

# Congenital malalignment of the great toenail and onychomadesis in monozygotic twins



Benjamin Buttars, OMS-III,<sup>a</sup> Mark A. Braswell, DO,<sup>b</sup> Thy Huynh, MD,<sup>c</sup> Hannah Badon, MD,<sup>d</sup>  
C. Ralph Daniel III, MD,<sup>d</sup> and Robert T. Brodell, MD<sup>d</sup>

*Dothan, Alabama; St. Clair County, Illinois; Boston, Massachusetts; and Jackson, Mississippi*

**Key words:** Beau lines; congenital malalignment of the great toe; nail deviation; onychomadesis; onychomycosis; paronychia; retronychia.

## INTRODUCTION

Congenital malalignment of the great toenails (CMGTs) may result in onychocryptosis, a disappearing nail bed, paronychia, Beau lines, onychomadesis, retronychia, or erosion of the nail bed.<sup>1,2</sup> Onychomadesis is a spontaneous arrest of the nail plate that leads to nail shedding and is a more severe manifestation of the same pathophysiologic process that produces Beau lines.<sup>3</sup> Onychomadesis is commonly seen as a delayed onset effect of hand, foot, and mouth disease.<sup>4</sup> Infectious diseases, trauma, autoimmune/systemic diseases, and medications may also injure the nail matrix, leading to onychomadesis.<sup>5-8</sup> Rare cases of idiopathic forms of onychomadesis include familial autosomal dominant inheritance, as well as sporadic genetic mutation.<sup>3</sup> This case illustrates the possible link between CMGT and onychomadesis.

## CASE REPORT

Eleven-year-old male monozygotic twins presented with a lifelong history of abnormal growth and discoloration of the bilateral great and second toenails. The past medical history for the first twin was significant for ventriculoperitoneal shunt placement for communicating hydrocephalus at birth and surgical occlusion of a patent ductus arteriosus and unremarkable for the second twin. During infancy,

### Abbreviation used:

CMGT: congenital malalignment of the great toenail

both the twins had abnormal first and second toenail plates. Neither twin had any toenail pain or pain with ambulation. Fungal cultures of the great toenail clippings demonstrated no fungal growth. There was no improvement following a therapeutic trial of griseofulvin 20 mg/kg for 4 months and oral terbinafine 125 mg daily for 12 weeks.

Physical examination of both twins revealed yellow discoloration, lateral deviation of the great toenail, and multiple complete transverse indentations with nail plate separations (Beau lines) involving the first and second toenail plates (Fig 1). Several nail plate separations were full thickness, illustrating a complete spontaneous arrest of the nail matrix. Due to the onset, presentation, and chronic condition of their toenails, both patients were diagnosed with onychomadesis secondary to CMGT.

The patients were instructed to wear properly fitted shoes, practice good nail hygiene, and trim nails horizontally and not round at the edges to decrease the chances of an ingrowing nail. Surgical intervention, such as complete nail matrixectomy,

From the Alabama College of Osteopathic Medicine, Dothan,<sup>a</sup> Scott Air Force Base, St. Clair County,<sup>b</sup> Boston Children's Hospital,<sup>c</sup> and Dermatology Department, University of Mississippi Medical Center, Jackson.<sup>d</sup>

Funding sources: None.

Disclaimer: The authors whose names are listed immediately below certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or nonfinancial interest (such as personal or professional relationships, affiliations, knowledge, or beliefs) in the subject matter or materials discussed in this manuscript. The views expressed in this case report are

those of the authors and do not necessarily reflect the official policy or position of the Department of the Air Force, Department of Defense, or the US Government.

IRB approval status: Not applicable.

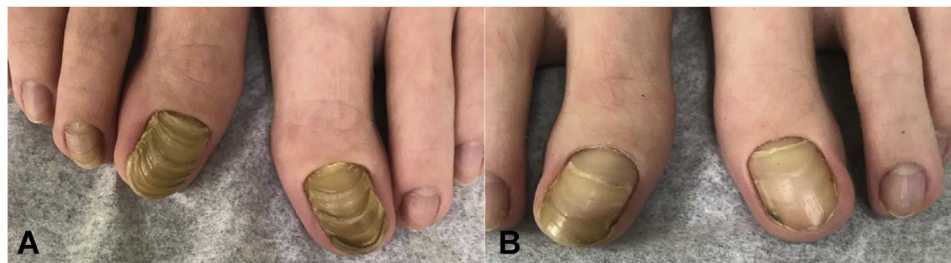
Correspondence to: Benjamin Buttars, OMS-III, Alabama College of Osteopathic Medicine, 445 Health Sciences Blvd, Dothan, AL 36303. E-mail: [buttarsb@acom.edu](mailto:buttarsb@acom.edu).

JAAD Case Reports 2021;14:■-■.

2352-5126

© 2021 Published by Elsevier on behalf of the American Academy of Dermatology, Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

<https://doi.org/10.1016/j.jidcr.2021.05.027>



**Fig 1.** A and B, Twins with bilateral congenital malalignment of the great toenails showed complete transverse indentations associated with arrest of nail growth.



**Fig 2.** Deep sulci and complete matrix arrest were associated with onychomadesis.

was not recommended for these asymptomatic twins.

## DISCUSSION

Most nail pathologies are slow healing and can be complicated to treat. Beau lines occur when the matrix ceases to produce nail keratin and the nail moves distally, resulting in depressions or breaks in the integrity of the nail plate.<sup>3,4</sup> Complete shedding of the nail occurs when the nail plate distal to the Beau line separates from the nail bed (Fig 2).

Onychomadesis of various causes is related to the arrest of nail matrix growth (Table I). In CMGT, lateral displacement of the nail apparatus causes recurrent, shearing microtrauma to the nail matrix that would not occur in properly aligned toenails. As such, CMGT increases the risk of various forms of nail dystrophy, including paronychia, retronychia, onycholysis, onychogryphosis, and onychocryptosis.<sup>8,9</sup> We report that the identical genetically determined malalignment of the 4 toenails in the twins

**Table I.** The causes of onychomadesis and their relationship with temporary stunting of nail matrix growth

Etiology	Condition resulting in stunted nail matrix growth
Single traumatic event	Acute trauma to the great toe
Recurrent microtrauma	Congenital malalignment of the great toe and/or toenail, traumatic nail malalignment, and iatrogenic malalignment due to wide lateral nail biopsies, ill-fitted shoes, and bunions
Systemic insults	High fever and systemic infectious diseases, COVID-19, nutritional deficiency
Local inflammatory or infectious involvement of the nail matrix	Hand, foot, and mouth disease; autoimmune diseases; cutaneous vasculitis drug reactions; varicella; bullous disease
Drug-induced arrest of nail matrix growth	Chemotherapy treatment, antiepileptic medications, antifungal treatment

that was associated with weight gain and activities of daily living during pubescence, increases the strain on the nail matrix causing recurrent arrest of the matrical growth. This results in horizontal nail depressions (Beau lines) and, ultimately, onychomadesis.

Efforts to prevent onychomadesis include minimization of trauma, treatment of any underlying systemic disease, and discontinuation of inciting medications such as chemotherapy. In CMGT, the shearing forces produced with normal ambulation probably cannot be prevented by measures such as avoiding ill-fitted shoes and choosing shoes with a wide toe box. Despite this, careful attention to properly sized and fitted footwear is mandatory to reduce the risk of secondary complications. Surgical options include removal and reimplantation of the nail matrix.<sup>10</sup> In adults with recurrent

onychomadesis and onychogryphosis, complete phenol nail matrixectomy may be required. Generally, surgical options become a priority only when patients become symptomatic with recurrent infection and painful onychocryptosis.<sup>1</sup>

This rare case shows a clear association between CMGT and onychomadesis. The extended period of the necessary treatment window and the imperfect nature of the available preventive treatment make it difficult to establish whether the patients will benefit from our suggestions.

#### Conflicts of interest

None disclosed.

#### REFERENCES

1. Wagner G, Sachse MM. Congenital malalignment of the big toe nail. *J Dtsch Dermatol Ges*. 2012;10(5):326-330.
2. Daniel R, Meir B, Avner S. An update on the disappearing nail bed. *Skin Appendage Disord*. 2017;3(1):15-17.
3. Hardin J, Haber RM. Idiopathic sporadic onychomadesis: case report and literature review. *Arch Dermatol*. 2012;148(6):769-770.
4. Giordano LMC, de la Fuente LA, Lorca JMB, Kramer HD. Onychomadesis secondary to hand-foot-mouth disease: a frequent manifestation and cause of concern for parents. *Rev Chil Pediatr*. 2018;89(3):380-383. Article in Spanish.
5. Podder I, Das A, Gharami RC. Onychomadesis following varicella infection: is it a mere co-incidence? *Indian J Dermatol* 2015;60(6):626-627.
6. Grover C, Vohra S. Onychomadesis with lichen planus: an under-recognized manifestation. *Indian J Dermatol*. 2015;60(4):420.
7. Damevska K, Gocev G, Pollozahani N, Nikolovska S, Neloska L. Onychomadesis following cutaneous vasculitis. *Acta Dermatovenerol Croat*. 2017;25(1):77-79.
8. Catalfo P, Musumeci ML, Lacarrubba F, Dinotta F, Micali G. Congenital malalignment of the great toenails: a review. *Skin Appendage Disord*. 2018;4(4):230-235.
9. Braswell MA, Daniel CR III, Brodell RT. Beau lines, onychomadesis, and retronychia: a unifying hypothesis. *J Am Acad Dermatol*. 2015;73(5):849-855.
10. Baran R, Haneke E. Etiology and treatment of nail malalignment. *Dermatol Surg*. 1998;24(7):719-721.