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# Salivary gland ultrasound in children: a useful tool in the diagnosis of juvenile Sjögren's syndrome

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## ABSTRACT

*Primary Sjögren's syndrome (pSS) is a systemic autoimmune disease uncommon in children, clinically characterised by recurrent parotitis at the onset, which is a common disorder in childhood, most of them of infectious origin. Juvenile pSS diagnosis is based on clinical symptoms and presence of autoantibodies, after exclusion of infectious or lymphoproliferative diseases. However, salivary gland ultrasound (SGU) shows typical features of pSS that can add useful information for the diagnosis of this disorder. We describe three patients who presented with recurrent parotitis in which characteristic autoantibodies and typical SGU pattern allow us to make the diagnosis of juvenile pSS. We suggest that in children with recurrent parotitis SGU and autoantibodies should be routinely performed.*

## Introduction

Primary Sjögren's syndrome (pSS) is a systemic autoimmune disease. In adults, this disease is characterised by ocular and mouth dryness; organ involvement is also frequent. The SG biopsy is included in the classification criteria for pSS. The positivity of SG biopsy is mandatory if autoantibodies are negative for diagnosing pSS. However, the sensitivity of SG biopsy is variable and the reliability has not been fully addressed (1, 2). Juvenile onset of pSS is uncommon, and clinical features are different from adults (3). In children, recurrent parotitis is the typical sign at the onset, and dryness appears after several years of disease (4-6). However, recurrent parotitis is a common disorder in childhood, most of them of infectious origin (*i.e.* viral or bacterial) (3, 5). These facts and the lack of validated diagnostic criteria in children make juvenile pSS a disease poorly known and probably underdiagnosed (7).

Some studies on adult pSS have described a typical pattern on salivary gland ultrasound (SGU) characterised by lost of the homogeneous internal echogenicity and echotexture, with multiple oval hypoechoic areas of different size (8, 9). These SGU features have been proposed to be included in the diagnostic criteria of adult pSS (10, 11). Similar SGU changes have been reported in juvenile pSS (3, 12). Herein we describe three juvenile pSS patients in which SGU provided important information for the diagnosis.

## Case reports

*Patient 1*

Patient 1, a 7-year-old girl, attended our hospital five years ago because of recurrent left parotitis. Lymphoproliferative and infectious diseases were ruled out by blood smears and appropriate microbiological test, respectively. She did not respond to antibiotics and was sent to paediatric rheumatology. The study was completed with SGU and autoantibodies. The immunologic study showed positivity of antinuclear antibodies (ANA), rheumatoid factor (RF), anti-SS-A and anti-SS-B and elevated immunoglobulin G. The SGU examination was performed with a liner probe (5-13 MHz), with a General Electrics Healthcare Logic E BT12, Wuxy, China. The SGU scanning technique consisted of a longitudinal sweeping for the submandibular glands and longitudinal and transverse sweepings for the parotid glands. SGU showed multiple oval hypoechoic areas inside both parotid and submandibular gland parenchyma. Although she did not complain of dryness salivary scintigraphy was requested, which was normal. Juvenile pSS was diagnosed and non-steroidal anti-inflammatory drugs (NSAID) were started. Afterwards, because of symptoms recurrence hydroxychloroquine (100 mg per day) was indicated. Symptoms improved after few months. Despite good clinical response, SGU findings are currently present (Fig. 1).

*Patient 2*

Patient 2, a 2-year-old boy, presented with bilateral recurrent parotitis four years ago. After ruled out infectious and lymphoproliferative diseases by standard methods he was sent to paediatric rheumatology. We performed an immunologic study and SGU. ANA and anti-SS-A were positive. The SGU examination was performed with a liner probe (5–13 MHz), with a General Electrics Healthcare Logic E

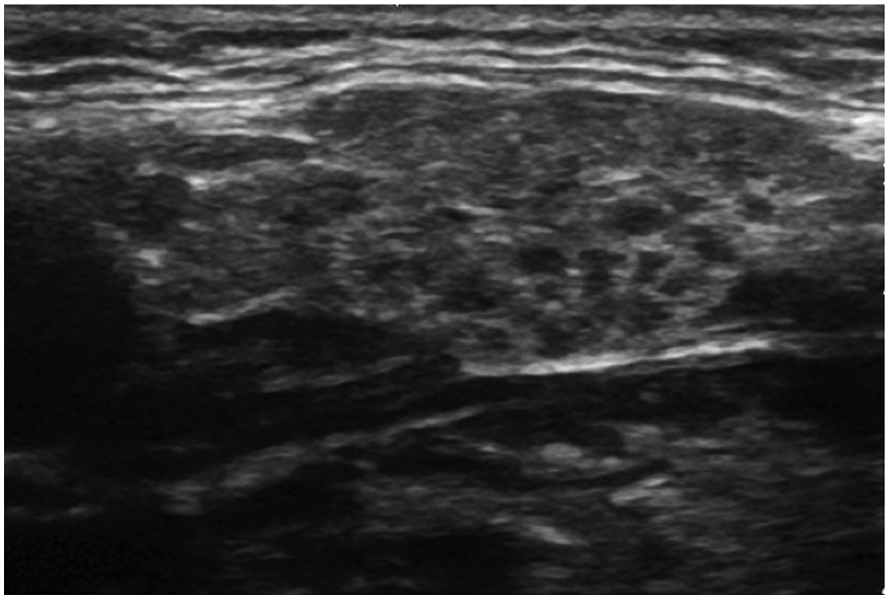

**Fig. 1.** Longitudinal ultrasound image of a submandibular gland showed multiple hypoechoic areas in the gland parenchyma.

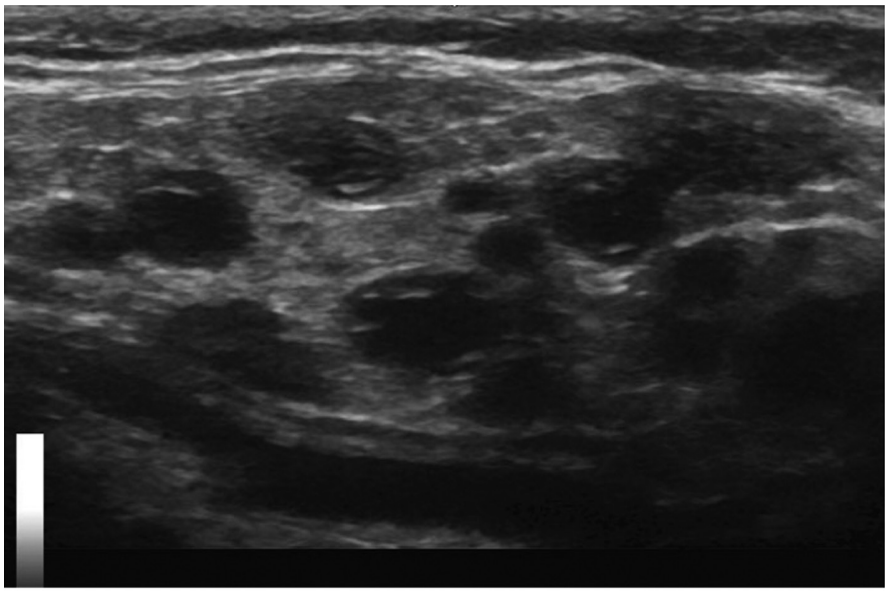

**Fig. 2.** Longitudinal ultrasound image of a parotid gland showed multiple hypoechoic cysts and dyshomogeneous gland parenchyma.

BT12, Wuxy, China. The SGU scanning technique consisted of a longitudinal sweeping for the submandibular glands and longitudinal and transverse sweepings for the parotid glands. SGU showed lymphadenopaties and multiple hypoechoic areas inside both parotid and submandibular gland parenchyma. He did not complain of ocular or mouth symptoms. However, salivary scintigraphy was typical for grade IV Sjögren syndrome. He was treated with hydroxycloroquine, 100 mg daily, with good clinical response. Currently, SGU pattern of pSS is still present (Fig. 2).

*Patient 3*

Patient 3, a 4 year-old girl attended our hospital three years ago because of recurrent right parotitis. Lymphoproliferative and infectious diseases were ruled out. We performed immunologic test and SGU. ANA, anti-SS-A, anti-SS-B, anti-Sm and anti-RNP/Sm autoantibodies were positives. SGU showed multiple hypoechoic areas inside both parotid and submandibular gland parenchyma. She was treated with NSAIDs and symptoms improved.

## Discussion

We describe three cases of recurrent parotitis, in which characteristic autoantibodies and typical SGU pattern allow us to make the diagnosis of juvenile pSS. Juvenile pSS is an uncommon disease. Cimaz *et al*. published the greater serie of juvenile pSS, with 40 patients from 6 different countries (3). Juvenile pSS may start with arthritis o renal involvement (13-14). However, recurrent parotitis is the most frequent symptom at the onset (3, 6). Diagnosis of juvenile pSS is usually based on clinical symptoms (*e.g*. recurrent parotitis) and presence of characteristic autoantibodies (ANA, RF, anti-SS-A and/or anti-SS-B) (3). However, SGU can add useful information for the diagnosis of this disorder.

Musculoskeletal ultrasound (MSUS) is an available, quick, safe, non invasive, friendly and relatively cheap imaging modality, which is increasingly used in paediatric rheumatology for improving diagnosis and management of patients with joint disease (15-16). In addition, high frequency transducers used for MSUS can be used for imaging other superficial soft tissues such as salivary glands. Our SGU findings were consistent with those reported in the literature (8, 9, 12).

Because parotitis, the most common symptom at juvenile pSS onset, can be mistaken for infections, we hypothetisised that the diagnosis of juvenile pSS can be delayed or underestimated. Thus, we suggest that in children with recurrent parotitis SGU and autoantibodies should be routinely performed.

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