

NAIL ANOMALIES IN CHILDREN

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DISCLOSURE

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I declare no conflicts of interests

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Nail diseases in children

- Hereditary
 - syndromic
 - non syndromic
- Congenital
 - Congenital anomalies are the result of one or more genetic, infectious, nutritional, environmental, or unknown factor*
 - Some congenital anomalies can be prevented***
- Acquired
 - Same as in adults*
 - Frequency may vary***

Present at birth
Appear later in life

Congenital anomalies are the result of one or more genetic, infectious, nutritional, environmental, or unknown factor
Some congenital anomalies can be prevented

Same as in adults
Frequency may vary

Telfer NR, et al. Congenital nail dystrophies: an embryological approach to classification. Clin Exp Dermatol 1988;13:160-3

Khan S, et al. Genetics of human isolated hereditary nail disorders. Br J Dermatol 2015;173:922-9

Itin P, et al. Nail changes in genodermatoses. Eur J Dermatol 2002;12:119-28

Viseux V, Plantin P. L'ongle du nouveau-né et du nourrisson. Ann Dermatol Venereol 2003;130:74-8

Chinazzo M, et al. Nail features in healthy term newborns: a single-centre observational study of 52 cases. JEADV 2017;31:371-5

Hereditary/Congenital Disorders limited to the nails

Type MIM Number	Clinical Manifestations	Mode of Inheritance, Gene, Mapping	Other Manifestations
NDNC-1, MIM161050	Trachyonychia	AD	Nails are variably involved. Thinning, thickening, pitting, opalescence, absence of thumb nails, longitudinal ridges, discoloration is possible. Koilonychia reported sometimes.
NDNC-2	Koilonychia	AD	
NDNC-3, MIM 151600	Leukonychia	AR and AD, <i>PLCD1</i> , 3p21.3	True, apparent, or pseudoleukonychia belong to this group.
NDNC-4, MIM 206800	Anonychia, hyponychia congenita	AR, <i>RSPO4</i>	Possible remnants or rudimentary nail plates.
NDNC-5, MIM 164800	Distal onycholysis	AD	Increased transverse curvature and absent lunulae are possible.
NDNC-6, MIM 107000	Partial absence of nails	AD	Thumbs and great toe are severely affected; anonychia is reported.
NDNC-7, MIM 605779	Longitudinal streaks	AD, 17p13	Thinning of nail plate, poorly developed, or absent lunulae, vulnerability of the free nail margins.
NDNC-8, MIM 607523	Isolated toenail dystrophy	AD, (3p21.3, <i>COL7A1</i>) ^a	Two compound heterozygous families.
NDNC-9, MIM 614149	Anonychia of toenails and onycholysis of fingernails	AR, 17q25	Normal fingers and toes at birth. When 7–8 years old, onychodystrophy started on finger and toenails.
NDNC-10, MIM 614157	Nail dysplasia	AR, 8q22.3, <i>FZD6</i>	Started at birth, slow rate of nail growth. When 10 years old, claw-like structure is reported.
ICNC, MIM 119900	Isolated congenital nail clubbing	AR, 4q32-q34, <i>HPGD</i>	Bilateral, symmetric, congenital, all nails affected



Congenital total / subtotal leukonychia

- TRUE form (no fading with pressure)
- Idiopathic, Inherited (AD/AR), Secondary
- Several genes involved (PLCD1, GJB2, CAST)
- Can be a sign in several Ectodermal Dysplasias
- Exclude syndromes characterized by **auditory abnormalities** (sensorineural deafness) as Bart-Pumphrey (142900) or **generalized peeling syndrome** as PLACK (616295)



Iorizzo M, Starace M, Pasch MC. Leukonychia: What Can White Nails Tell Us? *AJCD* 2022;23:177

Bart RS, Pumphrey RE. Knuckle pads, leukonychia and deafness: a dominantly inherited syndrome. *N Engl J Med* 1967;276:202

DeMille D, et al. Three novel GJB2 variants associated with autosomal dominant syndromic and nonsyndromic hearing loss. *Am J Med Genet A* 2018;176:945

Lin Z, et al. Loss-of-function mutations in CAST cause peeling skin, leukonychia, acral punctate keratoses, cheilitis, and knuckle pads. *Am J Hum Genet* 2015;96:440

Congenital anonychia/micronychia

Check the mother's history before asking for additional tests !!!



- Anticonvulsants
(*carbamazepine, hidantoin, valproic acid*)
- Anticoagulants (*warfarin*)
- Morphine

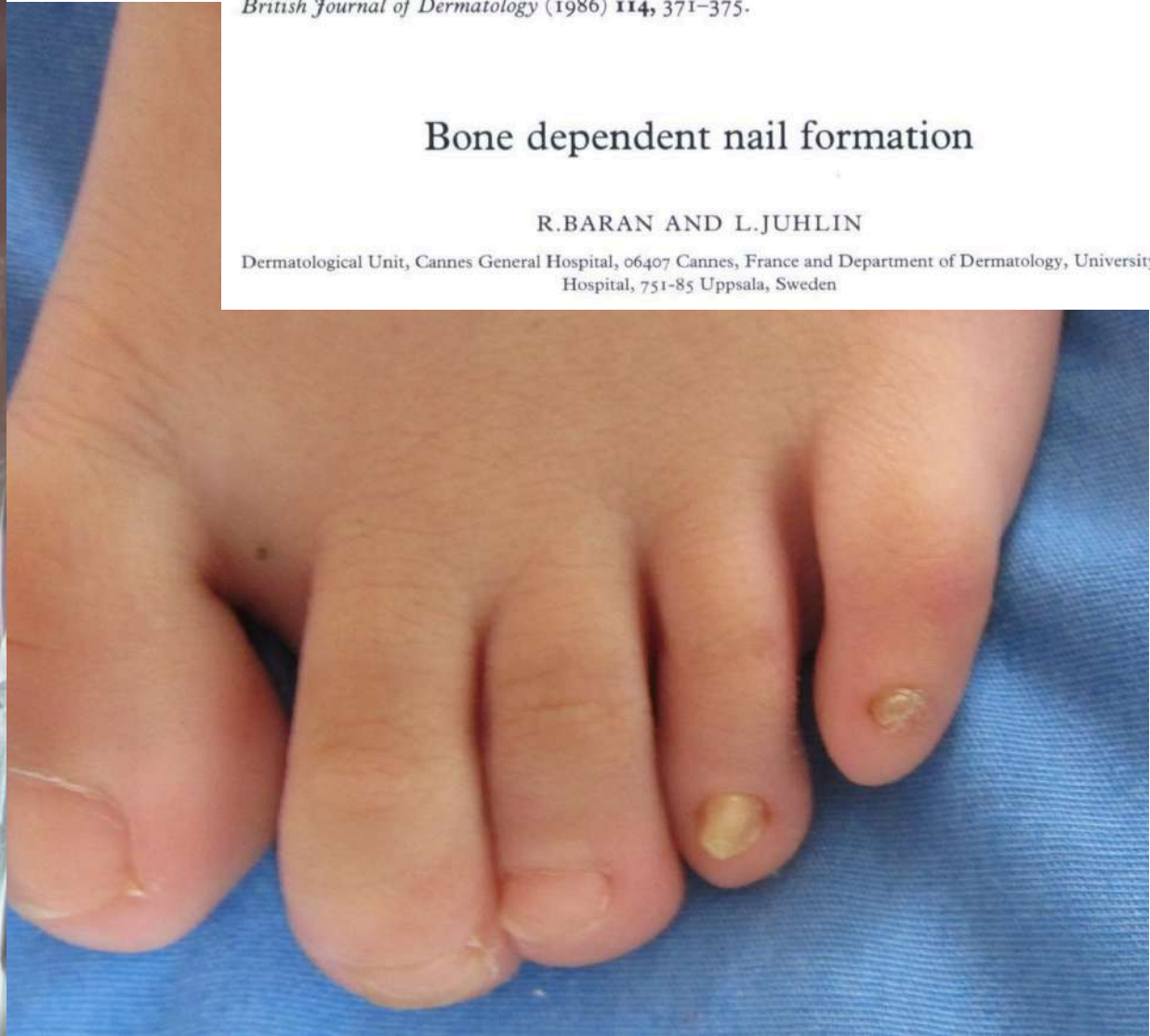
Peller AJ, et al. Physical features of newborns exposed during pregnancy to anticonvulsant medication and developmental monitoring. Birth Defects Res 2021;113:995-1000.

Jańczewska I, et al. Fetal alcohol spectrum disorders - diagnostic difficulties in the neonatal period and new diagnostic approaches. Dev Period Med 2019;23:60-6.

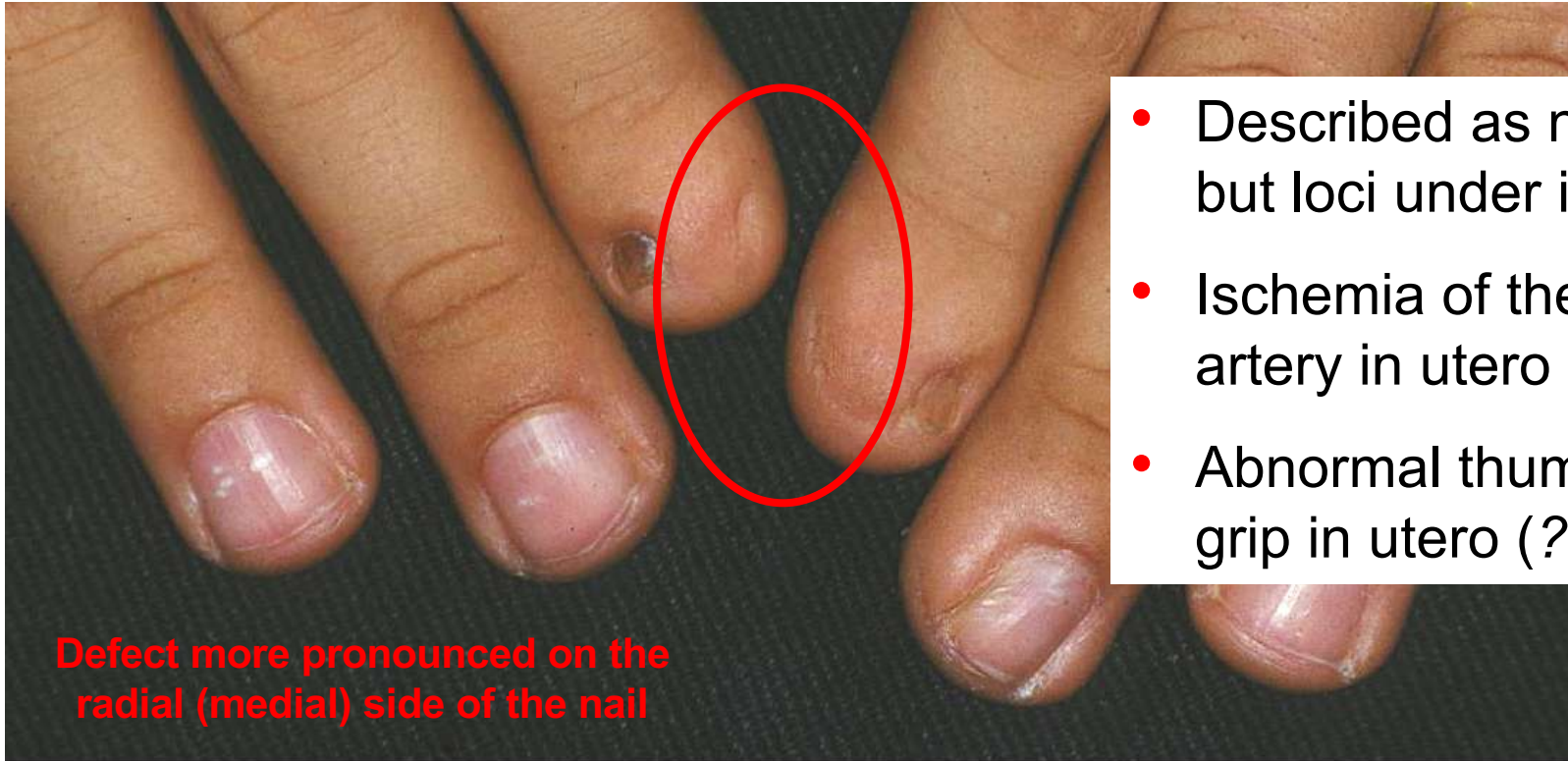
Bone dependent nail formation

R.BARAN AND L.JUHLIN

Dermatological Unit, Cannes General Hospital, 06407 Cannes, France and Department of Dermatology, University Hospital, 751-85 Uppsala, Sweden



Congenital Onychodysplasia of the Index Finger



- Described as non hereditary but loci under investigation
- Ischemia of the radial digital artery in utero (??)
- Abnormal thumb-on-finger grip in utero (??)

Iso R. Congenital nail defects of the index finger and reconstructive surgery. Seikei Geka 1969;20:1383-4.

Kikuchi I, et al. Congenital onychodysplasia of the index fingers. Arch. Dermatol 1974;110:743-6.

Baran R. Syndrome d'Iso et Kikuchi (C.O.I.F. syndrome) 2 cas avec revue de la litterature 44 cas. Ann Dermatol Venereol 1980;107:431-5.

Hamm HS, et al. Isolated congenital nail dysplasia: a new autosomal dominant condition. Arch Dermatol 2000;136:1239-43.

**X-ray DIAGNOSTIC (lateral projection):
Y-shaped bifurcation of the distal phalanx**



Nail Patella Syndrome

Defect more pronounced on the ulnar (lateral) side of the nail



- Lack of skin creases overlying the distal interphalangeal joints and hypoplasia of the distal phalanges

- Triangular lunula



Daniel CR, et al. Triangular lunulae: a clue to Nail-Patella syndrome. Arch Dermatol 1980;116:448-9.

Itin PH, et al. Missing creases of distal finger joints as a diagnostic clue of nail-patella syndrome. Dermatology 2006;213:153-5.

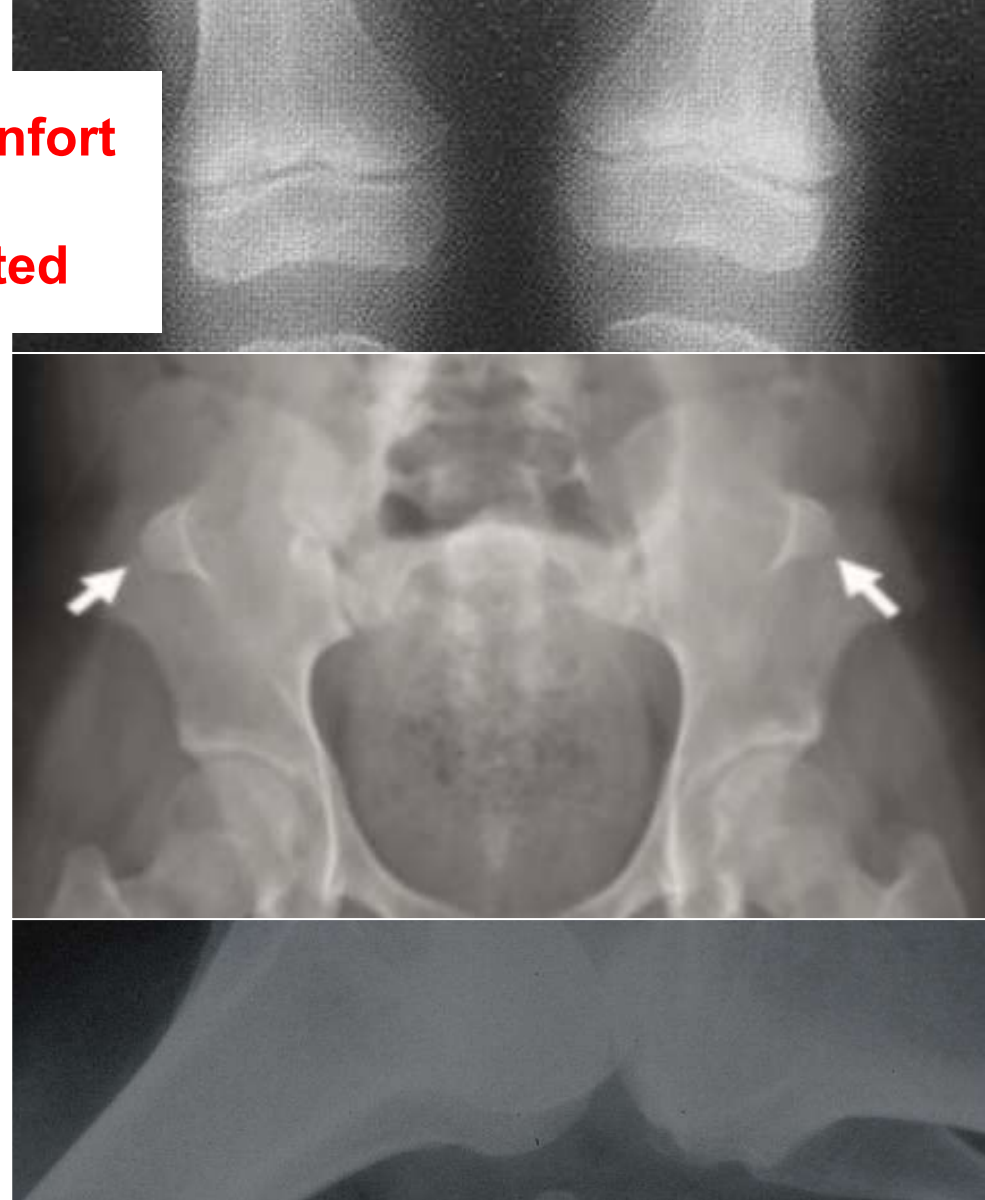
Starace M, et al. A double case of NPs in the same family: the importance of nail changes as diagnostic clues for renal involvement. SAD 2019;5:405-8.

Mild nail changes may cause minor discomfort
Diagnosis is delayed
Occult manifestations can go underreported

- **Kidneys impairment**
- Glaucoma / Deafness
- Mutation LMXB1 gene (AD)

X-rays DIAGNOSTIC

- Patellar absence
- Patellar hypoplasia
- Hyliac horns



- Kidney involvement in 30-50% of patients
- Presents with hematuria and proteinuria
- Up to 10% develop end stage renal failure

- LMX1B impairs the development and functioning of podocytes and glomerular filtration slits

Ectodermal Dysplasias

Christ-Siemens-Touraine Syndrome – Anidrotic / Hypohidrotic ED

Clouston Syndrome – Hidrotic ED

Dyskeratosis Congenita

Ectrodactyly Ectodermal Dysplasia Cleft Lip/Palate (EEC)

Goltz Syndrome – Focal Dermal Hypoplasia

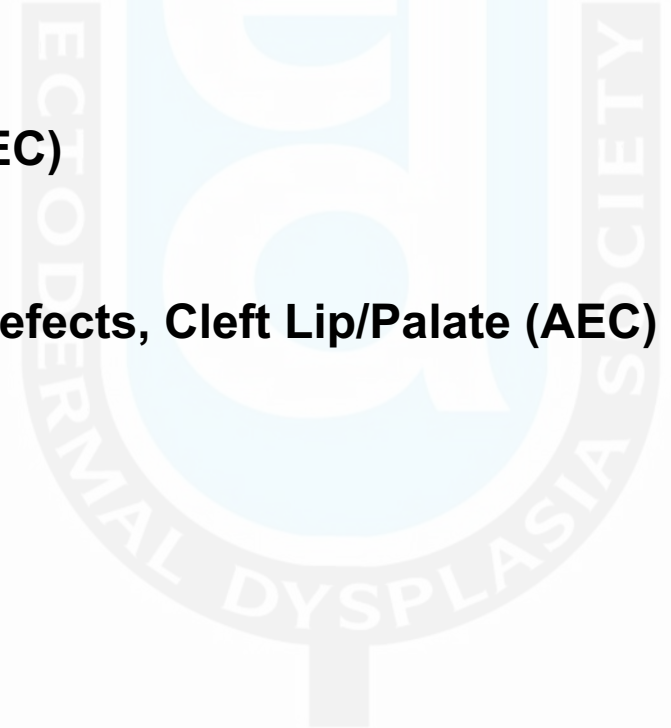
Hay Wells Syndrome – Ankyloblepharon Ectodermal Defects, Cleft Lip/Palate (AEC)

Incontinentia Pigmenti

Pachyonychia Congenita

Rothmund-Thomson's Syndrome

Trichorhinophalangeal Syndrome



Pachyonychia Congenita

- Thick nails
- Abnormally curved
- Onycholysis
- Nail bed hyperkeratosis
- Slow growth

- **Palmoplantar painful hyperkeratosis (*KRT6c*)**
- Follicular hyperkeratosis
- Oral leukokeratosis
- Hoarseness
- Natal teeth preservation



Changes present, at birth: 50% of cases

Changes present, after 5 years: 80% of cases

KRT6a and KRT17 mutations more severe on nails

Shah S, et al. PC in pediatric patients: natural history, features, and impact. JAMA Dermatol 2014; 150: 146–53.

Eliason MJ, et al. A review of the clinical & phenotype of 254 patients with genetically confirmed PC. JAAD 2012; 67:680–6.

Most children manifest the key features in the first year of life, but **the diagnosis is often delayed**

Iorizzo M, et al. PC Type 1 presenting with subtle nail changes. Ped Derm 2009;26:492-3.



*Candida
parapsilosis*

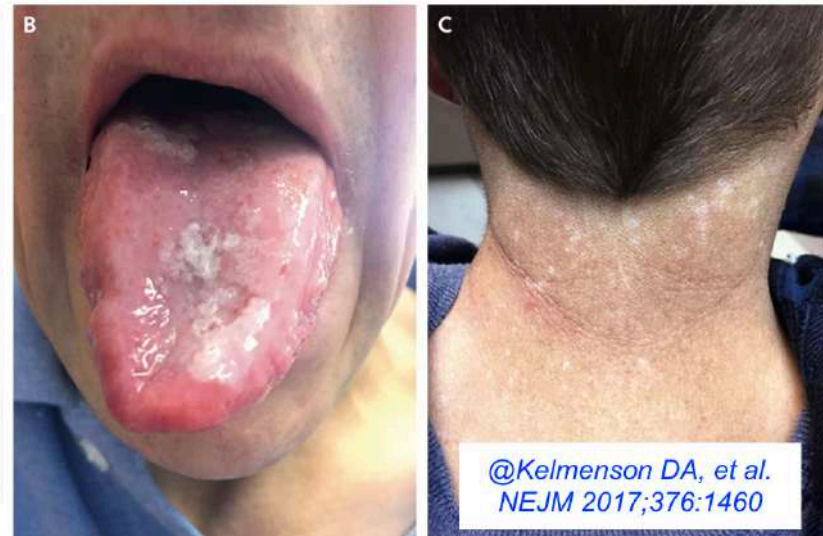
Mother



Grand-Mother



BEWARE of Dyskeratosis Congenita when you see lichenoid nail changes



- Oral leukoplakia, skin pigmentation before the age of 8-10 years
- Average life expectancy of 30 years - bone marrow failure (60%-70%)
- Non hereditary forms, Xrecessive, AR, AD

Hypertrophy of the lateral / distal nail folds

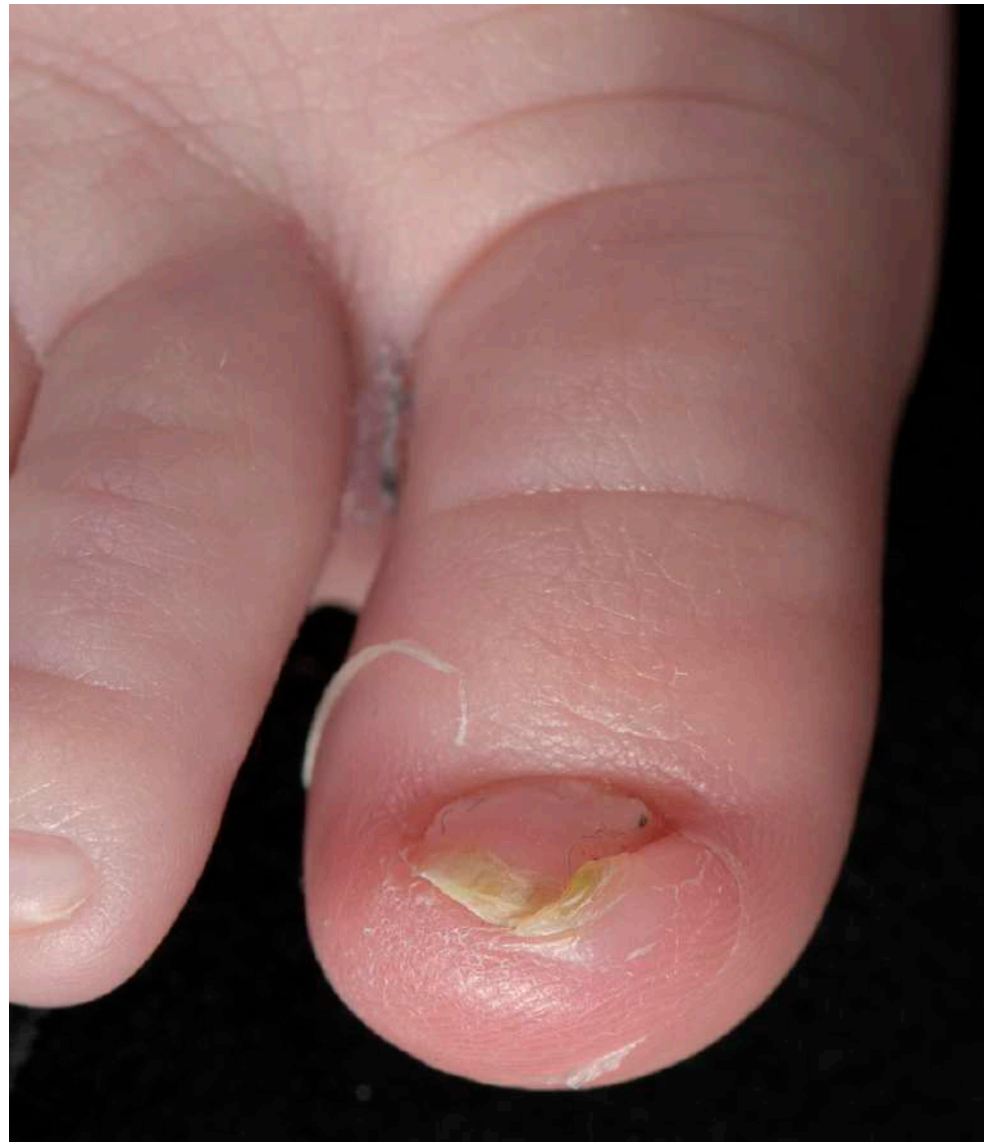
- the lateral / anterior nail fold is hypertrophic and forms a dome-shaped lip that partially covers the nail plate
- lateral nail embedding / nail ingrowing and paronychia
- concomitant koilonychia and congenital malalignment can be seen

POSSIBLE CAUSE:

Lack of pressure
of the thin nail plate



@ Piraccini BM, et al. Pediatr Dermatol 2000;17:348-51





Make the differential with **PSEUDO HYPERTROPHY**

- Normal periungual tissues
- Triangular nail plate



Arai H, et al. Improved conservative treatment of ingrown nail-acrylic affixed gutter treatment, sculptured nail, taping, antibiotic impregnated gauze packing, plastic nail brace, and nail iron. *Rinsho Hifuka* 2003;57:110.



before treatment



vertical taping



diagonal taping



anchor taping



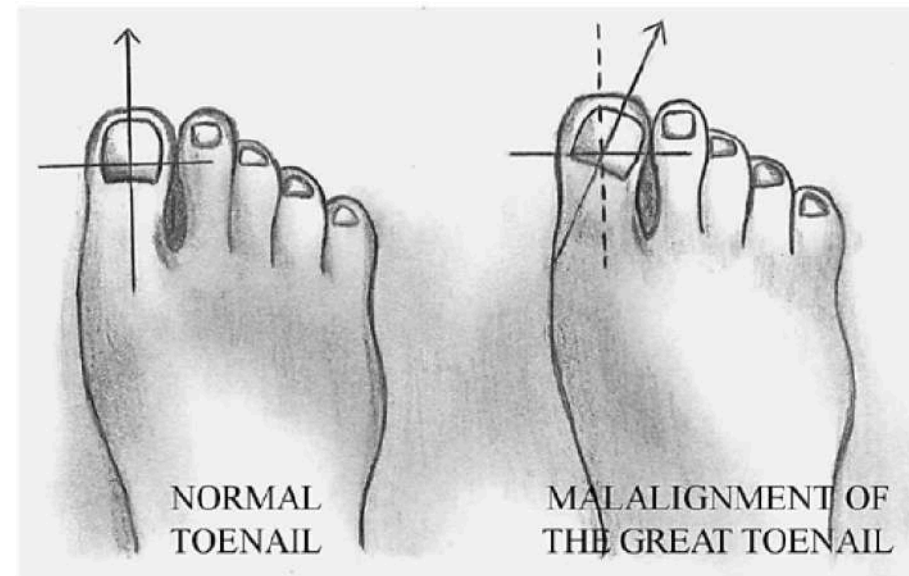
tape fixation



back site

Toenail malalignment

- nail plate deviates laterally from the longitudinal axis of the distal phalanx
- present within the 1st year of age
- different degrees of severity
- lateral nail embedding / nail ingrowing and paronychia as consequences
- typical aspect of the nail plate: short and thick triangular plate with transverse furrows (oyster nails)



Baran R, Bureau H, Sayag J: Congenital malalignment of the big toenail. Clin Exp Dermatol 1979;4:359–60.

Baran R, et al. Congenital malalignment of the big toe-nail as a cause of ingrowing toe-nail in infancy. Clin Exp Dermatol 1983;8:619-23.

Cohen PR. Congenital malalignment of the great toenails: case report and literature review. Pediatr Dermatol 1991;8:43.

POSSIBLE CAUSES:

- ✓ improper in utero positioning?
- ✓ increased tension of hallux extensor tendon?
- ✓ Autosomal Dominant?





Malalignment IS NOT hallux valgus!

Once the nail has lost attachment, the prognosis is worse



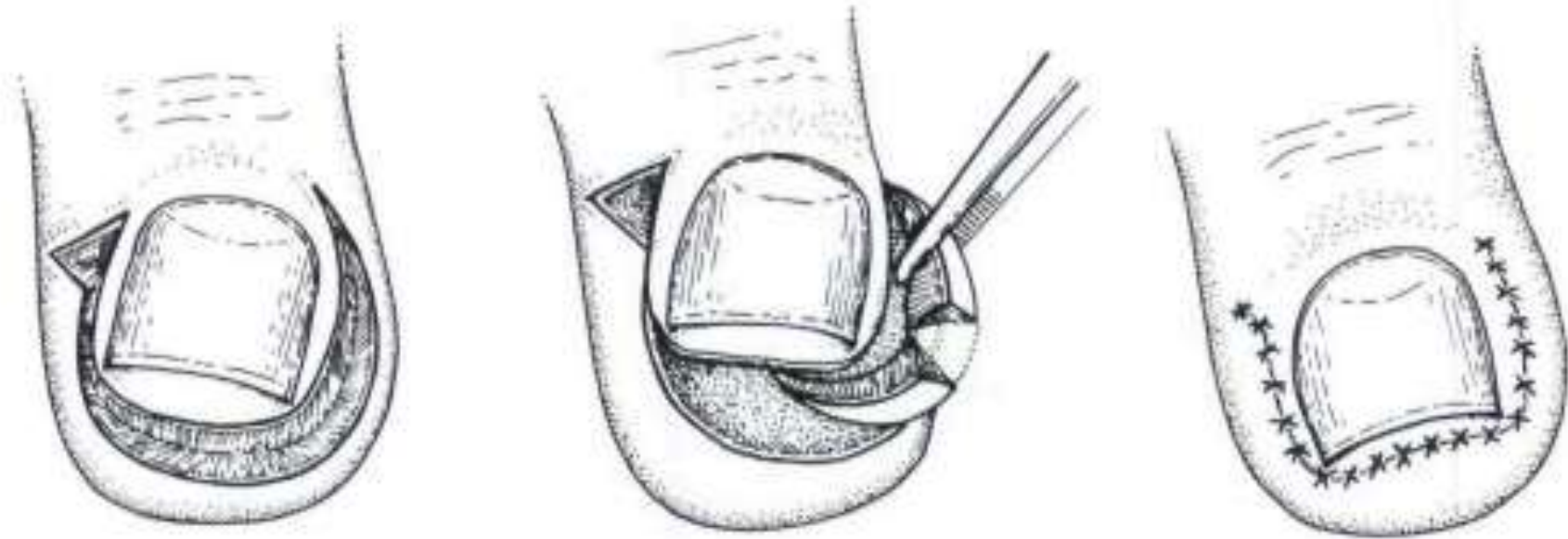


Figure 5. A schematic of the operation recommended for cure of realignment of the big toe nail.



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Parakeratosis pustulosa

- exclusively affecting children
- usually involves a single digit
- family / personal history + for psoriasis / atopy
- spontaneous improvement with age

Most children develop a mild nail psoriasis in adulthood
Patch tests can be useful to rule out contact dermatitis



Psoriasis

- *Pitting and trachyonychia are common in the fingernails*
- *Nail bed hyperkeratosis and onycholysis are common in the toenails*
- **Exclusive nail involvement is rare**
(Fingernails > Toenails)
- Clinical predictor for a more severe disease course over time or arthritis



Piraccini BM, et al. Nail Psoriasis in Children: Common or Uncommon? Results from a 10-Year Double-Center Study. SAD 2015;1:43-8.

Uber M, et al. Nail changes in psoriatic children. Indian J Dermatol Venereol Leprol 2016;82:314-6.

Pourchot D, et al. Nail Psoriasis: A Systematic Evaluation in 313 Children with Psoriasis. Pediatr Dermatol 2017;34:58-63.

Bronckers IMGJ, et al. Nail Involvement as a Predictor of Disease Severity in Paediatric Psoriasis. Acta Derm Venereol 2019;99:152-7.

1. Look at the detachment line

Irregular / dented margin
Whitish to yellow orange color
Absence of fungal spikes

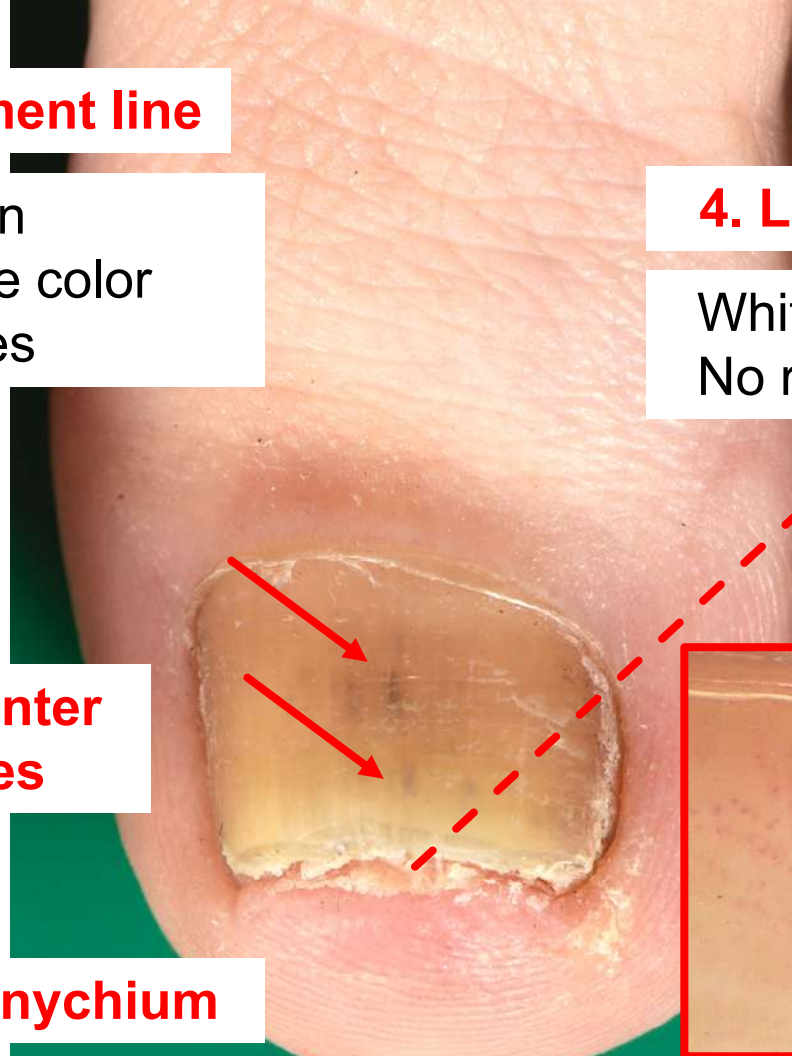
4. Look under the nail plate

Whitish hyperkeratosis
No ruin-like appearance

2. Look for splinter haemorrhages

3. Look at the hyponychium

Capillaries are always visible, tortuous, dotted and irregularly distributed



Consider NAIL CLIPPINGS for a definitive diagnosis

The act of cutting the distal portion of a nail (3mm) and processing the fragment for histopathologic analysis – quick and painless procedure

TABLE 1: Results of the findings of onychomycosis and data collected in the studies of normal nails and nail psoriasis

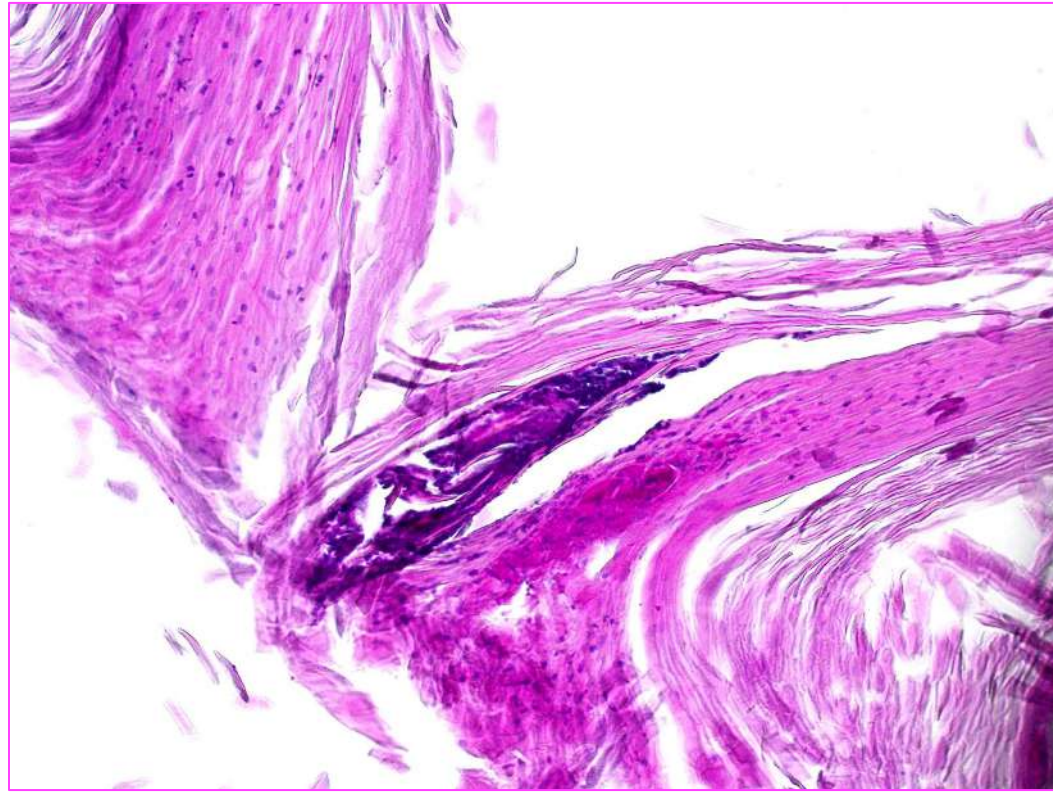
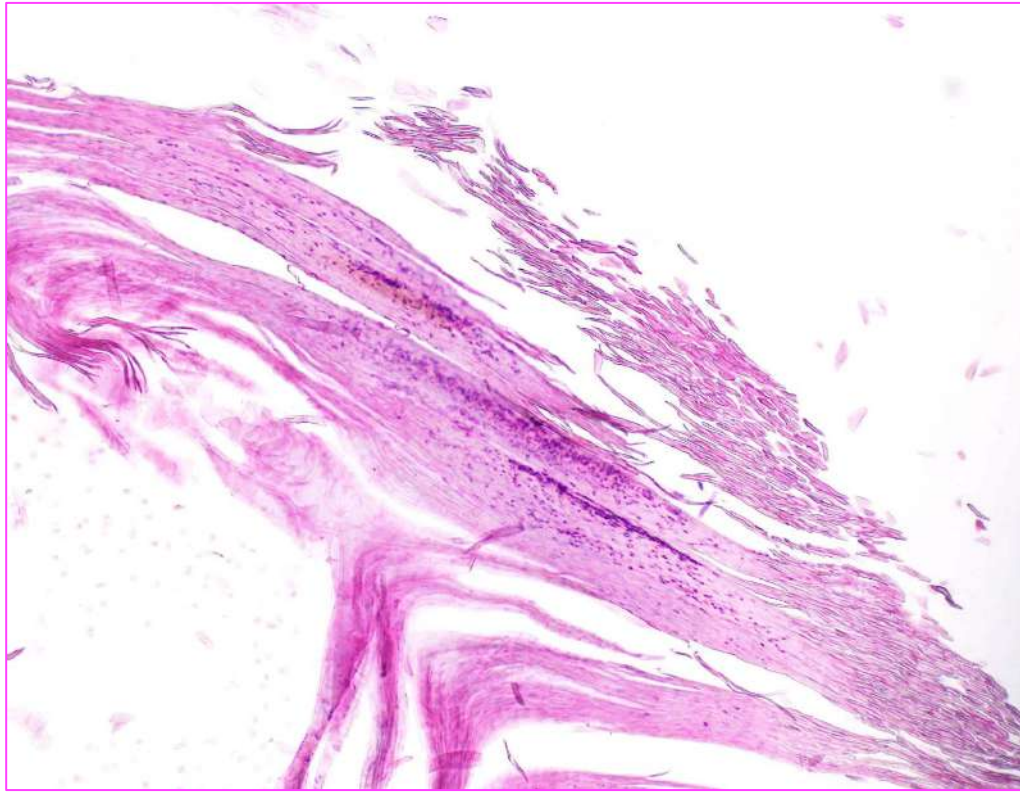
Criteria	Onychomycosis	Normal nails (4)	Nail psoriasis (7)
Number of samples	62	30	50
Plate thickness (mean in mm)	0.59 (0.39–1.3, SD \pm 0.26)	0.36 (0.25–0.50, SD \pm 0.06)	0.44 (0.26–0.85, SD \pm 3.50)
Onychocariosis (%)	53.33	41.00	7.00
Subungual thickness (mean in mm)	0.54 (0.2–1.2, SD \pm 0.09)	0.11 (0–0.31, SD \pm 0.07)	0.19 (0–0.7, SD \pm 0.15)
Serous lakes (%)	64.81	0.00	46.00
Neutrophils (%)	40.74	0.00	12.00
Layers of subungual parakeratosis (mean)	10.8 (0–44, SD \pm 11.8)	5.6 (1–13, SD \pm 3.5)	6.9 (0–36, SD \pm 8.12)
Bacteria (%)	32.73	60.00	74.00

SD: Standard deviation

Uber M, et al. Clinical features and nail clippings in 52 children with psoriasis. Pediatr Dermatol 2018;35:202-7.

Goktay F, et al. Histopathological nail-clipping examination for the diagnosis of isolated childhood nail psoriasis. Turkderm-Turk Arch Dermatol Venereol 2023;57:34-5.

Trevisan F, et al. Nail clipping in onychomycosis and comparison with normal nails and unguinal psoriasis. An Bras Dermatol 2019;94:344-7.



parakeratosis with clusters of polynuclear cells



EPIDERMOLYSIS BULLOSA: great challenge when the only clue is a history of acquired childhood nail dystrophy (big toes).

The AD forms start at birth

The AR appear between the ages of 6 and 8 years



Psoriasis – management

- AVOID traumas of any kind including nail cosmetics; overzealous manicure / pedicure; water contact as much as possible; manual activities
- KEEP THE NAILS SHORT
- CLIP AWAY THE ONYCHOLYTIC PLATE
- Moisturize
- Consider podiatric aid for gait and shoes analysis

*Plachouri KM, et al. Management of Pediatric Nail Psoriasis. Cutis 2021;108:292-4.
Iorizzo M, Tosti A. Updates in treatment and impact of nail psoriasis. Expert Rev Clin Immunol 2023;19:1091-1100.*



MATRIX PSORIASIS



Clobetasol propionate 0,05% (1/day)

Vitamin D derivatives (calcipotriol, calcitriol, tacalcitol) (1-2/day) – *alone or in combination*

Calcineurin inhibitors (1/day)
Azelaic acid (1/day)

Tofacitinib cream? Cyclosporin gel?
Tazarotene gel? Antralin cream? 40% urea?

~~/.L. Triamcinolone acetonide 2.5-5-10 mg/ml~~

~~/.L. Methotrexate 25 mg/ml~~
~~/.L. Cyclosporin 50 mg/ml~~

Methotrexate 5-15 mg/week
Acitretin 0.3-0.5 mg/kg/day

~~Apremilast 60 mg/day~~
~~JAK inhibitors /TYK2 inhibitors~~

Biologics (TNFa/IL17/IL12-23)*

BED PSORIASIS



Vitamin D derivatives (calcipotriol, calcitriol, tacalcitol) (1-2/day) – *alone or in combination*

Calcineurin inhibitors (1/day)
Azelaic acid (1/day)

Tofacitinib cream? Cyclosporin gel?

~~/.L. Triamcinolone acetonide 2.5-5-10 mg/ml~~
~~/.L. Methotrexate 25 mg/ml~~

Cyclosporin 2.5-5 mg/kg/day
Acitretin 0.3-0.5 mg/kg/day

~~Apremilast 60 mg/day~~
~~JAK inhibitors /TYK2 inhibitors~~

Biologics (TNFa/IL17/IL12-23)*

*infliximab, etanercept, adalimumab, ustekinumab, secukinumab and ixekizumab

Intralesional steroid injections for inflammatory nail dystrophies in the pediatric population

Matilde Iorizzo MD, PhD¹ | Nilton Gioia Di Chiacchio MD, PhD^{2,3} |
 Nilton Di Chiacchio MD, PhD² | Chander Grover MD⁴ |
 Shari R. Lipner MD, PhD⁵ | Bertrand Richert MD, PhD⁶ |
 Bianca Maria Piraccini MD, PhD⁷ | Michela Starace MD, PhD⁷ | Antonella Tosti MD⁸

- 3/30 patients < 12 years old
- **Beware of dose-related local adverse events**
- Periungual skin of children thinner than adults
- Max dose 0.2 ml per nail in <15yo ???
- Local anesthetic injections are one of the most anxiety-inducing stimuli in the pediatric patients
- Use should be discouraged

Age range, years (mean)	9-17 (14.6)
PSORIASIS (PSO) NAIL LICHEN PLANUS (NLP)	16 14
N° affected and treated nails	141 (129 Fingernail; 12 Toenail)
DRUG USED Triamcinolone acetonide 2.5mg/ml Triamcinolone acetonide 5mg/ml	6 24
Total dose per nail (mean)	0.1 ml – 0.4 ml (0.19 ml)
Total dose per session (mean)	0.1 ml – 1.8 ml (0.7 ml)
N° OF SESSIONS 1 every 4 weeks 1 every 6 weeks 1 every 8 weeks	18 7 5
ADVERSE EVENTS Systemic Pain mild Pain severe Proximal nail fold atrophy Proximal nail fold hypopigmentation Hematoma& Beau's lines	0 14 6 2 2 1
TREATMENT EFFICACY Resolution Improvement * Worsening	19 (9PSO/10NLP) 9 (6PSO/3NLP) 2 (1PSO/1NLP)

*PSO: from a baseline NAPSI<10 to NAPSI 1-2; NLP: from moderate to mild disease

How to reduce pain from procedure

- **The analgesic effect of vibration/pressure can be explained by the gate control theory of pain**
- Pain is transmitted from the peripheral to the central nervous system through the fast A-delta fibers carrying acute pain, and the unmyelinated slower C fibers carrying chronic pain signals
- When nerves receive non-painful signals such as vibration/pressure, the fast A-alpha and beta fibers activate inhibitor neurons acting on C and A-delta to close the gate on pain signals
- **The brain perceives the vibratory/pressure sensation before the sensation of pain, thus blocking ("gating") the sensation of pain**



Melzack R, Wall PD. Pain mechanisms: a new theory. Science 1965;150:971-9.

Duplisea MJ, Flores K. Using an electric toothbrush for vibration anesthesia during painful procedures. Pediatr Dermatol 2019;36:414-15.

Secil A, et al. Efficacy of vibration on venipuncture pain scores in a pediatric emergency department. Pediatr Emerg Care 2014;30:686-88.

Trachyonychia

- Average age of onset: 3-12 years old
- Hereditary forms described
- Fingernails > toenails - M > F
- Nail plate shows thinning and roughening due to multiple longitudinal ridging and superficial scaling ---> **sandpapered**
- Cuticle hyperkeratosis / Koilonychia associated
- **Diagnosis of the causative disease requires a biopsy**
- Possibly due to psoriasis, atopy/eczema, lichen planus, ...



- The insult to nail matrix keratinocytes impairs their maturative and differentiative activity without interrupting the mitotic activity
- Onychocytes retain their nuclei and, instead of maturing to form a compact layer of tightly adherent flat cells, produce a stratum corneum-like layer that easily desquamates

*Grover C, et al. Dermatol Surg 2003;29:1125-29.
Hur K, et al. J Cutan Pathol 2021;48:396-402.*



Trachyonychia – management

No treatment → evaluate QoL

Topical agents: mometasone 0.1% ointment; betamethasone dipropionate 0.05% ointment; tacrolimus 0.1% gel, tazarotene 0.1% gel; urea – once a day

Intralesional steroids: triamcinolone acetonide 2.5-5 mg/mL – once a month

Systemic agents: oral or i.m. steroids; retinoids



Mittal R, et al. Trachyonychia treated with oral mini pulse therapy. *Indian J Dermatol Venereol Leprol* 2001;67:202-3.

Lipner SR et al. Intralesional triamcinolone matrix injections for treatment of trachyonychia. *J Am Acad Dermatol* 2020;82:e121-2.

Park JM, et al. Efficacy and Safety of Calcipotriol/Betamethasone Dipropionate Ointment for the Treatment of Trachyonychia. *Ann Dermatol* 2015;27:371-5.

Lichen Striatus

- **Cutaneous mosaicism caused by a postzygotic somatic mutation**
- Tolerance until a triggering event induces a self-limiting autoimmune T lymphocyte inflammatory reaction
- Flesh-colored or slightly red flat-topped papules following Blaschko's lines / lichenoid nail changes
- Nail changes improves over the years
- No treatment necessary





Dealing with nail pigmentation

- Which pigmentation we are dealing with?
- We are facing activation or proliferation?
- How many nails are involved? Which ones?
- Age of the patient? Age of the band?
- Triggers? Traumas .. Ethnicity .. Drugs ..

Ronger S, et al. Dermoscopic examination of nail pigmentation. Arch Dermatol 2002;138:1327-33.

Tosti A, Piraccini BM, de Farias DC. Dealing with melanonychia. Semin Cutan Med Surg 2009; 28:49–54.

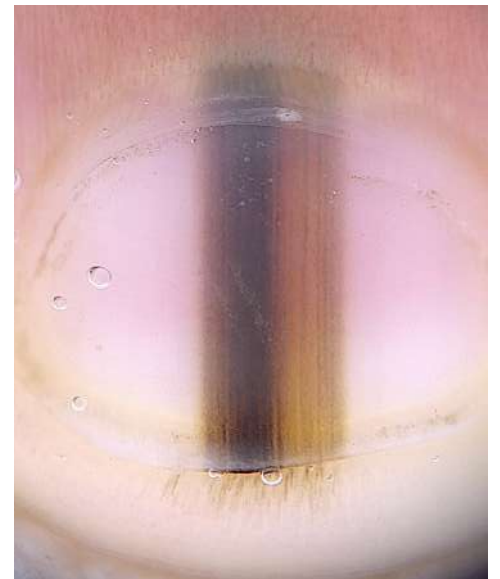




- background color – grey/brown/black
- size and shape of the band
- borders – regular/irregular
- aspect of the lines – homogeneous or not in color, spacing and thickness, regular or disrupted parallelism
- nail plate dystrophies including thinning and splitting
- periungual pigmentation / *vascular abnormalities* / *ulcerations*

Nevi in children have alarming features

- Band often irregular in color/heavily melanized
- Triangular shape (distally thinner)
- Progressive color change (globules/fading/zig-zag)
- Distal nail plate splitting
- Periungual pigmentation (brushing)



Diagnosis difficult even for nail expert pathologists:

- Nuclear atypia of a few melanocytes and mild degree of transepidermal melanocytic migration may be seen – not sufficient to be a sign of malignancy
- Acral benign nevi and Spitz nevi also present these features
- Number of melanocytes per millimeter not yet assessed in children

Goettmann-Bonvallot S, et al. Longitudinal melanonychia in children: a clinical and histopathologic study of 40 cases. JAAD 1999;41:17-22.

Cooper C, et al. A clinical, histopathologic, and outcome study of melanonychia striata in childhood. J Am Acad Dermatol 2015;72:773-9.

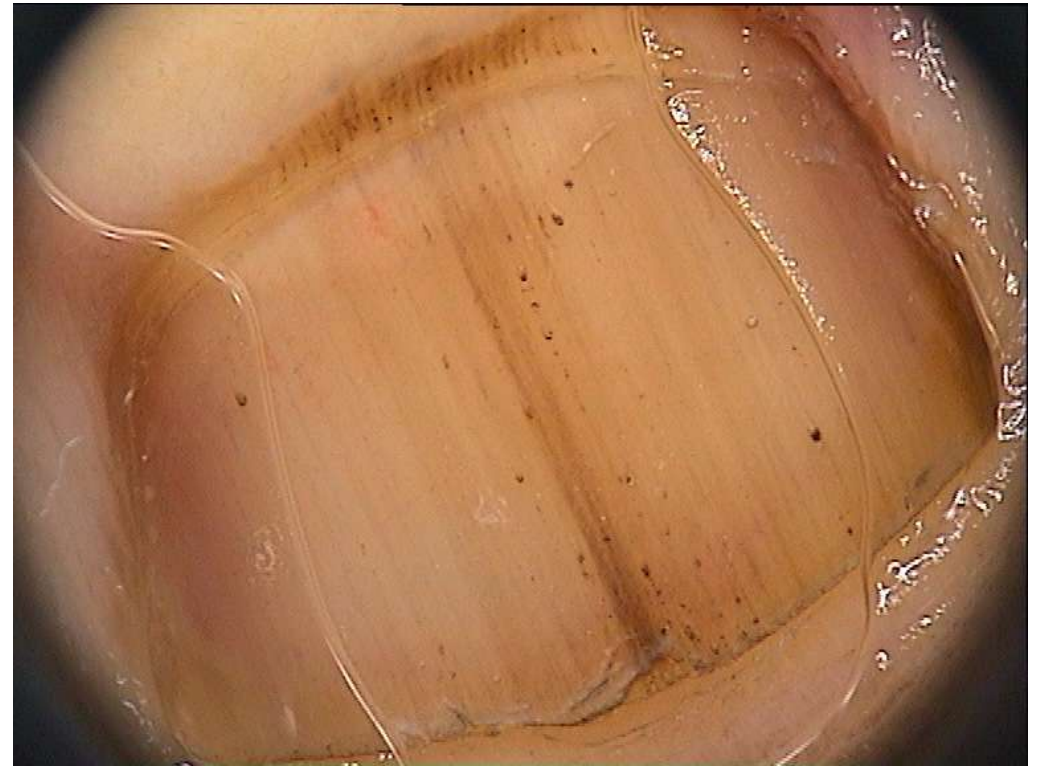
Ohn J, et al. Dermoscopic features of nail matrix nevus (NMN) in adults and children: a comparative analysis. JAAD 2016;75:535-40.

Lee JH, et al. Clinical and histopathologic features of pediatric longitudinal melanonychia: A multicenter retrospective study. J Am Acad Dermatol 2023;88:1393-6.

Lee JH, et al. Clinicopathologic features of 28 cases of nail matrix nevi in Asians: Comparison between children and adults. JAAD 2018;78:479-89.

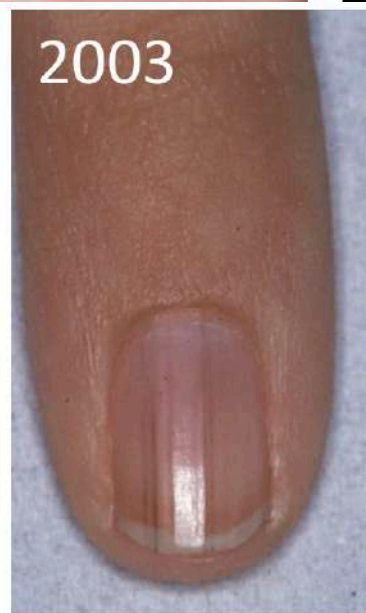
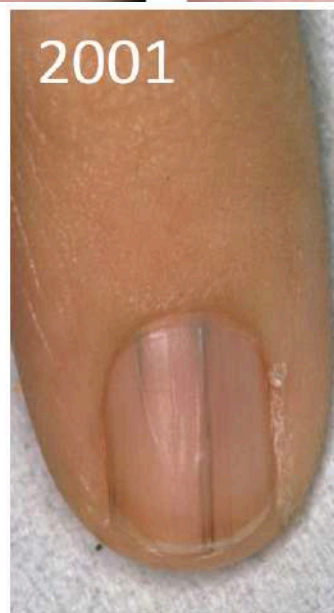
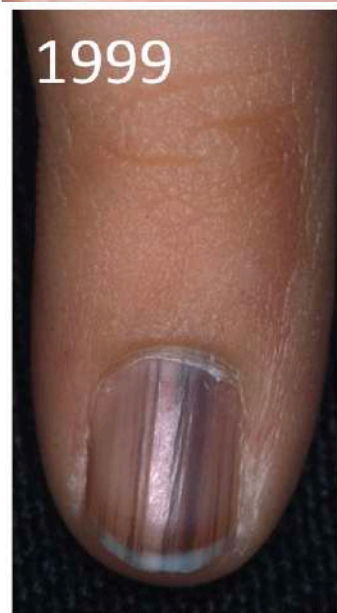
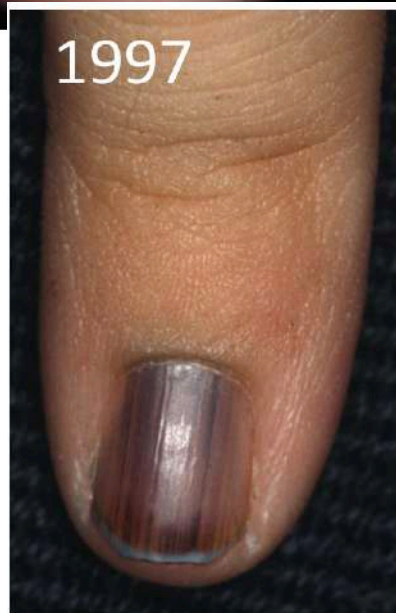
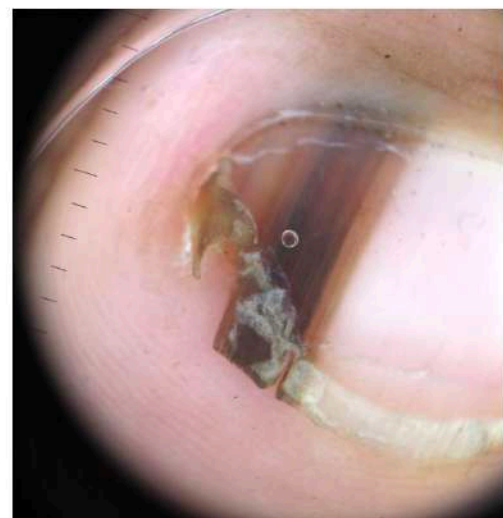
Colin Tan W, et al. Should we biopsy melanonychia striata in Asian children? A retrospective observational study. Pediatr Dermatol 2019;36:864-8.

Matsui Y, et al. Natural course of pediatric longitudinal melanonychia: A retrospective cohort study in Japan. JAAD 2021;86:946-8.



DOTS, cluster of nevus cells that migrate upward from the DE-junction, are a sign of regression in children

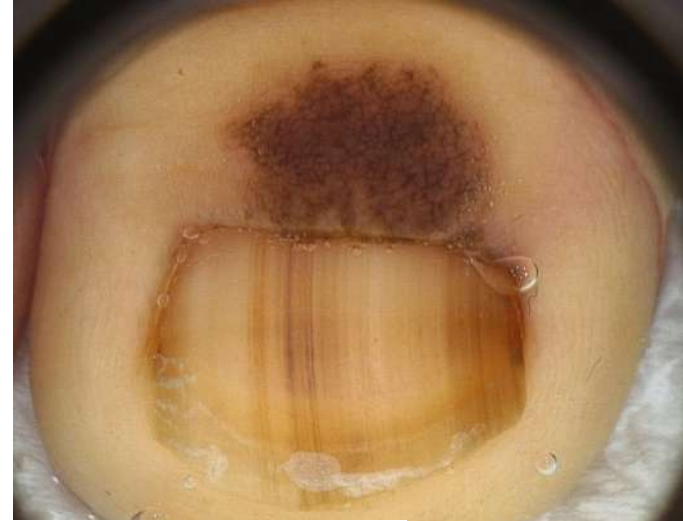
Murata Y, Kumano K. Dots and lines: a dermoscopic sign of regression of longitudinal melanonychia in children. Cutis 2012;90:293-6.



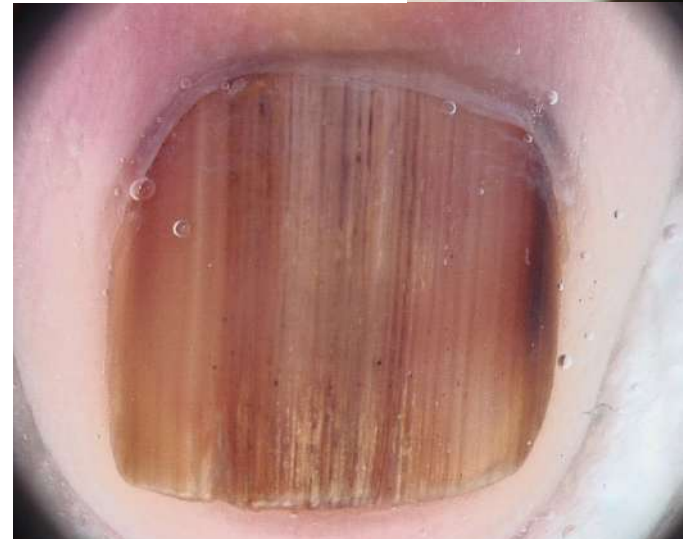
What is the best management?

- Wait and see - follow up every 3-6 months
- Excise when worrisome features are present
- Management should be individualized prioritizing clinical and dermoscopic follow up and considering the family level of anxiety
- If biopsy is chosen, the analysis by a nail expert pathology is mandatory

Thomas L et al. Dermatoscopic and clinical features of congenital (at birth) or congenital-type (< 5yo) nail matrix nevi: A multicenter prospective cohort study by the IDS. JAAD 2022;87:551-8.
Lohman ME, et al. An evidence-based approach to pediatric melanonychia. Dermatol Clin 2022;40:37-49.



Congenital nevus



Junctional nevus



Exostosis

- Outgrowth of normal bone or calcified cartilaginous remains
- Detection of a pathognomonic translocation suggests that exostoses may be true tumors and not merely reactive lesions
- Subungual firm/hard nodule, with superficial teleangiectasias, arising from the dorsomedial aspect of the distal phalanx (toenails > fingernails)
- Onycholysis and (pain) – 55% of cases < 18yo



Storlazzi CT, et al. Rearrangement of the collagen COL12A1 and COL4A5 genes in subungual exostosis: molecular cytogenetic delineation of the tumor-specific translocation t(X;6)(q13-14;q22). Int J Cancer 2006;118:1972-6.

Piccolo V, Starace M, et al. Dermoscopy of subungual exostosis: a retrospective study of 10 patients. Dermatology 2017; 233: 80-85.

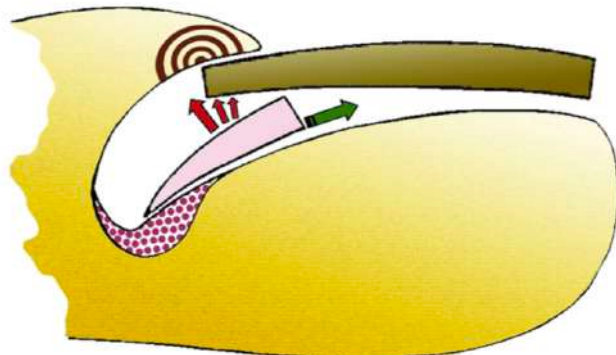
with ingrown toenail



Retronychia: Proximal ingrowing of the nail plate

David A. de Berker, MRCP,^a Bertrand Richert, MD, PhD,^b Edith Duhard, MD,^c
Bianca Maria Piraccini, MD, PhD,^d Josette André, MD,^c and Robert Baran, MD^f
Bristol, United Kingdom; Liège, Belgium; Tours and Cannes, France; and Bologna, Italy

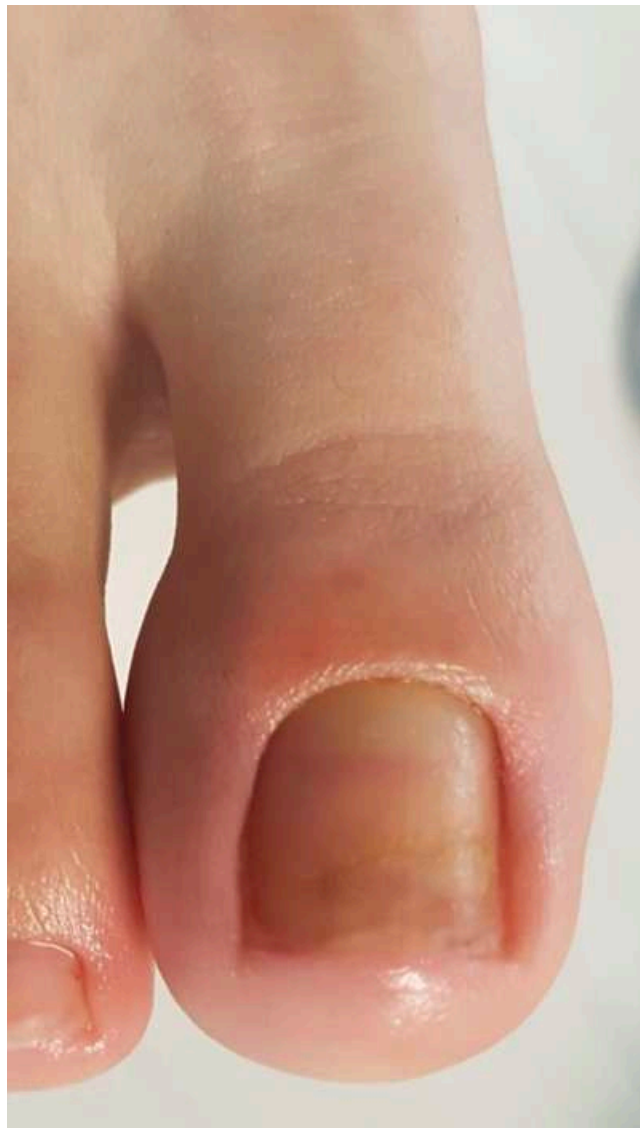
Background: Proximal nail fold inflammation can be caused by many diseases and has not previously been recognized as a result of posterior embedding of the nail. We describe a new pattern of ingrowth that we have termed retronychia (“retro”—Latin for backwards; “onychia”—Greek for nail). **The term describes a combination of proximal nail plate ingrowth into the proximal nail fold which is associated with multiple generations of nail plate misaligned beneath the proximal nail.**



- ✓ Repeated trauma in predisposed subjects
- ✓ Incomplete detachment
- ✓ Growth of a new plate unable to push away the old one incompletely detached and locked by the lateral nail folds
- ✓ Conflict between the plate and surrounding tissues with paronychia and granulation tissue formation



Piraccini BM, et al. Retronychia in children, adolescents, and young adults: a case series. J Am Acad Dermatol 2014,70:388-90.





Retronychia – management



SURGERY

PROXIMAL AVULSION

PRO - Eliminates proximal embedding and inflammation immediately

CONTRA - Patients will be in distress for a while; possible disappearing nail bed

CONSERVATIVE

TOPICAL / I.L. STEROIDS

PRO - Relief of inflammation and pain; at home treatment easy to perform

CONTRA - Long time to see results
Lost of patients at follow up

Sometimes quick decisions create regret ...



Daniel CR 3rd, Tosti A, Iorizzo M, Piraccini BM. The disappearing nail bed: a possible outcome of onycholysis. Cutis 2005;76:325-7





Letter to the Editor

Topical steroids for the treatment of retronychia

A. Lencastre ✉, M. Iorizzo, M. Caucanas, N. Cunha, M.G. Trakatelli, I. Zaraa, M. Henry, R. Daniel, S. Gregoriou, E. Haneke, C. Prevezas, P. Salphale, B.M. Piraccini, M. Starace, A. Tosti, B. Richert
... See fewer authors ^

First published: 01 April 2019 | <https://doi.org/10.1111/jdv.15603> |



- ✓ 56 patients (38 females)
- ✓ 0,05% clobetasol propionate cream under occlusion overnight on the PNF
- ✓ 41% cured & 28.5% partially cured
- ✓ **If no improvements in 10 weeks → avulsion!**



ORIGINAL ARTICLE

DERMATOLOGIC
THERAPY | WILEY

Intralesional steroid treatment for the intermediate stage of retronychia: A pilot study

Aurora Alessandrini  | Andrea Sechi | Yuri Merli  | Francesca Bruni |
Bianca Maria Piraccini | Michela Starace

- ✓ 28 patients (18 females)
- ✓ Intralesional 40mg/ml triamcinolone acetonide (diluted 1:3 with 0.9% NaCl) **1/month x 2-3 months**
- ✓ 27/28 cured & 1/28 avulsion due to a sudden relapse



Nail dystrophy due to toe malposition in children

Authors

[Authors and affiliations](#)

Matilde Iorizzo , Shari Lipner, Tracey C. Vlahovic

- abnormal toe nail plate shape (*hand-fan nail*)
- toe malposition
- difference in shape/size between feet
- abnormal gait
- improper shoes
-





- A healthy toe must lay straight in the shoe and during gait, but if it is uplifted or downlifted the distal free edge of the nail plate tap against the shoe or the ground and this produce thickening, dystrophy and hyperkeratosis as a form of protection
- Weight bearing radiographs better show the pedal joint relationships and position while standing
- **Static and dynamic gait analysis mandatory**
- Surgical methods to straighten the malpositioned toe possibly avoided



Tauveron PH, et al. Generalites sur le pied. Ann Dermatol Venereol 2005;132:741-7.

Sano H, et al Clinical evidence for the relationship between nail configuration and mechanical forces. Plast Reconstr Surg Glob Open 2014;2:e115.



T H A N K Y O U

