derived from keratinization of cells from proximal nail matrix; (2) intermediate nail plate, contributed by distal nail matrix; and (3) ventral nail plate, contributed by nail bed (Figure 2.4). Hence, proximal nail matrix pathology manifests itself in alterations in dorsal nail plate, which are both palpable and visible. On the other hand, distal nail matrix pathology results in alterations in intermediate nail plate, which are visible but not palpable. Understanding this concept is crucial in choosing site of nail biopsy in different conditions.