CASE REPORT

Congenitally missing primary and permanent maxillary lateral incisors

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SUMMARY

Congenitally missing tooth/teeth are a common developmental abnormality. It is defined as the developmental absence of tooth/teeth excluding the third molars. It is more commonly seen in permanent dentition but rarely in primary dentition. This paper reports a rare case of non-syndromic bilaterally missing primary and permanent lateral incisors in the maxillary region.

BACKGROUND

Developmental alterations in the number of teeth that develop are common. Hypodontia denotes the lack of development of one or more teeth. Hypodontia is uncommon in primary dentition when compared with permanent dentition with a prevalence that ranges from 0.5% to 0.9%.¹ The overall frequency of patients with congenitally missing teeth excluding the third molars was found to be 1.6–9.6% in permanent teeth, as reported by Graber.² After the third molars, second premolars and lateral incisors are absent most frequently.³ Agenesis is more frequently seen in maxillary primary teeth when compared with mandibular primary teeth.⁴ Women are more often affected when compared with men with a predominance of 1.5:1.¹ There is a close correlation between congenitally missing deciduous teeth and their permanent successors according to the study of Grahnen and Granath.⁵

Genetics has been suggested as an important aetiological factor and autosomal dominant pattern of inheritance has been considered to be predominant.⁶ It may also be associated with some environmental insult during development. The presence of missing teeth in primary dentition can cause a lot of clinical problems like reduced alveolar development, decreased facial height, impairment of speech, deep bite and asymmetry of the affected side of face.¹ Hypodontia is also associated with increased freeway space. The absence of a primary tooth is strongly correlated with increased prevalence of a missing successor.⁵ Furthermore, congenitally missing teeth in permanent dentition are considered to be the most common reason for primary tooth retention.⁷

A multidisciplinary approach may be indicated in the clinical management of problems associated with missing teeth. The aim of the present article is to present a rare case report of congenitally missing bilateral primary and permanent maxillary lateral incisors.

CASE PRESENTATION

A boy aged 7 years reported to the Department of Pedodontics and Preventive Dentistry with the chief symptom of missing teeth in the upper front teeth region. The family and medical history of the child patient regarding missing teeth was insignificant. Thorough intraoral examination showed the presence of all primary teeth with missing maxillary primary left and right lateral incisors. There was no history of extraction of teeth. The primary mandibular right and left central incisors were naturally exfoliated and were replaced by permanent mandibular right and left central incisors. The maxillary arch also revealed an asymmetry in the tooth number. There were four teeth each present on the left and right side (figure 1). External stains were present on primary maxillary central incisors and right first molar. The other teeth were of normal colour, size and shape. Orthopantomogram radiograph confirmed the absence of primary and permanent maxillary right and left lateral incisors (figure 2).

The primary maxillary lateral incisors were missing, thereby confirming that it was a case of congenitally missing teeth. The parents of the child patient gave no history of trauma in the anterior region, dental anomalies and consanguineous marriage. A thorough general examination was carried out to rule out the presence of any associated syndrome. No abnormality of skin, hair and nails was observed. The patient was advised to maintain the regular follow-up visits to observe any deviation from normal eruption pattern.

DISCUSSION

True anodontia or congenital absence of teeth can be found in almost any region of the dental arch

Figure 1 Clinical photograph showing missing maxillary lateral incisors.

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Figure 2 Orthopantomogram X-ray showing agenesis of primary and permanent maxillary lateral incisors.

and in primary and permanent dentitions. According to the most accepted theory, hypodontia is multifactorial and results from a combination of genetic and environmental influences.¹ True anodontia or congenital absence of teeth can be classified into two types, total and partial. Total anodontia is a rare condition in which all teeth are missing and is frequently associated with a more generalised disturbance, whereas partial anodontia involves one or more teeth and is a rather common condition.³ Many cases of congenitally missing primary dentition have been reported in the literature, but this case report presents a unique case of bilaterally missing lateral incisors in the maxillary region associated with agenesis of permanent tooth.

Missing teeth are associated with trauma, infection, radiation, chemotherapeutic medications, endocrine disturbances and severe intrauterine disturbances.¹ Somatic diseases like syphilis, rickets and scarlet fever have also been associated with tooth agenesis.⁸ Tooth agenesis has been associated with a lot of syndromes and dental anomalies like orofacial clefts, Downs syndrome (trisomy 21), Book syndrome, Coffin-Lowry syndrome, Goldenhar syndrome, Ellis-van Creveld syndrome, Marshall-White syndrome, Johanson-Blizzard syndrome, Gorlin-Chaudhary-Moss syndrome, Progeria, Tooth-and-Nail syndrome and Witkop syndrome.^{1 8 9} Hypodontia may be associated with dental anomalies such as enamel hypoplasia, taurodontism, delayed eruption, delayed tooth formation, prolonged retention of primary teeth, ectopic eruption, transposition, abnormal spacing of teeth and altered dimension of the associated gnathic regions.¹⁰ The tooth germs that give rise to permanent incisors, canines and premolars form as a result of further proliferative activity within the dental lamina at its deepest extremity. This increased proliferative activity leads to the formation of another tooth bud on the lingual aspect of the deciduous tooth germ, which remains dormant for some time. The molars of the permanent dentition have no deciduous predecessors, so their tooth germs do not originate in the same way.¹¹ In the present case, no systemic factors/diseases/syndromes were obvious which could be responsible as a possible reason of the missing teeth. However, any unnoticed trauma/infection in the maxillary anterior region in this case could have damaged the teeth germs leading to missing primary and permanent maxillary lateral incisors.

Regulatory homeobox genes found to be associated with tooth agenesis consists of *Msx-1*, *Pax-9*, *EDA*, *TGFA*, *Axin-2*.⁶ Recently, it has been found that mutations in gene encoding low-density lipoprotein receptor-related protein (LRP6) cause tooth agenesis in humans.¹²

The treatment of missing teeth depends on the clinical situation and severity of the case. A number of therapeutic options are available, including removable partial dentures, fixed

prosthodontics, resin-bonded bridges or implants in association with prosthetics crown. Recently, an alternative treatment modality has been reported in which a zirconia-based resin-bonded prosthesis is used to restore congenitally missing maxillary lateral incisors.¹³ Fibre-reinforced composite bridge also offers a good treatment alternative to conventional treatment options for replacement of permanent anterior teeth until a more definitive treatment can be planned.¹⁴ Recently, Rosa *et al*¹⁵ concluded that orthodontic space closure, including first premolar intrusion and canine extrusion in patients with missing lateral incisors does not incur risks of periodontal tissue deterioration or temporomandibular disorders.

A multidisciplinary approach can also be used to ensure functional occlusion and aesthetics at the same time. Genetic counselling may play an imperative role in children with multiple missing teeth. In the present case, the child patient was kept under follow-up to observe the pattern of eruption of permanent anterior teeth.

Learning points

- ▶ Congenitally missing deciduous teeth are uncommon, but if occurring, usually involve the maxillary lateral incisors.
- ▶ Orthopantomogram is necessary if congenitally missing teeth are present in primary dentition to detect the absence of permanent successors in permanent dentition.
- ▶ Paediatric dentists should be aware of the fact that hypodontia of permanent dentition is the most common problem associated with congenitally missing teeth in primary dentition.
- ▶ Hypodontia may also be associated with dental anomalies such as enamel hypoplasia, taurodontism, delayed eruption, prolonged retention of primary teeth, ectopic eruption, transposition, lack of alveolar bone and reduced vertical dimensions.
- ▶ As tooth buds are extremely sensitive to X-ray radiation and may be destroyed with relatively low dosages, they should be advised only if necessary.

Competing interests None declared.

Patient consent Obtained.

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REFERENCES

- 1 Abnormalities of teeth. In: Neville BW, Damm DD, Allen CM, *et al*, eds. *Oral and maxillofacial pathology*. 3rd edn. Elsevier, 2009:78–80.
- 2 Graber LW. Congenital absence of teeth: a review with emphasis on inheritance patterns. *J Am Dent Assoc* 1978;96:266–75.
- 3 Levi SH. Developmental disturbances of oral and paraoral structures. In: Rajendran R, Sivapathasundharam B, eds. *Shafer's textbook of oral pathology*. 6th edn. India: Elsevier (Reprinted), 2011:45–6.
- 4 Stewart RE, Witkop CJ Jr, Bixler D. The dentition. In: Stewart RE, Barber TK, Troutman KC, *et al*, eds. *Pediatric dentistry: scientific foundations and clinical practice*. St Louis, MO: C V Mosby Company, 1982:91–2.
- 5 Grahnen H, Granath LE. Numerical variations in primary dentition and their correlation with the permanent dentition. *Odontol Revy* 1961;12:348–57.
- 6 Galluccio G, Castellano M, La Monaca C. Genetic basis of non-syndromic anomalies of human tooth number. *Arch Oral Biol* 2012;57:918–30.
- 7 Robinson S, Chan MF. New teeth from old: treatment options for retained primary teeth. *Br Dent J* 2009;207:315–20.
- 8 AlShahrani I, Togoo RA, AlQarni MA. A review of hypodontia: classification, prevalence, etiology, associated anomalies, clinical implications and treatment options. *World J Dent* 2013;4:117–25.

- 9 Larmour CJ, Mossey PA, Thind BS, *et al.* Hypodontia—a retrospective review of prevalence and etiology. Part I. *Quintessence Int* 2005;36:263–70.
- 10 Cobourne MT. Familial human hypodontia—is it all in the genes? *Br Dent J* 2007;203:203–8.
- 11 Development of the tooth and its supporting tissues. In: Nanci A, ed. *Ten Cate's oral histology: development, structure, and function*. 8th edn. St Louis, MO: Elsevier, 2013:86–7.
- 12 Ockeloen CW, Khandelwal KD, Dreesen K, *et al.* Novel mutations in LRP6 highlight the role of WNT signaling in tooth agenesis. *Genet Med* 2016; doi:10.1038/gim.2016.10.
- 13 Stylianou A, Liu PR, O'Neal SJ, *et al.* Restoring congenitally missing maxillary lateral incisors using zirconia-based resin bonded prostheses. *J Esthet Restor Dent* 2016;28:8–17.
- 14 Gupta A, Yelluri RK, Munshi AK. Fibre-reinforced composite resin bridge: a treatment option in children. *Int J Clin Pediatr Dent* 2015;8: 62–5.
- 15 Rosa M, Lucchi P, Ferrari S, *et al.* Congenitally missing maxillary lateral incisors: long-term periodontal and functional evaluation after orthodontic space closure with first premolar intrusion and canine extrusion. *Am J Orthod Dentofacial Orthop* 2016;149:339–48.

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