



Pachyonychia Congenita Project

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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.

a supraclavicular incision, no matter in what position the arm is placed.

3. Because of this incision, we were able to milk out the thrombus from the entire length of the brachial artery by actual massage of the axillary artery, together with external manipulation, until dark blood, free from clots, could be expressed freely in a retrograde manner.

4. The thrombus that had formed in the second and third parts of the subclavian, axillary and brachial arteries was due to the stagnation of blood in the vessels below the embolus, which acted not unlike the cork in a bottle, and tended to produce a vacuum. This, with the venous blood pressure ahead, accounts for the formation of the thrombus.

5. Collateral circulation, which will develop perfectly following ligation of the subclavian, axillary or brachial artery at any one location, cannot take place when a thrombus extending from the subclavian to the lower end of the brachial artery blocks all the anastomosing arteries that go to make up the collateral circulation about the shoulder joint.

6. It is the sudden, complete occlusion of the vessel, with immediate block of the arterial flow, which leaves the arteries filled with blood that produces the thrombi and causes the ischemic gangrene, whereas an incomplete occlusion allows the vessels to accommodate themselves to the physiologic changes, and, in this instance, collateral circulation sufficient to prevent gangrene usually develops.

7. We believe that the site of lodgment of the embolus, whether in the upper or in the lower extremity, depends on the phase of respiration present; for example, during inspiration, the intra-abdominal pressure is increased, the thoracic pressure being decreased. At this time, an embolus passing along the periphery of the blood stream by the opening of one of the large vessels tends to be aspirated into the vessel. The pressure of the blood stream behind drives it forward until lodgment at a bifurcation of or point of narrowing in the vessel. In suturing a longitudinal incision in a large blood vessel, the lumen at this point is markedly narrowed; and, although enough blood reaches the extremity beyond for purposes of normal function, the arterial pressure may not be sufficient to produce a distinct palpable pulsation in a peripheral artery.

8. The element of time is an important factor in securing good results. For example, cases in which operation is performed before six hours have elapsed give almost uniformly good results, whereas those in which operation is performed after twelve hours have elapsed give a high percentage of failures. The reason is obvious: a thrombus forms rapidly following the insult, and after several hours it is difficult to remove because of its adherence to the vessel walls by prolongations that extend into the branching vessels. The condition, no doubt, reaches into the smaller arteries and arterioles, so that complete removal is impossible, and, therefore, permanent ischemia, with cell death and gangrene, is inevitable.

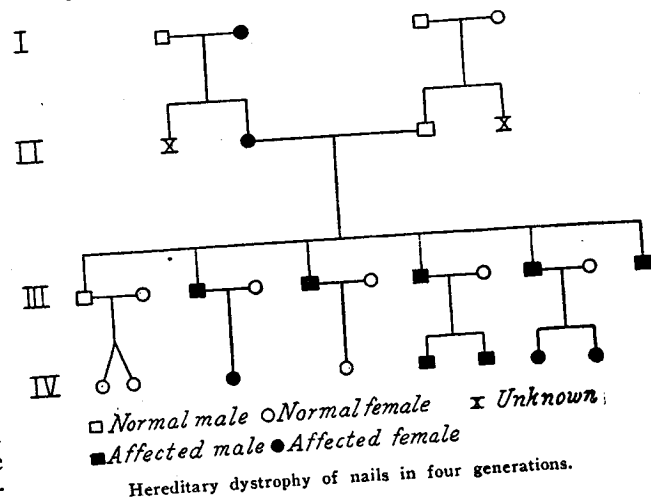
World Prevalence of Disease.—Health conditions throughout the world seem not to have been disturbed by any unusual or special epidemic prevalence during the months of October and November, 1924, judging by data made available in the December number of the epidemiologic report issued by the Health Section of the League of Nations' secretariat. Mortality from all causes in a number of large cities scattered over the world showed some seasonal increases, but agreed very closely with the corresponding periods of 1923.—*Pub. Health Rep.* 40:248 (Feb. 6) 1925.

HEREDITARY FAMILIAL DYSTROPHY OF THE NAILS

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Affections of the nails are frequent in dermatologic practice, but the hereditary conditions are relatively uncommon. Moreover, they are interesting from the standpoint of genetics. In the family reported here, the deformity has occurred with considerable frequency, the defect being present in twelve cases through four generations.

The onychoses may be congenital or acquired. The congenital types may consist of partial atrophy or complete absence. The etiology of the acquired types may be (a) inflammatory, (b) toxic, (c) parasitic or (d) trophic. Local causes are responsible for more than 50 per cent. of the cases of the disease. In every case of nail atrophy, syphilis should be ruled out by a Wassermann test, heredity by a careful history, and tinea infection by scrapings and cultures.



Congenital dystrophy of the nails may be limited to the fingers or toes, or may involve both and may be apparent at birth, or appear at puberty or later. Clinically, the disturbances of nail growth and nutrition give rise to a varied picture, from complete absence to thinning, thickening, irregular growth, splitting, discoloration, elevation or distortion from subungual hyperkeratosis.

Anomalies of other epidermal structures may coexist in these cases. Among these may be cutaneous manifestations, such as ichthyosis, epidermolysis bullosa and keratosis palmaris; hair affections, such as hypertrichosis and alopecia; and involvement of the teeth with premature or abnormally late eruption.

The association of dystrophy of the hair and nails with hypothyroidism has been observed by Barrett,¹ who reported fourteen cases in six generations. Inherited differences in the glands of internal secretion may be responsible for a whole series of changes in the growth and nutrition of certain tissues, and must be taken into consideration as having an important bearing on the study of heredity.

In two of our cases observed in the third and fourth generations, the nails of the fingers became involved

1. Barrett, A. M.: Hereditary Occurrence of Hypothyroidism with Dystrophies of the Nails and Hair, *Arch. Neurol. & Psychiat.* 2:628 (Dec.) 1919.

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early in life. The thumb nails were practically absent, being replaced by rough hyperkeratotic tissue. The other nails bore longitudinal fissures and were split at the ends. The skin under the free border of the nails was considerably thickened and verrucous. The Wassermann reaction and examination of the scrapings of the tinea were negative. Physical examination revealed no other abnormalities in the cases studied. In the course of a routine history, careful inquiry revealed the fact that there were several other cases in the family, and formed the basis for the accompanying chart.

In this study, comprising twenty-five persons in four generations, at least twelve positive cases of dystrophy of the nails have been determined. The disease has appeared in each generation, the defect having been handed down from parent to child through direct transmission by the mechanism of simplex inheritance, the offspring receiving a single or simplex dose of positive determiners from the affected parent. In spite of the small size of the families in the second and fourth generations, the defect definitely appears to be a mendelian dominant or positive characteristic and is inherited by the affected individual as a new trait. Dominant or positive types of a disease are relatively common in any given family, since one half or all of the offspring commonly show the trait.

Metropolitan Building.

A TREATMENT FOR WHITE PHOSPHORUS BURNS *

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Although white phosphorus has largely been replaced by various substitutes in the industries, this is not true in military practice. During the World War enormous quantities of white phosphorus were used on both sides, as a smoke screen producer, as an incendiary agent, and above all for its burning and terrorizing effect on hostile troops, when used in Stokes mortar shells, Livens projectors and hand grenades. The American production of white phosphorus for war purposes had reached the enormous figure of 1,006 tons at the time of the armistice. While the recent gas treaty prohibits the use of chemical agents such as the toxic smokes and lethal war gases, this prohibition does not cover the use of nontoxic smoke producers, and white phosphorus is still a permitted war agent. In consequence it is widely used in troop maneuvers, and a number of serious accidents have occurred, both in filling plants and in the field, as a result of premature bursts of grenades and shells.

Great difficulty has been experienced in treating these cases, owing to the fact that the phosphorus adhered to the flesh, where it continued to fume and occasionally burst into flame again. This necessitated the placing of such cases in a continuous bath, and immediately on removal from the bath the phosphorus would begin to fume and smolder again.

The following methods of treatment were considered:

1. To use a phosphorus solvent containing some neutralizing agent. This method at first glance appears to be the ideal one, but the only satisfactory solvent for phosphorus, carbon disulphid, is itself highly toxic. The same objections apply in less degree to the other

solvents; also, they would be highly painful on a burned area. The possibility of the neutralizing agent not acting immediately on the dissolved phosphorus has also to be considered, as the burned area would rapidly absorb the dissolved phosphorus, and acute phosphorus poisoning would inevitably result.

2. To use oxidizing agents that would rapidly oxidize the phosphorus. This idea was abandoned, since either the reaction would proceed so slowly as to necessitate long applications of the chemicals, or, if stronger reagents were used, the reaction would proceed with such violence as to cause further tissue damage, and in addition any reagents capable of acting rapidly would be too irritant in themselves.

3. To use agents that would rapidly place an insoluble, inert, nontoxic coating on any particles of phosphorus present, such particles to be later removed by irrigation with some nonirritant such as physiologic sodium chlorid solution.

A number of experiments were performed on phosphorus in vitro, using solutions of nontoxic chemicals, and it was found that copper sulphate acted the most rapidly. Copper sulphate has long been used as an antidote for phosphorus poisoning, coating the ingested phosphorus with a layer of insoluble and nontoxic copper phosphid and acting as an emetic. A lump of phosphorus dropped in a 1 per cent. solution of copper sulphate is immediately turned black, and no longer fumes when exposed to the air.

Copper sulphate is used in medicine for a number of purposes. Solid sticks are used in the treatment of trachoma, a disease of the eyes. Solutions of various strengths and also copper sulphate pastes have occasionally been used as local applications to stimulate indolent ulcers. Bevan¹ has used copper sulphate successfully in actinomycosis. He gave from 1 to 3 grains (0.065 to 0.2 gm.) daily internally, and applied a 1 per cent. solution to the ulcerated area. Cushny states that a 1 per cent. solution is occasionally used as a lotion in wounds, and that it is much less dangerous to man than many other remedies used as parasiticides and disinfectants. Sollmann states that its toxicity to animals is low. Experiments on mice showed us that the toxicity of a 2 per cent. solution of copper sulphate for mice by subcutaneous injection was 625 milligrams per kilogram.

The following experiments on animals were then performed: Areas were shaved on two guinea-pigs; the pigs were then morphinized, and anesthetized with chloroform. Lumps of phosphorus were placed on their skin and ignited. The flames were then quenched with water, but the phosphorus continued to fume. "Milk of Magnesia" had no effect on the phosphorus, which continued to fume. A 1 per cent. solution of copper sulphate was then applied, and the fuming immediately ceased, the phosphorus having a heavy black film of copper phosphid on its surface.

A small black dog with abdomen shaved was morphinized and then anesthetized with chloroform. Lumps of phosphorus the size of a pea were applied to the skin and ignited. Cotton pads soaked in water were applied, and the flames extinguished. The phosphorus remaining fumed markedly. A pad of 3 per cent. copper sulphate was applied for two minutes. Fuming ceased, and the phosphorus lumps were black.

The foregoing experiment was repeated with 1 per cent. copper sulphate with the same favorable results.

* From the Medical Research Division, Edgewood Arsenal.

1. Bevan: *Semaine méd.*, 1905, p. 558.