but on examination of the marriage certificates of these two sibs the names of their parents were found to be identical, and their relationship was confirmed from the 1861 census when they were living in the same household and were recorded as brother and sister. Unfortunately, we have no documentary evidence that the brother was affected. Their mother (I.1, fig 2) was born in 1819 and can be quite confidently presumed to be a carrier whose abnormal X chromosome was inherited by these two children. Accordingly, the proband had not inherited his disease from his normal father's side of the family but from his carrier mother. This case shows the importance of taking a full family history of both sides of the family and of enquiring about consanguinity. We might have missed this diagnosis if the maternal grandmother had also been a carrier (with a normal, instead of an affected, husband), a possibility often forgotten in relation to X linked disease. Conversely, without the details available from Register House, we might have postulated some highly speculative mechanism whereby the proband inherited his X linked disease through an unaffected father.

In summary, we were greatly exercised to try to find an explanation for X linked retinosis in a male proband with a typical X linked pedigree on the paternal side, especially as his own father was unaffected (though a paternal uncle was blind). The solution to the problem was achieved by gentle persistence in history taking from the mother, who believed that her father who died when she was very young was blind, and that she had once been told that she was a distant relative of her husband. Tracing of both families in the public records system of Scotland established that the proband's paternal great great grandmother was a sister of the maternal great great grandfather, with X linked recessive blindness in descendants of both.

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CHILD naevus is not ILVEN

In a recent article, Moss and Burn¹ advanced the hypothesis that CHILD syndrome and ILVEN are "polar groups on a clinical spectrum, both reflecting an ectodermal defect variable in site and extent". They proposed the new descriptive term 'psoriasiform epidermal naevus (PEN)', sometimes associated with 'congenital ipsilateral limb defects (PENCIL)'.

For the following reasons, however, the equation 'CHILD naevus + ILVEN = PEN or PENCIL' is mistaken. The epidermal naevus associated with the CHILD syndrome is definitely not ILVEN but a distinct cutaneous entity that should be called 'CHILD naevus'.²

(1) CHILD naevus can be distinguished from ILVEN by the presence of yellow, wax-like scales, resulting in a distinctive 'ichthyosiform' appearance. (2) This naevus shows a tendency to non-linear arrangement, often involving one half of the trunk in a diffuse manner. By contrast, ILVEN is always linear. (3) This naevus displays a pronounced affinity for the body folds, or ptychotropism³; by contrast, ILVEN is not ptychotropic. (4) CHILD naevus causes no, or only minimal, pruritus whereas in patients affected with ILVEN itching often constitutes a serious problem. (5) CHILD naevus may show the histopathological features of 'verruciform xanthoma', a phenomenon characterised by abundant foamy histiocytes occupying the dermal papillae. Such

xanthomatous transformation has so far not been observed in ILVEN. (6) CHILD naevus is a well defined genetic entity inherited as an X linked dominant trait, constituting the cutaneous hallmark of the CHILD syndrome. It occurs almost exclusively in females because the underlying X linked mutation is lethal in male embryos. By contrast, the genetic basis of ILVEN is unclear and possibly heterogeneous.

Furthermore, I disagree with Drs Moss and Burn that CHILD should apply only to the extreme 'hemidysplastic' form of a variable condition. It is true that the term was originally suggested as an acronym for 'congenital hemidysplasia with ichthyosiform erythroderma and limb defects',³ but it has now become evident that the associated skin disease should be classified more appropriately as a naevus, and therefore the following modified interpretation of the acronym has been proposed: 'congenital hemidysplasia with ichthyosiform naevus and limb defects'.⁴

In conclusion, there are different epidermal naevi giving the impression of a psoriasiform skin lesion, and it seems unjustifiable to lump them together under the term 'psoriasiform epidermal naevus'.

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The variable clinical spectrum and mental prognosis of the acrocallosal syndrome

In the August 1990 issue of this journal we read with interest the paper 'How wide is the clinical spectrum of the acrocallosal syndrome? Report of a