EULAR Issues Behçet Disease Recommendations

BY MIRIAM E. TUCKER
Senior Writer

Eye involvement is a core element of new recommendations on the management of Behçet disease from the European League Against Rheumatism.

The recommendations were developed by a EULAR task force of the Standing Committee for Clinical Affairs, which included nine rheumatologists, three ophthalmologists, one internist, one dermatologist, and one neurologist. Members were from six European countries plus Tunisia and Korea. A patient representative also participated. The lead author was Dr. Gulen Hatemi, of the division of rheumatology at the Cerrahpasa Medical School, Istanbul (Turkey) University.

Among the recommendations:

► Any patient with BD and inflammatory eye disease affecting the posterior segment should be on a treatment regime that includes azathioprine and systemic corticosteroids. Azathioprine is widely accepted as the initial agent for ocular involvement. Local and systemic corticosteroids rapidly suppress the inflammation but potential side effects, including cataracts and glaucoma, cause concern.

► Patients with severe eye disease, defined as more than a drop of more than two lines in visual acuity on a 10/10 scale and/or retinal disease (retinal vasculitis or macular involvement) should receive either cyclosporine A or infliximab in combination with azathioprine and corticosteroids. Alternatively, interferon-α (INF-α) with or without corticosteroids could be used.

Each of the nine recommendations was voted on both by the group as a whole and by individual members who were experts in each field. While recommendations related to the eye, skin/mucosa, and arthritis were mainly evidence based, those pertaining to vascular disease, neurologic, and gastrointestinal involvement were based largely on expert opinion and uncontrolled evidence from open trials and observational studies.

"The need for further properly designed controlled clinical trials is apparent," Dr. Hatemi wrote in the December issue of Annals of the Rheumatic Diseases (Ann. Rheum. Dis. 2008;67:1656-62).

Further recommendations held:

► There is no firm evidence to guide the management of major vessel disease in BD. For acute deep vein thrombosis in BD, agents such as corticosteroids, azathioprine, cyclophosphamide, or cyclosporine A are recommended. Cyclophosphamide and corticosteroids are recommended for the management of pulmonary and peripheral arterial aneurysms.

► Similarly, there are no controlled data—and no evidence of benefit from uncontrolled experience—with anticoagulants, antiplatelet, or antifibrinolytic agents in the management of deep vein thrombosis or for the use of anticoagulation for the arterial lesions of BD.

► There is no evidence-based treatment that can be recommended for the management of gastrointestinal involvement of BD. Agents such as sulfasalazine, corticosteroids, azathioprine, tumor necrosis factor–α (TNF-α) antagonists, and thalidomide should be tried first before moving to surgery, except in emergencies.

► Arthritis can be managed with colchicine in most BD patients.

► There are no controlled data to guide the management of central nervous system involvement in BD. For parenchymal involvement, agents to be tried may include corticosteroids, INF-α, azathioprine, cyclophosphamide, methotrexate, and TNF-α antagonists. For dural sinus thrombosis, corticosteroids are recommended.

► Cyclosporine A should not be used in BD patients with central nervous system involvement unless necessary for intraocular inflammation.

► The decision to treat skin and mucosa involvement will depend on the perceived severity by the physician and the patient. Mucocutaneous involvement should be treated according to the dominant or codominant lesions present:

Topical measures, such as local corticosteroids, should be the first line of treatment for isolated oral and genital ulcers.

Acne-like lesions are usually of cosmetic concern only. Thus, topical measures as used in acne vulgaris are sufficient.

Colchicine should be preferred when the dominant lesion is erythema nodosum. Leg ulcers in BD might have different causes. Azathioprine, INF-α, and TNF-α antagonists may be considered in resistant cases.

The work was financially supported by EULAR, with no competing interests. ■