**Sjogren’s syndrome - A retrospective service evaluation of diagnosis and management in paediatric patients**

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**Background**

Sjogren's syndrome is an inflammatory disorder involving the exocrine glands, characterised by dry eyes and mucosal surfaces, fatigue and arthralgia. Sjogren's may occur in isolation (primary) or secondary to another autoimmune condition and is considered rare in the paediatric population (incidence and prevalence unknown).

Anecdotal reports of differences in clinical presentation and absence of validated paediatric classification criteria render reliable diagnosis challenging and optimal treatment pathways difficult to ascertain.

The American-European Consensus Group (AECG) criteria are considered gold standard for diagnosis of primary Sjogren’s syndrome in adults. A 2017 BSR Sjogren’s guideline recommends treating children according to the adult pathway.

We report a multicentre service evaluation exploring the presentation and management of children with Sjogren’s presenting to UK paediatric rheumatology services, with reference to existing adult criteria.

**Methods**

Retrospective study conducted using a standardised data collection proforma under local service evaluation permissions. Inclusion criteria: pre-existing diagnosis of either primary or secondary Sjogren’s syndrome under 16 years of age. Each case was assessed against the current adult criteria and guidance (AECG and BSR).

**Results**

20 patients (16 female) in eight UK centres with an mean age at diagnosis of 11 (range 2-16 years). 15 patients had primary Sjogren’s, 3 secondary Sjogren’s and 2 were not categorised.

No primary Sjogren’s patients fulfilled AECG diagnostic criteria. Dry eye symptoms were present in 13%, dry mouth symptoms in 53% and 14/15 patients were tested for anti Ro/La (100% anti-Ro positive, 43% anti-La positive). Objective ocular dryness (Schirmers or van Bijsterveld positive), objective oral dryness and labial gland focus score were not formally assessed in any patients. No primary Sjogren’s patients fulfilled ACR EULAR diagnostic criteria and one patient fitted exclusion criteria.

The most common primary Sjogren’s symptoms included parotid pain or swelling (73%), dry mouth (53%), arthralgia (47%) and fatigue (40%). Rash and headaches were also described whereas fever and lymphadenopathy were not reported. Sicca symptoms were reported in 100% secondary Sjogren’s patients (66% had parotid pain or swelling.)

Management of all patients was assessed against 2017 BSR guidelines; There was documented evidence in 75% of receiving education, diet and lifestyle advice. 50% were prescribed moisturising eye drops. Hydroxychloroquine treatment was considered in 75%; of these, 4% were not referred for annual eye screening.

**Conclusion**

Sjogren’s syndrome is rare in children and specific paediatric guidance does not exist. Despite extensive investigations no primary Sjogren’s patients fulfilled the AECG or ACR EULAR diagnostic criteria due to difficulty in obtaining objective evidence of oral or ocular dryness. These data suggest that primary paediatric presentations may differ from adult cohorts with frequent parotid pain or swelling and less self-reporting of sicca symptoms. Further work is needed to support development of paediatric specific guidelines for Sjogren’s syndrome.