

## BRIEF REPORT

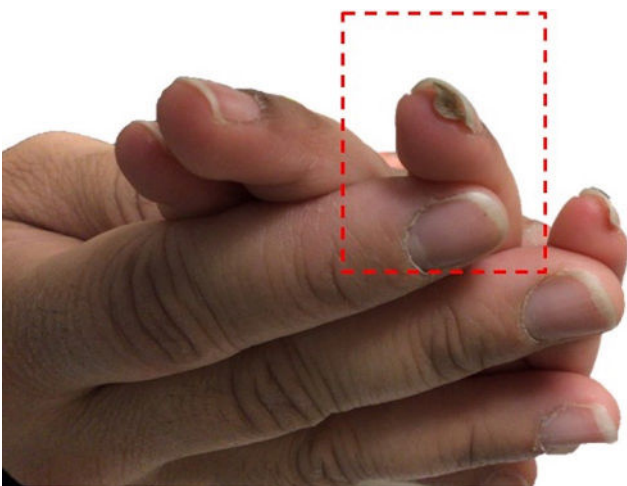
# Congenital onychodysplasia of the ring finger presenting as a bifid nail

## Abstract

Congenital onychodysplasia, or Iso-Kikuchi syndrome, is classically defined as a congenital nail abnormality of one or both index fingers that is often associated with a bone abnormality in the affected finger. We report an unusual case of a 6-year-old girl who presented with an S-shaped, bifid nail of the left ring finger that had been present since birth. X-ray findings were used to confirm a diagnosis of congenital onychodysplasia of the ring finger.

## 1 | CASE REPORT

A 6-year-old girl presented for evaluation of a congenital nail dystrophy of the left ring finger. The dystrophic nail was located just ventral to a normally developed fourth nail plate, appearing to project outward from the hyponychium. The anomaly had an S-shaped curvature with longitudinal ridges throughout (Figure 1). The patient reported recurrent pain with clipping of the nail. X-ray showed brachydactyly of the fourth and fifth distal phalanges, terminal widening of the fourth distal phalanx, and Y-shaped bifurcation of the fourth distal phalanx (Figure 2A, B, C). The patient had no history of



**FIGURE 1** A bifid nail growing from the left ring finger of a child with congenital onychodysplasia

trauma to the hand, complications during gestation, similar medical history in the family, or familial history of consanguineous marriage. Past medical history was unremarkable other than a left ear abnormality requiring tube placement. Based on the symptomatology, congenital onychodysplasia of the ring finger was diagnosed.

Congenital onychodysplasia, or Iso-Kikuchi syndrome, has classically been characterized as a congenital nail abnormality of one or both index fingers, often with an associated bone anomaly and without family history.<sup>1</sup> Since first being defined by Iso in 1969 and Kikuchi in 1974, cases of congenital onychodysplasia of the third finger and second toe have been reported.<sup>1,2</sup> However, to the best of our knowledge, no previously reported cases of congenital onychodysplasia of the ring finger were found. Associated congenital nail abnormalities include micronychia, polyonychia, anonychia, hemionychogryphosis, malalignment, and irregularities of the lunula.<sup>3</sup> Abnormalities in the bone have ranged from distal phalanx abnormalities of the affected finger, most commonly enlargement with bifurcation (often described as Y-shaped or clawlike); deformities of the hand, such as syndactyly or brachymesophalangy; and functional deformities, such as restricted flexion in the distal interphalangeal joint of the affected finger.<sup>1,2,4</sup> Bony abnormalities can also be present in a finger that shows no visible nail dysplasia, as noted in the left fifth digit of our patient.<sup>1</sup> In our patient, each wing of the bony bifurcation was likely associated with its own nail matrix, leading to the clinical presentation of a ventrally oriented S-shaped bifid nail.

Limited evidence of any further medical associations with congenital onychodysplasia outside of the abnormalities of the hand was found. However, three cases have been reported with associated ear abnormalities, congruent with our patient.<sup>1,5</sup>

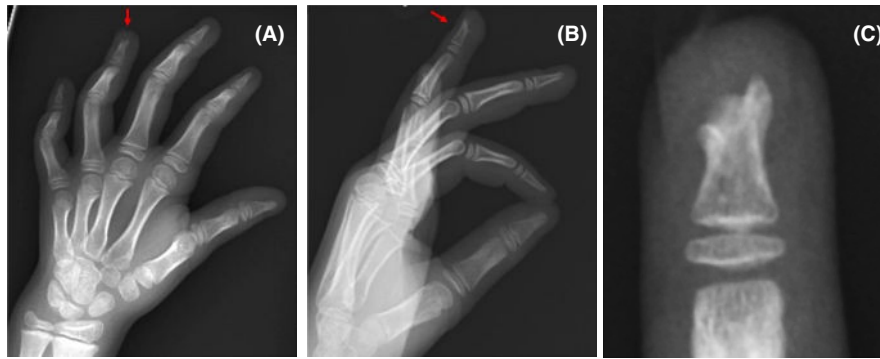
Treatment of congenital onychodysplasia is optional and usually dictated by symptoms or cosmetic concerns. Due to the pain associated with our patient's dystrophic nail, urea cream has been used to soften the nail; however, if unresponsive, partial matricectomy of the curved portion of the nail will be considered.

## KEYWORDS


congenital abnormalities, nails, malformed, radiography

## CONFLICT OF INTEREST

The authors have no conflict of interest to declare.



**FIGURE 2** X-ray findings of a 6-year-old child with congenital onychodysplasia of the ring finger. A, Left hand with terminal widening and brachydactyly of the fourth distal phalanx. Brachydactyly of the fifth distal phalanx is also seen. B, Left hand with Y-shaped bifurcation of the fourth distal phalanx. C, Left ring finger showing Y-shaped bifurcation of the distal phalanx

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#### REFERENCES

1. Kitayama Y, Tsukada S. Congenital onychodysplasia: report of 11 cases. *Arch Dermatol.* 1983;119(1):8-12. <https://doi.org/10.1001/archderm.1983.01650250012007>

2. Yang MY, Ko HC, Kim BS, Kim MB, Jo G, Mun JH. Image gallery: congenital onychodysplasia of the toenails. *Br J Dermatol.* 2017;177(1):e3. <https://doi.org/10.1111/bjd.15478>
3. Milani-Nejad N, Mosser-Goldfarb J. Congenital onychodysplasia of index fingers: Iso-Kikuchi syndrome. *J Pediatr.* 2020;218:254.e1. <https://doi.org/10.1016/j.jpeds.2019.10.008>
4. Kikuchi I, Abe S, Amano F. Congenital onychodysplasia of the index fingers. *Arch Dermatol.* 1976;112(12):1788. <https://doi.org/10.1001/archderm.1976.01630370068016>
5. Haneke E, Kienlein-Kletschka B. Kongenitale onychodysplasie: Iso-Kikuchi-syndrom [Congenital onychodysplasia: Iso-Kikuchi syndrome]. *Hautarzt.* 1984;35(9):468-471.

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