Congenital Malalignment of the Great Toenail, the Disappearing Nail Bed, and Distal Phalanx Deviation: A Review

Benjamin Butters^a Steven Gates Scott^a Danielle Glinka^a C. Ralph Daniel^b
Robert T. Brodell^b Mark A. Braswell^c

^aAlabama College of Osteopathic Medicine, Dothan, AL, USA; ^bUniversity of Mississippi Medical Center, Jackson, MS, USA; ^cScott AFB, Belleville, IL, USA

Keywords

Congenital malalignment of the great toenail ·
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Abstract

Importance: Congenital malalignment of the great toenail (CMGT) is an idiopathic deviation of the nail apparatus. CMGT predisposes patients to recurrent stress forces, microtrauma, and secondary complications. The purpose of this study was to review the current published photographs to determine the relationship between variants of CMGT and the disappearing nail bed (DNB). **Observations:** A search in PubMed and Google using the terms congenital malalignment of the great toenail, disappearing nail bed, and lateral nail deviation was performed. Of the 53 photographs found in a total of 35 articles, 23 were disqualified due to low picture resolution or poor angle. The remaining 30 photographs were evaluated. Pure nail malalignment with associated dystrophy and DNB was found in 22 of 30 photographs. Four of 30 cases demonstrated pure deviation of the distal phalanx, with nail dystrophy but minimal DNB. The remaining 4 cases demonstrated a combination of toe deviation and nail unit deviation with varying degrees of DNB. **Conclusions and**

Relevance: DNB was associated with all forms of pure CMGT. Moreover, a variant of malalignment of the distal phalanx was noted in 8 photographs. This has potential implications for further studies and treatment to correct secondary complications.

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Introduction

Congenital malalignment of the great toenail (CMGT) is an idiopathic lateral deviation of the nail apparatus. The displacement of the nail unit may result in recurrent microtrauma leading to various nail pathologies including retronychia, paronychia, onycholysis, onychomadesis, and onychomycosis [1–3]. CMGT was first reported in the 1970s [4, 5]. There may be a similar pathophysiologic basis to nail dystrophy associated with hallux valgus (malalignment of the MTP) and CMGT associated with the lateral displacement of the distal digit (see Fig. 1a–c). One of the end results of chronic onycholysis in CMGT is epithelization of part of the nail bed. This has been termed the disappearing nail bed (DNB) [6].

Fig. 1. Illustrations of classic CMGT (**a**), MDPGT associated with lateral displacement of the distal phalanx (**b**), and hallux valgus (**c**) demonstrate anatomic deformities leading to nails that are not oriented parallel to the foot. CMGT, congenital malalignment of the great toe; MDPGT, malaligned distal phalanx of the great toe.

Fig. 2. Forces applied to CMGT (**a**) and CMGT associated with lateral displacement of the distal phalanx (**b**) lead to torque that damages the matrix and nail bed that would not be present in normally oriented toenails. CMGT, congenital malalignment of the great toe.

Nail pathologies are often difficult to treat due to the requirement of long-term patient compliance and recurrent macro- or microtrauma. Further insight into CMGT concerning treatment and clinical presentation provides the possibility of offering the best possible solutions for patients. One of this article's main goals is to better understand the pathophysiology of CMGT. This may lead to more efficacious forms of treatment. Certainly successful resolution in some cases has been documented. A review of all published cases of CMGT was performed to assess its association with underlying lateral displacement of the distal phalanx, nail dystrophy, and DNB.

Methods

A search in PubMed and Google for reports up to January 31, 2021, was (with no restriction on earliest search date) performed by the authors. Search terms used were congenital malalignment of the great toenail, disappearing nail bed, and lateral nail deviation.

Thirty-five articles reporting or reviewing cases of CMGT from 1978 to 2019 were identified. All cases with clinical photographs published were included in this review [1, 2, 4, 7–34]. Duplicate images were not included. When multiple photographs of the same patient were available, the image that most clearly demonstrated features of CMGT was used in the assessment. Photographs were analyzed for the presence of CMGT, lateral deviation at the interphalangeal joint of the great toe, nail dystrophy, and DNB.

Fifty-three clinical photographs were included in the final set for review separately by each author. Of these photographs, 23 were disqualified because of the angle of the image which made it difficult to assess or the image demonstrated poor resolution. Of the remaining photographs, the digits were evaluated for nail alignment, toe alignment, nail dystrophy, and DNB.

Discussion/Observations

Nail unit malalignment in the setting of a straight digit was identified in 22 of 30 (73%) photographs. In each case, the CMGT was associated with nail dystrophy and a DNB. Of the 8 remaining images, 4 cases (13%) demonstrated only deviation of the distal phalanx, with no DNB associated. The remaining 4 cases (13%) demonstrated a combination of toe deviation and nail unit deviation, with varying levels of DNB being seen with each. Radiologic images were not present in any of the cases which would have been the best way to confirm bone deviation.

Pathophysiology

CMGT is an idiopathic lateral deviation of the nail apparatus. It is most often a congenital/inherited disorder. The displacement of the nail unit increases risks of recurrent microtrauma leading to various nail pathologies including retronychia, paronychia, onycholysis, onychomadesis, and onychomycosis [1–3]. The microtrauma of the mispositioned nails produces torque that damages the nail matrix and nail bed. There may be a similar pathophysiologic basis to nail dystrophy associated with hallux valgus (malalignment of the MTP) and CMGT associated with either the lateral displacement of the distal digit or lateral deviation of the toenail in the setting of a straight digit (see Fig. 1). Walking, crawling, and trauma from footwear impacting a malaligned toenail create torsion stress forces that would not be present in a normally positioned nail (see Fig. 2).

Clinical Presentation

Patients with CMGT dystrophy often present with dystrophic nails related to recurrent retronychia, paronychia, onychomycosis, or onychomadesis. One of the end results of chronic onycholysis in CMGT is the DNB [6]. DNB is a relatively new concept, with the first article being published in 2005 and the second being published in 2017. It has been seen in chronic cases of onychomycosis and advancing age [6, 35]. In this study, DNB was identified in all presumably due to the same mechanisms noted above. While this study demonstrated that the majority of patients have straight toes, deviation of the distal phalanx was present in 4 cases (13%) which has not been noted previously.

Assessment and Diagnosis

In patients with nail dystrophy of the great toenail, the alignment of the great toenail and the digit itself should be evaluated. Physical evaluation of the misalignment of the nail unit in the setting of a normally aligned digit delineates a diagnosis of classic CMGT. The distal digit is laterally displaced, and the nail is growing parallel with the bone in a variant best termed MDP of the great toe. The presence of DNB in a patient, though highly sensitive in the set of photographs studied, is not specific to CMGT.

Treatment and Prognosis

Treatment options should focus on reducing the mechanical stress impacting the nail by wearing well-fitted footwear, trimming at-risk nails, and even surgical removal or realignment of the toenail [36]. In some cases, CMGT may spontaneously resolve, but many patients will continue to have continuing complications such as pain with ambulation [21].

Further Discussion

The Nail Unit or the Digit

This review of all published photographs demonstrated a new variant of CMGT that has not been previously recognized. In 4 of 30 patients, the toenail is properly oriented parallel to the axis of the toe, but the distal phalanx itself is malaligned, and the nail is therefore deviated by proxy. MDP leads to the same abnormal forces on the nail and the same onycholysis, DNB, and nail dystrophy. In fact, the same forces would seemingly be present on the nail in hallux valgus where the entire digit is laterally displaced.

In 4 additional patients, there was a combination of both lateral displacement of the distal phalanx and nail unit malalignment. Further research should focus on these 2 subsets of CMGT to determine their frequency and the severity of nail changes in each situation.

Therefore, when considering treatment, especially with pure phalangeal deviation, alignment of the distal toe may prove helpful in the manner bunion surgery can be performed to realign hallux valgus [37]. In theory, this procedure might minimize the trauma that produces nail dystrophy. This should be studied further as more cases of distal phalangeal deviation are recognized and evaluated clinically.

Limitations

This study was limited by the wide variation in quality of photographs in publications describing CMGT. Furthermore, many of the clinical pictures were taken at angles that made it difficult to determine orientation of the distal interphalangeal joint. Radiologic images to confirm the joint alignment were not available in any published case. Gathering a series of cases of CMGT with radiologic imaging and careful photography may well lead to a better understanding of the pathophysiologic basis of CMGT, the DNB, and the presence of MDP.

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Conflict of Interest Statement

Benjamin Buttars, Gates Scott, and Danielle Glinka have no relevant financial conflicts of interest to disclose. Robert Brodell is a principal investigator for a clinical trial (Novartis) and for the Corevitas psoriasis biologic registry. He serves on editorial boards of *American Medical Student Research Journal* (faculty advisor); *Practice Update Dermatology* (Editor-in-Chief); *Journal of the American Academy of Dermatology* (Associate Editor); *Practical Dermatology*; *Journal of the Mississippi State Medical Society*; *SKIN: The Journal of Cutaneous Medicine*; and *Archives of Dermatological Research* and is a staff physician at the GV “Sonny” Montgomery Veterans Administration Hospital in Jackson, MS, USA. C Ralph Daniel serves on an advisory board for Ortho Pharmaceutical and has an ownership interest in Medimetrics. None of these disclosures are relevant to the subject matter of this article.

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Author Contributions

Benjamin Buttars (primary author) contributed to development of research concept, management, data acquisition, interpretation, and construction of the original manuscript with confirmation of final draft approval. Gates Scott was responsible for data acquisition/analysis contribution to the Methods section and original draft for intellectual content. Danielle Gates was responsible for data acquisition/analysis contribution to the Methods section and original draft for intellectual content. C. Ralph Daniel was responsible for interpretation/analysis of data, confirmation of primary research conception, data acquisition/analysis, manuscript review and editing for intellectual content, and confirmation to the final draft. Robert Brodell (corresponding author) was responsible for interpretation/analysis of data, confirmation of primary research conception, data acquisition/analysis, manuscript review and editing for intellectual content, and confirmation to the final draft. Mark Braswell (participating investigator) was responsible for data acquisition/analysis contribution to the Methods section and original draft for intellectual content.

References

- 1 Wagner G, Sachse MM. Congenital malalignment of the big toe nail. *J Dtsch Dermatol Ges*. 2011;10(5):326–30.
- 2 Catalfo P, Musumeci ML, Lacarrubba F, Dinotta F, Micali G. Congenital malalignment of the great toenails: a review. *Skin Appendage Disord*. 2018;4(4):230.
- 3 Braswell M, Daniel CR, Brodell RT. Beau lines, onychomadesis and retronychia. A unifying hypothesis. *J Am Acad Dermatol*. 2015; 73:849–55.
- 4 Samman PD. Great toe nail dystrophy. *Clin Exp Dermatol*. 1978;3(1):81–2.
- 5 Baran R, Bureau H, Sayag J. Congenital malalignment of the big toe nail. *Clin Exp Dermatol*. 1979;4:359–60.
- 6 Daniel CR 3rd, Tosti A, Iorizzo M, Piraccini BM. The disappearing nail bed: a possible outcome of onycholysis. *Cutis*. 2005;76:325–7.
- 7 Bailie FB, Evans DM. Ingrowing toenails in infancy. *Br Med J*. 1978;2(6139):737–8.
- 8 Balci S, Bostanci S, Ekmekci P, Cebeci I, Bokeysoy I, Bartsch O, et al. A 15-year-old boy with Rubinstein-Taybi syndrome associated with severe congenital malalignment of the toenails. *Pediatr Dermatol*. 2004;21:44.
- 9 Baran R, Bureau H. Congenital malalignment of the big toe-nail as a cause of ingrowing toenail in infancy. Pathology and treatment (a study of thirty cases). *Clin Exp Dermatol*. 1983;8(6):619–23.
- 10 Baran R, Bureau H. Congenital malalignment of the big toenail: a new subtype. *Arch Dermatol*. 1987;123(4):437.
- 11 Baran R. Congenital malalignment of the big toenail. *Arch Dermatol*. 1980;116(12):1346.
- 12 Baran R. Significance and management of congenital malalignment of the big toenail. *Cutis*. 1996;58(2):181–4.
- 13 Barth JH, Dawber RP, Ashton RE, Baran R. Congenital malalignment of great toenails in two sets of monozygotic twins. *Arch Dermatol*. 1986;122(4):379–80.
- 14 Chaniotakis I, Bonitsis N, Stergiopoulou C, Kiorpelidou D, Bassukas ID. Dizygotic twins with congenital malalignment of the great toenails: reappraisal of the pathogenesis. *J Am Acad Dermatol*. 2007;57(4):711–5.
- 15 Cohen PR. Congenital malalignment of the great toenails: case report and literature review. *Pediatr Dermatol*. 1991;8(1):43–5.
- 16 Dawson TA. An inherited nail dystrophy principally affecting the great toe nails. *Clin Exp Dermatol*. 1979;4(3):309–13.
- 17 Dawson TAJ. An inherited nail dystrophy principally affecting the great toenails: further observations (letter). *Clin Exp Dermatol*. 1982;7:455–6.
- 18 Decker A, Scher RK, Avarbock A. Acquired congenital malalignment of the great toenails. *Skin Appendage Disord*. 2016;1(3):147–9.
- 19 Domínguez-Cherit J, García-Galaviz R, Gatica-Torres M. Successful surgical treatment of bilateral congenital malalignment of the great toenail and hHypertrophic lateral nail folds. *Dermatol Surg*. 2019;45(9):1211–3.
- 20 Fierro-Arias L, Morales-Martínez A, Zazueta-López RM, Ramírez-Dovala S, Bonifaz A, Ponce-Olivera RM. Congenital malalignment of the great toenail. *Skinmed*. 2015;13(6):433–7.
- 21 Handfield-Jones SE, Harman RR. Spontaneous improvement of congenital malalignment of the great toe nails. *Br J Dermatol*. 1988;118(2):305–6.
- 22 Harper KJ, Beer WE. Congenital malalignment of the great toe-nails: an inherited condition. *Clin Exp Dermatol*. 1986;11(5):514–6.
- 23 Hendricks WM. Congenital ingrown toenails. *Cutis*. 1979;24(4):393–4.
- 24 Ko D, Lipner S. Congenital malalignment of the great toenail. *J Cutan Med Surg*. 2018; 22(3):326.
- 25 Kus S, Tahmaz E, Gurunluoglu R, Candan I, Uygur T. Congenital malalignment of the great toenails in dizygotic twins. *Pediatr Dermatol*. 2005;22(5):434–5.
- 26 Lipner SR, Scher RK. Congenital malalignment of the great toenails with acute paronychia. *Pediatr Dermatol*. 2016;33(5):e288–9.
- 27 Nakouri I, Litaïem N, Jones M, Zegloui F. Retronychia clinical features and surgical treatment. *J Am Podiatr Med Assoc*. 2018; 108(1):74–6.

- 28 Ozdemir E, Bostanci S, Akyol A, Ekmekci P, Gürgey E. Congenital malalignment of the great toenails in a pair of monozygotic twins. [J Am Podiatr Med Assoc](#). 2005;95(4):398–400.
- 29 Peralta L, Morais P. Great toenail deformity: case studies. [Aust Fam Physician](#). 2012;41(6):408–9.
- 30 Perlis CS, Telang GH. Congenital malalignment of the great toenails mimicking onychomycosis. [J Pediatr](#). 2005;146(4):575.
- 31 Richert B, Choffray A, de la Brassinne M. Cosmetic surgery for congenital nail deformities. [J Cosmet Dermatol](#). 2008;7(4):304–8.
- 32 Vasily DB. Dystrophy of the great toenail. [Arch Dermatol](#). 1980;116(1):19–20.
- 33 Wang CY, Kern J, Howard A. Late-onset malalignment of the great toenails. [Australas J Dermatol](#). 2019;60(4):315–7.
- 34 Wollina U, Wollina K. Kongenital dysplasie des Grosszehen nagels: eine Ursache chronisch-rezidivierend er Paronychien des Kindesalters. [Z Hautkr](#). 1995;70:35–7.
- 35 Daniel R, Meir B, Avner S. An update on the disappearing nail bed. [Skin Appendage Disord](#). 2017;3(1):15–7.
- 36 Baran R, Haneke E. Etiology and treatment of nail malalignment. [Dermatol Surg](#). 1998;24:719–21.
- 37 Andrews NA, Ray J, Dib A, Harrelson WM, Khurana A, Singh MS, et al. Diagnosis and conservative management of great toe pathologies: a review. [Postgrad Med](#). 2021 May; 133(4):409–20.