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Dental findings of a child with Wolf–Hirschhorn syndrome

N. J. JOHNSTON & D. L. FRANKLIN
Department of Child Dental Health, Bristol Dental Hospital, Lower Maudlin Street, Bristol, BS1 2LY

Summary. The case presented is that of a 5-year-old female with Wolf–Hirschhorn syndrome. Dental findings were severe hypodontia, late dental development, taurodontism of the primary molars, microdontia, and spacing. Hypodontia has previously been reported and therefore this case adds to the evidence that hypodontia may be a common feature of this syndrome. It also suggests that other dental anomalies could occur in children with Wolf–Hirschhorn syndrome.

Introduction

In 1965, Wolf *et al*. [1] and Hirschhorn *et al*. [2] first reported a syndrome associated with the deletion of the short arm of chromosome 4. These cases were simultaneously published and resulted in the identification of Wolf–Hirschhorn syndrome. The syndrome was initially associated with 'cri du chat' syndrome. Alhough the syndrome shows similarity to cri du chat, there are important differences in a number of phenotypic characteristic such as the lack of midline cranial defect and a cat-like cry [3]. Studies revealed that cri du chat is associated with the deletion of chromosome 5, whereas in Wolf–Hirschhorn syndrome the deletions are on chromosome 4. In Wolf–Hirschhorn syndrome the amount of deleted material varies, from a small break to 50% of the short arm of chromosome 4p.

The most common characteristics of this syndrome are growth retardation and learning difficulties, microcephaly, hypotonia, muscle hypotrophy, seizures, feeding difficulties, and craniofacial features. The most striking feature is represented by the 'Greek warrior helmet appearance' of the nose, which is the broad bridge of nose continuing to the forehead [4]. Other craniofacial features are short philtrum, micrognathia, hypertelorism, high-arched eyebrows, ear anomalies, and bifid uvula. Eye, heart, or hearing defects may occur and 25–50% patients also have cleft lip and/or palate [4].

Correspondence: D. Franklin, Department of Child Dental Health, Bristol Dental Hospital, Lower Maudlin Street, Bristol BS12LY, UK. E-mail: deborah.franklin@ubht.swest.nhs.uk

Children with this syndrome show marked growth deficiency and slow weight gain. This may result from many features of the syndrome, e.g., heart defect. It may also be related to feeding factors such as oral clefts with related difficulty in sucking, poorly coordinated swallow with consequent aspirations, and gastroesophageal reflux [4].

Heart defects are a common finding. The most common defects are atrial septal defect (ASD), pulmonary stenosis, and ventricular septal defect (VSD). They are not usually complex and can be surgically repaired if necessary [4].

The severity of affectation varies. Some children can walk and verbalize well, whereas others require full-time care. This syndrome occurs in about 1 in 50,000 births [4,5] and affects females more than males; there is no ethnic predilection.

It is a rare syndrome and dental literature is limited. Burgersdijk and Tan [6] were the first to describe the dentition of a patient with Wolf–Hirschhorn syndrome. They noted fusion of the mandibular central and lateral primary incisors and severe hypodontia of the permanent dentition. There are only three other reports regarding the dentition of children with Wolf–Hirschhorn syndrome. Breen [5] reported on a case with this syndrome in which he described severe hypodontia and was also the first and only case so far to report taurodontism of the primary molar teeth. A larger research report was published by Nieminen *et al*. [7] in 2003 and this showed hypodontia in five of seven patients studied with Wolf–Hirschhorn syndrome, and associated it with the deletion of the *MSX1* gene found in those patients. They also reported hypoplasia and discoloration of the permanent dentition. Kozma *et al*. [8]

in 1999 reported hypodontia and cone-shaped teeth as part of this syndrome.

Case report

The patient is a 5-year-old white female with multiple, classical features of this syndrome and has been seen for regular examinations and dental advice since she was 1 year old. She presented with severe learning difficulties, cardiac lesions (ASD and pulmonary stenosis), seizures, poor sight (severely long-sighted), and other craniofacial features including cleft palate and uvula.

At 2 years old, the cleft palate and uvula were successfully surgically repaired. This was later than usual because of medical needs and social issues prior to this. This was followed by cardiac surgery at 3 years of age, when the ASD was closed and the pulmonary stenosis corrected.

She has progressed well from birth. She has learning difficulties and her development level is not that of a child aged 5 years; however she walks and is verbalizing well. Currently she attends mainstream nursery. There are no hearing problems and for her long sightedness she wears strong spectacles but appears to see reasonably well. Feeding has not been easy but she has gained weight. Her diet consists of only soft food as she will not eat anything 'lumpy'.

She has a class 3 facial skeletal pattern, a large forehead with a high hairline and a short upper lip (Fig. 1). She has the Greek warrior helmet appearance of the nose with associated hypertelorism (Fig. 2). The appearance of the nose is not as profound as in other children with this syndrome and is disguised when she wears her spectacles.

Dentally the primary teeth erupted late. The mandibular primary central incisors erupted at about 18 months and these were spaced and slightly misshapen. The other primary teeth (excluding the maxillary primary lateral incisors) erupted over the next 18 months. The final primary dentition is spaced and all the teeth are microdont (Fig. 3a,b). The maxillary left primary incisor was lost through trauma. She also has a prominent mandibular and maxillary midline frenum between the primary central incisors.

The primary dentition at age five is

EDC A	CDE
EDCBA	ABCDE

(55, 54, 53, 51, 63, 64, 65, 75, 74, 73, 72, 71, 81, 82, 83, 84, 85)

Fig. 1. Facial profile illustrating maxillary retrognathia, high hairline, and a short upper lip and philtrum.

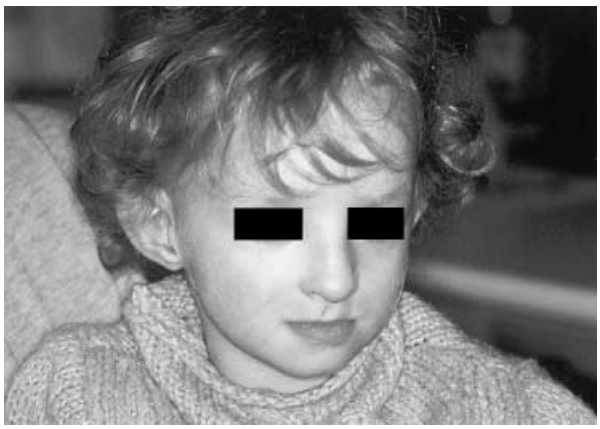

Fig. 2. Facial appearance showing 'Greek warrior helmet' appearance of the nose.

Patient cooperation was only sufficient for left and right lateral oblique radiographs and these show severe hypodontia (Fig. 4a,b). There is absence of both upper lateral primary incisors at present and the only permanent teeth seen developing are in the more anterior part of the mouth. There are no permanent molar teeth present. The radiographs also reveal taurodontism in the posterior primary dentition.

The oral hygiene is excellent. Tooth brushing it not easy but the mother manages to brush the patient's teeth once or twice a day with an electric toothbrush and children's toothpaste. The patient

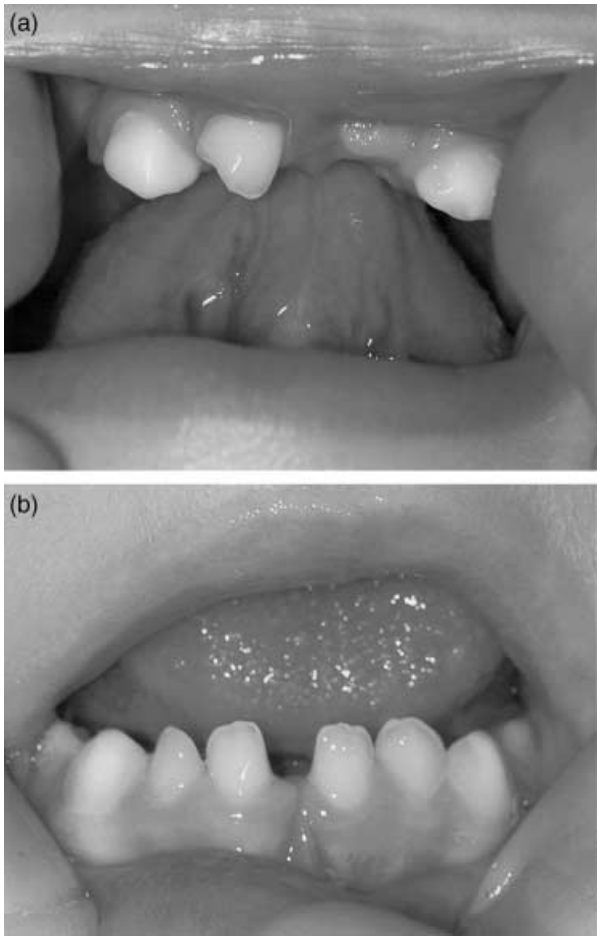

Fig. 3. (a) Maxillary dentition showing microdontia, spacing, and prominent frenum. (b) Mandibular dentition showing microdontia, spacing, and prominent frenum.

was prescribed fluoride drops at 4 years old and these continue to be taken regularly. At present she is caries free.

Discussion

The previously mentioned case shows a young child with a class 3 skeletal pattern, delayed dental development, severe hypodontia, taurodontism of the primary molars, spacing, and microdontia. Hypodontia has previously been reported in children with Wolf–Hirschhorn syndrome [5–8], and this case adds to the evidence that hypodontia is likely to be a common feature of this syndrome. The patient follows the same pattern of missing permanent teeth previously reported by Nieminen *et al*. [7] who noted that the more posterior teeth are more commonly absent.

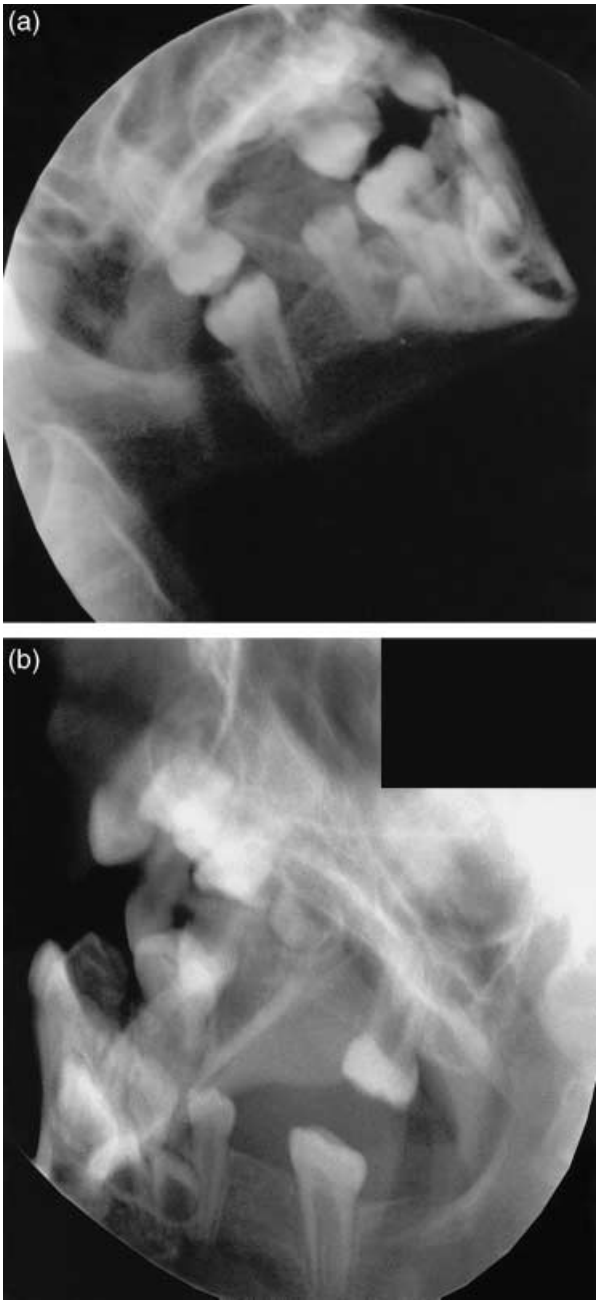

Fig. 4. (a) Right lateral oblique radiograph showing severe hypodontia and taurodontism of the primary molars. (b) Left lateral oblique radiograph showing severe hypodontia and taurodontism of the primary molars.

It is important that hypodontia is recognized as a potentially important feature of this syndrome and that preventative methods are put in place as soon as the dentition first erupts. These patients usually require a lot of medical care, especially in the first few years of life but it is essential that their oral

health is not overlooked. Regular dental examinations and advice are necessary to ensure that the dentition remains healthy. This child had regular examinations every six months, oral hygiene advice and fluoride drops that have maintained a caries-free dentition. In this case the teeth that have erupted are well formed and healthy; however, the patient is still very young and the full effect of the syndrome of the permanent dentition is not known. Because of the lack of permanent teeth it is desirable to maintain the primary dentition for as long as possible. This is even more important as improving function and aesthetics in the future will be challenging because of difficulties in cooperation. A multidisciplinary approach will be required.

This case reveals taurodontism of the primary molar teeth that has only been reported once before by Breen [5]. This indicates that taurodontism may be an additional dental characteristic of the syndrome.

It has been thought that children with Wolf–Hirschhorn syndrome have severe learning and developmental difficulties, with minimal speech, and difficulties or inability to walk. This case shows a happy, active child who walks well and has begun to communicate effectively. It has to be concluded that not all these children are so severely affected by this syndrome. This is supported by Battalgia *et al*. [4] who found severe learning difficulties in only 66·6% patients, whereas it was moderate in 25% and mild in 8·3%.

What this paper adds

- This paper adds to the evidence that hypodontia may be a consistent feature of this syndrome.
- It also indicates other dental anomalies that may be part of the syndrome.
- The paper will increase awareness and knowledge about Wolf-Hirschhorn syndrome.
- Discussions about prevention, treatment and future care that may help with this syndrome are noted.

Why this paper is important for paediatric dentists

- This paper will be of value to paediatric dentists as Wolf-hirschhorn syndrome is a rare syndrome with little written on dental findings.

Conclusion

It can be concluded that an important dental characteristic of a child with Wolf–Hirschhorn syndrome is likely to be hypodontia. Others probably include taurodontism of the primary molars, microdontia, spacing, and late dental development. Further reports will determine if these other characteristics are a consistent finding.

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