INTRODUCTION.

Neural tube defects (NTDs) are birth defects that occur in the early stages of fetal development. They involve the primary structure that forms the brain and the spinal cord (encephalocele) and cause deficiencies that affect the spinal structure (spina bifida). NTDs have a multifactorial etiology including environmental and genetic factors. Genetic predisposition is caused by an autosomal recessive pattern, having a high risk of recurrence. Other causes include exposure to teratogens (drugs and environmental pollution), gestational diabetes, hyperthermia and obesity.1-3,5-7

Spina bifida (SB) is a congenital malformation of the spinal cord associated with several vertebral abnormalities caused by incomplete neural tube closure. It is one of the most common malformations of the central nervous system, with an incidence of 1:800 live births.8 In Venezuela, prevalence is 1.8 per 1000 live births. There are two known clinical types: aperta and occulta. In aperta, the defect is clearly identified at birth, in occulta, the abnormality is not apparent and has no clinical significance at birth.2,3,5,6,9 There are two types of SB aperta depending on their onset time: meningocele (MC), onset occurring after the fourth week of pregnancy, and myelomeningocele (MMC), with an earlier onset time.2,3,6,9

Patients with SB may have cognitive deficits, lung function abnormalities, scoliosis, deformed hips, feet and legs, short stature and bladder dysfunction.10,11 Sensorimotor alterations are variable, but 90% of patients must use wheelchairs.1,6,9 The 70-90% of newborns with MMC suffer from hydrocephalus and eventually develop cognitive deficits. Hydrocephalus is usually treated with...
an implanted valve, and antibiotic prophylaxis should be indicated in case of high-risk dental procedures.\textsuperscript{1,2,6}

Children with SB are more prone to dental caries and periodontal disease because of multiple factors such as poor diet, lack of oral hygiene and prolonged use of oral medications that contain sugar.\textsuperscript{1,2,6} Despite the special care these patients need, there are no recent reports describing their oral health status. Therefore, the aim of this paper is to report the oral health status of a sample of Venezuelan patients with SB.

**MATERIALS AND METHODS.**

An observational cross-sectional study was performed in 30 Venezuelan patients aged 1 to 16 years diagnosed with SB. Patients were enrolled from the Venezuelan Association of Spina Bifida located in the city of Caracas. No exclusion criteria were considered.

All patients were examined after their legal guardian signed an informed consent accepting the dental evaluation. Ethical approval for the study was obtained from the Bioethics Committee of the School of Dentistry at Universidad Santa Maria. Sex distribution was 12 male patients (40\%) and 18 female (60\%).

Once the sample was selected, guardians were questioned in relation to patients’ medical history to confirm the presence of hydrocephalus or a valve for draining cerebrospinal fluid. In addition, they were asked about the presence of oral parafunctions and food intolerances.

All patients were examined by a single and previously calibrated researcher who evaluated the presence of caries, gingivitis and calculus using artificial light and a clinical mirror. Caries history was assessed by dmft and DMFT indices, using criteria proposed by the World Health Organization. The presence or absence of gingivitis was assessed by the Löe and Silness index. The Simplified Oral Hygiene Index and the patient’s occlusion type according to Angle’s classification were also determined.

Statistical analysis was performed using SPSS version 20.0 (IBM, USA).

**RESULTS.**

Mean age of the whole sample was of $4.67 \pm 3.78$ years. The 76.7\% had associated hydrocephalus, and 60\% had an implanted valve to drain excess cerebrospinal fluid.

Three patients had food intolerances; all three were lactose-intolerant, one gluten-intolerant, and one intolerant to food dyes and colorings.

The 46.7\% of the patients had a history of caries: 22\% in the 1-4 year group, 71.4\% in the 5-7 year group, and 100\% in the 8-16 year group. The dmft and DMFT indices were 1.55 and 3.50, respectively. A 46.7\% of the patients had gingivitis, 30\% had dental calculus, with an OHI-S of 2. The 83\% had Angle Class II and 17\%, Angle Class I. A 40\% had parafunctional habits such as digital suction, use of pacifiers and onicophagia. The 70\% had deep palate.

**DISCUSSION.**

The 76.7\% of the patients had hydrocephalus, commonly treated with the use of valves, as seen in the present study. Kabani \textit{et al.}\textsuperscript{12} suggest the need for antibiotic prophylaxis in patients who have implanted valves to prevent bacteremia while performing high-risk dental procedures.

In this study, 60\% of patients were female, coinciding with Salid \textit{et al.}\textsuperscript{13}, who reported a female: male ratio of 2:1.

Caries prevalence in patients with SB is usually higher than in the rest of the population because their neurological disorder affects their oral hygiene.\textsuperscript{1,14} Likewise, some authors as Queiroz \textit{et al.}\textsuperscript{2} and Solanki \textit{et al.}\textsuperscript{15} suggest that a poor diet and prolonged use of sugary medicines contribute to the development of caries at an early age. In the present study only 46.7\% of the patients had cavities, a figure which increases with the child’s age. This finding coincides with data reported in other studies.\textsuperscript{16}

Regarding the presence of cleft lip and palate, they rarely appear simultaneously with neural tube defects.\textsuperscript{17,18} Accordingly, in the population studied none of the patients had orofacial clefts. Recent studies relate SB and orofacial clefts with different genes. \textit{PAX3} haploinsufficiency has been linked to SB while the \textit{EPHA4} gene has been linked to palatal development.\textsuperscript{19}
Estado de salud bucal de una muestra de pacientes venezolanos con espina bífida. Estudio transversal.

Resumen: La espina bífida (EB) es una malformación congénita de la médula espinal con alteraciones vertebrales simultáneas, debido al cierre incompleto del tubo neural. El objetivo de este trabajo es reportar el estado de salud bucal de una muestra de pacientes venezolanos con EB. Materiales y métodos: se realizó un estudio observacional de corte transversal donde se evaluaron 30 pacientes con EB para determinar su estado de salud bucal y otras variables de interés. Resultados: Un 46,7% de los pacientes presentó historia de caries, con un 22,2% en el grupo de 1-4 años, 71,4% en el grupo 5-7 años y del 100% en los pacientes de 8 a 16 años. El índice ceod fue de 1.55 y el COPD de 3.50. Un 46,7% presentó gingivitis, el 30% presentó cálculo dental, siendo el IHOS de 2. El 83% presentó clase II de Angle y el 17% clase I de Angle. El 40% tenía hábitos parafuncionales, siendo éstos, la succión digital, uso de pacificadores y la onicofagia. El 70% de los pacientes presentó paladar profundo. Conclusiones: Los pacientes con EB presentan características bucales particulares y factores de riesgo que deben ser considerados durante la consulta odontológica para brindarle una atención de adecuada y mejorar su calidad de vida.

Palabras clave: Espina bífida, Disrafía espinal, Hidrocefalia, Caries dental, Maloclusión.

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