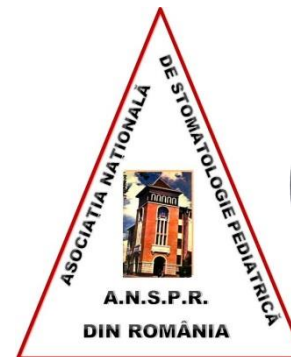




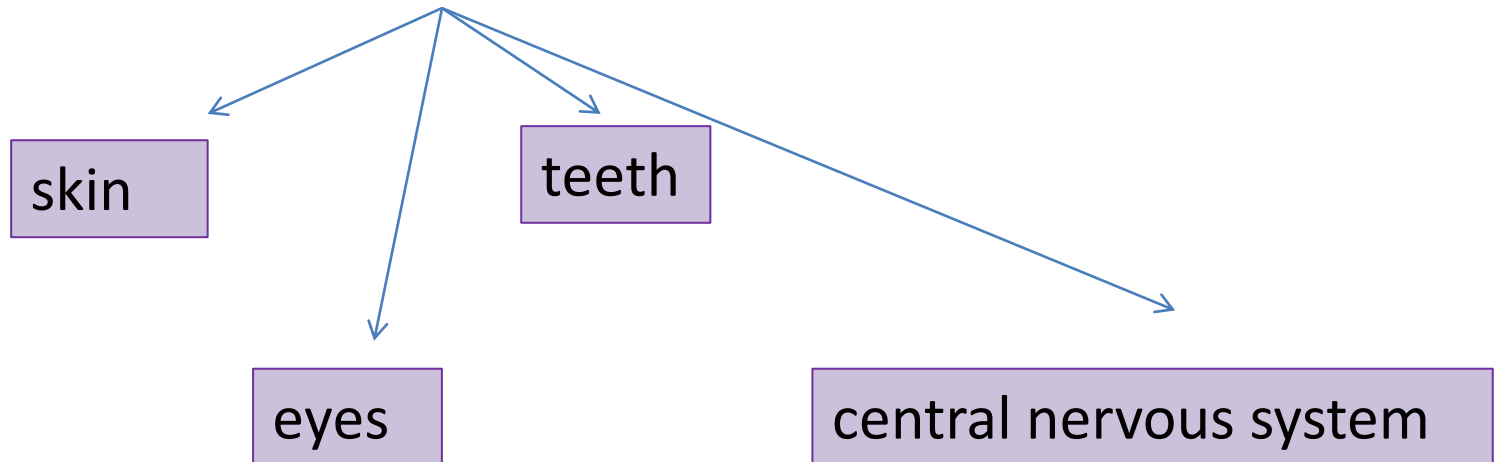
the Child Dental Patient with

Incontinentia Pigmenti (Bloch-Sulzberger syndrome)



Definition

- a rare X-linked genodermatosis, of autosomal dominant character, which affects ectodermal and mesodermal tissues:



Frequency

- 1-9 /100.000 newborns
- Appears almost exclusively in female
- Is usually lethal in males in utero, although survival has been reported in males with a XXY genotype (Klinefelter syndrome) or somatic mosaicism
- Male:female = 20:1
- No ethnic or geographic predominance
- Most cases – in Caucasians

Etiology

- Mutation of the IKBKG (Inhibitor of Kappa B Kinase Gamma) (formerly NEMO) gene, localized on the X-chromosome
- IKBKG gene is required for NF-KB activation pathways that played important roles in inflammation, immune response, cell growth control, and protection against apoptosis.

Cutaneous manifestations

- Stage 1 - vesicular
- Stage 2 - verrucous
- Stage 3 - hyperpigmented
- Stage 4 –atrophic

!! Stages may overlap or not occur at all in a same patient (stages 1 and 3 are more common)

Cutaneous manifestations

Stage 1 (vesicular)

- erythema, vesicles and blisters anywhere on the body but usually spare the face and often respect the midline
- typically linear pattern, along Blaschko's lines
- the lesions develop within the first few weeks of life
- in general, stage 1 has cleared completely by four months.



Borges J et al. Incontinentia pigmenti or Bloch-Sulzberger syndrome: a rare X-linked genodermatosis. *An Bras Dermatol.* 2014;89(3):486-9. Open Access license

Cutaneous manifestations

Stage 2 (verrucous)

- papulae, verrucous lesions and hyperkeratosis
- they usually appear on the distal limbs, as the blisters begin to heal
- the lesions clear completely by six months in over 80% of cases



Ocaña Jaramillo S *et al.* Incontinencia pigmenti. Estudio descriptivo de la experiencia en dos centros hospitalarios. *An Pediatr (Barc)*. 2020;92:3–12. Open Access license

Cutaneous manifestations

Stage 3 (hyperpigmented)

- Linear hyperpigmentation
 - more often apparent on the trunk than the limbs
 - fades and disappears by the end of the second decade



Iordan-Dumitru AD, Luca R - Development dental abnormalities in a child with Incontinentia Pigmenti. Case presentation. *Ro J Stomatol* 2013; LIX(4): 316-9. Open Access license



Kitakawa D *et al.* Incontinentia pigmenti presenting as hypodontia in a 3-year-old girl: a case report. *J Med Case Reports* 3, 116 (2009). Open Access license

Cutaneous manifestations

Stage 4 (atrophic)

- Hypopigmentation and cutaneous atrophy
- Lesions are often present before the hyperpigmentation has disappeared completely
- Until 2 to 3 decades



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Nails & hair abnormalities

- Scarring alopecia, usually on the vertex
- Absence or hipoplasia of eyebrows and eyelashes
- Onychodystrophy – ranges from mild ridging or pitting to onychogryposis



Ocaña Jaramillo S *et al.* Incontinencia pigmenti. Estudio descriptivo de la experiencia en dos centros hospitalarios. *An Pediatr (Barc)*. 2020;92:3–12. Open Access license

Ocular manifestations

- Strabismus
- Microphthalmia
- Cataract
- Optic atrophy
- Retinal lesions – risk of retinal detachment

Neurologic manifestations

- Seizures
- Spastic paralysis
- Microcephaly
- Mental retardation
- Encephalopathy

!! >60% of patients are neurologically normal

Oral features

- Hypodontia
- Delayed eruption
- Impacted teeth
- Morphological anomalies
- Malocclusions

in 54-90% of IP cases



Iordan-Dumitru AD, Luca R - Development dental abnormalities in a child with Incontinentia Pigmenti. Case presentation. Ro J Stomatol 2013; LIX(4): 316-9. Open access license

Hypodontia

- Is the most frequent dental anomaly in IP
- Both in primary and permanent dentition
- Usually, multiple tooth germs congenitally missing
 - Maxilla: lateral incisors, second premolars
 - Mandible: second premolars
- Prolong retention of primary molars .



Chen AY, Chen K. Dental treatment considerations for a pediatric patient with incontinentia pigmenti (Bloch-Sulzberger syndrome). *Eur J Dent.* 2017;11(2):264-267. PMC Open access license

Atypical tooth morphology

- Conical, trapezoidal and peg-shaped teeth
- Additional cusps in posterior teeth
- Microdontia



Kitakawa D *et al.* Incontinentia pigmenti presenting as hypodontia in a 3-year-old girl: a case report. *J Med Case Reports* **3**, 116 (2009). Open access license

Malocclusions

- Class II malocclusions are the most frequent
- Class III malocclusions are developed due to a rotation of the jaw given the need to cover tooth loss
- Anterior cross-bite – as a result of poor maxillary growth



Chen AY, Chen K. Dental treatment considerations for a pediatric patient with incontinentia pigmenti (Bloch-Sulzberger syndrome). *Eur J Dent.* 2017;11(2):264-267. PMC Open access license

Skeletal problems

- Transverse maxillary deficiency associated with oligodontia in the maxilla and mandible
- Prominent chin
- Facial asymmetry, such as facial hemia-trophy and hemifacial hypoplasia.

Palate

- High-arched palate
- Cleft palate (1 of 3 cases) ± cleft lip
- Soft palate hypoplasia

Alterations of the stomatognathic system

- Esthetic appearance
- Masticatory function
- Swallowing disorders
- Speech and language disorders

Dental treatment

- Oral rehabilitation by a multidisciplinary team: pedodontist, orthodontist, prosthodontist
- Aims:
 - improve esthetic appearance
 - Improve occlusal function
 - reestablish masticatory function
- Primary molars has to be preserved as long as they can
- Reshaping of malformed teeth using composite
- Fissure sealant for non-carious molars and premolars.

Dental treatment

- Treatment of hypodontia:
 - fixed or removable prostheses
 - tooth auto-transplantation.

- Definitive treatment - after growth spurt for:
 - better prognosis
 - esthetic result
 - minimal tooth structure preparation.