Clinical diagnosis and management strategies of amelogenesis imperfecta variants

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Abstract

Amelogenesis imperfecta (AI) is a group of inherited disorders primarily affecting dental enamel. Variants of AI generally are classified as hypoplastic, hypocalcified, or hypomaturation types based on the primary enamel defect. The aim of this study was to analyze the clinical presentations, diagnostic features, and clinical complications of different variants of AI. Thirty-two patients from 17 families with several subtypes of AI were studied. The results showed that distinctive clinical features may be observed in each variant. However, all AI patients suffered common clinical problems of poor esthetics, teeth sensitivity, and loss of occlusal vertical dimension. The mildest problems were found in the pitted hypoplastic type whereas the most severe problems were encountered in the hypocalcified type of AI. Management strategies include composite resin veneers and jacket crowns for anterior teeth as well as steel crowns for posterior teeth. Knowledge of the clinical features and dental complications of each variant of AI helps in the diagnosis of the condition and allows institution of early preventive measures. (Pediatr Dent 15:384–93, 1993)

Introduction

The term "amelogenesis imperfecta" (AI) now is reserved for those developmental enamel defects inherited primarily as defects of the enamel only.[1] The prevalence of this condition has been estimated to range from 1 in 718[2] to 1 in 14,000,[3] depending on the population studied. The etiology of AI is thought to be alteration of the genes involved in complex processes of enamel formation and maturation.

A few classifications of AI, based on clinical appearance of the defects as well as the inheritance patterns have been proposed in the past,[1,4,5] the most recent and comprehensive being suggested by Witkop[1] (Table 1). In general, the defects in AI may be classified as hypoplastic, hypocalcified, or hypomaturation types, depending on the stage of enamel formation that is primarily affected.[1] The hypoplastic types are characterized by a deficiency in the quantity of enamel, which may be expressed clinically as thin enamel or pits or grooves on the enamel surface.[1,4,5] By contrast, the hypocalcified varieties are characterized by enamel that is insufficiently mineralized, and appear clinically as soft, discolored enamel that is easily removed.[1,4,5] The hypomaturation types of AI are associated with abnormalities of the maturation stages of enamel formation, resulting in the enamel being opaque and chalky in appearance.[1,4,5] As shown in Table 1, autosomal dominant, autosomal recessive, and X-linked modes of inheritance have been reported.

Although recent research has made significant advances into the diagnosis of a few types of AI by molecular[6–8] and biochemical[9] methods, these sophisticated techniques are not yet routinely available. Currently, diagnosis of the different AI variants rests mainly on the dental clinical presentations and their modes of inheritance as determined from family pedigrees. Accurate diagnosis is clinically important for several reasons. First, it is important to exclude the presence of certain systemic diseases that may show generalized enamel hypoplasia as accompanying signs.[10–14] Second, accurate diagnosis enables genetic counselling,[15] which is often sought by affected families. Third, accurate diagnosis leads to the recognition of clinical problems that are associated with the condition, so preventive measures may be instituted early. Fourth, diagnostic

Table 1. Classification of amelogenesis imperfecta according to Witkop (1989)

Type I		**Hypoplastic**
	IA	hypoplastic, pitted autosomal dominant
	IB	hypoplastic, local autosomal dominant
	IC	hypoplastic, local autosomal recessive
	ID	hypoplastic, smooth autosomal dominant
	IE	hypoplastic, smooth X-linked dominant
	IF	hypoplastic, rough autosomal dominant
	IG	enamel agenesis, autosomal recessive
Type II		**Hypomaturation**
	IIA	hypomaturation, pigmented autosomal recessive
	IIB	hypomaturation, X-linked recessive
	IIC	snow-capped teeth, autosomal dominant?
Type III		**Hypocalcified**
	IIIA	autosomal dominant
	IIIB	autosomal recessive
Type IV		**Hypomaturation-hypoplastic with taurodontism**
	IVA	hypomaturation-hypoplastic with taurodontism, autosomal dominant
	IVB	hypoplastic-hypomaturation with taurodontism, autosomal dominant

differentiation of the many variants of AI may help to determine the type of restorations[16–19] that are most successful.

Although the genetic defects in the X-linked form of AI now have been linked to amelogenin genes on the X-chromosome,[6–8] the molecular defects associated with the other types of AI are still unclear. Hence, the diagnosis of AI currently rests largely on clinical criteria.

With the exception of a few epidemiological investigations,[3, 20, 21] previous studies of AI have been mainly case reports of individuals or small numbers of families.[22–38]

The aim of the present study was to analyze the clinical presentations and dental complications in a group of affected patients to determine the distinct clinical features of each variant.

Patients and methods

The study subjects were all referred to the author over the past few years for dental management of enamel hypoplasia, and diagnosed as having AI by the author. A total of 32 subjects (16 males and 16 females) from 17 different, unrelated families were available for study. At the time of initial dental examination, their mean age was 12.8 ± 5.6 years (range 7.2–34.5 years).

All the patients were examined at the University of Queensland Dental School. The teeth were dried, and a mirror and probe used for the dental examination. Erythrosin disclosing dyes were painted on the enamel of some patients to demonstrate the surface defects. Bite-wing and panorex radiographs were exposed as part of their routine dental management. The results of the dental examinations were recorded in comprehensive charts.

Table 2. Characteristics of families with the hypoplastic variants of amelogenesis imperfecta

Family	*Hypoplastic Variant*	*Likely Mode of Inheritance*	*Clinical Features*	*Clinical Problems*		
				Poor Esthetics	*Sensitivity of teeth*	*Loss of OVD*
1	Pitted	AD	Small discrete pits on all surfaces. Normal contact between teeth. Normal radiographic contrast of enamel and dentin.	+	0	0
2	Pitted	AD	As above.	+	0	0
3	Pitted	AD	As above. Extensive loss of enamel on occlusal of primary molars.	+	+	+
4	Smooth	AD	Thin, smooth, hard, glossy enamel. White to yellow-brown in color. No contact between teeth. Radiographs show thin enamel.	++	++	+
5	Smooth	AD	As above. No contact between teeth. Radiographs show thin layer of enamel.	++	+	+
6	Smooth	XL	Females show vertical bands of alternating normal thick, and abnormal thin enamel in both primary and permanent dentitions.	+++	0	0
7	Smooth	XL	Females show above. Males shows uniformly thin, smooth enamel.	+++	++ (Males)	++ (Males)
8	Smooth	XL	All females show above.	+++	0	0
9	Smooth	XL	In addition to above, one male shows anterior open bite.	++	+	+
10	Rough	AD	Thin, hard enamel with rough surfaces. Minimal contact between teeth. Radiographs show thin enamel.	++	0	0
11	Rough	AD	As above. Primary dentition shows thin, less rough enamel.	++	0	0
12	Local	AR	Horizontal pits and grooves of missing enamel in the middle third of the crowns of all permanent teeth. The enamel present shows hypomaturation defects.	++	+	0

AD = autosomal dominant; AR = autosomal recessive; XL = X-linked; OVD = occlusal vertical dimension; + = mildly affected; ++ = moderately affected; +++ = severely affected.

Table 3. Characteristics of families with the hypocalcified and hypomaturation variants of amelogenesis imperfecta

Family	*Likely Mode of Inheritance*	*Clinical Features*	*Clinical Problems*		
			Esthetics Affected	*Sensitivity of Teeth*	*Loss of OVD*
Hypocalcification					
14	AR/XL	Enamel appears soft, opaque white-yellow upon eruption. Early loss of enamel. Minimal contact between teeth. Radiographs show enamel loss and lack of contrast between enamel and dentin.	+++	++	++
15	AD	As above.	+++	+++	++
16	AR/XL	As above.	+++	+++	++
Hypomaturation					
17	XL/AR	Thin enamel with mottled opaque-white discoloration. Enamel may chip away. Normal contact between teeth. Radiographs show thin enamel and less contrast between enamel and dentin. Mild anterior open bite present.	+	0	+

AD = autosomal dominant; AR = autosomal recessive; XL = X-linked; OVD = occlusal vertical dimension; + = mildly affected; ++ = moderately affected; +++ = severely affected.

For every proband, a family pedigree chart was constructed. In an affected family, examination of as many family members as possible was performed. Dental management outcomes are not part of the study design.

Diagnosis of amelogenesis imperfecta

A diagnosis of AI was based on the following criteria: 1) generalized enamel hypoplasia of both the primary and permanent dentition; 2) family history of the condition, although in the recessive forms, or new mutations, there may be no previous history; 3) absence of systemic diseases that may cause generalized enamel hypoplasia resembling AI (e.g. systemic disorders involving calcium metabolism such as renal and liver disorders).[10, 11]

In addition, the trichodentoosseous (TDO) syndrome (kinky hair, dysplastic nails, sclerotic bones, enamel hypoplasia, severe taurodontism),[39–41] which shows hypocalcification enamel defects, was excluded.[13] Variants of ectodermal dysplasia, which may also show generalized enamel hypoplasia,[12, 42] as well as fluorosis[43] also were excluded.

Results

Tables 2 and 3 show the 17 families in the study, and the type of variant diagnosed in each case. Twelve families showed the hypoplastic variety. Three of these were classified further as having the pitted hypoplastic type, another seven, the smooth hypoplastic type, and two, the rough hypoplastic type. In addition, one family showed the local hypoplastic variety.

There were three families with the hypocalcification type of AI, and another one that showed the hypomaturation variety.

Pitted hypoplastic AI (autosomal dominant)

Clinical features. Five affected children from the three families with the pitted type of AI (Table 2) all showed classical features of small, discrete, pinpoint-to-pinhead sized pits, which were arranged in horizontal or vertical rows (Fig 1a). In areas of the teeth subjected to occlusal stresses, there were localized areas of enamel loss. Contacts between the teeth were normal.

Defects in the primary dentition in this form of AI may be demonstrated in an affected female child from family #3 (Fig 1b). The defects in the thinner enamel of

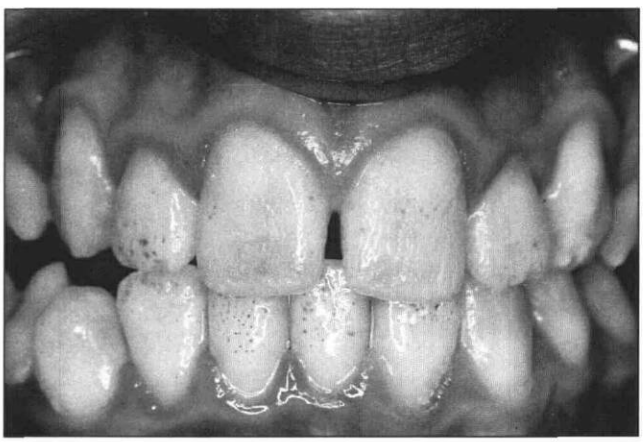

Fig 1a. Permanent teeth of a male patient from family #1 with the pitted type of AI.

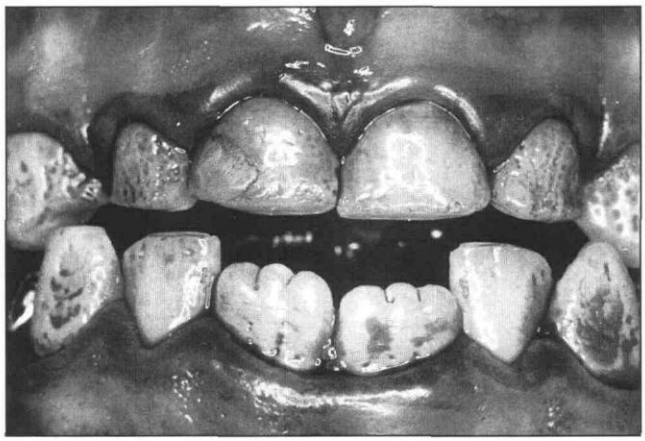

Fig 1b. Anterior teeth of patient from family #2 stained with erythrosin dye. The surface pits may occur in vertical rows as shown on the labial surfaces of the primary lateral incisors.

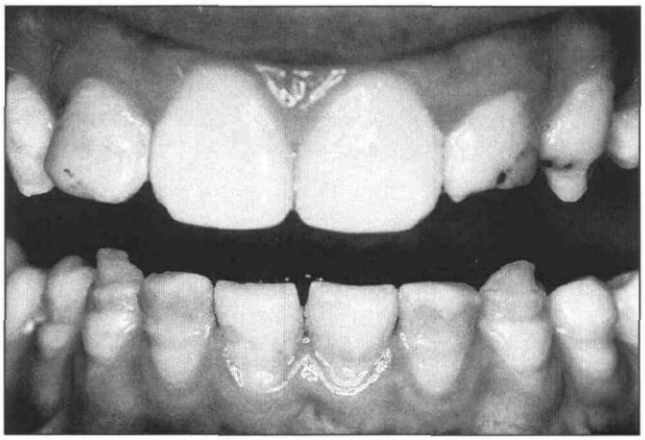

Fig 2a. Dentition of a male patient (family #12) affected with the local hypoplastic type of AI. Restorations had been placed previously on the labial surfaces of the maxillary central incisors.

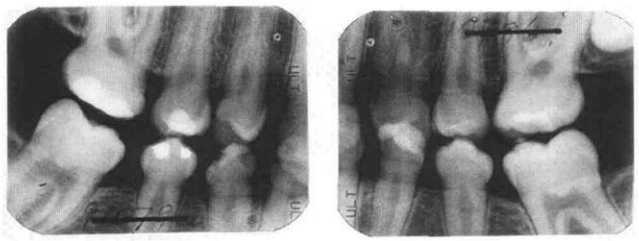

Fig 2b. Bite-wing radiographs of the patient depicted in Fig 5a, showing bulbous appearance of the crowns and constricted cervical areas due to lack of enamel at these areas of the crowns. There is also complete coronal pulp calcification.

primary teeth may be less conspicuous compared with the permanent dentition. In this patient, staining the teeth with erythrosin-disclosing dye clearly demonstrated the pitted hypoplastic defects.

Radiographs of affected teeth in the pitted type of AI showed normal enamel thickness and normal contrast between enamel and dentin.

In all the families, autosomal dominant modes of inheritance were demonstrated.

Clinical problems. Minor esthetic problems were encountered in two patients who showed mild staining of the enamel pits. Except for one patient who had extensive loss of enamel of her primary teeth, none complained of sensitivity. There was also little potential for loss of occlusal height. Mild gingivitis was noted in many patients.

Local hypoplastic AI (autosomal recessive)

Clinical features. One male patient presented with the local hypoplastic type of AI (Family #12, Table 2). All his permanent teeth showed hypoplastic defects that occurred as horizontal bands of pitted or missing enamel (Fig 2a). In the entire dentition, the enamel appeared opaque. Radiographs revealed normal enamel thickness (Fig 2b). Of interest is the complete calcification of the coronal parts of the pulp chambers, as well as the bulbous appearance and constricted cervices of the crowns, which may be related to the lack of enamel at the cervical areas (Fig 2b).

As there was no history of the condition in other members of the family, it was postulated that the mode of transmission was autosomal recessive or X-linked recessive. Alternatively, it may represent a new genetic mutation in the family.

Clinical problems. The patient complained of poor esthetics and some sensitivity to hot and cold. There was little potential for loss of occlusal vertical height, and minimal gingivitis.

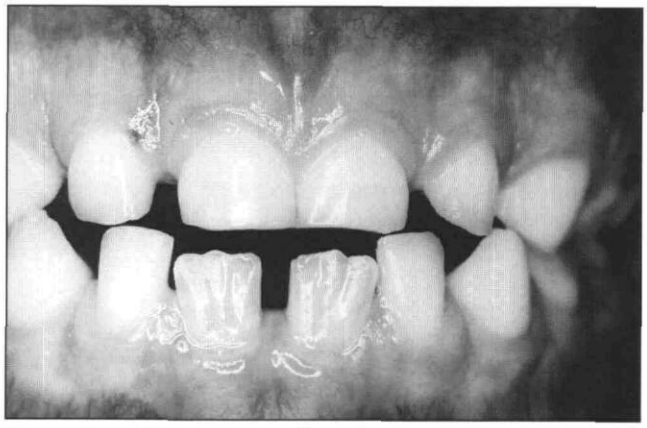

Fig 3a. Dentition of patient from family #4 affected with the smooth thin hypoplastic type of AI.

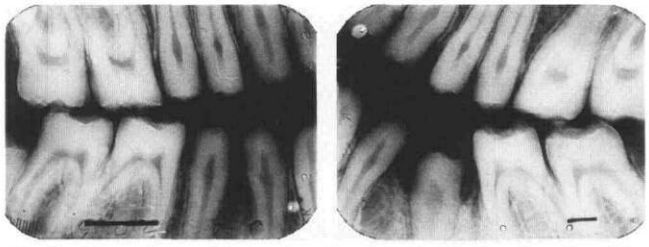

Fig 3b. Bite-wing radiographs of patient in Fig 2a, showing thin enamel and spacing between the teeth.

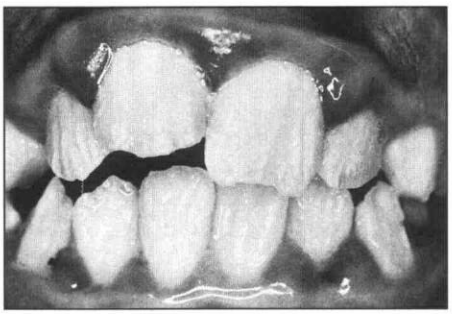

Fig 4a. Mixed dentition of a female patient (family #6) affected by the smooth hypoplastic (X-linked) type of AI showing the classical vertical striping effect of alternating bands of normal and affected enamel.

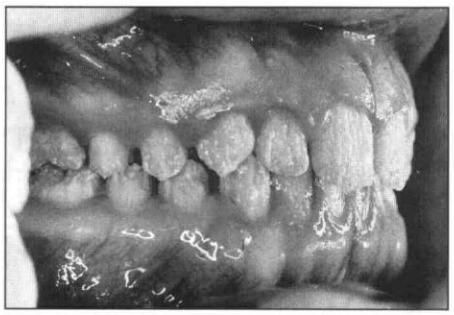

Fig 4b. Permanent dentition of an affected female patient from family #8 (X-linked smooth hypoplastic AI) clearly depicting the vertical striping effect on the facial surfaces of the entire dentition.

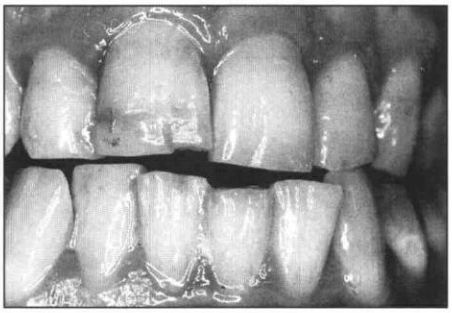

Fig 4c. Dentition of mother of proband in family #7 affected by the X-linked smooth hypoplastic type of AI. Note the mild vertical ridging of the enamel surface.

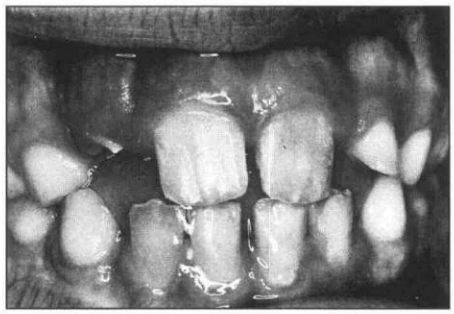

Fig 4d. Early mixed dentition of male proband in family #9 showing uniformly thin enamel surface, in direct contrast to the vertically ridged enamel seen in his mother (Fig 4c).

Smooth, thin hypoplastic AI (autosomal dominant)

Clinical features. Seven patients from three families with the autosomal dominant type of smooth hypoplastic type of AI were available for examination (Table 2). All the patients showed thin, hard, smooth, and glossy enamel, which varied in color from white to cream-brown (Fig 3a). The teeth appeared narrow in all dimensions and there were no contacts between the teeth. Radiographs of the affected patients showed a thin layer of enamel outlining the crowns (Fig 3b).

In the three families studied, autosomal dominant patterns of inheritance were observed. This variant may be distinguished from the smooth, hypoplastic X-linked dominant type by the fact that in the autosomal dominant variant, both sexes are affected equally to the same extent, whereas in the X-linked type, males are affected more severely.

Clinical problems. All affected patients with the smooth hypoplastic AI complained of poor esthetics as well as moderate dental sensitivity. Enamel loss on occlusal surfaces in the older, untreated patients was severe, leading to potential problems of loss of vertical dimension. Gingival health in this group of patients appeared better compared with the rough hypoplastic types, most probably due to the enamel surfaces being relatively smooth.

Smooth hypoplastic AI (X-linked)

Clinical features. A total of seven affected females and two males from four separate families were diagnosed as having the X-linked type of hypoplastic AI (Table 2). In this variant, the affected females classically showed alternating vertical bands of normal and hypoplastic enamel (Fig 4a), an effect known as Lyonization,[15,16] which may be seen in X-linked conditions. The expression of the enamel defect in vertical ridges and grooves was seen also in the primary dentition although the effects may not be as pronounced. Radiographs of the affected females revealed the enamel to be thin. Contacts between the teeth may vary, depending on severity of the defect. In two of the families (#6 and #8), the expression in the females was severe (Figs 4a, 4b). However, in the other two families (#7 and #9), only faint vertical ridging of enamel was noted in the mother (Fig 4c) and maternal grandmother of the male proband, indicating that in this family, the females were affected only to a mild degree. The teeth had normal contacts and the radiographs showed only mild thinning of the enamel.

By contrast, the enamel defects in affected males in families #7 and #9 were severe, and manifested as uniformly thin and smooth enamel (Fig 4d). Furthermore, the teeth appeared small and no contacts existed between the teeth. In addition, radiographic appearance of the teeth in affected males usually showed the enamel to be extremely thin or nonexistent.

The classical differences in clinical manifestations between the sexes, as well as the family pedigrees, indicated that in these families the most likely mode of inheritance was X-linked. For example, in family #6, an affected father has transmitted the condition to all of his daughters and none of his sons. By contrast, an affected mother may transmit the condition to half her daughters and half her sons.

Clinical problems. Poor dental esthetics was suffered by all male and female patients who showed extensive vertical grooving of the enamel. However, females tended to complain less of dental sensitivity and suffered less dental destruction and loss of occlusal vertical dimension. Most affected patients showed severe gingival inflammation.

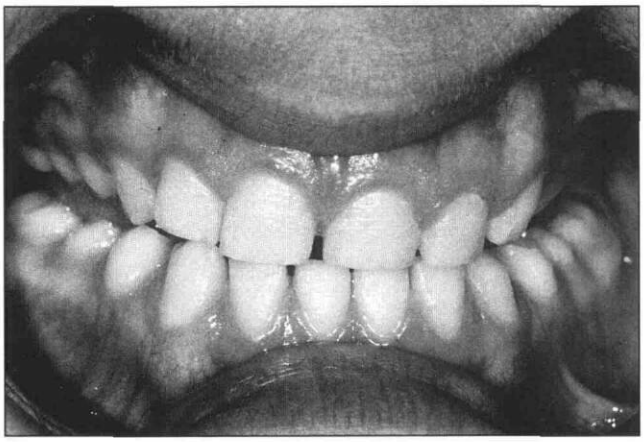

Fig 5. Dentition of an affected female from family #11 with hypoplastic type of AI showing thin, rough enamel.

Rough hypoplastic AI (autosomal dominant)

Clinical features. Two families (#10 and #11, Table 2) presented with the autosomal dominant, rough hypoplastic type of AI. In this variant, both sexes are affected to the same degree and presented with similar features of thin, hard, rough-appearing enamel (Fig 5). There were minimal contacts between the teeth, and radiographs revealed thin enamel that had normal radiographic contrast with dentin.

In the primary dentitions of the study patients, the defects were not as obvious as in the permanent dentition, particularly in the anterior teeth. However, in the posterior primary teeth, moderate loss of tooth structure and dental caries were noted on the occlusal surfaces. In all the three families showing this type of AI, the most likely mode of inheritance was autosomal dominant.

Clinical problems. Poor dental esthetics resulting from stained and rough teeth was the chief complaint of most patients. Sensitivity of the teeth, as well as loss of occlusal vertical dimension, were not as great as the hypocalcified types. Severe gingivitis was noted in most patients.

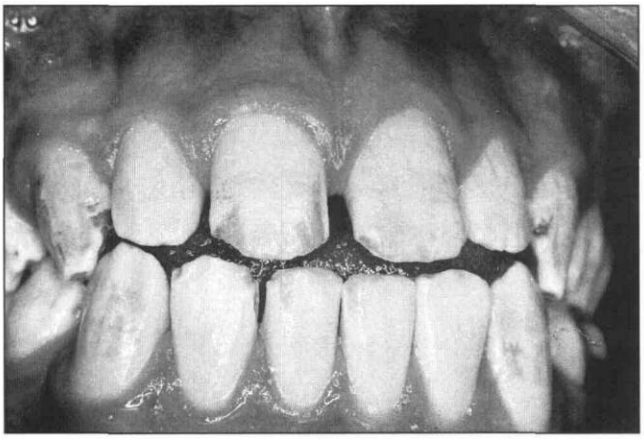

Fig 6. Dentition of a male patient (family #17) affected by the hypomaturation type of AI. Note similar appearance of teeth to dental fluorosis.

Hypomaturation AI (autosomal recessive/ X-linked recessive)

Clinical features. A male patient (family #17, Table 3) presented with the hypomaturation-type AI. He showed typical features of opaque white enamel with areas of hypoplasia in the entire permanent dentition (Fig 6). There were normal contacts between the teeth. A mild anterior open bite was present. Radiographs revealed lack of contrast between enamel and dentin, although the enamel thickness appeared normal. Since the appearance of the enamel defects was similar to dental fluorosis, this possibility was excluded from history, as well as the radiographic appearance of enamel.

The patient appeared to be the only affected member of his family. Thus the mode of inheritance may be postulated to be either autosomal recessive or X-linked recessive or a new genetic mutation in the family.

Clinical problems. Dental esthetics of the patient was only mildly affected. There was no sensitivity of the teeth, and little potential for loss of occlusal vertical dimension through loss of tooth structure.

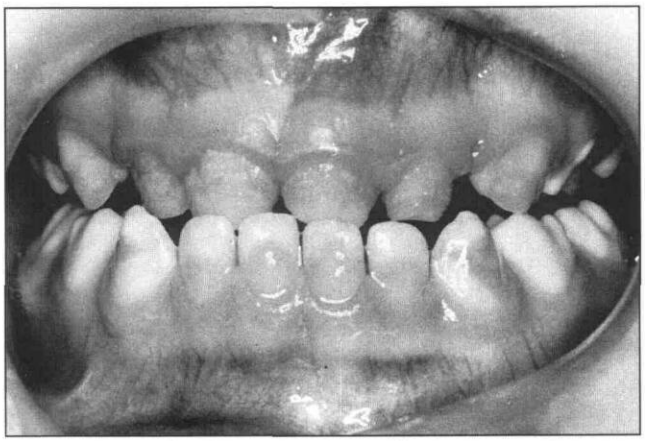

Fig 7. Primary dentition of patient with the hypocalcified AI (autosomal dominant) showing total loss of enamel on maxillary anterior teeth and opaque discoloration of remaining enamel on other teeth.

Hypocalcified AI (autosomal recessive/ autosomal dominant)

Clinical features. Three families (#14–#16, Table 3) presented with the hypocalcification-hypoplastic type of AI. In all affected members of families showing this variant of AI, the enamel typically appeared soft, opaque, and yellow-white upon eruption (Fig 7). It tended to chip away easily, particularly on the facial surfaces, exposing large areas of dentin. There were adequate contacts between the teeth. The primary dentition appeared as severely affected as the permanent dentition with large areas of enamel missing from most of the primary teeth.

Radiographically, in all cases, there was minimal contrast between enamel and dentin, and the enamel thickness ranged from normal to thin.

In one of the families (#15, Table 3) with this type of AI, an autosomal dominant mode of inheritance was evident. However, in the two remaining families (#14 and #16, Table 1), the proband in each case was male and there were no previous histories of the condition in the families. It may be postulated that in each of these cases, an autosomal recessive, or an X-linked recessive mode of inheritance or a new genetic mutation is possible.

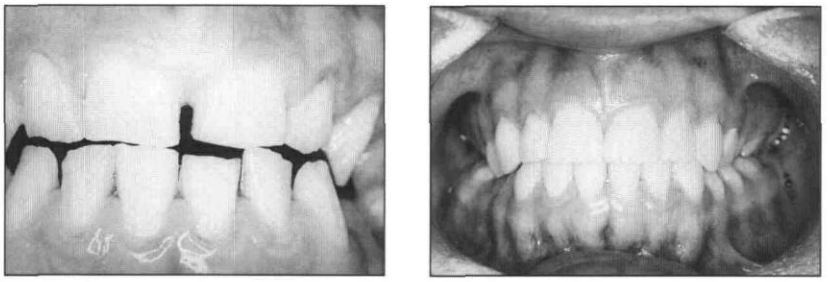

Fig 8a, 8b. Anterior teeth of patient presenting with the rough, hypoplastic AI(left) successfully restored with composite resin facings (right).

Clinical problems. All affected patients complained of extremely poor dental esthetics and moderate levels of sensitivity to hot and cold. In the older, untreated patients, there was excessive loss of occlusal vertical height. Also, it was noted that margins around previous amalgam restorations were defective due to the fracture of supporting tooth structure.

Discussion

Diagnostic difficulties

Since the current classifications of AI variants are based mainly on clinical presentations and patterns of inheritance of relatively few patients, revision may be necessary as new knowledge becomes available. The current classification systems dividing the enamel defects into hypoplastic, hypocalcified, and hypomaturation types may cause difficulties in identifying some variants that simultaneously show clinical features of two or more groups (e.g., hypoplasia is often noted in the hypocalcified groups). Overlapping features also have been identified both micromorphologically[18–20,22] and microradiologically.[44,45] Some AI variants such as the pitted hypoplastic type are clinically distinctive and easily diagnosed. Others such as the X-linked variants may be more difficult to diagnose due to their presentations in a few phenotypes, as well as the existence of striking differences in expression between males and females.

In addition, the unavailability of dental data from certain family members, as well as incompleteness of pedigrees, may compromise accurate diagnosis in many AI patients. Furthermore, the modes of inheritance in many small families may be difficult to determine, particularly in the recessive types. Also, in many variants of AI, such as the X-linked varieties, the modes of inheritance are still not clearly established.[46,47]

Management of oral complications

The families in this study represented the hypoplastic subtypes IA, IC-IF, hypomaturation subtype IIB, and hypocalcification subtypes IIIA and IIIB in Witkop's classification[1] (Table 1). The relative prevalence of each type is comparable to those found in previous reports.[2,4] In addition to delineating further the distinctive phenotypic features of AI variants, this study compared the different complications that may be encountered in each type. This may have value in planning effective preventive and restorative strategies for managing each variant.

In this study, it was found that the main clinical problems of AI in general were esthetics, dental sensitivity, and loss of occlusal vertical dimensions through loss of dental structure. The severity of dental problems experienced by the patients, however, varied with each type of AI. The hypoplastic variants tended to be associated with less severe clinical problems, with the mildest problems encountered in the pitted hypoplastic type of AI. By contrast, the patients with the hypocalcified type of AI usually presented with the most severe clinical problems.

Poor dental esthetics. Poor dental esthetics in AI was usually the result of surface roughness, staining, and abnormal crown shapes from enamel loss. Several strategies may be used to overcome the compromised esthetics. In the patients with hypoplastic types of AI, there is usually sufficient enamel available for bonding so that composite resins veneers may be used to mask the staining and improve the crown morphology (Fig 8a, 8b). However, in patients affected by the hypocalcified varieties of AI, enamel is usually insufficient for direct bonding, and dentin bonding resins[23] or glass ionomer cements[34] are first required to bond to the underlying dentin before applying the veneer of composite resins. Other anterior veneers using porcelain are also likely to be useful, particularly if sufficient enamel is available for bonding; however, their use in AI teeth has not been evaluated.

Porcelain jacket crowns, which provide esthetic permanent restorations, are probably the restoration of choice for AI and have been reported to be successful in affected adults,[19] but their use in young patients usually is contraindicated due to the presence of large pulps.

In the primary dentition, anterior primary teeth may be restored with strip crowns, using glass ionomer cements as an intermediary material underneath the composite resin veneers. Alternatively, anterior stainless steel crowns with composite resin facings have been tried successfully.[48]

Dental sensitivity. Sensitivity of the teeth to hot

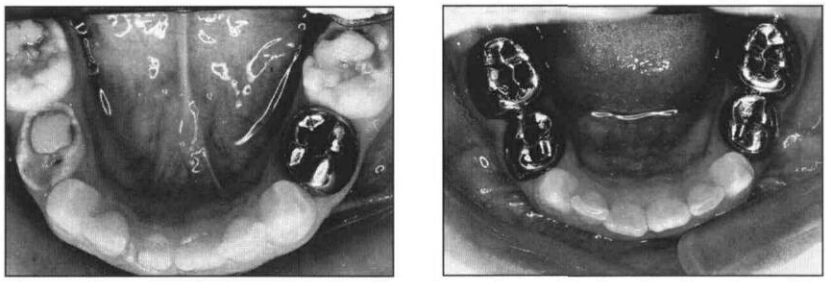

Fig 9a, 9b. Hypoplastic primary molars of a patient with the pitted variant of AI (left), successfully restored with stainless steel crowns (right).

and cold is a common complaint of patients with AI. The severest problems are encountered in the variants presenting with the least amount of enamel, such as the hypocalcified and the smooth and thin hypoplastic types. In the young permanent dentition, as well as the primary dentition, the most effective method to manage dental sensitivity is full coronal coverage using stainless steel crowns in the posterior teeth (Fig 9).

In constructing steel crowns, a conservative technique of tooth separation using separating elastics prior to the insertion of the crowns is recommended.[49,50] This technique, which obviates the need for proximal reduction of tooth structure, allows the stainless steel crowns to be inserted with minimal tooth reduction.

Furthermore, glass ionomer cements are likely to be better luting agents for the crowns compared with zinc phosphate if there are large areas of exposed dentin.

Dental caries and intracoronal restorations. Although a few studies have suggested that patients with AI have less dental caries[20] due to a lack of proximal contacts and elimination of fissures through enamel loss, it is equally likely that in many forms of AI, the rough enamel surfaces predispose to increased plaque retention and greater caries susceptibility. Furthermore, the loss of enamel and the presence of large areas of exposed dentin also may increase caries. Therefore, caries-preventive measures such as frequent topical applications and dietary control are strongly recommended for all AI patients.

Except for mildly affected teeth, intracoronal restorations with amalgam are usually unsuccessful due to fracture of the weak enamel margins. In this study, it was found that for small restorations, adherent materials such as glass ionomer cements and composite resins[17] are better retained compared to amalgam restorations. However, in most cases, full coverage is required for posterior teeth due to extensive enamel loss, as well as for the prevention of further loss of tooth structure. In the primary and early mixed dentition, stainless steel crowns are effective restorations.

Anterior open bite. Alteration of the occlusal vertical dimensions may occur in AI. Anterior open bite has been associated in AI, particularly in the hypocalcified types,[20, 21, 51–53] although its etiology remains unclear. Theories include the suggestion that it has resulted from abnormal tongue positioning caused by teeth sensitivity as well as the possibility that the anterior open bite is a feature of the AI syndrome.[52, 53] Whatever its cause, the open bite often is difficult to treat. Types of corrective treatment that have been suggested range from routine orthodontic banding[47] to orthognathic surgery,[46] all with varying degrees of success.

In contrast to anterior open bite, collapse of the posterior occlusal segments, leading to deep anterior overbite also has been reported in some types of AI.[2, 20, 30, 35] In this study, the patients most predisposed to this problem belonged to the hypocalcified AI group (Table 3). In addition, affected male patients of the X-linked type of AI, as well as all patients with the smooth hypoplastic type of AI also demonstrated this propensity. Loss of occlusal vertical dimension is best prevented as early as possible, preferably in the primary dentition by fabricating posterior steel crowns.[50] In the case of patients who have lost extensive interocclusal height, rehabilitation may be achieved by posterior full crowns and/or by overlay dentures.[30, 35]

Gingival inflammation. All AI patients are predisposed to poor gingival health. There is enhanced plaque retention and calculus formation resulting from the rough enamel surfaces, which may extend subgingivally. Increased preventive oral health practices as well as frequent professional prophylaxis form an important component of management strategies for these patients.

Other clinical problems that have been reported previously in AI include delayed eruption and/or tooth impaction.[23] This problem was noted in only one male patient in this study who had the hypocalcified type of AI. Resorption of unerupted teeth also has been reported previously,[33] but was not noted in this patient series.

Future studies

Future research into several aspects of AI are required to improve the understanding of this condition. Molecular studies of the genetic aspects of the disease would provide important insight into its pathogenesis. Comparative biochemical, clinical, and electron microscopic studies of affected teeth from different variants of AI would lead to better understanding of the differences in defects found in each type. Furthermore, while previous prevalence studies have provided useful information, further epidemiological studies of other populations/racial groups are necessary. In these studies, improved diagnostic criteria based on current understanding of the phenotypic expressions of the different variants may provide more accurate figures of prevalence.

In conclusion, this clinical study has provided further insight into the diagnostic features and clinical complications of the different AI variants. Accurate diagnosis and appreciation of associated clinical problems in each case enable the institution of early preventive measures and management techniques using a multidisciplinary approach.

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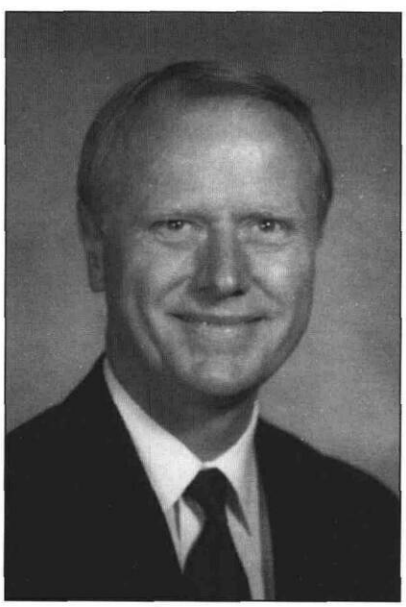
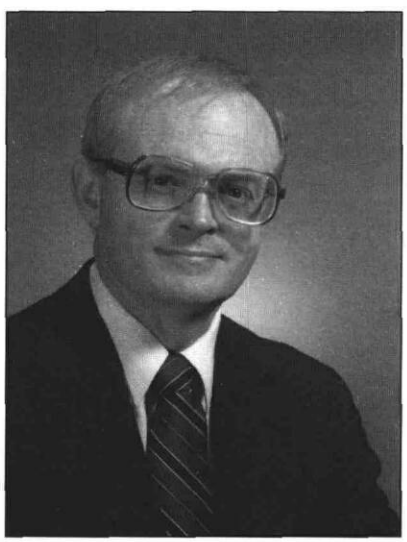
New chairman of the board and board director of the American Board of Pediatric Dentistry



Thomas J. Wickliffe

During the Annual Meeting of the American Board of Pediatric Dentistry at Richmond, Virginia, Dr. Thomas J. Wickliffe, a pediatric dentist in Billings, Montana, was installed as chairman of the board. Dr. Wickliffe received a DDS and an MSD in pediatric dentistry from Indiana University. He is a past president of the Ninth District Dental Society and the Montana Academy of Pediatric Dentistry, and is a fellow of the American Academy of Pediatric Dentistry. Dr. Wickliffe has served on the Membership Committee of the Academy.



Michael W. Roberts

Dr. Roberts received his DDS from the University of Texas—Houston. He completed a general practice residency at the U.S. Public Health Service Hospital in Boston and received a MScD in pediatric dentistry from the Boston University School of Graduate Dentistry. In 1989, Dr. Roberts joined the University of North Carolina School of Dentistry and School of Medicine faculties as graduate program director, pediatric dentistry, following a career in the U.S. Public Health Service. He is a fellow of the American Academy of Pediatric Dentistry, American Society of Dentistry for Children, and American College of Dentists. Dr. Roberts has served on numerous committees of the American Academy of Pediatric Dentistry and is currently chairman, membership.