SIR, We read with interest the Editorial by Chambers et al. [1] regarding treatment adherence in systemic lupus erythematosus (SLE). It has been estimated that up to 50% of patients who suffer from chronic diseases do not adhere to treatment recommendations [2]. We have recently reviewed our experience of compliance with immunosuppressive treatment amongst patients with connective tissue diseases (CTDs) at The Royal London Hospital. We used a physician-designed questionnaire, which was distributed to consecutive patients attending the CTD clinic. The questionnaires were filled anonymously. Data collected included age, sex, ethnicity, immunosuppressant prescribed, clinic attendance and degree and reasons for non-adherence. Our catchment area in East London has a large ethnic-minority group from the Asian subcontinent.

One hundred questionnaires were given. All were filled but 10 were subsequently discarded as eight patients were on no immunosuppressants and two had a diagnosis of rheumatoid arthritis. Ninety questionnaires were analysed. Seventy per cent of patients had SLE. The remainder had other CTDs including systemic lupus erythematosus. Arthritis 2005;52:865–76.

Table 1. Ethnicity distribution and adherence to immunosuppressants

<table>
<thead>
<tr>
<th>Ethnic background</th>
<th>Number (%) of patients</th>
<th>Number (%) with poor adherence</th>
</tr>
</thead>
<tbody>
<tr>
<td>White Caucasian</td>
<td>37 (41)</td>
<td>15 (40.50)</td>
</tr>
<tr>
<td>Afro-Caribbean</td>
<td>27 (30)</td>
<td>12 (44.50)</td>
</tr>
<tr>
<td>Asian</td>
<td>19 (21)</td>
<td>12 (63)</td>
</tr>
<tr>
<td>Oriental</td>
<td>3 (3)</td>
<td>3 (100)</td>
</tr>
<tr>
<td>Others</td>
<td>4 (4)</td>
<td>1 (25)</td>
</tr>
</tbody>
</table>

Amongst non-English-speaking patients; thus Asians and Orientals reported missing tablets far more frequently compared with English-speaking Afro-Caribbean and Caucasian patients (Table 1). Analysis of poor compliance according to drug taken showed patients were least compliant with HCQ (53.5%) followed by AZA (47%), steroids (42%) and MMF (30%). Non-compliance was not reported with other immunosuppressants. Patients taking only HCQ tended to have the mildest disease and this may have been a factor in their lack of adherence. The main reasons given for not taking medication were: forgetting (70%), side-effects (14%), concerns about side-effects (14%), feeling well (5%), prescription problems (5%) and not enough explanation (5%). Our data highlight the problem of treatment adherence to long-term medications in patients with CTDs. We would reinforce the call from Chambers et al. to engage in measures aimed at improving compliance amongst this group of patients.

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Specialist interventions at the rheumatology outpatient clinic

SIR, As rheumatologists we are under significant pressure to reduce the waiting times for new patients in the outpatient clinic. As there is restricted clinic time available for new and review patients, it has been suggested that 'stable' review patients should be discharged to primary care. We believe there exists a perception that review patients with rheumatic disease require little intervention or adjustment to treatment and do not need to continue to re-attend the outpatient clinic on a long-term basis. We collected data on specialist interventions required by patients with chronic
rheumatic disease over a 6-week period on 253 consecutive patients who had been attending the rheumatology outpatient clinic for >12 months. A record was made of their diagnosis, current disease-modifying-drugs (DMARDs) and all interventions that had been undertaken in the clinic in the preceding 12 months. Anti-rheumatic ethical approval and informed patient consent was not required for this case-review-based study as confirmed by the trust research office.

The majority of patients had a chronic inflammatory arthritis; the largest subgroup being rheumatoid arthritis (47%) with psoriatic arthritis the next biggest group (14%), followed by osteoarthritis (13%), systemic lupus erythematosus (6%), ankylosing spondylitis (4.5%), seronegative arthritis (3%), polymyalgia rheumatica (2%), fibromyalgia syndrome (1.5%), juvenile idiopathic arthritis (1%) and others (8%).

In the previous 12 months, 90% of patients had required at least one specialist intervention including the following:

- Thirty percent of the patients had received an intra-articular or soft-tissue steroid injection.
- Thirty percent of the patients had required a dose adjustment or change of DMARD.
- Twenty-six percent had a change or new prescription of an analgesic or anti-inflammatory drug.
- Thirty-two percent of patients had a radiological investigation, five percent had diagnostic musculoskeletal ultrasound performed at the clinic.
- Twenty percent were referred to another member of the multidisciplinary team or medical specialty.
- Other interventions included adjustment of oral steroids (11%), adjustment of other rheumatological treatments (10%), blood testing other than for routine monitoring (11%), pulse of methylprednisolone (10%), admission to day ward or inpatient ward (10%).

Only 25 patients (10%) had no interventions in the preceding 12 months. Ninety percent of patients had one intervention, 31% had two, 23% had three, 10% had four, 4% had five, 2% had six and one patient (<1%) had seven interventions. The median number of investigations per patient was 2, with a mean of 2.07 interventions per patient. Of the 25 patients who had no intervention, 22 had a chronic inflammatory arthritis, and 15 were maintained on DMARD therapy.

This study was conducted in a general rheumatology outpatient clinic in a teaching hospital. The staff involved were Consultant, Specialist Registrar (SpR), Senior House Officer (SHO) and Nurse Specialist. The review patients are largely seen every 6–8 months during a 15-minute appointment. There were no complex connective tissue disease/vasculitis patients and few patients on biological therapies as they are seen in separate clinics. The patients at this clinic could therefore be considered to be a ‘stable’, less ill population and yet 90% of them required a specialist intervention in a 12 month period, 64% requiring between two and four interventions. Even the majority of those who had no intervention had chronic inflammatory disease and most were on DMARDs.

These results suggest that very few of our review patients could be discharged without the need for further ongoing care. In this practice, the consultant already receives on average 2–3 referrals per week as well as 3–4 letters or phone calls per week from GPs seeking advice.

It appears that those involved in these management decisions consider the surgical ‘one stop shop’ model of care to be appropriate, suggesting a fundamental lack of understanding of the management of chronic disease.

In the era of biological therapies, early aggressive treatment, ‘tight control’ of inflammatory disease [1], cardiovascular risk assessment, etc., we are now able to influence the natural history of rheumatic disease more than ever. It is likely, therefore, that patients will require more frequent review by trained specialists rather than less.

In our view, the care of review patients cannot be sacrificed to free up clinic time for new patients who may in turn become review patients. This study has highlighted the need for long-term specialist care for all patients with chronic rheumatological disease.

**Rheumatology key message**

- Patients with chronic rheumatological disease require long-term specialist review.

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**Mevalonate kinase deficiency syndrome with structural damage responsive to anakinra**

Sir, mevalonate kinase deficiency (MKD) syndrome is an autosomal recessive autoinflammatory disorder caused by mutations in the MVK gene. We describe the first case of MKD syndrome with structural damage successfully treated with anakinra.

A 32-year-old French Caucasian woman experienced relapsing flares of progressively destructive bilateral and symmetrical polyarthritis involving small and large joints with morning stiffness (lasting 2–4 h). Synovial fluid, drained several times, was inflammatory and contained an average of 30 000 leucocytes/mm³ (98% neutrophils), without crystals or bacteria. X-rays detected joint-space narrowing and erosions in several joints (Fig. 1). She had three or four polyarthritis attacks/year, each lasting 2–3 weeks, associated with fever (38–39°C), chills, fatigue, headaches, maculopapular rash, diarrhoea, vomiting with severe abdominal pain (macroscopically inflammatory peritonitis without bacteria), splenomegaly, bronchopneumopathy, elevated ESR (40–100 mm/1st h) and high CRP (50–200 mg/l). Since childhood, no autoantibodies (rheumatoid factor, antinuclear antibodies, anti-citrullinated antibodies) have ever been detected.

The extra-articular symptoms started at 3 months (1974) with flares occurring at irregular intervals; atypical systemic juvenile idiopathic arthritis (sJIA) was diagnosed, but was considered in 2004 when laboratory investigations showed high serum immunoglobulin D (IgD) [166.4 mg/l (28.2× normal range <141 mg/l)], elevated mevalonacida (19.2 mmol/mol of creatinine (normal range <1.3 mmol/mol)) and low mevalonate kinase activity in lymphocytes [0.7 μkat/kg of protein (2.5× normal range <7.8 μkat/kg)]. Sequencing of the 10 MVK-gene exons detected a four-base deletion (c.475-478 delACTG) and an