# Oral Manifestations and Treatment of a Child With Sjögren's Syndrome

*Fernanda Sardenberg, DDS, MSc* *Daniela Goursand, DDS, MSc*
*Luiz Thadeu Polletto, DDS, MSc, PhD* *Miriam Pimenta Vale, DDS, MSc, PhD*
*Patrícia Maria Zarzar, DDS, MSc, PhD* *Saul Martins Paiva, DDS, MSc, PhD*

## ABSTRACT

Sjögren's syndrome (SS) is a chronic inflammatory systemic autoimmune disease affecting the exocrine, salivary and lacrimal glands. The condition occurs more often in adults and is rare in childhood. SS should be considered in the differential diagnosis of recurrent parotitis and keratoconjunctivitis sicca. Oral manifestations include early tooth decay and xerostomia. Treatment consists of operative dentistry and saliva management. This paper reports a case of a 10-year-old Brazilian boy with SS, stressing the oral manifestations, treatment conduct, clinical importance and need for an early diagnosis in order to improve the patient's quality of life.

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Sjögren's syndrome (SS) is a chronic, inflammatory, systemic autoimmune disease affecting the exocrine, salivary, and lacrimal glands. The condition occurs more often in adults and is rare in children. SS should be considered in the differential diagnosis of recurrent parotitis and keratoconjunctivitis sicca. Oral manifestations include early tooth decay and xerostomia. Treatment consists of operative dentistry and saliva management. The purpose of this paper was to report the case of a 10-year-old Brazilian boy with SS, stressing the oral manifestations, treatment conduct, clinical importance, and need for an early diagnosis to improve the patient's quality of life.

*Drs. Sardenberg and Goursand are Masters in Pediatric Dentistry, Department of Pediatric Dentistry and Orthodontics; Dr. Polletto is an associate professor, Department of Operative Dentistry; and Drs. Vale, Zarzar, and Paiva are associate professors, Department of Pediatric Dentistry and Orthodontics, all at the Federal University of Minas Gerais, Belo Horizonte, Brazil.*

*Correspond with Dr. Sardenberg at fesardenberg@hotmail.com*

The purpose of this paper was to describe the case of a child with Sjögren's syndrome. Symptoms included parotid swelling, xerostomia, and xerophtalmia. Treatment for the oral manifestations is presented and proved effective in resolving the child's condition.

Sjögren's syndrome (SS) is an autoimmune disease of the exocrine glands characterized by chronic lymphocytic and plasma cell infiltration of lacrimal and salivary glands, leading to a progressive reduction in saliva (xerostomia) and tear flow (xerophtalmia).[1,2] The primary form of the syndrome (pSS) has characteristic inflammatory cell involvement of the exocrine glands, while the secondary (sSS) form involves various systemic autoimmune disorders and connective tissue diseases in addition to exocrine gland involvement. Some of the most common autoimmune diseases associated with SS are rheumatoid arthritis, systemic lupus erythematosus, renal tubular acidosis (RTA) and scleroderma.[2-6]

SS affects approximately 0.2% of the general adult population and occurs approximately 9 times more often in women than men. Average age at the onset of the clinical symptoms is around 40 years.[7,8] The European

Study Group on Classification Criteria for SS validated classification criteria for this syndrome between 1989 and 1996.[9] In 2002, a revised version of the European criteria was proposed by the American-European Consensus Group.[10] These criteria require the evaluation of xerophthalmia and xerostomia, salivary gland biopsy evaluated by an expert histopathologist, and laboratory tests evaluating serum autoantibodies, such as rheumatoid factor, antinuclear antibody (**ANA**), anti-Ro (**SS-A**) and anti-La.[1,3,5]

The diagnosis of SS in childhood is difficult, as the condition is rare in children and can exhibit unexplained RTA and recurrent parotid swelling, which may contribute to a higher frequency of undiagnosed cases.[6,11-15] Examination procedures and diagnostic criteria for pediatric SS are based on a physical examination (dryness of the eyes or mouth and enlarged parotid glands), clinical history findings (such as recurrent parotid swelling), and laboratory findings of lymphocytic infiltration in a minor salivary gland.[13,14,16,17]

Evaluating the oral component in SS should be included as part of the standard health questionnaire, addressing symptoms of dryness such as the frequent use of liquids to assist in swallowing food and changes in taste.[1,6] Difficulties in mastication and swallowing are most specifically related to advanced salivary gland hypofunction.[18]

Easy-access body fluids such as saliva may provide a less invasive diagnosis and a source of predictive markers for various diseases, including SS.[19] A multidisciplinary approach should be used to treat these patients, with the aim of improving their quality of life.[12]

## CASE REPORT

A 10-year-old Brazilian boy presented to the Clinic of Pediatric Dentistry of the Federal University of Minas Gerais, Belo Horizonte, Brazil, complaining of dry mouth, dental caries, pain, and discomfort while eating. According to his mother, the patient had several episodes of recurrent parotitis in the previous year. Following clinical diagnosis, treatment with antibiotics and anti-inflammatory agents was established. At presentation, the patient had a history of xerostomia for several months, complained of his eyes feeling sandy, and experienced symptoms of fatigue. After these episodes of recurrent parotitis, the pediatrician sent the patient to a rheumatologist, who requested laboratory tests. The laboratory findings revealed that the patient was antinuclear antibody positive (titer 1:1280) and SS-A positive. Eye involvement was evident in the form of positive Schirmer and Rose Bengal tests and xerophtalmia is been treated with artificial tears (Restasis, Allergan Inc, Irvine, Calif) prescribed by his ophthalmologist from September 2005 up to now, before coming to our department. The patient was diagnosed with Pss by a rheumatologist.

Oral examination revealed dry, peeling lips and angular cheilitis as well as dry and erythematous mucosa with mucosal sloughing. The tongue appeared furrowed, atrophic and depapillated (Figure 1). Sialometry, a test for measuring saliva flow, was used to confirm the xerostomia.[20] Passively expectorated saliva was collected every 2 minutes for a total of 6 minutes, allowing deglutition of saliva only between the 2 minutes intervals. The amount of saliva collected was 0.16 mL per minute. The child was then given a paraffin tablet to chew on for 6 minutes, with saliva collected every 2 minutes. This salivary stimulation provided 0.25 mL of saliva per minute, demonstrating that there was salivary stimulation, albeit of low intensity.[20] This procedure was performed on the morning on the same day by a dentist.

To reduce oral dryness, a salivary flow stimulation protocol was implemented through mastication stimulus and high levels of hydration (intake of at least 2 liters of water per day).

Clinical examination revealed that in the maxillary, permanent incisors, both permanent canines, and left first and second premolars had decalcification. Both mandibular lateral incisors also had decalcification. In structions regarding oral hygiene and a protocol for caries prevention were implemented, including plaque

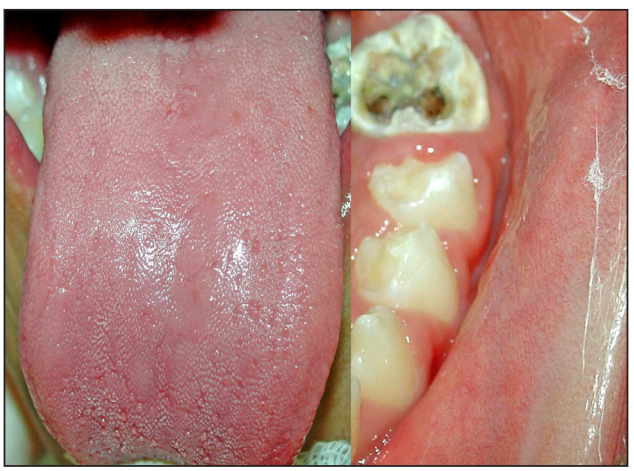

*Figure 1. Depapillated tongue and dry mucosa.*

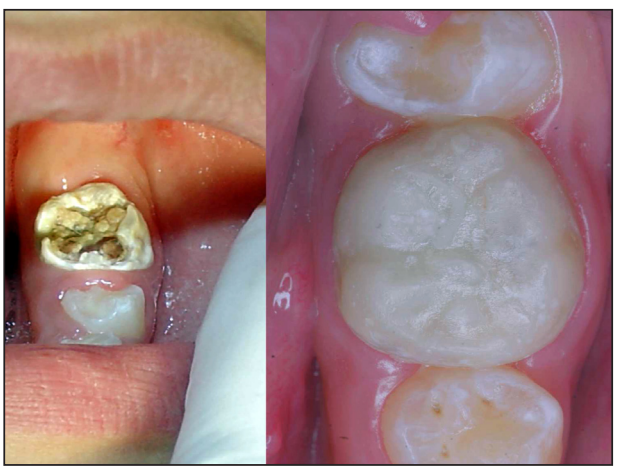

*Figure 2. Carious lesion in the permanent mandibular left first molar and corresponding restoration.*

control, diet modification, and regular fluoride application. Restorative treatment was also performed.

The permanent maxillary right central incisor had caries in the dentin on the labial surface, and the permanent mandibular left first molar had dentinal carious lesions on the occlusal surface. Restorative treatment for these 2 teeth was conducted with a composite resin (Z100, 3M, St. Paul, Minn). The final results of the restorations were considered satisfactory from both the normative standpoint as well as the standpoint of the child and parents (Figure 2).

The patient has been followed up with every 3 months for 2 years, with the monitoring of saliva flow, application of topical fluoride gels, and evaluations of carious activity. There have been no complications or new carious lesions. The patient is being monitored for caries in a recall program every 6 months.

## DISCUSSION

SS is frequently an undiagnosed disease in children, as it is a very rare condition in the pediatric age and clinical manifestations may be different from those found in adults.[11,13-17,21] It is difficult to obtain adequate patient history data on children, as initial clinical manifestations in childhood are nonspecific signs and symptoms such as fever, exanthema, arthralgia, and pain. The classic diagnostic criteria successfully used for adults[10] prove to be limited in diagnosing children.[16,17,21,22]

Recurrent parotid swelling seems to be the most common clinical manifestation in children in early stages of SS and is defined as recurrent parotitis (**RP**)—inflammation characterized by recurrent episodes of painful unilateral or bilateral parotid enlargement associated with swelling, fever, redness, and reduction in salivary flow. Therefore, a pediatric patient with possible RP should be evaluated for SS.[16,21,23,24]

RTA, an extraglandular manifestation, is an underrecognized complication of pediatric SS that should be considered in all older children and adolescents. The possible severe consequences include interstitial nephritis, glomerulonephritis, and hyposthenuria.[6]

Clinical and serologic findings from pediatric patients with pSS demonstrate that nearly 80% are female and the mean age of pSS children is 9.7 years. The most common sign of pediatric pSS is parotid enlargement (present in 70% of cases), followed by signs or symptoms of eye involvement (present in 70%) and dry mouth (present in 66% of cases).[23,25,26] These findings agree with data on our patient, who also exhibited persistent swelling of the parotid glands, reduced salivary flow, and symptoms of dry mouth and dry eye.

Values of nonstimulated salivary flow under of 0.1 mL/minute and values of stimulated salivary flow under 0.5 mL/minute signify that the patient has xerostomia.[20] In the case presented here, the patient exhibited 0.16 mL/minute of nonstimulated salivary flow and 0.25 mL/minute of stimulated salivary flow. The saliva collection techniques proposed by Sreebny and Valdini (1987) were used due to easier collection and lower cost. The loss of saliva in pSS increases vulnerability to dental caries and impairs all oral functions, which may lead to social withdrawal.[20,27] Since saliva helps prevent dental caries by promoting dental remineralization and maintaining a physiological oral pH level.

The form of the syndrome (pSS or sSS), specific signs or symptoms, and severity of clinical manifestations generally determine the type of treatment for pediatric SS patients. To be effective, a multidisciplinary team (dentist, primary care physician, ophthalmologist, and rheumatologist) should be involved in the patient's health care. Our patient was under the care of his ophthalmologist, who treated the xerophthalmia, and was referred for treatment on his dental needs. Including: restorative dental treatment with the goal of restoring the function and aesthetics of the affected teeth; topical fluorides to help control the activity of carious lesions; physiotherapy; and the intake of at least 2 liters of water per day to increase the amount of saliva.

SS's oral symptoms have a negative effect on quality of life.[28] An oral health program should be implemented in all cases. Clinical follow-up is important, as this syndrome's signs, symptoms and clinical course are complex and early diagnosis and proper treatment can prevent complications.[7,29,30] Problems involving the mouth and teeth have been shown to have a key impact on many aspects of daily living.[28,31] In the case presented here, the treatment of the oral symptoms (xerostomia and dental caries) improved the child's quality of life.

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