



# ACTAS Derma-Sifiliográficas

Full English text available at  
[www.actasdermo.org](http://www.actasdermo.org)



## E- CASE REPORT

# Iso-Kikuchi Syndrome: Report of 3 Pediatric Cases<sup>☆</sup>



L.L. Tirelli,\* P.C. Luna, R. Cano, J.P. Giraldo, M. Larralde

Servicio de Dermatología, Hospital Alemán, Ciudad Autónoma de Buenos Aires, Argentina

### KEYWORDS

Iso-Kikuchi;  
Anonychia;  
Nail;  
Index finger;  
Phalanx

### PALABRAS CLAVE

Iso-Kikuchi;  
Anoniquia;  
Uña;  
Dedo índice;  
Falange

**Abstract** Iso-Kikuchi syndrome, or congenital onychodysplasia of the index finger, is an uncommon condition characterized by total anonychia or dysplasia of the nail of the index finger. It is occasionally accompanied by underlying bone abnormalities and is rarely associated with other conditions. Although various hypotheses have been put forward to explain the pathophysiology of the syndrome, its etiology remains unknown.

We report the cases of 3 pediatric patients (2 boys and 1 girl) with nail changes and bone abnormalities consistent with Iso-Kikuchi syndrome. We highlight the importance of recognizing this entity early to avoid the need for additional tests and unnecessary treatment.

© 2017 Elsevier España, S.L.U. and AEDV. All rights reserved.

### Síndrome de Iso-Kikuchi: 3 casos en la edad pediátrica

**Resumen** El síndrome de Iso-Kikuchi, onicodisplasia congénita del dedo índice, es una entidad poco frecuente caracterizada por la anoniquia total o displasia de la uña del dedo índice, acompañado, en algunas ocasiones, de alteraciones óseas subyacentes, por lo general, en ausencia de otras anomalías. Si bien se han planteado distintas hipótesis fisiopatológicas, la etiología sigue siendo desconocida.

Describimos los casos de 3 pacientes pediátricos, 2 varones y una niña, con alteraciones ungueales y óseas compatibles con el síndrome de Iso-Kikuchi. Destacamos la importancia de reconocer esta entidad tempranamente para evitar la realización de estudios complementarios y terapéuticos innecesarios.

© 2017 Elsevier España, S.L.U. y AEDV. Todos los derechos reservados.

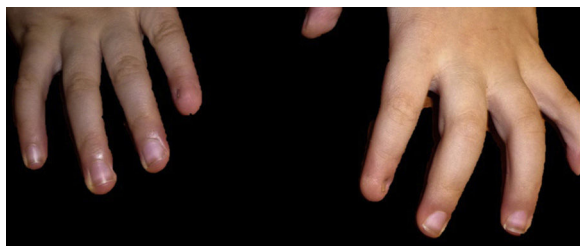
## Introduction

Iso-Kikuchi syndrome is characterized by anonychia or onychodysplasia of the index finger, accompanied or not by underlying bone abnormalities. It is a benign condition that is not associated with alterations to other organs or systems. Very few cases have been reported to date.

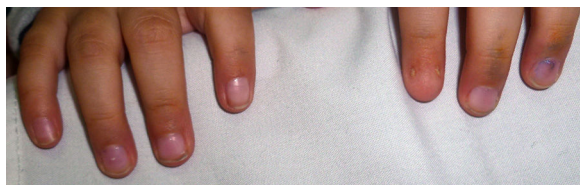
<sup>☆</sup> Please cite this article as: Tirelli LL, Luna PC, Cano R, Giraldo JP, Larralde M. Síndrome de Iso-Kikuchi: 3 casos en la edad pediátrica. Actas Dermosifiliogr. 2018;109:e33–e36.

\* Corresponding author.

E-mail address: [lucianatirelli@yahoo.com.ar](mailto:lucianatirelli@yahoo.com.ar) (L.L. Tirelli).



**Figure 1** Patient #1. Note the micronychia on the radial side of both index fingers.



**Figure 2** Patient #2. Unilateral involvement of the left index finger with micronychia on the ulnar and radial sides of the nail bed. The rest of the nails are unaltered.



**Figure 3** Patient #3. Polyonychia of the left index finger and nail dystrophy of the right index finger.

None of the patients' relatives had Iso-Kikuchi syndrome. All the parents denied consanguinity and reported that their children's nail alterations had been present since birth.

## Discussion

Iso-Kikuchi syndrome was first described by Dr. Iso in 1969 and Dr. Kikuchi in 1974,<sup>1-3</sup> and very few cases have been reported since (Table 2). It affects both sexes equally and can be sporadic or familial.

Iso-Kikuchi syndrome runs an indolent course that is characterized by anonychia or onychodysplasia of the index finger possibly accompanied by bone abnormalities in the underlying phalanx.<sup>4</sup> It can affect one or both hands or feet. Based on their observations and analysis of different clinical cases, Baran and Stroud<sup>5</sup> proposed the following diagnostic criteria for Iso-Kikuchi syndrome: 1) unilateral or bilateral hypoplasia (up to complete anonychia) of the index finger and/or other fingers or toes, 2) radiographic changes in the distal phalanx of the affected digit; and 3) sporadic or hereditary congenital occurrence. Transmission of Iso-Kikuchi syndrome is autosomal dominant. No associations with systemic diseases have been reported.<sup>4,5</sup>

Onychodysplasia has multiple presentations (Table 2), the most common of which are anonychia, micronychia (characteristic deviation of the nail to the ulnar side of the nail bed),<sup>6</sup> polyonychia (2 nails, 1 on each side of the nail bed),

## Case Descriptions

Two boys and 1 girl presented with Iso-Kikuchi syndrome for the first time between the ages of 2 and 4 years.

Patient #1, a 2-year-old boy, had bilateral index finger involvement and a Y-shaped deformity of the underlying phalanx (Fig. 1). His past medical history was remarkable for extreme preterm birth (30 weeks), which is why we have included other diseases associated with this condition in Table 1.

Patient #2, a 4-year-old girl, had involvement of the left index finger and a Y-shaped bifurcation of the affected phalanx (Fig. 2).

Patient #3 was 2 years old and had bilateral index finger involvement (Fig. 3) without radiographic changes (Table 1).

**Table 1** Cases Presented.

Patient	1	2	3
Sex	Male	Female	Male
Age	2 y	4 y	2 y
Perinatal history	Born at 30 wk; nail changes present at birth	Term delivery; nail changes present at birth	Term delivery; nail changes present at birth
Digit involved	Both index fingers	Left index finger	Both index fingers
Appearance of other nail	Micronychia on radial side	Micronychia on radial and ulnar sides	One finger with polyonychia, dystrophic homolateral finger
Radiographic changes	Bilateral Y-shaped deformity	Y-shaped deformity in affected finger	None
Other alterations	Grade 3 intraventricular hemorrhage, bilateral inguinal bleeding, bilateral pelviureteric junction stenosis	None	Gastroesophageal reflux
Family history	No	No	No

**Table 2** Cases Published in the Literature.

Publication	Reference	No. of Patients	Age	Sex	Characteristics	Other
Di Chiacchio et al.	3	1	13 y	Not specified	Micronychia and altered lunula of right index finger Micronychia, malalignment, and irregular lunula of left index finger	No
Valerio et al.	4	1	Neonate	Not specified	Complete anonychia of left middle finger	Mother: consumption of mebendazole during pregnancy
Baran & Stroud	5	1	41 y	Male	Hemionychogryphosis of right and left index fingers and abnormal lunula	No
Hussein et al.	7	1	52 y	Female	Ulnar micronychia of both index fingers	Leukonychia of both hands
Thappa & Shivaswamy	8	1	20 y	Male	Polyonychia of right index finger	No
Iso	1					
Kikuchi et al.	2	2	5 y 29 y	Female Female	Polyonychia of right and left index fingers and absent lunulae Polyonychia of left index finger; hemionychogryphosis of contralateral nail	Radiography: narrowing of distal phalanx of both index fingers, brachymesophalangy of thumbs Radiography: brachymesophalangy of both thumbs
Kikuchi et al.	6	2	24 y 4 mo	Female Female	Ulnar micronychia of both index fingers Polyonychia of left index finger	Radiography: Y-shaped bifurcation Radiography: hypoplasia of underlying phalanx

hemionychogryphosis, irregular lunula, and nail malalignment.

Narrowing of the distal phalanx is a characteristic radiographic finding, but it is not always present. This narrowing will be seen on anteroposterior radiographs, whereas lateral projections will show the corresponding Y-shaped bifurcation.<sup>3-8</sup> Other, less common, bone changes include brachydactyly, brachymesophalangia, and brachymetacarpia.<sup>7,8</sup>

The etiologic and pathogenic mechanisms of Iso-Kikuchi syndrome are not fully understood, and while several theories have been put forward, none of them have been fully accepted. Notable causes are mutations in the Wnt signaling pathway during embryogenesis, in utero damage to arterioles that depend on the radial artery (which has a smaller diameter than the ulnar artery), osteodystrophy of the phalanx during embryogenesis, and consumption of teratogenic drugs (anticonvulsants) during pregnancy.<sup>4</sup>

Diagnosis is mostly clinical. Iso-Kikuchi syndrome should be suspected in patients with congenital nail changes affecting one or both index fingers in the absence of

other hand alterations, particularly when characteristic radiographic alterations are observed. The differential diagnosis should include other congenital nail changes, such as ectodermal dysplasia, pachyonychia congenita, nail-patella syndrome, and deafness-onychodystrophy syndrome. Although nail changes are present from an early age in all these diseases, they have distinctive clinical characteristics and are often accompanied by extracutaneous alterations. The differential diagnosis may also include acquired nail changes due to trauma or infection, but these can be distinguished by their postnatal onset.

Iso-Kikuchi syndrome is a rare, benign syndrome with characteristic clinical features. We have presented 3 new pediatric cases and highlight the importance of early recognition to avoid the need for unnecessary tests and treatment.

### Conflicts of Interest

The authors declare that they have no conflicts of interest.

## Acknowledgments

We thank Bruno Mentasti for his help with editing the photographs.

## References

1. Iso R. Congenital nail defects of the index finger and reconstructive surgery. *Seikei Geka*. 1969;20:1383–4.
2. Kikuchi I, Horikawa S, Amano F. Congenital onychodysplasia of the index fingers. *Arch Dermatol*. 1974;110:743–6.
3. Di Chiacchio N, Jasso-Olivares JC, di Chiacchio NG, Jacinto JA, Restrepo MV. Syndrome in question. *An Bras Dermatol*. 2015;90:423–5.
4. Valerio E, Favot F, Mattei I, Cutrone M. Congenital isolated Iso-Kikuchi syndrome in a newborn. *Clin Case Rep*. 2015;3:866–9.
5. Baran R, Stroud JD. Congenital onychodysplasia of the index fingers-Iso and Kikuchi syndrome. *Arch Dermatol*. 1984;120:243–4.
6. Kikuchi I, Ishi Y, Idemori M, Ogata K. Congenital nail defects of the index finger. A possible explanation of radially pronounced involvement of the nail in this disorder. *J Dermatol*. 1981;8:145–9.
7. Hussein TP, Brandt HRC, Gabbi TVB, Nico MMS. Malformations of the index nails. *Clin Exp Dermatol*. 2008;34:890–1.
8. Thappa DM, Shivaswamy KN. Polyonychia in Congenital onychodysplasia of the Index Finger (Iso and Kikuchi Syndrome): A Report from India. *J Dermatol*. 2002;29:603–5.