Dokumenttyp: journal article

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Titel des Beitrags: The International Criteria for Behçet's Disease (ICBD): a collaborative study of 27 countries on the sensitivity and specificity of the new criteria.

Abstract: Behçet's disease (BD) is a chronic, relapsing, inflammatory vascular disease with no pathognomonic test. Low sensitivity of the currently applied International Study Group (ISG) clinical diagnostic criteria led to their reassessment. An International Team for the Revision of the International Criteria for BD (from 27 countries) submitted data from 2556 clinically diagnosed BD patients and 1163 controls with BD-mimicking diseases or presenting at least one major BD sign. These were randomly divided into training and validation sets.
Logistic regression, 'leave-one-country-out' cross-validation and clinical judgement were employed to develop new International Criteria for BD (ICBD) with the training data. Existing and new criteria were tested for their performance in the validation set. For the ICBD, ocular lesions, oral aphthosis and genital aphthosis are each assigned 2 points, while skin lesions, central nervous system involvement and vascular manifestations 1 point each. The pathergy test, when used, was assigned 1 point. A patient scoring ≥4 points is classified as having BD. In the training set, 93.9% sensitivity and 92.1% specificity were assessed compared with 81.2% sensitivity and 95.9% specificity for the ISG criteria. In the validation set, ICBD demonstrated an unbiased estimate of sensitivity of 94.8% (95% CI: 93.4-95.9%), considerably higher than that of the ISG criteria (85.0%). Specificity (90.5%, 95% CI: 87.9-92.8%) was lower than that of the ISG criteria (96.0%), yet still reasonably high. For countries with at least 90%-of-cases and controls having a pathergy test, adding 1 point for pathergy test increased the estimate of sensitivity from 95.5% to 98.5%, while barely reducing specificity from 92.1% to 91.6%. The new proposed criteria derived from multinational data exhibits much improved sensitivity over the ISG criteria while maintaining reasonable specificity. It is proposed that the ICBD criteria to be adopted both as a guide for diagnosis and classification of BD.