

Dr. Baer, *et al* reply

To the Editor:

We thank Drs. Prabhakaran and Handler for their interest in our report¹ and their important observations. “Sjrupus” is certainly a novel disease name, but even more difficult to pronounce than “Sjögren’s”! The name accurately reflects the frequent overlap of connective tissue diseases. In our opinion, their Patients 1 and 3 would be best classified as having systemic lupus erythematosus (SLE) with secondary Sjögren’s, and Patient 2 as having undifferentiated connective tissue disease. The latter patient may have primary Sjögren’s syndrome, but this diagnosis would require that the patient have one or more objective measures of keratoconjunctivitis sicca (e.g., Schirmer’s test, conjunctival and/or corneal staining with rose Bengal or lissamine green) or salivary hypofunction (e.g., sialometry, sialography, or parotid scintigraphy) in addition to demonstration of focal lymphocytic sialoadenitis (with a focus score ≥ 1) on a minor labial gland biopsy. It is uncommon for these additional, but necessary, diagnostic steps to be carried out in routine practice, limiting the utility of the current diagnostic criteria for Sjögren’s syndrome². Patients 1 and 3 illustrate the occasional acceleration of autoimmune disease that can occur in the context of drug therapy (i.e., estrogen for Patient 1 and tumor necrosis factor antagonist for Patient 3).

The primary indication for rituximab treatment in these 3 patients was polyarthritis, refractory to a variety of other immunosuppressive treatments. The clinical benefit of rituximab in these patients is noteworthy and is in accord with the emerging experience of other investigators who have been treating SLE patients with rituximab for a variety of different disease manifestations. Of note, Garcia-Carrasco, *et al* observed remission of refractory polyarthritis in 19/25 patients with SLE treated with rituximab. It was not stated whether these patients had sicca symptoms or signs³.

In the placebo-controlled trial of rituximab in Sjögren’s syndrome recently reported by Meijer, *et al*, the primary outcome of a significant improvement in stimulated whole saliva flow rate was achieved at 12 weeks, but not at the end of the study⁴. This trial included only patients

with residual salivary secretory capacity, since earlier trials failed to demonstrate improvement in salivary function in patients with advanced disease. It would thus be of interest to know if Drs. Prabhakaran and Handler made similar observations in their rituximab treatment of 22 patients with Sjögren’s syndrome.

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