BJD
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## Re: Digital compression of facial arteries facilitates cutaneous nasal surgery

DOI: 10.1111/bjd.12464

Dear Editor, We read with interest the technique described recently by Moran *et al.*[1] for controlling intraoperative haemorrhage during cutaneous nasal surgery.

We would like to suggest a similar technique that we have found useful in our department for skin surgery in the temporal region.

The superficial temporal artery runs through the temporoparietal fascia, and supplies a wide region of soft tissue superficial to the temporal fascia via the frontal and parietal branches. The trunk of this artery can be found reliably by palpation anterior and superior to the tragus, superficial to the root of the zygomatic arch.

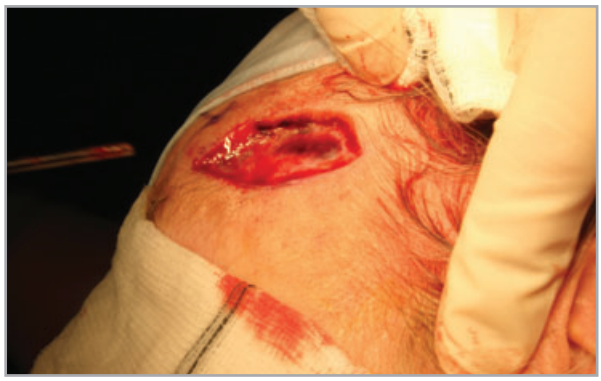

**Fig 1.** Firm digital pressure applied to the trunk of the artery facilitates haemostasis.

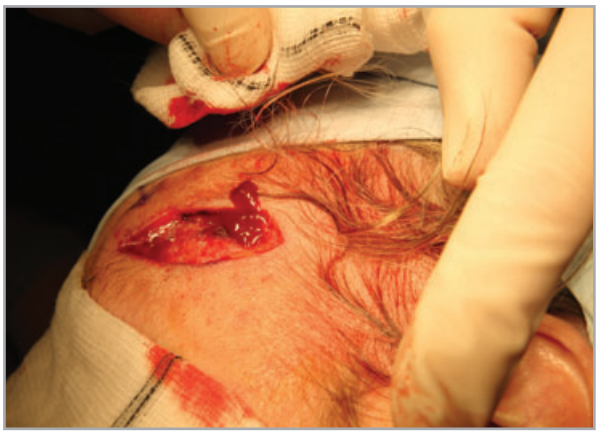

**Fig 2.** Release of pressure results in haemorrhage from the proximal wound margins.

Firm digital pressure at this point reduces bleeding from the proximal wound margin of excisions in the temporoparietal region. This can be achieved without discomfort to the patient (Figs 1 and 2) and facilitates haemorrhage control. A short video clip (Video S1; see Supporting Information) is included to illustrate the efficacy of this technique.

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1 Moran B, Foley C, Ormond P. Digital compression of facial arteries facilitates cutaneous nasal surgery. *Br J Dermatol* 2013; **169**:186–7.

### Supporting Information

Additional Supporting Information may be found in the online version of this article at the publisher's website:
**Video S1.** Digital pressure.

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Conflicts of interest: none declared.



## Resolution of the plantar hyperkeratosis of pachyonychia congenita during chemotherapy for Ewing sarcoma

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Dear Editor, A 15-year-old girl with pachyonychia congenita (PC) confirmed by genetic testing presented with pain and a localized subcutaneous swelling on her right back. Biopsy confirmed Ewing sarcoma of the posterior 10th rib. Preoperative tumour-reduction chemotherapy was commenced as per the Euro EWING 99 protocol, with vincristine, ifosfamide, doxo-

rubicin and etoposide.[1] Ifosfamide was subsequently replaced by cyclophosphamide, as per protocol, due to potential renal tubule toxicity (Table 1).[1] Surgical resection was performed followed by radiotherapy to the primary site.

Prior to chemotherapy, our patient had painful focal plantar keratoderma, in a distribution characteristic of PC, affecting the weight-bearing areas of both soles. Each area had a rim of surrounding erythema. Mild toenail dystrophy was present (Fig. 1a). The painful keratoderma had a significant impact on the girl's quality of life, with pain and impaired mobility. She had a known keratin 16 mutation: *KRT16* c.374A>G; K16 p.Asn125Ser. Previous treatment with keratolytic agents, including urea, salicylic acid and tar-based preparations, along with regular intense podiatric therapy, had resulted in very limited improvement. Six-monthly plantar botulinum toxin injections helped with pain and were ongoing prior to her illness.

A dramatic improvement in the keratoderma was noted following chemotherapy induction. Six weeks into chemotherapy, shedding of large areas of hyperkeratotic plaques commenced. A marked improvement was noted at 12 weeks (Figs 1b and 2), and this improvement continued at 16 weeks (Fig. 1c). There was virtually no residual keratoderma during maintenance chemotherapy and on completion of treatment. The clearance was maintained for a further 9 months following remission of the Ewing sarcoma and cessation of all chemotherapy (Fig. 1d), with minimal areas of keratoderma redeveloping at pressure sites after this.

The precise mechanism of this observed effect is not clear, and as far as we know this clinical observation has not been previously noted. The mechanisms may have some analogies to the better understood chemotherapy-induced alopecia (CIA). CIA is a frequent toxic side-effect of cytotoxic cancer therapy. It results as a direct toxic effect on the rapidly dividing cells of the hair follicle. A major characteristic of the anagen hair follicle is that the epithelial compartment undergoes proliferation, with the greatest proliferation activity at the bulb matrix cells building up the hair shaft. The abrupt cessation of mitotic activity leads to the weakening of the partially keratinized proximal portion of the hair shaft, with narrowing and subsequent breakage within the hair canal.[2] As a result, hair shedding begins at 1–3 weeks from anagen follicles (anagen effluvium), and is complete at 1–2 months after chemotherapy initiation. The frequency and severity of CIA differs among chemotherapeutic drug classes, with the commonest agents including topoisomerase agents such as doxorubicin and daunorubicin, antimicrotubule agents such as paclitaxel, and alkylators such as cyclophosphamide and ifosfamide.[3] CIA with permanent alopecia has been reported, particularly with taxanes.[4]

The epidermis of ridged or palmoplantar skin expresses a complex pattern of keratins due to the greater stress it must withstand. Specific expression patterns of keratins K6, K16 and K17 have been demonstrated in ridged skin, suggestive of regional adaptation patterns resulting in high proliferative activity of normal ridged skin.[5] Mutations in suprabasal keratins lead to hyperkeratosis of specific epithelia. Mutations in the site-specific keratins K6a, K6b, K6c, K16 or K17 lead to phenotypic variants of PC, with painful hyperkeratosis of the soles and other sites where these keratins are expressed, such as the nails and oral mucosa.[6] Thus, chemotherapy may improve hyperkeratosis primarily by an antiproliferative mechanism. The disease mechanisms in PC are thought to initiate with cell lysis (blistering, when lysed cells coalesce) due to dominant negative mutations in the above keratins impairing the structural mechanics of the cytoskeleton of the suprabasal cells in the relevant skin sites. This observed effect suggests that a hyperproliferative response may have a greater role in pachyonychia than previously recognized. The same hyperproliferative mechanism may also be important in other dominant negative keratin disorders such as keratinopathic ichthyosis. We recognize that other mechanisms could possibly explain this observed effect, for example bed rest may reduce

**Table 1** Chemotherapy regimen

| Week of regimen | Euro EWING 99 Protocol | |
|---|---|---|
| Week 12 (Figs 1b and 2, after cycle 4) | Cycles 1–4 | Vincristine, ifosfamide, doxorubicin, etoposide (VIDE) × 4 |
| Week 16 (Fig. 1c, after cycle 6) | Cycles 5–6 | Vincristine, cyclophosphamide, doxorubicin, etoposide (VCDE) × 2. Ifosfamide replaced by cyclophosphamide due to potential tubulopathy |
| | Surgical resection performed after cycle 6 | |
| | Cycles 7–15: maintenance cycles. Radiotherapy performed after cycle 8 | Vincristine, actinomycin, cyclophosphamide (VAC) × 8 |
| | Cumulative doses | Vincristine, ifosfamide 36 g $m^{-2}$, doxorubicin 360 mg $m^{-2}$, etoposide 2340 mg $m^{-2}$, cyclophosphamide 11·625 g $m^{-2}$ |
| Week 92 (Fig. 1d), 36 weeks after completion of chemotherapy | | |

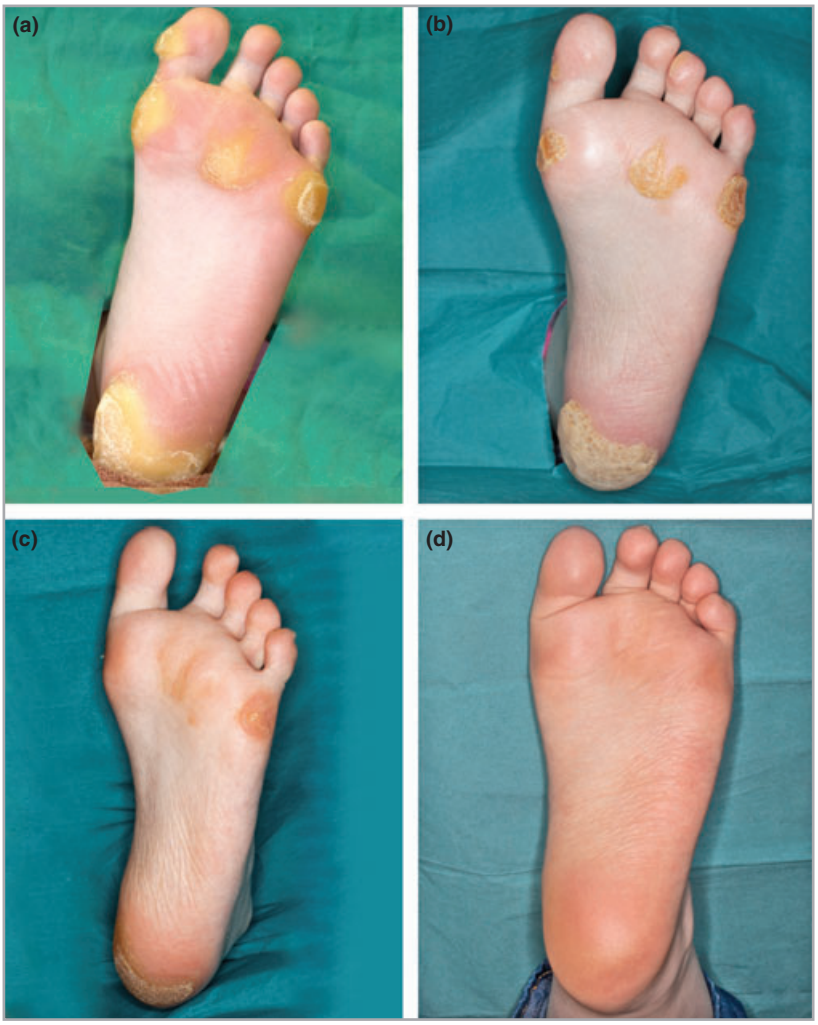


**Fig 1.** Clinical features of the focal keratoderma of pachyonychia congenita, (a) prior to treatment, (b) after 12 weeks, (c) after 16 weeks and (d) after 92 weeks (36 weeks after chemotherapy completion).

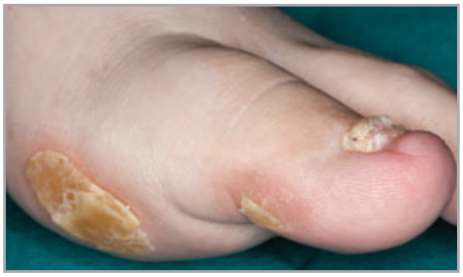

**Fig 2.** Close-up detail of hyperkeratotic skin shedding after 12 weeks of chemotherapy.

local stress and friction to hyperkeratotic areas; however, anecdotal data collected through the International Pachyonychia Congenita Registry (IPCRR) suggest that bed rest results in little improvement in most cases. Chemotherapy could also inhibit inflammatory cytokines implicated in pain pathogenesis and possibly hyperkeratosis.

While our patient experienced a dramatic resolution of her symptoms during chemotherapy, we are aware of other patients with PC on different chemotherapy regimens (for breast cancer) who have not experienced this dramatic benefit (IPCRR, unpublished data), so the response may be regimen specific. Clearly such a regimen is too toxic to be applied to patients with PC in the broader sense, but our hope is that this observation may stimulate thinking as to how to harness this positive effect in a safe and localized fashion.

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Conflicts of interest: none declared.



## Dermatology training in the U.K.: does it reflect the changing demographics of our population?

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Dear Editor, The U.K. has seen an increase in the proportion of the 'nonwhite' population by 61% between 2001 (8·7%) and 2011 (14%), with the proportion of black Africans doubling over this period.[1]

Studies have shown that among different ethnic and racial groups, cutaneous disorders may vary in both prevalence and clinical manifestation.[2] In light of this, as well as the observed changes in the U.K. demographics, it is important that dermatologists are adequately trained to diagnose and manage dermatoses pertinent to our ethnic population.

For this reason, we conducted a questionnaire-based study to ascertain whether the current U.K. dermatology training programme for specialist registrars provides adequate training in the diagnosis and management of diseases pertinent to the ethnic population.

We identified 217 dermatology trainees (specialty training level ST3–ST6, including those out of programme) and 77 junior consultants (within 2 years of receiving their Certificate of Completion of Specialist Training) from the database of the U.K. Joint Royal Colleges of Physicians Training Board (JRCPTB) as being eligible for inclusion into the study. A web link to our online questionnaire was sent to this target group via: (i) the monthly e-mail bulletin of the British Association of Dermatologists (BAD); (ii) letters posted to all National Health Service hospitals with a dermatology department in the U.K.; and (iii) an e-mail bulletin distributed twice to dermatology trainees via regional trainee representatives.

Questions focused on participant demographics and clinical and procedural experience pertaining to dermatoses pertinent to the U.K.'s ethnic population. Junior consultants were asked to complete the same questionnaire as trainees; however, their responses were regarding their previous dermatology training programme.

In total, 78/294 participants (26·5%) completed the survey – 65/78 were trainees (83%) and 13/78 were junior consultants (17%) – of whom 63/78 (81%) were female and 15/78 (19%) male. The mean age was 33·3 years (range 27–51 years). The largest proportion of respondents (24/78, 31%) were training/trained in London.

Only 3/78 (4%) recognized an expert conducting clinics specializing in 'ethnic dermatology' in their current or previous training programmes, and only 17/78 (22%) had formal teaching sessions/lectures focused in this field as part of their training programme (Table 1). Despite this, most respondents did cite clinical and procedural experience in relation to specific diseases pertinent to the U.K.'s ethnic population (Table 2), the exception being the management of central centrifugal cicatricial alopecia (CCCA). Over half (56%) had experience in managing this disease; however, only 27% had performed intralesional injections in the setting of CCCA. A significant number of respondents (74/78, 95%) acknowledged that individuals with 'ethnic skin' had unique and specific dermatological problems, and 55/78 (71%) thought that formal outcomes pertinent to this field should be incorporated into the training curriculum (Table 1).

Only 38/78 respondents (49%) thought they were/would become competent in treating conditions pertinent to the U.K.'s ethnic population following completion of their training programme, with the highest proportion of 'yes' responses coming from the West Midlands (88%) and London (71%). The latter regions correspond to the same two U.K. regions with the highest proportional nonwhite populations.[3]

A free text part of our questionnaire asked participants to identify dermatoses pertinent to the ethnic population, which were considered to be poorly understood and/or worst man-

 