ARTICLE

Dental findings in children with West syndrome

Neusa Barros Dantas-Neta, DDS;[1] Carlos Henriquede Carvalho e Souza, DDS;[1] Suyá Moura Mendes Alencar, DDS;[1] Raimundo Rosendo Prado Júnior, PhD;[2]* Regina Ferraz Mendes, PhD[2]

[1]Student of the Dental Postgraduation Program at Federal University of Piauí (UFPI) – Teresina – PI, Brazil; [2]Associate Professor of the Dental Postgraduation Program at Federal University of Piauí (UFPI) – Teresina – PI, Brazil.
*Corresponding author e-mail: rosendo_prado@ig.com.br

Spec Care Dentist XX(X): 1-4, 2014

ABSTRACT

Introduction: West syndrome (WS) is a rare age-related syndrome of epilepsy. The oral manifestations of WS are still unknown.
Objective: To assess the oral health status and oral findings of a sample of WS children being treated at a specialized referral center.
Material and Methods: The dental record forms of 528 children were searched at this center. Eight of the children had been diagnosed with WS.
Results: The ages of those with WS ranged from 6 months to 13 years, and 62% of them were male. All of them were taking antiepileptic medication. Twenty-five percent of them had tongue interposition between the dental arches and a deep palate. The mean DMFT was .25 and dmft was 1.12.
Conclusion: Patients with WS seem to have low caries experience, can have a deep palate, and may have their tongue positioned between the arches or on the incisive papilla.

KEY WORDS: infantile spasms, epilepsy, special patients, West syndrome

Introduction

In 1841, the physician Dr. William James West published the first report of West syndrome (WS) in a letter to the editor of *The Lancet*. In this letter, the doctor described the epilepsy and its consequences, such as developmental delay and intellectual disability.[1,2] WS is a rare childhood syndrome of epilepsy related to age and includes the triad: epileptic spasms, EEG with hypsarrhythmia, and arrest or regression of psychomotor development.[3–5]

WS most frequently manifests itself from 3 to 8 months of age, most frequently in the fifth month.[6] The syndrome is responsible for 1.4% of the causes of childhood epilepsies, is more frequent in males, and can be associated with other systemic conditions or syndromes.[6]

According to Silveira,[7] the etiology of WS is heterogeneous, and can be classified as cryptogenetic or idiopathic and as symptomatic or secondary. When idiopathic, the child's development is normal until the onset of symptoms and, from then on, it deteriorates. When symptomatic, there is a distinct disease process and a delay in development precedes the spasms.[8–10]

The determination of the etiological factor is important if therapy is to be effective. When treated, children with infantile spasms can have an improvement in their quality of life.[2,8] Specific information about prognosis and treatment response is directly related to the etiology of WS. In spite of treatment, however, patients with WS usually have an unfavorable prognosis. Comprehensive health care from a multidisciplinary team is essential, and this team should include a dentist.

There are hardly any studies investigating aspects related to dentistry in WS patients; therefore it is not known whether oral disorders are present. Thus, the aim of this study was to analyze the oral conditions of a sample of patients with WS who were being treated at a referral center in order to characterize and describe the oral findings.

Methods

Patients with WS were searched for among 528 dental records at the Integrated Center for Special Education (CIES), a public institution specialized in the care of patients with special needs in Piauí State, Brazil.[11,12] Eight medical records with a diagnosis of WS were found and the variables collected were: socioeconomic data, family history, review of pregnancy complications, and the analysis of oral health conditions such as oral hygiene,

© 2014 Special Care Dentistry Association and Wiley Periodicals, Inc.
DOI: 10.1111/scd.12068

Table 1. Socioeconomic characteristics of families of children with West syndrome in Teresina, 2012.

	Socioeconomic status		
		Female	Male
Child age group (years)	<1 Between 1 and 6 Between 7 and 13	0 2 1	1 0 4
Birth	At term Preterm	3 0	5 0
Mothers schooling (years)	7 to 9 ≥10	1 2	2 3
Family income	<2MS ≥2MS	2 1	2 3
Government aid	Yes No	1 2	2 3

nonnutritive oral habits, dmft, and DMFT.

This study was carried out according to the norms and regulations of research on human subjects, contained in Resolution No. 196/96 of the National Health Council and the Declaration of Helsinki II (2000). The study was approved by the Ethics Committee of the Federal University of Piauí (protocol 0151.0.045.000-08). Each participant, represented by their legal guardian, was clear about the objectives and signed an informed consent form.

The information collected was stored in SPSS (Statistical Package for Social Sciences) version 18.0 (SPSS Inc., Chicago, IL, USA). A descriptive analysis of the data was carried out and the variables were expressed as absolute and relative frequencies and means.

Results

Eight patients (1.5%) had been diagnosed with WS among those whose records were examined. Their age ranged from 6 months to 13 years, 5 were male and 3 were female (Table 1). This study found that WS was more prevalent in boys than girls: a ratio of 3/5. Also, there were no previous reports of the syndrome in the family. Three children had other syndromes associated with WS (Down syndrome, autism, and cerebral palsy). All of the children were born at term and 5 (62.5%) of the children had complications at birth, such as fetal anoxia, cyanosis, convulsions, jaundice, or required a period of time in an incubator after birth. Most mothers (7–87.7%) had more than 6 prenatal consultations.

The children in the study had a low socioeconomic status, with mean household income of 1.81 times the minimum wage. However, most mothers had more than 10 years of schooling (Table 1). Among the family characteristics, 50% of mothers reported a history of intellectual disability in the family, but there were no cases of WS.

The health care of these children included systemic daily use of antiepileptic medication, the most common of which was valproic acid (Figure 1).

All of the mothers reported cleaning their children's teeth using a toothbrush and toothpaste. The majority of the mothers stated that they insisted on brushing when the child was uncooperative. Most children (87.5%) were reported as having difficulty in chewing; half of the children had mixed (mouth and nose) breathing.

The most striking oral characteristics found in these children were deep palate, and tongue interposition between arches or on the palatine papilla, observed in 25% of children (Table 2).

The mean DMFT was .25 and dmft was 1.12. The sample had nine decayed teeth, one missing, and one filled tooth. Both the tooth extraction and filling were carried out at CIES and the decayed teeth were to be restored at the same institution.

Discussion

The oral health status of patients with special needs has hardly ever been studied and reliable data are scarce in Brazil.[11,12] This is also the case for WS patients; no studies on the oral health of WS patients were found in the literature.

Many patients with special needs find it difficult to maintain good oral health and to access dental care because of their disability or medical condition.[12,13] However, patients treated at CIES receive multidisciplinary and integrated health care, which includes dental care that focuses on promoting oral health.

CIES is an institution specializing in the care of children with special needs and has been operating since 2006. It develops nonprofit work with children and adolescents with intellectual disability (with or without other disabilities), aged 0–14 years. Since 2007, an extension project of the Federal University of Piauí has been developed at CIES with the participation of undergraduate and postgraduate dental students. The students gain experience in the practice of dental care for special needs patients under the supervision of a university professor and one dentist. At the time of this study, there were more than 400 children being treated.[12]

CIES aims to facilitate the social inclusion of people with special needs, providing them with education and multidisciplinary healthcare. The fields of health care that it focuses on are: psychology, speech therapy, dentistry, educational psychology, physiotherapy, nursing, and social work.[14]

As in other studies,[15–17] there was a predominance of males in our sample; however, this might have been due to the limited number of individuals assessed, which in turn may have been to the low prevalence of the syndrome of its

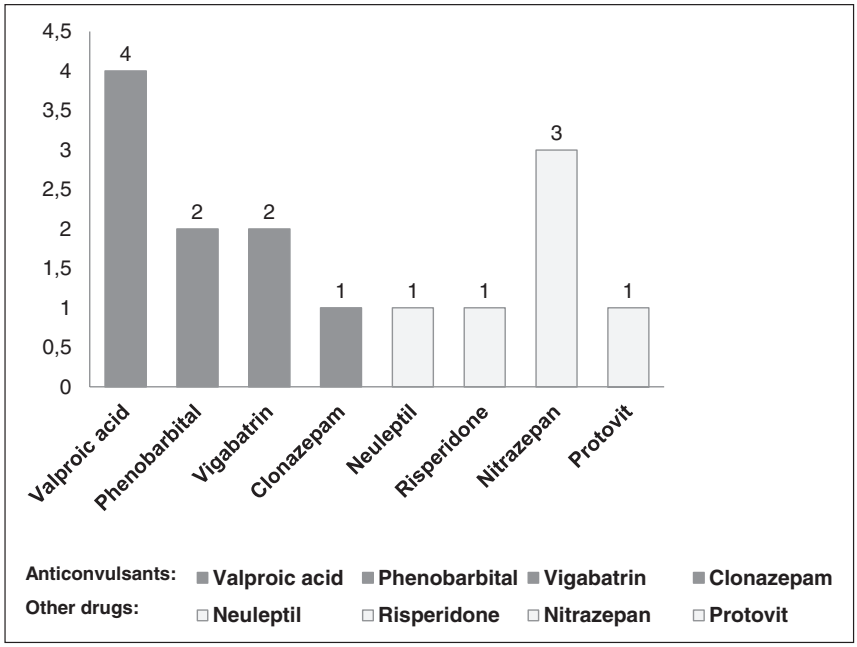

Figure 1. Medications used by children with West syndrome in Teresina, 2012.

Table 2. Oral characteristics of patients with West syndrome.

Tongue position	N	%
On the palatine papilla	1	12.5
Interposition between arches	2	25.0
Mouth floor	5	62.5
Palate		
Deep	2	25.0
Normal	6	75.0
DMF-T		
0	6	75.0
1	2	25.0
dmf-t		
0	5	62.5
1	1	12.5
4	2	25.0

subnotification and or unawareness of family members of what the disease is.

The etiologic factors that may lead to symptomatic WS can be classified as: pre-, peri- or postnatal.[1,8] Postnatal factors that might have led to the development of WS were not observed in the study. However, the mothers reported factors such as chromosomal (Down syndrome) and perinatal abnormalities (fetal anoxia and neonatal jaundice).

In this study, one child (16.7%) had WS associated with autism. The prevalence of epilepsy in children with autism spectrum disorder varies between 5% and 39%, with a bimodal distribution of the onset of seizures; the first peak is before the age of five, and the second, after 10 years of age. Despite being a very heterogeneous group, it is estimated that one-third of these cases will present at least one epileptic crisis before adolescence.[4]

Pharmaceutical treatment of epilepsy aims to reduce mortality and morbidity and improve quality of life. This treatment begins earlier in patients with WS and decreases the high risk of cognitive impairment.[18]

Patients who use medication require rigorous care from health professionals. Dentists must have knowledge of the medications used due to the possible orofacial and systemic reactions. They must also be aware of adverse interactions with other drugs used in dental procedures, such as anesthetics. Good communication between the dentist and the physician is important to ensure safe treatment planning.

The following antiepileptic drugs were used by patients in this study: valproic acid, phenobarbital, vigabatrin, and clonazepam. Valproic acid interacts with benzodiazepines and with nervous system depressants by strengthening the effects of both. Therefore, it should be used with caution. Among the adverse effects of phenobarbital are respiratory depression and reduction of heart rate. Thus, the interaction of phenobarbital with benzodiazepines can increase central nervous system depression. Clonazepam and vigabatrin are not known to interact with drugs used in dentistry.[18]

There are reports that the use of phenobarbital and valproic acid can induce gingival hyperplasia.[16,19–21] Despite being an extremely rare condition[22] three factors can be important in the manifestation of gingival hyperplasia: the use of a variation of the drug, plaque-induced inflammatory changes, and genetic factors.[21] The patients evaluated in this study did not present any gingival hyperplasia, maybe due to the oral care that had been provided by professionals and students at CIES. According to Loureiro *et al.*,[23] meticulous oral hygiene at the beginning of the administration of medication may decrease the incidence of gingival hyperplasia.

To date, no oral changes that are specific to WS patients have been reported. However, some case reports have identified the following oral findings as being common: mouth breathing, deep palate, gingival hyperplasia, severe gingivitis, anterior open bite, alteration in the chronology of tooth eruption, widespread tooth wear, fissured tongue, and lingual interposition between arches.[1,16] Furthermore, the presence of multiple white spot carious lesions, poor oral hygiene, and a diet high in carbohydrate have also been reported.[16]

In this study, deep palate and tongue interposition were observed in two

children. In addition, there was a low caries experience in the primary and permanent dentition, up until the time of evaluation. This may be due to the low frequency of sugar intake and the fact that the mothers routinely cleaned their children's teeth.

The main limitations for the dental treatment of these patients are similar to any patient with impaired psychomotor and social development, such as impaired communication, difficulties in cooperation with the treatment, and fear of strangers. Therefore, health promotion should be emphasized in the treatment of patients with WS. Oral health status can also be improved by stressing the importance of the family's involvement in daily oral hygiene and the rational consumption of fermentable carbohydrates.[16]

One of the limitations of this study is its small sample, which may not represent the reality for all patients. Nevertheless, this study is important because of the scarcity of studies on WS and dentistry. Studies that assess the quality of care for WS patients and the impact on the lives of individuals and their families should also be carried out.

Conclusions

The analyzed patients with WS showed deep palate, tongue interposition between the arches and tongue disposition on the incisive papilla. It is suggested that a study be conducted with a larger sample to represent a population with this problem.

References

1. Aguiar SMF, Torres CP, Borsatto MC. West syndrome. *J Bras Odontopediatr Odontol Bebê* 2003;6(30):123-6.
2. Shields WD. Infantile spasms: little seizures, BIG consequences. *Epilepsy Curr* 2006;6(3):63-9.
3. Lux AL, Osborne JP. A proposal for case definitions and outcome measures in studies of infantile spasms and West syndrome: consensus statement of the West Delphi group. *Epilepsia* 2004;45(11):1416-28.
4. Souza VMA, Pereira AM, Palmini A, *et al*. West Syndrome, autism and epilepsy: resolution of the epilepsy and recover of autism after surgery. *J Epilepsy Clinneurophysiol* 2008;14(1):33-7.
5. Jansen K, Vandeput S, Huffel SV, Lagae L. Cardiac autonomic dysfunction in West syndrome. *Epilepsy Res* 2012;102(3):167-72.
6. Albuquerque M, Muszkat M, Vergani MIC, Campos CJR. West syndrome: report of 25 cases. *Rev Paul Pediatra* 1990;8(29):76-8.
7. Silveira PR. West Syndrome. *Pediatr Modern* 1997;33(10):793-6.
8. Matta APC, Chiacchio SVB, Leyser M. Possible etiologies of West syndrome: evaluation of 95 patients. *Arq Neuropsiquiatr* 2007;65(3a):659-62.
9. Osborne JP, Lux AL, Edwards SW, *et al*. The underlying etiology of infantile spasms (West syndrome): information from the United Kingdom Infantile Spasms Study (UKISS) on contemporary causes and their classification. *Epilepsia* 2010;51(10):2168-74.
10. Wong V. West syndrome – the University of Hong Kong experience (1970–2000). *Brain Dev* 2001;23:609-15.
11. de Carvalho RB, Mendes RF, Prado Jr RR, Moita Neto JM. Oral health and oral motor function in children with cerebral palsy. *Spec Care Dentist* 2011;31(2):58-62.
12. Oliveira JS, Prado Jr RR, Lima KRS, Amaral HO, Moita Neto JM. Intellectual disability and impact on oral health: a paired study. *Spec Care Dentist* 2013;33(6):262-8.
13. Castro AM, Marchesoti MGN, Oliveira FS, Novaes MSP. Analysis of dental treatment provided under general anesthesia in patients with special needs. *Rev Odontol UNESP* 2010;39(3):137-42.
14. Tomita NE, Fagote BR. Programa educativo em saúde para pacientes especiais. *Odontol e Soc* 1999;1(1/2):45-50.
15. Pereira Filho AL, Malucelli DAB, Ferreira LLA, Gonçalez-D'Ottaviano F, Silveira JAM. Avaliação dos achados ao exame dos potenciais evocados do tronco cerebral em indivíduos com síndrome de West. Rev. Bras. *Otorrinolaringol* 2004;70(1):90-3.
16. Regis RR, Rocha CT, Torres CP, Queiroz SE, de Queiroz AM. Oral findings and dental treatment in a child with West syndrome. *Spec Care Dentist* 2009;29(6):259-63.
17. Scantlebury MH, Galanopoulou AS, Chudomelova L, Raffo E, Betancourth D, Moshé SL. A model of symptomatic infantile spasms syndrome. *Neurobiol Dis* 2010;7(3):604-12.
18. Sander JW. The use of antiepileptic drugs—principles and practice. *Epilepsia* 2004; 45(Suppl 6):28-34.
19. Anderson HH, Rapley JW, Williams DR. Gingival overgrowth with valproic acid: a case report. *ASDC J Dent Child* 1997;64(4):294-7.
20. Lafzi A, Farahani RM, Shoja MA. Phenobarbital-induced Gingival Hyperplasia. *J Pract Dent Contemp* 2007;8(6):50-6.
21. Lin K, Guilhoto LMFF, Yacubian EMT. Drug-induced gingival enlargement – Part II. Antiepileptic drugs: not only phenytoin is involved. *J Epilepsyclin Neurophysiol* 2007;13(2):83-8.
22. Sinha S, Kamath V, Arunodaya GR, Taly AB. Phenobarbitone induced gingival hyperplasia. *J Neurol Neurosurg Psychiatry* 2002;73(5):601.
23. Loureiro CCS, Adde CA, Perez FEG, Penha SS. Efeitos adversos de medicamentos tópicos e sistêmicos na mucosa bucal. *Rev Bras Otorrinolaringol* 2004;70(1): 106-11.