

Sjögren's Syndrome: An Underestimated Disease Despite its High Prevalence: A Narrative Review

Farah Sabah Rasheed^{1*}, Zainab M Hussain², Farah Ghassan Ibrahim³

Abstract

Sjögren's syndrome (SS) is a chronic autoimmune disease characterized mainly by dysfunction of the exocrine glands, particularly the salivary and lacrimal glands. The resulting reduction in salivary and tear secretion produces the characteristic symptoms of dry mouth and dry eyes. However, the disease has a broader clinical spectrum and may involve several organs outside the exocrine glands.

Oral manifestations are particularly important because salivary gland dysfunction can produce persistent xerostomia and hyposalivation, followed by changes in the oral mucosa, increased dental caries, periodontal problems, oral candidiasis, and recurrent salivary gland inflammation. These complications may have a significant effect on oral health and quality of life. Systemic manifestations may also occur, including neurological and pulmonary involvement, while a proportion of patients are at increased risk of lymphoproliferative complications.

Sjögren's syndrome occurs predominantly in women and is commonly recognized during middle age. Despite its relatively frequent occurrence among autoimmune diseases, diagnosis may be delayed because the early symptoms can be nonspecific and may remain unrecognized for years. Early identification is therefore important, particularly when persistent oral or ocular dryness is present.

This narrative review discusses the epidemiology, pathogenesis, clinical manifestations, oral complications, systemic involvement, diagnosis, and management of Sjögren's syndrome, with particular emphasis on the oral manifestations and their relevance to early recognition of the disease

Keywords

Sjögren's syndrome; autoimmune disease; xerostomia; hyposalivation; oral manifestations; dental caries; salivary glands.

^{*1}Ph.D. Lecturer of Oral Pathology, Department of Oral Diagnosis, College of Dentistry, Uruk University, Baghdad, Iraq.

²Department of Oral Medicine, College of Dentistry, Al-Iraqia University, Baghdad, Iraq.

³M.Sc. Oral Pathology, Ministry of Health, Al Imamein Al Kadhimein Medical City, Baghdad, Iraq

INTRODUCTION

Sjögren's syndrome is a chronic systemic autoimmune disease that mainly affects the exocrine glands. The salivary and lacrimal glands are the principal sites of involvement, leading to reduced secretion of saliva and tears and consequently to xerostomia and xerophthalmia [3,4,8,9].

The disease may occur as a primary disorder or in association with another autoimmune connective tissue disease. Rheumatoid arthritis, systemic lupus erythematosus, and systemic sclerosis are among the conditions reported in association with secondary Sjögren's syndrome [3,4]. Although dryness of the mouth and eyes is the most recognizable clinical presentation, SS should not be considered a disease limited to the exocrine

glands. Neurological, pulmonary, and other systemic manifestations have also been described [1,4,8,11,12].

The disease predominantly affects women, with a female-to-male ratio of approximately 9:1. It is generally diagnosed around the fifth decade of life, although symptoms may begin several years before diagnosis [3–5]. This difference between the onset of symptoms and formal diagnosis is one of the reasons that SS may remain unrecognized for a considerable period.

The oral cavity is particularly affected by impaired salivary function. Saliva is important for lubrication, swallowing, maintenance of the oral mucosa, and protection of the teeth. A persistent reduction in salivary secretion can therefore

***Corresponding Author:** Farah Sabah Rasheed

© The Author(s) 2026. This is an open-access article distributed under the terms of the Creative Commons Attribution License (CC-BY-NC)

produce a range of oral complications, including dental caries, mucosal discomfort, candidiasis, and salivary gland problems [8,10].

Recognition of these findings is important for both dental and medical practitioners because oral manifestations may be among the first clinical clues suggesting an underlying systemic autoimmune disease.

EPIDEMIOLOGY

Sjögren's syndrome predominantly affects women and is most frequently recognized during middle age. The marked female predominance is one of the most consistent epidemiological characteristics of the disease [3].

The reported frequency of SS differs between populations. These differences may be related to geographical variation as well as differences in diagnostic approaches and the criteria used to identify affected patients [3]. The epidemiological assessment of SS is also complicated by the variability of its clinical presentation and the possibility of delayed diagnosis.

Primary SS occurs independently, whereas secondary SS develops in patients who already have another systemic autoimmune disease. The latter is particularly associated with rheumatoid arthritis, systemic lupus erythematosus, and systemic sclerosis [3,4].

The clinical burden of SS cannot be assessed solely by the presence of glandular dryness. Systemic involvement occurs in some patients and may affect organs outside the salivary and lacrimal glands [4,11]. Consequently, recognition of the disease requires attention to both glandular and extra glandular manifestations.

ETIOLOGY AND PATHOGENESIS

The etiology of Sjögren's syndrome remains incompletely understood. The available evidence supports a multifactorial process in which genetic susceptibility interacts with environmental factors and immune dysregulation [3,8,9].

A characteristic feature of the disease is lymphocytic infiltration of the exocrine glands. This inflammatory process interferes with normal glandular function and progressively reduces salivary and lacrimal secretion. The resulting glandular dysfunction accounts for the main symptoms of xerostomia and xerophthalmia.

The immune response involves both cellular and humoral mechanisms. B-cell activation and autoantibody production are important features of the disease and may contribute to manifestations beyond the exocrine glands [8,9]. Persistent B-cell stimulation is also relevant to the development of lymphoproliferative disorders in a subset of patients with SS [2].

The pathogenesis therefore provides a link between the local glandular manifestations and the systemic nature of the disease. The same autoimmune process responsible for glandular dysfunction may be associated with involvement of other organs and with complications that require long-term follow-up.

CLINICAL MANIFESTATIONS

The clinical presentation of Sjögren's syndrome is variable. Some patients mainly experience symptoms related to glandular dysfunction, while others develop systemic manifestations that may become an important part of the disease course [4,8,11].

Glandular manifestations

The principal glandular manifestations are dry mouth and dry eyes. Oral dryness may be persistent and can interfere with eating, swallowing, speaking, and general oral comfort.

Salivary gland enlargement may also occur, particularly involving the major salivary glands. Recurrent swelling and inflammation can be seen in some patients and may be associated with impaired glandular function [4,11].

Ocular involvement results from reduced lacrimal secretion and may produce persistent dryness and irritation. The severity of glandular symptoms

varies among patients and does not necessarily reflect the extent of systemic involvement.

Extraglandular manifestations

Sjögren's syndrome is a systemic autoimmune disease, and manifestations outside the exocrine glands are well recognized [4,8,11]. Their clinical presentation is variable and may involve different organ systems.

Neurological involvement has been reported in patients with SS and may affect both the peripheral and central nervous systems [1]. In some patients, neurological manifestations may represent an important part of the clinical presentation.

Pulmonary involvement is another recognized extraglandular manifestation. Respiratory abnormalities may occur in patients with SS and should be considered when respiratory symptoms are present [12].

Lymphoproliferative complications represent another important concern. Patients with Sjögren's syndrome have an increased risk of lymphoma, reflecting the importance of persistent immune and B-cell activation in the disease [2].

ORAL MANIFESTATIONS

The oral manifestations of Sjögren's syndrome are mainly related to reduced salivary secretion. Because saliva has several protective functions in the oral cavity, persistent hyposalivation can produce substantial changes in the oral environment [8–10].

Xerostomia and hyposalivation

Xerostomia is one of the most common complaints in patients with SS. Patients may experience a persistent sensation of dryness, difficulty swallowing dry foods, increased thirst, difficulty speaking for prolonged periods, and oral discomfort.

The clinical importance of xerostomia extends beyond subjective discomfort. Reduced salivary flow compromises the normal cleansing and protective functions of saliva and creates an

environment in which several oral complications become more likely.

Changes in the oral mucosa and tongue

Patients with reduced salivary secretion may develop dry, thin, or shiny oral mucosa. The tongue may become smooth or depapillated, and some patients report burning or soreness.

These changes can interfere with eating and may make the oral cavity more sensitive to certain foods. Persistent mucosal discomfort can also have an effect on the patient's daily activities and overall oral quality of life.

Dental caries

Dental caries is an important oral complication of primary Sjögren's syndrome. The reduction in saliva decreases several protective mechanisms that normally help maintain tooth integrity.

Patients may therefore develop increased caries activity and lesions in sites that are not usually highly susceptible to decay. The relationship between primary SS and caries risk has been specifically examined, emphasizing the need for regular dental assessment and preventive care [10].

The prevention of dental caries in these patients requires particular attention because the underlying reduction in salivary function may persist for a long period.

Periodontal problems

Periodontal problems may occur in patients with Sjögren's syndrome. Changes in salivary flow and the oral environment may influence oral hygiene and the balance of microorganisms within the mouth.

However, periodontal disease is multifactorial. Its assessment in patients with SS should therefore include the usual periodontal risk factors as well as the effects of salivary gland dysfunction.

Oral candidiasis

Reduced salivary secretion can increase susceptibility to oral fungal infection. Candidiasis

may present with oral soreness, burning, mucosal changes, or altered taste.

The presence of recurrent candidiasis in a patient with persistent oral dryness should therefore raise attention to possible underlying salivary gland dysfunction and, when appropriate, to an underlying systemic condition such as Sjögren's syndrome [8].

Salivary gland enlargement and sialadenitis

Major salivary gland enlargement is a recognized manifestation of SS. Recurrent swelling may occur as a consequence of the underlying glandular disease and impaired salivary function [4,11].

Patients may also experience bacterial sialadenitis. Persistent or progressive glandular enlargement requires appropriate assessment because of the established association between Sjögren's syndrome and lymphoproliferative disease [2].

SYSTEMIC INVOLVEMENT

The systemic nature of Sjögren's syndrome is an important feature that distinguishes it from isolated disorders of salivary or lacrimal gland function. Clinical manifestations outside the exocrine glands have been described in several organ systems [4,8,11].

Neurological involvement

Neurological manifestations represent an important extraglandular component of SS. Both central and peripheral neurological involvement have been described [1].

The presentation may vary between patients, and neurological symptoms can occasionally complicate the recognition of the underlying autoimmune disease. For this reason, unexplained neurological manifestations in a patient with other features suggestive of SS warrant appropriate clinical evaluation.

Pulmonary involvement

Pulmonary manifestations are another recognized feature of Sjögren's syndrome. Respiratory

involvement may affect different parts of the respiratory system and can contribute to morbidity in affected patients [12].

Dryness of the respiratory tract may contribute to respiratory symptoms, while more extensive pulmonary involvement may occur in some patients. The presence of persistent cough or other unexplained respiratory symptoms should therefore be considered in the overall assessment of patients with established SS.

Lymphoproliferative complications

One of the most clinically important complications of Sjögren's syndrome is the increased risk of lymphoma. The relationship between SS and lymphoproliferative disease is associated with chronic immune stimulation and B-cell activation [2].

Persistent or unusual enlargement of the salivary glands is therefore clinically significant. Although glandular enlargement is common in SS, changes in its pattern or persistence should not be overlooked.

DIAGNOSIS AND EARLY RECOGNITION

The diagnosis of Sjögren's syndrome requires consideration of the clinical presentation together with appropriate laboratory and, when indicated, histopathological assessment. Because symptoms may develop gradually and vary considerably between patients, early recognition can be challenging [5,7].

Persistent xerostomia or xerophthalmia should not automatically be considered an isolated local problem, particularly when accompanied by salivary gland enlargement, recurrent oral infection, extensive dental caries, or systemic manifestations.

Serological assessment has an important role in the evaluation of suspected patients. The overall clinical and serological profile may also be associated with differences in disease presentation and systemic involvement [4].

When the diagnosis remains uncertain, evaluation of the minor salivary glands may provide additional information. The diagnostic approach should therefore be based on the combination of clinical findings and appropriate investigations rather than on a single symptom or laboratory result [5,7].

Early diagnosis is particularly relevant because recognition of SS allows appropriate management of glandular symptoms, prevention of oral complications, assessment of systemic involvement, and identification of patients who require monitoring for important complications.

MANAGEMENT

Management of Sjögren's syndrome is directed toward controlling symptoms, preserving glandular and oral function, preventing complications, and addressing systemic disease when present. Treatment should therefore be individualized according to the clinical manifestations and severity of disease [6].

Management of glandular symptoms

Management of dryness focuses primarily on improving lubrication and relieving symptoms. Patients with oral dryness require measures aimed at maintaining adequate oral moisture and reducing the consequences of hyposalivation.

Ocular symptoms require appropriate local measures, while patients with significant systemic involvement may require additional medical therapy according to the organs affected [6].

ORAL MANAGEMENT

Oral care is a major component of the long-term management of patients with SS. Because reduced salivary flow increases the risk of caries and mucosal complications, preventive dental care should be maintained regularly [6,10].

Particular attention should be given to:

- regular dental examinations;
- prevention and early detection of dental caries;

- maintenance of good oral hygiene;
- management of oral fungal infection when present;
- measures to relieve oral dryness;
- monitoring for recurrent salivary gland swelling or infection.

The dental management plan should be adapted to the patient's degree of salivary dysfunction and existing oral disease.

Role of Dental Professionals in Early Recognition

The dental professional has an important role in the recognition of Sjögren's syndrome because oral manifestations may be prominent and may appear before the systemic diagnosis is established.

A patient presenting with persistent xerostomia, extensive or recurrent dental caries, oral burning, candidiasis, mucosal changes, or recurrent salivary gland enlargement should be assessed carefully. When these findings are associated with ocular dryness or other systemic complaints, an underlying autoimmune disorder should be considered.

Early recognition by dental professionals may facilitate referral for appropriate medical assessment. This is particularly relevant because delayed diagnosis may allow oral complications to progress and may postpone the recognition of systemic disease [5,7].

Management also requires cooperation between dental and medical professionals. The oral findings should not be treated in isolation when they form part of a broader systemic disorder.

DISCUSSION

Sjögren's syndrome represents a clinical challenge because its most recognizable symptoms, dry mouth and dry eyes, can appear relatively simple compared with the complexity of the underlying systemic disease. The available literature demonstrates that SS involves a wide clinical spectrum, ranging from predominantly glandular

disease to significant systemic manifestations [4,8,11].

The oral consequences are especially relevant because salivary dysfunction directly affects the integrity of the oral environment. Dental caries, mucosal discomfort, candidiasis, and salivary gland complications may become persistent problems in affected patients [8,10]. These manifestations are not merely secondary complaints; they can be an important part of the patient's disease burden.

The systemic nature of SS further supports the need for early recognition. Neurological and pulmonary involvement have been documented, while lymphoma represents a serious long-term complication [1,2,12]. Therefore, persistent glandular symptoms or unusual clinical findings should be considered within the wider context of the disease.

Another important issue is the variability of clinical presentation. Patients may not present with the full classical picture, and the severity of oral, ocular, and systemic manifestations can differ substantially. This variability may contribute to delayed diagnosis and emphasizes the value of a careful clinical history and multidisciplinary assessment [5,7].

From a dental perspective, recognizing the relationship between hyposalivation and oral disease is particularly important. Prevention and early treatment of dental complications can reduce the burden of disease and improve the patient's long-term oral health. At the same time, recurrent or disproportionate oral manifestations may provide an opportunity to identify patients who have not yet been diagnosed with SS.

Overall, the literature supports viewing Sjögren's syndrome as a systemic autoimmune disease with substantial oral consequences rather than as an isolated disorder of salivary and lacrimal secretion.

CONCLUSION

Sjögren's syndrome is a chronic systemic autoimmune disease with prominent glandular and oral manifestations and a variable range of extraglandular involvement. Although xerostomia and xerophthalmia are the characteristic features, the clinical consequences of the disease extend to dental caries, mucosal changes, candidiasis, salivary gland inflammation, neurological and pulmonary manifestations, and an increased risk of lymphoma.

The oral manifestations are of particular importance because they may become evident early in the disease and can significantly affect oral health. Persistent dry mouth, recurrent oral infection, unexplained salivary gland enlargement, and extensive caries should therefore be assessed carefully, especially when accompanied by ocular or systemic symptoms.

Early recognition, appropriate investigation, preventive dental care, and cooperation between dental and medical professionals are important components of the long-term management of patients with Sjögren's syndrome. Greater awareness of the disease may help reduce diagnostic delay and improve the management of both its oral and systemic consequences.

REFERENCES

1. Andonopoulos, A. P., Lagos, G., Drosos, A. A., & Moutsopoulos, H. M. (1990). The spectrum of neurological involvement in Sjögren's syndrome. *British Journal of Rheumatology*, 29(1), 21–23.
2. Baldini, C., Pepe, P., Quartuccio, L., et al. (2014). Primary Sjögren's syndrome as a multi-organ disease: Impact of the serological profile on the clinical presentation of the disease in a large cohort of Italian patients. *Rheumatology*, 53(5), 839–844.
3. Berman, N., Vivino, F., Baker, J., Dunham, J., & Pinto, A. (2019). Risk factors for caries development in primary Sjögren syndrome. *Oral Surgery, Oral Medicine*,

- Oral Pathology and Oral Radiology, 128, 117–122.
4. Brito-Zerón, P., Theander, E., Baldini, C., et al. (2016). Early diagnosis of primary Sjögren's syndrome: EULAR-SS task force clinical recommendations. *Expert Review of Clinical Immunology*, 12(2), 137–156.
 5. Brito-Zerón, P., Theander, E., Baldini, C., Seror, R., Retamozo, S., Quartuccio, L., Bootsma, H., Bowman, S. J., Dörner, T., Gottenberg, J.-E., et al. (2016). Early diagnosis of primary Sjögren's syndrome: EULAR-SS task force clinical recommendations. *Expert Review of Clinical Immunology*, 12, 137–156.
 6. Mariette, X., & Criswell, L. A. (2018). Primary Sjögren's syndrome. *New England Journal of Medicine*, 378, 931–939.
 7. Mavragani, C. P., & Moutsopoulos, H. M. (2010). The geoepidemiology of Sjögren's syndrome. *Autoimmunity Reviews*, 9(5), A305–A310.
 8. Negrini, S., Emmi, G., Greco, M., Borro, M., Sardanelli, F., Murdaca, G., Indiveri, F., & Puppo, F. (2022). Sjögren's syndrome: A systemic autoimmune disease. *Clinical and Experimental Medicine*, 22, 9–25.
 9. Nocturne, G., & Mariette, X. (2015). Sjögren syndrome-associated lymphomas: An update on pathogenesis and management. *British Journal of Haematology*, 168, 317–327.
 10. Ramos-Casals, M., Brito-Zerón, P., Bombardieri, S., et al. (2020). EULAR recommendations for the management of Sjögren's syndrome with topical and systemic therapies. *Annals of the Rheumatic Diseases*, 79(1), 3–18.
 11. Ramos-Casals, M., Brito-Zerón, P., Solans, R., Camps, M.-T., Casanovas, A., Sopena, B., Díaz-López, B., Rascón, F.-J., Qanneta, R., Fraile, G., et al. (2014). Systemic involvement in primary Sjögren's syndrome evaluated by the EULAR-SS Disease Activity Index: Analysis of 921 Spanish patients (GEAS-SS Registry). *Rheumatology*, 53, 321–331.
 12. Stojan, G., Baer, A. N., & Danoff, S. K. (2013). Pulmonary manifestations of Sjögren's syndrome. *Current Allergy and Asthma Reports*, 13, 354–360.

Conflict of Interest: No Conflict of Interest

Source of Funding: Author(s) Funded the Research

How to Cite: Rasheed, F. S., Hussain, Z. M., Ibrahim, F. G. (2026). Sjögren's Syndrome: An Underestimated Disease Despite its High Prevalence: A Narrative Review. *Journal of Advanced Medical Research and Innovation*, 2(4), 9-15.