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J Pharm Bioallied Sci. 2014 Jul; 6(Suppl 1): S200–S203.
doi: 10.4103/0975-7406.137469
PMCID: PMC4157270
PMID: 25210374

Nonsyndromic oligodontia in siblings: A rare case report

Veerabadhran Mahesh Mathian, Murugesan Gawthaman, Ramachandran Karunakaran, Selvaraj Vinodh, Sundaram Manikandan, and Alagappan Meenakshi Sundaram[1]

Abstract

A syndrome is a medical condition that is characterized by a particular group of signs and symptoms, involving several organ systems. Oligodontia is defined as the developmental absence of six teeth or more, excluding third molars. Oligodontia can be classified as syndromic or nonsyndromic. Nonsyndrome oligodontia is a developmental dental anomaly without involvement of other organs. The consequences of missing teeth include abnormal occlusion or altered facial appearance, which can cause psychological distress in some patients. The management of oligodontia usually requires a multidisciplinary approach. The present article reports a case of oligodontia in siblings of the same family with no apparent systemic abnormalities.

KEY WORDS: Hypodontia, oligodontia, permanent teeth, primary teeth, tooth agenesis

Hypodontia – missing of one or few teeth is more common in the permanent dentition. Oligodontia – is agenesis of numerous teeth. Anodontia – is the extreme of oligodontia were there is a total absence of any dental structure. The incidence of oligodontia usually varies from 0.08% to 0.16%. It is a rare condition and the occurrence is common in girls in the ratio of 3:2.[1] The most commonly missing teeth are the mandibular second premolars followed by maxillary second premolar and maxillary lateral incisors and the rarely missing teeth are the maxillary central incisors, maxillary and mandibular first molars and canines.[2]

Several studies have reported that bilateral agenesis of the tooth was more common than the unilateral form.[3,4] Grahnen found that if either parent had one or more congenitally missing teeth, there was an increased likelihood that their children also would be affected.[5] Syndromic and nonsyndromic form of oligodontia can be differentiated by conducting thorough physical examination of hairs, nails, sweat glands, eyes, and to check for any congenital disorders.

Case Reports

Two male patients of 16 and 14 years reported to the Department of Pedodontia with similar complaints of noneruption of teeth in upper and lower jaws [Figure 1]. Both patients were children of same parents. No history of consanguineous marriage is reported between parents. The patients had no history of trauma or extractions. The patients on questioned revealed that their mother also had numerous teeth missing extra oral examination revealed a face with normal facial profile and normal skeletal dental base relations. Physical examination of both the patients revealed no abnormality in either hairs or nails, perspiration was normal and no congenital clefts of the lip or palate was seen.



Figure 1

Clinical photograph of siblings

Case 1: 16 years boy

On intra oral examination the following teeth were present clinically 17, 16, 55, 53, 52, 63, 26, 27, 75, 34, 33, 31, 41, 43, 44, and 85 [Figures 2 and 3]. Dental caries was found in 16, 26, and 75. The retained deciduous teeth were attrited and 31 and 41 were conical shaped. The oral mucosa and gums are normal. Except dental anomalies the boy had normal physique. Suspecting the congenital absence of permanent teeth panoramic radiograph was taken, which showed absence of 17 permanent teeth [Figure 4]. The caries teeth were treated conservatively and removable partial dentures were given for the missing teeth [Figure 5].



Figure 2

Intaoral maxillary view showing 17, 16, 55, 53, 52, 63, 26, and 27



Figure 3

Intraoral mandibular view showing 75, 34, 33, 31, 41, 43, 44, and 85

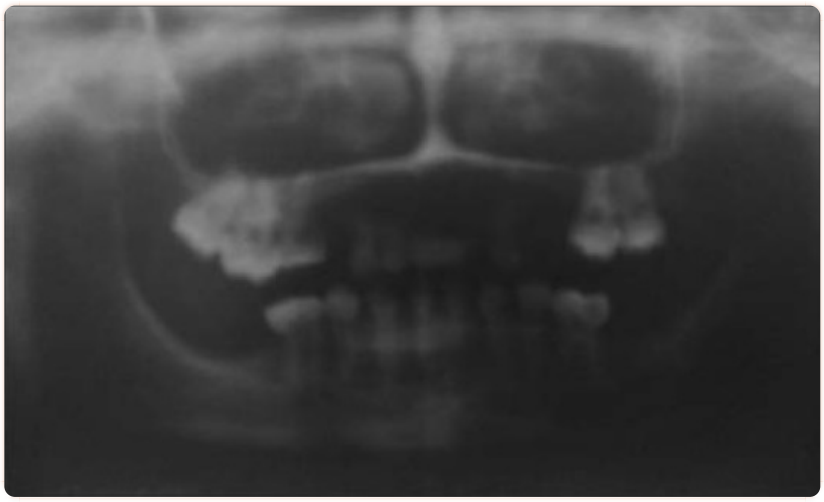


Figure 4

Orthopantomogram of 16-year-boy



Figure 5

Removable partial denture inserted

Case 2: 14 years boy

On examination, the following teeth were present 16, 52, 63, 64, 25, 26, 35, 34, 73, 72, 42, 43, and 44 [Figures 6 and 7]. Dental caries was noted in 63 and 52 was mobile. The retained deciduous teeth were attrited. Suspecting the congenital absence of permanent teeth panoramic radiograph was taken which showed absence of 21 permanent teeth [Figure 8]. 52 and 63 teeth were extracted under local anesthesia. Removable partial dentures were given once the extracted sites healed [Figure 9].

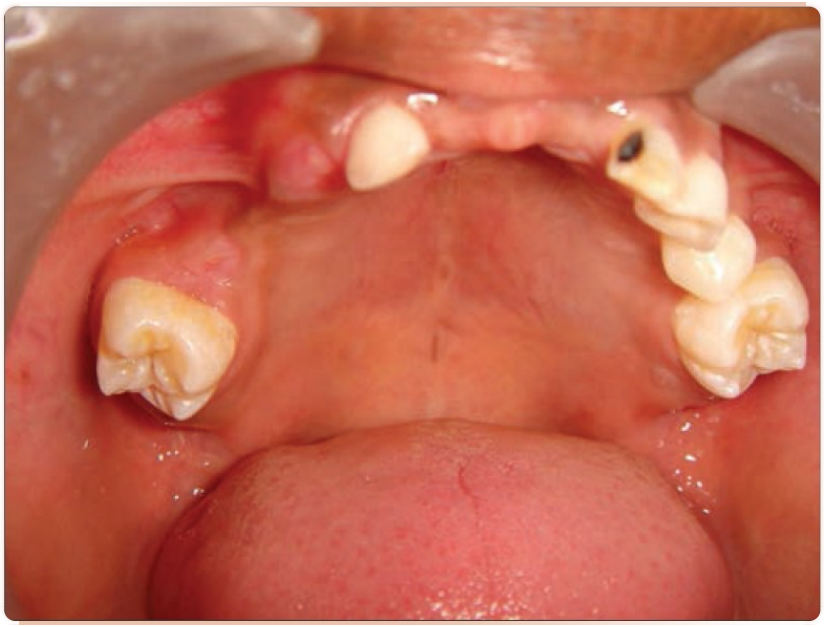


Figure 6

Intaoral maxillary view showing 16, 52, 63, 64, 25, and 26



Figure 7

Intraoral mandibular view 35, 34, 73, 72, 42, 43, and 44



Figure 8

Orthopantomogram of 14-year-boy



Figure 9

Removable partial denture inserted

Discussion

Oligodontia can occur in association with various genetic syndromes, such as ectodermal dysplasia, incontinentia pigmenti, Down syndrome, Rieger syndrome, Wolf-Hirschhorn syndrome, Van der Woude syndrome, ectrodactyly-ectodermal dysplasia-clefting syndrome, cleft lip palate ectodermal dysplasia syndrome, oral facial digital syndrome type I, Witkop tooth-nail syndrome and Fried syndrome. When oligodontia is associated with a syndrome there may be abnormalities of the skin, nails, eyes, ears, and skeleton.[1] In this case report, oligodontia is not associated with any syndrome, which is a rare finding. Oligodontia is often related with conical shaped teeth, microdontia, delayed eruption of permanent teeth, an increased freeway space, and retention of deciduous teeth.[6] Our case report also had conical shaped teeth and retention of deciduous teeth.

The biologic basis for the congenital absence of permanent teeth is partially explained by the failure of the lingual or distal proliferation of the tooth bud cells from the dental lamina. In many cases, multiple genetic and environmental factors are acting together. The causes of hypodontia are attributed to environmental factors such as irradiation, tumors, trauma, hormonal influences, rubella, and thalidomide or to hereditary genetic dominant factors, or to both.[7] MSX1 and PAX9 genes play a key role in early tooth development. All mutations of PAX9 identified to date have been associated with nonsyndromic form of tooth agenesis.[8]

Tsai *et al*. have reported a case of oligodontia in a 6-year-old girl with congenital absence of 16 permanent teeth.[9] Rasmussen P reported nonsyndromic 9 cases with the absence of 14-24 teeth excluding third molar.[10] The present report shows congenitally missing 17 permanent teeth in 16-year-old boy and 21 permanent teeth in 14-year-old boy. No identifiable etiology in both the patients was found. Since both patients were young and growing, the possibility of implants was ruled out and was treated with removable partial dentures.

Conclusion

Patients with oligodontia should be evaluated carefully by a geneticist for the presence of any syndromes and managed appropriately. Oligodontia should not be neglected as it may result in various disturbances such as abnormal occlusion, altered facial appearance which may cause psychological distress, difficulty in mastication and speech. These patients should be evaluated carefully by specialists of various disciplines and early diagnosis and treatment planning should be made to achieve both prosthetic and aesthetic functionality of teeth.

Footnotes

Source of Support: Nil

Conflict of Interest: None declared.

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