

Incontinentia Pigmenti - Dental Manifestations: A Report of Two Cases

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ABSTRACT

Introduction: Incontinentia Pigmenti or Bloch-Sulzberger syndrome is a rare form of ectodermal dysplasia, affecting the skin, hair, eyes, central nervous system and the teeth. This variant of genodermatosis is most prevalent in females, with a reported high mortality rates in males.

Case report: Dental abnormalities include hypodontia in both, the primary and permanent dentitions, peg shaped incisors and canines, and delayed eruption. This article reports the clinical features of two children, a 6 year old girl and a rare occurrence of a 12 year old boy diagnosed with Incontinentia pigmenti.

Conclusion: Functional needs of such cases are usually very high, hence a multidisciplinary team is needed to achieve sufficient dental rehabilitation.

Keywords: Incontinentia Pigmenti, Genetic, Dental, Hypodontia, Multidisciplinary

INTRODUCTION

Incontinentia Pigmenti [IP] is a rare X linked dominant neurocutaneous syndrome with cutaneous, neurologic, ophthalmologic and dental manifestations. It displays a high prevalence for females (95%) and is usually lethal in males.¹ Dermatological manifestations usually present at or shortly after birth and is manifested in a series of stages -- first, vesicular or erythematous; second, verrucous; third, hyperpigmented and fourth, scarred.² Dental anomalies are the next most commonly observed findings. Most of the patients presented with hypodontia involving both primary and permanent dentitions; pegging of incisors, canines and premolars; and delayed eruption. IP is also said to affect the nervous system with epilepsy, partial paralysis and retinal detachment not uncommon findings. It is pertinent to note that more than 80% of patients presented with normal or above average intelligence. This case report describes the various dental anomalies observed in two children with a documented history of IP.

CASE REPORT

Case 1: A 6 year old girl reported to the pediatric dental clinic with a complaint of missing primary teeth since birth. The medical history revealed that the patient had a known history of Incontinentia Pigmenti. The family history was insignificant with the mother reporting that the delivery was without complications. The mother reported that a unilateral erythematous rash was noticed in the first week of life on the infant's face and neck. The rash persisted, for the first year and gradually resolved after the first birthday. The child also suffered from ophthalmologic complications, including a constantly increasing power and two retinal detachments

in the past. History revealed that the child had regular milestones.

Intraoral examination revealed multiple missing teeth in the primary dentition. The missing teeth included the maxillary primary lateral incisors and the mandibular right primary second molar. Also the upper and lower incisor teeth showed an irregular conical appearance, characteristic of ectodermal dysplasia (figure 1A,1B,1C). There was no previous history of extractions done in the past; hence it was assumed that the teeth were congenitally missing.

The mother also gave a history of delayed eruption of primary teeth, reporting that the first tooth erupted, at the age of 20 months. A panoramic X-ray was requested to evaluate tooth germ development, and to determine the reason for the missing and irregularly shaped teeth. The orthopantomograph revealed the absence of several primary teeth (52, 62,85) and multiple permanent tooth germs (12,17,22,25,27,35,37,42,44) as well. Insufficient root development of the permanent teeth also indicated that the delayed eruption pattern would seem to continue in the permanent dentition as well (Figure 2A). No evidence of caries was present on examination and the oral hygiene status of the child was satisfactory.

Case 2: A 12 year old boy reported to the pediatric dental clinic with a complaint of multiple missing permanent teeth. The parents gave a history of previously detected Incontinentia Pigmenti characterized by retinal abnormalities, normal intelligence and multiple missing primary and permanent teeth. Incontinentia pigmenti in males is usually lethal and is associated with a survival rate of only 1 of 5 cases. Intraoral examination revealed the absence of all permanent premolars and molars excluding the permanent first molar (Figure 3A,3B,3C). Also there was no previous history of the patient having undergone multiple extractions in the past. Hence it was assumed that the teeth were congenitally missing. An

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Figure-1: A) Anterior view; B) Intraoral view of the Maxillary arch; C) Intraoral view of the Mandibular arch



Figure-2: A) OPG displaying radiographic view of Patient 1; B) OPG displaying radiographic view of Patient 2

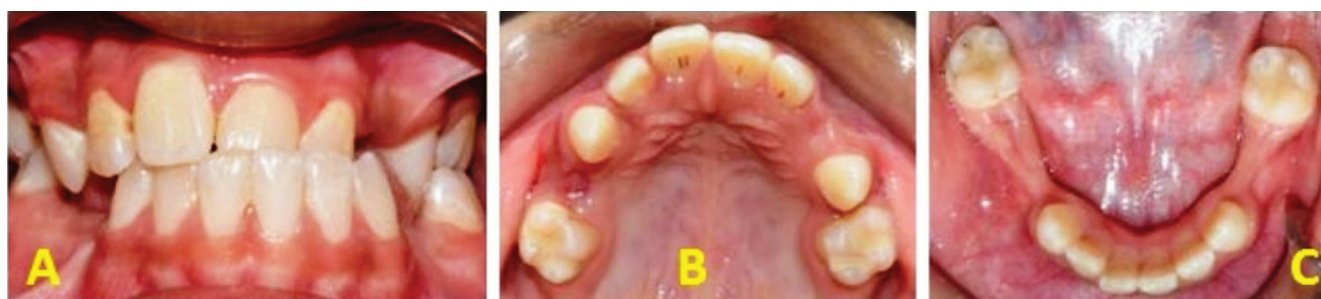


Figure-3: A) Anterior view; B) Intraoral view of the Maxillary arch; C) Intraoral view of the Mandibular arch

orthopantomograph confirmed the clinical findings (Figure 2B). The parents also gave a history of delayed eruption and exfoliation of all primary and permanent teeth.

Intraoral examination also revealed the presence of multiple root pieces and decayed permanent first molars. Dental restorations of the decayed teeth was done using a calcium hydroxide liner (Dycal®, LD Caulk Co, Milford, DE) at the cavity base, followed by a glass Ionomer (GC Fuji VII-GP, GC, Tokyo, Japan) restoration.

DISCUSSION

The name incontinentia pigmenti is derived from the histological characteristics of the disease, that is, melanin incontinence by melanocytes in the basal epidermal layer. IP is a single-gene disorder caused by mutations in the NEMO/IKK- γ gene.¹

Dermatological manifestations are the most common findings and are usually present at or shortly after birth. These are evident in a series of stages -- first, vesicular or erythematous; second, verrucous; third, hyperpigmented and fourth, scarred. The pigmentary disturbance, is usually preceded by a phase suggesting inflammation in the skin. The pigmentation and other residue of skin manifestations gradually resolve and usually disappear by adulthood.³ Ocular abnormalities such as strabismus, myopia are also frequently seen associated with IP, as seen in this case. The

mother also gives a history of retinal detachment and almost complete loss of vision in the right eye. IP is also said to affect the nervous system with epilepsy and partial paralysis not uncommon findings. It is pertinent to note that more than 80% of patients presented with normal or above average intelligence. Neurological manifestations include mental deficiency, microcephaly, spasticity and /or seizures in about 30% of the cases. Hair, nails and eyes are affected in most cases and musculoskeletal disorders may be seen.⁴

Following dermatologic alterations, dental manifestations are the most frequent, which are observed in 80% of patients and usually affects both dentitions.⁵ Hypodontia or partial anodontia is most commonly seen in up to 43% of patients, followed by microdontia, pegged or conically crowned teeth, seen in 30% of cases. Hamartomatous dental cusps and single maxillary incisors have also been reported in earlier literature. These manifestations are important because they persist throughout the patient's life, thus requiring an adequate dental treatment plan from the time of diagnosis of the disease to oral rehabilitation by a multidisciplinary team.⁶⁻⁸

CONCLUSION

Pediatric dentists are probably the first dental practitioners to come in contact with such cases, and it is imperative that emphasis is placed on the need for early diagnosis and

prevention of any further dental problems. Incontinentia pigmenti usually displays numerous symptoms, encompassing various systems, hence it is important that the dentist works in close association with ophthalmologists, dermatologists as well as neurologists. The esthetic and functional needs of such cases are usually very high, hence a multidisciplinary team comprising of a pedodontist, orthodontist and prosthodontist is usually the best option to achieve sufficient dental rehabilitation.

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