

Dental defects in incontinentia pigmenti: case report

Deborah Ann Himelhoch, DDS, MS Bonnie June Scott, DDS
Richard Allen Olsen, DDS

Abstract

Incontinentia pigmenti is an uncommon type of ectodermal dysplasia involving abnormalities of the skin, hair, central nervous system, and teeth. The literature is reviewed and a case with a variety of dental abnormalities is presented.

Incontinentia pigmenti (IP) is one of the lesser known ectodermal dysplasias with fewer than 800 cases reported in the world literature. This genodermatosis affects mostly females and may involve the skin, hair, nails, eyes, and central nervous system.

Diagnosis usually is made from the clinical presentation and occasionally skin biopsy. Characteristic changes in the dentition of some patients also have been observed. Dental abnormalities include hypodontia, pegged incisors and canines, and delayed eruption (Butterworth and Ladda 1981; Freira-Maia and Pinheiro 1984). The authors report a case of a 12-year-old female with incontinentia pigmenti associated with enamel hypocalcification and hypoplasia of the permanent teeth.

Literature Review

Incontinentia pigmenti is a developmental defect involving many structures of ectodermal and mesodermal origin (Morgan 1971). Carney (1976), analyzing 682 cases described in the world literature, found the disorder to be present in all races with a 37:1 female-to-male ratio. Inheritance may be dominant with lethality in males, but this has been disputed (Curth and Warburton 1965; Hecht et al. 1982).

Onset of IP skin lesions usually occurs at or shortly after birth and is manifested in a series of stages — first, vesiculobullous or erythematous; second, hypertrophic or verrucous; and third, pigmented (Carney and Carney 1970; Ianco et al. 1975).

Most children with IP have other abnormalities including partial alopecia of the scalp, strabismus, and dental anomalies. Less common manifestations include nail dystrophies, serious ocular abnormalities, epilepsy,

mental retardation, spastic or paralytic disorders, and leukocyte dysfunctions.¹ It is important to note, however, that more than 80% of patients have normal or above average intelligence.

Fewer than half of the reported cases of IP make any mention of the dentition. Gorlin (1960) reviewed 25 cases with dental abnormalities and reported three new cases. Noted were: hypodontia involving both primary and permanent dentitions; pegging of incisors, canines and premolars; and delayed eruption. Of the 306 cases reviewed by Carney (1976), 64.7% had some major dental abnormality. Of these, 43.1% had hypodontia, 30.4% malformed or pegged teeth (mostly incisors or canines), and only six had enamel disorders. These six patients were reported to have multiple caries and "crumbly" teeth.

Most recently, Burgess (1982) reported on hypodontia in five children with incontinentia pigmenti, three of whom were from the same family. The four older children had hypodontia of the permanent dentition. The infant and three of the four older children also showed hypodontia of the primary dentition. All five of the children had conical teeth and the four older children showed notching of the permanent incisors. Four of the children also showed delayed eruption of some teeth. Changes in the quality of the enamel were not reported.

In the present report, a 12-year-old black female patient was first seen at the University of Chicago Walter G. Zoller Memorial Dental Clinic at the age of three years. Referral was made by the outpatient pediatrics clinic which had been following the patient since age four months. At the time of the initial visit, a diagnosis of incontinentia pigmenti was established. The patient had returned sporadically for dental care and at her last examination presented with numerous carious lesions and other dental problems.

¹ Butterworth and Ladda 1981; Carney and Carney 1970; Carney 1976; Ianco et al. 1975; Jensen et al. 1978; Morgan 1971.

Medical History

The patient was the middle of three children, with one older and one younger brother. The mother denied difficulty with any of her pregnancies and had no miscarriages. Both boys are reported to be free of any signs of systemic illness or abnormalities. Family history was negative for any relative with IP. The mother was a diagnosed epileptic who took anticonvulsant medication sporadically. It is uncertain whether or not she was taking phenytoin during her second pregnancy. The mother has fewer than 10 teeth remaining, and reported that many teeth were extracted due to "cavities." She denied any history of skin lesions and at the time of examination had no visible abnormalities of her face, neck, arms, or nails. Oral soft tissues demonstrated evidence of gingivitis and periodontal disease.

The patient was born full term at an uneventful delivery. A rash was noted in the first week of life on the infant's shoulders and neck. The rash initially was described as bilateral and erythematous. She was sent home at the age of four days although the rash persisted, gradually resolving in the fifth or sixth month of life. The child was left with a "swirled" appearance of her skin with areas of hypopigmentation where the rash had occurred previously.

Currently, the patient has numerous darkly pigmented, slightly elevated lesions of the face extending onto her neck (Fig 1). She has raised areas of hypopigmentation on her hands (Fig 2), but no abnormalities of the nails. Her hair is of normal color, texture, and pattern.

Coats' disease, an ocular disorder characterized by exudation beneath or in the retina, was noted in both eyes at the age of four years. Esotropia of both eyes was present with the right being greater than the left. The patient was legally blind and wore corrective glasses.

A submucosal cleft of the palate was noted in infancy and the patient is followed by the University of Chicago Craniofacial Anomalies Team. She had a long history of chronic otitis media and several cholesteatomas were removed from both ears. The patient currently exhibits normal speech without hypernasality, although speech acquisition was delayed, beginning around age three years. Speech delays were felt to be due to mental retardation. The patient is behind her chronological age in both mental and motor development. She attends a special school in the Chicago Public School System.

Oral Examination and Dental Treatment

An oral examination, including panoramic, periapical, and bite-wing radiographs was completed. Other radiographs dating from 1977 were also available. Soft tissue examination revealed a bifid uvula and gingivitis secondary to poor oral hygiene. All other soft tissues were normal and 25 permanent teeth were present. Radiographs revealed an additional six unerupted per-

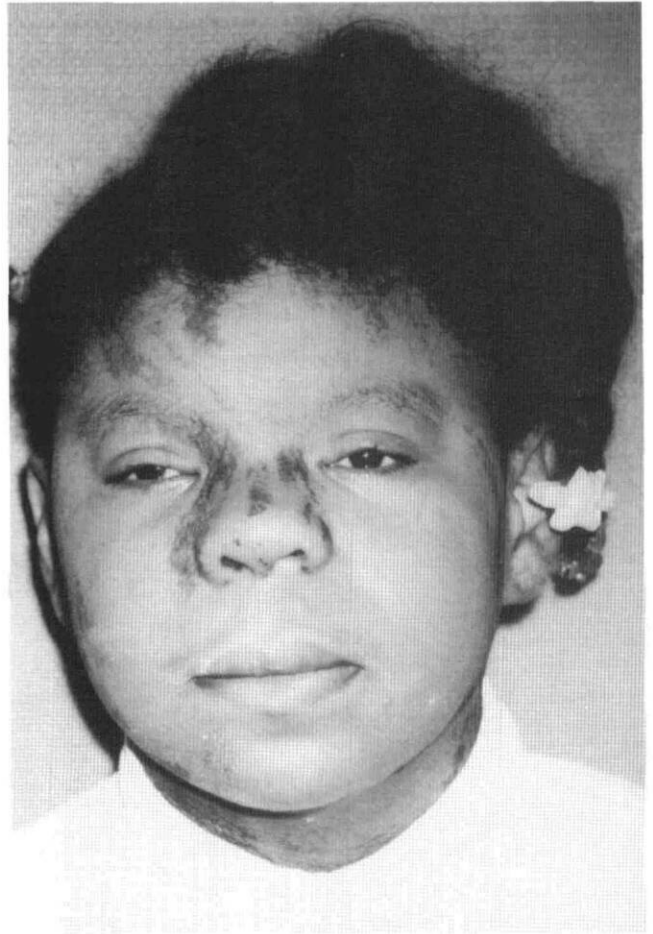


FIG 1. Note the pigmented streaks and whorls characteristic of the third phase of incontinentia pigmenti.

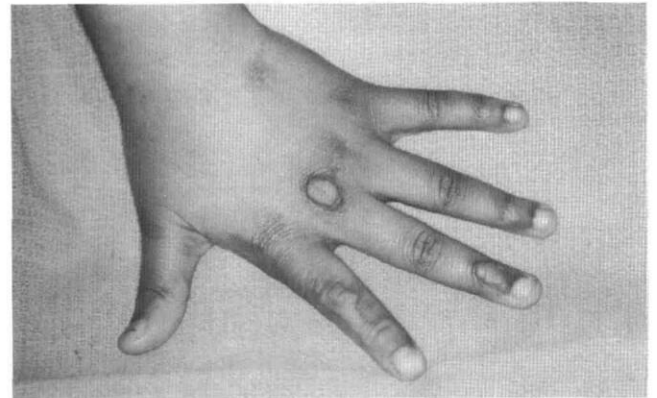


FIG 2. Note the characteristic raised, hypopigmented areas present on the patient's hands.

manent teeth, including four third molars and the maxillary second molars. Radiographs of both primary and permanent dentitions exhibited "tulip-shaped" teeth with bulbous crowns and short, thin roots (Fig 3, next page). A microdont supernumerary tooth was noted in the region of the unerupted maxillary right second and third molars. The mandibular right central incisor was congenitally absent. No primary teeth were present.

First molar occlusion was end-to-end with an edge-to-edge incisor relationship. Crowding was present in the maxillary anterior region, with rotation of the maxillary left central and lateral incisor. There was no residual space in the mandibular arch despite the missing incisor (Fig 4). The maxillary right first molar was somewhat microdontic, resembling a large premolar (Fig 5). Vertical notching of the incisal edges of the maxillary right central and left lateral incisors was noted. Areas of hypocalcification, chalky white or milky yellow in appearance, were present on maxillary right lateral and left central incisors, left canine, maxillary right first and left second premolars, maxillary left first molar, mandibular right and left canines, and mandibular right first molar. Enamel pitting was noted on the maxillary right canine and lateral incisor, maxillary left first premolar, and both mandibular canines. Several restorations were present and new carious lesions were noted on the maxillary and mandibular molars and premolars. These lesions were restored in amalgam. Carious lesions on the buccal surfaces of maxillary right canines and left second premolar were restored with composite resin.

Discussion

The patient in this report demonstrated several of

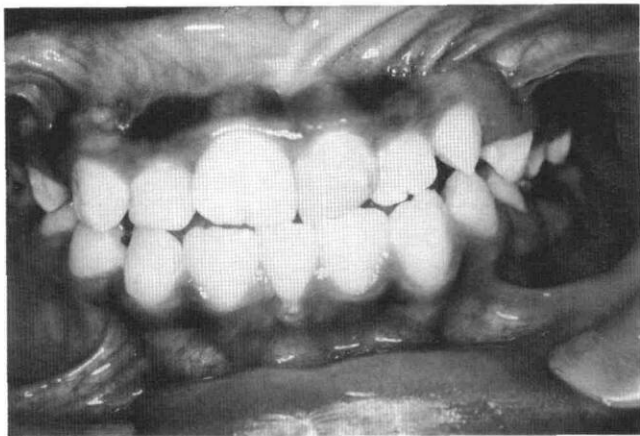


FIG 4. Note the congenitally absent mandibular central incisor and notching of the maxillary right central and left lateral incisors.

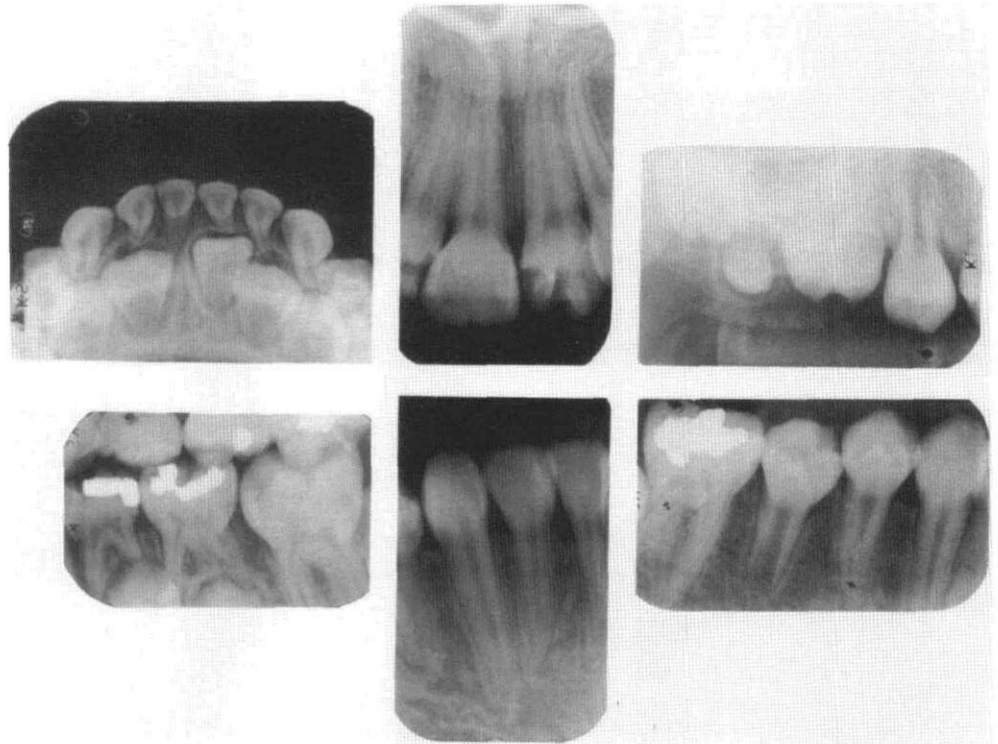


FIG 3. Selected radiographs from ages 3-12 showing patient's unusual "tulip-shaped" teeth as well as the impacted supernumerary tooth distal to the small maxillary right first permanent molar.

the typical features of incontinentia pigmenti. The erythematous stage began shortly after birth and persisted 5-6 months. The patient is currently in the third or pigmented stage, although verrucous lesions persist on her hands. She does not have abnormalities of the hair or scalp, but ocular abnormalities are present and severe. The patient exhibits some mental and motor retardation, an uncommon but not unreported finding.

The finding of a submucosal cleft palate is unusual and is not a reported part of the disease. It may be a coincidental finding, but cleft lip and palate have been reported in association with other forms of ectodermal dysplasia (Freire-Maia and Pinheiro 1984). It is possible that the submucosal cleft palate in this case is an uncom-

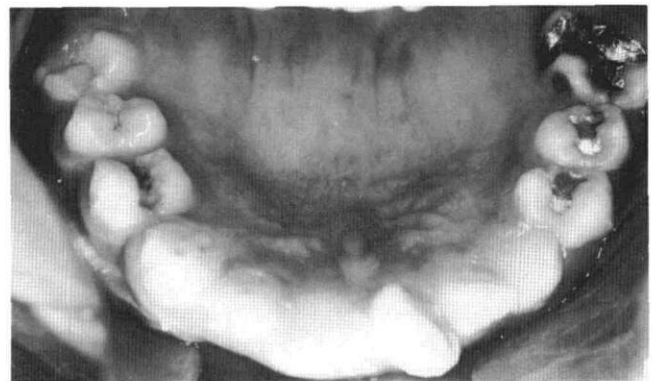


FIG 5. Note the small maxillary right first molar compared to the normal-sized left first molar.

mon manifestation of the genetic defect that causes incontinentia pigmenti.

Etiology of incontinentia pigmenti in this case is probably a sporadic mutation. Although the mother had lost most of her teeth, she had no evidence of enamel defects or malformations of the remaining teeth. No skin abnormalities were present, nor was there a history of any abnormalities of ectodermal structures. Finally, no family members were reported to have incontinentia pigmenti or other dermatological diseases.

The most commonly reported dental anomalies in IP patients are hypodontia and malformed or pegged teeth (usually anteriors). This patient presented with enamel hypoplasia, enamel hypocalcification, a supernumerary tooth, abnormally formed teeth, and a congenitally missing tooth. Most striking, however, was the extensive pitting, hypoplasia, and hypocalcification present on 12 of the 25 erupted teeth. Previously restored carious lesions on another 10 teeth may have been due to similar enamel defects.

Due to the patient's youth, conservative alloys and composites were chosen to restore carious teeth. Non-carious, unrestored molars and premolars were protected with occlusal sealants. The apparently unaffected enamel surfaces seemed to etch normally and sealants were intact several months after placement. However, because of fear that the enamel may be abnormal, care was taken to achieve mechanical retention for all acid-etch composite restorations. The supernumerary tooth was removed and orthodontic treatment is in progress. The child has received oral hygiene instruction and has been advised to rinse daily with a neutral sodium fluoride rinse. Plaque indices are improving and it is hoped that there will be little future dental decay.

At the time of writing Dr. Himelhoch was a clinical assistant professor of pediatric dentistry, Dr. Olsen was an assistant professor and head of general dentistry, and Dr. Scott was a general practice resident, University of Chicago's Zoller Dental Clinic. Dr. Scott also is in private practice in Lake Zurich, Illinois. Reprint requests should be sent to: Dr. Deborah A. Himelhoch, 46 Waverley St., 1st Floor, Belmont, MA 02178.

Burgess MC: Incontinentia pigmenti: 6 cases of Bloch-Sulzberger syndrome. *Br Dent J* 152:195-96, 1982.

Butterworth T, Ladda RL: *Clinical Genodermatology*, vol 1. New York; Praeger Scientific, 1981 pp 65-70.

Carney RG, Carney RG Jr: Incontinentia pigmenti. *Arch Dermatol* 102:157-62, 1970.

Carney RG: Incontinentia pigmenti: a world statistical analysis. *Arch Dermatol* 112:535-42, 1976.

Curth HO, Warburton D: The genetics of incontinentia pigmenti. *Arch Dermatol* 92:229-35, 1965.

Freire-Maia N, Pinheiro M: *Ectodermal Dysplasia: A Clinical and Genetic Study*. New York; Alan R. Liss Inc, 1984 pp 77-79.

Gorlin RJ, Anderson JA: The characteristic dentition of incontinentia pigmenti. *J Pediatr* 57:78-83, 1960.

Hecht R, Hecht BK, Austin WJ: Incontinentia pigmenti in Arizona Indians including transmission from mother to son inconsistent with the half chromatid mutation model. *Clin Genet* 21:293-96, 1982.

Iancu T, Komlos L, Shabtay F, Elian E, Halbrecht I, Book JA: Incontinentia pigmenti. *Clin Genet* 7:103-10, 1975.

Jessen RT, Van Epps DE, Goodwin JS, Bowerman J: Incontinentia pigmenti: evidence for both neutrophil and lymphocyte dysfunction. *Arch Dermatol* 114:1182-86, 1978.

Morgan JD: Incontinentia pigmenti (Bloch-Sulzberger syndrome): A report of four additional cases. *Am J Dis Child* 122:294-300, 1971.

Hepatitis B traced to dentist

The ninth outbreak of hepatitis B virus (HBV) traceable to a dentist since 1974 resulted in 4 cases of the disease in a New Hampshire town.

The outbreak was traced to an oral surgeon who had performed tooth extractions, without gloves, 3-5 months before each patient became ill with the virus. All 4 patients claimed no other risk factors for HBV; all were serologically confirmed.

While the oral surgeon did not wear gloves, he was careful to scrub his hands between surgical procedures. Following discovery of the outbreak, the doctor, in practice for 25 years, discontinued practicing and sent letters to all patients informing them of possible exposure and offering free testing for HBV.

The Centers for Disease Control, in its report about this incident, concluded, "Recurrent, avoidable outbreaks such as this one should prompt dentists and oral surgeons to seek hepatitis B vaccination and to use gloves routinely when treating patients."