Diseases of the Nails and their Management

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AND

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Preface

Since the earliest publication by Heller there have been several books written on diseases of the nail: in particular the works of Alkiewicz & Pfister, Pardo-Castello & Pardo, Samman, Sertoli and Zaïas must be mentioned as they are of high quality and extremely useful, mainly to dermatologists.

For many years we have felt that there is a need for a comprehensive reference book on all aspects of the nail in health and disease. It is evident that in different cultures nail abnormalities are often seen by a variety of specialists, e.g. traumatic and genetic dystrophies are rarely seen initially by dermatologists whilst cosmetic and industrial problems may be handled by dermatologists, industrial health experts, cosmetologists or chiropodists. These are a few examples of the need for a reference book to 'cross' speciality, and even more important, parochial, national medical barriers. We believe that a satisfactory book on the nail must do this. The world is small! We have both travelled widely in recent years and do hope that the content and style of the book succeeds in this aim.

Some people may be surprised to find a Frenchman and Englishman apparently having agreed with each other for long enough to produce a book of this nature—not all French and English are enemies! We have worked diligently to benefit from our language differences and to combine the differences in training and interests and hope that this first truly international book, including authors from France, Germany, Sweden, UK and USA, will be of use world wide.

Though the chapters have been contributed by specific authors, we must point out that the book is very much a group activity; in particular, the editors have contributed much from their own files to every section. This applies to the script, references and figures, and therefore any errors of fact, emphasis or quality of picture may be the fault of the editors rather than the named chapter writers!

The inclusion of colour pictures has obviously made the book more expensive than with black and white pictures alone. We gave considerable thought to this and decided to include them as important diagnostic aids because of the photogenic nature of the nail; and the fact that between the ten authors we had a unique opportunity to pool material collected over many years.

Robert Baran
Rodney Dawber

Acknowledgements

An undertaking of this kind is quite impossible without the help of a vast number of colleagues the world over who have encouraged, cajoled and constructively disagreed with us over the many years that we have been interested in nails; and more specifically we must thank those who have provided details and pictures of their patients—these are acknowledged in the script.

We are deeply indebted to Georges Achten, Peter Samman and Nardo Zaïas who at various times have stimulated our interest in this field; without their help in our careers this book would not have materialised in any shape or form.

We are very grateful to Dr Gerald Godfrey and Chris Gummer who gave great assistance in formulating the final text.

Robert Baran
Rodney Dawber

Contents

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Chapter 1
Structure, Embryology, Comparative Anatomy and Physiology of the Nail

R.P.R. DAWBER & R. BARAN

The major part of the nail apparatus develops *in utero* from the primitive epidermis (Zaïas, 1963); in consequence it has many similarities both in health and disease to the hair and stratum corneum. As early as the second century BC Galen remarked on the similarity of hair and nails. In generalised integumentary diseases such as psoriasis, the nail apparatus, the hair follicle and the epidermis may all be structurally and functionally affected, presumably because of their common tissue of origin.

The primary function of the nail apparatus is to produce a strong, relatively inflexible, keratinous nail plate over the dorsal surface of the end of each digit. The nail plate affords a protective covering for the fingertip; by exerting counter pressure over the volar skin and pulp, the flat nail plate adds to the precision and delicacy of touch, the ability to pick up small objects and many other subtle finger functions. Fingernails typically cover approximately one seventh of the dorsal surface, whilst on the great toe the nail may cover up to 50% of the dorsum of the digit.

STRUCTURE

Gross anatomy (Lewis, 1954; Lewin, 1965)

The component parts of the nail apparatus can be seen in Figs 1.1a & b and Fig. 1.2. The rectangular nail plate is the largest structure, resting on and being firmly attached to the nail bed; the attachment is less firm

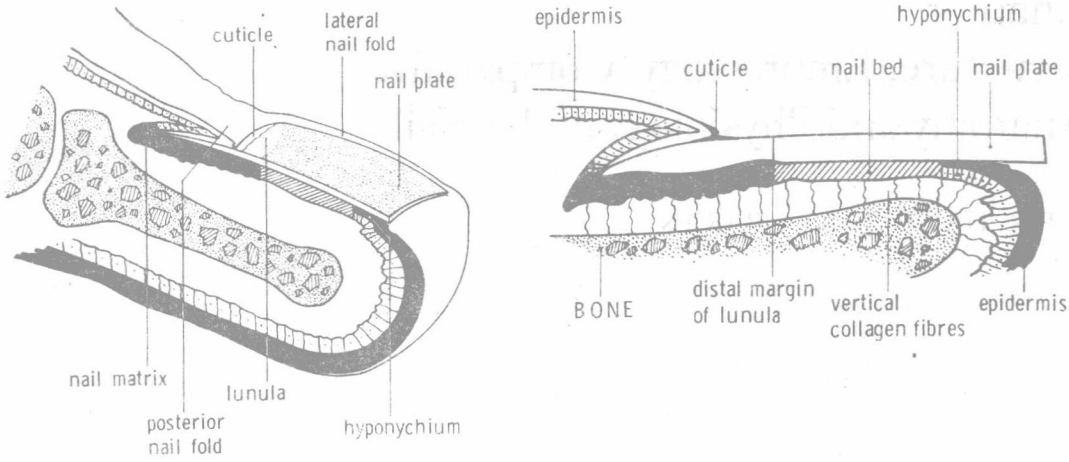

Fig. 1.1. a & b. Longitudinal section of a digit showing the dorsal nail apparatus.

proximally, apart from at the posterolateral corners. Approximately one fourth of the nail is covered by the proximal nail fold whilst a small margin of the sides of the nail plate is often occluded by the lateral nail folds. Underlying the proximal part of the nail is the white lunula (Burrows, 1917). The nail plate distal to the lunula is usually pink in colour because it is translucent, allowing the redness of the vascular nail bed to be seen through it. The free nail plate is white to grey in colour due to the intrinsic colour of the nail. In longitudinal section (Fig. 1.2) the proximal nail fold is seen to be in continuity with the skin over the dorsum of the terminal phalanx. The epidermis of this fold continues around the base of the nail as the germinative matrix; this can be clearly differentiated from the nail fold structurally, but functionally, as can be seen in the section on nail dynamics (p. 000), the boundary is less distinct. The proximal nail fold

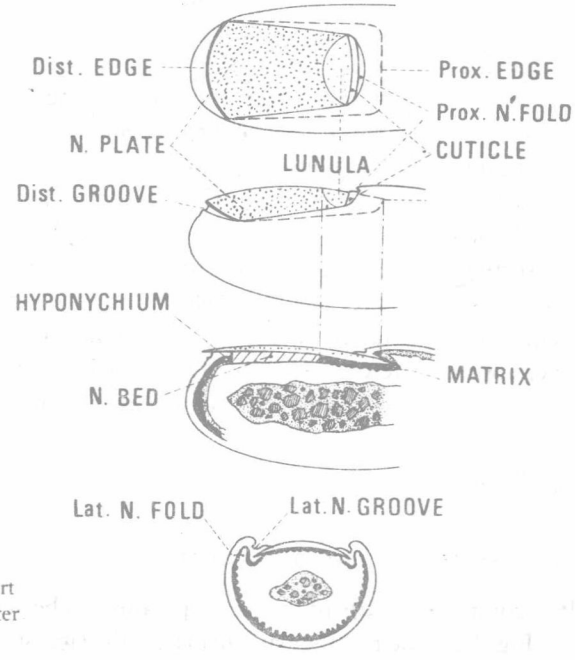

Fig. 1.2. Component part of the nail apparatus (after R. Iso).

has two epithelial surfaces, dorsal and ventral; at the junction of the two the cuticle projects distally onto the nail surface. Many authorities propose the term eponychium for various parts of the dorsal area of the nail base and posterior nail fold (Achten, 1968; Samman, 1978; Zaïas, 1980). Since there is no uniformity of opinion regarding the use of the term, and all the component parts of the nail apparatus can be defined without it, it is not used in this text; also there is no definite embryological, comparative anatomical or physiological basis for it cf. the hyponychium. The lateral nail folds (Fig. 1.2) are in continuity with the skin on the sides of the digit laterally and medially they are joined by the nail bed. Some authorities term the lateral nail fold and adjacent finger tissue lateral to the nail fold, the nail wall.

The nail matrix can be subdivided into a dorsal section, an intermediate section which underlies the proximal nail plate to the distal border of the lunula; some authorities prefer the terms proximal and distal matrix respectively: also a so-called ventral section contributed from the nail bed. At the point of separation of the nail plate and the nail bed, the sub-ungal epidermis may be modified as the solehorn (p. 7) or sohlenhorn (Pinkus, 1927). In humans, this structure may be only vestigeal in health, its original significance only being evident from comparative anatomical studies (p. 14); however, in certain diseases, it could be the seat of distal sub-ungual hyperkeratosis or parakeratosis e.g. in pachyonychia congenita (p. 40) and pityriasis rubra pilaris (p. 174). Beyond the solehorn region which is probably the proximal part of the hyponychium, the hyponychium terminates at the distal nail groove; the tip of each digit beyond this ridge assumes the structure of the epidermis elsewhere.

Microscopic anatomy: (Horstmann, 1957; Lewin, 1965; Achten, 1968)

The nail folds

The external epidermal surface of the proximal nail fold is essentially similar in structure to the adjacent normal skin, though it is devoid of dermatoglyphic markings and hair follicles; the same applies to the lateral nail folds. The proximal (or posterior) and lateral nail folds keratinise via keratohyalin formation, possessing a granular layer which is absent in all parts of the nail matrix. The lateral nail folds are only loosely apposed to the nail plate apart from their proximal and medial parts which connect to the intermediate and ventral matrices respectively. From the distal area of the proximal nail fold, the cuticle reflects onto the surface of the nail plate to which it is firmly attached. The cuticle typically consists of modified stratum corneum. It serves to protect the structures of the nail base, particularly the germinative matrix. Loss of the cuticle often allows acute and chronic inflammatory and infective processes to involve the nail matrix, leading to secondary nail plate dystrophies such as chronic paronychia (p. 136) and pri irritant contact dermatitis (p. 274).

Nail matrix (intermediate and dorsal)

The nail matrix is the germinative part of the nail apparatus. As in the epidermis of the skin, the matrix possesses basal cells which undergo cell division; many of these cells, like keratinocytes in the skin, undergo differentiation as they move cut of the basal layer and finally harden, keratinise, die, and become incorporated into the nail plate; the nail matrices produce this change without the formation of a keratohyalin granular layer.

As with light microscopy, much of the cytological structure seen with transmission electron microscopy (Hashimoto, 1971a, b & c) is similar to the skin epidermis. The basal cells possess desmosomes and hemi-desmosomes and interdigitate freely. Differentiating cells are rich in ribosomes and polysomes and contain more RNA than equivalent epidermal cells. As cell differentiation proceeds towards the nail plate, there is a progressive build up of cytoplasmic microfibrils (~75 Å). These fibrils are haphazardly arranged within the cells up to the transitional zone; subsequently, the fibrils become vertically orientated in relation to the axis of growth of the hardened nail. Membrane coating granules (Odland bodies) are formed within differentiating cells; these discharge their contents into the intercellular space in the transitional zone; research has at various times suggested that these organelles may contribute to the thickening of the plasma membranes, and in desquamation and separation of squamous cells, or secrete material which aids in the firm adhesion of cells to each other to form the relatively impermeable membrane of the stratum corneum. In view of the firm adherence of nail plate cells to each other throughout the length of the nail, one can speculate that membrane coating granules contribute to this extracellular adhesiveness. Mitochondria are degraded during the transitional phase whilst RNA-containing ribosomes are evident up to the stage of plasma membrane thickening. Vacuoles containing lipids and other products of cytolysis are seen in cells in the transitional stage; at this point in differentiation, dorsal matrix cells demonstrate shrinkage of nuclei with a decrease in DNA. This breakdown continues until the exposed nail plate nuclear remnants can no longer be detected cf. the intermediate matrix in which prominent nuclei are evident up to the time of cell death.

The intermediate matrix continues forward from the nail root under the nail and is visible from the surface as the lunula (syns: lunule; 'half-moon'). Even though this sub-ungual part of the intermediate matrix always exists, it may not be visible as a lunula. Various anatomical factors have been suggested as reasons for the white colour: (Burrows, 1917; 1919; Ham & Leeson, 1961; Achten, 1963; Lewin, 1965; Baran & Gioanni, 1969):

1 The matrix epithelial cells are rich in nuclei in the lunula region, appearing parakeratotic.

2 The surface of the nail is smoother and more shiny than the distal part.

3 The nail plate is thinner at the lunula and the underlying epidermis is

thicker and thus the underlying capillary network cannot be seen from the surface.

4 The attachment of the nail and nail bed at the lunula is less firm, giving more reflection of light at the nail plate- nail bed interface.

5 The nail is more opaque in the white area.

6 The underlying dermis has less capillaries within it.

7 The underlying dermal connective tissue is of looser texture.

Most of these factors are hypothetical. When the nail is removed from the nail bed, both the nail plate and nail bed retain the pallor. The lunula would appear to be of no functional significance; in many individuals, a lunula is only seen on the thumbs and in others on the hands and not the feet. In such individuals nail structure appears to be the same with or without a lunula.

The overall shape of the dorsal and intermediate matrix in relation to surrounding structures is not the same in all digits, this is very important with regard to possible surgical procedures. For example, in toenails the germinative matrix is curved almost in a semi-circle around the terminal phalanx; the lateral limits lie as far round the sides as its coronal plane. Also the matrix is separated from the dorsum of the phalanx by connective tissue but the lateral edges and proximal corners lie very close to the periosteum. It is these less accessible parts which may get left behind by the surgeon and cause regrowth of horns of nail (Austin, 1970).

The nail matrix contains melanocytes in the lowest two cell layers (Higashi, 1968; Higashi & Tadeo, 1969). Thus the nail plate contains melanin granules and pigmentation from melanocytic donation of mature melanosomes to differentiating matrix cells. This is often barely evident in caucasoids. Pigment therefore arrives in the nail plate as in the keratinised cells of the stratum corneum and hair cortex. (Jimbow *et al.* 1971; Zelickson & Mottaz, 1974). Nail pigmentation is most evident in negroid individuals when it is commonly seen as longitudinal linear streaks; such linear pigment may occur in many pathological states (p. 71). Whether hormonal factors control melanocyte production and donation of pigment in the normal nail matrix is not known. In the epidermis, each melanocyte has a relationship with a determined pool of adjacent keratinocytes to which it donates pigment, usually via dendritic processes (Hadley & Quevado, 1966). At present there is no evidence to show whether a similar state pertains in the nail matrix.

Nail bed (and ventral matrix)

This consists of an epidermal part (ventral matrix) and the underlying dermis closely apposed to the periostium of the distal phalanx. There is no subcutaneous fat in the nail bed.

The nail bed epidermis is relatively thin, consisting of no more than two or three cell layers. In this region, the transitional zone in which living cells become keratinised and incorporated into the ventral nail plate is very abrupt. In this respect it is similar to the Henle layer of the internal root sheath of the hair follicle (Roth, 1967; Achten, 1968). It has been

noted (Achten, 1968) that the nail apparatus is similar in many respects to half a hair follicle cut longitudinally and laid on its side (Fig. 1.3) the hair bulb being analogous to the nail root matrix and the definitive hair cortex the equivalent of the upper nail plate. As the cells differentiate, vertical movement is limited by the horizontal and distal movement of the intermediate (and dorsal) nail plate. The keratining cells of the nail bed thus assume the same direction of movement as they near, or form the deepest (ventral) layer of the nail plate. These cells do not move distally in the absence of the nail plate Zaias, 1967). Keratinisation occurs in the nail bed without keratohyalin formation and it has been suggested that pressure changes may effect keratinisation, as seen in rat tail epidermis.

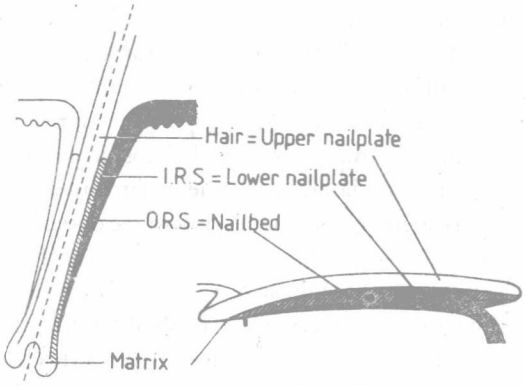

Fig. 1.3. Comparison of the nail apparatus and hair follicle (Achten, 1968): I.R.S. = internal root sheath, O.R.S. = outer root sheath.

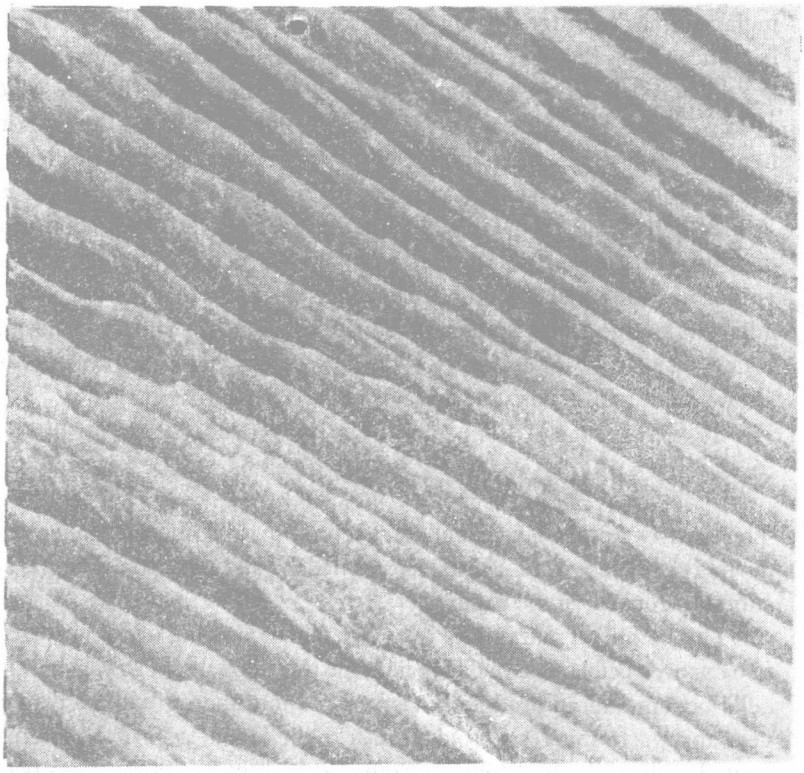

Fig. 1.4. Nail bed, showing the regular longitudinal folds.

The dermo/epidermal junction of the nail bed is thrown into regular longitudinal folds (Fig. 1.4) which give firm adhesion of the nail bed to the nail plate. The capillary network of the dermis is therefore orientated in relation to these folds. This is the reason for splinter haemorrhages being longitudinal (p. 362). The major connective tissue network radiates from the phalangeal periosteum vertically to connect with the nail bed epidermis (Fig. 1.1b) binding the two structures firmly together. A less prominent network of collagen fibres, orientated horizontally, is also present. Within the connective tissue networks lie blood vessels, lymphatics, a fine network of elastic fibres and a small amount of fat; there are no pilo-sebacceous units.

Bound phospholipids are present throughout the nailbed epidermis. Acid phosphatase and non-specific esterase are absent cf. dorsal and intermediate zones; bound cysteine can be detected in the transitional zone (Jarrett & Spearman, 1966).

In some nails, at the distal border of the nail bed the epidermis thickens (proximal hyponychium) and the dermo-epidermal junction becomes papillary; in keratinising, this narrow band, the solehorn (Pinkus, 1927), may contribute soft keratin to the nail plate ventrally.

Nail plate

The nail plate is made up of three layers; a dorsal layer which is thin, the thickest intermediate layer, and the ventral layer from the nail bed. The shape of the terminal phalanx plays an important role in determining the shape of the nail plate. Indeed, the nail is always changed if the phalanx is abnormal. Microscopically it consists of closely-knit horny, squamous cells arranged in lamellae; acidophilic masses are occasionally seen in older age groups, the pertinax bodies of Lewis and Montgomery (1955) which result from the drawing up of the epidermis into the substance of the nail plate.

The fully hardened nail is made up of flattened cells (Fig. 1.5) with closely apposed tortuous and interlocking plasma membranes (Hashimoto, 1971a & b; Breathnach, 1971); some in folded parts of the

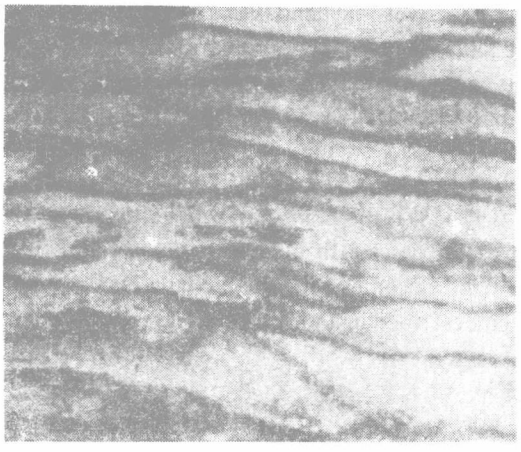

Fig. 1.5. Flattened nail plate cells with prominent plasma membranes.

membranes may appear to be within the cytoplasm. At high magnification the contents of each cell show a uniform fine granularity untrastructurally similar to the staining pattern seen in the hair cuticle.

Onychodermal band (Fig. 1.6)

When the attached nail plate is viewed from above, its colour can typically be divided into two clearly defined zones, the proximal lunule and the larger pink zone. On close examination two further distal zones can often be identified, the distal yellowish-white margin, and immediately proximal to this, the onychodermal band (Terry, 1955). This band forms a barely perceptible pale, narrow band running transversely across the

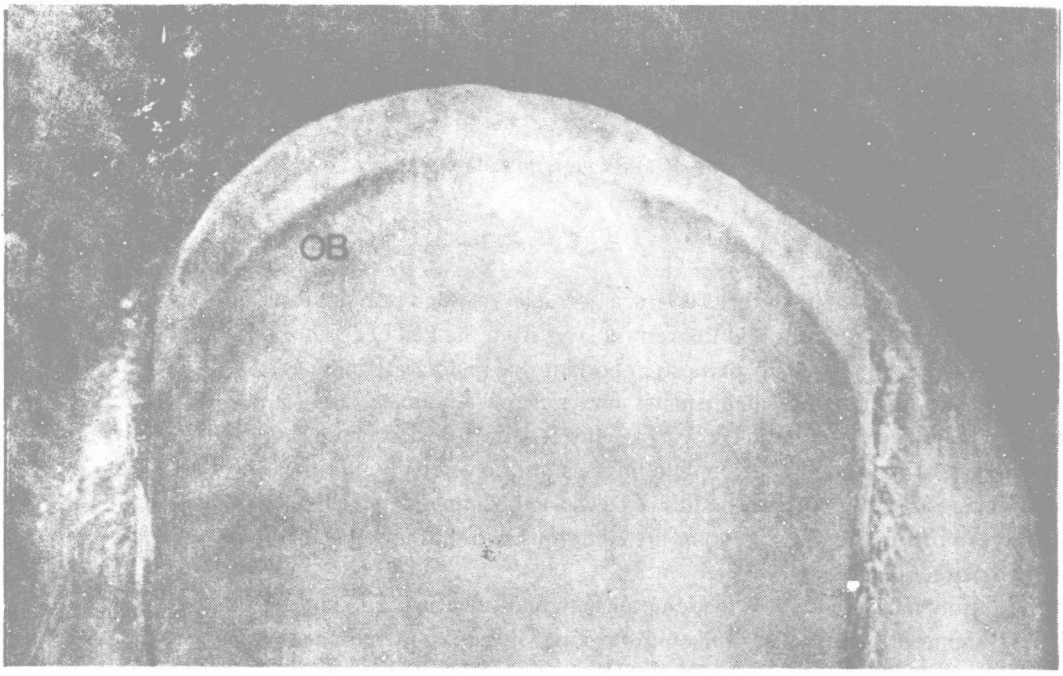

Fig. 1.6. Terminal part of the nail showing the onychodermal band (OB).

distal portion of the nail. On fingernails the band is usually no more than 0.5–1.5 mm wide; on some nails it is narrower and may even be absent, e.g. on thumb nails. It is more obvious in acrocyanosis. In some individuals the pale colour may appear yellow, whilst brown pigmentation may be present in negroes. The exact anatomical basis of the onychodermal band is not known, but it appears to have a different blood supply than the main body of the nail bed; if the point of the finger is pressed firmly, the band and an area proximal to it blanch. If the pressure is repeated several times the band becomes reddened. Many changes of colour of this band have been described in a variety of diseases (p. 76).

Biochemistry and histochemistry (Jarrett and Spearman, 1966; Achten, 1968; Vellar, 1970)

Keratin

It has been shown histochemically that cystine, containing disulphide bonds is concentrated mainly in the intermediate plate at the periphery of individual cells; the dorsal plate contains the lowest concentration. The reverse applies with regard to bound sulphydryl groups, the dorsal plate containing the highest concentration. Total sulphur concentration is similar in the dorsal and intermediate plates. Nail keratin analysis has revealed essentially the same fractions present in nail as in hair:

1 α-fibrillar, low sulphur protein;
2 globular, high sulphur matrix protein;
3 high glycine-tyrosine rich matrix protein.

Amino-acid analytical studies on nail keratin show higher cysteine, glutamic acid, serine and less tyrosine in nail compared to hair and wool (Baden *et al,* 1973; Dawber, 1974; Dawber, 1982). Most cysteine is probably in the intermediate plate. Proline and threonine are in greatest concentrations in the high sulphur proteins of the keratin (Gillespie & Frenkel, 1974). Moving boundary electrophoresis, used to compare high sulphur fractions of hair and nail has shown that there are marked differences, suggesting that a different mixture of proteins comprise the high-sulphur fractions of hair and nail. Unlike the epidermis which contains relatively soft keratin, both hair and nails contain hard keratin mainly due to the high sulphur matrix protein.

Calcium

This is found as the phosphate in hydroxyapatite crystals in the cytoplasm and bound to phospholipids, particularly in the dorsal and ventral nail plate. Using alizarin red staining, Cane and Spearman (1967) showed calcium to be the chief metal in nail. The relevance of other metals such as copper, manganese zinc, iron and many others found in small quantities is not exactly known; the subject is well reviewed by Zaïas (1980). Calcium concentration is approximately 0.1 per cent by weight i.e. ten times greater than in hair (Pautard, 1963). It has been suggested that nail calcium is not part of the intrinsic structure but absorbed into the nail from extrinsic sources such as soaps; nail is relatively porous and calcium could enter as ionic calcium or bound to fatty acids. In support of this is the finding of significantly greater quantities of calcium in the terminal free edge of the plate. Pautard (1964) believes that calcium is a constituent part of the nail structure. Supporting this is the finding of the same calcium-magnesium ratio (4.5:1) in the nail as in the blood (Goldblum *et al,* 1953). Both ideas are probably relevant. The authors agree with Forslind *et al,* (1976) that calcium contributes little to the hardness of the nail plate in man.

Phospholipids (Spearman, 1976)

The nail plate contains significant amounts of phospholipids, mainly in the dorsal and ventral layers. These are probably important in maintaining flexibility in the nail (Chapter 3). Free fats and long chain fatty acids are detectable but like calcium it has been suggested that such constituents are of extrinsic origin.

	Matrix		Nail Bed		Nail Plate			Nail Folds & Hyponychium			Dermis
	Dorsal	Intermediate	Basal Layer	Malpig. Layer	Ventral	Intermediate	Dorsal	Basal Layer	Malpig. Layer	Keratinised Layer	
Glycogen	–	–	–	±	–	–	–	–	±	–	
Mucopolysacch.	+	+	±	+	++	–	+	±	+	±	+
Ribonucleic acid	+	+	+	+	–	–	–	+	+	–	
Sulphydryl Groups					++	–	+		+	+	
Disulphide Bonds					++	++	–			+	
Acid Phosphatase	+		±	±	+	++	–	+	+	+	
Alkaline Phosphatase	–		–		–	–	–		+		+
Amylophosphorylase	+	+	+	+	–	–	–				
Cholinesterase											+

Fig. 1.7. Histochemistry of the nail apparatus (modified from Savag & Jancovici, 1980).

In the various sections on the ultrastructure of the component parts of the nail, histochemical changes are described briefly. Fig. 1.7 summarises the histochemistry and histoenzymology of the nail apparatus. A considerable amount of work has been carried out in defining these changes in normal nails but their organised use in aiding diagnosis on nail biopsy tissue has not been fully explored as yet.

Blood supply (Flint, 1955; Ryan, 1973a)

The nail apparatus has a very rich blood supply received from the digital arteries (Fig. 1.8). The two ventral digital arteries are the most important. They give rise to the cruciate anastomoses in the ventral pulp space. Where the two arterial arcades anastomose, branches pass dorsally around the terminal phalanx. On reaching the dorso-lateral surfaces the vessels divide to give rise to a proximal and distal branch which

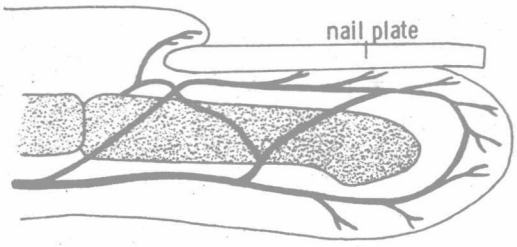

Fig. 1.8. Blood supply to the tip of the finger and nail apparatus showing many anastomoses.