Laboratório de Genética Humana, Universidade Federal do Paraná, Curitiba

# Dermatoglyphic Analysis in a Highly Mutilating Syndrome *

**E. A. Chautard, N. Freire-Maia**

SUMMARY

This note describes the dermatoglyphic disturbances verified in two patients with a very complex and highly mutilating syndrome. The parents of the affected individuals present, however, normal dermatoglyphics.

An apparently new syndrome was recently described (Freire-Maia et al, 1969, 1970; Freire-Maia, 1970) as composed of extensive bone deficiencies in the four extremities, scarcity of hair in the whole body surface, abnormal dentition, protruding and deformed ears, hypoplastic nipples and areolas, metabolic abnormalities, etc. This syndrome is not associated with any major chromosome abnormality. The hypothesis of an autosomal recessive gene, with variable expressivity, has been advanced to explain its etiology.

This note will present an analysis on the dermatoglyphics of the two investigated patients. Anatomical landmarks will not always be referred to, due to the impossibility of their precise recognition. For a knowledge of the skeleton of the remaining distal parts of the extremities of the affected individuals, see Figs. 1-4. Data will also be presented as regards dermatoglyphics of the parents.

AFFECTED SON. The right elongated distal appendage (Fig. 5) presents two areas with uncharacteristic lines (some of them interrupted) surrounded by nonridged skin. One of the fingers of the left hand (Fig. 6) shows a tented arch; the other shows a radial loop (RC 7). The remaining part of the palmar region without the normal flexion creases and with only one digital triradius. Near the most distal bracelet crease, there is a simple whorl with an atypical core.

AFFECTED DAUGHTER. The ball-like right appendage (Fig. 7) is characterized by interrupted and dissociated ridges. The terminal left appendage (Fig. 8) presents palm-like interrupted lines and a figure resembling a double whorl without triradii.

Two photographs of the patients are shown in Figs. 9 and 10.

* Research project sponsored by the National Research Council (Brazil), the Rockefeller Foundation, the Calouste Gulbenkian Foundation, the Research Council of the Federal University of Paraná, and WHO.

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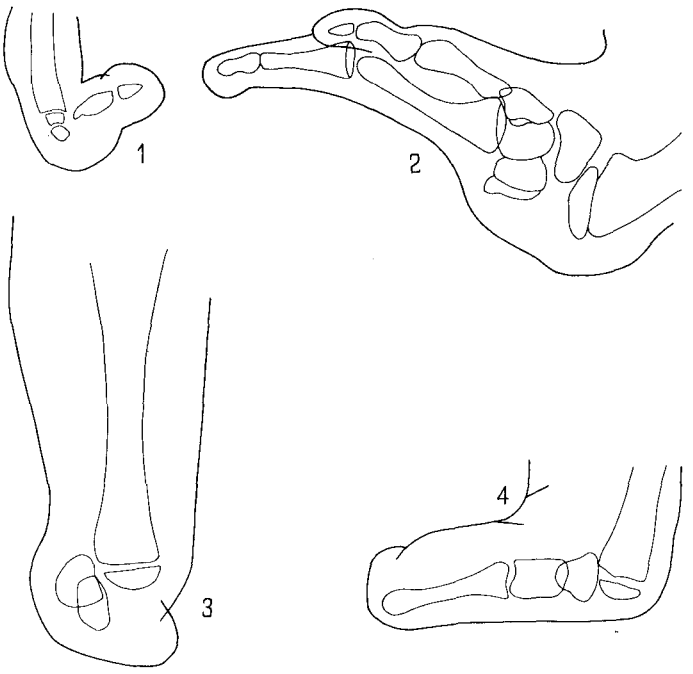
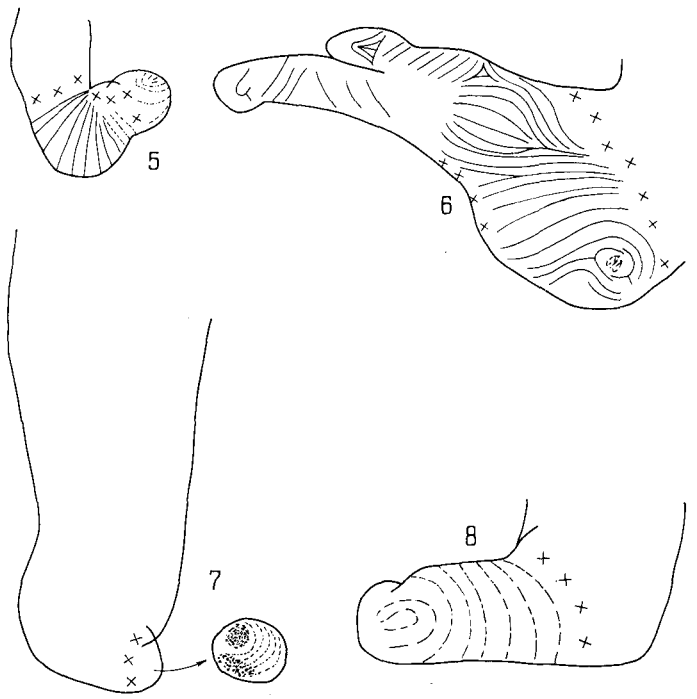
Acta Genet. Med. Gemellol. (1970), 19: 421-424




Figs. 1-4. Bone elements of the remaining distal parts of the upper extremities of the affected boy (1-2) and the affected girl (3-4). Figs. 1 and 3: right extremities; 2 and 4: left extremities




Figs. 5-8. Dermatoglyphics in the upper extremities of the patients. For a knowledge of the bone elements of these regions, cf. Figs. 1-4. The sign " × " indicates nonridged skin

https://doi.org/10.1017/S1120962300014797 Published online by Cambridge University Press

FATHER. The palmar configurations of the right hand present the following main-line formula: 7/10.X.5′ ′.5′ ′–t′–$A^u$.0.0.0.0. The axial triradius is displaced to a more distal position: *atd* angle of 52°; Walker ratio of 39.8%. On the left hand, the main-line formula is: 7.X.5′ ′.5′–t′–$A^u$/$A^c$.0.0.0.L/D. The axial triradius is also displaced to a more distal position: *atd* angle of 48°; Walker ratio of 32.0%. The

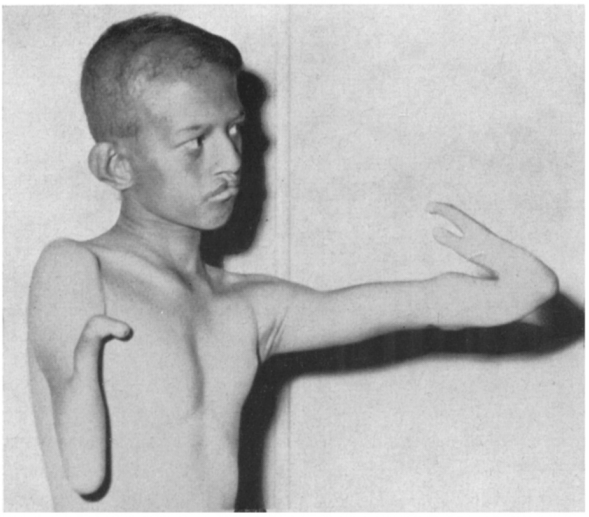


Fig. 9. The affected boy

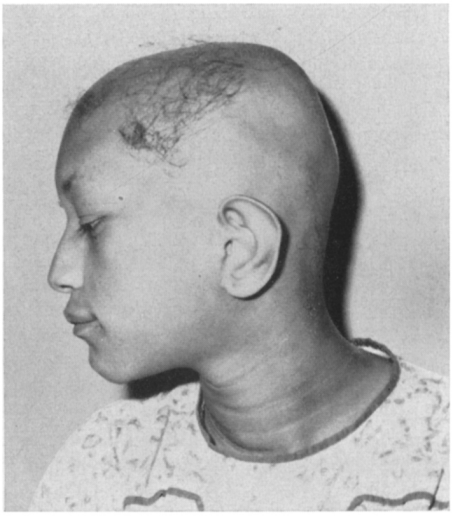


Fig. 10. The affected girl

fingerprint patterns are: right hand — four ulnar loops and one simple whorl; left hand — three ulnar loops, one simple whorl and one double whorl (TRC 135).

MOTHER. On the right palm, the main-line formula is: 11.9.7.5′–t–$L^r$.0.0.L.0. Walker ratio of 19.8%; *atd* angle of 44°. The main-line formula of the left palm is: 10.7.6.5′–t–$L^r$.0.0.0.L. In this hand, the *atd* angle is of 47° and the Walker ratio is 25.0%. The right fingerprint patterns are: two ulnar loops, two central pocket loops and one simple whorl. The first finger presents a large dissociation. The patterns of the left fingers are: four ulnar loops and one simple whorl (TRC 144).

The dermatoglyphic disturbances verified in the two patients are probably an effect of the process that led to the limb malformations; the parents of the patients show, however, normal dermatoglyphics on the basis of the data from a normal Brazilian population (Toledo et al, 1969).

*Acknowledgment.* We are very grateful to Dr. S. P. A. Toledo, for a critical reading of the manuscript.

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Riassunto

Vengono descritte le alterazioni dermatoglifiche in due pazienti affetti da una sindrome malformativa complessa. I genitori, tuttavia, presentano dermatoglifi normali.

Résumé

Les auteurs décrivent les altérations dermatoglyphiques chez deux patients atteints par un syndrome malformatif complexe. Leurs parents, toutefois, présentent des dermatoglyphes normaux.

Zusammenfassung

Beschreibung der Hautleistenveränderungen bei zwei Patienten mit schwierigem Missbildungssyndrom. Bei den Eltern waren die Hautleisten jedoch normal.

Eleidi Alice Chautard, M.Sc., Laboratório de Genética Humana, Universidade Federal do Paraná, Caixa Postal 756, Curitiba, Paraná, Brasil.