International Journal of Dental Research
www.dentaljournal.net
Online ISSN: 2664-9055, Print ISSN: 2664-9047
Received: 10-12-2023, Accepted: 25-12-2023, Published: 10-01-2024
Volume 6, Issue 1, 2024, Page No. 1-3

Hypodontia: In search of elucidation

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Abstract
Hypodontia is a developmental and a congenital condition characterised by the fewer than normal teeth. There are different terms used for this condition such as severe hypodontia, oligodontia and anodontia. It is one of the most common dental anomalies and has a negative impact on looks and functions. It may occur as a part of a recognised genetic syndrome or as a nonsyndromic isolated trait. Reported prevalence of hypodontia varies from 1.6% to 6.9% and this range does not include missing third molars. Both genetic and environmental factors play an important part in its eiology but the role of genetic factors is more justified. Hypodontia is one factor in the clinical indices used by orthodontists when prioritising treatment, so reflecting the clinical importance of the condition for the patient concerned. The main aim of this review is to develop a greater knowledge and understanding of the causes, features and management of this dental condition.

Keywords: Hypodontia, oligodontia, anodontia, developmental anomaly

Introduction

Disturbances during the early stages of tooth formation may result in the developmental or congenital absence of one or more teeth. This condition has been described in the literature using a range of terms and the most widely used term is hypodontia, used by many authors to describe the whole spectrum of the disorder from the absence of a single tooth to the rare absence of all teeth (termed anodontia). However absence of third permanent molars is generally not considered while assessing the presence and severity of hypodontia. To assist in diagnostic classification, the degree of severity of hypodontia has been arbitrarily described as:

- **Mild:** 1–2 missing teeth
- **Moderate:** 3–5 missing teeth
- **Severe:** 6 or more missing teeth

Some authors have given the suggestion that the absence of one to six teeth should be termed hypodontia, while the absence of more than six teeth should be termed oligodontia (Arte and Pirinen, 2004; Polder *et al*. 2004). [1] Others have proposed that the term oligodontia should be further limited to describe the absence of six or more teeth with associated systemic manifestations, as seen in several syndromes (Nunn *et al*, 2003). [2] To reflect the differences in terminology, a further sub - division of hypodontia and oligodontia has been proposed into isolated hypodontia/oligodontia (non - syndromic) and syndromic hypodontia/oligodontia (associated with syndromes) (Schalk van der Weide *et al*, 1992; Arte and Pirinen, 2004) [1, 3]. Current terminology also demonstrates geographical variations. The term oligodontia is often preferred in Europe, whereas the descriptive terms agenesis or multiple dental agenesis are often used in the USA. Partial anodontia is the term which was once widely used is now considered largely obsolete (Jones 2009) [4].

Prevalence

Hypodontia is relatively uncommon in deciduous dentition. The prevalence of 0.1 – 0.9% is equally distributed between males and females. It is most common in the anterior maxilla, with the lateral incisors being most frequently affected. In mild cases, hypodontia of the primary dentition often goes unnoticed or may be wrongly dismissed as of some interest but seemingly unimportant. Diagnosis in a younger patient is frequently made by general dental practitioners [5]. Prevalence of hypodontia in permanent dentition has mainly suffered because of small sample sizes and varies with wide range from 0.3% to 36.5%. It is more commonly seen in females than males. (Polder *et al*, 2004) [6]. In order to increase the sample size and thus improve the reliability of population data, Polder *et al*. (2004) conducted a meta - analysis which has added significantly to our knowledge. It included data from 33 studies, with a total sample size of approximately 127,000 individuals, and concluded that the prevalence of hypodontia in the permanent dentition varied between continents, racial groups and genders.

The frequency of absent teeth in descending order conducted by Polder *et al* 2004 was:

Mandibular second premolar (3.0%), Maxillary lateral incisor (1.7%), Maxillary second premolar (1.5%), Mandibular central incisor (0.3%), Mandibular lateral incisor and maxillary first premolar (0.2%), Mandibular fi rst premolar (0.15%), Mandibular second molar and maxillary canine (0.1%), Maxillary second molar (0.05%), Maxillary first molar (0.03%), Mandibular canine (0.02%), Mandibular first molar (0.01%), Maxillary central incisor (0.005%). This supports one of the widely accepted sequences of missing teeth as: Mandibular second premolar > Maxillary lateral incisor > Maxillary second premolar > Mandibular incisor [6].

Etiology

1. **Environment & Genetic factors:** Several theories say that both genetic and environmental factors may play a role in the etiopathogenesis of hypodontia. Butler's Field Theory for the evolutionary development of mammalian teeth (Butler, 1939) suggested that the most

mesial tooth in each morphological series was the most genetically stable and consequently was rarely missing [7]. Such teeth were designated as ' key teeth ' and included the central incisors, canines, first premolars and first molars. In contrast, teeth at the end of each field showed less genetic stability. This led to the concept of stable and unstable elements of the dentition (Bailit, 1975) [8]. This principle was further supported by Bolk's Theory of Terminal Reduction (Rózsa *et al*, 2009). This proposed that the evolutionary process was leading to the reduction of the distal element of tooth groups, resulting in the more frequent absence of second premolars, lateral incisors and third molars [9]. Postnatal nutrition, disease, general health and climatic conditions had little influence on hypodontia. The intra-uterine effects of drugs such as thalidomide have been associated with the development of hypodontia (Axrup *et al*., 1966) [10]. Other environmental factors include trauma, jaw fracture or surgery or itrogenic damage to the tooth germ from traumatic extraction of the overlying primary tooth (Nunn *et al*, 2003) [2]. Hypodontia has also been associated with cleft lip and palate usually localised to the maxillary lateral incisor. It is considered due to physical obstruction in the developing dental lamina and more recently a defect in the Msx 1 gene has been identified [11]. (Alappat *et al*. 2003).

Inheritance patterns

Examination of monozygotic twins and triplets indicates there is a familial pattern in hypodontia. This is thought to occur by an autosomal dominant process with incomplete penetrance of up to 86% (Arte and Pirinen, 2004) [11]. There is also an association between hypodontia and microdontia which was based on an underlying continuum of tooth size with thresholds, whereby there is a progressive reduction in the size of reaches a certain threshold below which the developing tooth germ degenerates, so producing hypodontia.

Tooth development

Development of the dentition is a complex process involving a series of epithelial – mesenchymal interactions, and involving growth factors, transcription factors, signalling pathways and other morphogens (Thesleff, 2000) [12]. With such complexity, it is not surprising that disturbances can occur in the process, potentially resulting in tooth agenesis. At the molecular level during odontogenesis, epithelial – mesenchymal signalling is under the control of members of the Wnt (wingless), Hh (hedgehog), Fgf (fi broblast growth factor) and Bmp (bone morphogenic protein) gene families. Defects in any of these pathways can result in disorders of tooth number (hypodontia or supernumerary teeth), tooth morphology (tooth size and shape) and tooth mineralisation. Of particular interest in hypodontia are the genes called Msx1 (muscle segment homeobox 1) and Pax9 (paired box 9), which are homeobox transcription factors involved in early odontogenesis under the control of Bmp and Fgf signalling. More recently, defects in a third gene, Axin2, have been identified as having a possible association with severe hypodontia [13].

Syndromic associations

Several syndromes exhibit hypodontia as one of their features, and many of these have demonstrated gene defects (Online Mendelian Inheritance in Man (OMIM) database).

Mutations in the homeobox transcription factor Pitx2 (paired - like homeodomain transcription factor 2) are associated with Rieger syndrome, an autosomal dominant disorder with ocular, umbilical and dental defects. Mutations in p63 are associated with syndromes involving hypodontia that include digital disorders like syndactyly and ectrodactyly, facial clefts, cleft lip and palate, and ectodermal dysplasia. Mutations in Msx1 have also been associated with isolated cleft lip and palate, and Witkop (tooth and nail) syndrome. The genetic inheritance of the family of ectodermal dysplasias has been investigated. There are over 190 different types of this condition, and while several genes have been implicated, the exact numbers of genes have yet to be determined. Hypohidrotic ectodermal dysplasia (HED) is a disorder in which the sweat glands are reduced in number, which has received the greatest attention. Defects in the Xq12 – Xq13 site on the X chromosome, which encodes for the protein ectodysplasin - A (Eda), have been shown to be associated with an X - linked inheritance pattern (XHED). The same chromosome site defects have been identified in non - syndromic isolated X - linked hypodontia. Mutations in the modulator gene Nemo, a downstream target of Eda signalling, have also been associated with X - linked HED. Eda has a role in epithelial – mesenchymal signalling, and is expressed in the development of the ectodermal structures that develop from epithelial placodes, including skin, sweat glands, hair, nails and teeth. In severe cases, the dental effects can result in anodontia. Hypohidrotic ectodermal dysplasia is also associated with both autosomal dominant and autosomal recessive patterns of inheritance through mutations in the ectodysplasin - A receptor (Eda - R) [14]. Understanding the genetics of hypodontia is important for diagnostic and counselling purposes. It also presents the challenges of employing tissue engineering and stem cell technology as therapeutic alternatives [15].

Clinical features

Missing teeth is inherent to the condition. Patterns of missing teeth in hypodontia are very variable with regards to number and form and the jaw that is affected, even within siblings [16]. Some 5% of populations reported to date have at least one missing tooth (excluding third molars) with a range of between 2.2% and 7.7%. The majority of patients with hypodontia have one or two teeth missing and the percentage with larger numbers of missing teeth is much smaller. These data relate principally to the permanent dentition and there is little information available for prevalence in the primary dentition, although it does appear to be much less common with a reported prevalence of approximately 0.5%. There are very limited published data for the prevalence of anodontia which is very uncommon. While hypodontia appears to be more frequently reported than was historically the case, meta - analysis of the data on prevalence has not demonstrated an increase in its incidence in Caucasian people. Missing teeth define hypodontia, but other features may be present, including:

- Microdontia
- Conical teeth
- Ectopic eruption
- Retained primary teeth
- Tooth surface loss
- Reduced alveolar development
- Abnormally large freeway space
- Delayed eruption of permanent teeth
- Altered craniofacial morphology

Patients complaints

The most common complaints are:

- Appearance (frequent)
- Speech problems (less frequent)
- Mastication difficulties (less frequent)

Management

The treatment for patients with hypodontia is often complex, with the delivery of care best delivered through an integrated hypodontia clinic with access to the expertise of a range of dental and medical specialties. The roles of a hypodontia clinic include the within - unit treatment of patients, the provision of treatment plans for outreach care, patient and family counselling, education and training, and research. It is also important to recognise that hypodontia is a lifetime problem, and frequently cannot be managed completely by early intervention. Treatment must be planned on a longitudinal basis to give optimised outcomes over a lifetime, and often requires phases both of active treatment and long - term clinical maintenance. The clinical team must therefore possess suffi cient skills to plan treatment with a perspective on current and future needs. A number of patient support groups have been set up world - wide by affected families, providing a valuable service for patients through specialist advice, counselling and hypodontia research funding [18].

Conclusion

Various terms have been used to describe the developmental absence of teeth, including hypodontia, oligodontia, anodontia and dental agenesis. Hypodontia may present as an isolated condition, or may be associated with syndromes including the ectodermal dysplasias. Prevalence varies between continents, racial groups and genders. In Caucasians, the prevalence is 4 – 6% with a female to male ratio of approximately 3:2. A number of homeobox genes associated with tooth development have been implicated in the aetiology of hypodontia, including Msx1, Pax9 and Axin2. Gene therapy may offer the potential for bioengineering of replacement teeth as a novel approach to managing hypodontia [19].

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