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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.
at least four

5. ZINNAMON, B. L., SEEBERG, V. F., Penicillin in water-in-oil emulsion as compared with those obtained by single or multiple injections of aqueous penicillin solutions. J. F. M. D. Infor. 27:725-730. August '46.


PACHYONYCHIA CONGENITA

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Jadassohn and Lewandowski in Monographie Dermatologica, Fasc. 1, 1906 page 28, tab. VI, Figs. 8, 9, described a patient, girl, 15 years of age under the title, pachyonychia congenita; keratosis disseminata circumscripta (follicularis); tylomata; leukokeratosis linguae. Individual case reports and reviews of the literature have been made in the past forty years. The clinical symptoms are unmistakable and the history almost repetitious. The mystery of the etiology remains. Therapy has seemed unavailing. The relation of the condition of the nails to the definite pathology of the skin has not been clarified.

The possibility of failure of hormonal output has been suggested. The relationship to lack of vitamin D had not been previously cited in the case reports studied. The appearance of the patient offered the opportunity of consultation with the director of the New York City Nuti- tion Institute, Dr. Norman Jolliffe after blood studies reported all findings normal except for ascorbic acid. The findings here were 0.3 mgs percent with normal given as 0.8 mgs percent.

It was possible also to secure abstracts from two hospitals where the patient had been under observation when much younger. The case presentation of Dr. Capollaro is on this patient. For ten years, the patient was under intermittent observation. In those ten years, his nail condition was recognized. Despite diagnosis, treatment was unavailing.

Joseph P. American born youth of Italian parentage now 17 years of age, was the youngest of four children. He knows many cousins and other relatives. He is the only with the skin and nail disturbance to be described.

The nails were noted to be abnormal at birth. They were thick, with rounded upper surface, and extended beyond the tip of the fingers and toes. All the nails were lost when the infant was 40 days old. The new nails grew thicker, flatter, and longer than normal nails. A physician consulted said there would be great difficulty with them later in life. The child always had repeated loss of nails. Each new growth of nail was more stubborn and thicker than the previous. From about the age of two to about ten or eleven years, inflammation about the nails caused repeated loss of the nails. The nails always regrew with the same abnormalities. It was noted the nails of the two fifth fingers most closely approximated normal nails. They were the thinnest. The normal activities of the boy and youth have never been curtailed by the presence of the abnormal nails. He can write, write with tools and draw. Only in infancy and early childhood has it been possible to cut or trim the nails with scissors or clippers. A contrivance of grinding wheel on shaft of a hobby motor has been utilized to trim the nails for many years. From two to four years of age the patient had kidney trouble and diseases of childhood. The boy has noted the presence of chicken skin or permanent goose pimples on the extremities. Wart-like projections formed on the tips of the elbows and less prominent warty-like ones on the knees.

When the patient was seven or eight years of age, he developed new skin troubles. After wearing a pair of rubber sneakers, he noted a larger blister on one foot. He leaned on the other foot to balance himself and blisters formed on both feet. He has never been without blisters since. About eighteen months ago, when fifteen and one-half years old, he noticed pimples on his face with enlarged pores. Fever blisters appear from time to time at the corners of the lips. When presented before the Atlantic Dermatologic Conference by Dr. Anthony C. Capollaro, March 9, 1948, changes were noted in the oral mucous membranes. The inner surfaces of the lips and cheeks and on the dorsum and edges of the tongue. Dull whitish gray patches, irregular in shape and slightly elevated, were described. The patient mentioned in the historical survey of his condition as present since four years of age following very severe whooping cough. The mother insists patient was 'nearly dead' and survived by means of strong medicine, chiropractic treatment and inhalation from steam kettle. The mother believes strong medicine given caused the white wasting and mushroom of the tongue and cheeks and perianal...
regions. The mother gave Kepler's cod liver oil regularly—says more than 200 bottles.

Appearance of the skin—March, 1915:
The extensor surface of upper arms and forearms present non-inflammatory perifollicular hyperkeratosis. Normal skin exists between the projections. The hyperkeratotic lesions cannot be removed by expression but can be rubbed off with firm agitation. The projections are highest on the tips of each elbow, and only slightly less marked over the knee caps. On the extensor surfaces of the legs, the hair projects from the keratotic lesion.

An erosion is present below the left external malleolus. It measures 2 x 2 cm. The edges are limited by soggy masses of corneum. On the dorsum of the left foot is an erosion 6 x 2 cm. with thin overhanging epidermis at the periphery. Another erosion, 3 x 3 cm. exists on the inner side of the foot below the malleolus. The soggy corneum extends from its lower border to the sole. An erosion, 3 x 3 cm. was present below the external malleolus of the right foot. The callus-like projection extended to the sole and to the Achilles tendon. A soggy adherent mass of corneum was present on the inner malleolus of the right foot. It was impossible to remove the keratin. The sole of the right foot at the arch showed an erosion, 3 x 4 cm. with irregularly thick soggy keratin at the periphery. Erosions were present between the first and second toes of each foot, and between the second and middle toe of each foot with extensions to the contiguous skin of the sole.

The surface of the tongue was white, and thick with definite linear thickening at the periphery of the upper surface and the sides of the tongue. The sides and the under surface of the tongue were pink, smooth and moist. At the mid-point of the tongue extending to each side of the median raphe was an erosion, or smooth section about 1 x 1 cm. in area. The other papillae seemed exaggerated in height, but forcible agitation did not remove any of them. The inner surface of the cheek evidences white irregular patches along the line of teeth.

The child has been vaccinated and given toxoid. At 7 months of age the patient had a first attack of a urinary infection and was unable to void for 3 days. On subsequent visits to a hospital in Brooklyn the mother was told that the trouble was still active over a period of 4 years. At 14 months he had diphtheria, followed by pneumonia, and the patient has been subject to bronchitis since. At two years the patient had measles, pertussis at 3½ years. Since the fourth year the patient has been in fairly good health, generally.

Present illness: In infancy nails were noted to be very small; when they first grew out they were thickened and repeatedly fell off. Since the age of 2 years the patient has had lesions on his skin. These began from measles rash and spread over his body with breakdown of large areas of skin. Since that time new lesions have occurred, beginning with discrete yellow papules occurring singly, then in crops with coalescence to form an open sore which drains pus. The lesions are made worse by cold weather and seem to have some relation to fatty foods and sweets. They are irritated by clothing. Various ointments have been used on the lesions, but clearing is...
A second hospital sent the following case report:

The above named patient registered in our skin department on 11-12-37 at which time a diagnosis of Darier's disease (?) Pachyonychia congenitalis (?) was made. Local medication was prescribed.

Patient: Joseph R. On February 7, 1940 the patient returned to clinic after a lapse of 3 years and a diagnosis of "dermatitis" was made. The patient made routine visits to clinic through August 23, 1940. During this time the patient received 6 Alpine treatments to the toes, and seven 1/2 in units of unfiltered x-rays to the outer lateral side of the foot, dorsum of all toes. Biopsy done on 3-4-40 reported: "Eczema with contact dermatitis."

On January 6th, 1941, the patient returned to clinic with an eruption on the right ankle. Local medication was given. Another visit was made on March 29, 1943. Patient has not been seen in clinic since that time.

Resume: A youth of 17 was observed for some months in whom the clinical diagnosis of pachyonychia congenita was made. The condition had been recognized for years. The skin eruption of the youth was one found associated with pachyonychia congenita, and had features of both keratosis follicularis and epidermolysis bullosa. Studies for vitamin content of the blood indicated normal ascorbic acid content, with other findings normal. Treatment was of no help. The patient was the same as discussed in a case presentation of Dr. Cipollaro.

No solution to the problem of pachyonychia congenita and the associated skin and mucous membrane disorders has been found.

Note: The discrepancy of history given by the mother in 1936 and ten years later should be noted. The 1946 history was taken in stages, repeated, and given to the mother to amend after consultation with other members of the family.

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P O M P H O L Y X

Its etiology, pathogenesis and vaccine therapy, with particular reference to its military importance and post-war significance.

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Chicago, Illinois

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**Formerly Lt. Colonel, Medical Corps, Army of the United States. This paper was ready for publication in October, 1944; however, due to shortage of paper it could not be published during the war. The text has remained substantially unchanged except for the addition based on recent literature and on additional observations made in the Central and Western Pacific, while the author was stationed at Oahu, Territory of Hawaii and on Okinawa, the Ryukyus, Japan with the 23rd General Hospital.