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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.
Correspondence

the UVB source, at the exposure dose used, an effect was obtained which was slightly greater than that for 1 J/cm² of UVA. However, this UVB dose would produce a severe sunburn reaction in the majority of normal subjects and the emission from the source contains significant amounts of UVA.

It is evident that UVA wavelengths, in significant amounts, penetrate the dermis where 20% of the cardiac output is directed. Because sunbeds and sunbathing may expose patients taking ciprofloxacin to more than 10 J of UVA in a day (UVA at noon in midsummer in Dundee is approximately 5 mW/cm², i.e. 18 J/cm²/h), it is possible that environmental exposure to these wavelengths may result in a reduced antibiotic effect of this drug. The preliminary results reported here are being followed up by precise monochromatic wavelength dependency studies, as well as pharmacokinetic work to determine their significance in vivo.

The photochemical change in ciprofloxacin may be monitored very simply by following the UV-induced change in its absorption spectrum or, with more sophistication, using high performance liquid chromatography (HPLC). Our own studies with HPLC have revealed a number of photoproducts. However, until these compounds have been identified and their own antibiotic potential determined, the bioassay remains the main indicator of UV-induced loss of therapeutic activity. The role of photoproducts, as potential local and systemic toxic agents, also requires further study. It may well be that simple avoidance of irradiation or use of appropriate photoprotection will minimise both the toxicity and the possible loss of therapeutic activity.

Department of Dermatology, and
*Department of Medical Microbiology,
University of Dundee
†Drug Development (Scotland) Ltd., Ninewells Hospital,
Dundee, U.K.

J. Ferguson
G. Phillips*
J. McEwan†
T. Moreland†
B. E. Johnson

REFERENCES

2 Kocherar IE. Possible mechanisms of toxicity due to photochemical products of protriptyline. Toxicol Appl Pharmacol 1980; 54: 258.

Etretinate-responsive pachyonychia congenita

MADAM, We were interested to read the recent correspondence regarding the treatment of pachyonychia congenita.¹,² Unlike Drs Soyuer and Candan¹ we have seen definite improvement in a patient with type I pachyonychia congenita following treatment with etretinate.

A 46-year-old man was first seen at St John's Hospital in 1964 at the age of 22. His finger nails and toenails had been long and thickened since infancy, the latter becoming wedge-shaped (Fig. 1). When 4 years old he was troubled by recurrent blistering and crust formation on the soles of his feet. This was followed by the gradual development of hyperkeratosis of the soles (Fig. 2), accompanied by profuse sweating and a pungent odour. His tongue later became white and fissured. Painful fissuring of the soles made walking very difficult without the aid of a walking stick. There was no family history of pachyonychia congenita.

A number of topical keratolytics were tried without success. In 1980, etretinate therapy was commenced at 25 mg/day (body weight 68 kg) and this was increased to 50 mg daily after 2 weeks. After 3 months of
Correspondence

FIGURE 1. Wedge-shaped nails in pachyonychia congenita.

FIGURE 2. Hyperkeratosis of the soles in pachyonychia congenita.

therapy comfortably for the first 3 weeks, followed by degenerative therapy for a further 3 weeks. Thus, our follow-up of 3 years would have required a total of 6 weeks of treatment.

St John's Hosp.
5 Lisle Street

1 Tidman M
2 Soyuer U

MADAM, In no cases did I advise the use of UVA lamps in the treatment of pachyonychia congenita. However, it is possible that the use of UVA lamps may lead to a reduction in the incidence of this condition. Further studies are needed to evaluate the long-term effects of UVA lamp therapy.

Regional N
Dryburn H
Durham D

1 Diffey B
1062: 665

MADAM, The reasons for selecting the UV lamp for Diffey's study were: the UV lamp was shown to be effective in the treatment of pachyonychia congenita. Thirdly, the example of the UV lamp's ability to induce a reduction in the incidence of this condition provides further evidence for its use.
Correspondence

therapy there was a reduction in the amount of hyperkeratosis on the soles, and he was able to walk comfortably without the use of his walking stick. After a further 2 months of treatment, he walked 5 miles for the first time in his life. Temporary discontinuation of the etretinate during the past 8 years has been followed by exacerbation of his symptoms. X-rays of the vertebral column have revealed minor degenerative changes only and he remains improved on etretinate.

Thus, our patient experienced marked symptomatic improvement with decreased hyperkeratosis following 3–5 months of etretinate therapy. It may be that the patient described by Drs Soyuer and Candan would have responded to more prolonged treatment with etretinate, had the drug been tolerated for more than 5 weeks.

St John’s Hospital for Diseases of the Skin
5 Lisle Street, London WC2H 7BJ, U.K.

F.CARABOTT
C.B.ARCHER
W.A.D.GRIFFITHS

REFERENCES


The suitability of sunglasses worn by PUVA patients

MADAM, In a recent paper (British Journal of Dermatology 1988; 118: 247), Dr Moseley and colleagues advised against the practice of checking the optical suitability of sunglasses worn by PUVA patients by using UVA radiation from the opened door of the PUVA cubicle and the departmental UVA meter. I examined this practice critically some time ago and concluded that this simple technique was unlikely to lead to a misleading impression concerning the ultraviolet opacity of the patients’ sunglasses in natural sunlight. Given that a spectrophotometer is not readily to hand in most PUVA departments, I commend this quick and simple checking procedure to staff as a worthwhile alternative to spectral transmission measurements.

Regional Medical Physics Department,
Dryburn Hospital,
Durham DH1 5TW, U.K.

B.L.DIFFEY

REFERENCES


Reply

MADAM, Dr Diffey raises an interesting point in his letter, which we are pleased to comment on. The reasons for our advising against dermatologists placing undue reliance on the procedure described by Dr Diffey are as follows: Firstly, the ‘transmission’ so measured will vary depending on the spectral emission of the UVA lamps and the spectral sensitivity of the UVA meter. Secondly, PUVA lamps usually have a peak emission at a wavelength of around 360 nm. However, we have suggested that shorter wavelengths are likely to be more important for PUVA-induced cataract from daylight exposure. If this is the case, then UVA lamps and a UVA meter would be measuring ‘transmission’ in the wrong part of the spectrum. Thirdly, we have examined eyewear where transmission peaks occurred within the UV region. For example, one pair had a transmission of 5% of 400 nm and 15% at 340 nm. A UVA lamp-UVA meter...