

CASE REPORT

Idiopathic Congenital Agenesis of Primary Teeth: A Report of Two Cases with Review of Literature

¹NB Nagaveni, ²P Poornima, ³Anila Valsan, ⁴Mebin G Mathew

ABSTRACT

Nonsyndromic idiopathic agenesis of primary teeth is a rare finding. Clinician's sound knowledge and preparedness for the early diagnosis and appropriate treatment are important to prevent consequences of esthetic and functional problems in children with congenitally absent primary teeth. This article reports two such cases of missing primary teeth in children of Asian origin.

Keywords: Congenital absence, Hypodontia, Oligodontia, Primary teeth.

How to cite this article: Nagaveni NB, Poornima P, Valsan A, Mathew MG. Idiopathic Congenital Agenesis of Primary Teeth: A Report of Two Cases with Review of Literature. *CODS J Dent* 2016;8(2):121-126.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

In dentistry, there are plenteous literature describing the congenitally missing permanent teeth; however, the literature is underfed regarding the number, frequency, and location of congenitally missing primary teeth.¹ Congenitally missing teeth have been defined as either absence of one or more teeth or total absence of teeth. The severity may be manifested in varying degrees. Anodontia is a total absence of teeth, whereas oligodontia designates the congenital absence of six or more teeth, and hypodontia, an absence of less than six teeth.^{2,3}

A primary tooth is defined to be congenitally missing if it has not erupted in the oral cavity and is not visible even in the radiographic examination after supportive past history of number of lost teeth due to trauma/infection or any other reasons.⁴ By around 3 years of age, all primary teeth would have erupted, therefore 3- to 4-year-old children are suitable for diagnosis of congenitally missing primary teeth by clinical examination.⁵ The use of panoramic radiography

is recommended, together with clinical examination in detecting or confirming dental development.⁶

In the present article, the prevalence, possible etiological factors, clinical implications, and management of congenitally missing primary teeth are briefly reviewed along with presentation of two clinical case reports.

CASE REPORTS

Case 1

A 4-year-old boy reported along with his parents for routine dental checkup. The patient was healthy with no evidence of any syndromic features. Intraoral examination revealed missing primary both right and left maxillary lateral incisors resulting in gap between teeth (Fig. 1). No other positive findings were observed in the patient. Orthopantomograph and intraoral periapical radiographs were made to rule out agenesis of any other teeth or any other developmental abnormalities (Figs 2 and 3). Parents were informed about the existing condition.

Case 2

A 3-year-old male patient reported with parents complaining of decayed teeth. The patient was healthy with no evidence of any syndromic features. On intraoral examination, we noticed missing of primary lower right central incisor (Fig. 4). Radiographic examination revealed the same finding, thereby confirming the congenital agenesis of primary tooth (Fig. 5).



Fig. 1: Intraoral photograph showing missing maxillary primary right and left lateral incisors

¹Professor, ²Professor and Head, ^{3,4}Postgraduate Student

¹⁻⁴Department of Pedodontics and Preventive Dentistry, College of Dental Sciences, Davangere, Karnataka, India

Corresponding Author: NB Nagaveni, Professor, Department of Pedodontics and Preventive Dentistry, College of Dental Sciences, Davangere, Karnataka, India, Phone: +918971695506 e-mail: nagavenianurag@gmail.com



Fig. 2: Orthopantomograph showing congenital agenesis of primary maxillary lateral incisors

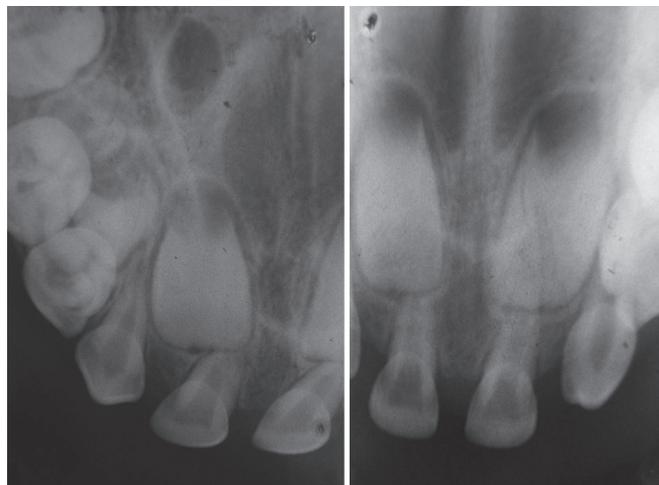


Fig. 3: Periapical radiographs confirming the agenesis of primary lateral incisors



Fig. 4: Missing primary mandibular right lateral incisor



Fig. 5: Intraoral periapical radiograph showing congenital agenesis of mandibular right lateral incisor

DISCUSSION

Etiology

Oligodontia is a rare condition that can occur in association with genetic syndromes, or as a nonsyndromic isolated familial trait, or as a sporadic finding.⁷ Although tooth agenesis is associated with more than 49 syndromes (such as hypohidrotic ectodermal dysplasia, Down's syndrome, incontinentia pigmenti, Rieger's syndrome, and chondroectodermal dysplasia.), several case reports describe nonsyndromic forms that are either sporadic or familial in nature, as reviewed by Gorlin et al.⁸ Various studies suggest both genetic and environmental etiology toward this anomaly.⁹

Hypodontia and oligodontia may be inherited as an autosomal dominant trait with incomplete penetrance and variable expression. Mutation of genes, such as Msh homeobox 1 (MSX1) and paired box protein 9 (PAX9) have been shown in families with nonsyndromic familial oligodontia.¹⁰ Certain studies suggest that the two members of the tooth family (primary teeth and its permanent successor) function rather as separate modules by the

observation that in some species, primary teeth form, but fail to be replaced. This is exemplified by studies revealing differential involvement of genes in primary *vs* permanent teeth.¹¹

The dental lamina is extremely sensitive to external insults which may lead to its severe damage or disruption. Occasional environmental factors may be trauma in the dental region (such as fractures and surgical procedures on the jaws), tumors, infections (like rubella and syphilis), physical obstruction, endocrine disturbances, multiagent chemotherapy, and radiation therapy. Based on the age of patient and dosage used, radiation is found to have more serious effects when compared with chemotherapy.^{9,11} The explanation for the presented case can be genetic, environmental, or more likely the combination of both.

Prevalence

The prevalence of hypodontia in the permanent dentition ranges between 2 and 10%, while in primary dentition, hypodontia is uncommon, with a prevalence of 0.1 to 0.9%.^{2,3} Oligodontia has a population prevalence of

Table 1: Epidemiological studies on prevalence of congenitally missing primary teeth

Author	Place	Sample size	Age group	Hypodontia prevalence	Most prevalent missing primary teeth
Ravn ¹³	Europe	4,564	3–3½ years	0.5%	17 maxillary and 7 mandibular lateral incisors, 1 mandibular primary canine
Järvinen and Lehtinen ¹⁴	Finland	1,141	3–4 years	0.9%	7 maxillary and 4 mandibular lateral incisors
Whittington and Durward ¹⁵	New Zealand	1,680	5 years	0.36%	Mandibular incisors with 1 case of missing mandibular primary canine
Daugaard-Jensen et al ²⁰	Denmark	193 cases	1.2–9 years	193 cases	119 maxillary and 53 mandibular lateral incisors
Yonezu et al ¹⁸	Japan	2,733	3 years	2.38%	Mandibular lateral incisors
Kramer et al ¹⁶	Brazil	1,260	2–5 years	0.6%	Maxillary lateral incisors followed by other incisors and canines
Chen et al ¹⁷	Taiwan	2,611	2–6 years	2%	Mandibular incisors
Kapdan et al ²²	Turkey	1,149	2–5 years	0.2%	Mandibular central incisors
Mukhopadhyay and Mitra ¹⁹	India	2,757	4–6 years	0.5%	Maxillary lateral incisors

0.3% in permanent dentition and 0.5 to 0.9% in primary dentition.¹² Prevalence of missing teeth in the primary dentition differs among different populations; a few epidemiological studies have given the incidence (Table 1), i.e., of 0.4 to 0.9% in the European^{13,14} and Brazilian population,¹⁶ and about 2¹⁷ and 2.38%¹⁸ reported among Asian children. According to a study conducted in West Bengal, India, the prevalence of hypodontia was found to be 0.5%.¹⁹ The prevalence of nonsyndromic oligodontia in Danish schoolchildren has been reported to be 0.16% and in Indore, India, prevalence of oligodontia was reported as 0.36%.⁷

Epidemiological studies suggest that there exists a difference in the prevalence of hypodontia in the primary dentition between maxilla and mandible among different races (Table 1). Agenesis was found twice as frequently in the maxillary lateral incisor region than in the mandibular lateral incisor region. Congenital absence of primary molars, canines, and maxillary central incisors was extremely rare.²⁰ Absence of more than two primary teeth was found in 8% of children with hypodontia in the primary dentition. Prevalence rates of agenesis in both dentitions are significantly higher in females compared with that in males, but gender apparently does not affect dental agenesis patterns.^{11,21}

Clinical Implications

In hypodontia among the primary dentition, the absence of teeth may be unilateral or bilateral.²³ It has been hypothesized that if hypodontia is primarily genetic, then bilateral absence would be expected to occur; and when hypodontia occurs unilaterally, it may be associated with structural anomalies of the contralateral tooth (such as microdontia or conical teeth).²⁴

Congenital dental agenesis has been reported to be associated with anomalies like a tendency toward

delayed tooth formation, reduced tooth size, ectopic maxillary canines, ectopic eruption of other teeth, short roots of teeth, enamel hypoplasia, hypocalcification, dentinogenesis imperfecta, and taurodontism.^{11,25,26} Absence of multiple teeth in primary dentition can cause impaired growth of alveolar process, constricted arches, deep bite, asymmetry of face, reduced lower facial height, reduced orofacial space, lowered tongue position, high frenal attachment, and speech and masticatory impairment, all of which can have a physiological and psychological impact on the individual.^{22,27} Cases with unilateral agenesis (especially of primary maxillary lateral incisors with missing permanent successor) was reported to be associated with dental midline shift and a molar class II relation on the same side of the agenesis. Missing mandibular incisors has been suggested as one of the chief factors affecting mandibular symphysis growth and morphology as found commonly in the Japanese populations.^{17,28}

Agenesis of a primary incisor was often but not always followed by agenesis of the succedaneous tooth. Most authors report 100% absence of the permanent successor.^{13,14} According to Daugaard-Jensen et al,²⁹ agenesis in the primary dentition is often but not always followed by agenesis in the permanent dentition. Oligodontia in the primary dentition with presence of permanent successors was reported by Ooshima et al.³⁰ However, the pattern of agenesis in the permanent dentition differed from that in the primary dentition. Incisors were most frequently missing in the primary dentition and premolars in the permanent dentition.²⁹

A summary of few case reports in dental literature on congenitally missing primary teeth and associated clinical findings is outlined in Table 2. These highlight the importance of proper and early diagnosis followed by a sequential treatment plan, for long-term stable results in the function and esthetics of such individuals.

Table 2: Case reports on congenitally missing primary teeth with associated clinical and radiographic findings

Author	Sample size	Age/ gender	Missing primary teeth	Absent permanent tooth buds	Other anomalies
Ooshima et al ³⁰	1	2/M	51, 52, 61, 71, 72, 73, 81, 82	NA	Small malformed—53, 54, 74, 84. Small and tapering—62, 63.
Shashikiran et al ¹	1	3/M	52, 53, 62, 63, 71, 72, 73, 82, 83	12, 22, 31, 32, 41, 42	Ankyloglossia and high frenal attachments with very thin alveolar ridges
Cho and Lee ³¹	3	6/M	53, 63	21: delayed eruption	A premaxillary supernumerary tooth
		6/M	53	25, 35, 45	—
		6/M	63	—	—
Venkataraman et al ³²	1	4/M	51, 52, 53, 54, 61, 62, 63, 64, 71, 72, 73, 74, 75, 81, 82, 83, 84, 85	NA	Improper speech, developing permanent tooth buds showing defective formation of dentin and roots of the permanent molars and lower central incisors
Shilpa et al ³³	1	2/M	52, 53, 54, 62, 63, 64, 71, 72, 73, 74, 81, 82, 83, 84	31, 32, 41, 42	Thin alveolar ridge
Shilpa et al ³⁴	1	2½/F	51, 52, 53, 54, 61, 62, 63, 64, 71, 72, 81, 82, 83, 84	All absent except 16, 26, 36, and 46	Severely atrophic alveolar ridges
Navin et al ⁵	1	7/F	51, 52, 54, 61, 62, 64, 71, 72, 73, 74, 81, 82, 83, 84	12, 14, 15, 22, 24, 25, 31, 32, 33, 34, 35, 41, 42, 43, 44	Conical shaped maxillary primary canines, thin and knife-edged mandibular anterior alveolar ridge and decrease in vertical dimension of occlusion
Gaur et al ²³	1	4/F	52, 62, 71, 72, 73, 81, 82, 83	12, 13, 15, 17, 22, 23, 25, 27, 31, 32, 33, 35, 37, 41, 42, 43, 45, 47	A very thin lower alveolar ridge with highly placed abnormal lingual frenum
Selva Kumar et al ³⁵	1	5/F	52, 62	12, 22, 31, 32, 41	Space in the upper front teeth
Goswami et al ³⁶	1		72, 82	35, 45, and 22	
Moses et al ⁷	1	3/F	52, 53, 62, 63, 71, 72, 73, 81, 82, 83	13, 23, 31, 32, 33, 41, 42, 43	Delayed formation of permanent tooth buds, different positional change of right second permanent molar, difficulty in speaking and chewing
Nirmala et al ³⁷	1	6/F	51, 52, 53, 54, 61, 62, 63, 64, 71, 72, 81, 82, 83, 84	11, 12, 13, 14, 15, 21, 22, 23, 24, 25, 31, 32, 33, 41, 42, 43, 44	Facial asymmetry, thin alveolus
Krishnapriya et al ⁹	1	2½/F	62	—	—
Birnboim-Blau et al ³⁸	1	7/M	73, 83	—	All first permanent molars showed taurodontism and short roots
Ephraim et al ²⁶	1	10/M	83, 84, 85	41, 42, 43, 44, 45, 46, 47, 16, 17	Ankyloglossia, difficulty in speech, asymmetry of right side of the face with lower lip placed inward, narrow V-shaped upper arch with supraeruption and deep bite and psychological impact

Management

A child with hypodontia or oligodontia should be under proper treatment planning, treatment, and coordination of treatment and review appointing. In addition to the loss of function and esthetic compromise, psychosocial development is an important concern in the oral rehabilitation of growing patients.⁷ The successful management of hypodontia is achieved by an integrated multidisciplinary approach of pedodontist, orthodontist, oral maxillofacial surgeon, prosthodontist, and speech therapist.

The general principle in management is to deal with the space within the dental arches, i.e., a space closure

in less severe cases, while prosthetic replacements (in the form of dentures, crowns, bridges, autotransplantation, dental implants, etc.) as well as some orthodontic tooth movement in more severe cases.³⁹ Nagaveni et al⁴⁰ reported a rare occurrence of concomitant occurrence of canine transmigration and symmetrical agenesis of mandibular permanent incisors.

A prosthodontic rehabilitation, with a removable partial denture at an early stage and later rehabilitation of teeth with osseointegrated implants, is fundamental in these situations, attempting to provide a functional and esthetic solution that will allow the child as normal

a lifestyle as possible. A removable partial denture which can be modified according to the eruption status of the teeth can be fabricated, providing reasonable esthetic result and an acceptable masticatory function without unnecessary damage to tissues, which needs to be changed every 6 months serving as interim prosthesis.⁷ The correction of associated anomalies like expansion of narrow maxilla, correction of ankyloglossia, etc., along with space maintenance should be sequenced in a way that it produces a long-term outcome.

In those cases of existing permanent successor teeth, it is advised to examine the child periodically in order to avoid possible space loss due to tipping of adjacent teeth toward the space created by the missing tooth. If space is already lost, it might be necessary to gain it back and then to use a space maintainer, till the permanent tooth is erupted.³⁷

Management in cases of congenitally missing primary teeth will depend on the pattern and severity of tooth absence, the amount of spaces present, eruption pattern, growth potential, health of teeth and surrounding structures, occlusion and interocclusal space, and, of course, the patient's and caretaker's attitude.²¹

CONCLUSION

Hypodontia requires great care with extensive and complex treatments. Early identification of missing primary dentition anomalies can guide the dentist to prevent future permanent dentition ailments and planning interceptive dental treatment at the appropriate time. Therefore, in the mixed dentition period, it is very essential to monitor these patients, thereby various treatment options can be proceeded in future, creating less psychological distress for such patients.

REFERENCES

1. Shashikiran ND, Karthik V, Subbareddy VV. Multiple congenitally missing primary teeth: report of a case. *Pediatr Dent* 2002 Mar-Apr;24(2):149-152.
2. Mc Donald, RE.; Avery, DR.; Hartsfield, JK Jr. Acquired and developmental disturbances of the teeth and associated oral structures. In: Mc Donald RE, Avery DR, Dean JA, editors. *Dentistry for the child and adolescent*. 8th ed. St. Louis (MO): CV Mosby Co.; 2011. pp. 107-147.
3. Sauk, JJ Jr. Defect of the teeth and tooth bearing structures. In: Braham RL, Morris ME, editors. *Textbook of pediatric dentistry*. 2nd ed. New Delhi: CBS Publishers and Distributors; 1990. pp. 72-104.
4. White, SC.; Pharaoh, MJ. *Oral radiology principles and interpretation*. 5th ed. Philadelphia (PA): Mosby Co.; 2004. p. 332.
5. Navin HK, Nagarathna C, Shakuntala BS, Mathew S. Hypodontia of primary and permanent dentition: a case report. *Int J Contemp Dent* 2011 Jan;2(1):76-79.
6. Pirinen, S.; Thesleff, I. Development of the dentition. In: Thilander B, Ronning O, editors. *Introduction to orthodontics*. Stockholm: Lic Forlag; 1995. p. 41-43.
7. Moses J, Gurunathan D, Rangeeth BN, Kannan KS. Non-syndromic oligodontia of primary and permanent dentition: 5 year follow up—a rare case report. *J Clin Diagn Res* 2013 Apr;7(4):776-779.
8. Gorlin, RJ.; Cohen, MM Jr.; Levin LS. *Syndromes of the head and neck*. New York: Oxford Univ. Press; 1990.
9. Krishnapriya V, Kumar S, Singh V, Mehrotra N. Unilateral hypodontia in primary dentition: does it affect the permanent successor: a literature review and case report. *J Young Med Res* 2014 Dec;1(2):1-7.
10. Mostowska A, Kobiela A, Trzeciak WH. Molecular basis of non-syndromic tooth agenesis: mutations of MSX1 and PAX9 reflect their role in patterning human dentition. *Eur J Oral Sci* 2003 Oct;111(5):365-370.
11. De Coster PJ, Marks LA, Martens LC, Huysseune A. Dental agenesis: genetic and clinical perspectives. *J Oral Pathol Med* 2009 Jan;38(1):1-17.
12. Vastardis H. The genetics of human tooth agenesis: new discoveries for understanding dental anomalies. *Am J Orthod Dentofacial Orthop* 2000 Jun;117(6):650-656.
13. Ravn JJ. Aplasia. Supernumerary teeth and fused teeth in the primary dentition. An epidemiologic study. *Scand J Dent Res* 1971 Feb;79(1):1-6.
14. Järvinen S, Lehtinen L. Supernumerary and congenitally missing primary teeth in Finnish children. An epidemiologic study. *Acta Odontol Scand* 1981 Feb;39(2):83-86.
15. Whittington BR, Durward CS. Survey of anomalies in primary teeth and their correlation with the permanent dentition. *N Z Dent J* 1996 Mar;92(407):4-8.
16. Kramer PF, Feldens CA, Ferreira SH, Spiguel MH, Feldens EG. Dental anomalies and associated factors in 2- to 5-year old Brazilian children. *Int J Paediatr Dent* 2008 Nov;18(6):434-440.
17. Chen YH, Cheng NC, Wang YB, Yang CY. Prevalence of congenital dental anomalies in the primary dentition in Taiwan. *Pediatr Dent* 2010 Nov-Dec;32(7):525-529.
18. Yonezu T, Hayashi Y, Sasaki J, Machida Y. Prevalence of congenital dental anomalies of the deciduous dentition in Japanese children. *Bull Tokyo Dent Coll* 1997 Feb;38(1):27-32.
19. Mukhopadhyay S, Mitra S. Anomalies in primary dentition: their distribution and correlation with permanent dentition. *J Nat Sci Biol Med* 2014 Jan;5(1):139-143.
20. Daugaard-Jensen J, Nodal M, Kjær I. Pattern of agenesis in the primary dentition: a radiographic study of 193 cases. *Int J Paediatr Dent* 1997 Mar;7(1):3-7.
21. Nagaveni NB, Bajaj M, Muthusamy K, Poornima P, Pai SM, Subba Reddy VV. Concomitant mandibular hypo-hyperdontia: report of two rarest cases with the literature review. *Int J Contemp Dent Med Rev* 2014;6:1-6.
22. Kapdan A, Kustarci A, Buldur B, Arslan D, Kapdan A. Dental anomalies in the primary dentition of Turkish children. *Eur J Dent* 2012 Apr;6(2):178-183.
23. Gaur A, Mallikarjuna RM, Sujana SG. Multiple congenital missing primary teeth: a case report. *J Int Oral Health* 2011 Aug;3(4):29-32.
24. Pinho T, Tavares P, Maciel P, Pollmann C. Developmental absence of maxillary lateral incisors in the Portuguese population. *Eur J Orthod* 2005 Oct;27(5):443-449.

25. Poornima P, Shruthi AS, Nagaveni NB, Roopa KB. A case report: nonsyndromic familial oligodontia. *Int J Infor Res Rev* 2015 Jul;2(7):853-855.
26. Dhanrajani PJ. Hypodontia: etiology, clinical features and management. *Quintessence Int* 2002 Apr;33(4):294-302.
27. Ephraim R, Rajamani T, Feroz TM, Abraham S. Agenesis of multiple primary and permanent teeth unilaterally and its possible management. *J Int Oral Health* 2015 May;7(5): 68-70.
28. Endo T, Ozoe R, Kojima K, Shimooka S. Congenitally missing mandibular incisors and mandibular symphysis morphology. *Angle Orthod* 2007 Nov;77(6):1079-1084.
29. Daugaard-Jensen J, Nodal M, Skovgaard LT, Kjær I. Comparison of the pattern of agenesis in the primary and permanent dentitions in a population characterized by agenesis in the primary dentition. *Int J Paediatr Dent* 1997 Sep;7(3):143-148.
30. Ooshima T, Sugiyama K, Sobue S. Oligodontia in the primary dentition with permanent successors: report of case. *ASDC J Dent Child* 1988 Jan-Feb;55(1):75-77.
31. Cho SY, Lee CK. Congenitally missing maxillary primary canines: report of three cases. *Int J Paediatr Dent* 2006 Nov;16(6):444-447.
32. Venkataraghavan K, Anantharaj A, Prasanna P, Sudhir R. Oligodontia in the primary dentition: report of a case. *J Dent Child (Chic)* 2007 May-Aug;74(2):154-156.
33. Shilpa, Thomas AM, Joshi JL. Idiopathic oligodontia in primary dentition: case report and review of literature. *J Clin Pediatr Dent* 2007 Fall;32(1):65-67.
34. Shilpa, Mohapatra A, Reddy CP, Sivakumar N. Congenital absence of multiple primary teeth. *J Indian Soc Pedod Prev Dent* 2010 Oct-Dec;28(4):319-321.
35. Selva Kumar H, Swaminathan K, Thomas E, Varghese JS. Congenitally missing primary lateral incisors along with permanent successors: a case report. *Int J Dent Case Rep* 2012;2(1):47-49.
36. Goswami M, Chaitra TR, Singh S, Kulkarni AU. Congenitally missing primary mandibular lateral incisors: a case of rare occurrence. *BMJ Case Rep* 2012 Aug;2012:006472.
37. Nirmala SV, Gokhale N, Sivakumar N, Quadar MA. Agenesis of multiple primary teeth and its rehabilitation: a case report. *Int J Clin Pediatr Dent* 2013 Jan-Mar;6(1):55-57.
38. Birnboim-Blau G, Spierer S, Keinan D. Congenital bilateral missing primary mandibular canines while their successors exist: a case report. *Dentistry* 2014 Mar;4:224.
39. Kokich V Jr. Early management of congenitally missing teeth. *Semin Orthod* 2005 Sep;11(3):146-151.
40. Nagaveni NB, Radhika NB, Satisha TS. Concomitant occurrence of canine transmigration and symmetrical agenesis of mandibular incisors. A case report. *Bangladesh J Med Sci* 2011 Apr;10(2):132-136.