JRD Journal of Rheumatic Diseases Vol. 23, No. 1, February, 2016
http://dx.doi.org/10.4078/jrd.2016.23.1.19

Original Article

Behçet's Disease Current Activity Form as a Patient's Derived Measure

Hyo Jin Choi, Mi Ryoung Seo, Hee Jung Ryu, Han Joo Baek
Division of Rheumatology, Department of Internal Medicine, Gachon University Gil Medical Center, Incheon, Korea

Objective. This study measured the reliability of the Behçet's Disease Current Activity Form (BDCAF) questionnaire used as a patient self-report form. **Methods.** A study was conducted among 63 patients with Behçet's disease who attended our rheumatology clinic. First, a physician administered a BDCAF questionnaire. Second, the patient completed a self-administered questionnaire at home within 24 hours of the visit. The test–retest reliability was analyzed using kappa tests. Kappa scores of >0.6 indicated good agreement. The BDCAF score was compared with the patient's/clinician's perception of disease activity and the Korean version of Behçet's Disease Quality of Life (BDQOL). **Results.** The study included 17 males and 46 females. The mean age of participants was 47.7 years and the mean disease duration was 5.3 years at the first assessment. Fifty-three patients (84.1%) returned the questionnaires to us by mail. For test–retest reliability, good agreement was achieved with the items including headache, oral/genital ulceration, erythema, arthritis, and diarrhea with altered/frank blood per rectum; moderate agreement with skin pustules, arthralgia, and eye involvement; fair agreement with nausea/vomiting/abdominal pain, nervous system, and major vessel involvement. Significant associations were observed between BDCAF scores with the patient's/clinician's perception of disease activity and BDQOL ($p<0.05$). **Conclusion.** The BDCAF appears useful as a patient self-report instrument for assessment of disease status. **(J Rheum Dis 2016;23:19-22)**

Key Words. Behçet's Disease Current Activity Form, Self-report measure, Reliability

INTRODUCTION

Behçet's disease (BD) is a chronic, multisystem disorder that is a fluctuant chronic disease like rheumatoid arthritis (RA). It is difficult to define the disease activity in BD [1,2]. We validated a Korean version of Behçet's Disease Current Activity Form (BDCAF) [3] from the revised version of 2006 (http://www.behcet.ws) of original study [4]. The Iranian Behçet's Disease Dynamic Activity measure (IBDDAM) and BDCAF are the most widely used indices in BD, but are designed for administration by physicians, who are usually pressed for time during busy clinics [5]. Behçet's Syndrome Activity Score (BSAS) is a patient-administered measure [6]. Self-assessment tools, especially in RA, are advantageous in clinical practice [7,8], but many physicians consider a patient questionnaire subjective, not an objective tool [8]. This study evaluated the reliability of BDCAF as a patient-derived measure.

MATERIALS AND METHODS

Sixty-three patients with BD were collected prospectively from March 2013 through August 2013. All patients were older than 18 years and fulfilled the criteria of International Study Group for BD or Chang's criteria [9,10]. All patients agreed to a written informed consent to participate in this study, and the Institutional Review Board of Gachon University Gil Medical Center approved the study protocol. The first BDCAF questionnaire was administered by a physician to the patient during his or her visit to the rheumatology clinic (test 1) and the sec-

Received : May 20, 2015, **Revised :** (1st) June 29, 2015, (2nd) July 13, 2015, (3rd) July 14, 2015, **Accepted :** July 15, 2015

Corresponding to : Han Joo Baek, Division of Rheumatology, Department of Internal Medicine, Gachon University Gil Medical Center, 21 Namdong-daero 774beon-gil, Namdong-gu, Incheon 21565, Korea. E-mail : baekhj@gilhospital.com

pISSN: 2093-940X, eISSN: 2233-4718

ond BDCAF was scored by the patient at home within 24 hours after first BDCAF and was sent to us via mail (test 2). Patients were asked to assess overall perception of disease activity at test 1 and 2. The clinician's overall perception of disease activity was recorded when the first BDCAF was administered. The questionnaire takes 5 to 10 minutes to complete. The Korean version of Behçet's Disease Quality of Life (BDQOL) was also taken by patient both times that the BDCAF was filled out [11]. The agreement between the first and second assessments was assessed by calculating kappa statistics. Kappa score higher than 0.6 is considered good agreement, 0.41 to 0.6 as moderate agreement, 0.21 to 0.4 as fair agreement, and below zero as poor agreement. The correlations between BDCAF score (transformed), patient's/physician's visual analogue scale (VAS), and BDQOL were calculated by Spearman's coefficient method. Null hypotheses of no difference were rejected if p-values were less than 0.05. Statistical analysis was performed using PASW Statistics ver. 18.0 (IBM Co., Armonk, NY, USA).

RESULTS

Seventy-three percent (n=46) of the patients were female. The mean age of sixty-three patients was 47.7 years and the mean disease duration was 5.3 years at the first BDCAF assessment. The mean patient VAS was 3.9 and that of physician was 4.1. The mean of transformed BDCAF score was 6.3 and of BDQOL score was 8.6. Fifty-three (84.1%) patients completed the second BDCAF questionnaire. Demographic and clinical characteristics of patients are summarized in Table 1. There was no stat-

Table 1. Demographic and clinical characteristics of study patients

Characteristic	Physician-derived BDCAF (n=63)	Patient-derived BDCAF (n=53)
Age (yr)	47.7±11.0	47.5±11.0
Sex (M:F)	17:46	14:39
Disease duration (yr)	5.3±4.9	4.9±4.8
Clinical symptoms		
Oral ulcer	63 (100.0)	53 (100.0)
Genital ulcer	40 (63.5)	32 (60.4)
Skin lesions	41 (65.1)	36 (67.9)
Organ involvement		
Eye	12 (19.0)	10 (18.9)
Vascular	5 (7.9)	5 (9.4)
Joints	24 (38.1)	18 (34.0)
Gastrointestinal	7 (11.1)	5 (9.4)
Central nervous system	5 (7.9)	5 (9.4)
Medication patterns and dose (mg)		
Colchicine	40 (1.3±0.4)	34 (1.3±0.4)
NSAIDs	18 (689.7±384.8)	16 (743.8±363.3)
Steroids	33 (9.8±9.7)	27 (10.1±10.3)
Immunosuppresants		
Azathioprine	6 (91.7±20.4)	5 (100.0±0.0)
Methotrexate	7 (12.9±3.7)	7 (12.9±3.7)
Sulfasalazine	5 (1,300.0±670.8)	3 (1,166.7±763.8)
Hydroxychloroquine	4 (350.0±100.0)	4 (350.0±100.0)
Patient's VAS	3.9±1.8	4.1±1.7
Physician's VAS	4.1±1.5	
BDCAF score (transformed)	6.3±3.2	6.0±3.2
BDQOL score	8.6±7.7	8.7±8.4
ESR (mm/h)	14.9±15.7	
CRP (mg/dL)	0.6±2.1	

Values are presented as mean±SD, number only, number (%), or number (mean±SD). NSAIDs included six different drugs. Dose of methylprednisolone was converted to equivalent prednisolone dose. BDCAF: Behçet's Disease Current Activity Form, BDQOL: Behçet's Disease Quality of Life, CRP: C-reactive protein, ESR: erythrocyte sedimentation rate, F: female, M: male, NSAIDs: non-steroidal anti-inflammatory drugs, SD: standard deviation, VAS: visual analogue scale (1 to 7).

istically significant difference between BDCAF 1 and 2 scores. At the first assessment, the percentage of patients answering "yes" to the each 12 items were 82.5% for joint symptoms (arthralgia/arthritis), 77.7% for oral/genital ulcer, 47.6% for skin (erythema/pustules), 44.4% for headache, 33.3% for gastrointestinal symptoms (nausea/vomiting/abdominal pain or diarrhea with altered/frank blood per rectum), 15.9% for eye involvement, 11.1% for major vessel involvement, and 9.5% for nervous system involvement.

The agreement between BDCAF 1 and 2 items is shown in Table 2. Good agreement (kappa scores >0.6) was observed for six manifestations of BD: headache, oral/genital ulceration, erythema, arthritis, and diarrhea with altered/frank blood per rectum. Moderate agreement was observed for skin pustules, arthralgia, and eye involvement. For nausea/vomiting/abdominal pain, nervous system, and major vessel involvement, we noted fair agreement.

Table 2. The agreement between physician and patient-derived questionnaires of Behçet's Disease Current Activity Form

Item	Kappa	p-value
Headache	0.883	<0.001
Oral ulceration	0.775	<0.001
Genital ulceration	0.797	<0.001
Erythema	0.651	<0.001
Skin pustules	0.588	<0.001
Joints- arthralgia	0.586	<0.001
Joints- arthritis	0.652	<0.001
Nausea/vomiting/abdominal pain	0.332	0.005
Diarrhea with altered/frank blood per rectum	0.737	<0.001
Eye involvement	0.511	<0.001
Nervous system involvement	0.291	0.033
Major vessel involvement	0.291	0.033

The test-retest reliability of BDCAF score was good with a high correlation between the time points (Spearman's correlation coefficient, $r=0.829$, $p<0.001$). The BDCAF1 score significantly correlated with patient's VAS ($r=0.391$, $p=0.004$), physician's VAS ($r=0.366$, $p=0.010$), and BDQOL ($r=0.506$, $p=0.013$) at test 1 and BDCAF 2 score also correlated with patients' VAS ($r=0.472$, $p<0.001$) and BDQOL ($r=0.389$, $p=0.013$) at test 2 (Table 3). There was also good correlation between BDQOL 1 and 2 ($r=0.905$, $p<0.001$).

DISCUSSION

We performed this study to evaluate the reliability of BDCAF as a self-administered measure. BD presents with heterogeneous organ involvement and has a fluctuating course [1]. Quantitative measurement in BD is rare; there is no single gold standard to assess disease activity, and a lack of laboratory tests. With RA, the self-report questionnaire gives a highly reliable quantitative analysis of the disease course, and is simple to administer [7,8]. Houssien et al. [7] reported that a patient-derived disease activity score (DAS) can substitute for a physician-derived DAS in RA. Kappa analysis showed good agreement between physician and patient assessments of individual joint tenderness. Patient self-reported questionnaires, which take about 10 minutes to complete while waiting to see a physician in routine care, provide valuable data representing clinical status, monitoring disease progression or predicting outcomes. The disadvantages are cultural differences in interpretation of data, literacy requirement, motivation, and possibility of manipulation by patient [8,12].

In BD, there are few tools to measure disease activity, especially self-administered measures [6]. We investigated

Table 3. Spearman's correlation coefficient between BDCAF score and other variables

Correlation coefficient	Evaluator	Physician-derived BDCAF	Patient-derived BDCAF
BDCAF score (transformed)	By physician		0.829*
	By patient	0.829*	
Patient's VAS	By physician	0.391*	
	By patient		0.472*
Physician's VAS	By physician	0.366*	
	By patient		
BDQOL score	By physician	0.506*	
	By patient		0.389*

BDCAF: Behçet's Disease Current Activity Form, BDQOL: Behçet's Disease Quality of Life, VAS: visual analogue scale (1 to 7).
*$p<0.05$.

BDCAF as a patient self-report measure, although it was originally was developed for scoring by clinician [4]. We noted good agreement (kappa scores >0.6) for most items, but like reports of physician assessment [4,13,14], the agreement of nervous system and major vessel involvement was relative low. There were significant positive correlations between total BDCAF scores and patient's VAS, physician's VAS, and BDQOL at each time, which represent reliability and reproducibility of our study. We conclude that a patient-derived BDCAF gives results comparable to a physician-administered BDCAF, and is correlated with BDQOL and physician/patient's VAS. In addition, we questioned the patients (n=47/50) in our concurrent study [15], "which questionnaire do you think easy to fill out, BDCAF or BSAS?" 73.1% of patients selected BDCAF, 17.3% BSAS, and 9.6% responded 'I don't know'. Patient-derived BDCAF could as useful in usual clinical practice as the physician-administered BDCAF, with the advantage of not taking physician time. If the only clinical need is monitoring changes in disease activity or in patient function, this self-administered BDCAF may be sufficient.

We have some limitations in this study. First, there was a lack of generalizability and possible selection bias because our subjects are patients at one tertiary hospital. Second, because the second assessment occurred within 24 hours after physician-administered BDCAF, the patient's decision might change over time. On the other hand, patient recall could lead to high kappa agreement, although a physician decided and filled out patient's symptoms for each item at the first questionnaire. For more valid test of a self-administered measure, this point should be controlled. Third, unfortunately we did not compare patient-derived BDCAF with BSAS, because BSAS validation work [15] was performed after this study.

CONCLUSION

The patient-derived BDCAF was reliable for measuring current activity in BD patients. It can substitute for a physician-derived BDCAF.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. Sakane T, Takeno M, Suzuki N, Inaba G. Behçet's disease. N Engl J Med 1999;341:1284-91.
2. Alpsoy E, Zouboulis CC, Ehrlich GE. Mucocutaneous lesions of Behçet's disease. Yonsei Med J 2007;48:573-85.
3. Choi HJ, Seo MR, Ryu HJ, Baek HJ. Cross-cultural adaptation and validation of the Behçet's Disease Current Activity Form in Korea. Korean J Intern Med 2015;30:714-8.
4. Bhakta BB, Brennan P, James TE, Chamberlain MA, Noble BA, Silman AJ. Behçet's disease: evaluation of a new instrument to measure clinical activity. Rheumatology (Oxford) 1999;38:728-33.
5. Hatemi G, Merkel PA, Hamuryudan V, Boers M, Direskeneli H, Aydin SZ, et al. Outcome measures used in clinical trials for Behçet syndrome: a systematic review. J Rheumatol 2014;41:599-612.
6. Forbess C, Swearningen C, Yazici Y. Behçet's syndrome activity score (BSAS): a new disease activity assessment tool, composed of patient derived measures only, is strongly correlated with the Behçet's disease current activity form (BDCAF). Arthritis Rheum 2008:58(Suppl 9);S854.
7. Houssien DA, Stucki G, Scott DL. A patient-derived disease activity score can substitute for a physician-derived disease activity score in clinical research. Rheumatology (Oxford) 1999;38:48-52.
8. Pincus T, Sokka T. Complexities in the quantitative assessment of patients with rheumatic diseases in clinical trials and clinical care. Clin Exp Rheumatol 2005;23(5 Suppl 39):S1-9.
9. International Study Group for Behçet's Disease. Criteria for diagnosis of Behçet's disease. Lancet 1990;335:1078-80.
10. Chang HK, Kim SY. Survey and validation of the criteria for Behçet's disease recently used in Korea: a suggestion for modification of the International Study Group criteria. J Korean Med Sci 2003;18:88-92.
11. Yi SW, Kim JH, Lim KY, Bang D, Lee S, Lee ES. The Behçet's Disease Quality of Life: reliability and validity of the Korean version. Yonsei Med J 2008;49:698-704.
12. Pincus T. Why should rheumatologists collect patient self-report questionnaires in routine rheumatologic care? Rheum Dis Clin North Am 1995;21:271-319.
13. Hamuryudan V, Fresko I, Direskeneli H, Tenant MJ, Yurdakul S, Akoglu T, et al. Evaluation of the Turkish translation of a disease activity form for Behçet's syndrome. Rheumatology (Oxford) 1999;38:734-6.
14. Neves FS, Moraes JC, Kowalski SC, Goldenstein-Schainberg C, Lage LV, Gonçalves CR. Cross-cultural adaptation of the Behçet's Disease Current Activity Form (BDCAF) to Brazilian Portuguese language. Clin Rheumatol 2007;26:1263-7.
15. Choi HJ, Seo MR, Ryu HJ, Baek HJ. Validation and reliability of a Behçet's Syndrome Activity Scale in Korea. Korean J Intern Med 2016;31:170-5.