Odontodysplasia: report of two cases

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In 1947, a problem of "arrested tooth development" was reported by McCall and Wald.[1] Rushton[2] introduced the term "shell teeth" in 1954, which he used to describe the radiographic findings. The term "odontodysplasia" was introduced by Zegarelli[3] in 1963, which he defined as "a rare developmental anomaly involving both mesodermal and ectodermal dental components in a group of contiguous teeth." It is characterized by a deficiency or abnormality in the formation of dentine and enamel and involves the primary and/or permanent dentitions.[4] Odontodysplasia appears to be slightly more prevalent in females, has no racial predilection, and is seen in the maxillary dentition twice as often as in the mandibular dentition.[5–7]

Etiology

Many factors have been suggested as being involved in the etiology, as shown in Table 1.[8–13] Although no one factor has been positively identified as the single cause of the condition,[14–18] there is evidence to suggest that vascular defects are involved in the pathogenesis of odontodysplasia.[12] Walton et al.[19] described three cases where vascular nevi were present in the facial skin overlying the area where defective teeth developed and in 1991, Steiman et al.[20] presented a case associated with a vascular nevus of the face and neck. However, it has also been suggested that ischemia due to insufficient blood supply may be a cause.[17]

TABLE. ETIOLOGY OF ODONTODYSPLASIA

1. Local trauma
2. Local ischaemia caused by vascular defect
3. Infection
4. Irradiation
5. Metabolic and nutritional disturbances/vitamin deficiency
6. Hyperpyrexia
7. Genetic/hereditary factors
8. Local somatic mutations
9. Neural crest migration disorder
10. Rhesus incompatibility
11. Systemic disease
12. Associated with hemangiomas

Clinical presentation

The condition manifests during primary tooth eruption, often with delay or failure of eruption. Most often, the central and lateral incisors and canines are involved,[6, 12] but any teeth (usually consecutive) may be affected. If a primary tooth is affected, so is the permanent successor.[6, 7] Although the condition rarely crosses the midline,[10, 21, 22] odontodysplasia has been reported to involve both sides of the midline.[23] Clinically affected teeth appear discolored, hypoplastic, and hypocalcified—an appearance that can resemble dental caries. The teeth tend to be shorter, have short roots with open apices, and abnormally wide pulp chambers.[11, 22]

Radiographic appearance

Affected teeth are poorly discernible, having a reduced radiodensity.[11, 16] The enamel and dentin layers are thin and defective,[12, 19] producing a faint, fuzzy outline. This appearance gives rise to the frequently used term of "ghost teeth".[24] The pulp chambers and canals appear correspondingly large. The roots appear short and stubby with open apices.[5, 12, 25, 26]

Histologic appearance

The connective tissue that corresponds to the dental sac is loose and fibrillar, with calcified bodies scattered throughout.[27, 28] Embryologically, abnormalities are seen in all derivatives of the tooth germ.[5, 6] Differentiation of odontoblasts and ameloblasts is abnormal, which produces defective dentin and enamel.

The enamel is hypoplastic, hypomineralized, and contains degenerated globular calcifications.[22, 29, 30] The dentin is hypoplastic with clefts and interglobular dentin. A widened predentine zone and a reduced number of dentinal tubules are also characteristic of odontodysplasia.

Fig 1. Orthopantograph of Case 1.

Fig 2. Intraoral radiograph of 12 in Case 1.

Foci of calcifications are seen in the dental follicles. These are thought to represent either degenerative changes within the reduced enamel organ or dystrophic calcification mediated by connective cells.[2, 12] Rushton[15] reported that the pathological changes related to early life, with the dentinal changes being less severe in the later-formed dentin nearer the pulp.

In this report, we highlight the clinical and radiographic findings associated with odontodysplasia, and discuss various options in managing these patients.

Case 1

History

A 5-year-old Caucasian girl, now 11, was referred from her general dental practitioner requiring extraction of grossly carious I and J. Her medical history contained nothing of note. Her dental history indicated that she had regular dental examinations, with no previous restorative treatments. A survey of her diet history indicated that she rarely had sweets, as they were restricted by her mother.

Examination

A clinical examination revealed that both I and J were grossly carious, but no further dental caries was present. A routine radiographic examination of the dentition confirmed the presence of caries localized to I and J (Fig 1).

Management

Due to the patient's age, her lack of dental experience, and the condition of I and J, it was decided to arrange an outpatient general anesthetic appointment to extract the teeth.

The patient was re-examined 6 months later. The area where I and J had been extracted had healed uneventfully. At this appointment, L was noted to be infraoccluded. This condition progressively worsened over subsequent recall appointments.

An orthopantograph taken 1 year later revealed that 12 was abnormal in size with incomplete calcification of the crown. Other findings highlighted the fact that 13 and 29 were absent. An intraoral radiograph of 12 clearly showed bony trabeculae present in place of the crown and that 14 was developing abnormally.

At age 8, we did a routine radiographic examination to determine a cause for the absence of teeth 12, 13, and 14, which revealed a ghost-like appearance for 12. This led to a diagnosis of odontodysplasia.

At 9 years of age, 14 had erupted enough to allow inspection of almost all the occlusal surface. The enamel was very pitted, and the contour of the tooth most irregular. Teeth 13 and 29 were still absent and L, K, S, and T were infraoccluded.

The patient was seen by an orthodontist, who advised the extraction of the infraoccluded mandibular first and second primary molars. In order to achieve some resolution of the patients' developing anterior crowding, it was also decided to extract the maxillary primary canines. The poor, long-term prognosis of 14 necessitated its extraction. An outpatient general anesthetic appointment was made for the extraction of the teeth.

Visual inspection of the extracted 14 showed that the buccal roots were stunted and the enamel irregular and hypoplastic, while histopathological examination revealed that much of the enamel was dysplastic, had an irregular, lobulated surface, and lacked the scalloped attachment to dentin. There were also large areas of interglobular and irregular dentin. This confirmed the diagnosis of odontodysplasia.

Follow-up

Regular follow-up of the patient revealed that at age 16, 12 had not erupted (Fig 2) but remained just beneath the gingival surface. To encourage 12 to erupt,

we decided to surgically expose and examine it. An appointment was made to expose 12 under local analgesia, which was carried out uneventfully.

The patient was again orthodontically assessed, with the determination to use fixed appliances to correct the malocclusion, align the teeth, and close the residual spaces. Regular review of 12 will allow monitoring of its eruption. Should any problems develop, it may be necessary to bond a gold chain to the tooth and extrude it orthodontically.

Case 2

History

An 8-year-old, Caucasian boy was referred by his dental practitioner with failure of eruption of 7, 8, and 9. Tooth 10 had erupted normally.

His medical history revealed that he was a mild asthmatic who occasionally used an inhaler (Salbutamol®). He was also being assessed by a neurologist regarding an apparent right-sided, hemifacial paresis of unknown cause.

The patient had no experience of dental treatment with local analgesia. However, the carious E and F had been removed under general anesthesia. Unfortunately, neither the patient nor the mother could recall at what age this procedure had been done.

Examination

Oral hygiene was good and there were no problems except for the absent incisors. Spacing was present where 7, 8 and 9 were absent, with little tilting or space closure by the adjacent teeth.

Radiographic examination was performed to assess 7, 8, and 9 and to eliminate any local causes for their noneruption. An orthopantograph (Fig 3) revealed that all teeth were present except for the third molars. A periapical radiograph of the unerupted teeth (Fig 4) showed a ghostly appearance consistent with that seen in odontodysplasia.

Management

The radiographic findings revealed that the affected teeth are poorly developed, and the prognosis for further development and eruption is poor. Treatment options for this child included maintaining the teeth or their extraction with restoration of the space with a removable prosthesis. Once a stable gingival margin is attained, at about the age of 17 or 18, restoration with a fixed prosthesis can be considered.

The decision was made to leave the teeth in situ based both on clinical reasons and the wishes of the child and parent. Removal of the teeth at this age would lead to a substantial reduction in alveolar ridge height/mass and the subsequent defect would pose immense problems for future restorations should the patient want a fixed prosthesis. By maintaining the teeth for as long as possible, we hope to achieve a normal alveolar ridge. Another advantage to maintaining the alveolar ridge is that the defect can be restored with implants once the growth period is over. For the present, an appointment was made to take impressions so a removable acrylic prosthesis could be constructed to replace the absent teeth.

The patient will be reviewed regularly to assess 7, 8, and 9. One problem that may arise is the partial eruption of the teeth beneath the prosthesis, so monitoring of the gingiva/alveolus will be maintained. At a later date, restoration with a fixed prosthesis will be considered. The decision will then be whether to construct a fixed prosthesis and retain the teeth, or to extract them prior to placement.

Should implants be considered, consultation with

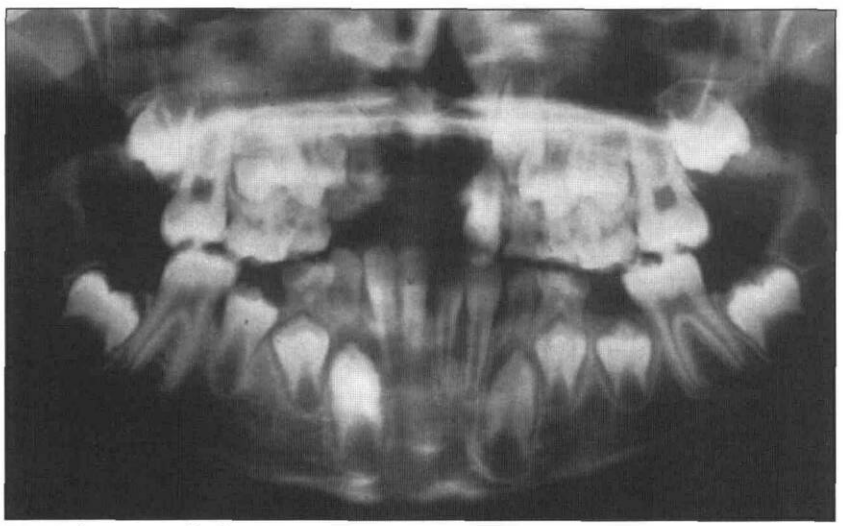

Fig 3. Orthopantograph of Case 2.

Fig 4. Intraoral periapical radiograph of 7, 8, 9 in Case 2.

an implantologist will be necessary to determine whether to extract the teeth prior to or at the time of implantation.

Discussion

In determining the best treatment options for a child with teeth affected by odontodysplasia, several factors must be considered.

1. Age of patient. Age has multiple effects on the future treatment options for the child. The ability to cooperate with treatment may be limited so it may be possible to provide only simple restorative care initially. The main question to answer is whether the affected teeth should be left in situ in the hope that they will eventually develop and erupt normally. Maintaining the teeth, even if they do not erupt, will preserve the alveolar ridge, achieving optimal esthetics for any future prosthesis or, should implants be considered at a later date, maintain adequate bone. For young patients, it may be justifiable to leave the teeth in situ and monitor them regularly. However, the longer the unerupted teeth are retained (i.e., increased in younger children), the higher the chances of developing pathosis. Not only will this necessitate removal of the teeth, but their removal may be more difficult.
2. Any relevant medical history. The patient's medical history (e.g., medical, mental, or physical handicap) may affect the ability of the child to cooperate with, or the clinician to provide, dental treatment. Any cardiac condition will also add to the problems. While the teeth are unerupted, no untoward effects on the patient can be expected, so leaving them in situ may be the best treatment. However, the development of pathosis may have serious consequences for the child's health, thus immediate extraction—which could be a problem in itself—may be indicated.
3. Previous dental experience. This will determine the ability of the patient to cooperate with any future dental treatment and thus determine the treatment provided.
4. Attitude/wishes of child and parent regarding dental treatment. The wishes of the child and parent must be taken into account when formulating the treatment plan. What may appear to be a problem to the dentist may not be to the child and vice versa. It is important to determine what the patient wishes to gain from any treatment provided and the extent to which they are willing to undergo treatment. Does the patient want comprehensive treatment that may involve many visits over a long period of time aimed at correcting aesthetics and malocclusion, or are they interested in a short course of treatment aimed at correcting a minor aesthetic concern? Tailoring the treatment plan to the wishes of the patient will increase patient compliance and improve the chances of a successful outcome for both patient and dentist.
5. Number of affected teeth. The number and position of affected teeth will determine the prosthetic, restorative, and orthodontic difficulties in the future management of the spacing should extractions be necessary.

For a patient, the best treatment is that which requires the minimum intervention and minor restorative care in the future while adhering to their wishes. Preservation of symptom-free, unerupted teeth, though they are malformed, would appear to be the best option. The child is not subjected to complicated, invasive treatment at an early age, so the risks of inducing a negative attitude towards dental care—particularly when future care may warrant complicated and demanding treatment—are reduced.

Removal of the teeth is irreversible and has a detrimental effect on the alveolar ridge, due not only to the absence of teeth but also because of the surgical procedure. Therefore, retention is necessary until such time as removal of the teeth is indicated; for example partial eruption, development of pathology, placement of implants, or impedance to orthodontic alignment.

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