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Epidemiological and Clinical Features of Behçet's Disease in Korea

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Behçet's disease occurs with a high prevalence in the Far East including Korea. In this report we inspected 1,155 patients with Behçet's disease and collected information concerning the clinical and epidemiologic features of Behçet's disease in Korea. In summary, patients in their 30s were the most common, with the most common age of onset in the 20s; the sex ratio was 0.63:1 with female predominance; according to revised Shimizu's classification, the order of frequency of the different types was incomplete (38.2%), suspected (25.4%), possible (20.3%) and complete (16.1%); oral ulcers were the most frequent major symptom (97.5%) and the most common initial symptom (78.5%); erythema nodosum-like lesion (55.3%) was the most common type of skin lesion; a seasonal aggravation of the disease was observed in 21.3% of patients; and the most common type of childhood onset Behçet's disease was suspected type.

Key Words: Behçet's disease, clinical and epidemiologic features

Behçet's disease is a chronic and systemic disease that may involve multiple organs (Shimizu *et al.* 1979). It was first described in 1937 by the Turkish dermatologist Hulusi Behçet as a symptom complex of recurrent oral ulcers, genital ulcers, and uveitis (Behçet, 1937). The cause of Behçet's disease is unknown and its clinical course is characterized by exacerbations and remissions of unpredictable duration and frequency.

Although Behçet's disease is distributed worldwide, its incidence is known to be high in eastern Mediterranean areas and Asia, especially Japan and China (Shimizu *et al.* 1979; Ohno, 1986). Since the first report on Behçet's disease in Korea was published in 1961 (Joo, 1961), the number of patients in Korea with Behçet's disease has been increasing. The fact that Behçet's disease is especially prevalent in Korea as well as in Japan and China has led us to suspect that Behçet's disease in Korea shares geographical, racial, or cultural intimacy with Japan and China. Previous reports (Kang, 1971; Park, 1973; Lee, 1979; Rhim *et al.* 1980; Eun *et al.* 1984; Hong *et al.* 1985) in our country support this hypothesis in parts.

The purpose of this report is to give a general outline of the clinical and epidemiological features of Behçet's disease in Korea. This is a summary of clinical and epidemiologic data from 1,155 patients with Behçet's disease in Korea, supplemented by other data from Korea, Japan and China reported independently.

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MATERIALS AND METHODS

A study group composed of 1,155 patients with

Behçet's disease was selected. They were diagnosed at the Behçet's Disease Specialty Clinic of Severance Hospital at Yonsei University College of Medicine from November 1983 to February 1990. The symptoms were classified according to the revised Shimizu's classification (Shimizu *et al.* 1974); the major symptoms included oral ulcers, genital ulcers, eye lesions, and skin lesions, and the minor symptoms included arthritic signs, neurologic signs, gastrointestinal signs, and vascular lesions. Medical records concerning clinical and epidemiologic features of Behçet's disease were made by dermatologists, entered into a computer database, and analyzed by SPSS statistical package program.

RESULTS

Classification

According to the revised Shimizu's classification, the frequency of the different types in patients with Behçet's disease was as follows: complete type 16.1%, incomplete type 38.2%, suspected type 25.4%, and possible type 20.3% (Table 1).

Sex distribution

The male to female ratio was 0.63:1 with female predominance. The male to female ratios in the complete, incomplete, suspected, and possible types of Behçet's disease were 1.05:1, 0.54:1, 0.52:1, and 0.69:1, respectively (Table 1).

Age distribution

The age of onset was most common in the 20s (37.1%). The second most common age of onset was in the 30s (25.7%), followed by teens (15.7%) and 40s (11.0%) (Table 2).

The age at the time of diagnosis was most common in the 30s (39.7%), followed by 40s (27.1%), 20s (14.0%), and 50s (13.2%) (Table 3). There was no significant difference between males and females.

Initial symptom

Oral ulcer (78.5%) was the most frequent initial symptom of Behçet's disease, followed in decreasing order by skin lesion (11.2%), genital ulcer (6.9%), and ocular symptom (3.4%)(Table 4).

Table 1. The number of patients according to Shimizu's classification

Type	No. of patients(%)		M:F ratio
Complete	186	16.1	1.05:1
Incomplete	441	38.2	0.54:1
Suspected	294	25.4	0.52:1
Possible	234	20.3	0.69:1
Total	1,155	100.0	0.63:1

Table 2. The number of patients by age of onset and sex

Age(yr)	Male		Female		Total	
	No.	%	No.	%	No.	%
Under 9	27	6.1	46	6.2	73	6.3
10~19	64	14.4	117	16.4	181	15.7
20~29	156	35.2	272	38.4	428	37.1
30~39	121	27.3	175	24.6	296	25.7
40~49	49	11.1	78	10.9	127	11.0
50~59	23	5.2	22	3.1	45	3.9
Over 60	3	0.7	2	0.2	5	0.5
Total	443	100.0	712	100.0	1,155	100.0

Table 3. The number of patients by age of diagnosis and sex

Age(yr)	Male		Female		Total	
	No.	%	No.	%	No.	%
Under 9	3	0.7	5	0.7	8	0.7
10~19	12	2.7	10	1.4	22	1.9
20~29	50	11.3	112	15.7	162	14.0
30~39	181	40.9	277	39.1	458	39.7
40~49	129	29.1	184	25.9	313	27.1
50~59	54	12.2	99	13.9	153	13.2
Over 60	14	3.1	25	3.3	39	3.3
Total	443	100.0	712	100.0	1,155	100.0

Table 4. Initial major symptom to appear

Symptom	No.	%
Oral Ulcer	723	78.5
Genital Ulcer	64	6.9
Skin Lesion	103	11.2
Ocular Lesion	31	3.4
Total	921	100.0

Table 5. Positive rate of major and minor symptoms in Behçet's disease in Korea, Japan and China

	Korea (n=1155)	(%)	Japan† (n=3316)	China‡ (n=98)
Oral Ulcer	1126	97.5	98.2	100.0
Genital Ulcer	655	56.7	73.2	81.6
Skin Lesions	700	60.6	97.1	
Erythema nodosum				70.4
Folliculitis				30.6
Ocular Symptom	329	28.5	69.1	21.4
Arthritis/Arthralgia	279	24.2	56.9	30.6
Vascular Lesion			8.9	16.3
Gastrointestinal Symptom	46	4.0	15.5	35.7
Neurologic Symptom	66	5.7	11.0	9.2
Pathergy Test			43.8	62.2

†: complete and incomplete types, ‡: type not specified. (modified from references: Dong *et al.* 1991; Nakae *et al.* 1993)

Table 6. Major manifestation combinations in incomplete and suspected types

Type	Oral	Genital	Skin	Ocular	Total(%)
Incomplete					441(100.0)
	+	+	+	−	298(67.6)
	+	+	−	+	39(8.8)
	+	−	+	+	75(17.0)
	+	−	−	+	24(5.4)
	−	−	+	+	5(1.2)
Suspected					294(100.0)
	+	−	+	−	192(65.3)
	+	+	−	−	97(33.0)
	−	+	+	−	5(1.7)

Table 7. Mean duration of major manifestations

Type	Sex	Oral ulcer	Genital ulcer	Skin lesion	Ocular lesion
Complete	M	6.4±5.2	4.8±4.2	5.5±4.7	3.6±3.2
	F	7.6±5.8	3.8±3.0	3.8±3.4	3.2±3.9
Incomplete	M	6.4±6.1	3.8±3.1	4.1±3.9	3.7±3.4
	F	7.7±6.2	4.2±3.3	4.5±4.2	4.6±5.2
Suspected	M	6.3±5.7	4.8±4.1	4.5±4.7	
	F	7.9±6.4	3.7±3.6	4.2±4.1	
Possible	M	6.1±5.9			
	F	8.3±6.6			

Values are Mean±SD (years)

General features of major and minor symptoms

Oral ulcers were seen in 97.5% of patients, skin lesions in 60.6%, genital ulcers in 56.7%, ocular lesions in 28.5%, articular symptoms in 24.2%, neurologic symptoms in 5.7%, and gastrointestinal symptoms in 4.0% (Table 5).

Among the major manifestation combinations in incomplete and suspected types, the combination of oral ulcer, genital ulcer, and skin lesion (67.6%) and the combination of oral ulcer and skin lesion (65.3%) were the most frequent combinations respectively (Table 6).

The mean duration of oral ulcer was the longest, followed by skin lesion, genital ulcer, and ocular symptom in decreasing order (Table 7). There was no significant difference among the types of the disease or between the two sexes.

The mean interval between the first major symptom, oral ulcer, and the second symptom was 5.0, 5.4, and 5.1 years when the second symptom was genital ulcer, ocular symptom, and skin lesion, respectively in the cases of complete type; 5.9, 7.9 and 6.3 years when the second symptom was genital ulcer, ocular symptom, and skin lesion, respectively in the cases of incomplete type; and 6.9 and 6.1 years when the second symptom was genital ulcer and skin lesion in the cases of suspected type (Table 8).

Oral ulcer

Oral ulcer was the most common clinical manifestation (97.5%) (Table 5).

Table 8. Average time interval between the initial and the second major manifestations

Type	Initial symptom	Second symptom	Interval (years)
Complete	oral ulcer	genital ulcer	5.0
	oral ulcer	ocular lesion	5.4
	oral ulcer	skin lesion	5.1
Incomplete	oral ulcer	genital ulcer	5.9
	oral ulcer	ocular lesion	7.9
	oral ulcer	skin lesion	6.3
Suspected	oral ulcer	genital ulcer	6.9
	oral ulcer	skin lesion	6.1

Table 9. Type of oral ulcers

Type	No.	%
Minor	508	44.0
Major	300	26.0
Herpetiform	29	2.5
Combined	289	25.0
Total	1,126	100.0

The more detailed classification of oral ulcers was described as minor, major, herpetiform, and combined types, and these were present in 44.0%, 26.0%, 2.5% and 25.0% respectively (Table 9). Frequent sites of oral ulcers were tongue (83.2%), buccal mucosa (72.2%), lip (63.7%), and gingiva (60.7%) (Table 10).

Genital ulcer

Genital ulcers were found in 55.5% of female patients and 58.7% of male patients (Table 5). The frequent sites of genital ulcers were vulva (66.1%), vaginal mucosa (35.7%), anus (9.6%), cervix (4.1%), and groin area (0.8%) in females (Table 11); penis (46.5%), scrotum (38.5%), anus (9.2%), and groin area (5.0%) in males (Table 12).

Among the female patients with genital ulcer, 25.6% of patients showed aggravation of the genital ulcer at some time related to menstruation, most frequently during the premenstrual stage (Table 13).

Table 10. Site of oral ulcers in 1,126 cases

Site	No.	%
Tongue	937	83.2
Buccal mucosa	811	72.2
Lip	717	63.7
Gingiva	683	60.7
Pharynx	266	23.6
Palate	257	22.8
Tonsil	226	20.1

Table 11. Site of genital ulcers in 395 females

Site	No.	%
Vulva	261	66.1
Vaginal mucosa	141	35.7
Anus	38	9.6
Cervix	16	4.1
Groin	3	0.8

Table 12. Site of genital ulcers in 260 males

Site	No.	%
Penis	121	46.5
Scrotum	100	38.5
Anus	24	9.2
Groin	13	5.0

Cutaneous manifestations

Cutaneous involvement was seen in 60.6% of patients (Table 5). The most frequent skin lesion was an erythema nodosum-like lesion observed in 55.3% of patients; less frequent symptoms were folliculitis (25.3%), pustule (11.1%), acneiform eruption (10.9%), furunculosis (6.0%), and erythema multiforme-like lesion (5.7%) in decreasing order (Table 14).

Ocular symptom

Ocular symptoms were found in 28.5% of patients (Table 5).

Table 13. Exacerbation of genital ulcers in relation to menstruation

Stage of menstruation	No.	%
Premenstruation	56	55.4
During menstruation	23	22.8
Postmenstruation	22	21.8
Total	101	100.0

Table 14. Types of skin lesions in 700 cases

Skin lesions	No.	%
Erythema nodosum-like	387	55.3
Folliculitis	177	25.3
Pustules	78	11.1
Acneiform eruption	76	10.9
Furunculosis	42	6.0
Erythema multiforme-like	40	5.7
Thrombophlebitis	18	2.6
Skin ulcer	14	2.0
Abscess	10	1.4
Papules	6	0.9

Table 15. Familial relationship to patient of affected members of 178 patients

Family member	No.	%
Brother, sister	63	35.4
Mother	62	34.8
Father	30	16.9
Partner	9	5.1

Table 16. Distribution of seasonally aggravated symptoms of Behçet's disease in 246 patients

Season	No.	%
Spring	88	35.8
Summer	80	32.5
Autumn	20	8.1
Winter	58	23.6

Family history

Family history was detected in 15.4% of patients. Siblings and mothers of patients comprised a great portion of relatives, while fathers were less often affected (Table 15).

Seasonal variation

Among patients with Behçet's disease, 21.3% showed seasonal variation in the amount of aggravation. The symptoms seem to be aggravated more frequently in the spring, summer, and winter and less frequently in the fall (Table 16).

DISCUSSION

Classification

To make the diagnosis of Behçet's disease, classifications according to Mason and Barnes (1969), Lehner (1977), Lehner and Barnes (1979), or Shimizu *et al.* (1979) were applied. Based on our clinical experiences we believed that the guideline by the Behçet's Disease Research Committee of Japan (the revised Shimizu's classification) was most clinically relevant. According to the revised Shimizu's classification, incomplete type was most common in our patients (Shimizu *et al.* 1974).

Although a positive pathergy test has been recently accepted as a new diagnostic criterion, its positivity was reported in only 39.6% of Korean patients with Behçet's disease (Yoon *et al.* 1987). This percentage was similar with 43.8% of Japanese cases (Nakae *et al.* 1993), but was much lower than that of Turkish patients (Saylan *et al.* 1986). The positive rate of a pathergy test was once reported as an even lower occurrence rate than that of minor symptoms (Kim *et al.* 1988). In addition, it was claimed that the test had a decreased positivity and was influenced by the diameter of the needle (Ozarmagan *et al.* 1991). Again we prefer to use the revised Shimizu's criteria (Shimizu *et al.* 1974), which requires the presence of four major mucocutaneous signs regardless of the results of a pathergy test.

Sex distribution

The male to female ratio in Korea was previously

reported to be between 0.94:1 and 0.71:1 (Lee, 1979; Eun *et al.* 1984; Cho *et al.* 1988; Kim *et al.* 1988). In our study, the sex ratio was 0.63:1. These ratios did not exactly coincide with one another, but female predominance was commonly noted in the occurrence of Behçet's disease in Korea. Exceptionally, the sex ratio was reversed to male predominance (1.05:1) in complete type in our study. With a few exceptions in studies from the U.S. and Britain (Wong *et al.* 1984), most countries reported a male to female ratio of 1-2:1 with male predominance (Baserer *et al.* 1979; Dilsen *et al.* 1979; Nakae *et al.* 1993). On the other hand, the sex ratio in Chinese patients was 0.77:1 (Ohno, 1986). The fact that female predominance was commonly noted in both Korean and Chinese patients suggested that genetic factors were important in expressing the disease (Cho *et al.* 1988). But it may be possible that the pathogenesis of Behçet's disease is also influenced by environmental factors because Japanese patients who are closely related genetically and racially did not show female predominance and the detection rate of HLA-B5 is commonly high in Japanese, English, Turkish, and in some others who did not show female predominance (Lee *et al.* 1988).

Age distribution

The age of onset has been most commonly reported in the 20s (Baserer *et al.* 1979; Dilsen *et al.* 1979; Shimizu *et al.* 1979; Wong *et al.* 1984). In our study, the age of onset was most common in the 20s (37.1%) with a mean age of 29.7 years in males and 28.2 years in females, which showed significant difference between the two sexes. In contrast with Japanese data that showed no significant sexual difference in the age of onset, there was a significant difference in the ages of onset between males and females in Korea (Oshima *et al.* 1963).

The age at the time of diagnosis has been known to be most commonly in the 30s or 40s (Baserer *et al.* 1979; Dilsen *et al.* 1979; Shimizu *et al.* 1979; Nakae *et al.* 1993). And this was the same in our data.

Therefore the age distribution of Behçet's disease in Korea showed similar features with those reported in other countries.

Oral ulcer

Oral ulcer was not only the most frequent initial manifestation (78.5%) but also the most common clinical manifestation (97.5%) noticed in our patients with Behçet's disease. Previous studies reported that the frequency of oral ulcer was 52-78% (Oshima *et al.* 1963; Lehner, 1967; Mason and Barnes, 1969; Chajek and Fainaru, 1975). Recently the frequency of oral ulcer in Japanese and Chinese patients was reported as 98.2% and 100%, respectively (Dong *et al.* 1991; Nakae *et al.* 1993) (Table 5).

Recurrent oral ulceration is a common clinical manifestation of many disorders and also the most frequent initial symptom of Behçet's disease. Because recurrent oral ulceration in Behçet's disease cannot be distinguished from those in other disorders on the basis of the clinical characteristics of oral ulcers, recurrent oral ulcerations without any other manifestation of Behçet's disease have resulted in a diagnostic problem. According to the Behçet's Disease Research Committee of Japan (Shimizu *et al.* 1979), recurrent oral ulceration without any other manifestation was formalized as a possible type of Behçet's disease; however, an international study group suggested a new diagnostic criteria which excluded recurrent oral ulceration without other manifestations as a diagnostic criterion for Behçet's disease (International Study Group for Behçet's disease, 1990). In our previous studies we found that the average time interval between the initial oral ulceration and the second major manifestation of Behçet's disease was 6.8 years (Cho *et al.* 1988), and 52.2% of patients with recurrent oral ulceration alone developed overt manifestations of Behçet's disease over an average of 7.7 years (Lee *et al.* 1993; Bang *et al.* 1995). Therefore oral ulceration with a high frequency of recurrence should be considered as a warning signal of Behçet's disease.

Oral ulcers were found at any site in the oral cavity, and minor ulcers were most frequently encountered in our patients.

Recurrent tonsillitis, smoking, dental caries, and alcohol consumption were known to be irritating factors affecting the oral cavity (Cho *et al.* 1988).

Genital ulcer

The facts that genital ulcers are more common in females than in males and that genital ulcers tend to become worse during the premenstrual stage led us to speculate that hormonal factors may be closely related to the pathogenesis of Behçet's disease. And this is demonstrated by a report that showed possible influences of immunologic and hormonal factors on the clinical course of Behçet's disease during pregnancy (Bang *et al.* 1993).

Cutaneous manifestations

Cutaneous involvement was the second most common symptom in our patients with Behçet's disease. An erythema nodosum-like lesion was the most frequent skin lesion. The order of frequency of skin lesions coincided with other studies (Shimizu *et al.* 1979; Rhim *et al.* 1980; Eun *et al.* 1984). Although cutaneous manifestations of Behçet's disease are protean and not characteristic of the disorder, the high incidence of skin lesions makes them a major criterion in the diagnosis of Behçet's disease.

Ocular symptom

Ocular symptoms were found in 28.5% of the patients in our study. In 1992 Kang and Kim analyzed ocular symptoms in 1,454 patients with Behçet's disease. According to their study, the mean duration between the initial symptom and ocular symptom was 44 months. Ocular involvements were bilateral in more than half of the cases as reported in other studies (Colvard *et al.* 1977; Michelson and Chisari, 1982). The most common ocular symptom was iritis (64.0%). Other findings were occlusive vasculitis (34.2%), cataract (23.1%), chorioretinitis (19.6%), retinal degeneration (13.9%), glaucoma (6.9%), retinal hemorrhage (5.2%), and retinal detachment (0.5%). About 30% of patients had corrected visual acuity of 0.01 or worse on their first visit. It was known that 74% of patients with Behçet's disease lost their visual ability within 6 to 10 years (BenEzra and Cohen, 1986), and this was similar to the situation in Korean patients whose visual acuities deteriorated in 50.9% of the eyes followed over 24 months.

General features of major and minor symptoms

As shown in our data, the relative frequency of ocular lesion and genital ulcer is somewhat lower than in other studies (Baserer *et al.* 1979; Dilsen *et al.* 1979; Shimizu *et al.* 1979). The lower rate of articular involvement (24.2%) also seems to be unusual compared with another report (Mason and Barnes, 1969).

A comparison of data taken from Korean, Japanese, and Chinese patients showed some differences among these 3 countries (Dong *et al.* 1991; Nakae *et al.* 1993) (Table 5). Genital ulcer, gastrointestinal symptom, and neurologic symptom were more common in both Japan and China than in Korea. Skin lesion, ocular symptom, and articular symptom were more common in Japan than in the other 2 countries. The positive rate of pathergy test was remarkably higher in China than in the other 2 countries. In Japan genital ulcers were found in females 2 times more than in males but ocular symptoms were more frequently found in males (Nakae *et al.* 1993). Apart from oral ulcer the reason for the lower frequency of major symptoms than was found in other countries was thought to be due to the inclusion in our data of patients with suspected or possible type of Behçet's disease.

It was shown that patients with a shorter interval before manifesting a second symptom had an increased risk of becoming the more serious type, and this was reinforced by another report (Cho *et al.* 1988).

Family history

The reports concerning familial occurrence of Behçet's disease are rare (Lehner, 1967; Mason and Barnes, 1969). In our data family history was detected in 15.4% of patients, whose siblings and mothers were frequently affected.

Although correlation between HLA antigens and pathogenesis of Behçet's disease is still obscure, a highly significant association of HLA-B5 has been found (Takano *et al.* 1976; Erosy *et al.* 1977; Brautbar *et al.* 1978; Adorno *et al.* 1979; Hamza *et al.* 1979; Lehner *et al.* 1982; Ohno *et al.* 1982). In our previous report, HLA-B5 and DRw3 presented significantly high frequencies in all of patients with Behçet's disease examined, and HLA-B5 was asso-

Table 17. Frequency of major symptoms in childhood-onset patients in Korea and Japan*

Symptom	Korea(n=40)		Japan(n=31)	
	No.	%	No.	%
Oral ulcer	40	100.0	31	100.0
Genital ulcer	33	82.5	18	58.1
Skin lesion	29	72.5	17	54.8
Ocular lesion	11	27.5	8	25.8

*: modified from references: Kim *et al.* 1994; Fujikawa and Suemitsu, 1997

ciated with the severity of the disease (Lee *et al.* 1988).

Behçet's disease in children

Behçet's disease is very uncommon in children and there is little in the literature on this aspect (Shimizu *et al.* 1974; Mundy and Miller, 1978; Lang *et al.* 1990; Kim *et al.* 1994). According to Kim *et al.* the male to female ratio was 0.67:1 and the most frequent major sign was oral ulcer (Kim *et al.* 1994) (Table 17). The average time interval between the initial oral ulcer and the second symptom was 8.8 years. After the second major manifestation, the third and fourth features rapidly developed within one or two years. According to another report complete type was 19.3% of 57 childhood patients with Behçet's disease, incomplete 33.3%, and suspected type 47.4% (Cho *et al.* 1989). Since the most common type was suspected type, this is the most characteristic feature of early onset Behçet's disease distinguishable from adult onset Behçet's disease. Probably this difference is due to the fact that suspected type may progress to more severe types later. In Japanese children with Behçet's disease, complete type was found in 9.7% of patients, incomplete type was 77.4%, and possible type was 12.9%. Oral ulcer was the most frequent major manifestation and gastrointestinal signs were more frequent in children than in adults, while ocular symptoms were less frequent (Fujikawa and Suemitsu, 1997).

REFERENCES

Adorno D, Pezzi PP, Bonini S, Masala C, Bonini S, Amendolea MA, Casciani CU: HLA-B5 and Behçet's disease. *Tissue Antigens 14: 444-448, 1979*

Bang D, Haam IB, Lee ES, Lee S: *The influence of pregnancy on Behçet's disease.* In Godeau P, Wechsler B, eds. *Behçet's disease.* Amsterdam, Excerpta Medica, *1993, 163-166*

Bang D, Hur W, Lee ES, Lee S: Prognosis of recurrent oral ulceration in Behçet's disease. *J Dermatol 22: 926-929, 1995*

Baserer T, Mirzatas C, Basar B: *A clinical study of 147 patients with Behçet's disease.* In Dilsen N, Konice M, Övül C, eds. *Behçet's disease.* Amsterdam-Oxford, Excerpta Medica, *1979, 120-123*

Behçet H: Über rezidivierende, aphthöse durch ein Virus verursachte Geschwüre am Mund, am Auge und an den Genitalen. *Dermatol Wochenschr 36: 1152-1157, 1937*

BenEzra D, Cohen E: Treatment and visual prognosis in Behçet's disease. *Br J Ophthalmol 70: 589-592, 1986*

Brautbar C, Chajek T, Ben-Tuvia S, Lamm L, Cohen T: A genetic study of Behçet's disease in Israel. *Tissue Antigens 11: 113-120, 1978*

Chajek T, Fainaru M: Behçet's disease. Report of 41 cases and a review of the literature. *Medicine 54: 179-196, 1975*

Cho MY, Lee S, Bang D, Lee ES, Lee SH: *Clinical analysis of 57 cases of Behçet's syndrome in children.* In O'Duffy JD, Kokmen E, eds. *The Fifth International Conference on Behçet's Disease.* Minnesota, Rochester, Marcel Dekker, Inc, *1989, 41-47*

Cho MY, Lee SH, Bang D, Lee S: Epidemiologic findings of Behçet's syndrome. *Korean J Dermatol 26: 320-329, 1988*

Colvard DM, Robertson DM, O'Duffy JD: The ocular manifestations of Behçet's disease. *Arch Ophthalmol 95: 1813-1817, 1977*

Dilsen N, Konice M, Övül C: *Clinical evaluation of 106 cases of Behçet's disease.* In Dilsen N, Konice M, Övül C, eds. *Behçet's disease.* Amsterdam-Oxford, Excerpta Medica, *1979, 124-129*

Dong Y, Ming QX, Zhang NZ, Li CH, Wu QY: *Testing different diagnostic criteria of Behçet's syndrome in Chinese patients.* In O'Duffy JD, Kokmen E, eds. *Behçet's disease: basic and clinical aspects.* New York, Marcel Dekker, Inc, *1991, 55-59*

Erosy F, Berkel AI, Firat T, Kazokoglu H: HLA antigens associated with Behçet's disease. *Arch Dermatol 113: 1720-1721, 1977*

Eun HC, Chung H, Choi SJ: Clinical analysis of 114 patients with Behçet's disease. *J Korean Med Assoc 27: 933-939, 1984*

Fujikawa S, Suemitsu T: Behçet's disease in children: a nationwide retrospective survey in Japan. *Acta*

Paediatrica Japonica 39: 285-289, 1997

Hamza M, Sohier R, Betuel H, Ayed HB: *HLA-B5 and Behçet's disease.* In Dilsen N, Konice M, Övül C, eds. *Behçet's disease.* Amsterdam-Oxford, Excerpta Medica, *1979, 265*

Hong WP, Park IY, Cung MH, Lee JG, Park HJ: Clinical and immunological study on oral lesions of Behçet's syndrome. *Korean J Otolaryngol 28: 57-62, 1985*

International study group for Behçet's disease: Criteria for diagnosis of Behçet's disease. *Lancet 335: 1078-1080, 1990*

Joo CR: Two cases of Behçet's syndrome. *J Cathol Med College 5: 393-400, 1961*

Kang HJ: Behçet's syndrome. *Korean J Dermatol 9: 25-30, 1971*

Kang SJ, Kim HB: Behçet's disease in Korean. *J Korean Ophthalmol 33: 118-127, 1992*

Kim DK, Chang SN, Bang D, Lee ES, Lee S: Clinical analysis of 40 cases of childhood-onset Behçet's disease. *Pediatr Dermatol 11: 95-101, 1994*

Kim HJ, Bang D, Lee SH, Yang DS, Kim DH, Lee KH, Lee S, Kim HB, Hong WP: Behçet's syndrome in Korea-a look at the clinical picture. *Yonsei Med J 29: 102-108, 1988*

Lang BA, Laxer RM, Thorner P: Pediatric onset of Behçet's syndrome with myositis. *Arthritis Rheum 33: 418-425, 1990*

Lee ES, Hur W, Bang D, Lee SH, Lee S, Kim HB, Hong WP: *Prognosis of recurrent oral ulceration in Behçet's disease.* In Godeau P, Wechsler B, eds. *Behçet's disease.* Amsterdam, Excerpta Medica, *1993, 167-170*

Lee JK: Statistical study of Behçet's disease. *J Korean Ophthalmol Soc 20: 167-174, 1979*

Lee S, Koh YJ, Kim DH, Bang D, Nam IW, Lee KH, Park K: A study of HLA antigens in Behçet's syndrome. *Yonsei Med J 29: 259-262, 1988*

Lehner T: Behçet's syndrome and autoimmunity. *Br Med J 1: 465-467, 1967*

Lehner T: Oral ulceration and Behçet's syndrome. *Gut 18: 491-511, 1977*

Lehner T, Barnes CG: *Criteria for diagnosis and classification of Behçet's syndrome.* In Lehner T, Barnes CG, eds. *Behçet's syndrome.* New York, Academic press, *1979, 1-9*

Lehner T, Welsh KL, Bachelor JR: The relationship of HLA-B and DR phenotypes to Behçet's syndrome, recurrent oral ulceration and the class of immune complexes. *Immunology 47: 581-587, 1982*

Mason RM, Barnes CG: Behçet's syndrome with arthritis. *Ann Rheum Dis 28: 95-103, 1969*

Michelson TB, Chisari FV: Behçet's disease. *Surv Ophthalmol 26: 190-203, 1982*

Mundy TM, Miller JJ: Behçet's disease presenting as chronic aphthous stomatitis in a child. *Pediatrics 62: 205-208, 1978*

Nakae K, Masaki F, Hashimoto T, Inaba G, Mochizuki M, Sakane T: *Recent epidemiological features of Behçet's disease in Japan.* In Godeau P, Wechsler B, eds. *Behçet's disease.* Amsterdam, Excerpta Medica, *1993, 145-151*

Ohno S: *Behçet's disease in the world.* In Lehner T, Barnes CG, eds. *Recent advances in Behçet's disease.* London, Royal Society of Medicine Services, *1986, 181-186*

Ohno S, Aoki K, Sugiura S, Nakayama E, Itakura K, Aizawa M: Close association of HLA-Bw51 with Behçet's disease. *Arch Ophthalmol 100: 1455-1458, 1982*

Oshima Y, Shimizu T, Yokohari R, Matsumoto T, Kano K, Kagami T, Nagaya H: Clinical studies on Behçet's syndrome. *Ann Rheum Dis 22: 36-45, 1963*

Ozarmagan G, Saylan T, Azizlerli G: Re-evaluation of pathergy test in Behçet's disease. *Acta Derm Venereol 71: 75-76, 1991*

Park BI: Behçet's disease. *J Korean Ophthalmol Soc 14: 114-151, 1973*

Rhim KJ, Choi JS, Son SJ: Clinical observation on Behçet's syndrome. *Korean J Dermatol 18: 561-571, 1980*

Shimizu T, Ehrlich GE, Inaba G, Hayasi K: Behçet's disease. *Semin Arthritis Rheum 8: 233-260, 1979*

Shimizu T, Inaba G, Hashimoto T: Diagnostic criteria and their problems of Behçet's disease. *Intern Med 33: 278-282, 1974*

Takano M, Miyajima T, Kiuchi M, Ohmori K, Amemiya H, Yokoyama T, Hashizume H, Iwasaki Y, Okamoto S, Sato H: Behçet's disease and the HLA system. *Tissue Antigens 8: 95-99, 1976*

Wong RC, Ellis CN, Diaz LA: Behçet's disease. *Int J Dermatol 23: 25-32, 1984*

Yoon MS, Lee SH, Bang D, Lee S: Cutaneous manifestations of Behçet's syndrome. *Yonsei Med J 28: 291-296, 1987*