

IN THIS ISSUE...

Comments on the Proposed Term Pleomorphic Ichthyosis

In this issue, Professor Anders Vahlquist (1) intends to introduce the category pleomorphic ichthyosis (PI) into the clinical classification of this group of cornification disorders on the basis of his extensive experience with affected adults displaying mild or moderate generalized phenotypes, linked to multiple gene mutations (2). Paediatric dermatologists would rather adopt a different view because the neonatal/early phenotype is usually more pronounced, and favour a more specific clinical separation of patients. The major conditions grouped under the umbrella term PI, namely ichthyosis prematurity syndrome (IPS) and self-healing collodion baby/self-improving collodion baby (SICB), have distinct neonatal presentations, as detailed in Professor Vahlquist's review. Our group has recently proposed giving IPS a more descriptive, dermatological name. In fact, we published our case under a different name because we were unaware of the neonatal phenotype observed by the Nordic authors (3), but the causative gene for IPS was recently identified and a new mutation confirmed in our case (4). Based on the photographs in Vahlquist's review and our report, this neonatal phenotype would be better termed "self-healing congenital verruciform hyperkeratosis" (SHCH). However, we all, as experts convening at the Sorèze meeting, accepted, after a lengthy debate, to retain, because of its history and striking meaning, the umbrella term ichthyosis as a descriptor for the entire group of heritable generalized early-onset cornification disorders, even if fish-like scales are not always apparent (2).

So, being exposed to the same patient at different periods of his/her life makes physicians happy, or not, with clinical classifications. Pediatric dermatologists are

more on the splitter side and adult dermatologist on the lumpier side, but we both need expert genetic testing. The development of gene chip techniques for specific groups of disorders such as the congenital ichthyoses would be welcome in this field because the current approach to diagnosis is tedious. However, our very simple clinical message would be: first, get informed about the neonatal phenotype. The parents may agree to show you just a photograph of their baby's hand at birth and this could be sufficient to make a diagnosis of mild collodion baby (5).

REFERENCES

1. Vahlquist A. Pleomorphic ichthyosis: A new name for a heterogeneous group of congenital ichthyoses characterized by phenotypic shifting and mild residual scaling. *Acta Derm Venereol* 2010; 90: 454–460.
2. Oji V, Tadani G, Akyiama M, Blanchet-Bardon C, Bodemer C, Bourrat E, et al. Revised nomenclature and classification of inherited ichthyosis: Results of the first Ichthyosis Consensus Conference in Sorèze 2009. *J Am Acad Dermatol* 2010 (in press).
3. Leauté-Labreze C, Boralevi F, Cony M, Maleville J, Lacombe D, Surlève-Bazeille JE, Taieb A. Self-healing congenital verruciform hyperkeratosis. *Am J Med Genet A* 2004; 130A: 303–306.
4. Morice-Picard F, Léauté-Labreze C, Décor A, Taieb A, Fischer J. A novel mutation in the Fatty Acid Transport Protein 4 Gene in a patient initially described as affected by self-healing congenital verruciform hyperkeratosis. *Am J Med Genet A* 2010 (in press).
5. Taieb A, Enjolras O, Vabres P, Wallach D, editors. *Dermatologie Néonatale*. Paris: Masson, 2009, p. 303.

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