Figure 1 Discordance between doctors concerning the level of serum uric acid that is thought to inflect adequate control of gout.

within 2 weeks. Sixteen per cent of respondents would treat asymptomatic hyperuricaemia with ULT, with a marked difference in practice being observed between rheumatologists and internal medicine specialists (15% versus 59%, respectively). Importantly, opinions varied considerably on what sUA level might be regarded as representing adequate control in a gout patient (fig 1).

It is interesting to consider some of these findings in light of the recent EULAR recommendations. These make clear, for example, that sUA levels do not confirm or exclude a diagnosis of gout. Despite this, nearly all respondents (97%) stated that they would rely on measuring sUA for precisely that purpose. For gout management the recommendations assign a central role to sUA and define the therapeutic goal of ULT as being to promote crystal dissolution, prevent crystal formation and improve clinical symptoms by maintaining sUA below 6 mg/dl. The survey findings show, however, that most physicians perceive a reduction in the frequency of acute attacks as the central goal of therapy and that the ability to cure the condition by maintaining sUA below 6 mg/dl is not yet universally appreciated.

A caveat of any such survey is that physicians may have responded on the basis of what they know should be done rather than what they actually do in practice. They may also have completed the survey in haste and possibly misinterpreted some of the questions. Nevertheless, a large number of practitioners representing several countries in Europe participated in this simple survey.

The authors hope that, as awareness of the EULAR recommendations grows, and the goal of maintaining sUA below 6 mg/dl becomes more widely recognised, all patients with gout will be spared the burden of this eminently treatable condition.

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REFERENCES

- 1 **Zhang W**, Doherty M, Pascual E, *et al*. EULAR evidence based recommendations for gout. Part I: diagnosis. Report of a task force of the EULAR Standing Committee for International Clinical Studies Including Therapeutics (ESCISIT). *Ann Rheum Dis* 2006;**65**:1301–11.
- 2 **Zhang W**, Doherty M, Bardin T, *et al*. EULAR evidence based recommendations for gout. Part II: management. Report of a task force of the EULAR Standing Committee for International Clinical Studies Including Therapeutics (ESCISIT). *Ann Rheum Dis* 2006;**65**:1312–24.

Like father, like son

Jeffrey Lee, Peter Merry, Richard Ball, Karl Gaffney

We present a unique case of familial primary Sjögren's syndrome (pSS) involving a father and son that challenge several key features of this disease onset.

A 20-year-old man (fig 1) was referred with a 4-year history of recurrent parotitis and persistent bilateral parotid swelling. He also had increasing fatigue and night sweats for 3 months and cosmetically unacceptable parotid swellings. Although Schirmer's test was normal, he was found to have positive Ro, La and antinuclear antibodies. Computerised tomography scans of his abdomen and chest showed no abnormality. Subsequent parotid biopsy showed a low-grade mucosa-associated lymphatic tissue (MALT) lymphoma. A diagnosis of pSS that was complicated by MALT lymphoma was made. The parotid swellings were reduced by 50% with no further fatigue or night sweats after the introduction of prednisolone 30 mg once daily and hydroxychloroquine.

On subsequent inquiry, his 55-year-old father was found to have had a 15-year history of bilateral parotid swellings for

which he never sought medical attention (fig 2). He was a mechanical engineer who gave a 15-year history of dry eyes and mouth and recurrent cervical lymphadenopathy. His Schirmer's test was strongly positive, with measurements of 2 mm in his left eye and 0 mm in his right. He also had positive Ro and La antibodies along with hypergammaglobulinaemia (immunoglobulin (Ig)G 25.2 (4.9–16.1) g/l, IgM 3.95 (0.60–2.1) g/l), lymphopenia (lymphocyte 0.4×10^9 (1.1–3.5)/l) and an erythrocyte sedimentation rate of 92 (1–10) mm/h. As he remained relatively asymptomatic apart from his ocular and oropharyngeal dryness, he was treated with artificial tears only.

This case report highlights several unusual features. Although familial clustering of different autoimmune diseases in individuals with pSS have been reported, familial cases of

Abbreviations: MALT, mucosa-associated lymphatic tissue; pSS, primary Sjögren's syndrome

Figure 1 A 20-year-old man with recurrent parotitis and persistent bilateral swelling.

Figure 2 A 55-year-old man presented with bilateral parotid swelling.

pSS at any age are uncommon.¹ A literature search shows no previous documented cases of familial pSS involving father and son. pSS has a peak incidence in the fourth and fifth decades of life, with a female:male ratio of 9:1.² Both of our patients were male members and one patient presented with symptoms since his teenage years.

Table 1 Classification criteria as defined by the European Study Group on classification of Sjogren's Syndrome⁸ in index case and parent

Criteria	Index case	Parent
Ocular symptoms	—	+
Oral symptoms	+	+
Positive Schirmer's test	—	+
Autoantibodies (to Ro and La)	+	+

+, presence of criteria; — absence of criteria.

Although a genetic predisposition to pSS appears to exist, the level of genetic contribution is unknown.¹ Family members with pSS seem to share similar phenotypes, with almost identical clinical presentation and serological data.^{3,4} In our case, the proband presented in the patient's early 20s with MALT lymphoma, whereas his father had lived with the symptoms of pSS undiagnosed (table 1). Lymphoma is usually a late complication of pSS, with a median time from pSS diagnosis to lymphoma diagnosis of 7.5 years.^{5,6}

Genetic studies of pSS already show strong associations with major histocompatibility complex genes, including HLA-DR3, which may also determine the severity of the autoimmune disease.⁷ Future studies comparing the clinical, serological and genetic features of familial pSS are needed to gain further understanding of this complex multifactorial disorder.

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REFERENCES

- Bolstad AI**, Jonsson R. Genetic aspects of Sjogren's syndrome. *Arthritis Res* 2002;**4**:353–9.
- Jonsson R**, Haga H-J, Gordon T. Sjogren's syndrome. In: Koopman WJ, eds. *Arthritis and allied conditions: a textbook of rheumatology*, 14th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001:1736–59.
- Scofield RH**, Kurien BT, Reichlin M. Immunologically restricted and inhibitory anti-RO/SSA in monozygotic twins. *Lupus* 1997;**6**:395–8.
- Bolstad AI**, Haga HJ, Wassmuth R, Jonsson R. Monozygotic twins with primary Sjogren's Syndrome. *J Rheumatol* 2000;**27**:2264–6.
- Kassan SS**, Thomas TL, Moutsopoulos HM, Hoover R, Kimberley RP, Budman DR. Increased risk of lymphoma in sicca syndrome. *Ann Intern Med* 1978;**89**:888–92.
- Voulgarelis M**, Dafni UG, Isenberg DA, Moutsopoulos HM. Malignant lymphoma in primary Sjogren's syndrome. *Arthritis Rheum* 1999;**42**:1765–2.
- Foster H**, Stephenson A, Walker D, Cavanagh G, Kelly G, Griffiths I. Linkage studies of HLA and primary Sjogren's syndrome in multicase families. *Arthritis Rheum* 1993;**36**:473–84.
- Vitali C**, Bombardieri S, Moutsopoulos HM, et al. Preliminary classification criteria for Sjogren's syndrome. Results of a prospective concerted action supported by the European Community. *Arthritis Rheum* 1993;**36**:340–7.