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Hypodontia in an adolescent with the HbSC genotype: a case report

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Summary. The haemoglobin SC (HbSC) genotype is a variant form of haemoglobin disorder that often presents less frequent clinical symptoms of sickle cell disease (SCD) than the homozygous state (HbSS). Various dental abnormalities, such as enamel hypoplasia, hypomineralization, midline diastema and delayed eruption of the teeth, have been associated with SCD. This report presents the case of an adolescent patient with HbSC who had hypodontia, transposition of a permanent canine, retained primary teeth, multiple carious lesions and poor oral hygiene complicated by sickle cell crisis. This report focuses on the need for preventive care in such patients to reduce functional and aesthetic complications.

Introduction

Hypodontia, an anomaly of odontogenesis, may occur in isolation, or as a component of genetic or chromosomal defects [1,2] such as Down syndrome or ectodermal dysplasia. The majority of individuals with hypodontia have other signs which could be linked to their condition [3]. The developmental absence of teeth presents varying clinical challenges in the management of affected patients [4]. These challenges can be greater in the case of the medically compromised.

The haemoglobin SC (HbSC) genotype is a variant form of haemoglobin disorder that often presents less frequent systemic symptoms of sickle cell disease (SCD) in affected individuals than the homozygous state (HbSS). It is less common than HbSS, and is mostly found in northern Ghana and Burkina Faso [5]. Among the Yoruba tribe of south-western Nigeria, HbSC has an incidence of 1·3% [6–8]. The clinical syndrome of SCD results from the susceptibility of the red blood cells in such patients to become sickle-shaped under conditions of hypoxia, and the consequent vaso-occlusive complications [9]. In addition to its systemic manifestations, dental abnormalities have been reported in individuals with SCD. These include enamel hypoplasia, hypomineralization, multiple interdental spacing, midline diastema and delayed eruption of teeth [10–13]. These findings have been attributed to systemic disturbances, and hyperplasia of the marrow in the maxilla and mandible, which may disturb amelogenesis and eruption of the teeth.

The objectives of this report are to document a case of hypodontia in an adolescent with the HbSC genotype and to highlight the importance of preventive care in such individuals.

Case report

A 15-year-old girl of the Yoruba tribe of south-western Nigeria known to have HbSC was referred to the paediatric dental outpatient clinic of the Lagos University Teaching Hospital, Lagos, Nigeria. She had a 3-day history of toothache and swelling of the left side of her face. It was the subject's first visit to the dentist. Two weeks before presentation, she had been hospitalized for sickle cell crisis. She was the youngest of eight siblings and had an older brother who had also been diagnosed with HbSC.

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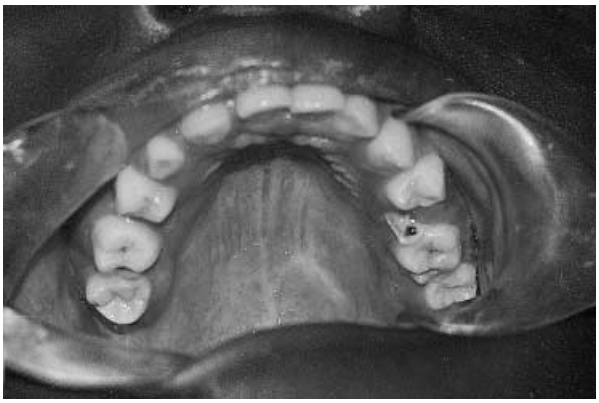

Fig. 1. Intraoral view of the subject's upper teeth showing a retained maxillary right primary canine (53), a right permanent canine (13) transposed to the position of the missing lateral incisor (12), and a retained, carious left second primary molar (65).

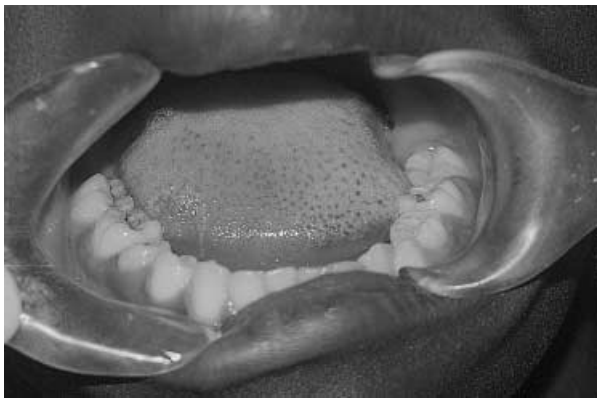

Fig. 2. Intraoral view of the subject's lower teeth showing a carious left first permanent molar that was also mobile with an associated dentoalveolar abscess (36), a retained left second primary molar (75), a retained and carious right second primary molar (85), and carious right first and second permanent molars (46, 47).

The following teeth were present on examination:

7 6 4 C 3 1	1 2 3 4 E 6 7
7 6 E 4 3 2 1	1 2 3 4 E 6 7

There was poor oral hygiene and the following carious teeth:

6	E 6
7 6 E	6

The maxillary right permanent canine was transposed to the position of the missing maxillary right lateral incisor (Fig. 1). The mandibular left first permanent molar was also mobile with an associated dentoalveolar abscess (Fig. 2). The maxillary right primary canine and mandibular left second primary molar were firm and caries-free. Periapical radiographs revealed that all second premolars were missing, and that there was an apical and furcation radiolucency of the mandibular left first permanent molar (Fig. 3).

A panoramic radiograph confirmed agenesis of the missing teeth, including the maxillary left third molar (Fig. 4):

5 2	5 8
5	5

On further inquiry and oral examination, the maxillary right permanent lateral incisor was also found to be absent in an older brother with HbSC. No other members of the family had any missing teeth.

Treatment plan consisted of:

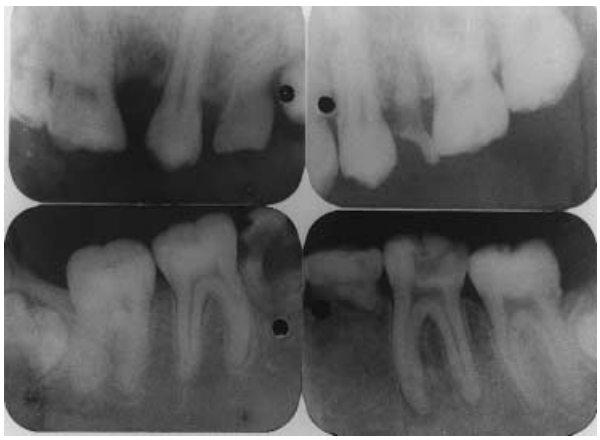

Fig. 3. Periapical radiographs showing a retained maxillary right primary canine, maxillary left and mandibular left and right second primary molars (53, 65; 75; 85), missing second premolars (15; 25; 35; 45), and a carious 36 with furcation involvement.

- extractions of the following mobile and carious teeth (with antibiotic prophylaxis):

	E
E	6

- a comprehensive oral hygiene and dietary programme, including parental counselling because of the subject's poor oral hygiene (the girl has been on regular professional oral prophylaxis since presentation);
- restoration of the carious teeth;
- an orthodontic consultation for realignment of the posterior teeth;

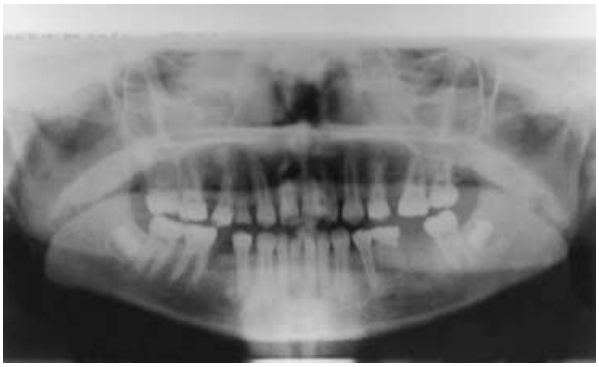

Fig. 4. Panoramic radiograph showing agenesis of all second premolars, the maxillary right permanent lateral incisor and the left third molar (12, 15; 25, 28; 35; 45), and transposition of the maxillary right permanent canine.

- a prosthodontic consultation for the construction of a conversion crown for the transposed tooth, and a removable prosthesis to fill the space left by the extracted mandibular left first permanent molar until exfoliation of the adjacent primary molar for permanent prosthesis;
- regular monitoring of the retained primary teeth for any signs of ankylosis; and
- management of the patient with the cooperation of her physician.

Discussion

Congenital absence of teeth may arise from disruption of the dental lamina, functional abnormalities of the dental epithelium and failure of induction of the underlying mesenchyme [14]. A significant genetic component has been reported in the aetiology of missing teeth, as with other developmental dental anomalies [15]. Hypodontia of this type in SCD has not been reported in the literature. The significantly variable clinical expression of the HbSC genotype has been previously reported [16], and is demonstrated in this case, where the older sibling had only one missing tooth.

The most commonly reported absent teeth are the third molars, maxillary lateral incisors and second premolars [17]. This is confirmed in this patient: a maxillary third molar, a lateral incisor and all second premolars were missing. Congenitally missing teeth may result in dental malpositioning, periodontal damage, and lack of development of maxillary and mandibular bone height; it also has significant aesthetic and functional consequences [18]. In this case, the missing lateral incisor caused the transposition of the canine. The missing maxillary right lateral incisor in an older brother with the HbSC genotype suggests a genetic link between HbSC and hypodontia, although this has not been previously reported in literature.

Space loss was further complicated by the extraction of a first permanent molar, and the spaces of the missing second premolars were already being taken up by the mesial drift of the first molars. This was made worse because appointments were rescheduled many times because of a recurrent illness caused by sickle cell crisis and noncompliance. The presence of a medical condition has been reported to act as a barrier to receiving dental care [19], which was the case in this patient. The dental infection from the carious mandibular first molar was suspected to be the cause of the crisis experienced by the patient before presentation at the dental clinic. Infections, dehydration, cold and exhaustion all cause sickling of the red blood cells and promote vaso-occlusion, which precipitates a crisis in affected patients [20–22]. Regular professional oral prophylaxis is being undertaken for the patient, together with an improved dietary programme, with her cooperation. This cannot be overemphasized since it will reduce the incidence of oral infection and new caries lesions. Good standards of oral health and motivation will have to be maintained throughout the subject's developing years [23].

Early diagnosis of the problem can reduce orthodontic complications and assist in treatment planning, with the aims of treatment being improvements in oral aesthetics, health and function, as well as the achievement of stable results. This involves a multidisciplinary approach, as in this case, involving the physician, paediatric dentist, hygienist, orthodontist and prosthodontist.

It is concluded that the adverse aesthetic and functional effects of hypodontia and other dental abnormalities can be minimized by early diagnosis and treatment planning, especially in the medically compromised, in whom chronic illness can interfere with the treatment plan.

What this case report adds

- The haemoglobin SC genotype is a haemoglobin disorder that presents with less frequent symptoms of sickle cell disease.
- This case presents with hypodontia involving all second premolars, one maxillary lateral incisor and one maxilliary third molar.

Why this case report is important to paediatric dentists

- Children with sickle cell disease exhibit enamel hypoplasia and hypomineralisation, interdental spacing, midline diastema and delayed eruption of teeth.
- Dental infections may contribute to sickle cell crisis. A programme for dental prophylaxis is of importance for this group of patients.

Résumé. Le génotype SC de l'hémoglobine (HbSC) est une variante des désordres de l'hémoglobine, souvent avec des symptômes cliniques moins fréquents de drépanocytose (SCD) que dans l'état homozygote (HbSS). Différentes anomalies dentaires telles que hypoplasie de l'émail, hypominéralisation, diastème médian et éruption retardée ont été associées au SC. Cet article rapporte le cas d'un adolescent HbSC avec agénésie, transposition de la canine permanente, rétention de dents temporaires, des caries multiples et une hygiène buccale défectueuse. Ce cas porte l'attention sur le besoin de prévention chez de tels patients afin de réduire les complications esthétiques et fonctionnelles.

Zusammenfassung. Hämoglobin SC (HbSC) ist eine Form der Hämoglobinopathie mit einer geringeren Ausprägung klinischer Symptome als der homozygote Status (HbSS) der Sichelzellanämie (SZA). Verschiedenartige Zahnanomalien wie Schmelzhypoplasie, Hypomineralisierung, Diastema mediale, oder verzögerter Zahndurchbruch wurden mit SZA assoziiert. Dieser Fallbericht stellt einen jugendlichen SZA-Patienten vor mit Hypodontie, Transposition eines bleibenden Eckzahns, retinierten Milchzähnen, zahlreichen kariösen Läsionen und insuffizienter Mundhygiene, der eine eine Sichelzellkrise duchmachte. Dieser Fallbericht zeigt die Erfordernis von präventiver Betreuung bei solchen Patienten, um funktionelle und ästhetische Folgen zu reduzieren.

Resumen. El genotipo de hemoglobina SC (HbSC) es una variante de las alteraciones de la hemoglobina que suele cursar con una menor frecuencia de síntomas clínicos de la enfermedad de células falciformes que en la forma homocigota (HbSS). Se han asociado con SCD varias anomalías dentales tales como hipoplasia del esmalte, hipomineralización, diastema de la línea media y retraso de la erupción de los dientes. Este informe muestra el caso de un paciente adolescente con HbSC que presenta hipodoncia, transposición de un canino permanente, dientes primarios retenidos, lesiones cariosas múltiples e higiene bucal pobre, complicado con crisis de células falciformes. Este caso se centra en la necesidad de un cuidado preventivo en estos pacientes para reducir las complicaciones funcionales y estéticas.

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