New Behçet's Management Guidelines Issued

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Nine new recommendations for the management of Behçet's disease have been issued by the European League Against Rheumatism, based on a literature review from 1966 through 2006.

Guidelines relating to the oral, dermatologic, ocular, and joint manifestations of Behçet's disease (BD) were mostly evidence based, but recommendations on BD-associated vascular, neurologic, and gastrointestinal problems were "mainly based on observational studies, retrospective analyses, and clinical experience of the experts" (Ann. Rheum. Dis. 2008 Jan. 31 [doi:10.1136/ard.2007.080432]). The guidelines are applicable to many different specialties; the authors hailed from rheumatology, ophthalmology, internal medicine, dermatology, and neurology.

Dr. Yusuf Yazici, who was not on the task force, said in an interview the lack of randomized clinical trials can be explained by the fact that "these are rare manifestations and hard to recruit for, and also vascular and neurologic involvement can be life threatening, and [it's] hard to do a RCT in that situation." Dr. Yazici is the director of the Behçet's Syndrome Evaluation, Treatment and Research Center at the New York University Hospital for Joint Diseases. His father, Dr. Hasan Yazici, was one of the report's authors.

Dr. Yusuf Yazici added there are "no good numbers" to describe the prevalence of the disease in the U.S. BD affects between 1 and 6 people per 100,000, but "these are old numbers; no recent numbers are available," he said.

The nine recommendations are as follows:

► Treat posterior inflammatory eye disease with azathioprine and systemic corticosteroids. The authors cite a study (N. Engl. J. Med. 1990;322:281-5) where 2.5 mL/kg per day of azathioprine was efficacious in visual acuity and in halting disease progression.

► Severe eye involvement—greater than a 2-point drop in visual acuity on a 10/10 scale, or retinal disease—calls for a second immunosuppressive. "Cyclosporine A 2-5 mg/kg per day shows its effect rapidly and is, here, usually the treatment of choice," wrote the authors. Infliximab and interferon-α are also candidates, though the latter is considered a second choice.

► For BD-associated acute deep vein thrombosis, corticosteroids, azathioprine, cyclophosphamide, or cyclosporine A are recommended. However, "there are no RCTs addressing this issue." The same treatment is recommended for pulmonary and peripheral artery aneurysms.

► Pulmonary embolism is rare, so anticoagulants, antiplatelets, and fibrinolytic agents are not recommended. This is doubly true because of the chance of a coexisting pulmonary arterial aneurysm. Again, however, "controlled trials are needed."

► Immunosuppressants should be the first-line treatment over surgery in case of gastrointestinal ulcers, though no controlled trials exist to support one treatment specifically. "One study reported that azathioprine decreased reoperation rates and suggested that it should be used as maintenance therapy in patients who require surgery (Dis. Colon Rectum 2000;43:692-700)," wrote the authors.

► In most patients, arthritis can be managed with colchicine.

► For parenchymal involvement, "3-7 pulses of intravenous methylprednisone 1 g/day is given during attacks, followed by maintenance oral corticosteroids which is tapered over 2-3 months." However, the authors caution that central nervous system (CNS) involvement in BD is mostly based on anecdotal reports.

► Neurotoxic cyclosporine A should not be used in BD patients with CNS involvement unless intraocular inflammation makes it an unavoidable choice.

► Regarding skin involvement, perceived severity should determine treatment. Topical steroids should be first-line treatment in genital and oral ulcers, while acnelike lesions can usually be treated with standard acne vulgaris treatments. In the literature, azathioprine was effective against resistant skin and mucosa lesions.

"With proper management, remission is frequent in eye disease, skin-mucosa disease, and arthritis," said Dr. Yazici. He added that though CNS disease and thrombotic manifestations pose difficulties, "the disease usually gets better with time. The aim of treatment is to prevent any long-term damage while it is active, since in the long term most patients are doing better and require less medication." ■