Eur. J. Prosthodont. Rest. Dent., Vol.15, No. 2, pp 55-60
Printed in Great Britain

Oligodontia and Associated Characteristics: Assessment in View of Prosthodontic Rehabilitation

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Abstract - *Bibliographic evidence regarding oligodontia was reviewed to extract information necessary for a systematic review for a prosthodontic approach to mangagement. Syndromic oligodontia, appearing as a symptom of many syndromes, was distinguished from isolated, or non-syndromic, which is an independent trait. Although a rare disorder, oligodontia has always been considerably researched, especially concerning its prevalence and genetic background. Non-syndromic oligodontia has been associated with the presence of small and misshapen natural teeth, orofacial clefting and reduced saliva secretion. A typical maxillofacial morphology has also been reported, which seems to result from the lack of dental and functional compensation and not from an altered growth pattern. Syndromic oligodontias also exhibit the above basic features but are complicated by each syndrome's specific characteristics. Prosthodontic treatment of individuals with oligodontia must anticipate the dental and oral clinical characteristics and provide with continuing support and preservation of proper maxillofacial relationships.*

KEY WORDS: Oligodontia, Oral abnormalities, Prosthodontic treatment

INTRODUCTION

Undisrupted eruption and settlement of the primary and permanent dentition contributes significantly to a harmonic growth and development of the face and supports proper function and aesthetics. Oligodontia, the congenital absence of many permanent teeth, constitutes a considerable burden during the early years of life, both from a functional and an aesthetic aspect; it also induces a risk for psychological disturbances.

It is now well established that young individuals with oligodontia must be systematically followed during their childhood and adolescence years by a team of closely collaborating specialists[1]. A key part of the treatment is taken by the prosthodontics, which supports appearance and function during the stages of growth[2]. Prosthodontic treatment of edentulous children is a long process; it deals with the problem of edentulism providing a series of sequential temporary or provisional appliances and it only delivers a final restoration by the time the patient reaches adulthood. It is also a challenging assignment; besides the practical difficulties and specific characteristics, it must also be coordinated with the complicated and sensitive procedures involved in craniofacial growth. The prosthodontist must therefore possess a concise knowledge of its background and manifestations of the conditions. The aim of the present review was to provide basic information on oligodontia, emphasizing issues that may affect the course of prosthodontic intervention. Characteristics and manifestations related to lack of teeth are highlighted and their impact on the planning of the prosthodontic interventions is discussed.

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Terms and definitions

A preliminary search was conducted to identify proper terms and provide relevant definitions. Anodontia is defined as the congenital absence of all teeth[3-5]. Hypodontia is the congenital absence of one or more teeth, excluding the third molars[3,5-8]. The same definition applies to the term "dental" or "teeth agenesis", which refers to the developmental disorder involved[9]. Lack of a considerable number of teeth has been termed advanced hypodontia, severe hypodontia or oligodontia. Definitions differ slightly; advanced or severe hypodontia is defined as congenital absence of four or more[10-13], five or more[12], or six or more [6,8,14-19] permanent teeth, third molars excluded.

The term "oligodontia" seems to be preferred recently to denote congenital lack of six or more teeth (again, except third molars)[6,8,14-17,19-21]. The authors of the present study have adopted this term. A further classification identifies oligodontia as part of a syndrome (syndromic oligodontia, oligodontia/S) as opposed to isolated or non-syndromic oligodontia (oligodontia/I), where congenitally missing teeth are not accompanied by any other ectodermal symptom[17,18]. Non-syndromic oligodontia should be distinguished from induced or false anodontia, a term used to describe clinically missing teeth as a result of extractions, and pseudoanodontia or pseudohypodontia, the condition in which permanent teeth are absent because of impaction, delayed eruption or early exfoliation[18,22].

NON-SYNDROMIC (ISOLATED) OLIGODONTIA

Non-syndromic oligodontia has been thoroughly researched concerning its prevalence, aetiology and characteristics. In many publications it is simply referred as oligodontia. Consequently, the search terms used in the present review were "oligodontia", in combination with

attributes of interest such as "prevalence", "aetiology", "craniofacial morphology", "teeth size" etc. An additional search was also performed, using the terms "hypodontia" and "teeth/dental agenesis" instead of "oligodontia".

Prevalence

The prevalence of permanent dentition hypodontia amounts up to 2.6-8%[5,19,21,23-29]. Racial differences affect greatly the reported percentages[9], but most reports agree that second premolars and upper lateral incisors are the most common absent teeth[13,18,19,25-31]. Severe hypodontia with 4 or more absent teeth appears in a percentage of 0.2-1.1%[13], which drops to 0.5%[20] or even less[9,19,32] for oligodontia cases (6 or more missing teeth).

No specific pattern of agenesis has been convincingly identified. Neither gender, nor jaw (upper or lower) or guadrant seem to prevail in teeth absence. Also, the Butler's field theory, according to which distal teeth within each morphogenic class are the most variable, is not universally supported[33]; at least, it does not seem to apply in cases of severe hypodontia[13,19,20].

Primary dentition hypodontia is rarely observed (0.1-0.9%)[4,6,21,34-36]. Whenever it is encountered, it usually refers to incisors[4,26,30,34,36] and is followed by permanent dentition hypodontia[4,6,21,34,36-38].

Aetiology

By definition, oligodontia is a congenital disorder; it exists from birth[39] and originates from failure of teeth sperms to develop during the embryonic stages. A hereditary nature can be reasonably assumed and several publications look into the heredity of the trait by examining familial cases. All types of inheritance mode (autosomal –dominant and recessive- and X-linked) have been noted[40]. Local factors such as infections[6,18,21,31,41,42], mechanical trauma or accidental removal of a tooth germ[13,21,31], effects of cytostatica and radiation[6,13,18,21,41,42] and hormonic or metabolic disturbance[6,18,21], acting prenatally, are also reported to induce agenesis of teeth, probably via mutagenic activity.

The complexity of the inheritance patterns and the variability of the resulting phenotypes led to the assumption that teeth agenesis is a polygenic trait. Brook[41] reported an association between teeth number and size and proposed a multifactorial model of a continuous scale with thresholds, which linked microdontia and hypodontia in terms of cause[41]. Kjaer[43] also proposed an aetiological model connecting teeth agenesis with nerve, mucosal and bone abnormalities.

The recent advent of gene technology, which gave developmental biology research a new momentum, promoted the understanding of the mechanisms involved in teeth agenesis, through transgenic mice research and genetic screening. Early stage tooth morphogenesis is regulated by sequential and reciprocal signalling between the embryonic epithelium and mesenhyme[44]. More than 100 gene transcription factors and genes in signalling networks are involved in the process[40]; their deficient function can arrest the procedure before or at the bud stage. Indeed, mutations of two of these genes (MSX1 and PAX9) have been associated with tooth agenesis in humans[40]. These mutations result in a characteristic pattern of absent teeth; third molars and second premolars for MSX1, and molars for PAX9[40,45]. However, the agenesis pattern may be expressed with a great phaenotypical variation, depending on the localization of mutations[45]. A good example of the complexity of the issue is the recent discovery of a mutation in *AXIN2*, a Wnt-signaling receptor, which causes teeth agenesis together with a predisposition for colorectal cancer[46]. Considering that a lot more genes, besides the above, are expressed in early dental epithelium, and all of them are potential candidates for tooth agenesis, it becomes obvious that the genetic background of teeth agenesis is complicated and not yet clearly understood[40]. Nevertheless, the current results have been enlightening and the rigorous research being conducted is promising for future progress, concerning not only aetiology, but also prevention methods and therapeutic potential[47].

Oral abnormalities accompanying oligodontia

Oligodontia has been studied in relation to teeth abnormalities such as reduced size and delayed teeth development[6,10,15,16,48]. A relationship between hypodontia and microdontia has been found by a number of reports[10,16,20,41]. Mesiodistal reduction is mostly referred, but an analogous labiolingual reduction was also observed[16,20]. Microdontia becomes more pronounced as oligodontia becomes more severe[17,29], and seems to affect anterior more than posterior teeth[10] . The total length of teeth appears also affected; shorter molars and, for females only, canines often accompany oligodontia[14]**.** The prevalence of taurodontism is significantly higher among individuals with oligodontia (about 34% compared to about 7%)[14,26,49] .

It has also been reported that persons with oligodontia show a tendency for delayed teeth formation[10,15]. Delayed formation occurs mostly in the early stages of tooth development[15] and is less marked in the mandible and more frequent in the teeth contralateral to missing teeth[10] .

An increased percentage (30%) of missing permanent teeth was also reported for patients with cleft palate, with a positive correlation between cleft size and hypodontia[50]. Genetic research has already shown that orofacial clefting and hypodontia share some genetic mechanisms[51], and Vieira[40] suggests that oral clefts may be the best models for isolated teeth agenesis.

Studies on the salivary function of patients with oligodontia reported reduced salivary secretion in association both with non-syndromic and syndromic (connected with ectodermal dysplasia) cases[6,32]. Although it was acknowledged that in ectodermal dysplasias the salivary glands are more likely to be involved, it was hypothesized that [non-syndromic] oligodontia in some cases may represent a microform of ectodermal dysplasia[6].

Craniofacial morphology and expected growth for hypodontia and oligodontia children

Whether or not genetical1y induced hypodontia is linked with abnormal craniofacial morphology and impaired growth, has been widely debated. There are a number of publications, which look into the craniofacial morphology of children with agenesis; some of them proceed to an evaluation of the expected growth of the lower face.

Cephalometric studies examining craniofacial morphology in cases of teeth agenesis, but not considering the number of missing teeth, present conf1icting results. In an early study of the craniofacial morphology of children with up to 6 missing teeth, Wisth et al.[52] reported reduced maxil1ary prognathism. Dermaut et al.[37] and Yukcel and Usem[53] however, reported normal cephalometric values and concluded that teeth agenesis has little effect on dentofacial structures.

The issue becomes clearer when focusing on severe hypodontia cases. Chung et al.[7] examined patients with 1-21 missing teeth and found normal mean values for the whole sample, but tendencies to class ΠΙ skeletal relationship and reduced maxil1ary –mandibular planes angle only when severe hypodontia patients were considered. This result confirmed the early report of Sarnas and Rune[11], who found a retrognathic maxilla and upright incisors in children with 4 or more missing teeth. Nodal et al.[12] found a significant association between the number of missing teeth and cephalometric variables: an increased number of missing teeth corresponded to reduced vertical jaw relation and a prognathic mandible. A threshold of about 12 missing teeth was identified, beyond which the cephalometric values were considerably affected. Using a similar design for children with 3 or more missing teeth, Ben-Bassat and Brin[54] observed a characteristic skeletodermal pattern, which was more evident in the cases with more than 10 teeth missing.

Literature evidence suggests, therefore, that cases of oligodontia are frequently accompanied by a retrognathic/underdeveloped maxil1a, a reduced vertical jaw relationship and a forward rotation of the mandible. The mechanisms underlying these clinical findings may involve an abnormal growth pattern, which means that the genetic factors that induce agenesis of the teeth may also affect the growth process of the lower face. Equal1y possible, however, is that the absence of teeth *per se* could block or deviate the growth pattern; in other words, the lack of dental growth may result in impaired craniofacial development.

To investigate the role of dental growth in the development of the jaws and craniofacial structures, Yamashita et al[55] examined cephalometrically an 8-year-old girl with permanent teeth anodontia and reported that the role of dental growth was significant in the development of maxillary and alveolar bone, but not in the development of the mandible. Roald et al[56] re-examined the sample of Wisth et al[51], 7 years after the first measurements and reported that the characteristic craniofacial morphology was not accompanied by abnormal growth pattern.

Ogaard and Krogstad[57] examined a sample of children with mild, moderate and severe hypodontia and compared them with a control group. Bondarets and McDonald[58] examined the facial skeleton of young individuals with oligodontia patients aged from 6 to 18 years and compared it with a matched control. They both reported that in persons with oligodontia the anterior lower face height was reduced and concluded that "the typical dentofacial structure in persons with advanced hypodontia may be due to dental and functional compensation rather than to a different growth pattern" and thus these cases are "dependent on the teeth for vertical growth". As will be discussed later, these observations emphasize the need for a timely prosthodontic intervention, in order to avert the anticipated consequences in vertical and transversal growth.

SYNDROMIC OLIGODONTIA

Oligodontia is frequently encountered as part of a syndrome. A syndrome is defined as a known complex group of defects or dysmorphic features occurring in one patient, due to genetic, chromosomal or environmental causes[59]. In some of the many syndromes that are referred to include teeth agenesis, such as X-linked ectodermal dysplasia and Rieger syndrome[22,59], the genetic background has been already studied. The researchers' efforts are again focusing on providing prevention or even treatment measures by interventions on malfunctioning genes[60]. In fact, encouraging results of correction of genetic impairment in transgenic mice have already appeared[61].

In syndromes, the clinical features may involve many different systems; however, as far as teeth are concerned, common characteristics with isolated oligodontia can often be found. For example, most syndromes exhibit a characteristic facial morphology, derived from specific skeletal deformities[58]. Those syndromes that include teeth agenesis commonly present a reduced lower face height and altered maxillofacial relationships; these findings are similar to those found in non-syndromic oligodontias and probably originate from the same pathogenetic mechanism, i.e. lack of teeth support[62].

The group of inherited disorders known as ectodermal dysplasias (ED), which seems to comprise the main bulk of syndromic oligodontias, can be used as a good example of the similarities with non-syndromic cases. Originating from failure of ectodermal development, they are characterized by an easily recognizable set of abnormalities involving skin, hair and nails[22,59,63], along with hypodontia, which, more often than in other syndromes, extends to oligodontia or even anodontia.

Many of the characteristics accompanying isolated oligodontia are met in ED-associated oligodontia: microdontia, mishapen teeth, and cleft palate are distinguishing symptoms in some Eds[59]. The maxillofacial relationships may correspond to the pattern described for non-syndromic oligodontias[62,64,65]. Impaired salivary secretion was recently found to be shared by both syndromic and non-syndromic cases[32]. It would seem reasonable to assume that the pathogenetic mechanisms of the two pathological entities, the idiopathic and the syndromic, are met at some, not yet identified point, and genetic research may provide a clarifying information in the future. In the mean time, identification of the phaenotypal similarities between non-syndromic and syndromic oligodontias is extremely helpful in providing a unified concept of the pathologic entity to be treated.

The prosthodontic insight: anticipating the need and complications of prosthodontic intervention

The necessity of a streamlined treatment plan to meet the needs of oligodontia cases, especially the syndromic ones, has been convincingly proposed[2,66]. The prosthodontic approach can be facilitated by codifying the cases into three groups according to their background and requirements:

Firstly, cases in which the lack of teeth is the only abnormal manifestation; secondly, cases where oligodontia is clearly part of a syndrome but comprises the main symptom requiring treatment, all other manifestations being under control. Most ectodermal dysplasias fall in this category, as by early childhood, symptoms like hypohidrosis are effectively faced. A third group could accommodate the more severely handicapped cases, in which oligodontia coexists with impairments such as mental retardation or general growth failure. These present more complicated clinical situations, varying according to the special characteristics of the syndrome.

From a prosthodontic aspect, the above grouping can be helpful for diagnosing the complexity of each case and the need for close collaboration with other specialists. The main differentiation lies in that in the first two groups, the prosthodontic restoration constitutes the backbone of the therapeutic design, all other interventions, such as prophylaxis measures being built around it. For cases falling in the third group, however, the prosthodontic rehabilitation may have to be modified and perhaps postponed in order to cooperate with the ongoing therapeutic measures. Non–syndromic cleft cases are a good example of this group: hypodontia frequently accompanies orofacial defects of varying severity[50,67]. Prosthodontic rehabilitation often has to follow the surgical intervention and even so, it is complicated because of the need to restore missing tissue along with missing teeth.

Incorporating the prosthodontic rehabilitation in a broad and diverse treatment plan is often challenging, as it requires a flexible protocol, balancing the cost and benefits at each treatment level, and meeting at the same time the specific needs of the patient. Resourceful treatment planning could take advantage of sophisticated strategies, such as orthodontic space closure, tooth movement to induce bone growth, or autotransplantation. Also, the potential of implant placement, even in the young, seems more and more promising. Overall, the means and methods of modern restorative dentistry (a detailed description of which falls beyond the scope of the present review) can help substantially to minimize or even negate the need of prosthodontic therapy, at least in its classical form.

In terms of clinical appearance, all cases of oligodontia seem to share some characteristics, besides the extensive edentulous areas. Convincing bibliographic evidence links non-syndromic oligodontia with reduced dimensions of remaining teeth and points to a common genetic background for these two characteristics. Therefore, unfavourable size and shape of abutment teeth can be anticipated in the course of prosthodontic restoration planning.

Saliva secretion may be affected in oligodontia. Many syndromes, ectodermal dysplasias the most common among them, are characterised of reduced salivary secretion; there seems to be a strong possibility that an analogous malfunction may also be present in non-syndromic oligodontias and has been left undiagnosed until the first prosthodontic examination. From a prosthodontic aspect, this is a critical prognostic sign, as the quantity and quality of saliva is crucial for the adaptation to removable prosthetic devices.

Concerning the morphology of the lower facial skeleton, there seems to be a common background in all oligodontia cases. In syndromic oligodontias this pattern is often complicated by extended facial and scull abnormalities, characteristic of each particular syndrome[58]. The underlying maxillofacial pattern which was studied in non-syndromic cases is basically characterised by a small maxilla, reduced height of the lower face and forward mandibular rotation[12,54]. Reports to date seem to indicate that the characteristic morphology is not genetically dictated, but created, maintained or even promoted during growth because of the lack of dental growth and subsequent functional compensation[55,58,65]. Prosthodontically improving maxillofacial relationships and restoring the functionality could therefore help the lower face to grow normally[12,66,68,69]. Cephalometric values can immediately be corrected by the placement of a prosthodontic device[70]. Though it is not feasible to prove these assumptions using strict research criteria, there are long-term case reports supporting its validity[2,64,71]. Accordingly, the standard prosthodontic principles and techniques serve well in restoring persons with oligodontia. The prosthodontic responsibility lies in that every treatment measure should be applied cautiously, so that the success of the final, post-growth result is not jeopardised; also the interventions must be adapted to the wishes and demands of the patient[2].

In conclusion, the search of the literature disclosed the complexity of the oligodontia issue, in that oligodontia is referred both as a symptom and as an individual trait. Bibliographic evidence has shown that the solitary congenital absence of more than 6 permanent teeth is a rare incident; nevertheless, its weak prevalence percentage is incremented by the syndromic cases. Whenever it is met, oligodontia presents a challenge to the prosthodontist, as it requires an early-scheduled, yet long-term oriented restoration plan. Therefore, knowledge of the characteristics that accompany, or are expected to accompany, oligodontia is essential. Not only does such knowledge help to properly design and effectively execute a long- term treatment plan, but it also contributes to a timely response to growth changes and promotes the communication with the other members of the multidisciplinary treatment team.

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