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Congenital Malalignment of the Great Toenails: A Review

Piera Catalfo Maria Letizia Musumeci Francesco Lacarrubba Franco Dinotta Giuseppe Micali

Dermatology Clinic, University of Catania, Catania, Italy

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Nail · Dystrophy · Congenital malalignment · Great toenail

Abstract
Congenital malalignment of the great toenail is an underestimated dystrophic disorder of unknown origin characterized by lateral deviation of the nail plates, which are not parallel to the major axis of the distal phalanx. It usually presents in infancy or childhood, while late onset is uncommon. Treatment depends on the degree of deviation. If minimal, a conservative and expectant attitude, based on prevention and treatment of possible complications, is recommended because of the possibility of spontaneous regression of the nail deviation. Surgical therapy may be considered in patients with severe or complicated forms.

Introduction

Nail dystrophy is a general term referring to altered or disturbed nail growth due to a wide range of conditions that may affect the matrix and/or bed; among them, congenital or acquired (traumatic, iatrogenic) nail deviation with consequent malalignment of the nails may be responsible for nail dystrophy [1]. Congenital malalignment usually involves great toenails, even if it may rarely involve other toenails [2]. Congenital malalignment of the great toenail is a dystrophic disorder characterized by lateral displacement of the nail apparatus, resulting in altered growth of the nail plate that is not directed along the major axis of the distal phalanx [2, 3] (Fig. 1, 2). It usually presents in infancy or childhood, while late onset is uncommon; it may not be identified until the child is older or develops related complications caused by repeated microtraumas to the toenail. A review of the existing literature on congenital malalignment of the great toenails is presented and discussed.

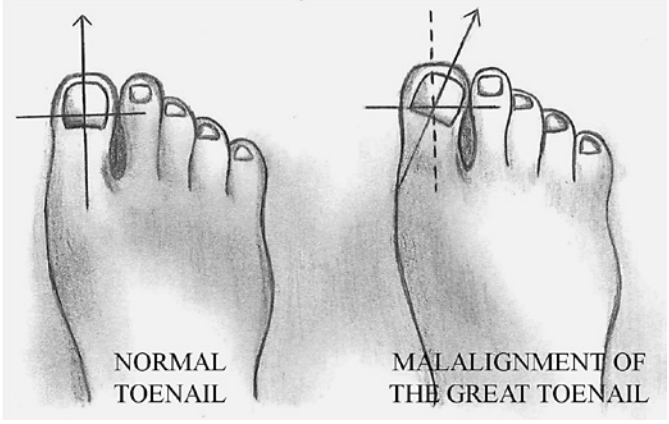

Fig. 1. Graphic rendering of a toenail affected by malalignment compared to a normal toenail.

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E-Mail karger@karger.com
www.karger.com/sad

Giuseppe Micali, MD
Dermatology Clinic, University of Catania
A.O.U. Policlinico-Vittorio Emanuele, Via Santa Sofia, 78
IT–95123 Catania (Italy)
E-Mail cldermct@gmail.com

Methods

Articles for this review were identified through searches in PubMed for papers published from 1950 to 2016 using the following specific search terms: *nail malalignment* OR *congenital malalignment* OR (*great toenail* AND *dystrophy*). The electronic searches were supplemented with hand searches of references in related articles identified in PubMed citations. Articles published in English, Italian, French, and German were considered.

All pertinent case reports and case series were included and are summarized in this review.

Results

From 1978 to 2016, 24 reports were published [2–25]. Among these, 1 report lacked clinical and epidemiological data [4], another report dealing with 30 patients lacked details about gender, location, and family history [5], and 2 reports, dealing with the same kindred, focused exclusively on inheritance with incomplete clinical details [6, 7], which is why they have not been included in the present review.

In the 20 selected articles, a total of 33 patients were described [2, 3, 8–25] (Table 1). The age at onset was at birth in 25 cases [2, 9, 10–13, 15–18, 21, 22, 24, 25], during the first 2 years of life in 2 cases [2, 20], and later in life in 2 cases [19], whereas it was not specified in 4 cases [3, 8, 14, 23]. Familial occurrence was confirmed in 5 cases [10, 12, 14, 18], likely in 2 cases [11], and negative in 9 cases [2, 13, 15, 16, 25], whereas in the remaining ones it was not specified [3, 8, 9, 17, 19–24]. Three sets of monozygotic twins [11, 21], 2 of dizygotic twins [12, 16], and 2 identical twins of triplets [13] were reported. Considering those cases for which the gender was specified, 13 females were affected compared to 11 males (female-to-male ratio 1.1:1).

Clinically, congenital malalignment of the toenail was usually characterized by lateral deviation of the nail plates, which were not parallel to the distal phalanx, as seen in 30 cases [2, 3, 8, 10–17, 19–25, 28]; medial deviation has rarely been observed, with only 3 cases reported [9]. As regards the localization, the disorder more commonly and synchronously involved both great toenails, as ob-

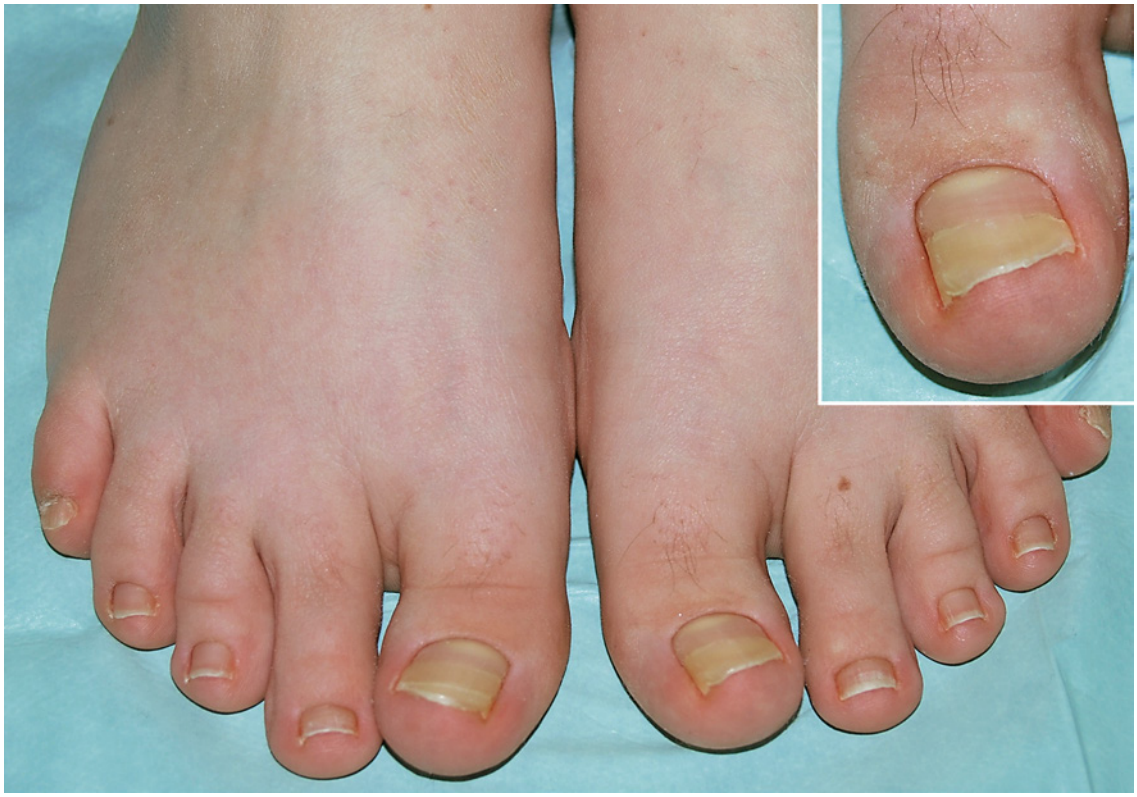

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Fig. 2. Lateral deviation of great toenail plates in a 14-year-old girl. Nail thickening, transverse ridges, and partial onycholysis may also be observed. Note the regular alignment of the other toenails.

Table 1. Cases of congenital malalignment of the great toenail reported in the literature

Author(s) [Ref.], year	Patients	Age	Age at onset	Gender	Family history	Location: bilateral (B) unilateral (U)	Treatment/ follow-up
Baran et al. [3], 1979	1	2 years	NS	NS	NS	B	Surgical intervention; no recurrence after 18 months
Harper and Beer [13], 1986	2 (identical twins of triplets)	Newborn	At birth in both cases	F F	Negative	B B	NS
Barth et al. [21], 1986	4 (two sets of monozygotic twins)	3 years 8 years	At birth in all cases	NS	NS	B B	NS
Baden and Mizner [22], 2003	1	20 months	At birth	NS	NS	B	NS
Baran and Bureau [9], 1987	3	Newborn	At birth in all cases	NS	NS	NS (medial deviation)	NS
Handfield-Jones and Harman [24], 1988	2	19 months 20 months	At birth in both cases	M F	NS	B B	Spontaneous normalization at 5 years in both cases
Cohen et al. [14], 1991	1	6 years	NS	M	Father affected	B	NS
Cohen [23], 1991	1	10 years	NS	M	NS	B	NS
Dyall-Smith [10], 1996	1	5 years	At birth	M	Brother with congenital thumbnail dystrophy	B	Conservative approach
Balci et al. [15], 2004	1	15 years	At birth	M	Negative	B	Cauterization; no recurrence after 2 years
Perlis and Telang [8], 2005	1	2 years	NS	F	NS	B	NS
Kus et al. [16], 2005	2 (dizygotic twins)	19 months	At birth in both cases	M F	Negative	B B	NS
Özdemir et al. [11], 2005	2 (monozygotic twins)	3 years	At birth in both cases	F F	Mother and aunt possibly affected	B B	NS
Chaniotakis et al. [12], 2007	2 (dizygotic twins)	1.5 months	At birth in both cases	F M	Father and grandfather affected	B B	Spontaneous improvement at 10 months in both cases
Senayli et al. [17], 2008	1	2.5 years	At birth	M	NS	B	NS
Wagner and Sachse [2], 2012	2	2.5 years 6 years	18 months At birth	M F	Negative	B B	Conservative approach; no changes after 1 year in both cases

Table 1 (continued)

Author(s) [Ref.], year	Patients	Age	Age at onset	Gender	Family history	Location: bilateral (B) unilateral (U)	Treatment/ follow-up
Batalla and Curto [25], 2014	2	3 years 5 years	At birth in both cases	F M	Negative	U B	Conservative approach
Decker et al. [19], 2016	2	21 years 20 years	15 years 16 years	F F	NS	B B	No intervention in both cases; no follow-up
Lipner and Scher [20], 2016	1	12 years	Early childhood	F	NS	B	Nail avulsion; no follow-up
Çayırlı et al. [18], 2016	1	6 months	At birth	M	Brother and sister affected	B	Spontaneous improvement after 6 months in both cases

NS, not specified.

served in 29 cases [2, 3, 8, 10–25]; in 1 case, one toenail only was affected [25], and in 3 cases the localization was not specified [9].

The severity of congenital malalignment of the great toenail was variable, ranging from mild to severe. As described in Table 2, nail plate thickening [8–12, 14, 16, 19, 22–25], discoloration ranging from brownish-black to green-brown and yellow [2, 8, 10, 11, 14, 16, 18, 19, 25], and transversal grooves or ridges [2, 8, 10, 11, 16, 23–25] were the most frequently observed abnormalities. The involved nails were sometimes shorter and trapezoidal in shape [2, 22, 23]. The distal nail plate was usually pointed [2, 25] or sometimes curved [2, 23]. Onycholysis [8, 14, 25], acute or chronic paronychia [11, 15, 18, 20], and ingrown nails [10, 11, 15, 16, 18] were other frequently reported features. Persistence of the condition usually resulted in severe nail dystrophy and nail plate thickening, which might sometimes cause complete [21] or partial onychogryphosis [10]. An unusual complication is the development of a firm, tender, noninflammatory nodule in the lateral part of the proximal nail region, possibly as a result of misplacement and/or cystic transformation of matrix portions [2, 18]. Bacterial and fungal infections were reported in some cases.

No associated findings have been reported in patients with congenital malalignment, with the exception of one patient showing ocular melanocytosis [17] and another one with Rubinstein-Taybi syndrome [15], but this occurrence was considered to be coincidental.

Follow-up data are generally not reported in the articles. When performed, the period of observation was short, ranging from 6 months to 2 years. Only 1 study reported a long-term follow-up of 5 years for 2 children in whom spontaneous improvement occurred [24].

Discussion

Congenital malalignment of the great toenails may be observed in 1–2% of children [8], although we believe along with other authors that this entity is probably underdiagnosed. The cause of malalignment is still a matter of debate. Genetic factors, i.e., hereditary embryological abnormalities, and extrinsic factors have been considered [25]. The occurrence of nail dystrophy in monozygotic and dizygotic twins and in several generations of a single family seems to suggest a role of genetic factors, and a possible autosomal dominant transmission with variable expressivity has been hypothesized [11–13, 16, 21]. Extrinsic factors include constriction of the toes by amniotic bands, increased intrauterine pressure, or vascular abnormalities during fetal life [22]. It has also been suggested that a key role in the pathogenesis of toenail malalignment is played by an increased tension of the extensor tendon of the hallux, which, by pulling the lat-

Table 2. Clinical features of great toenails affected by malalignment reported in the literature

Nail abnormalities	Patients, *n* (%)	Ref.
Discoloration		
Brownish-black	2 (6)	14[a–c], 16
Green-black	1 (3)	10
Green to yellow-brown and reddish	1 (3)	25[a, b]
Brown-green	1 (3)	18
Yellow	2 (6)	8[c], 19[a]
Blue-green	1 (3)	19[a, b]
Gray-brown	2 (6)	11[a], 2[b]
Nail plate thickening	12 (36)	8–12, 14, 16, 19, 22–25
Transversal grooves or ridges	11 (33)	2, 8, 10, 11, 16, 23–25
Onycholysis	4 (12)	8, 14, 25
Paronychia	5 (15)	11, 15, 18, 20
Ingrown nail	5 (15)	10, 11, 15, 16, 18
Shorter and trapezoidal shape	4 (12)	2, 22, 23
Narrow, curved shape	3 (9)	2, 23
Pointed shape	2 (6)	2, 25
Beau's lines	2 (6)	19
Lack of luster	2 (6)	2, 8
Onychogryphosis	2 (6)	21
Onychodystrophy	2 (6)	14, 19
Hemionychogryphosis	1 (3)	10
Tender nodule	1 (3)	2
Circumscribed leukonychia	1 (3)	2

[a] Fungal infection. [b] Bacterial infection. [c] Subungual hemorrhage.

eral portion of the matrix proximally, may cause a lateral rotation of the nail matrix, and, hence, a longitudinal displacement of the nail plate axis [12]. A recent hypothesis suggests that the pathogenesis may be due to desynchronization in growth between the nail and the distal phalanx of the hallux, resulting in larger nail plates, which must grow laterally to fit into the underlying bony space [12, 19]. It is also possible that the dystrophy is due to the genetically determined malalignment of the toenails plus superimposed cumulative effects of postnatal trauma [21, 26].

Generally the condition is present at birth, but, in case of minimal deviation, it may go unnoticed until childhood or puberty, when nail dystrophic changes occur as a result of mechanical stress, due to wearing tight shoes or to activities such as dancing or active sports [2, 19, 25]. As very few data on long-term follow-up are available, the exact percentage of spontaneous realignment is unknown, although it has been suggested to occur in about one-half of the patients before the age of 10 years [25].

A correct diagnosis is required to prevent errors and unnecessary treatments. The differential diagnosis includes onychomycosis, connective tissue disorders, nail apparatus tumors, and dermatoses with nail involvement (e.g., psoriasis, lichen, and ectodermal dysplasia) [8].

Treatment depends on the degree of deviation. As spontaneous realignment may occur up to 10 years of age, a conservative and expectant attitude, based on prevention (e.g., wearing comfortable footwear) and treatment of complications, is recommended [2, 12, 14, 19, 25, 27]. Surgical therapy may be considered, until up to 2 years of age, in patients with severe deviation of the nail plate or early complications, such as recurrent episodes of acute paronychia and/or infections [2, 9, 11, 15, 16, 19, 26]. Rotation of the whole nail unit, allowing the nail plate to grow in parallel to the distal phalanx, represents the main treatment [2, 11, 15, 26]. Nail matrixectomy, which involves the complete removal of the nail plate and matrix, leads to complete resolution of symptoms, but may not be cosmetically acceptable [19]. Surgical elongation of the extensor tendon of the phalanx has also been suggested [1, 9].

Statement of Ethics

The present research complies with the guidelines for human studies. The subjects have given their informed consent.

Disclosure Statement

The authors have no conflicts of interest to disclose.

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