Research Highlights

A case of differing oligodontia in monozygotic twins

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Abstract

There are multiple aetiological factors for the developmental absence of teeth. Oligodontia is the congenital absence of six or more teeth and presents most commonly in association with a syndrome. Non-syndromic oligodontia may occur as a result of genetic factors, environmental factors or, most frequently, a combination of the two. This report presents a case of discordant, non-syndromic oligodontia in monozygotic male twins. The twins have a positive family history of developmentally absent teeth, suggesting a genetic cause for the condition, whilst the difference in clinical presentations between the twins may be explained by environmental and epigenetic factors. Awareness of the aetiology and recognition of a positive genealogy of hypodontia may play an important role in achieving early diagnosis and multidisciplinary treatment for patients with hypodontia. This may prevent the various functional and psychological difficulties associated with the condition.

Abbreviations

DPT – Dental panoramic tomograph
ISO – International Standards Organisation
MSX1 - Msh homeobox 1
PAX9 - Paired box 9

Introduction

Tooth agenesis refers to the failure of a tooth to form during development, and affects the general population at a prevalence rate of 1.6-6.9%. Females are more commonly affected than males.[1] Hypodontia refers to the agenesis of one to five teeth, excluding the third molars, whilst oligodontia is the congenital absence of six of more teeth and anodontia is the absence of all teeth.[2] The second tooth in each tooth series is most frequently missing, specifically the second premolars and upper lateral incisors. Moderate to severe levels of tooth agenesis, the absence of three or more teeth, can have significant aesthetic, psychological and functional consequences on patients of all ages.[3] Therefore, the primary aim when treating a patient with hypodontia is the development of a functional and aesthetic dentition. These goals may be met through a series of surgical, restorative, orthodontic, paediatric and prosthodontic work, which may take several years to complete.

Case report

This report defines a case of identical male twins with discordant oligodontia. Informed and valid consent was obtained from both twins to present and publish the findings in this case report.

At the age of 10, both twins were medically fit and well, and did not suffer from any systemic disorders. Intraoral examinations showed both twins were a class 2 division 2, a relationship where the maxillary jaw is positioned more anteriorly to the mandibular jaw than in an ideal occlusion, and where the upper anterior incisors were retroclined. They had severely displaced lower permanent lateral incisors, gingivitis and displayed signs of microdontia. Following referral for orthodontic treatment at the Great Ormond Street Hospital, UK, radiographs showed one twin to be missing teeth 15, 25, 34, 35, 44 and 45, whilst the other was missing teeth 15,13,12, 25, 35 and 45 **(Figure 1).** During the process of treatment to correct their displaced lower incisors with removable appliances, the twins were referred to the Hypodontia Clinic at a London teaching hospital where they received treatment from specialist dentists.

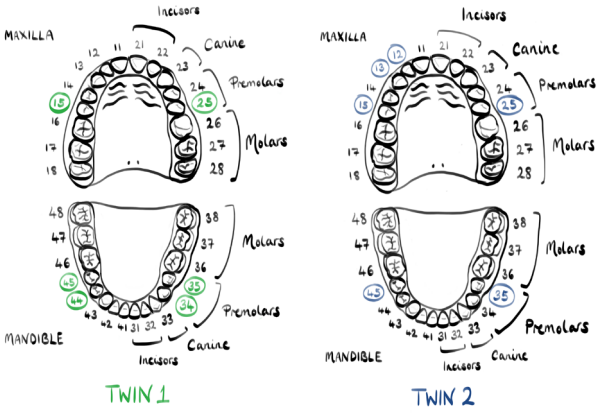

Figure 1. Labelled diagrammatic representation of the maxilla and mandibular arch, with teeth numbered according to the International Standards Organisation (ISO) numbering system. Green-coloured teeth are missing teeth from twin 1 and blue-coloured teeth are missing teeth from twin 2.

Further investigation, including a history from the patients' mother, revealed that the pregnancy and birth stages of the twins' lives were uneventful, but at 12 weeks old, one twin suffered from meningitis.

> *Investigation of the dental history from other family members revealed a positive history of moderate hypodontia in the twins' oldest brother.*

Following oral hygiene instruction and extensive orthodontic work to re-align, open and close spaces, at the age of 20, the twins had dental panoramic tomographs DPTs **(Figure 2)** and clinical photographs **(Figure 3)** taken prior to the commencement of the restorative aspect of their treatment.

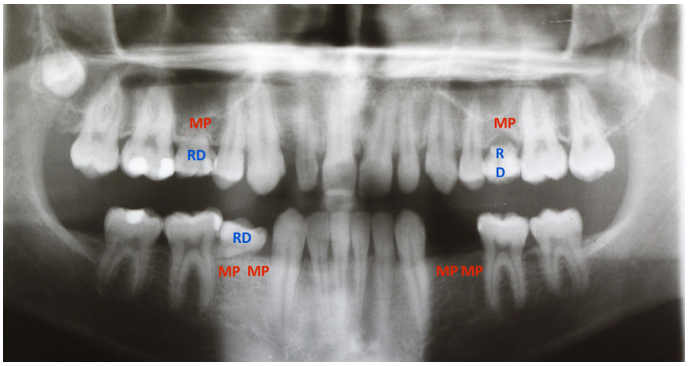

Figure 2. DPT of twin 1. The DPT of twin 1 shows multiple missing permanent (MP) teeth and three retained deciduous (RD) second molars.

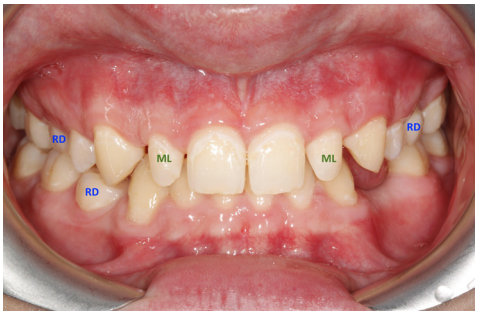

Figure 3. Coloured clinical photograph of twin 1. The photograph shows retained deciduous (RD) teeth, spacing and upper microdont lateral (ML) incisors. Photo by Mr Jagdip Kalsi, Consultant in Restorative Dentistry at Croydon University Hospital (Surrey, UK), who has given permission for use.

Discussion

Tooth agenesis occurs due to failure of initiation within the dental lamina during odontogenesis and can be classified as being syndromic or non-syndromic. Syndromes that hypodontia have been associated with include ectodermal dysplasia and Down syndrome, among others.[2] Non-syndromic hypodontia and oligodontia are rare presentations that have been previously reported in twin studies.[4]

Hereditary genetic factors, epigenetics and various environmental conditions, including drugs, trauma and exposure to chemotherapy, have been found to be predisposing factors for non-syndromic hypodontia.[5] Hereditary hypodontia may be a result of autosomal dominant or recessive genetic traits. Mutations of numerous genes, particularly those encoding the paired box 9 (PAX9) and Msh homeobox 1 (MSX1) transcription factors, which are involved in the regulation of craniofacial development, have been found to be associated with missing premolar and molar units. Some studies have linked PAX9 to altered tooth size, specifically microdontia, which presents commonly in patients with hypodontia.[6]

Identical twins develop from one zygote and, therefore, share an almost identical set of genes.[7] Regarding the phenotypic differences that are notable between identical twins, evaluation of twin studies suggest interactions between genes, common environmental variance and unique environmental variance to be the causes.[8] Epigenetics refers to the variation in phenotype that occurs as a result of gene expression modification as opposed to a change in DNA. This may occur in response to an environmental assault, hormones, inflammation or stochasticity.[9] A single mutation in developmental regulatory genes, or an environmental insult, possibly meningitis in twin 1 in this particular case, may affect the balance between activating and inhibiting genes involved in odontogenesis. The macroscopic consequences are alterations in tooth eruption patterns, tooth structure or tooth formation.[10]

Severe hypodontia can have a significant impact on quality of life.[11]

> *Primary aims when treating a patient with hypodontia are to improve aesthetics, maintain or increase vertical dimension and enhance dental function to improve phonetics and mastication.*

Treatment of patients with hypodontia can prove to be complicated, requiring a multidisciplinary approach including involvement from paediatric dentists, orthodontists, restorative specialists and implantologists. **Figure 4** and **Figure 5** show the stages in the placement of dental implants following restorative planning and the orthodontic movement of teeth. Delayed treatment referral has been found to have 'social and educational' implications for patients with hypodontia,[12] and studies have shown that older patients with a better understanding of their condition are more concerned about their appearance.[3] This has, in turn, had a greater impact on their psychological welfare than younger patients.

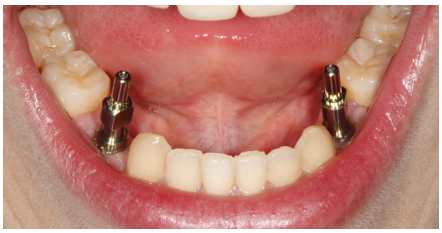

Figure 4. Coloured clinical photograph showing placement of dental implant and abutment as further treatment carried out by the multidisciplinary team. Photo by Mr Jagdip Kalsi (Croydon University Hospital, Surrey, UK), who has given permission for use.

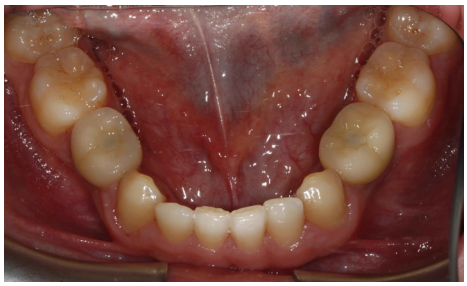

Figure 5. Coloured clinical photograph of twin 1 showing a final implant restoration. Implant restoration was used to replace teeth 34, 35, 44 and 45, with two large molar shaped implant crown prosthesis. Photo by Mr Jagdip Kalsi (Croydon University Hospital, Surrey, UK), who has given permission for use.

Conclusion and clinical significance

The genetic and environmental factors that play a role in the aetiology of non-syndromic oligodontia have been reported in several case reports.[13] Twins share most of their genetic variants which accounts for their similarity in presentation. Despite this, differences in medical conditions, levels of obesity, or dental anomalies present in many monozygotic twin studies. These studies show phenotypic differences between identical twins to be influenced by environmental and epigenetic factors.[9] Although rare, dentists should be aware of the various syndromic and non-syndromic causes for oligodontia. They should take into consideration the significance that a positive family history of hypodontia has on the chance of subsequent family members inheriting the condition, as early referral and successful treatment may result in fewer psychosocial and functional implications for the patient.

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