Journal of Medical & Surgical Pathology

Davatchi et al., J Med Surg Pathol 2016, 2:1
DOI: 10.4172/2472-4971.1000141

Research Article Open Access

Behcet's Disease Criteria

Fereydoun Davatchi[1,2*], Bahar Sadeghi Abdollahi[1], Farhad Shahram[1], Cheyda Chams Davatchi[1], Abdolhadi Nadji[1], Hormoz Shams[1], Massoomeh Akhlaghi[1], Tahreh Faezi[1], Zahra Ghodsi[1], Farimah Ashofteh[1], Negin Mohtasham[1], Hoda Kavosi[1] and Mariam Masoumi[1]

[1]Behcet's Disease Unit, Rheumatology Research Center, Tehran University of Medical Sciences, Iran
[2]Chair Behcet's Disease, Iran National Elite Foundation, Iran

[*]**Corresponding Address:** Fereydoun Davatchi, Rheumatology Research Center, Shariati Hospital, Kargar Avenue, Tehran 14117, Iran, Tel: (9821) 8802-6953; E-mail: fddh@davatchi.net

Received date: July 27, 2016; **Accepted date:** August 19, 2016; **Published date:** August 24, 2016

Abstract

Objective: From 1946 to 2014, 16 different sets of Classification/Diagnosis criteria, from different countries, have been made for Behcet's Disease. Among them, there are two International criteria sets, the International Study Group (ISG) criteria in 1990 by the collaboration of 7 countries, and the International Criteria for Behcet's Disease (ICBD) in 2010 by the collaboration of 27 countries. The aim of this study is to compare the performance of them, in new patients and controls, from the Iran registry of Behcet's Disease.

Methods: Patients (1323) and controls (2438) are consecutive patients, seen from 2010 to 2016. The diagnosis was clinical and by expert opinion. Sensitivity, specificity, accuracy, optimization, efficiency, positive predictive value, negative predictive value, positive likelihood ratio, negative likelihood ratio, diagnostic odds ratio, and Youden's index were calculated.

Results: The sensitivity of ISG was 64% versus 96.9% ICBD (a difference of 32.9%), while specificity was 99.9% versus 97.4% (a difference of 2.5%), and accuracy 87.3% *vs.* 97.2% (a difference of 9.9%). The optimization (100-difference between sensitivity and specificity. The best score is 100) was 64.1 versus 99.7 (a difference of 35.6). The efficiency ((optimization/100) × accuracy. The best score is 100) was 56 versus 96.9 (a difference of 40.9).

Conclusion: The differences for all the items was statistically highly significant, even for the specificity, although numerically the difference was small, just 2.5%. However, clinically, the difference was not relevant. The performance of ICBD is much higher than that of ISG. The ICBD criteria have a much higher sensitivity, accuracy, optimization, and efficiency, with clinically, approximately the same specificity.

Keywords: Behcet's disease; Vasculitis

Introduction

Behcet's Disease (BD) is classified among vasculitides. In the majority of cases, it is easily recognizable from other vasculitis. One of the major differences is its progression by repetitive attacks and remissions. Depending on the involved organ, the healing process may result in a return to normal of the tissue, like oral mucosa or joints. Some organs, on the contrary, may progress toward fibrosis, or sequela, like the eyes, and the brain [1,2].

BD has no characteristic laboratory tests, different imaging, and no specific pathological patterns on tissue biopsy. Therefore, the diagnosis is only clinical, while classification/diagnosis criteria may be of help. However, even when a patient fulfils the criteria [3-5]. As many symptoms of BD can be seen in other diseases too, the association of two or more can happen fortuitously, without being BD [2].

BD is one of the diseases having many Classification/Diagnosis criteria. The first was created in 1946 by Curth [6], followed by Hewitt [7], Mason and Barnes [8], Japan [9], Hubault and Hamza [10], O'Duffy [11], Cheng and Zhang [12], Dilsen [13], Japan revised [14], International Study Group (ISG) [15], Iran [16], Classification Tree [17], Dilsen revised [18], Korea [19], International Criteria for Behcet's Disease (ICBD) [20], and ICBD revised [21] in 2014. Among these criteria, two were made by an International collaboration. The first was ISG criteria in 1990, made by the collaboration of 7 countries (France, Iran, Japan, Tunisia, Turky, UK, and USA). The ICBD was created by the collaboration of 27 countries.

We showed previously [4] that ISG has an excellent specificity in different studies, 97% in its original study [15], 97.5% in Iran [16], 79.8% in China [22], 99.3% in 1998 APLAR study [23], 99.8% in 2000 Russia study [24], 96% in 2006 ICBD database [20], 89.5% in 2008 Germany report [25], 99.2% in 2008 China report [26], and 98.8% in 2010 Iran report [27]. Unfortunately, the sensitivity of the ISG was low in the majority of reports. It was 92% in its original study [15], 86.2% in 1993 Iran [16], 72.2% in APLAR 1998 [23], 79.8% in Russia 2000 [24], 75.6% in USA 2000 [28], 72% in India 2004 [29], 46% in Singapore 2004 [29], 81% in China 2004 [29], 58% in Korea 2004 [29], 82% in Iran 2004 [29], 82.4% in ICBD 2006 [20], 83.7% in Germany 2008 [25], 65.4% in China 2008 [26], and 78.1% in Iran 2010 [27]. Due to the low sensitivity, the accuracy was low too, despite the excellent specificity [4].

The preceding study showed the performance of ISG and ICBD from the beginning to 2010 [4]. The aim of this study is to show their performance (in Iranian patients) from 2010 to 2016, and compare the performance ISG with the original ICBD and its revised form.

Methods

Patients and controls

From 2010 to July 2016, 1323 BD and 2438 control patients (mimicking Behcet's Disease) were selected as consecutive patients, from the Behcet's Disease Registry of Iran.

Statistics

Sensitivity was calculated as the number of BD patients, classified by the criteria, multiplied by 100, and divided by the total number of BD patients (here 1323). Specificity was calculated by the number of patients, who were correctly recognized as not having BD, by the criteria, multiplied by 100, and divided by the total number of control patients. Accuracy (Percent Agreement) was calculated by the number of BD patients, correctly classified as having BD, by the criteria+the number of patients, who were correctly recognized as not having BD, by the criteria. The total is then multiplied by 100, and divided by the total number of BD patients and the total number of control patients. The optimization is how the criteria recognize the BD patients and the control patients. The ideal is to have the same rate of error for patients and controls. For that, the difference between sensitivity and specificity is subtracted from 100 to find the optimization as a percentage. The best score will be 100% optimization. The efficiency is calculated by dividing the optimization by 100, and then multiplying the result by the Accuracy.

Comparison of results was done by the Pearson's chi square test.

Results

In the cohort of patients (from January 2007 to mid-July 2016) 3761 patients were seen. Among them, 1323 were BD and 2438 were control patients.

The sensitivity of the ISG was 64% with a 95% confidence interval (95%CI) of 2.6. For ICBD, the sensitivity with the original criteria was 97.9% (95%CI 0.8) and with revised version 96.9% (95%CI 0.9).

The specificity of the ISG was 99.9% (95%CI 0.1). For ICBD, the specificity with the original criteria was 97.3% (95%CI 0.6) and with revised version 97.4% (95%CI 0.6).

The accuracy of the ISG was 87.3% (95%CI 1.1). For ICBD, with the original criteria, it was 97.5% (95%CI 0.5) and with the revised version 97.2% (95%CI 0.5).

The performance of the other Classification/Diagnosis criteria is shown in Table 1.

	Sensitivity			Specificity			Accuracy		
Criteria	Number	%	CI	Number	%	CI	Number	%	CI
Curth	1310	99.0	0.5	2312	94.8	0.9	3622	96.3	0.6
Mason	696	52.6	2.7	2436	99.9	0.1	3132	83.3	1.2
Hewitt	465	35.1	2.6	2436	99.9	0.1	2901	77.1	1.3
Japan Original	1046	79.1	2.2	2390	98.0	0.6	3436	91.4	0.9
Hubault	657	49.7	2.7	2434	99.8	0.2	3091	82.2	1.2
O'Duffy	762	57.6	2.7	2435	99.9	0.1	3197	85.0	1.1
Zhang	1261	95.3	1.1	2280	93.5	1.0	3541	94.2	0.7
Dilsen	901	68.1	2.5	2392	98.1	0.5	3293	87.6	1.1
Japan Revised	1059	80.0	2.2	2388	97.9	0.6	3447	91.7	0.9
ISG	847	64.0	2.6	2435	99.9	0.1	3282	87.3	1.1
Iran	1107	83.7	2.0	2389	98.0	0.6	3496	93.0	0.8
Classification Tree	1284	97.1	0.9	2375	97.4	0.6	3659	97.3	0.5
Dilsen Revised	873	66.0	2.6	2434	99.8	0.2	3307	87.9	1.0
Korea	1038	78.5	2.2	2417	99.1	0.4	3455	91.9	0.9
ICBD	1295	97.9	0.8	2371	97.3	0.6	3666	97.5	0.5
ICBD revised	1282	96.9	0.9	2375	97.4	0.6	3657	97.2	0.5

Table 1: Performance of the other Classification/Diagnosis criteria.

The optimization of the ISG was 64.1%, while the original ICBD was 99.4%, and the revised ICBD 99.5%.

The efficiency of the ISG was 56% for ISG, 96.9% for the original ICBD, and 96.7% for the revised ICBD.

Discussion

The new study has approximately the same results as the older study. The ISG, in the older data, up to 2010, had a sensitivity of 78.1%

against the original ICBD with 98.2%. The specificity was 98.8% against 95.6%. The accuracy was 85.5% against 97.3% [4]. The sensitivity of the ISG was lower in newer data than in the old data, while with ICBD, both were very close. For specificity, the difference was minimal between the new and the old data for both ISG and ICBD. For the accuracy, it was the same as for specificity, minimal differences between the new and the old data, for ISG and ICBD.

References

1. Davatchi F, Shahram F, Chams-Davatchi C, Shams H, Nadji A, et al. (2010) Behcet's disease: from East to West. Clin Rheumatol 29: 823-833.
2. Davatchi F, Chams-Davatchi C, Shams H, Shahram F, Nadji A, et al. (2016) Behcet's disease: epidemiology, clinical manifestations, and diagnosis. Expert Rev Clin Immunol 11:1-9.
3. Davatchi F, Sadeghi Abdollahi B, Shahram F, Nadji A, Chams-Davatchi C, et al. (2010) Validation of the International Criteria for Behçet's disease (ICBD) in Iran. Int J Rheum Dis 13: 55-60.
4. Davatchi F (2012) Diagnosis/Classification Criteria for Behcet's Disease. Patholog Res Int 2012: 607921.
5. Davatchi F, Sadeghi Abdollahi B, Chams-Davatchi C, Shahram F, Shams H, et al. (2015) The saga of diagnostic/classification criteria in Behcet's disease. Int J Rheum Dis 18: 594-605.
6. Curth HO (1946) Recurrent genito-oral aphthosis and uveitis with hypopyon (Behcet's syndrome). Arch Derm Syphilol 54: 179-196.
7. Hewitt J, Escande JP, Lauret P, Perlemuter L (1969) [Criteria for diagnosis of Behcet's syndrome]. Bull Soc Fr Dermatol Syphiligr 76: 565-568.
8. Mason RM, Barnes CG (1969) Behçet's syndrome with arthritis. Ann Rheum Dis 28: 95-103.
9. Behcet's Disease Research Committee of Japan (1974) Behcet's Disease guide to the diagnosis of Behcet's Disease (1972). Jpn J Ophthalmol 18: 291-294.
10. Hubault A, Hamza M (1974) La maladie de Behçet en 1974: In: L'actualité Rhumatologique 15: 43-55.
11. O'Duffy JD (1974) Critères proposés pour le diagnostique de la maladie de Behçet et notes therapeutiques. Rev Med (Paris) 36: 2371-2379.
12. Cheng SP, Zhang XQ (1980) Some special clinical manifestations of Behçet's disease-report of illustrative cases and review of literature (author's transl) (in Chinese). Zhonghua Nei Ke Za Zhi 19: 15-22.
13. Dilsen N, Konice M, Aral O (1986) Our diagnostic criteria of Behcet's Disease-an overview: In: Recent Advances in Behcet's Disease 177-180.
14. Mizushima Y (1988) Recent research into Behçet's disease in Japan. Int J Tissue React 10: 59-65.
15. [No authors listed] (1990) Criteria for diagnosis of Behçet's disease. International Study Group for Behçet's Disease. Lancet 335: 1078-1080.
16. Davatchi F, Shahram F, Akbarian M, Gharibdoust F, Nadji A, et al. (1993) Accuracy of existing diagnostic criteria for Behcet's Disease: In: Behcet's Disease 225-228.
17. Davatchi F, Shahram F, Akbarian M, Gharibdoust F, Chams C, et al. (1993) Classification Tree for the Diagnosis of Behcet'sd Disease: In: Behcet's Disease 245-248.
18. Dilsen N (2000). About diagnostic criteria for Behcet's Disease: our new proposal: In: Behcet's Disease, Design Mecca Publishing Co. Seoul 101-104.
19. Chang HK, Lee SS, Bai HJ, Lee YW, Yoon BY, et al. (2004) Validation of the classification criteria commonly used in Korea and a modified set of preliminary criteria for Behçet's disease: a multi-center study. Clin Exp Rheumatol 22: S21-26.
20. International Team for the Revision of the International Criteria for Behcet's Disease (2006) Revision of the International Criteria for Behcet's Disease (ICBD). Clin Exp Rheumatol 24 (Suppl 42): S14-S15.
21. International Team for the Revision of the International Criteria for Behçet's Disease (ITR-ICBD), Davatchi F, Assaad-Khalil S, Calamia KT, Crook JE, et al. (2014) The International Criteria for Behçet's Disease (ICBD): a collaborative study of 27 countries on the sensitivity and specificity of the new criteria. J Eur Acad Dermatol Venereol 28: 338-347.
22. Dong Y, Yao Q, Wang M (1996) Behcet's disease in China, in Proceedings of the 8th APLAR Congress of Rheumatology. Melbourne, Australia S14.
23. APLAR subcommittee for Behcet's Disease (1998) APLAR evaluation of Behcet's disease diagnosis criteria. APLAR J Rheumatol 1: 237-240.
24. Prokaeva T, Reshetnjak T, Kuzin A, Rabinovich I, Ermakova N, et al. (2000) Evaluation of existing diagnosis criteria for Behcet's disease: In: Behcet's Disease 598-603.
25. Altenburg A, Bonitsis NG, Papoutsis N, Pasak M, Krause L, et al. (2008) Evaluation of diagnostic criteria including ICBD (2006) in Adamantiades-Behcet's disease patients in Germany. Clin Exp Rheumatol 26: S3.
26. Zhang Z, Zhou W, Hao Y, Wang Y, Dong Y (2008) Validation of the International criteria for Behcet's disease (ICBD) in China. Clin Exp Rheumatol 26: S6-S7.
27. Davatchi F, Sadeghi Abdollahi B, Shahram F, Nadji A, Chams Davatchi C, et al. (2010) Validation of the international criteria for Behcet's Disease in Iran. Int J Rheum Dis 13: 55-60.
28. Calamia KT, Davatchi F (2000) Sensitivityofdiagnosiscriteria in United States patients with Behcet's disease. In Behcet's Disease 121-124.
29. Davatchi F, Shahram F, Kumar A, Cheng YK, Cheong CT, et al. (2004) Comparative analysis of Behcet's disease in the APLAR region. APLAR J Rheumatol 7: 38-43.