



# Pachyonychia Congenita Project

15 March 2005

## Use of Articles in the Pachyonychia Congenita Bibliography

The articles in the PC Bibliography may be restricted by copyright laws. These have been made available to you by PC Project for the exclusive use in teaching, scholarship or research regarding Pachyonychia Congenita.

To the best of our understanding, in supplying this material to you we have followed the guidelines of Sec 107 regarding fair use of copyright materials. That section reads as follows:

Sec. 107. - Limitations on exclusive rights: Fair use

Notwithstanding the provisions of sections 106 and 106A, the fair use of a copyrighted work, including such use by reproduction in copies or phonorecords or by any other means specified by that section, for purposes such as criticism, comment, news reporting, **teaching (including multiple copies for classroom use), scholarship, or research**, is not an infringement of copyright. In determining whether the use made of a work in any particular case is a fair use the factors to be considered shall include - (1) the purpose and character of the use, including whether such use is of a commercial nature or is for nonprofit educational purposes; (2) the nature of the copyrighted work; (3) the amount and substantiality of the portion used in relation to the copyrighted work as a whole; and (4) the effect of the use upon the potential market for or value of the copyrighted work. The fact that a work is unpublished shall not itself bar a finding of fair use if such finding is made upon consideration of all the above factors.

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.

inflammation of the matrix, as in scarlatina, syphilis, &c.; and chronic inflammation of that organ, induced by local injury, burn, frost-bite, or arrest of nutrition, as in elephantiasis Græcorum.

*Faulty shape of the nails, deformitas unguium*, usually results from some disturbance of the secreting organ, the matrix, and sometimes from malnutrition, the consequence of deficient innervation, as in cases of paralysis. The nail may be unnaturally long or short, too broad or too much compressed at the sides, *arcuata unguis*, too prominent, too flat, or too much arched or curved. Sometimes the nail projects longitudinally, like the angular ridge of a house-top, and, when thickened, has rather the character of a talon than a nail; sometimes it is concave on the surface, the direction of the concavity being longitudinal, or horizontal; and sometimes it is remarkable for its curve

over the end of the finger, as in the *ungues adunci*, the arched and hooked nails so frequently seen in scrofulous and consumptive persons. Now and then we meet with *supernumerary nails*, generally in the form of two rudimentary nails blended together, evincing an intention of bifurcation of the finger or toe. And, occasionally, from some accident of development, we find the nail occupying an *abnormal situation*. The most remarkable illustration of the latter phenomenon is the production of a nail-like growth at the extremity of amputated fingers; even on the stumps of the first and second phalanges, where no rudiment of the original matrix can have existed.

ABNORMAL GROWTH of the nails is illustrated, sometimes by deficient growth, sometimes by growth in excess, and sometimes by unnatural growth. We have met with instances in which the growth of the nail has been so remarkably slow, that they might almost be taken for examples of complete arrest of growth. In other cases, the growth of the nail has been as striking for its rapidity or extent; the nails sometimes attain an enormous size in boucnemia; and an instance of extraordinary growth of the nails is recorded by Saillant, as occurring in a woman named Melin, and named from this peculiarity the "femme aux ongles."\* Where there has been neglect, as in bedridden and elderly persons, the nails are apt to acquire an extravagant size. We have in our possession several toe-nails of this kind, measuring two and three inches in length. Rohout, in a paper addressed to the Academy of Sciences of Paris, has described a toe-nail which

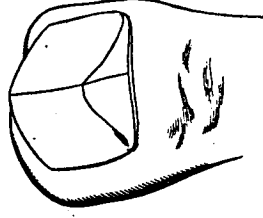
\* Saillant, Memoire sur la maladie de la femme dite aux ongles. Paris, 1776, 8vo.

The disorders of the nails are referrible to alterations in the disposition of the skin around the margin of the nail; in the development, growth, colour, and texture of the nails themselves; and to inflammation, suppuration, and ulceration of the matrix and adjoining soft parts.

In reference to the first of these alterations, we sometimes meet with cases in which the epidermis of the margin of the nail-follicle remains attached to the surface of the nail, and advances with its growth, until the nail is more or less completely covered with a thin film. This is termed *pterygium unguis*. When less complete, the epidermis is apt to break up into little bands, which curve back and project from the skin around the root of the nail, giving rise to much inconvenience, and often, from being accidentally torn, causing soreness and tenderness of the skin. These little ragged bands of cuticle are called *agnails*, on account of the pain and suffering which they not unfrequently occasion. At other times, instead of growing forward, the cuticular margin of the follicle recedes, and exposes the root of the nail. This affection is rare, but has received the name of *scius unguium*. Rayet remarks that he has observed it among curriers; we have seen it a few times where there existed a chronic state of inflammation of the nail-follicle.

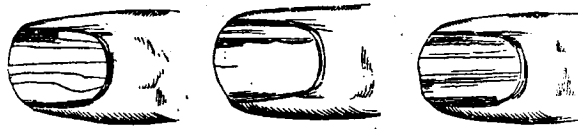
ABNORMAL DEVELOPMENT of the nails is evinced in the occasional congenital absence of those organs; in their disposition to shed from time to time, with and without apparent cause; in their irregular shape, their occurrence in greater number than natural, and their abnormal position. The *congenital absence* of one or more of the nails is rare, but such cases are sometimes seen. Of the *fall of the nails* (*lapsus unguis*), we have met with several examples, in which they were regularly shed; new nails being formed beneath, while the old ones were becoming loosened previously to falling off. This morbid peculiarity has been recognised by pathology, and has received the name of *alopecia unguibus*. More frequently, the fall of the nail results from

Fig. 1.



almost everything she touches; and when torn, they occasion bleeding and soreness of the matrix. Sometimes the nails are brittle from a morbid alteration of texture, being converted into a white earthy matter which breaks up into shapeless granules on the application of a slight degree of pressure.

Fig. M.



Another change in the nails, for which we have been several times consulted, is a fibrous state of those organs, which appear to be made up of a thick stratum of fibres, closely packed together, but becoming loosened here and there, so that separate fibres are met with on the surface. The surface of the nail is necessarily rough, ragged, discoloured, and marked by numerous dark, longitudinal lines; and, besides being very unsightly, adheres like a burr to any rough material with which it comes in contact.

Another variety of degenerated nail has the appearance of being *eroded*, or worm-eaten, *tinea unguium*, and sprinkled over more or less abundantly with hollow pits. The state of the nail, like the preceding, is unsightly, and the sufferer applies for relief, rather on account of the ugliness of appearance, than from any real inconvenience. Unhappily, medicine can afford very little aid in these cases.

A. B. C. Fore-finger, middle, and ring-finger, showing the broken and disfigured state of the nail, in degenerative unguium; fissura unguium.

## ONYCHIA.

ONYCHIA is an inflammation of the matrix of the nail, sometimes confined to that structure only, but more frequently involving the immediately surrounding soft parts. The inflammation is succeeded by suppuration and ulceration to a greater or less extent, the production of granulations of large size and unhealthy texture, of a fungous character, and an ichorous, sanious, and fetid discharge. The inflammation is sometimes superficial; sometimes extends to the deeper parts; sometimes terminates in the loss of the nail, or a part of the nail; and sometimes runs on to disease of the periosteum and bone. It is always a painful, and occasionally a troublesome and rebellious disease.

Onychia may be partial, that is, may attack a single nail only, or it may affect a greater number; it may also involve a part or the whole of the nail in destruction. It may occur along the edge of the nail, or at its root; and it may be either acute or chronic in respect of its progress.

measured nearly five inches. Rayer mentions two great toe-nails which measured three inches, and were spirally twisted like the horns of a ram; and Saviard "saw a patient in the Hôtel Dieu, who had a horn instead of a nail upon each great toe." Musæus, in his "Dissertatio de Unguibus Monstrosis," records a case of unnatural growth of the nails, in which those organs resembled talons, and were five inches in length; while similar horny growths were developed on the skin of other parts of the body.

DISCOLORATION OF THE NAILS is met with in some diseases of the skin affecting the matrix of the nails, as in eczema and alphas; and it also results from injury, as in ecchymosis beneath the nail, the consequence of a bruise, *ecchymoma unguis*. Not unfrequently the nail is speckled with small roundish white spots, *selene unguis*, figuratively named by the ancient writers, *flores unguium*; and, by the moderns, less elegantly termed *merdacia*, or lies. These spots are more common in the nails of children than adults, and result from some slight injury done to the matrix of the nail during the progress of growth.

ABNORMAL TEXTURE of the nail may present itself in the three-fold form of increased thickness, altered density, and altered smoothness, constituting the state of disease known as *degeneratio unguium*. Or the matrix of the nail may be the subject of inflammation, suppuration, and ulceration, constituting *onychia*. We may now proceed to the consideration of these two forms of disease.

## DEGENERATIO UNGUIUM.

Syn. *Defœditio unguium*; *scabrities unguium*.

The nails sometimes acquire an excessive degree of *thickness*, and the increase of bulk is accompanied with a yellow and dirty discoloration, the nail resembling horn, rather than its own natural texture, both in colour and density. In this state it is not uncommon to find the nail separating from its matrix, and a dry, whitish, broken substance collected in large quantity beneath it. Two such cases are now before us, in which the greater part of the fingers of both hands are affected in this manner.

In another case, also under our observation at the present time, the nails are reduced to a mere film, and so soft and brittle in texture, that they split and break with the slightest pressure, *mollities unguium*; the texture of the nail is healthy, but its quantity deficient, and with the absence of quantity there is also, as a matter of course, an equal defect of firmness and tenacity. This state of the nails is a subject of much annoyance to the lady who is the sufferer from the affection; the ends of the nails are always ragged and broken, they catch in her clothes, and in

It varies also according to the cause which shall have given rise to it; whether that cause be one of *external injury*, as a bruise, a splinter, a puncture, or a foreign body lodged beneath the nail. Whether it have been produced and kept up by the pressure of the side of the nail against the soft parts, giving rise to the *growing in* of the nail. Or whether its cause may be *internal* and constitutional, as that occasioned by eczema, scrofula, or syphilis.

In onychia resulting from external injury the treatment should be such as is applicable to a similar injury, when occurring in any other situation. If pus be formed beneath the nail, the nail should be thinned by scraping until reduced to a film, and then it should be punctured to allow the escape of the matter; the same manœuvre is applicable to the relief of the inflamed matrix when swollen by congestion; or to the removal of a foreign body lodged beneath the nail.

Onychia from in-growing nail originates frequently, and indeed generally, in interference with the growth of the nail in length by contact with the boot or shoe. Prevented from growing forward, the nail is apt to spread laterally, and the skin pressed forcibly against it, becomes inflamed; the inflammation increases from day to day; or, after much walking the skin becomes red, swollen, and painful, adding still further to the pressure; then suppuration takes place, next ulceration, the production of fungous granulations and an unhealthy discharge. The medical management of this case consists in the withdrawal of the cause by rest and position; then the removal of the offending border of the nail; and the general relief of the inflammation by scraping the whole surface of the nail, until it become thin and pliable, and capable of yielding to the swelling of the inflamed parts.

Eczematous onychia is usually accompanied with eruption on other parts of the fingers and hands, and frequently on several regions of the body. It does not call for special treatment, but usually gets well when the general eczema is relieved.

Scrofulous onychia commonly attacks the nail follicle, and the inflammation spreads to the whole of the skin immediately bordering the nail; by degrees the entire end of the finger is inflamed and enlarged, and the nail looks as if it were sunk in a deep hollow, surrounded by a tumid border of a deep red or purplish hue. The finger has the appearance which is commonly denominated *clubbed*, and, as the disease advances, and fungous granulations spring up from the ulcerated surface, the nail is sometimes completely concealed from view.

Syphilitic onychia has already been considered in the chapter on syphilitic eruptions; like scrofulous onychia it may attack the follicle of the nail, and is not confined to the lateral border, as in

the case of in-growing nail. It is always accompanied by other signs of syphilis, and frequently with syphilitic eruption in different regions of the body.

Onychia maligna is a name assigned to a peculiarly obstinate and severe form of the disease, apparently of idiopathic origin. It begins in the follicle of the nail, and is attended with excessive pain, and the secretion of an offensive discharge. Malignant onychia sometimes extends so deeply as to effect the periosteum and bone; and is often many months under treatment.

Onychia is not unfrequently accompanied with irritative fever, the consequence of severe pain and loss of sleep, and the constitutional disorder tends materially to the aggravation of the local disease.

The *diagnosis* of onychia is self-evident, the only disease with which it can possibly be confounded being *paronychia*, or common whitlow. Whitlow, however, begins in the skin at the end of the finger, or in the vicinity of the nail, and does not attack the matrix; it is simply a phlegmon in this situation; but when of large size, or, in its commencement, attacking the skin close to the border of the nail, the pus not unfrequently extends beneath the latter.

The *causes* of onychia are, direct violence or injury, continued irritation from in-growing nail, and the constitutional and specific affections, eczema, scrofula, and syphilis. The cause of onychia maligna must be referred to some general derangement of the constitution, probably mal-assimilation and cachexia.

*Treatment*.—In *degeneratio unguium*, we have derived advantage from the local application of iodide of potassium and chloride of zinc in solution.

The treatment of onychia must be local and constitutional; local, to subdue the congestion of the inflamed part, to relieve the pressure, to give early exit to pus, and to stimulate to a healthy healing process; and constitutional, to remove internal causes of irritation, and maintain and support the powers of the system against the morbid influence of the local disease.

The general principles of local treatment are, position, maintenance of an uniform temperature, and exclusion of air by a dressing of basilicon ointment. If foreign bodies are present, they must be removed in the first instance; and if the evil be kept up by the nail, either in consequence of its thickness and density or of its pressure upon the inflamed part, the nail must be scraped, and the offending parts gently cut away. It must be remembered that by scraping, the nail may be reduced to the thinness and softness of cuticle, and that in this way one of the chief difficulties in the treatment of onychia, and one of the

chief causes of its virulence, may be swept away. We may thin the nail so as to reach the bed of the pus, and liberate the matter with ease; and the same operation practised along the middle line permits of the folding of the nail, and at once relieves the pressure of the edges on the lateral walls. It is useful in some instances to introduce a piece of dry lint between the tumid lateral wall of the matrix and the border of the nail. Exuberant granulations must be kept down by the application of a strong solution of nitrate of silver or sulphate of copper, and the part must be dressed with a desiccative or mildly stimulative ointment, such as the benzoated ointment of oxide of zinc, the unguentum resinæ flavæ, or an ointment of Peruvian balsam.

The *constitutional* treatment of onychia must be regulated according to the indications presented by the general system. If the evil in the constitution be the eczematous or strumous diathesis, if it be syphilitic or cachectic, the treatment must be adapted to those states of the organism. Under all circumstances, digestion and secretion should be regulated, and we should keep up the powers of the system by nutritious diet and by tonic medicines, to enable it to resist the attack of disease, and to furnish the means of restoration of the disorganised parts to a healthy status. The remedies the most likely to be needed are, quinine and iron, iodide of iron, the mineral acids with vegetable bitters, and cod-liver oil.

## CHAPTER XXIII.

### AFFECTIONS OF THE HAIR SYSTEM.

THE HAIR SYSTEM is composed of two parts, the *hair* itself, and the *follicle* in which the hair is implanted; and the diseases of these organs are represented by their aberration from the normal standard of structure and function. In the case of the hair, the aberration may be one of *quantity, length, colour, direction,* and *texture*; and in the case of the follicles, there may be *excess, or diminution, or alteration* of secretion, or an organic change of structure.

The disorders of the hair having reference to quantity and colour are, *hirsuties, hairiness, or excess of hair; alopecia, or deficiency of hair; trichonosis decolor, or discoloration of hair; and canities, or whiteness of hair.* The disorders of texture, remarkable especially for brittleness of structure, are *fragilitas crinium, and trichonosis tonsurans or ringworm.* And the disorders of the follicles are, simple alteration of secretion, and an organic change in the structure of the follicle constituting *favus, kërion or scalled head, and sycosis.*

In a tabular scheme the diseases of the hair system may be arranged under four heads, as follows:—

#### 1. Diseases of quantity.

Hirsuties, Alopecia areata,  
Alopecia vulgaris, " calva, vel calvities.

#### 2. Diseases of colour and direction.

Trichonosis decolor, Canities, Trichiasis.

#### 3. Diseases of texture.

Fragilitas crinium, Trichonosis tonsurans, Plica polonica.

#### 4. Diseases of follicles.

Morbi sebacei, Kërion,  
Favus, Sycosis.