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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.
Sebum excretion rate (SER) was reduced by 70–80% in 4 weeks, and maintained at that level during treatment. The reduction was dose dependent. On cessation of therapy the SER of the 0.1 mg/kg group returned to the pre-treatment value in 4–8 weeks. The SER of the other two groups had returned to only 50–60% of the pre-treatment value by the end of the 16-week follow-up period (longer follow-up of the initial groups shows this takes 6–7 months).

Assessment of improvement in acne by acne grading and individual lesion counting of the face show a similar pattern. There was a steady improvement of 60–80% over the 16 weeks but with no difference between the doses. The acne continued to improve off treatment with the 1.0 mg/kg group, but the 0.1 mg/kg group had a high relapse rate requiring further therapy.

Clinical side effects occurred in all patients. They were dose related, and only one patient had to stop treatment. Laboratory side effects have been transient, and dose related.

13-cis-retinoic acid appears to be an effective treatment for patients with moderate or severe acne, and we suggest that it is now possible to begin to formulate schemes of management.

The dermatology services in England and Wales over the last twenty years

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Medical statistics must not be hidden from those who generate them, but should be used to improve the present and to plan for the future. The figures covering dermatology, however, are as hard to find, and as indigestible as sixpences in the Christmas pudding of official publications. This paper presents and combines information found in the Compendium of Health Statistics, Morbidity Statistics from General Practice, and the Hospital In-Patient Enquiries.

About 10% of the population consult their general practitioners each year with diseases of the skin, and more than one in twelve of these are now referred on to skin clinics where the number of new referrals has risen steadily from 357,000 in 1958 to 426,000 in 1974. Consultant numbers have risen by more than enough to balance this, the number of new patients per consultant per working day having fallen slightly from 10.8 in 1963 to 9.6 in 1974.

However the in-patient service has fared less well. The fall in the actual number of dermatology beds and the increase in consultants have combined to reduce the average number of beds used by each consultant from 11.3 in 1963 to 7.1 in 1974. To compensate for this, and in fact largely maintaining the total number of ward admissions, the mean stay of patients in these beds has fallen from 30.5 days in 1958 to 20.3 days in 1977. Less satisfactory have been the marked fall in the occupancy of available beds, the increased number of patients with skin diseases being coped with in non-dermatological beds, and changes in waiting times before admissions.

Pachyonychia congenita: a clinical study of eleven cases

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Eleven cases of pachyonychia congenita have been reviewed. The mode of inheritance is autosomal dominant.

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The clinical features of these cases included: thickened nails, hyperkeratosis of the palms and soles, thinning or alopecia of hair, painful bullae and/or ulcerations of the palms and soles, leukokeratosis oris, verrucous lesions of the extremities, hyperhidrosis, premature eruption of teeth, paronychial infections, sebaceous cysts with milia, and corneal dyskeratosis at times associated with cataracts. Biopsy from the planter lesions usually revealed marked hyperkeratosis, acanthosis, moderate hypergranulosis, and minimal dermal inflammatory infiltration. Treatment with keratolytic agents and lubricants is indicated to areas of palmar and plantar hyperkeratosis but usually produces only transient benefit.

One of the patients developed squamous cell carcinoma over the site of chronic plantar ulceration. Areas of chronic bullous formation or ulceration should be observed for possible occurrence of skin malignancy.

Evaluation of the skin surface by macrophotography and scanning densitometry

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Objective measures of scaling and surface roughness would be an invaluable aid in the diagnosis of scaly dermatoses and in the assessment of the response of skin disorders to treatment.

This study has employed a technique in which photographic negatives of the sites examined (taken at a ×4 magnification under standardized photographic conditions) are scanned in a densitometer. The resulting trace is a measure of the reflected light along the track of the scan and may be quantified by measuring line length or by Fourier analysis.

In the first part of the study, twenty normal volunteer subjects had their forearms photographed before and after application of various occlusive emollient agents in four separate experiments. The resulting densitometric scans (Fig. 1) demonstrate changes in the 4-h period following application of the emollients.

The second part of the study has been concerned with the examination of patients with scaling disorders. Four patients with chronic plaque type psoriasis have been followed serially during treatment and the response correlated with the clinical improvement. Four patients with autosomal dominant ichthyosis and four patients with the xeroderma of atopic dermatitis have also been examined. The scans demonstrate characteristic alterations for the various disorders examined (Fig. 2).

The method provides objective evidence for the response of scaling disorders to treatment and allows the pattern of scaling to be used as a diagnostic discriminant.

A comparative study of toxic erythema of pregnancy and herpes gestationis

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Toxic erythema of pregnancy (pruritic urticarial papules and plaques of pregnancy; Lawley et al., 1979) is a pruritic urticarial eruption which complicates as many as one in 120 pregnancies (Bourne, 1962). It has striking clinical similarities to the pre-bullous stage of herpes gestationis (HG). Indeed, it has been suggested that it may be a forme fruste of HG and that it may herald the development of