Dental Anomalies

13

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Learning Objectives
By the end of this chapter, readers will:

- Be able to describe the features and complications of hypodontia and supernumerary teeth.
- Recognise the presentation of microdontia, macrodontia and double teeth.
- Recognise the features of MIH and be aware of when a referral to specialist services is indicated.
- Understand the clinical presentations of enamel abnormalites and developmental disorders affecting dentine.

13.1 Number Anomalies

13.1.1 Hypodontia

Hypodontia is the absence of one or more primary or permanent teeth, excluding third molars. It results from developmental failure of teeth, either in isolation or as part of a syndrome. It can be mild (1–2 missing teeth), moderate (3–5 missing teeth) or severe (6 or more missing teeth, with variable prevalence) (see Table 13.1).

Table 13.2 shows possible aetiological factors for non-syndromic hypodontia. Common syndromes include ectodermal dysplasia (see Fig. 13.1a, b), incontinentia pigmenti and cleft lip and palate-ectodermal dysplasia syndrome (EEC1). Oral features are presented in Table 13.3.

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Table 13.1 Prevalence of hypodontia in the primary and permanent dentitions

	Primary dentition	Permanent dentition
Prevalence	1.5%–1.78%	6.4%
Most affected teeth	Upper lateral incisors Lower central incisors	Lower premolars Upper lateral incisors
Male:female ratio	No difference	Girls are 1.22 times more likely to have hypodontia

Table 13.2 Aetiological factors for non-syndromic hypodontia

Genetic	Environmental
• Pattern of inheritance: Autosomal dominant (AD), autosomal recessive (AR) or X-linked • Common genes: PAX9 (paired box gene 9), MSX1 (muscle segment homeobox 1), AXIN2 (axis inhibition protein 2), EDA (ectodysplasin A)	• Thalidomide treatment, rubella infections during pregnancy • Cleft lip and palate • Chemotherapy or radiotherapy in early infancy • Trauma (weak evidence)

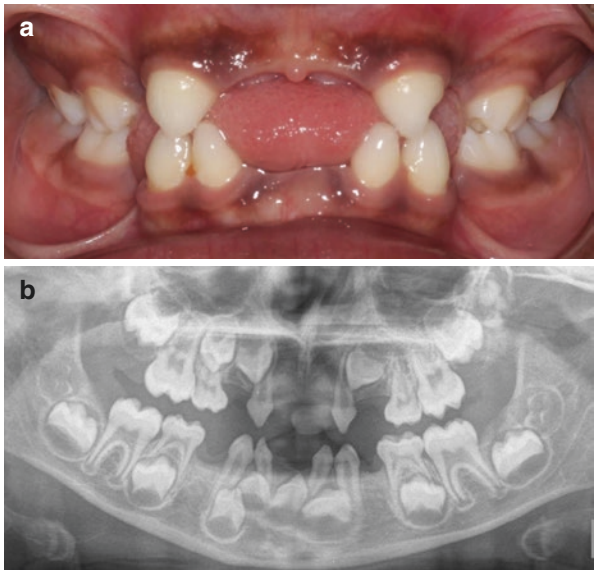

Fig. 13.1 (**a**, **b**) Photograph and DPT of a 7-year-old child in the mixed dentition and ectodermal dysplasia. Images showing hypodontia of UR4, UR2, UR1, UL1, UL2, UL4, UL5, LL1, LR1, LR4 and the presence of multiple conical teeth

Table 13.3 Oral features associated with hypodontia

Tooth morphology	Exfoliation/eruption	Bone development
• Microdontia (small teeth) • Conical teeth	• Primary teeth: Delayed exfoliation, Ankylosis/infraocclusion • Permanent teeth: Delayed eruption Ectopic eruption	• Reduced alveolar development • Altered craniofacial morphology (usually class III and reduced lower anterior facial height)

Adapted from Gill and Barker

Treatment is often complex, requiring multidisciplinary care as described in Tables 13.4, 13.5, and 13.6. Good communication between specialised centres and the general dental practitioners is essential as some treatment can be carried out locally in shared care arrangements. Figure 13.2 gives an example of one restorative option replacing missing teeth in a child with hypodontia.

Table 13.4 Management options for hypodontia in the primary dentition

Prevention: Diet, fluoride and oral hygiene instructions as per the DoH prevention toolkit	
Issue	*Management*
No issue with aesthetics or function No tooth wear	• Diet and oral hygiene instructions, 1350–1500 ppm fluoridated toothpaste, periodic fluoride varnish applications • Periodic monitoring: Radiographs may not be indicated in young children if it is thought that diagnosis would not change the treatment plan
Missing teeth	• Removable prosthetics; may need clasps for additional retention
Tooth wear (due to attrition)	• Composite restorations. If possible, full coverage using crown forms

Adapted from Hobkirk et al.

Table 13.5 Management options for hypodontia in the mixed dentition

Prevention: Diet, fluoride and oral hygiene instructions as per the DoH prevention toolkit	
Issue	*Management*
No issue with aesthetics or function No tooth wear No malocclusion	• Diet and oral hygiene instructions, 1350–1500 ppm fluoridated toothpaste, fluoridated mouthwash (from 7 years old) and periodic fluoride varnish applications • Periodic monitoring
Microdont permanent teeth or primary teeth with tooth wear	• Composite restorations to address aesthetic concerns
Missing teeth	• Removable prosthetics • Overdentures
Spaced dentition	• Simple orthodontic treatment to close spacing (frenectomy may be required for diastema closure if low insertion maxillary labial frenum) Long-term retention may be needed
Ectopic palatal canines	• Interceptive extraction of primary canines may be indicated following orthodontic assessment
Infraoccluded primary molars	• Mild/moderate: Monitor • Moderate with tipping of adjacent teeth or over-eruption of opposing teeth: Consider build up to occlusal height and monitor further infraocclusion • Severe: Extraction (may need to be surgical extraction)
Microdont permanent teeth, primary teeth with tooth surface loss	• Composite resin restorations

Adapted from Hobkirk et al.

Table 13.6 Management options for hypodontia in the permanent dentition

Prevention: Diet and oral hygiene instructions, 1350–1500 ppm fluoridated toothpaste, 2800 ppm (above 10 years old), 5000 ppm (above 16 years old), fluoridated mouthwash, periodic fluoride varnish applications	
Issue	*Management*
No issue with aesthetics or function No tooth wear No malocclusion	• Prevention as above • Maintain primary teeth if no issues • Periodic monitoring
Microdont permanent teeth or retained primary teeth with tooth wear	• Composite restorations to address aesthetic concerns
Missing teeth	• Orthodontic treatment • Pontics may be placed on fixed appliances or retainers as temporary measure • Resin bonded bridges or tooth auto transplantation following orthodontic assessment/treatment • Overdentures if severe hypodontia *Long term (adult management):* • Single tooth implant, implant-retained bridge or implant-retained removable prosthetics • Orthodontics and orthognathic surgery

Adapted from Hobkirk et al.

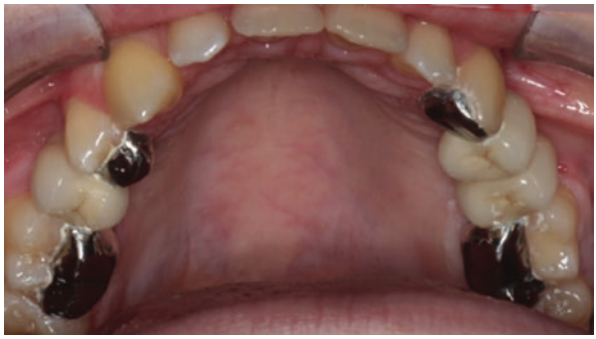

Fig. 13.2 Fixed-fixed resin retained bridges to replace upper premolars on a child with hypodontia

Clinical Tip: Management of Missing Upper Lateral Incisors

Open space: Keep or optimise space orthodontically followed by prosthetic replacement (resin-bonded bridges or implant in adults).

Close space: Mesialise canines and restore them to mimic lateral incisors.

13.1.1.1 Ectodermal Dysplasia

Ectodermal dysplasia (ED) is a group of over 200 inherited disorders characterised by congenital defects on one or more ectodermal structures (most commonly skin, hair, nails, teeth and sweat glands).

80% of ED patients have hypohidrotic ectodermal dysplasia (HED), which follows an X-linked recessive inheritance pattern—males are affected and females are carriers. Its characteristic triad features include hypotrichosis, hypohidrosis and hypodontia (Table 13.7). Dental management is similar to that of severe hypodontia.

Clinical Tip:
Keep the dental environment cool for patients with HED as they have difficulties regulating body heat due to reduced sweating.

13.1.2 Supernumerary Teeth

Supernumerary teeth are defined as teeth or tooth-like structures in addition to the normal number. Primary supernumerary teeth are quite rare, whereas they occur more often in the permanent dentition. If a child has supernumerary primary teeth, they are more likely to develop supernumerary teeth in their permanent dentition. The aetiology of supernumerary teeth remains unclear, but it is likely to be a multifactorial process with strong genetic and environmental influences. Table 13.8 describes the prevalence of supernumerary teeth.

13.1.2.1 Features

Supernumerary teeth may be diagnosed as radiographic findings, associated with syndromes (see Table 13.9) or, more often, following failure of eruption of permanent teeth. They may be classified according to morphology or location as described in Tables 13.10 and 13.11. Figure 13.3a, b feature a form of supernumerary tooth found during radiographic examination.

Clinical Tip
Delayed exfoliation of upper primary incisors should alert to possible presence of a supernumerary tooth.

Table 13.7 Clinical features of hypohidrotic ectodermal dysplasia (HED)

Hair	Hypotrichosis: Sparce, thin, dry and curly hair
Teeth	Hypodontia: Missing large numbers of teeth; conical teeth present
Sweat glands	Hypohidrosis: Reduced sweating

Table 13.8 Prevalence of supernumerary teeth

	Primary dentition	Permanent dentition
Prevalence	0.3–0.8%	0.1–3.8%
Male:female ratio	No difference	Two times more likely in boys

Table 13.9 Syndromes with associated supernumerary teeth

Associated syndromes
Cleidocranial dysplasia
Cleft lip and palate
Gardner's syndrome
Ehlers Danlos
Fabry Anderson's syndrome
Incontinentia pigmenti
Trico-rhino-phalangeal syndrome

Table 13.10 Classification of Supernumerary teeth according to morphology

Classification: Morphology				
Type		Appearance	Location	Prevalence (%)
Conical		• Small and conical • Normal root • Often can erupt without intervention	Pre-maxilla	75
Tuberculate		• Barrel-shaped crown	Pre-maxilla	12
Supplemental		• Normal tooth	Any	7
Odontome	Compound	• Separate, small tooth structures	Pre-maxilla	6
	Complex	• Single, irregular mass of dental tissue	Posterior mandible	

Table 13.11 Classification of supernumerary teeth according to location

Classification: Location	
Type	Location
Mesiodens	Between maxillary central incisors Usually palatal (but can be in the line of the arch or buccal)
Paramolar	Buccal or lingual/palatal to molars Rarely in the line of the arch, between the second and third molars
Distomolar	Distal/distolingual to the third molar
Parapremolar	In the premolar region Resembles a premolar

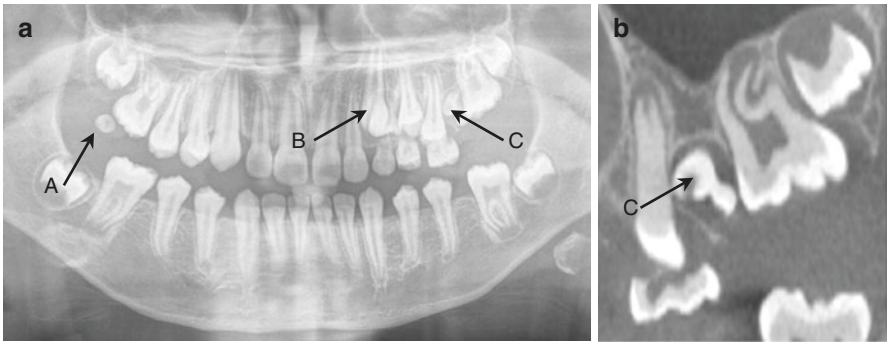

Fig. 13.3 (**a**, **b**) DPT and CBCT showing one paramolar (A) and two parapremolar (B) supernumerary teeth present on the same child

Clinical Tip:
Consider further views to determine the presence of multiple supernumerary teeth (e.g. DPT).

If doing a CBCT, choose a small field of view to limit exposure.

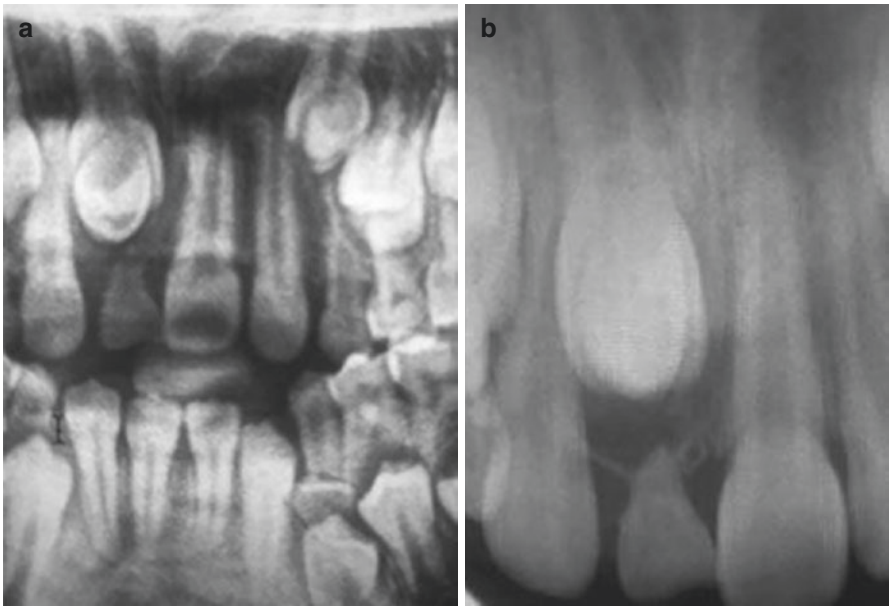

Fig. 13.4 (**a**, **b**) Section of DPT and USO showing a supernumerary tooth palatal to UR1. Note how the supernumerary tooth in (**a**) appears to move up in relation to the incisal edge of UR1 (**b**) from the DPT to USO

The position of supernumerary teeth may be determined by:

– Parallax view as described in Fig. 13.4a, b. Refer to Chap. 9 for explanation of parallax view.
– Cone beam computed tomography (CBCT) to obtain a three-dimensional image, may be justified for surgical planning of complex cases (see Fig. 13.3b).

The presence of supernumerary teeth may lead to:

- Delayed or failed eruption of permanent teeth (28–60%).
- Crowding.
- Rotation or ectopic position of permanent teeth.
- Permanent root malformation (dilacerations), delayed root development or root resorption.
- Cyst formation of unerupted supernumerary teeth (4–9% cases), migration into the nasal cavity/maxillary sinus (rare).

General principles for management include removal of supernumerary teeth and creating enough space for the eruption of the impacted permanent teeth, with or without orthodontic traction as described in Table 13.12. Management is often multidisciplinary. Figure 13.5b–d demonstrate surgical management of supernumerary teeth causing failure of eruption of the permanent incisor.

Table 13.12 Management options for supernumerary teeth

Monitor	• If supernumerary teeth are not causing complications, orthodontic treatment is not planned or tooth's position will not interfere with orthodontic treatment (e.g. supernumerary is beyond the apex of adjacent teeth) • Yearly radiographic reviews are advised
Removal of supernumerary tooth only	• Between 49% and 91% impacted teeth erupt spontaneously up to 18 months after extraction of supernumerary teeth • Good planning is essential prior to surgical extraction of unerupted supernumeraries. Pharmacological behaviour management (inhalation sedation, intravenous sedation) or general anaesthetics (GA) are often required
Removal of supernumerary tooth with creation of space	• Spontaneous eruption of impacted permanent teeth is more likely to occur if adequate space is available • Orthodontic treatment may involve removable or fixed appliances
Removal of supernumerary tooth with permanent tooth exposure or traction	• Exposure and bonding of permanent successor. This technique involves bonding of a gold chain followed by orthodontic traction • Royal college of surgeon guidelines consider this to be good practice to avoid the need for repeat GAs

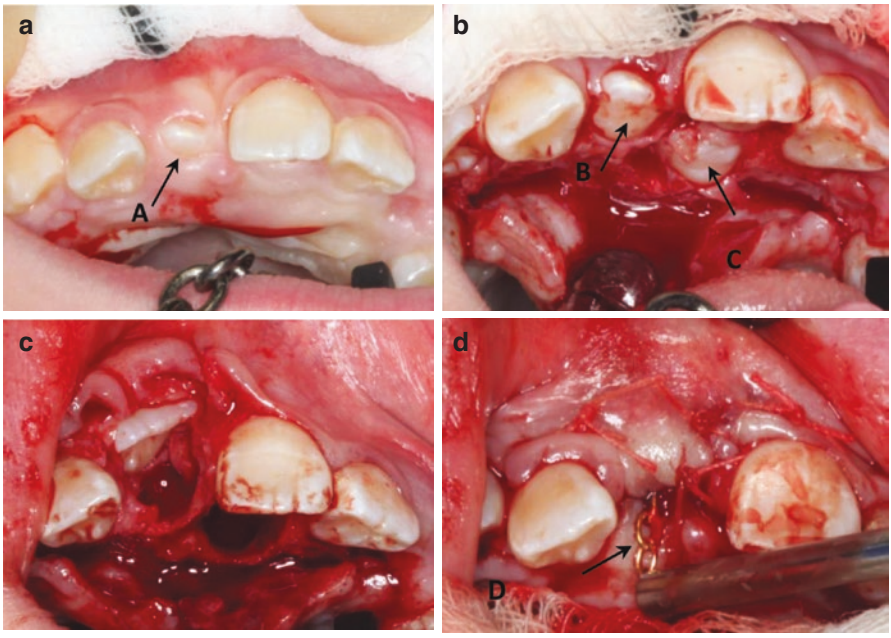

Fig. 13.5 (**a**) Failure of eruption of the upper right central incisor due to the presence of two supernumerary teeth (arrow A). (**b**) Two supernumerary teeth visible after raising a mucoperiosteal flap (arrows B and C). (**c**) Identification of the unerupted upper right central incisor. (**d**) Gold chain visible following suturing (arrow D). (Acknowledgement: Prabhleen Anand)

13.2 Shape Anomalies

13.2.1 Dens Evaginatus

Dens evaginatus (DE) are rare cusp-like formations that contain enamel, dentine and occasionally pulp. They are most common on the palatal and occlusal surfaces and most frequently affect upper incisors (called talon cusps) and lower premolars (see Table 13.13). DE have unknown aetiology and may be associated with other dental anomalies.

DE are rare (0.06–7.7%,), but prevalence varies around the globe, being more common on patients with Asian backgrounds. DE are very rare on primary teeth. Figure 13.6a, b show a talon cusp managed surgically.

Premolar evaginatus may initially be missed and tend to present following fracture, often resulting in dentine or pulp exposure and subsequent necrosis as seen in Fig. 13.7a, b.

Clinical Tip:
50% of dens evaginatus are bilateral so examine all teeth carefully.

Table 13.13 Hattab's talon cusp classification

Type I	Cusp projects at least half way between CEJ and incisal edge
Type II	Cusp projects less than half way between CEJ and incisal edge
Type III	Trace talon or enlarged tooth cingulum

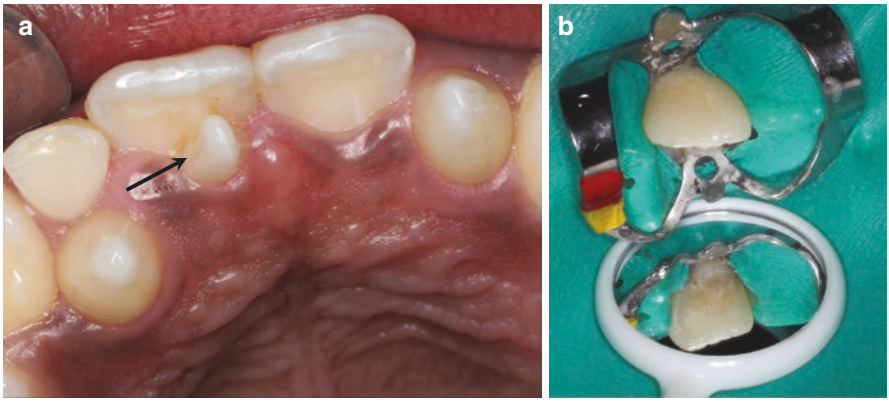

Fig. 13.6 (**a**) A Talon cusp on the upper right central incisor (see arrow). Note the presence of other anomalies as conical upper laterals and crowding. (**b**) The same upper right central incisor immediately following surgical excision due to occlusal interference

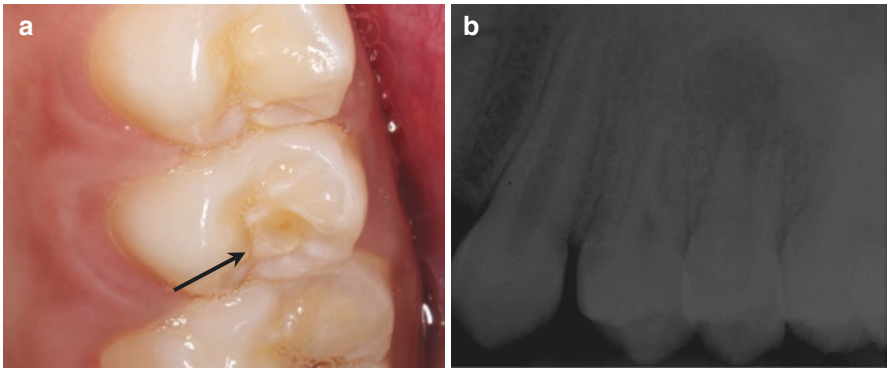

Fig. 13.7 (**a**) Fractured dens evaginatus on the upper left second premolar. (**b**) Periapical radiograph of the same child, showing a periapical radiolucency related to ED on the immature tooth

Table 13.14 Management options for dens evaginatus

Prevention	Selective reduction	Surgical excision
• Fissure sealant • Monitor	• Usually 1–1.5 mm reduction over 6–8 weeks (3–6 visits)—Use high speed with water cooling, no LA needed • Do not cover the dentine with bond or composite. Fluoride and tooth mousse (CPP-APP) may be used • After ideal reduction seal dentine with composite • Review: At least 1 year	• LA and rubber dam use are essential, especially if pulp exposure is expected • Direct pulp cap or Cvek pulpotomy if pulp is exposed (as described in Chap. 11) • Composite restoration following excision • Review: At least 1 year[a]

[a]If it is required to excise it on a single visit, clinicians must be prepared for possible pulp exposure and be aware of increased risk of pulp necrosis

The presence of a tooth evagination may cause a number of complications, including aesthetics, caries, cusp fracture, pulp necrosis, occlusal interference, displacement of the talon tooth, attrition, apical periodontitis or periodontal problems from excessive occlusal forces.

Periapical radiographs and, more recently, CBCT are advised for the assessment of the radicular anatomy before treatment, in order to avoid pulp exposure.

If no concerns, sealing and monitoring may be enough. If cusp removal is required, selective grinding in sequential visits seems to have a better prognosis than excision (see Table 13.14).

13.2.2 Dens Invaginatus

Invagination is an infolding of the enamel, dentine and pulp that may extend into the root. It is rare and more common in the permanent dentition, with reported prevalence between 0.3% and 10%, depending on the population. Upper lateral incisors are the most affected teeth. Aetiology remains unclear. Table 13.15 shows the Oehlers 1957 classification of dens invaginatus. Figure 13.8 features an example of dens invaginatus as seen on a periapical radiograph.

Table 13.15 Oehlers 1957 classification of dens invaginatus

Oehlers classification				
Type I: Invagination confined to the crown	 Type I	 Type II	 Type IIIa	 Type IIIb
Type II: Invagination invading root, as a blind sac. May connect to pulp				
Type IIIa: Invagination through the root to apical region—Opens laterally				
Type IIIb: Invagination through the root to apical region—Opens apically				

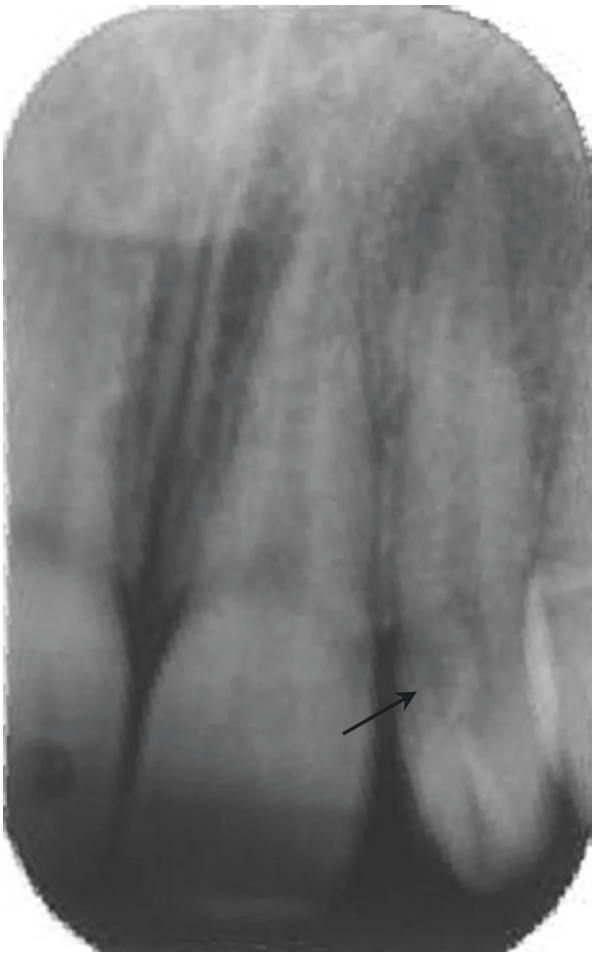

Fig. 13.8 UL2 with dens invaginatus Oehlers type II (see arrow)

Table 13.16 Management of dens invaginatus according to Oehlers classification (adapted from Gallacher et al.)

Type I	Type II	Type III
• Fissure sealant • If necrotic: Remove invagination with gates giddens or ultrasonic and proceed with root canal treatment	• Fissure sealant if not carious following thorough assessment[a] • If carious and vital: Remove caries and restore • If necrotic: Remove invagination with gates giddens or ultrasonic and proceed with root canal treatment	• Fissure sealant if not carious following thorough assessment[a] • If carious and vital: Restoration may be difficult as caries removal is likely to lead to pulp exposure • If necrotic: Endodontic treatment is often necessary in the invagination and on the rest of the root canal system, as histologically they are often interconnected • Bioceramics and thermoplastic GP are required • Referral to specialised services must be considered if necrosis is present

[a]It may be difficult to assess caries as they may be deep within the lesion

Tooth invaginations may range from deep pits or palatal grooves to grossly deformed teeth. Bacteria may contaminate these tooth pockets leading to caries and rapid necrosis. It is, therefore, important to assess vitality and prevent complications by intervening early. Assessment and treatment planning require good quality periapical radiographs taken at different angles or cone beam computer tomography (CBCT) due to complex anatomic variations. Root canal treatment may be challenging, often warranting referral to specialist services. Table 13.16 describes the management of dens invaginatus according to its classification.

13.2.3 Size

Teeth that are too small (microdontia) or too big (macrodontia/megadontia) are rare and may be associated with a syndrome. It is important to look for any other anomalies, as microdontia is often seen in hypodontia, and macrodontia can be linked to supernumerary teeth.

13.2.3.1 Microdontia

- Microdont teeth are smaller than normal and may be tapered (often called 'peg shaped') or normal in shape (see Fig. 13.9).
- Prevalence varies from 0.5% to 2.5%, with females more affected than males.
- Upper permanent lateral incisor is the most common tooth affected.
- Generalised microdontia affecting all the teeth is very rare.

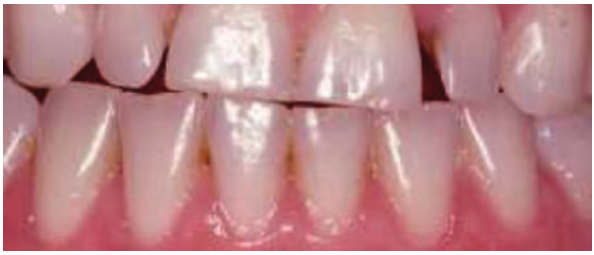

Fig. 13.9 Microdontia of UL2

Clinical Tips

If upper permanent lateral incisors are microdont or missing, the permanent canine may be ectopic.

Remember to palpate/investigate position of upper permanent canines by 9 years of age.

Management of microdontia depends on crown/root size, developing dentition, patient preferences and malocclusion. Options include monitoring (if no treatment required), restoring the tooth with composite (without prepping the tooth) or extracting as part of an orthodontic plan.

13.2.3.2 Macrodontia

- Macrodont teeth are larger than normal variation and are usually normal morphology.
- Usually affects the upper permanent central incisors and bilateral (Fig. 13.10).
- Affect 1% of population (permanent teeth).
- Often confused with double teeth.

Management of macrodontia is complex and depends on crown/root size, developing dentition, patient preferences and malocclusion. Options include monitoring (if no treatment required), trimming and restoring the tooth with composite (but this can be difficult if the teeth are wide at the cervical margin) or extracting as part of an orthodontic plan.

Clinical Tips

Macrodont permanent incisors may present with disturbed eruption due to lack of space.

Patients may require CBCT to assess root morphology and need MDT management.

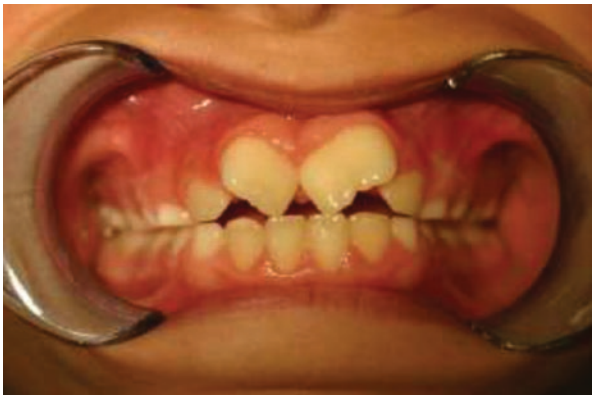

Fig. 13.10 Macrodontia of UR1 and UL1

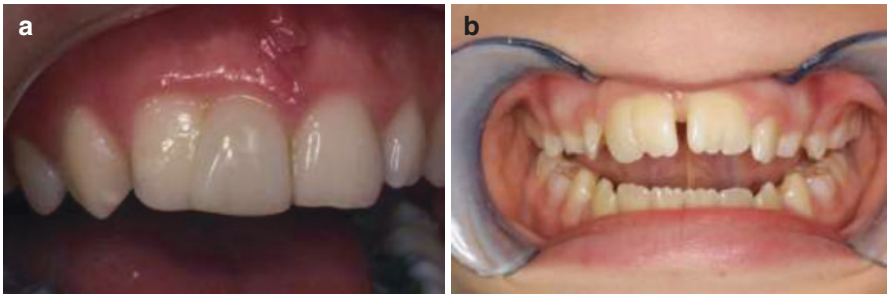

Fig. 13.11 (**a**) Double tooth on UR1. (**b**) Notched double teeth on UR1 and UL1

13.2.3.3 Double Teeth

- Wide variation in appearance—from minor notch as shown in Fig. 13.11b to two separate crowns (see Fig. 13.11a), usually affecting incisors, as result of either germination or fusion.
- Prevalence: 0.5–1.6% primary, 0.1–0.2% permanent dentition, males and females are equally affected.
- About 50% of double teeth in primary teeth have anomalies in permanent dentition.
- Gemination—incomplete attempt of tooth germ to divide into two.
- Fusion—complete or partial fusion of enamel/dentine of two separate teeth.

Management of double teeth is complex and depends on:

- Malocclusion
- Crown width
- Root morphology (i.e. extent of fusion)
- May need CBCT

Options include crown modification, hemisection or extraction.

13.3 Enamel Abnormalities

Enamel formation is tightly controlled by various gene interactions, and genetic and/or environmental disturbances can cause localised, or generalised abnormalities or defects. It is important to be able to distinguish between hypoplastic (missing enamel due to quantity defect) and hypomineralised (discoloured enamel due to qualitative defect), as this will affect the management.

Clinical Tips
Key questions to ask for enamel defects:
Does anyone else in the family have similar teeth?
Are all of the teeth affected in a similar manner?
Is there a chronological distribution to the appearance seen?
Is there anything in the past medical history which might have caused sufficient metabolic disturbance to affect enamel formation?
What is the fluoride history?

13.3.1 Localised and Chronological Enamel Defects

Localised enamel defects on permanent teeth may result as sequelae to trauma to the primary dentition (especially severe intrusions), due to primary tooth sepsis (localised defect or Turner's tooth) or idiopathic. Treatment aims to reduce pain and infection and improve aesthetics. Often localised hypoplastic enamel defects are easily managed with composite restorations. Turner's teeth need to be assessed for restorability, and long-term plans may involve orthodontic input.

Chronological enamel defects may be associated with acquired or genetic conditions of infancy. Disturbances occurring during tooth development may lead to hypoplastic or hypomineralised defects. Permanent teeth may be affected differently, according to the stage of tooth development when the general medical condition was present. Common causes of chronological enamel defects include severe or chronic childhood conditions such as coeliac disease, vitamin D deficiency (see Fig. 13.12), kidney or liver diseases. Acquired aetiology includes tetracycline antibiotic use.

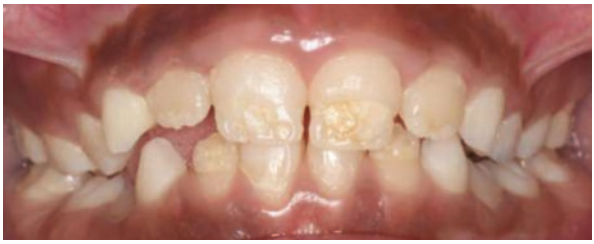

Fig. 13.12 Chronological hypoplasia resulting from vitamin D deficiency. Note that only certain areas of the crowns are affected, showing the chronological aspect of this condition that resolved at the age of 3 years old

13.3.2 Molar Incisor Hypomineralisation (MIH)

Molar incisor hypomineralisation (MIH) was first described in 2001 as 'hypomineralisation of systemic origin of one to four permanent first molars frequently associated with affected incisors'. It is the most common enamel defect, with an estimated 1 in 6 children affected worldwide. Aetiology is unknown, but thought to be systemic.

13.3.2.1 Features

- First permanent molars (FPM) can rapidly undergo post-eruptive breakdown (PEB), and it can be difficult to distinguish between enamel hypoplasia and hypomineralisation (see Fig. 13.13). Looking for discolouration on other FPM and incisors will help distinguish between hypoplasia and hypomineralisation.
- Hypomineralised second primary molars are associated with MIH and can be used as a predictor. This is now a known named condition as HPSM.
- Permanent canine tips can also be affected, as these teeth develop by 10.5 months of age.
- Children may present with sensitivity, pain or concerns with discolouration soon after the permanent teeth erupt, and FPMs can rapidly become carious. Hypersensitivity may make adequate anaesthesia with LA challenging.
- Children with MIH have ten times more dental treatment compared with a group of children with clinically healthy FPMs. Multiple treatment sessions may lead to an increased risk of developing behaviour management problems as well as dental fear and anxiety.
- Bonding with adhesive restorations can be an issue in hypomineralised enamel.
- A recent guide has been developed to aid diagnosis and classification of MIH.
- Management of anterior teeth in MIH depends on age of child, psychological impact on child, developing dentition, patient preferences and malocclusions. Options include monitoring (if no treatment required), or minimally invasive treatment such as vital bleaching (currently controversial in under 18 year olds), microabrasion (see Fig. 13.14a, b) and ICON resin infiltration. If desired result is not achieved, composite restorations may be necessary, although often need to remove hypomineralised area before restoring with composite.

Clinical Tips

To distinguish between hypoplasia and hypomineralisation:

Look for colour changes in the remaining enamel or other FPMs and incisors to indicate PEB due to hypomineralisation.

Radiographic changes:

Poor contrast between enamel and dentine may indicate hypomineralisation.

If unerupted teeth have missing enamel suggests genetic or chronological hypoplasia.

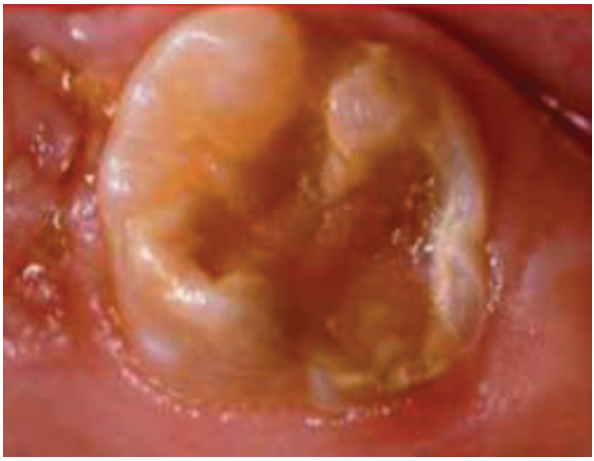

Fig. 13.13 Hypomineralised FPM with PEB (note colour changes in enamel, indicating hypomineralisation)

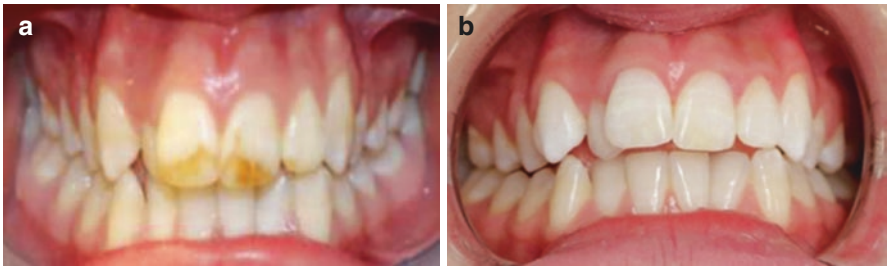

Fig. 13.14 (**a**, **b**) MIH affecting UR1 and UL1 before (**a**) and after (**b**) microabrasion

FPMs affected by MIH are a challenge and depend on:

- Age of patient
- Number of teeth affected
- Extent of damage/severity signs and symptoms
- Type of defect (MIH)
- Occlusion
- Teeth developing
- Cooperation

If the patient has no sensitivity or PEB, fissure sealants to protect porous enamel is recommended. Planned extraction of poor prognosis FPMs in MIH may be an elegant long-term solution for the patient, but may need orthodontic input regarding timing. If extractions are required, do not underestimate the difficulty of extracting badly broken down FPMs where LA may be compromised. If FPMs need to be maintained, adhesive restorations for minimal defects or preformed metal crowns (PMC) for larger defects are advised (Fig. 13.15). Further details on assessment and treatment planning can be found in Chap. 10.

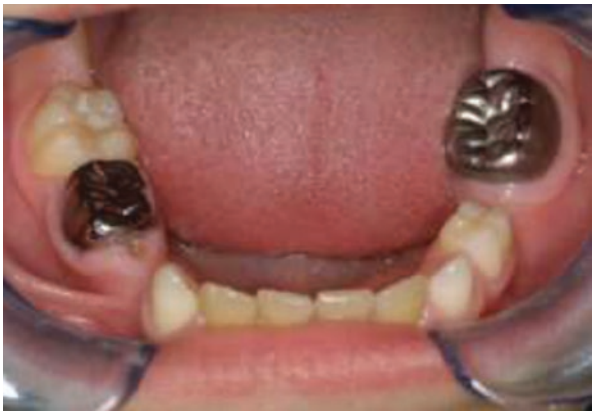

Fig. 13.15 MIH affecting LL6 with PMC

Clinical Tips

Compromised FPMs—principle of treatment:
Don't wait for problems—plan early, ideal window usually between 8 and 10 years of age.
Need a DPT radiograph.
Assess prognosis of the FPMs.
May require orthodontist involvement.

13.3.3 Fluorosis

The benefits of fluoride for reducing caries have been known since the 1940s, but dental fluorosis can cause hypomineralisation and subsequent discolouration, especially if the insult occurs below the age of 6 years. There is no threshold of fluoride that may cause fluorosis, although there is a relationship between dose and response. Teeth affected by fluorosis may present with bilateral opaque white areas to stained yellow dark brown pitted surfaces as seen in Fig. 13.16.

It is important to distinguish between fluorosis and other enamel defects, such as amelogenesis imperfecta (AI) by taking a detailed fluoride history:

- Where was the patient born? Did they grow up in the same area?
- Are any other family members affected?
- Where other people in the area affected with similar teeth?
- What toothpaste did the child use? What amount? Did they spit out after brushing?
- Any other supplements?

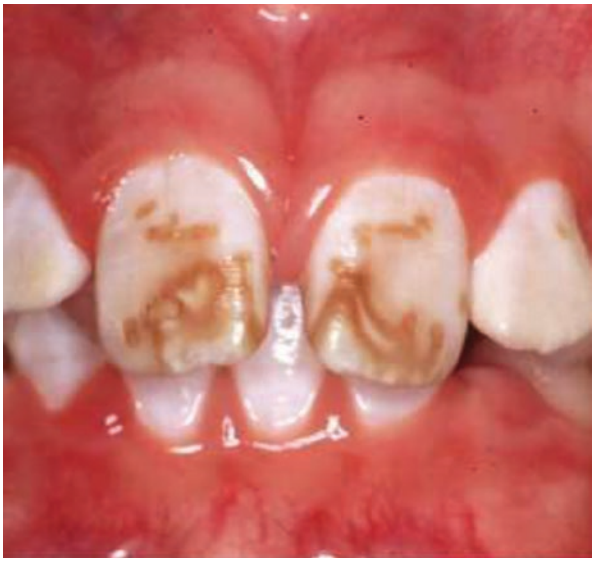

Fig. 13.16 Fluorosis affecting UR1 and UL1

Management of fluorosis can be complex and often requires specialist care. The management options are discussed in detail in the section on AI below.

13.3.4 Amelogenesis Imperfecta

Amelogenesis imperfecta (AI) is a group of genetically inherited conditions, presenting with defective enamel. Although both dentitions are affected, clinically it may only be noticeable on the permanent teeth. AI is rare, affecting between 1 in 700 and 1 in 14,000 people—depending on the population reported.

AI may be inherited as autosomal dominant (AD), autosomal recessive (AR) or X-linked patterns. Defective genes may cause isolated AI (such as AMELX, ENAM, MMP20 and KLK4), AI as part of a syndrome (such as Jalili syndrome, caused by CNNM4 or enamel renal syndrome FAM20A) or may be involved in both types (e.g. LM332). Genetic diagnosis of AI is a growing field and is already provided by some UK centres. More commonly, diagnosis is based on clinical presentation as well as medical and family histories.

13.3.4.1 Features

There are two main clinical presentations, resulting from disturbances at different stages of enamel formation: hypoplastic (see Figs. 13.17, and 13.18a, b) and hypomineralised. Hypomineralised AI may be subdivided into hypocalcified (Fig. 13.19a, b) and hypomature (Fig. 13.20). Both presentations may occur on the same individual. Tables 13.17, 13.18 and 13.19 describe the clinical characteristics of different forms of AI.

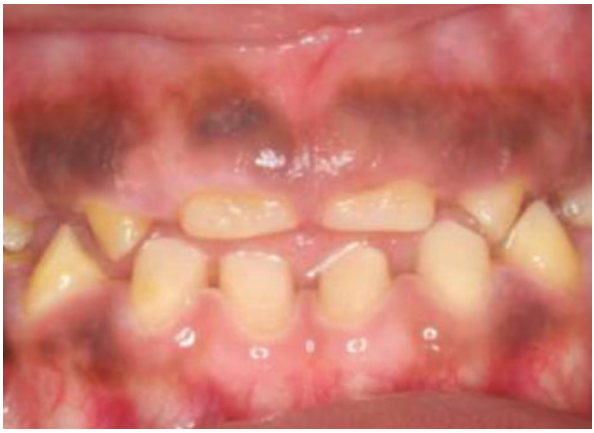

Fig. 13.17 Hypoplastic AI in a 13-year-old child, showing thin enamel and smaller, spaced teeth

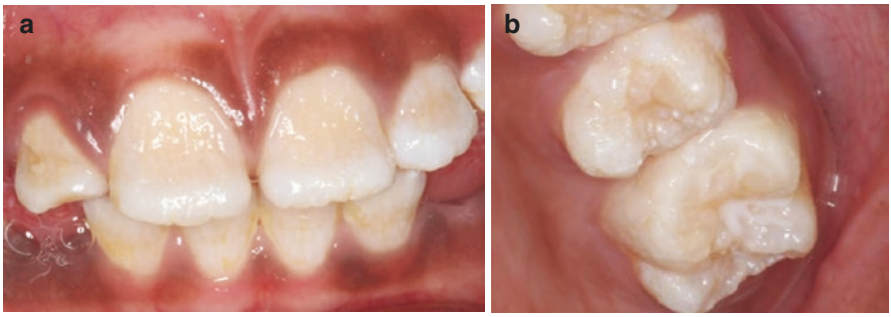

Fig. 13.18 (**a**, **b**) Hypoplastic AI showing grooves/pits on all teeth

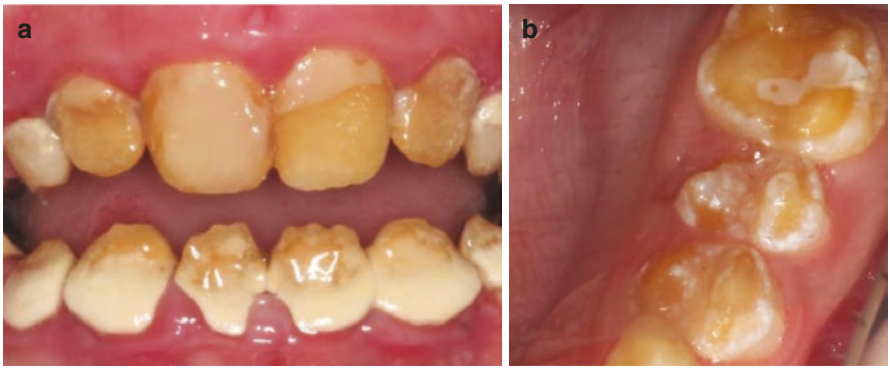

Fig. 13.19 (**a**, **b**) Hypocalcified AI showing enamel loss due to post-eruptive breakdown. The anterior teeth show calculus deposits caused by poor oral hygiene due to sensitivity and failed composite restorations due to difficulties bonding

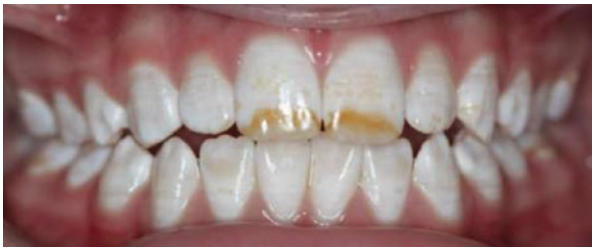

Fig. 13.20 Anterior view of patient with hypomature AI

Table 13.17 Clinical and radiographic characteristics of hypoplastic AI

Amelogenesis disturbance	Clinical presentation	Radiographic findings
Reduced quantity of enamel matrix	• Enamel: Thin, absent or pits/grooves • Normal quality (mineralised) • Abnormal shape and size: Small and may be spaced; colour variation: Normal/yellow/brown • Breakdown: Less likely	• Normal contrast between enamel and dentine • Enamel may appear thinner or unusual shape • Taurodontism may be present

Table 13.18 Clinical and radiographic characteristics of hypocalcified AI

Amelogenesis disturbance	Clinical presentation	Radiographic findings
Stage 1 of mineralisation	• Enamel: Initially normal size and shape but poorly mineralised and brittle, prone to post-eruptive breakdown resulting in missing enamel • Darker teeth: Dark yellow, brown	• Difficult to distinguish between enamel and dentine • Taurodontism may be present

Table 13.19 Clinical and radiographic characteristics of hypomature AI

Amelogenesis disturbance	Clinical presentation	Radiographic findings
Stages 2 and 3 of mineralisation	• Enamel: Initially normal size and shape but poorly mineralised and brittle, prone to post-eruptive breakdown resulting in missing enamel • Colour variation: White diffuse opacities, mottled, yellow/brown	• Difficult to distinguish between enamel and dentine • Taurodontism may be present

Clinical Tips:
On radiographs, check the enamel thickness, shape and contrast of unerupted teeth to help distinguish between AI types.

Aesthetic issues and sensitivity have profound impact on children's and adolescents' quality of life. Children with severe AI report eating difficulties, sensitivity (during toothbrushing) or feeling self-conscious about aesthetics, with many being teased by peers. They often attend more dental visits over time including more dental emergency encounters. Parents often feel guilty or ashamed for passing AI to their children and fear that they will be bullied.

Treatment aims to reduce sensitivity, maintain tooth structure and occlusal height and address aesthetic concerns. Treatment of AI is complex and poses a number of challenges, often requiring a multidisciplinary approach. Tables 13.20, 13.21, 13.22, and 13.23 describe the management of AI in both primary and permanent dentitions. Figure 13.21a, b, show some challenges of treating these children, whereas Figs. 13.22, and 13.23a, b feature examples of restorative management of different forms of AI.

Clinical Tips:
If the child feels sensitivity during bleaching, consider alternating between bleaching gel and a desensitising agent (as Tooth Mousse) placed on the bleaching tray.
Be aware of current legislation regarding bleaching for children.

Table 13.20 AI challenges and possible management options

Challenge	Issue	Management options
• Poor oral hygiene • Chronic gingivitis • Increased calculus	Sensitivity, rough enamel	• Avoid using cold water for toothbrushing • Increase fluoride as per DoH prevention toolkit: 1450 ppm, 2800 ppm, 5000 ppm fluoridated toothpaste according to age; fluoride mouthwash; periodic fluoride varnish applications • Desensitising toothpaste • Tooth mousse (CPP-ACP or casein phosphopeptide amorphous calcium phosphate) • Local analgesia for scaling if needed • Temporary restorations to improve sensitivity and oral hygiene (once oral hygiene improves definitive restorations should be provided)
• Increased caries risk	Poor OH Enamel breakdown Soft enamel	• Diet advise, oral hygiene and fluoride regimens as above • Restore teeth with enamel breakdown or caries

Table 13.20 (continued)

Challenge	Issue	Management options
• Dark discolorations (yellow/brown) or very white enamel or dentine exposure may be difficult to mask with current restorative options	Discolouration	• Microabrasion may be effective for superficial discolouration • Tooth bleaching only or prior to restorations/mircoabrasion • Use opaque composite resins or indirect composite veneers • Ceramic veneers/crowns following gingival maturation (not recommended for children)
• Reduced bond strength to enamel • Need for dentine bonding due to enamel breakdown	Hypomineralised enamel	• Some authors suggest sodium hypochlorite applications prior to etching to increase bonding, but no strong evidence available
• Large pulp/crown ratio	Enamel breakdown, lack of secondary dentine	• Minimal or no tooth preparation prior to restorations in order to retain as much tooth structure as possible
• Exposed margins following full eruption of tooth.	Delayed eruption and gingival maturation	• If possible await full eruption, otherwise replace composite resin when required
• Loss of vertical dimension	Rapid post-eruptive breakdown or enamel hypoplasia with maxillary down growth	• Early interventions to keep posterior vertical dimension in children (preformed metal crowns in primary dentition if required) • Consider overdentures
• Difficulties providing orthodontic treatment	Poor enamel bonding, anterior or posterior open bites, delayed eruption, crown or root resorption, taurodontism, root malformations	• Multidisciplinary care is advised • Interceptive orthodontics, alternative bonding agents, use of bands or metal crowns for bracket attachment, functional appliances and orthognatic surgery have been suggested
• Behaviour issues, treatment burden	Increased number of dental appointments and treatment	• Behaviour management, including pharmacological interventions • Delay non-essential treatment

Adapted from Patel et al. and Arkutu et al.

Table 13.21 Management options for AI in the primary dentition

Prevention: Diet and oral hygiene instructions, 1350–1500 ppm fluoridated toothpaste, periodic fluoride varnish applications Tooth mousse for sensitivity	
Monitor or fissure sealants: If no caries or post-eruptive breakdown Consider GIC sealants for increased sensitivity	
Primary molars	*Composite resin restorations or preformed metal crowns* if caries or post-eruptive breakdown
Anterior teeth	*Composite resin restorations (flowable or conventional)* if caries or post-eruptive breakdown

Table 13.22 Management options for AI in the mixed dentition

Prevention: Diet and oral hygiene instructions, 1350–1500 ppm fluoridated toothpaste, fluoridated mouthwash (from 7 years old), periodic fluoride varnish applications Tooth mousse for sensitivity	
First permanent molars	• *Fissure sealants* and monitor if no breakdown • *Direct and indirect composite* restorations if some breakdown but enough tooth structure • *Metal onlays (traditionally gold)* • *Preformed metal crowns* when generalised breakdown or soft enamel, as a temporary measure until full gingival maturation
Anterior teeth	• *Microabrasion* may address aesthetic concerns for more superficial opacities • *Resin infiltration*: Icon® caries infiltrant is a new product that has been advocated for masking white, mild to moderate opacities • *Direct or indirect composite restorations* (including indirect composite veneers) only if post-eruptive breakdown or hypoplasia and aesthetic issues
• Monitor tooth eruption and refer to orthodontics if necessary	

Table 13.23 Management options for AI in the permanent dentition

Prevention: Diet and oral hygiene instructions, 1350–1500 ppm fluoridated toothpaste, 2800 ppm (above 10 years old), 5000 ppm (above 16 years old), fluoridated mouthwash, periodic fluoride varnish applications Tooth mousse for sensitivity	
Molars	• *Fissure sealants* and monitor if no breakdown • *Direct and indirect composite* restorations if some breakdown but enough tooth structure • *Metal onlays (traditionally gold)* • *Preformed metal crowns* when generalised breakdown or soft enamel, as a temporary measure until full gingival maturation
Premolars	• *Fissure sealants* and monitor if no breakdown and no aesthetic concerns • *Direct or indirect composite restorations*
Anterior teeth	• *Microabrasion* may address aesthetic concerns for more superficial opacities • *Vital bleaching* using a customised tray is a conservative approach to help blending of opacities and lighten dark enamel. It may be used on its own, in combination with microabrasion or prior to placement of restorations[a] • *Resin infiltration*: Icon® caries infiltrant is a new product that has been advocated for masking white, mild to moderate opacities • *Direct or indirect composite restorations* (including indirect composite veneers) only if breakdown, hypoplasia or aesthetic concerns not improved by above measures
• Multidisciplinary care involving orthodontics may be necessary • Transition to adult restorative services must be planned, as well as shared care with the general dental practitioner	

[a]A European union directive in 2012 determined that bleaching products releasing above 0.1% peroxide hydroxide cannot be used under 18 years of age. In 2014, the GDC released a statement allowing bleaching for under 18 year olds, if used for treating or preventing disease. At the time of printing this book, bleaching is covered by the European Union (withdrawal) Act 2018, and therefore no changes to regulations.

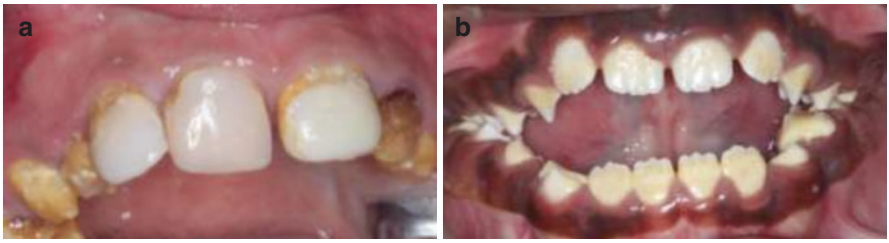

Fig. 13.21 (**a**) Hypocalcified AI with failing restorations due to difficulties in efficiently masking dark enamel, bonding difficulties and delayed gingival maturation. (**b**) Child with hypomature AI and anterior open bite

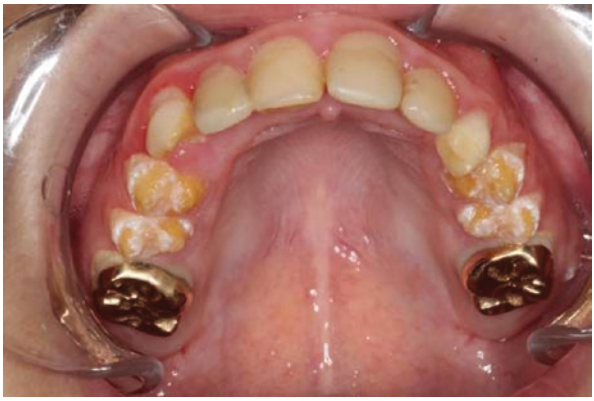

Fig. 13.22 Image of Gold onlays placed on the UR6 and UL6, of a patient with hypocalcified AI (see Fig. 13.19a, b for pre-operative photographs of the same patient)

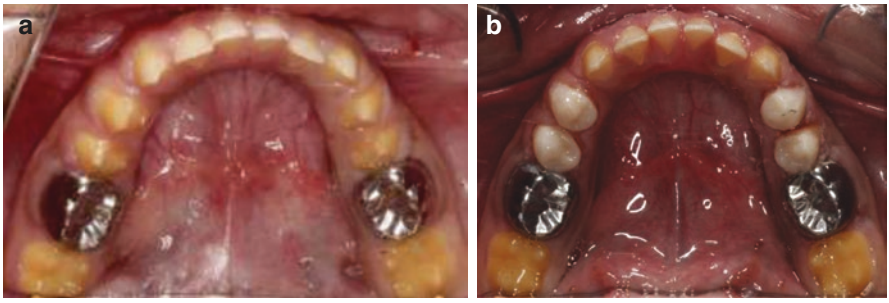

Fig. 13.23 (**a**, **b**) Pre- and post-op photographs of indirect composite onlays cemented on the lower premolars and performed metal crowns on the lower first permanent molars of an adolescent with hypoplastic AI

13.4 Dentine Abnormalities

Dentine is the most abundant tissue in teeth and consists of inorganic (mineralised) and organic (mostly collagen) components. Dentine defects can be caused by genetic or environmental factors and can be limited to the dentine (dentinogenesis

imperfecta or dentine dysplasia) or associated with a generalised disorder such as osteogenesis imperfecta.

13.4.1 Dentinogenesis Imperfecta

Dentinogenesis imperfecta (DI) is an autosomal dominant condition, meaning that one parent will likely be affected. Patients present with discoloured, opalescent dentine affecting both dentitions as seen in Fig. 13.24. Teeth have bulbous crowns that can wear rapidly, as the enamel chips off the underlying disorganised dentine, but pain/sensitivity and abscesses are rarely a problem as the pulps obliterate, which can be clearly seen on radiographs (Fig. 13.25).

Children with some types of osteogenesis imperfecta (OI), otherwise known as 'Brittle Bone disease', can also have DI, although the primary dentition is usually more affected than the permanent, except for OI types III and IV where both dentitions are affected. Many children with OI take bisphosphonates to strengthen their bones, and there has been concern about dental extractions resulting in bisphophonate-related osteonecrosis of the jaw (BRONJ), although no cases have been reported in children with OI.

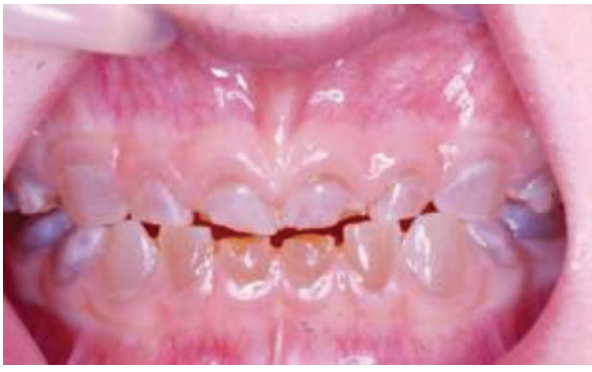

Fig. 13.24 DI in a child in the primary dentition with tooth wear

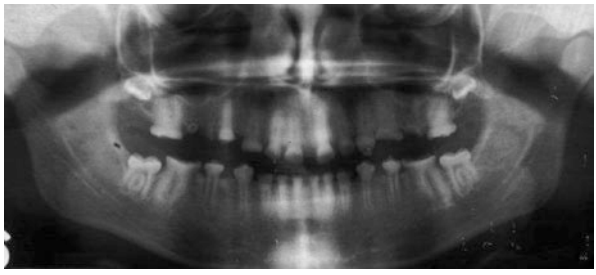

Fig. 13.25 DPT showing patient with DI showing bulbous crowns, tooth wear and pulpal obliteration

Clinical Tips
Dentinogenesis imperfecta is rare, but can be associated with OI. Clinical and radiographic examination will help you distinguish between DI and AI.

Patients with DI have lifelong treatment needs and will require specialist care for function and aesthetics. Management is aimed at maintaining tooth structure and vertical dimension into adulthood, and the use of preformed metal crowns, using the Hall technique, has transformed care for young children. Bonding remains an issue, with frequent repairs and replacements needed.

13.4.2 Dentinal Dysplasia

Dentine dysplasia (DD) is a rare autosomal dominant condition, which can present as two types:

- Type I: 'rootless teeth' as shown in Fig. 13.26a, b.
  - Present with mobile teeth or spontaneous abscesses.
  - Primary and permanent teeth affected.
- Type II: similar to DI type II in primary teeth but permanent teeth look normal with pulp stones and 'thistle-shaped pulp canals'.

Management of DD requires multidisciplinary care, as patients are likely to need advanced restorative care in adulthood.

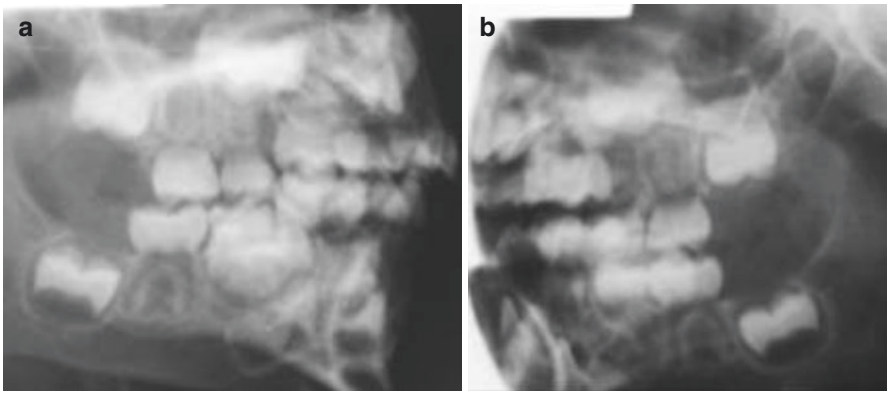

Fig. 13.26 (**a**, **b**) Lateral oblique radiographs showing patient with DD type I (rootless teeth)

13.5 Summary

Children with dental anomalies can be challenging to manage in the dental clinic, and many will need specialist care. However, GDPs are a vital part of the team providing care for these children and their families, as they are a local familiar presence reinforcing prevention and routine care.

Further Reading

Al-Ani AH, Antoun JS, Thomson WM, Merriman TR, Farella M. Hypodontia: an update on its etiology, classification, and clinical management. Biomed Res Int. 2017;2017:9378325. https://doi.org/10.1155/2017/9378325.

Almuallem Z, Busuttil-Naudi A. Molar incisor hypomineralisation (MIH) – an overview. BDJ. 2018;225(7):601–9.

Alqadi A, O'Connell AC. Parental perception of children affected by amelogenesis imperfecta (AI) and dentinogenesis imperfecta (DI): a qualitative study. Dent J (Basel). 2018;6(4) pii: E65 https://doi.org/10.3390/dj6040065.

AlQahtani SJ, Hector MP, Liversidge HM. Brief communication: the London atlas of human tooth development and eruption. Am J Phys Anthropol. 2010;142:481–90.

Arkutu N, Gadhia K, McDonald S, Malik K, Currie L. Amelogenesis imperfecta: the orthodontic perspective. Br Dent J. 2012;212(10):485–9. https://doi.org/10.1038/sj.bdj.2012.415.

Coffield KD, Phillips C, Brady M, Roberts MW, Strauss RP, Wright JT. The psychosocial impact of developmental dental defects in people with hereditary amelogenesis imperfecta. J Am Dent Assoc. 2005;136(5):620–30.

Dashash M, Yeung CA, Jamous I, Blinkhorn A. Interventions for the restorative care of amelogenesis imperfecta in children and adolescents. Cochrane Database Syst Rev. 2013;6:CD007157. https://doi.org/10.1002/14651858.CD007157.pub2.

Elfrink ME, ten Cate JM, Jaddoe VW, Hofman A, Moll HA, Veerkamp JS. Deciduous molar hypomineralization and molar incisor hypomineralization. J Dent Res. 2012;91(6):551–5.

European Union Directive. The cosmetic products (safety) (amendment) regulations. London: HMSO; 2012.

Evans D, Innes I. The Hall Technique A minimal intervention, child centred approach to managing the carious primary molar. 2010. https://dentistry.dundee.ac.uk/files/3M_93C%20HallTechGuide2191110.pdf.

Gallacher A, Ali R, Bhakta S. Dens invaginatus: diagnosis and management strategies. Br Dent J. 2016;221(7):383–7.

GDC Statement: http://www.gdc-uk.org/Search/Pages/results.aspx?k=position%20statement%20whitening. Accessed 07 Apr 2019.

Ghanim A, Silva MJ, MEC E, Lygidakis NA, Marin RJ, Weerheijm KL, Manton DJ. Molar incisor hypomineralisation (MIH) training manualfor clinical field surveys and practice. Eur Arch Paediatr Dent. 2017;18(Suppl 1):225–42.

Gill DS, Barker CS. The multidisciplinary management of hypodontia: a team approach. Br Dent J. 2015;218(3):143–9. https://doi.org/10.1038/sj.bdj.2015.52.

Hobkirk JA, Gill DS, Jones SP, Hemmings KW, Bassi GS, O'Donnell AL, Goodman JR. Hypodontia: a team approach to management. Chichester: Wiley-Blackwell; 2011.

Jälevik B, Klingberg GA. Dental treatment, dental fear and behaviour management problems in children with severe enamel hypomineralization of their permanent first molars. Int J Paediatr Dent. 2002;12(1):24–32.

Leith R, O'Connell AC. Selective reduction of talon cusps- a case series. J Clin Pediatr Dent. 2018;42(1):1–5. https://doi.org/10.17796/1053-4628-42.1.1.

Patel M, McDonnell ST, Iram S, Chan MF. Amelogenesis imperfecta - lifelong management. Restorative management of the adult patient. Br Dent J. 2013;215(9):449–57. https://doi.org/10.1038/sj.bdj.2013.1045.

Pousette Lundgren G, Hasselblad T, Johansson AS, Johansson A, Dahllöf G. Experiences of being a parent to a child with amelogenesis imperfecta. Dent J (Basel). 2019;7(1) pii: E17 https://doi.org/10.3390/dj7010017.

Public Health England. Delivering better oral health: an evidence based toolkit for prevention. 2017. Available at: https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/605266/Delivering_better_oral_health.pdf. Accessed 9 Apr 2019.

Rousseau M, Retrouvey JM. Members of the brittle bone disease consortium. PeerJ. 2018; https://doi.org/10.7717/peerj.5464.

Seehra J, Yaqoob O, Patel S, O'Neill J, Bryant C, Noar J, Morris D, Cobourne MT. National clinical guidelines for the management of unerupted maxillary incisors in children. Br Dent J. 2018;224(10):779–85. https://doi.org/10.1038/sj.bdj.2018.361.

Shah A, Gill DS, Tredwin C, Naini FB. Diagnosis and management of supernumerary teeth. Dent Update. 2008;35(8):510–2, 514–6, 519–20

Shah P, Chander JML, Noar J, Ashley PF. Management of 'double teeth' in children and adolescents. Int J Paediatr Dent. 2012;22(6):419–26.

Smith CEL, Poulter JA, Brookes SJ, Murillo G, Silva S, Brown CJ, Patel A, Hussain H, Kirkham J, Inglehearn CF, Mighell AJ. Phenotype and variant spectrum in the lamb3 form of amelogenesis imperfecta. J Dent Res. 2019;24:22034519835205. https://doi.org/10.1177/0022034519835205.

Venezie RD, Vadiakas G, Christensen JR, Wright JT. Enamel pretreatment with sodium hypochlorite to enhance bonding in hypocalcified amelogenesis imperfecta: case report and SEM analysis. Pediatr Dent. 1994;16:433–6.

Weerheijm KL, Jälevik B, Alaluusua S. Molar-incisor hypomineralisation. Caries Res. 2001;35(5):390–1.