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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.
Clinical Notes, New Instruments and Techniques

MOYONYCHIA CONGENITA
A Brief and New Approach to Treatment

L. FRED MULLINS, M.D.
NEVILLE MURRAY, M.D.
Joel SHAPIRO, M.D., Galveston, Texas

One of the most striking and distressing of the various dyskeratotic skin lesions is the rare and peculiar condition of moyonychia congenita. It is unusual, not only to the patient because of the severe disability involved, but also to the physician who sees it. A difficult problem to rehabilitate many times, the patient who is unfortunate enough to have this condition. The scope of this paper includes (1) a review of the various therapeutic modalities in moyonychia congenita, (2) the presentation of a case, and the use of hospital in the treatment. The success of this method of therapy was a report of Moyocytosis in which suggestive therapy and time was given benefit in a case of congenital dyskeratotic onychodystrophy of Reihl.

Moyonychia congenita is a type of dyskeratosis in which involving chiefly the nails and usually the skin and mucous membranes. Transmission is probably by a simple dominant gene (edwards, 1952), although it has been reported more in families in those of Jewish extraction.

The original and classic report of this entity has been attributed to Joffredson and Lewadowowsky in 1915. Many of the cases cited in the literature have had a resemblance to this case, in which a 17-year-old girl had nail changes from birth, i.e., the nails were thickened and had a sort of hardinless. They could not only with difficulty. The free borders were gradually thinned and showed grayishbrown stains, while the tips were thickened in color. Additional to the nail changes, she had many callosities of the soles which were associated with large plantar bulges during the foot summer months. There

From the Department of Dermatology (Dr. J. J. Hall, Division), The University of Texas Medical Branch, Galveston.

The appearance of dyskeratosis of the palms, soles, and the back. Bright red papules with central follicular plugs were noted on her elbows, knees, elbows, and on the back of the legs. The lesion was excised with irregular, keratotic plaques and had white streaks on the borders and the clinical surface. A similar broach presented only for English papules with follicular plugging over the bones and elbows.

A review of the cases reported to the English literature reveals that the treatment of moyonychia congenita have been mostly unsatisfactory or merely temporarily palliative with only a few notable exceptions. Early reports good results obtained in a 17-year-old boy by using a specially designed rubber base foot mold. This method also helped the mother, whose lesion had recurred after full-thickness skin grafts. The lesions in these cases, however, were confined to the soles. Vitamin A has been said to be, a beneficial effect, on the hyperkeratotic nail changes in this condition when used in high doses for long periods of time (Chustak, 1969); however, this was of no benefit to the hyperkeratotic lesions of the soles. Andrews has reported benefit from the surgical removal of the nails, followed by the scraping of the matrix to prevent regrowth, whereas Wright and Goepfert reported the removal of the distal phalanx with no resulting disability. Wright and Goepfert also reported the healing of painful lesions better with the use of keratinized tissue. When the condition did recur, as it frequently did, the therapy was again instituted, and the lesion healed.

A REPORT OF A CASE

J. L. N., a 17-year-old white boy, was normal at birth and remained so until the age of two, at which time a thickening of the skin on the palms and soles was noted. One year later his fingernails started to thicken and from the matrix. At this time he began to develop large bulges on his palms and soles, the support of which would lead to the formation of a thick layer of hyperkeratotic epidermides.

By the time of admission, the condition was so painful that he could not walk without the use of
crutches. While at home he would crawl about on his hands and knees in order to avoid pressure to his exquisitely tender soles.

Physical examination revealed a 38 in. (147 cm.), 96 lb. (43.5 kg.), slightly hypertrophic white boy, who did not appear acutely or chronically ill. The nails of his hands and feet were elevated from the matrix approximately 0.5 cm. by subungual, heretofore unknown material. There was a tendency toward a medial deviation of the great toenails and a lateral deviation of the nails on the other toes. The nails were extremely hard and could be pared only with hand-cutting instruments. The entire plantar surfaces were covered with a yellowish hyperkeratotic material, which in some places reached a thickness of 2.5 cm. The palmar surfaces were covered with a similar material reaching 1 cm. in thickness with linear extensions to the distal phalanges of each finger. There were erythromelalgia pustules with central hickey plugs on the elbows and knees, and several callouses were noted on the extensor surface of his forearms. A few whitish strüme ran vertically on the anterolateral aspect of the fingers; however, the mucous membranes were free of any hyperkeratoses. The hair was normal in distribution and texture, and the remainder of his integument was smooth and free of abnormalities.

The patient had previously been treated with numerous topical and systemic agents, including tinctures and 50 x-ray treatments of unknown dosage to each foot. Along with these measures, he had received 50,000 units of vitamin A daily for several years, but he had received no benefit from this therapy.

Case and Treatment of Pulmonary Congestive Heart Failure Reported in English Language

<table>
<thead>
<tr>
<th>Patient</th>
<th>Year of Born</th>
<th>Year of Treatment</th>
<th>Causative Agent</th>
<th>Treatment</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>29-year-old Jewish boy</td>
<td>1929</td>
<td></td>
<td></td>
<td>Cutting of nails</td>
<td>Not reported</td>
</tr>
<tr>
<td>30-year-old Jewish boy</td>
<td>1930</td>
<td></td>
<td></td>
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<td></td>
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<tr>
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<td>1931</td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td>31-year-old Jewish boy</td>
<td>1932</td>
<td></td>
<td></td>
<td>Not reported</td>
<td></td>
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<tr>
<td>32-year-old Jewish boy</td>
<td>1933</td>
<td></td>
<td></td>
<td>Not reported</td>
<td></td>
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<tr>
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<td>1934</td>
<td></td>
<td></td>
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<tr>
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<td>1935</td>
<td></td>
<td></td>
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<tr>
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<td>1936</td>
<td></td>
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<td>1938</td>
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<tr>
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<td>1939</td>
<td></td>
<td></td>
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<tr>
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<tr>
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<td>1944</td>
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<tr>
<td>44-year-old Jewish boy</td>
<td>1945</td>
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<tr>
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<td>1946</td>
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<tr>
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<td>1947</td>
<td></td>
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<tr>
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<td>1948</td>
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<tr>
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<td>1949</td>
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<td>1950</td>
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<td>50-year-old Jewish boy</td>
<td>1951</td>
<td></td>
<td></td>
<td>Not reported</td>
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</tbody>
</table>

On admission to Johns Hopkins Hospital, July 9, 1932, he was unable to walk and moved about inside of a wheel chair. The was treated with cold suds and petrolatum, which brought about relief and B.

E. coli. On Jan. 14, 1944, it was decided to try hypoglycemia treatment. In the aforementioned related congenital hereditary disease, congenital ichthyosiform erythroderma, he responded favorably to such treatment.

All other local and systemic therapy was discontinued, and the patient was seen by one of us.

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HYPONYCHIA CONGENITA

S. M.) at one- to three-day intervals during the
ens of his hospital stay. During these sessions
attempts were made to investigate the patient's
malade condition and to induce abractive phe-
ness by means of hypnotic procedures. The pa-
time was the same at the time measured, as well as
stage and extent of the lesions with the
action. Furthermore, posthypnotic suggestions were
to the effect that clearing of the lesions would
be, at first, in the left hand. Within three
g time there was noticeable softening of the keratotic
ail on the left hand and considerable subjective

John Sealy Hospital, July 16,
to walk and moved about the
He was treated with daily
which brought about im-
cions of the hyperkeratoses could
shape, and, later, on
was faster than re-
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which was probably due
to geology of keratin. On Aus-
thy hypnotherapy, insomuch as
mental congenital hyperkeratotic
ions erythromelalgia, had
to such treatment.

terapy was discon-
was seen by one of us.

improvement. By Aug. 24, the 10th day of treat-
ment, he was able to tolerate a pair of leather shoes.
Aug. 27, he stood on his feet without pain for the
first time in his life that he could remember, and
Aug. 31 he was able to walk the length of the
bed without pain. Discharged on Sept. 8.
Of the therapy, and psychological therapy was con-
ined at biweekly intervals. After two months this
therapy was discontinued. When he was last seen
be months later, he was walking with only slight

treatment.

The concept that there may be psychic influences
the condition of the skin is not surprising or
Sycobum (1861) wrote of "hysteric diseases," it
which he described angioneurotic edema affecting
Fig. 2. More attempts have been made more re-
cently to demonstrate a causal relationship between
psychic stresses and skin disorders. Becker and
amay (1947) list many diseases which they
have a more or less psychic component, and
recently Hall-Smith and Norton report significant
psychiatric factors in 78.5% of dermatologic pa-
ents selected at random. In view of the fact that
marked benefit resulted from suggestive hypno-
otherapy in this case of pachyonychia congenita
and in one instance of Bracy's ichthyosiform erythro-
dema, perhaps this form of treatment should be
ried in other congenital dyskeratoses when more
conserve treatment has been of no avail.

SUMMARY

The treatment of pachyonychia congenita as re-
presented in the English language is reviewed, and a
case of pachyonychia congenita in a 12-year-old
boy is reported. This case was treated by sug-
gestive therapy with improvement after failure
with the more conservative methods of therapy.

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J. L. N., before and after hypnotherapy.
A. M. A. ARCHIVES OF DERMATOLOGY


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J. B.

Clara, J. B.

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R. M.

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violence.