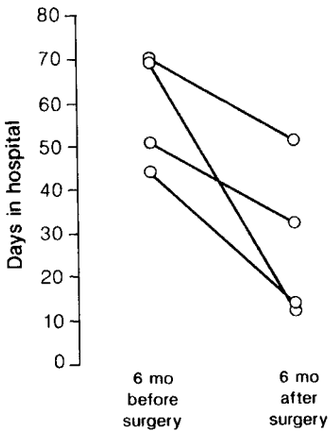

Days in hospital during 6 months before and 6 months after surgery.

58 days; $p < 0.04$; see accompanying figure). The observation periods were long, and they began at different months of the year for each patient, so it is unlikely that the improvement in the frequency of hospital admission was due to seasonal effects on lung disease.

Discussion

Our finding that aggressive management of sinus disease in patients with cystic fibrosis can reduce the need for hospital admission suggests that the presence of sinus disease in these patients may result in the exacerbation of lower respiratory tract symptoms. These findings are similar to those in patients with asthma, in whom medical and surgical management of sinusitis greatly alleviates asthma symptoms.[3-6]

Our results confirm that abnormal sinus X-rays can indicate infection in patients with cystic fibrosis,[11] but it is not clear which cystic fibrosis patients are likely to have infections. We believe, however, that the patients with a high degree of reactive airways, for example as shown by the use of corticosteroids for refractory wheezing, are most likely to have sinus infections and to benefit most from aggressive sinus management.

We found that intravenous anti-*Pseudomonas* antibiotics may not be sufficient to eradicate sinus infections in patients with cystic fibrosis. Sinusitis in these patients is very difficult to control even with surgery, and repeated mechanical lavage and direct instillation of anti-*Pseudomonas* antibiotics into the sinuses may be required. Nevertheless, we believe that aggressive management of sinusitis should prove useful in the treatment of respiratory disease in selected patients with cystic fibrosis.

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Criteria for diagnosis of Behçet's disease

INTERNATIONAL STUDY GROUP FOR BEHÇET'S DISEASE*

5 sets of criteria for diagnosis of Behçet's disease are in use—a problem which has hindered interpretation of different studies and collaborative research. An international study group, which included at least one proponent of 4 of the sets, was formed to derive new, internationally agreed diagnostic criteria for Behçet's disease. Data on 914 patients with Behçet's disease, from 12 centres in 7 countries, were compared with controls from the same centres. The new set of diagnostic criteria—which requires the presence of oral ulceration plus any two of genital ulceration, typical defined eye lesions, typical defined skin lesions, or a positive pathergy test—was simpler to use and had an improved discriminatory performance than its predecessors.

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Introduction

The clinical triad of uveitis with oral and genital ulceration was probably first recognised by Hippocrates,[1] but bears Behçet's name after his descriptions of the illness some 50 years ago.[2-4] The disease is heterogeneous with variable involvement of many organ systems, the exact cause is unclear, and there is no universally accepted diagnostic test: thus diagnosis of Behçet's disease has relied on identification of several of its more typical clinical features. However,

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TABLE I—DIAGNOSTIC VALUE OF INDIVIDUAL FEATURES IN 60% SAMPLE

Feature	Behçet's	Controls	Sensitivity	Specificity	Relative value	Log-likelihood ratio		Expected value
						Feature present	Feature absent	
Genital ulcers	530	55	77	95	172	+27	−14	17
Eye lesions	520	55	60	93	153	+22	−8	10
Pathergy test	497	52	58	90	148	+18	−8	7
Folliculitis, papulopustular lesions, acneiform nodules	514	55	70	76	146	+11	−9	5
Erythema nodosum	521	55	44	95	139	+22	−5	7
Folliculitis, papulopustular lesions, acneiform nodules/erythema nodosum	531	55	81	75	156	+12	−14	7
Subcutaneous thrombophlebitis	519	55	16	95	111	+12	−1	1
Deep vein thrombosis	519	55	13	96	109	+12	−1	1
Epididymitis	461	38	8	100	108	+18	−1	1
Arterial occlusion and/or aneurysms	509	55	3	98	101	+4	0	0
CNS involvement	513	54	10	89	99	−1	0	0
Arthralgia	524	53	56	38	94	−1	+2	0
Family history	363	52	20	73	93	−3	+1	0
Gastrointestinal features	519	55	9	82	91	−7	+1	0
Arthritis	517	55	38	38	76	−5	+5	1

confusion has arisen because 5 such sets of diagnostic criteria are in current use, each with its own list of clinical features and its own rules about the number and nature of criteria to be fulfilled to make a diagnosis of Behçet's disease (Mason and Barnes,[5] Behçet's Disease Research Committee of Japan,[6-8] O'Duffy,[9,10] Zhang,[11] and Dilsen[12,13]). These differences, which have hindered both interpretation of different trials and international multicentre collaboration, were addressed at the 4th International Conference on Behçet's disease, held in London in 1985. An international study group (which included proponents of 4 of 5 of the sets currently used, the exception being Dr X-Q Zhang from China) was set up to compare the performance of existing sets and to agree on one set of diagnostic criteria which would be quoted (though perhaps not alone) in all future published work.

Methods

914 unselected patients with Behçet's disease were recruited from the 12 centres, in 7 countries, which participated in the study. (366 were from Iran, 285 from Turkey, 141 from Japan, 50 from Tunisia, 21 from the UK, 14 from the USA, and 9 from France.) The diagnosis was based on the decision of an experienced clinician according to the diagnostic criteria with which they were most familiar. 28 (3%) did not have oral ulceration and were excluded from this analysis—although the study group accepts that, very rarely, patients with Behçet's disease may not have recurrent oral ulceration. This exclusion meant that a comparison group could be selected, which consisted of 97 individuals who also had recurrent oral ulceration and who were drawn from a panel of 308 patients from the same centres with connective tissue disease (eg, rheumatoid arthritis, systemic lupus erythematosus, ankylosing spondylitis, psoriatic arthropathy) which may show similar features to Behçet's disease.

Data for each patient or control were recorded by the participating physician on a standard form for the presence or absence of all features of Behçet's disease included in all current sets of diagnostic criteria. For each feature, where relevant, current and past presence, and whether it was observed by the patient or a physician, was recorded. Each patient also had a pathergy test performed which, where possible, was read by the physician in charge. This test of cutaneous hypersensitivity, when positive, consists of a sterile pustule that develops after 24–48 h at the site of a needle prick to the skin.[14]

Data on all patients notified were coded and entered onto a computer. Algorithms designed to allocate patients according to each available set of diagnostic criteria were used for computer-generated diagnostic assignments. Thus the sensitivity, specificity, and relative value of those sets were determined. Data from a randomly chosen 60% sample of the total study population with Behçet's disease (the training sample) were used to calculate an expected weight of evidence for each individual feature.[15] These were derived from the log-likelihood ratios for the presence or absence of that feature and its prevalence in the diagnosis of Behçet's disease. From these a new set of diagnostic criteria (that of the international study group [ISG]) was derived. The sensitivity and specificity of ISG diagnostic criteria were compared with previous sets in this 60% sample and then in the remaining 40% of patients with Behçet's disease (the validation sample).

Results

Preliminary analysis rapidly indicated that to rely only on current presence of any individual feature would be too insensitive for any reliable set of diagnostic criteria. Therefore all analyses allow for past or present symptoms or signs. Table I shows the relative performance of individual features in the 60% training sample: the first 5 individual items (genital ulceration, eye lesions, positive pathergy test, folliculitis, and erythema nodosum, and the last two skin lesions together) all showed good discrimination for Behçet's disease; none of the other features showed useful diagnostic value. Table I also shows the log-likelihood ratios and expected value for each feature, with similar results. In particular, the absence in a patient of such well-recognised complications of Behçet's disease as epididymitis or subcutaneous thrombophlebitis did not improve diagnostic accuracy, whereas absence of the major positive discriminants was much more important.

A new set of diagnostic criteria (ISG criteria), were derived from the 60% training sample. This set required the presence of two of the four features: genital ulceration, eye

TABLE II—COMPARATIVE PERFORMANCE OF SETS OF CRITERIA FOR DIAGNOSIS OF BEHÇET'S DISEASE

Set	Sensitivity (%)	Specificity (%)	Relative value
Dilsen et al[12]	93; 98	75; 91	168; 189
Japanese [6-8]	92; 94	89; 93	181; 187
Mason/Barnes[5]	86; 93	84; 95	170; 188
O'Duffy[9,10]	80; 85	80; 95	160; 180
Zhang [11]	98; 99	62; 74	160; 173
ISG	91; 95	96; 98	187; 193

Results shown for 60% training sample; 40% validation sample.

lesions, positive pathergy test, and either of the skin lesions listed above in addition to oral ulceration.

Table II shows the sensitivity, specificity, and relative value of ISG criteria in comparison with existing sets of diagnostic criteria for both the 60% training sample and the 40% validation sample of patients with a clinical diagnosis of Behçet's disease: ISG criteria showed better specificity with little loss of sensitivity in both groups of patients.

Discussion

Our main aim was to obtain an agreed set of diagnostic criteria for Behçet's disease to be quoted in all future published work on the disorder, to enable comparisons between studies and to be used as a basis for multicentre collaborative studies. It had been anticipated that such an exercise might introduce simplifications and generalisations that may have adversely affected specificity or sensitivity, and that some workers may have wanted to also quote data derived from earlier diagnostic criteria. Instead, the analysis yielded a single set of diagnostic criteria which are simpler to use, exclude rarer (and more subjective) manifestations, and are more specific than any previously available set, with little or no loss of sensitivity. Inclusion of recurrent oral ulceration as a non-optional criterion may exclude approximately 3% of patients considered to have definite Behçet's disease by an experienced clinician (although some centres now indicate that this might be an overestimate): we do not claim that Behçet's disease cannot exist without a history of oral ulceration, but suggest that exclusion of these very few patients from collaborative trials would not affect their conclusions. The ISG criteria were agreed at an international workshop;[16] the definitions used to define the features were:

Recurrent oral ulceration	Minor aphthous, major aphthous, or herpetiform ulceration observed by physician or patient, which recurred at least 3 times in one 12-month period
Plus 2 of:	
Recurrent genital ulceration	Aphthous ulceration or scarring, observed by physician or patient
Eye lesions	Anterior uveitis, posterior uveitis, or cells in vitreous on slit lamp examination; *or* Retinal vasculitis observed by ophthalmologist
Skin lesions	Erythema nodosum observed by physician or patient, pseudofolliculitis, or papulopustular lesions; *or* Acneiform nodules observed by physician in postadolescent patients not on corticosteroid treatment
Positive pathergy test	Read by physician at 24–48 h.

(Findings applicable only in absence of other clinical explanations.)

Several previous sets of diagnostic criteria included a long list of minor symptoms or signs which might accumulate towards the diagnosis. We accept that these include several important clinical features that may indicate a possible diagnosis of Behçet's disease, but suggest that they do not occur with sufficient frequency to be included in a set of diagnostic criteria, and that the simpler such lists are, the less the chance of subjective error. However, these minor features (the last 9 in table I) will be included in an appendix for training. Subgroup analysis indicated that the ISG criteria worked equally well in all countries but it is possible that, in countries where Behçet's disease is rare, the predictive value of any diagnostic criteria will be lower than in countries where Behçet's disease is the usual explanation for the clinical features described. Further validation of the criteria in individual countries is under way.

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From The Lancet

Solitary confinement and the silent system

SIR:—As I am led to understand that the Government are about to make an experiment upon the solitary confinement and silent system of punishment, in the model prison, which is now being erected, I beg to call your attention to the subject, lest it should be adopted and carried too far, without looking to the effects produced by the same system in other countries where it has been tried for some time, particularly in America and Belgium. From reports published in America we find that long solitary confinement has the effect of debilitating the mind, as well as the body, and after their time has expired, many criminals are thrown upon the world in a state of complete idiotcy, besides having contracted habits contrary to nature and prejudicial to health . . . I hope some of your readers, who have had opportunities of making personal observations upon this subject, will take it up, as there can be no doubt that it may be an excellent mode of punishment, when carried to a certain extent, but beyond that it is highly injurious . . . In the report I allude to, there is a great deal of religious feeling and self-laudation on the part of those interested in drawing it up; but there are many startling facts, which, if true, would make the refined cruelty of the silent system appear less humane and merciful than was the Spanish Inquisition. I am yours, most obediently,

110, Guilford Street. Wm Simpson

(6 June 1840)