Facilitating timely diagnosis of Sjögren's syndrome

By Linda Douglas¹

Introduction

Sjögren's syndrome is an autoimmune disease in which intense lymphocytic infiltration of the exocrine glands¹ and hyperactivity of B lymphocytes² cause inflammation, which damages glandular tissue and impairs function. Primary Sjögren's syndrome is diagnosed in the absence of any other connective tissue disease, while secondary Sjögren's syndrome is accompanied by other autoimmune diseases, such as rheumatoid arthritis, lupus erythematosus, Graft Versus Host Disease, or in rare cases, Behçet's disease.³ Secondary Sjögren's syndrome comprises approximately 60% of cases.

In January 2012, the Sjögren's Syndrome Foundation launched a 5-year Breakthrough Goal, to reduce the time between onset and diagnosis of Sjögren's by 50% in five years. When this Goal was launched, research showed the average diagnosis time was nearly six years.⁴

Research at the University of Toronto found a median delay of four years between onset and diagnosis (range 0-28 years).⁵ Prompt diagnosis allows access to treatment to relieve symptoms, minimise complications, and enhance vigilance for lymphoma: Sjögren's patients have a 16 times greater risk of developing lymphoma than a healthy individual.⁶

Much of the delay between onset and diagnosis is related to the diverse symptoms, which make diagnosis difficult, plus a lack of awareness of the condition on the part of various healthcare professionals encountered by many Sjögren's patients, who frequently fail to 'connect the dots' regarding the numerous signs and symptoms affecting multiple body systems.

Clinical features of Sjögren's syndrome

Sjögren's syndrome is mainly characterised

neuropathy,⁷ hypothyroidism, and lymphoma.

Sjögren's syndrome and mental health

In common with many chronic medical diseases, depression or anxiety might accompany Sjögren's syndrome, related to its adverse effect on the quality of life. This is shown by its score on the Devlin's Illness Intrusiveness Scale,⁸ where the negative impact on quality of life is comparable with that of multiple sclerosis, or kidney dialysis.⁹ In addition, mounting evidence indicates that chronic exposure to elevated inflammatory cytokines causes persistent alterations in neurotransmitter systems, which can lead to neuropsychiatric disorders and depression.¹⁰

'ORAL-RELATED COMPLICATIONS INCLUDE

SIALOLITHS, SALIVARY GLAND SWELLING, HIGH

CARIES RISK, PERIODONTAL DISEASE, SOFT

TISSUE INFECTIONS AND ANGULAR CHEILITIS'

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by dry eyes and a dry mouth, and may also affect other mucosal tissues such as the nose, larynx, gastro-intestinal tract and vagina. Dry eyes can lead to blurred vision. Oral-related complications include sialoliths (salivary stones), salivary gland swelling, high caries risk, particularly at the cusp tips and cervical sites, periodontal disease, and soft tissue infections such as candidiasis, and angular cheilitis. Dysphagia might also occur, which can lead to aspiration pneumonia.

The systemic autoimmunity associated with Sjögren's syndrome can also result in dry skin, fatigue, low grade fever, constipation, myalgia, and joint pain. Other conditions which might occur include small vessel vasculitis, Raynaud's phenomenon, pulmonary symptoms, nephritis,

Prevalence and incidence of Sjögren's syndrome

An estimated 221,583 Britons are affected,¹¹ and approximately 4 million Americans. Ninety percent of Sjögren's patients are women.¹² Most are diagnosed in their late 40s, but it can also affect children and males. Males are usually diagnosed later than females.

Diagnosis of Sjögren's syndrome

Accurate diagnosis of Sjögren's syndrome begins with thorough assessment to expedite specialist referral. This includes general appraisal, medical history review, and screening for symptoms using questionnaires, followed by head and neck examination, and comprehensive intra-oral examination.

CPD questions

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Questionnaires to assess symptoms

Oral symptoms

A positive response to one or more of the following questions is suggestive of Sjögren's syndrome:

1. Have you had a daily feeling of dry mouth for more than three months?
2. Have you had recurrently or persistently swollen salivary glands as an adult?
3. Do you frequently drink liquids to aid in swallowing dry food?

Ocular symptoms

A positive response to at least one of the following questions is suggestive of Sjögren's syndrome:

1. Have you had daily, persistent troublesome dry eyes for more than three months?
2. Do you have a recurrent sensation of sand or gravel in the eyes?
3. Do you use tear substitutes more than three times a day?

Table 1 Classification criteria for Sjögren's syndrome

Diagnostic Criteria Item	Scoring
Anti-SSA/Ro antibody positivity	3 points
Focal lymphocytic sialadenitis with a focus score of 1 foci/4 mm ² or greater	3 points
An abnormal ocular staining score of 5 or greater (or van Bijsterveld score of 4 or more)	1 point
A Schirmer's test result of 5 mm/5 minutes or less	1 point
An unstimulated salivary flow rate of 0.1 ml/minute, or less	1 point

established for use in individuals with signs and/or symptoms suggestive of SS.

These single set classification criteria performed well in validation analyses, demonstrating 96% sensitivity and 95% specificity, and are also suitable criteria

Management of the dry mouth needs to be multifaceted, to address the multiple oral symptoms and prevent complications. This includes stimulation of salivary flow, conservation of functional salivary gland tissue, and saliva substitutes as required to protect and lubricate, and facilitate speech, mastication and swallowing. Supplementation with Omega 3 1,000 mg has been shown to improve the lubricating quality of the saliva, and the tears.¹⁵

Topical fluoride applications, and calcium and phosphate remineralisation preparations reduce caries risk.¹⁶ Swelling of the parotid and submandibular salivary glands (Fig. 2) can be reduced by application of heat and massage to aid salivary flow through the ducts.

Conclusion

We are getting closer to our goal of early diagnosis. In 2016, the Sjögren's Syndrome Foundation annual survey of newly diagnosed patients revealed that for those diagnosed in 2015, it took an average of three years to receive an accurate diagnosis,¹⁷ compared with almost six years in 2012. This was achieved by increasing public awareness, and increasing education and awareness among healthcare professionals.

Increased awareness of Sjögren's syndrome in the medical and dental communities facilitates improved outcomes for the Sjögren's patient, by enhancing inter-professional collaboration and vigilance. For example: screening questionnaires utilised by both medical and dental professionals could include questions on ocular, oral and systemic symptoms of Sjögren's syndrome.

Raising public awareness with more widely disseminated information on Sjögren's syndrome, and increased availability of screening questionnaires for self-assessment could be valuable. Knowledge also empowers patients to advocate for themselves as they navigate the healthcare system.

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Diagnostic techniques include blood testing for anti-SSA/Ro antibody, biopsy to assess the minor salivary glands inside the lower lip for focal lymphocytic sialadenitis, ocular staining with Rose Bengal, or the van Bijsterveld score to assess surface damage of the conjunctiva and cornea, Schirmer's test for lachrymal flow, and measurement of unstimulated salivary flow.

Other conditions which cause dry eyes and dry mouth should be considered in differential diagnosis, and excluded. For example: pre-existing lymphoma, hepatitis C infection, HIV infection, sarcoidosis, use of anti-cholinergic drugs, or a history of head and neck radiation therapy.

Current diagnostic criteria

The diagnostic criteria for Sjögren's syndrome is evolving, and has been revised numerous times since 1965.

In 2016, the American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR) developed and validated an international consensus of data-driven classification criteria for primary Sjögren's syndrome (SS), which were

for enrollment in clinical trials. The final classification criteria are based on the weighted sum of five items.¹³

Each objective criterion is allocated points: individuals with signs and/or symptoms suggestive of Sjögren's syndrome and a total score of at least 4 points meet the criteria for diagnosis of primary SS (Table 1).

Management of the Sjögren's patient in the dental practice setting

Extra-oral and intra-oral examination needs to be performed on a regular basis, to assess for complications of Sjögren's syndrome. The Challacombe Scale of Clinical Oral Dryness¹⁴ (Fig. 1) is a valuable tool for clinically assessing and quantifying the severity of oral dryness, and for evaluating the outcomes of our interventions. This scale is based on a Clinical Oral Dryness Score (CODS), and lists ten key features of dry mouth; one point is allocated for each feature, and the patient's additive score indicates the severity of the dry mouth. There is an inverse relationship between salivary flow rates, and CODS: a high CODS is related to hyposalivation.

Fig. 1 The Challacombe Scale of Clinical Oral Dryness, courtesy of Professor Stephen Challacombe

The Challacombe Scale

of Clinical Oral Dryness

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The Challacombe Scale was developed from research conducted at King's College London Dental Institute under the supervision of Professor Stephen Challacombe*. The purpose of this scale is to be able to visually identify and quantify whether your patient has xerostomia (dry mouth) and if so, how it changes over time and the most appropriate therapy options. This scale is applicable whatever your profession.

The Challacombe Scale works as an additive score of 1 to 10 : 1 being the least and 10 being the most severe. Each feature scores 1 and symptoms will not necessarily progress in the order shown, but summated scores indicate likely patient needs. Score changes over time can be used to monitor symptom progression or regression.

1		Mirror sticks to buccal mucosa	An additive score of 1 - 3 indicates mild dryness. May not need treatment or management. Sugar-free chewing gum for 15 mins, twice daily and attention to hydration is needed. Many drugs will cause mild dryness. Routine checkup monitoring required.
2		Mirror sticks to tongue	
3		Saliva frothy	
4		No saliva pooling in floor of mouth	An additive score of 4 - 6 indicates moderate dryness. Sugar-free chewing gum or simple sialogogues may be required. Needs to be investigated further if reasons for dryness are not clear. Saliva substitutes and topical fluoride may be helpful. Monitor at regular intervals especially for early decay and symptom change.
5		Tongue shows generalised shortened papillae (mild depapillation)	
6		Altered gingival architecture (ie. smooth)	
7		Glassy appearance of oral mucosa, especially palate	An additive score of 7 - 10 indicates severe dryness. Saliva substitutes and topical fluoride usually needed. Cause of hyposalivation needs to be ascertained and Sjögrens Syndrome excluded. Refer for investigation and diagnosis. Patients then need to be monitored for changing symptoms and signs, with possible further specialist input if worsening.
8		Tongue lobulated / fissured	
9		Cervical caries (more than two teeth)	
10		Debris on palate or sticking to teeth	

* S Osailan et al "Investigating the relationship between hyposalivation and mucosal wetness". (2011) Oral Diseases volume 17, Issue 1, Pages: 109-114

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Fig. 2 Chronic salivary gland enlargement

‘INCREASED AWARENESS OF SJÖGREN’S

SYNDROME IN THE MEDICAL AND

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