LETTERS TO THE EDITOR

Atlantoaxial Subluxation in an Ankylosing Spondylitis Patient with Cervical Spine Ossification

Sir—Cervical subluxations can occur in ankylosing spondylitis (AS). We report here an anterior atlantoaxial subluxation (AAS) with ligamentous calcification and a previously unobserved ossification on the dens in an AS patient.

A 42-yr-old White man was referred to our department for neck pain. He had no pertinent medical history. He suffered from mild neck pain for 6 months without any prior trauma. He next developed inflammatory neck pain. Physical examination revealed limitation of cervical spine mobility without neurological disturbance. The patient had no psoriasis. He also complained of low back pain and heel pain responding to non-steroidal anti-inflammatory drugs, but he denied a history of peripheral arthritis. The laboratory data are remarkable for a mild inflammatory syndrome (erythrocyte sedimentation rate 30 mm/h) and positive HLA B27 antigen. Rheumatoid factors and antinuclear antibodies were negative.

Lateral radiographs of the cervical spine showed that the distance between the posterior margin of the anterior arch of C1 and the anterior margin of the odontoid was 10 mm. This diastasis was irreducible in flexion or extension of the neck (Fig. 1A and B). No calcification was observed in the disc or the ligaments of the cervical spine. There was no fracture of C1 and C2, and no syndesmophyte on the lower cervical spine. No transverse or rotatory subluxations were observed on open mouth view and anteroposterior projection of the cervical spine. Radiographs of the hands and wrists were normal. Radiographs of the dorsolumbar spine and pelvis showed a bilateral sacroiliitis with sclerosis and erosions on both sacroiliac joints, and syndesmophytes at the dorsolumbar junction. Magnetic resonance imaging (MRI) (Fig. 2) of the cervical spine also showed the diastasis with narrowing of the spinal canal, but without spinal cord compression. A soft-tissue mass was located between

![Fig. 1.—Lateral radiographs of the cervical spine in flexion (A) and extension (B): anterior atlantoaxial subluxation without reduction in extension. Motion from flexion to extension is very limited.](image-url)
F. 2.—Magnetic resonance imaging of the cervical spine (T1-weighted sequence): diastasis between the anterior arch of C1 and the odontoid process. Note the spinal space narrowing without neurological compression.

C1 and C2, and was assumed to be synovial pannus. Sagittal computed tomography (CT) of C1–C2 articulation (Fig. 3) revealed an erosion of the posterior surface of the odontoid at its junction with the body of C2. The atlantooccipital ligament was calcified. There was also ossification at the posterior margin of the anterior arch of C1 and at the anterior margin of the odontoid in front of the atlantodental joint. No surgical treatment was undertaken because of the normal neurological condition.

Cervical spine involvement in AS includes several lesions such as erosions, subluxations and ossification of ligaments [1].

Erosions in AS are most often described on the posterior margin of the dens [1]. Several types of subluxations are observed in AS [1, 2, 4, 5]—AAS, upward atlantoaxial subluxation, rotatory and transverse subluxations, backward subluxation—but AAS is the most frequent. Its frequency has been evaluated in a few studies as between 2 and 21% [3, 6], concerns male patients with longer disease duration. The mechanisms explaining atlantoaxial subluxations associate inflammatory lesions (atlantodental synovitis, erosions of the dens and adjacent ligaments) and physical stresses (kyphosis of the dorsal spine and weight of the head at the C1–C2 level).

New bone formation in AS consists of syndesmophytes and ligamentous calcifications, but the latter are curiously infrequent in the cervical spine as compared to the thoracic or lumbar spine [1]. They may be located at many parts of the cervical spine. In the case of atlantoaxial subluxation, bone tissue or calcification have not been previously observed at the level of the enlarged articular interspace [1]. We observed such an ossification in our patients,

Fig. 2.—Magnetic resonance imaging of the cervical spine (T1-weighted sequence): diastasis between the anterior arch of C1 and the odontoid process. Note the spinal space narrowing without neurological compression.

Fig. 3.—Sagittal computed tomography of the atlantodental joint: calcification of the atlantooccipital ligament (indicated with an asterisk), erosion of the posterior surface of the dens at its junction with the body of C2 and ossification (arrow) in front of the atlantodental joint.
which could be induced by the inflammatory process and/or correspond to a similar enthesopathic lesion.

Thus, an irreducible subluxation during neck motion in an AS patient can be explained by ossification near the atlantodental joint, as was observed in our patient, and will be better evidenced on CT.

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Undergraduate Education in Rheumatology

Sir—We are encouraged to see the issue of undergraduate education in rheumatology given prominence in the Journal. We concur with the views expressed by Steven and Lowry [1]. It is undeniably true that tomorrow’s doctors need more rheumatological knowledge and skills, and that rheumatology is an excellent vehicle for skills and attitudes which extend far beyond the confines of the specialty. We agree that communication skills and team work are prominent amongst these. In theory, at least, these aspects can be learned within a conventional undergraduate curriculum.

A more difficult, but no less important, issue is the appreciation of the implications of chronic disabling disease. As Brooks [2] points out, conventional undergraduate courses, even those which include relatively long attachments in rheumatology, can only provide a ‘snapshot’ of people and conditions at one point in time. This constraint even guides our teaching; it is easier to focus, in a ‘snapshot’, on the physical and interventional aspects rather than on a balanced view of both these and the social and emotional impact of an evolving and progressive disease. Whilst understanding is the province of the patient, a balanced appreciation and approach to chronic disease is an essential component of mature clinical practice and somehow the basis of it must be laid during the undergraduate course. A rational way to achieve this experience of chronic disabling illness is for students to undertake attachments with individuals who have chronic disabling diseases. Ideally, one or two students would visit a single patient at intervals throughout most, or all, of the undergraduate course. Contact between patient and student need not be frequent, but it should be structured and supervised, and could readily form the basis of a formally evaluated project. In this way, both the impact of the disease and the evolution of disability would gradually declare themselves to students who themselves are maturing and increasing in their ability to comprehend them. Such approaches appear to have been successful at some medical schools, whilst at others the effort, or perhaps the risk, of stepping away from the conventional approach seems to be too great. The General Medical Council [3] has quite explicitly invited medical schools to tackle issues such as this but, as we all know, the forces opposing change, not least the heavy clinical and administrative workload of NHS consultants, are in danger of stifling progress. Nevertheless, can there really be any doubt that this particular job is well worth doing?

What is true for the undergraduate also holds true for the graduate specialty trainee. Changes to speciality graduate training programmes envisaged by Calman [4] pose serious threats, but also hold out opportunities in this regard. It is likely that many training registrar posts will rotate annually from one hospital/post to another. Thus, they will effectively rotate from one group of patients with chronic disease to another. Even ‘snapshots’ of 12 months in length can hardly suffice to prepare doctors to guide the management of patients over decades or more. Thus, flexible training rotations must be devised that allow trainees to sample the delights of different institutions and trainers whilst remaining involved with the care of some patients over a minimum of 2 yr and ideally four or five.

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Respiratory Failure in Systemic Lupus Erythematous: Decisive Differentiation Between Acute Pneumonitis and Infection

Sir—We report the case of a 23-yr-old nurse with acute respiratory failure requiring intubation and mechanical ventilation. For 4 yr, she had been suffering from seropositive SLE. Two weeks before admission, lupus pneumonitis had been suspected on the basis of non-productive cough, fever and a rise of ANA titres.


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TABLE I

<table>
<thead>
<tr>
<th>Laboratory findings at admission. Electrolytes, other liver as well as pancreatic enzymes were within the normal range</th>
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<tbody>
<tr>
<td>White blood cells</td>
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<td>Red blood cells</td>
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<tr>
<td>Haemoglobin</td>
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<td>Platelet count</td>
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<td>Creatinine</td>
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<td>Blood urea nitrogen</td>
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<td>Bilirubin</td>
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<td>Total protein</td>
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<td>Cholinesterase</td>
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<td>Lactate dehydrogenase (LDH)</td>
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<td>Lactate</td>
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<td>Prothrombin time</td>
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<tr>
<td>C-reactive protein</td>
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<tr>
<td>Antinuclear antibodies</td>
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<td>Antibodies against double-stranded DNA</td>
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Histologically, single groups of macrophages and erythrocyte extravasates could be found in the lung parenchyma. The alveolar wall was somewhat fibrotic and infiltrated with mononuclear cells. There were no morphological signs of viral infection, bacteria, fungi or *P. carinii*. IgM immune staining revealed granular deposits of IgM in vessel and alveoli walls (Fig. 2).

Having ascertained the diagnosis ‘acute lupus pneumonitis’, treatment consisted of corticosteroids (2 g prednisolone on days 1, 2 and 3, 100 mg from day 4 to day 7, followed by a stepwise dose reduction to 25 mg/day), three cycles of plasmapheresis on days 3, 4 and 5 (substitution with human albumin), followed by 1 g of cyclophosphamide after the last cycle. Thirty grams of human immunoglobulins were administered after the last cycle. Antibiotic therapy was continued despite negative infection signs and cultures. Three days after admission, the patient could be extubated and was transferred to a general ward on day 6. The chest radiograph normalized.

Currently, more than 1 yr later, the patient suffers from minor proteinuria without the need for immunosuppression. ANA titres are in the normal range (1:20).

Acute lupus pneumonitis is a rare but severe complication of SLE [1]. However, owing to defective host defences and immunosuppressive therapy, infection is a common problem during the course of the disease, causing up to 33% of SLE-associated deaths [2, 3].

In our patient, all ‘acutely’ available diagnostic tools did not lead to a proven diagnosis. Transbronchial biopsy may be unsuccessful due to an insufficient amount of tissue and can lead to severe complications [4]. Open lung biopsy offers the possibility of a reliable histological evaluation with a low perioperative risk and should be the preferred diagnostic tool, especially when time is a decisive factor [5].

Although controlled studies are missing, corticosteroids are the mainstay of therapy in lupus pneumonia. In our patient, treatment with 200 mg cyclophosphamide, 40 mg methylprednisolone and 200 mg cyclosporine daily together with antibiotics had been initiated in a peripheral hospital. The patient was transferred to our intensive care unit.

Auscultation of the lungs revealed dry rales bilaterally, tachycardia with a frequency of 110 beats/min with no pathological extra sounds or murmurs was present. Blood pressure was 155/90 mmHg and oral temperature 38.9°C. Abnormal laboratory findings are listed in Table I. Within hours, the patient required invasive ventilation with an FiO₂ of 1.0 (Fig. 1). Gram staining of a bronchoalveolar lavage revealed no bacterial overgrowth, *Pneumocystis carinii* could not be isolated, cytomegalovirus immediate early antigen was negative. Clinically and radiologically, differentiating between lupus pneumonitis and pneumonia was not possible. Initiating massive immunosuppressive therapy could have been fatal in the case of infection; therefore, an open lung biopsy was performed.

![Fig. 1.—Chest radiograph at admission: intubated patient, central venous catheters via right subclavian and internal jugular vein. Extended patchy alveolar shadowing in both lungs, increasing from cranial to caudal with positive air bronchogram. Additional bilateral pleural effusions. Enlarged heart and possibly pericardial effusion. (Median sternotomy had been performed 3 yr earlier for thymectomy).](image-url)
pneumonitis [1, 6]. The role of plasmapheresis is less defined, although some positive case reports exist [7–9]. Synchronization with pulse cyclophosphamide has been proposed [10]. We decided not to withdraw prednisolone prior to and during the plasmapheresis period due to the patient’s severe impairment. High-dose cyclophosphamide was administered only once, because of the cyclophosphamide pre-treatment. In immunocompromised patients with cryptogenic respiratory insufficiency, time is a crucial factor and one should not hesitate to use early open lung biopsy to establish a timely diagnosis.

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Systemic Lupus Erythematosus Complicating Hyper IgE Syndrome

Sir—Connective tissue diseases such as systemic lupus erythematosus (SLE) can be associated with immunodeficiency, most notably complement component or IgA deficiency. We report a case of SLE complicating the course of an 11-yr-old boy with the hyper IgE syndrome (HIE). HIE is characterized by IgE levels >2000 IU/l, non-atopic dermatitis and eosinophilia. Impaired cellular immunity and poor antibody responses result in recurrent, often deep-seated infections with organisms such as Candida, Staphylococcus aureus, Streptococcus pneumoniae and Haemophilus influenzae [1].

An Asian male was diagnosed as having HIE aged 6 months on the basis of atypical eczema and persistently raised IgE levels and eosinophilia. Specific IgE to common allergens such as milk and house dust mite were not detected. Until the age of 11 yr, he suffered from persistent dermatitis, recurrent otitis media and externa, Candida paronychia and angular cheilitis, oral Candida, blepharitis and perianal abscesses. He also suffered from varicella–zoster when aged 10 yr. Treatment consisted of antimicrobial agents as appropriate, prophylactic co-trimoxazole and three surgical procedures to drain the perianal abscesses. He was anergic to intradermal testing with recall antigens and no significant abnormality of lymphocyte subsets or function was demonstrated. Serum total immunoglobulin levels were normal with the exception of IgE (>2000 IU/l), but specific IgG levels to pneumococcus and H. influenzae type b were low. The patient was immunized with Act-Hib and, 3 weeks later, with Pneumovax II. Protective levels of IgG to Hib were produced, but the IgG response to Pneumovax was poor.

Three weeks following the immunization with Pneumovax II, the patient presented with a pyrexia of 38.9°C, a non-productive cough and was found to have Candida paronychia of most of his toes, group B Streptococcus present on the external auditory meatus, a worsening of his facial rash, and a severe vasculitis present on the soles of both feet and at the site of the Pneumovax immunization. A list of results from investigations performed during the admission are presented in Table I. The fever and anaemia responded to prednisolone (30 mg/day), but azathioprine (75 mg/day) was required to obtain an improvement in the cutaneous vasculitis. Only minimal arthralgia had been
Bilateral Hip Replacements in a Man with Cystic Fibrosis and Episodic Arthritis

Sit—An association of cystic fibrosis with an inflammatory arthritis or hypertrophic pulmonary osteoarthropathy is recognized, although the pathogenic mechanisms are uncertain [1, 2]. We report a patient with an episodic arthritis who developed severe bilateral hip disease at the age of 30, resulting in hip replacements. To our knowledge, the association between cystic fibrosis and severe hip disease has not been reported previously.

A 36-yr-old man with cystic fibrosis presented at the age of 9 yr in 1968 with steatorrhea, erythema nodosum and synovitis of his knees and ankles. He had minimal respiratory symptoms and signs at presentation. Cystic fibrosis was diagnosed by chloride sweat testing. Rheumatoid factor was negative. During the next 17 yr, he developed increasing respiratory symptoms with recurrent infections and episodes of polyarthritis which tended to occur concurrently with his chest infections. The episodes of arthritis typically lasted from a few days to several weeks with symptom-free periods between attacks. The joints involved, in order of frequency, included his MCP joints, wrists, hips, knees, ankles and MTP joints. He was treated with short courses of oral corticosteroids, non-steroidal anti-inflammatories, joint aspiration of his knees and intra-articular corticosteroids. In early adulthood, he was fit enough to work as an agricultural labourer.

In 1985, his health deteriorated with a duodenal ulcer, respiratory infections requiring hospital admission, and an increase in the frequency and persistence of arthritis. He was forced to stop working. In 1988, he developed a severe haemoptysis requiring a four unit blood transfusion and was treated successfully with bronchial artery embolization. He developed increasingly severe bilateral hip pain and loss of function, and became wheelchair bound for 2 months prior to bilateral Charnley hip replacements in 1990. The pre-operative pelvic radiograph showed bilateral joint space narrowing, subchondral sclerosis and periarticular cysts. The sacroiliac joints were normal. In 1991, pulmonary tuberculosis was diagnosed and treated. He also developed chronic meconium ileus equivalent, a common gastrointestinal disorder in patients with cystic fibrosis in which the bowel becomes distended with faecal masses which cannot be expelled [3]. His arthritis became more frequent with no symptom-free periods. He was prescribed mycocrin in 1993 with a reduction in the severity and frequency of the episodes of arthritis.

Currently, the patient continues to have low-grade synovitis with swelling of the wrists and MCP joints bilaterally and knee effusions. He has mild clubbing of his finger nails, but has no other evidence of hypertrophic pulmonary osteoarthropathy (HPOA) clinically or radiographically. Joint function is well maintained despite mild ulnar deviation of the MCP joints. Hand and foot radiographs reveal joint space narrowing in his wrists, MCP joints and first MTP joints. Periarticular osteoporosis and periarticular bone cysts are prominent features. His respiratory illness is complicated by chronic Pseudomonas and Staphylococcus aureus infections. Blood tests show persistent elevation of his inflammatory markers: viscosity 1.95 (normal <1.72), CRP 0.064. Alkaline phosphatase is noted by the patient and a mild restrictive defect was detected on pulmonary function testing. There was never any evidence of impaired renal or central nervous system function.

The authors are aware of another definite [2] and a third possible [3] case of SLE complicating HIE which is insufficient to determine whether this is a chance association or if there is an association between SLE and HIE. Explanations for an association could be that the increased number of infections increases the likelihood of cross-reactions occurring between foreign and self-antigens or the impaired antibody responses seen in HIE generate insoluble immune complexes. In the case reported here, a third possibility is that the pneumococcal immunization generated an Arthus-type reaction which precipitated an episode of SLE.

TABLE I

<table>
<thead>
<tr>
<th>Investigation</th>
<th>Result</th>
<th>Normal range</th>
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<tbody>
<tr>
<td>Hb</td>
<td>8.3 g/dl</td>
<td>13.5–18.0</td>
</tr>
<tr>
<td>Neutrophils (10⁹/l)</td>
<td>2.0 × 10⁹/l</td>
<td>2.0–7.5</td>
</tr>
<tr>
<td>Lymphocytes (10⁹/l)</td>
<td>1.0 × 10⁹/l</td>
<td>1.5–4.0</td>
</tr>
<tr>
<td>Reticulocytes (10⁹/l)</td>
<td>3.0 × 10⁹/l</td>
<td>0–2</td>
</tr>
<tr>
<td>Haptoglobulin</td>
<td>&lt; 0.2 g/l</td>
<td>0.6–1.6</td>
</tr>
<tr>
<td>PV</td>
<td>1.74</td>
<td>1.5–1.72</td>
</tr>
<tr>
<td>Direct Coombs test</td>
<td>Positive</td>
<td></td>
</tr>
<tr>
<td>ANA (1:1000)</td>
<td>speckled</td>
<td>&lt; 1:16</td>
</tr>
<tr>
<td>DNA antibody</td>
<td>&gt; 300 IU/l</td>
<td>&lt; 30</td>
</tr>
<tr>
<td>Cardiolipin antibody</td>
<td>34 GPLU/ml</td>
<td>&lt; 14</td>
</tr>
<tr>
<td>ENA antibody</td>
<td>Weak positive*</td>
<td></td>
</tr>
<tr>
<td>IgG</td>
<td>16.0 g/l</td>
<td>5.4–16.1</td>
</tr>
<tr>
<td>IgA</td>
<td>3.0 g/l</td>
<td>0.8–2.8</td>
</tr>
<tr>
<td>IgM</td>
<td>8.0 g/l</td>
<td>0.5–2.0</td>
</tr>
<tr>
<td>IgE</td>
<td>4835 kU/l</td>
<td>&lt; 116</td>
</tr>
<tr>
<td>C3</td>
<td>1.39 g/l</td>
<td>0.85–1.98</td>
</tr>
<tr>
<td>C4</td>
<td>0.51 g/l</td>
<td>0.20–0.80</td>
</tr>
<tr>
<td>C-reactive protein</td>
<td>&lt; 1 mg/dl</td>
<td>&lt; 1</td>
</tr>
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</table>

*The antibody was shown to be distinct from Ro, La, Sm, RNP, Scl-70 or Jo-1 by counter-current immunoelectrophoresis against calf spleen and rabbit thymus extracts.

Direct immunofluorescence and histology of skin biopsies from the face and feet demonstrated changes consistent with SLE.

The authors are aware of another definite [2] and a third possible [3] case of SLE complicating HIE which is insufficient to determine whether this is a chance association or if there is an association between SLE and HIE. Explanations for an association could be that the increased number of infections increases the likelihood of cross-reactions occurring between foreign and self-antigens or the impaired antibody responses seen in HIE generate insoluble immune complexes. In the case reported here, a third possibility is that the pneumococcal immunization generated an Arthus-type reaction which precipitated an episode of SLE.

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raised (280, normal <92), but his bilirubin, alanine transaminase, calcium and phosphate are all normal. He has no autoantibodies relevant to joint disease and his HLA typing is HLA-A2,11; B21, 35/53; DR17(3), 24 (6); DQ 2, 5 (1).

An episodic arthritis associated with cystic fibrosis in children was first described in 1979 [1] and there have since been several multicase reports in the rheumatology literature [4–7]. Rush et al. [4] reviewed 533 patients attending a cystic fibrosis clinic and found 10 patients with an episodic arthritis, two with an erosive polyarthritis and positive rheumatoid factor, and 11 with HPOA. Four patients in the episodic group progressed, as in our case. Two patients had hip symptoms, but both had normal radiographs.

Our patient has several features of note: a temporal association between respiratory and joint symptoms, destructive hip disease, symmetrical joint involvement and subchondral cysts radiographically. A number of factors may be implicated in the aetiology of these features. First, acute-on-chronic respiratory infection may trigger the generation of immune complexes implicated in the pathogenesis [7, 8]. This may be analogous to the association of bronchiectasis with rheumatoid arthritis. Similarly, the bowel stasis of meconium ileus equivalent may result in bacterial overgrowth that causes the generation of immune complexes or serves as a source for molecular mimicry. Second, poor nutritional status may be implicated in the development of bone cysts, although specific nutritional deficiencies such as vitamin D deficiency, have not been documented in this patient. Third, developmental abnormalities of the hips, coupled with heavy physical work as an agricultural labourer, may be factors in causing our patient’s destructive hip disease. Finally, long-term antibiotic therapy and intermittent prednisolone therapy may also be implicated in bone remodelling. As the survival of patients with cystic arthritis increases to a median of 30 yr in some centres [9], progressive arthritis in adults may be more frequently seen in rheumatological practice.

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volume would be to express GAG in relation to a protein which is diluted by effusion volume. Accurate ways of measuring effusion volume, such as using endogenous albumin dilution after intra-articular injection of a known volume of normal saline [6], radioactive sodium [7], or the use of a magnetic resonance imaging scan [8], are possible in a research context. An alternative method that could be more informative than a pure concentration value would be to express two similar entities as a ratio [3], such as keratan sulphate to chondroitin sulphate (two similarly sized molecules of the same origin), which would reflect the relative amounts present.

In conclusion, GAG measurements are not affected by SF volume or needle bore size. Accordingly, for arthrocentesis, the needle size most appropriate for withdrawing SF with minimal discomfort to the patient should be used.

This work was supported by the Arthritis and Rheumatism Council.

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Polyarthritis Associated with Hepatitis B Vaccination

Sir—Many side-effects have been reported after vaccinations. They included fever, skin lesions (erythema nodosum, rash), uveitis, glomerulonephritis, arthralgias and polyarthritis. These manifestations are considered to be due to lesions of the vessel walls caused by circulating immune complexes. Side-effects of hepatitis B vaccination have rarely been reported [1–4]. We describe a patient who developed polyarthritis after the second dose of hepatitis B vaccine.

AG, a healthy 9-yr-old boy, was given the first dose of Engerix B vaccine in October 1994 and the second dose in December 1994. The patient, who weighed 42 kg, was given adult doses. Three weeks later, he developed polyarthritis involving the ankles, joints of the hands and feet, wrists, shoulders and hips. Fever to 40°C in the evening, and fatigue, were also present. One week later, lymphadenopathy appeared. Laboratory investigations showed: ESR of 128 mm/h, CRP of 104 mg/l (normal < 6 mg/l), alpha-2 globulins of 21.4% (normal < 12%), mild hypochronic anaemia (haemoglobin 10.5 g/dl), IgA of 450.4 mg/dl (normal 90–410), IgG of 1807 mg/dl (normal 800–1600), IgM of 322 mg/dl (normal 50–150), SGOT was 81 U/ml (normal < 40) and SGPT 181 U/ml (normal < 40).

Values for ANA and the titre of antistreptolysin O were normal. The results of biochemical tests were within normal limits. Serological tests for common organisms were negative. The boy was admitted to a paediatric hospital and juvenile chronic arthritis was diagnosed. Non-steroidal anti-inflammatory drug (NSAID) proved effective and arthritis and fever disappeared.

The patient presented to us 3 months later; all laboratory investigations showed normal values and test for rheumatoid factor was negative. The remission has lasted for 18 months and all laboratory investigations are still normal.

We think that an aetiological link is present between the appearance of arthritis and hepatitis B vaccination in our patient. In fact, the arthritis was similar to that of acute viral hepatitis with an analogous temporal
relationship between the development of articular symptoms and vaccination.

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Should Patients Presenting with Dry Eyes be Screened for Autoimmune Rheumatic Disorders?

Sir—Dry eyes are a common complaint, especially in the elderly, but may be caused by autoimmune processes and lymphocyte infiltration (e.g. Sjögren’s syndrome), granuloma (e.g. sarcoidosis) or damage by iron deposits (e.g. haemochromatosis of haem siderosis) [1, 2].

I wished to study whether it was worth screening patients presenting with dry eyes for the presence of either primary Sjögren’s syndrome (primary SS) or other autoimmune rheumatic disorders (AIRDs).

Fifty patients (17 male, 33 female) with dry eyes presenting to an ophthalmology clinic were screened for primary SS by means of a previously validated questionnaire [3] and for other rheumatic disorders by history and examination. Patients with known AIRDs were excluded. The questionnaire was also administered to 30 patients with known primary SS based on recognized criteria [1, 2, 4]. A mean score of 10.85 ± 2.89 s.d. was highly predictive of primary SS with specificity and sensitivity values of 98 and 95%, respectively [3].

All the patients had either an abnormal Schirmer’s test (<5 mm/5 min) and/or positive Rose Bengal staining, thus exhibiting features of significantly dry eyes [1, 2]. The study population (Table I) could be separated into two groups: group A having no evidence to suggest an AIRD and group B showing one or more minor abnormalities, such as raised erythrocyte sedimentation rate, serum immunoglobulins, antinuclear factor or rheumatoid factor titres. Interestingly the scores for primary SS patients were higher (P < 0.001) in group B (mean = 8.7) compared to group A (mean = 4.6) as in the case of group C with proven primary SS patients (mean = 10.9). Although none of the patients had any clinical or laboratory features to meet the required diagnostic criteria for either primary SS or any of the AIRDs at the time of assessment, I could not confidently exclude these disorders in the smaller group of five patients (group B) without resorting to further invasive tests such as lower lip biopsy [1, 2, 4]. This group may be harbouring subclinical disease and further follow-up may be necessary.

The screening questionnaire used in this study is simple, easily understood by patients and takes only a few minutes to complete. It is clear from this study that not all dry eye patients require full rheumatology assessment and certainly not the full set of serological tests described in this study. The most cost-effective way may be to use this screening questionnaire and select patients with a higher score for appropriate referrals. General practitioners and ophthalmologists especially may find this helpful to identify suitable patients for further assessment.

I would like to thank Lawrence Morgan, Consultant Ophthalmic Surgeon, Stepping Hill Hospital, Stockport, and Alison Webb, Medical Secretary, Wythenshawe Hospital, Manchester.

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