

CONGENITAL ONYCHODYSPLASIA OF THE INDEX FINGER IN A NEWBORN

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Case report. We present the case of a 1-month-and-15-day-old male newborn, the second child of his mother. Pregnancy was uneventful, with complete and normal prenatal checkups. He was born at 38.6 weeks of gestation via spontaneous vaginal delivery, weighing 4,286 g and measuring 51 cm in length. The mother denied taking any medications during pregnancy. Since birth, the patient had two small nail plates separated by normal skin on the second toe of the right foot, consistent with polyonychia (Fig. 1). No other abnormalities of the skin, hair, or sweating were noted. There was no family history of similar findings in the mother, father, or sibling.

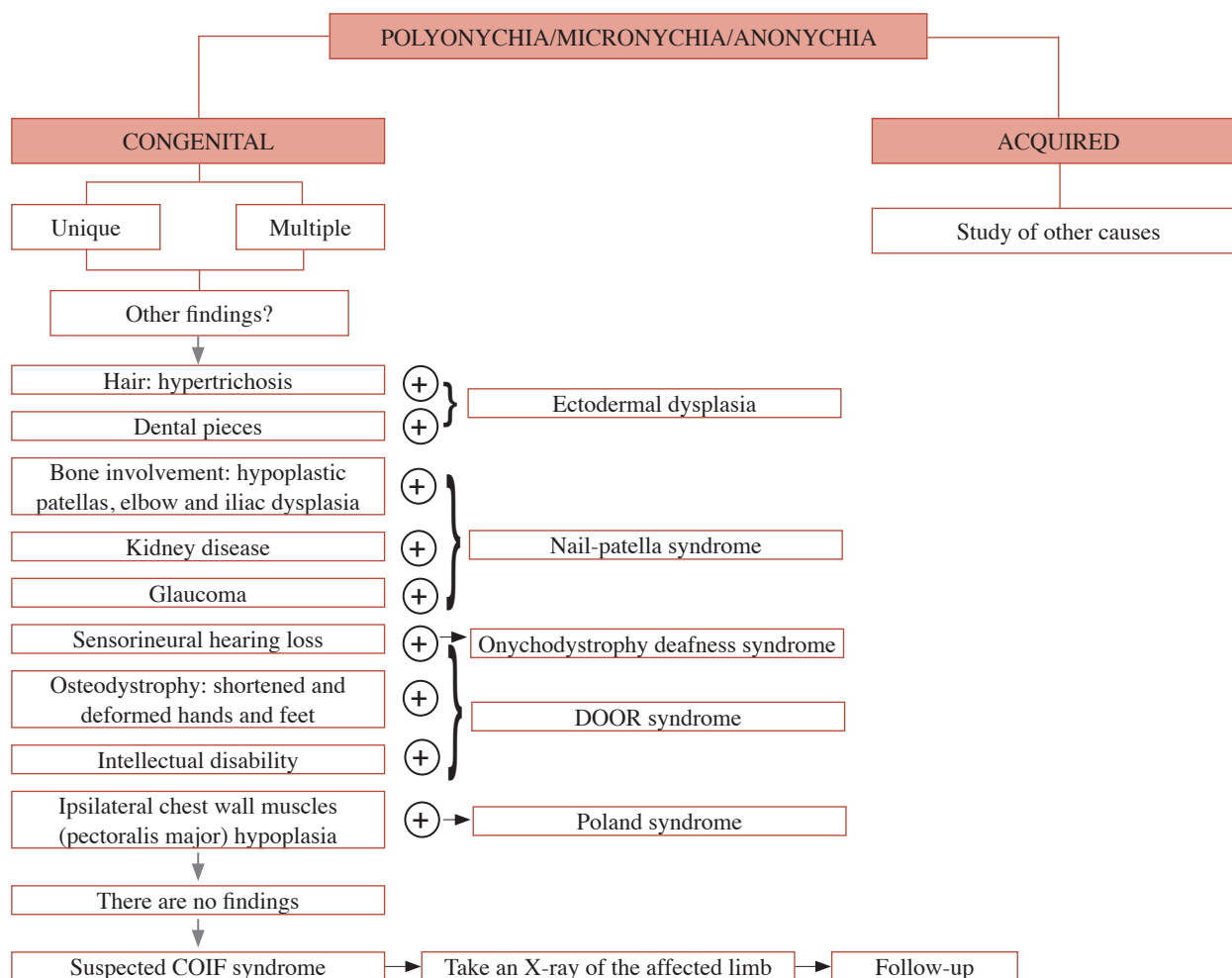


Fig. 1: Congenital onychodysplasia of the index finger.

Discussion. Congenital onychodysplasia of the index finger (COIF) is a congenital nail anomaly characterized by micronychia, polyonychia, anonychia, and irregular lunulae. It mainly affects the index finger, while involvement of the toes is rare. It was first described by Iso in 1969 and Kikuchi in 1974. The majority of reported cases come from Japan (1). It affects males and females equally, with no known racial predisposition (2).

The most common clinical manifestations include micronychia – typically on the ulnar side (3, 4) –, polyonychia (central anonychia with two lateral nail plates), anonychia, irregular lunulae, malalignment, and hemi-onychogryphosis.

Additional anomalies may include cutaneous or bony syndactyly, Y-shaped bifurcation of the distal phalanx, brachymetacarpia of the fifth digit, and limitation of distal interphalangeal joint movement (1, 2).



Although the pathogenesis remains debated, several hypotheses have been proposed (5, 6): intra-uterine exposure to teratogens (e.g., antiepileptics, antidepressants, abortifacients, insulin), intrauterine ischemic events (e.g., compression of the radial artery), hereditary transmission – autosomal dominant with variable expressivity (6) –, mutations in the Wnt signaling pathway (7), and associations with hepatic diseases such as hepatitis B (6).

The diagnosis is clinical. Baran and Stroud's diagnostic criteria (8) include: unilateral (42%) or bilateral (58%) hypoplasia of the index finger, presence at birth, onychodysplasia (micronychia, poly-onychia, etc.), radiographic abnormalities, and heredity (not mandatory for diagnosis).

Differential diagnosis should be made with other conditions, as outlined in the algorithm in Fig. 1. Nail-patella syndrome is characterized, in addition to micronychia or anonychia and triangular lunulae, by bilateral involvement, thumb abnormalities, hypoplastic or absent patellae, and renal disease. In cases of onychodystrophy associated with neurosensory hearing loss, nail-deafness syndrome should be considered. Toe nails may also appear dystrophic in the common condition of hypertrophy of the lateral nail folds, which usually resolves within months. Ectodermal dysplasia can be distinguished by the involvement of multiple nails along with hair abnormalities, sweating defects, and keratoderma. Pachyonychia congenita differs by progressive widening and thickening of the nail bed in multiple digits. DOOR syndrome presents with onychodystrophy along with deafness, osteodystrophy, and in-

tellectual disability, as summarized in the acronym DOOR. Poland syndrome is characterized by hand anomalies and absence or hypoplasia of the pectoralis major muscle.

Radiographs may be useful for assessing bony involvement. Management is conservative, and surgical intervention is generally not recommended (9).

Conclusion. This case of congenital onychodysplasia of the index finger was reported for its rarity and to highlight the importance of its differential diagnosis from complex syndromes.

Conflicts of interest

The authors declare that they have no conflicts of interest.

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