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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.
RESISTANT EROSI VE LESIONS IN PACHYONYCHIA CONGENITA OF JADASSOHN
TREATMENT WITH BUFFERED CYSTEINE HYDROCHLORIDE

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The purpose of this paper is not to report a case of pachyonychia congenita of Jadassohn, which has been excellently described by Tauber, Goldman and Claassen, Sohrweide, Andrews and Strumwasser and Diasio, or to summarize the thorough reports of Hammett and Reimann and Brunsting and Simonsen on the sulphydryl groups, but to demonstrate the value of buffered cysteine hydrochloride in other conditions than ulcerated lesions. I wish to report here the progress of a girl, aged 20, previously presented by Tauber and his associates. She had had persistent and resistant foul-smelling erosive lesions for a period of seven years and the condition had failed to respond to innumerable types of therapy. Figure 1 shows the type of lesion which was present, not only on the lateral aspects of the feet, but also on the plantar and posterior portions in symmetrical fashion. The lesions were palm sized, with polycyclic borders. They were covered with foul-smelling thick purulent crusts. The patient found it most difficult to walk with her feet in this condition.

The patient was first seen on June 18, 1936, at which time she was advised to go to bed, and wet dressings of alibour water (a solution of zinc and copper sulfates) were applied every four hours. At night the lesions were treated with Wilkinson's ointment (a compound sulfur ointment) and within seventy-two hours all the purulent and crustated material had been removed. There now remained oozing erosions the color of raw meat, devoid of epidermis, extremely tender and moder-

ately painful. Compresses saturated with solution of buffered cysteine hydrochloride, 1 Gm. to 100 cc. of distilled water, were applied directly to the erosions, which were then covered with oiled silk and moistened every thirty minutes. Fresh solutions were prepared daily, and the patient was taught all the necessary details in the preparation. Within

Fig. 1.—Type of lesions on the feet before treatment. (Photograph was furnished by Dr. H. L. Claassen.)

Fig. 2.—Appearance of the lesions on the feet (side view) three weeks after treatment.

three days islands of epithelial proliferation were seen about the sides and in the center of the large erosive areas. Within twelve days the areas on each foot were practically epitheliated, and the patient was permitted to walk to and from the lavatory, which was a few steps distant. After three weeks, except for the pigmented borders which separated the healthy from the previously diseased tissue, it was difficult to differentiate the new epidermis from the surrounding skin.
On further examination it was noted that the patient had unduly high arches, and in order to prevent undue trauma to the feet, it was deemed advisable to have an orthopedic surgeon examine the patient and prescribe a definite type of shoe. Although pachyonychia congenita is a disease associated with other ectodermal disorders, such as keratosis pilaris, leukokeratosis oris and the subjective symptom hyperhidrosis, there has never been any mention in the reported cases of a relationship between it and faulty arches. The following is the report of the orthopedic surgeon, Dr. C. W. Betzner:

On examination with glass and mirror the longitudinal arches were found to be in extreme cavus, with marked compensatory depression of the transverse arches, which resulted in the characteristic two island foot. Slight inversion of the feet was present when the patient was standing, and on a plumbline test the line of gravity fell 1¼ inches (4.45 cm.) outward on the left foot and ¾ inch (1.9 cm.) outward on the right foot from the normal point. The plantar fascia was extremely short and tense. The impression was of paralytic cavus of a severe type.

As a result of this examination a special shoe was fitted, and since that time the patient has expressed her pleasure in the comfort brought about by the new shoes.

Almost a year has now elapsed since the patient was discharged, and up to the time of writing there has been no relapse in the healed areas. Figures 2 and 3, taken after treatment, show complete epithelialisation.

Fig. 3.—Appearance of the soles three weeks after treatment.
In conclusion, I should like to emphasize the statement of Brunsting and Simonsen*: "... cysteine is advocated, not as a substitute... but as an adjunct to the treatment of more resistant cases."

**SUMMARY AND CONCLUSIONS**

Buffered cysteine hydrochloride is advocated in the treatment of resistant erosive lesions, as demonstrated in this report of the results in a case of pachyonychia congenita of Jadassohn.

A fresh solution must be prepared daily, and, as has been previously noted, secondary bacterial growth is inhibited.

Cases of pachyonychia congenita, particularly when there are associated erosive lesions of the feet, should be studied from an orthopedic point of view.

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