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 The fact that a work is unpublished shall not itself bar a finding of fair use if such finding is made upon consideration of all the above factors.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.

2386 East Heritage Way, Suite B, Salt Lake City, Utah 84109 USA
Phone +1-877-628-7300 • Email—Info@PC-Project.org
www.PC-Project.org

The Developmental Nature of Pachyonychia Congenita

A Twenty Year Study of a Case

Carroll S. Wright, M.D.

PROFESSOR OF DERMATOLOGY, TEMPLE UNIVERSITY SCHOOL OF MEDICINE

PHILADELPHIA, PA.

Pachyonychia congenita is a rare congenital anomaly originally described under this title by Jadassohn and Lewandowsky.[1] According to Ormsby and Montgomery,[2] it is characterized by dystrophic changes in the nails, palmar and plantar hyperkeratosis, anomalies of the hair, leukoplakia, follicular keratoses of the acneform type, particularly about the elbows and knees, and dyskeratosis of the cornea.

In 1947 Wright and Guequierre[3] published a report of 2 cases with studies on therapy. One of these cases, a boy, entered the Temple University Hospital in 1936 at the age of 6, and he has been under constant observation up to the present time. Between June 1942 and June 1945 there was a total of six admissions to the Temple University Hospital, varying in length from a few days to several weeks. In 1942, the picture presented was as follows: The skin was generally dry and thickened, with many elevated lesions resembling keratotic plugs, particularly over the shoulders, elbows, and knees. The feet had denuded, crusted patches extending from the soles up the medial and lateral aspects. Removal of the crusts revealed reddened bases having a shiny or somewhat glazed appearance. The hair was dry and lusterless. The buccal mucosa and tongue showed patches suggestive of leukoplakia. Most striking were the changes in the nails. The nails of the fingers and toes were greatly thickened, some to a depth of 0.5 inch (1.2 cm.). The growth of the nail appeared to be away from the nail matrix rather than from the nail root (fig. 1).

The patient and his parents were chiefly interested in therapy for the crusted inflammatory patches and the nails. Because of the overgrowth of the nail plate, he was able to make but little use of his fingers. Wet dressings of buffered cysteine hydrochloride, as described by Goldberg,[4] were continuously applied to the crusted areas, and within 10 days all lesions were healed. (It is worthy of note that during subsequent recurrent outbreaks of these inflammatory crusted lesions, various other applications were tried, but invariably it was necessary to use the buffered cysteine hydrochloride to bring about healing.)

A consultation with a member of the surgical staff, Dr. George Rosemond, was requested, and the decision was reached to remove the nail plates completely with the patient under general anesthesia. This was done Nov. 17, 1942, and the patient was sent home. In June 1943 the patient returned, showing only a partial recurrence of the thickening of the nail plates but with recurrent erosive and crusted lesions on the feet. These again cleared completely with wet dressings of buffered cysteine hydrochloride solution.

In October 1944 the patient returned, showing regrowth of all the nails to about

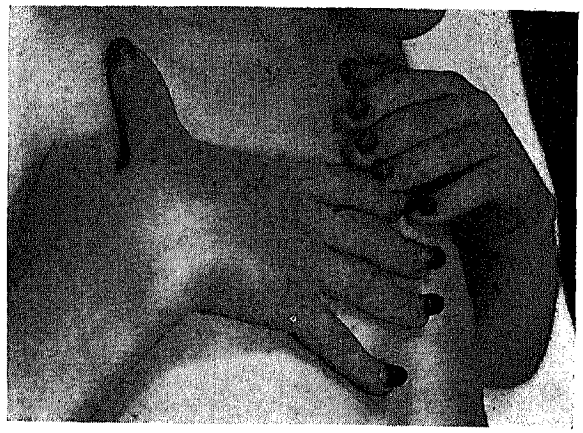

FIG. 1. Fingernails prior to operation, showing marked thickening.

one half of their original thickness. As it was realized that the only method of completely stopping the growth of the nail would mean complete removal of the matrix, which in the condition is apparently the root from which the nail grows, it was decided in consultation with Dr. Rosemond to remove completely the tip of one finger as a trial method of therapeusis. Obviously such a procedure would result in a finger completely lacking a nail (fig. 2).

The next admission to the hospital was in February 1945, and this time the boy requested that the same procedure of removing the distal phalanges be used for all the fingers and the thumb of the right hand, as he had had no discomfort in the one finger and could use it for the first time in his life. The operation was performed with the patient under general anesthesia. In June 1945 the same procedure was carried out on the left hand, the operation having been almost completely successful on the right hand. Apparently a small amount of matrix was not removed from the middle finger, and it resulted in a slight regrowth of the nail (fig. 3).

The results of the operation in which a sufficient amount of the distal phalanx was removed to prevent regrowth of the nails have been remarkable. After complete healing, use of the fingers became possible to an extent that the boy was able to button his shirt for the first time in his life and was able to take piano lessons.

FIG. 2. Removed portion of the distal phalanx of the finger.

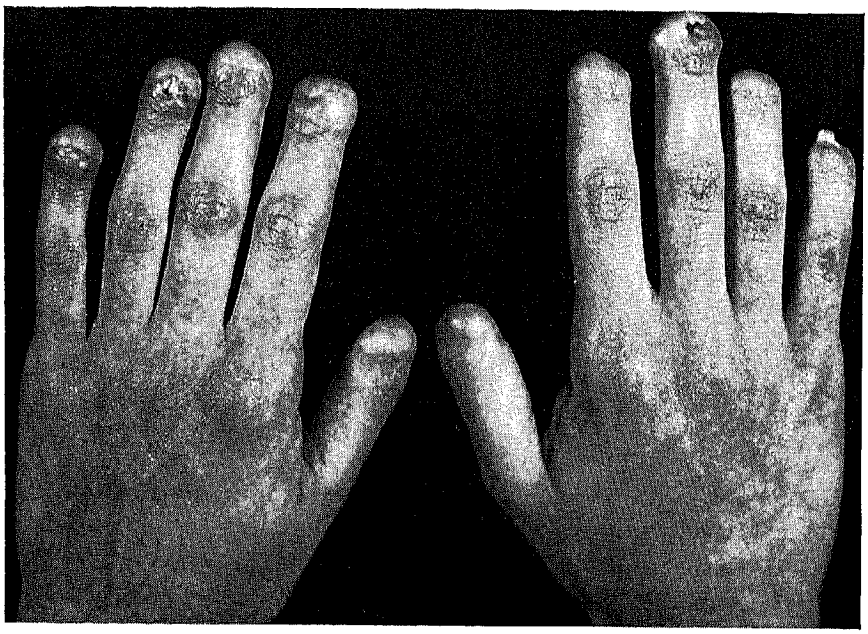

FIG. 3. Appearance of the fingers in 1956.

Now grown to manhood, he is a student in the Osteopathic Medical School and plans to specialize in pathology.

From time to time denuded areas of skin have appeared but have been limited to the soles, which also show areas of hyperkeratosis. By trial and error he has determined that wet dressings of Burow's solution in 1:10 dilution will heal the denuded areas and soften the hyperkeratotic areas to an extent that they are easily scraped off.

SUMMARY

A case of pachyonychia congenita, previously reported, is reviewed after 20 years of observation.

Surgical removal of a sufficient amount of the distal phalanx to prevent regrowth of the nails changed fingers that were previously incapacitating into useful digits.

BIBLIOGRAPHY

1. JADASSOHN AND LEWANDOWSKY. *In:* NEISSER, A., AND JACOBI, E.: Ikonographia dermatologica, Berlin, Urban and Schwarzenberg, 1907.
2. ORMSBY, O. S., AND MONTGOMERY, H.: Diseases of the Skin, ed. 8, Philadelphia, Lea & Febiger, 1943.
3. WRIGHT, C. S., AND GUEQUIERRE, J. P.: Arch. Dermat. & Syph. 55:819–825 (June) 1947.
4. GOLDBERG, L. C.: Arch. Dermat. & Syph. 36:331–377 (Aug.) 1937.