

Atopy, the barrier, urine and genital lichen sclerosis

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DEAR EDITOR, From their interesting data Becker *et al.*¹ induce that 'AD [atopic skin diathesis] seems to be a priming precondition for the development of LS [lichen sclerosis] in boys' and say their 'data are consistent with...a permissive...effect of atopy facilitating the development of LS'. Not unreasonably, they continue 'genetic defects causing a lack of filaggrin have been reported in 50% of all AD cases, underlining the pathogenetic hypothesis that a disturbed atopic skin barrier is more susceptible to infectious and allergic and possibly LS-triggering agents' but then take a tendentious wrong-turning to conclude that 'further studies are needed to illuminate the underlying immunological mechanisms in both entities to clarify the linkage between AD and LS'. This erroneous logic or bias arises because they have chosen to ignore, or are unaware of, the burgeoning and compelling evidence that LS is due to chronic occluded exposure of susceptible epithelium to urine. To rehearse that evidence: males circumcised at birth never get LS unless there is a congenital anomaly (e.g. hypospadias); later in life LS occurs after genital piercing for jewellery and surgery; it complicates urethrostomies and urostomies; it recurs in grafts (especially skin grafts, rarely mucosal grafts); the distribution of LS mirrors occlusional contact with urine so the anatomical distribution of LS differs strikingly between the sexes, with perianal disease commonplace in the female and vanishingly rare in the male; men with LS manifest urinary microincontinence (they dribble) and if examined attentively are found to have anatomically variant naviculomeatal fossae.²⁻⁷ In contradistinction, the evidence for autoimmunity is unconvincing. Why just the anogenitalia? Why never the perianal skin in men? It's urine! And the barrier! My urinary contact hypothesis requires a susceptible epithelium. Becker *et al.*¹ inadvertently provide a welcome piece of the jigsaw, for which we should all be grateful.

A final point concerning the methodology. Negative or nonspecific foreskin histology does not exclude LS. It is very specific but not at all sensitive. This is because the foreskin is permissive for LS (which is never seen in males circumcised at birth, as above) but the prepuce may not always be the seat or site of the disease. It is likely that many of the boys circumcised in this study needed the operation because of LS, which must be the one of the commonest causes of phimosis in this age group,⁸ so the 'control' group is likely to contain patients with LS and thus the 'control' (non-LS) denominator is unreliable; some of these cases almost certainly had LS, inferring to me the possibility that, if more tightly controlled, the difference could be more significant than it appears.

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Atopy, the barrier, urine and genital lichen sclerosis: reply from the authors

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DEAR EDITOR, We thank Dr Bunker for adding another condition that plays a role in the pathogenesis of lichen sclerosis (LS).¹

Indeed the 'warm, moist, urine-exposed environment' was postulated as predisposing towards balanitis xerotica obliterans in the observation of Depasquale *et al.*² We refer to this theory in our review of 225 cases of LS in boys.³ We confirmed the observation of recurrence of LS after circumcision in a boy whose obesity had led to a pseudoprepuce caused by the phenomenon of buried penis. Therefore exposure to urine is probably a factor. There are a lot of studies and data in the literature indicating a relationship between autoimmunity and LS.⁴ To our understanding the theory of occlusion does not rule out an autoimmune process started by exposure of the susceptible epithelium to urine.

In boys we have the opportunity to see cases of LS much earlier.³ The histologically confirmed cases of the beginning of LS in clinically 'normal' congenital phimosis and the apparently better and longstanding success of topical corticoid treatment in 'early' LS (own unpublished data) suggest in our eyes that the very beginning of LS history is a dynamic

inflammatory process, which can either regress or progress into a sclerosing process. It is therefore questionable if these cases account for the group of LS or rather the control group.

Pilatz *et al.*⁵ investigated a group of patients with congenital phimosis and found no specific gene expression patterns in contrast to the LS group. This rules out that all persistent congenital phimosis is due to a subclinical inflammation driven by the beginning of LS.

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Novel management of harlequin syndrome with stellate ganglion block

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DEAR EDITOR, Harlequin syndrome is a rare condition in which one half of the face fails to flush and sweat due to damage of the sympathetic fibres on the ipsilateral side.¹ This produces compensatory increased flushing and sweating on the opposite side of the face, with a clear line of demarcation between the two halves. Paradoxically, patients present with the compensatory flushing and sweating of the unaffected, sympathetically innervated side of the face. Surgical sympathectomy has been used to prevent compensatory flushing and sweating, but it is not always successful and can be complicated by further compensatory flushing and sweating elsewhere.² We describe a novel approach to the management of harlequin syndrome with repeated stellate ganglion blocks (SGBs), which might be considered either a screen for sympathectomy, or a less invasive alternative treatment.

A 53-year-old woman presented with a 5-year history of flushing and sweating affecting the right side of her face and neck, provoked by heat and exercise (Fig. 1). This had a

significant impact on her affect, with increased anxiety and consequent social withdrawal. Emotional flushing and sweating were preserved bilaterally, suggesting a central aetiology, either in the pons or cervical cord. Previous medical history included left migrainous headaches and an anxious personality complicated by agoraphobia. Neurological examination was normal, apart from an isolated long-standing tonic right pupil. Magnetic resonance imaging (MRI) of the head, neck and chest, and plasma glucose were normal. Antitreponemal serology was negative. MRI of the cervical spine showed minor bulges at C3–C7. A diagnosis of idiopathic harlequin syndrome was made.

The patient was reviewed by a neurologist, who confirmed that the patient had an isolated tonic right pupil due to a congenital abnormality, unrelated to her acquired harlequin syndrome.

The patient was referred to the pain clinic for management. A right-sided C7 fluoroscopy-guided SGB was performed with 18 mL levobupivacaine 0.5% injected in increments, with rapid onset of sympathetic block. Following the procedure there was a marked improvement in the duration (reduced from 20 min to 5 min) and extent of her flushing, reflected in a significant improvement in her Dermatology Life Quality Index from 20 to 7.³ After the initial block her agoraphobia improved and she enjoyed a holiday in the tropics without an exacerbation of symptoms. It was possible to repeat the SGB at progressively increasing intervals of 2–6 months, hypothesized to be due to the membrane-stabilizing effect of local



Fig 1. Patient with harlequin syndrome demonstrating absence of flushing of the left side of the face and compensatory increased flushing of the right side following exercise.