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CLINICAL STUDY

Uveitis in juvenile Behçet's disease: clinical course and visual outcome compared with adult patients

M Kramer[1,6], R Amer[2,3], M Mukamel[4,6], M Snir[5,6], T Jaouni[2] and R Friling[5,6]

[1]Department of Ophthalmology, Rabin Medical Center, Petach Tikva, Israel

[2]Department of Ophthalmology, Hadassah Medical Center, Jerusalem, Israel

[3]Hadassah School of Medicine, Hebrew University, Jerusalem, Israel

[4]Pediatric Rheumatology Unit, Schneider Children's Medical Center of Israel, Petach Tikva, Israel

[5]Pediatric Ophthalmology Unit, Schneider Children's Medical Center of Israel, Petach Tikva, Israel

[6]Sackler School of Medicine, Tel Aviv University, Tel Aviv, Israel

Correspondence: M Kramer, Department of Ophthalmology, Rabin Medical Center, Petah Tikva 49100, Israel
Tel: +972 3 937 6101; Fax: +972 3 921 9084.
E-mail: mkramer@netvision.net.il

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Abstract

Purpose **To characterize the disease course and visual outcome of uveitis in juvenile Behçet's disease (BD) compared with adults.**
Methods **The study population included 13 children (mean age 14 ± 2.4 years; 22 eyes) and 16 adults (mean age 30 ± 8.8 years; 27 eyes) with uveitis in BD diagnosed between 1997 and 2007.**
Results **The male/female ratio was 1.6:1 in the paediatric group and 3:1 in the adult group. Five children (38%) and four adults (25%) had complete BD. Mean duration of follow-up for both groups was 4.7 years. The children had more acute exacerbations (4.1 ± 2.7 *vs* 2.3 ± 1.5, $P = 0.054$). Treatment in both groups included systemic steroids and immunosuppressive agents. In children, mean initial visual acuity in the affected eyes ($n = 22$) was 0.6 ± 0.7 logMAR (range, 0–2.2). It decreased during exacerbations in 15 eyes (68%; mean, 1.6 ± 0.8 logMAR), severely reduced (worse than 1 logMAR) in 11 eyes (50%; mean, 2.0 ± 0.45 logMAR), and improved significantly in 12 of 13 promptly treated eyes (92%; 6/12 or better in 11; mean 0.2 ± 0.4 logMAR, $P < 0.001$). The visual outcome pattern was similar in the adults.**
Conclusions **Uveitis in juvenile BD is characterized by frequent exacerbations of explosive nature with profoundly reduced visual acuity. Similar disease pattern was observed in children and adults, as well as in patients with complete or incomplete disease. Early diagnosis, even before all systemic criteria are fulfilled, is important because early aggressive therapy can achieve long-term useful visual acuity.**

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Keywords: Behçet's disease; juvenile; uveitis

Introduction

Behçet's disease (BD) usually affects men in the second to fourth decade of life. The disease is uncommon in childhood, though it may occasionally present in incomplete form before the age of 16 years.[1–3] Because the disease is characterized by exacerbations and remissions, the complete form may be expressed only in adulthood.[4]

Uveitis is one of the major criteria for diagnosis of BD, occurring in 70 to 95% of patients.[5] In children, the reported rate of ocular involvement ranges from 27 to 80%.[3,4] Although not usually the presenting sign of BD (10–13%), ocular involvement in rare instances may serve as the only sign; these cases were classified as 'possible BD' by the Behçet's Disease Research Committee of Japan.[6] The International Study Group for Behçet's Disease (ISG) requires the presence of oral aphthae for diagnosis.[7]

Because BD is pathophysiologically an occlusive vasculitis that progresses by attacks and remissions, every attack of uveitis may cause irreversible damage to the eye, ultimately leading to blindness. In children, there is a 23% reported rate of significant deterioration of vision,[3] and thereby early diagnosis followed by aggressive treatment is of utmost importance. The aim of this study was to characterize the clinical features, course, and outcome of uveitis in juvenile BD (JBD) and compare the clinical course to adult patients.

Materials and methods

The files of patients with uveitis and BD at the age of 16 years or younger, treated between 1997 and 2007 at two general tertiary medical centres and one paediatric medical centre in Israel, were reviewed for diagnostic criteria, ocular examination, course of disease, complications, treatment, and visual outcome. The diagnosis in all cases was based on the criteria of the ISG[7] and the Behçet's Disease Research Committee of Japan.[6]

Eyes with active inflammation were referred as 'affected eyes'. Decrease or improvement in visual acuity (VA) was defined as ±0.1 change in logMAR. Severe decrease in VA was defined as VA of 1 logMAR or worse. Remission was defined as 'at least 6 months with no signs of active disease.'

These findings were compared with the data derived from the files of adult patients who were treated at the same general medical centres during the same time period. The cutoff age of 16 years was based on earlier studies of paediatric uveitis.[1–3,8] The research described adhered to the tenets of the Declaration of Helsinki and was approved by the Institutional Review Board.

Statistical analysis

Student's *t*-test was used for analysing visual outcome and the number of exacerbations. When the samples were small ($n<10$), the non-parametric Wilcoxon signed-rank test was used. Comparison between the groups for the other parameters was performed using χ^2-test.

Results

Thirteen children (22 eyes) and 16 adults (27 eyes) were diagnosed with uveitis in BD between 1997 and 2007. The paediatric group was consisted of eight male subjects and five female subjects (1.6:1). Mean age at presentation of ocular involvement was 14 ± 2.4 years (median, 15 years; range, 9–16); nine of them (69%) were aged 14–16 years. Eleven (85%) children were Jewish of Mediterranean origin and two (15%) were Arabs. The adult group consisted of 12 male subjects and four female subjects (3:1). The mean age of ocular presentation was 30 ± 8.8 years (range, 17–44). Nine (56%) were Jewish (eight from the Mediterranean region and one Romanian) and seven (44%) were Arabs. Mean follow-up time was 4.7 ± 2.8 years in the paediatric group and 4.8 ± 3.2 years in the adult group (range, 1–10 years for both). According to the ISG criteria, five children (38%) and four adults (25%) had complete BD. Eight children had uveitis and either recurrent oral or genital aphthae. The remainder of adult patients had uveitis with 1–2 additional signs, except for two patients diagnosed as possible BD. The demographic data and systemic features of the paediatric patients are summarized in Table 1.

HLA-B5 positivity was documented in 9 of the 12 children tested (75%) and 12 of the 15 adults tested (80%); no data were available for one patient in each group.

In the paediatric group, all patients had a history of recurrent aphthae, 12 oral and four genital. Albeit this, BD (complete or incomplete) was diagnosed at the time of appearance of the ocular involvement in 10 cases (77%), 6–12 months after the appearance of ocular

Table 1 Demographic and systemic features of paediatric patients

Patient no./sex	*Age at presentation*	*Time (years) between uveitis and diagnosis*	*Systemic signs*[a]	*HLA B5*	*Diagnosis: ISG*	*Diagnosis: RCJ*[b]
1/M	14	0	OA, pericarditis, elevated ICP	−	Uveitis + OA	Incomplete (2 M, 2 m)
2/M	13	0.5	OA	−	Uveitis + OA	Incomplete (2 M)
3/F	9	0	OA	−	Uveitis + OA	Incomplete (2 M)
4/M	10	0	OA, EN pulmonary embolism	+	Complete	Incomplete (3 M)
5/M	11	−2	OA, EN, arthralgia, pathergy +	+	Complete	Incomplete (3 M)
6/F	16	1	OA	+	Uveitis + OA	Incomplete (2 M)
7/M	16	0	OA	+	Uveitis + OA	Incomplete (2 M)
8/M	15	0	OA, GA, arthralgia	+	Complete	Incomplete (2 M)
9/F	16	0	OA, arthralgia	?	Uveitis + OA	Incomplete (2 M)
10/M	16	0	OA, GA	+	Complete	Incomplete (3 M)
11/M	16	0	OA	+	Uveitis + OA	Incomplete (2 M)
12/M	15	0	GA, Phlebitis	+	Uveitis + OA	Incomplete (2 M, 1 m)
13/M	15	0	OA, GA	+	Complete	Incomplete (3 M)

ISG = International Study Group; RCJ = Research Committee of Japan; *OA = oral aphthae; EN = erythaema NODOSUM; ICP = intracranial pressure.
[a]OA, EN, and ICP.
[b]M = major, m = minor.

symptoms in two cases, and 2 years before their appearance in one case only. In the adult group, seven patients (44%) were diagnosed at the time of appearance of ocular involvement ($P = 0.07$) and the others, from 6 months (six patients) to 2–4 years (three patients) later.

The clinical uveitis data are summarized in Tables 2 and 3. In the paediatric group, nine (69%) patients had panuveitis, three (23%) had posterior uveitis, and one (8%) had isolated anterior uveitis. In the adult group, nine (56%) patients had panuveitis and seven (44%) had posterior uveitis, including retinal vasculitis, retinitis, and neuroretinitis.

The ocular disease was remarkable for sudden, hyperacute exacerbations causing reduced VA, sometimes despite treatment. These recurrent attacks occurred in 9/13 (70%) paediatric group and in 8/16, (50%) adult group (NS). In the paediatric group, the mean number of acute exacerbations before remission was 4.1 ± 2.7 (range, 1–8), and the mean interval between exacerbations (corrected for variable duration of follow-up) was 1.4 ± 0.8 years (range, 4 months to 2.5 years). The mean number of attacks in the adult group was 2.3 ± 1.5. ($P = 0.054$) with mean interval between attacks of 2.9 ± 3.2 (NS). In both groups, reduced VA was caused by

Table 2 Ocular findings in children

Patient no.	*Laterality*[a]	*Type of uveitis*	*Clinical findings*					
			Anterior	*Vitritis*	*Retinal vasculitis*	*Retinitis*	*Macular oedema*	*ON involvement*
1	2	Panuveitis	+	+				+
2	2	Panuveitis		+	+			
3	2	Panuveitis	+	+ + +	+ +[b]	+		
4	2	Ant. uveitis	+ +					
5	2	Panuveitis	+ +	+	+ +	+ +	+ +	+ +
6	1	Panuveitis	+	+				
7	2	Panuveitis	+ +	+ + +		+		
8	1	Panuveitis	+ +	+ +	+	+	+ +	+ +
9	1	Post. uveitis			+ +[b]		+ +	
10	2	Post. uveitis		+		+		
11	1	Panuveitis	+ +	+	+	+		+ +
12	2	Post. uveitis		+	+	+		+
13	2	Panuveits	+ + +	+ +		+		

[a]Laterality: 1 = unilateral, 2 = bilateral.
[b]Multiple haemorrhages.

Table 3 Ocular findings in adults

Patient No	*Laterality*[a]	*Type of uveitis*	*Clinical findings*					
			Anterior	*Vitritis*	*Retinal vasculitis*	*Retinitis*	*Macular oedema*	*ON involvement*
1	1	Panuveitis	+	+ + +	+	+		+
2	2	Post. uveitis		+	+ +			
3	2	Panuveitis	+	+ +	+ +	+	+	+
4	2	Panuveitis	+	+	+ +	+ + +	+ +	
5	2	Panuveitis		+ +	+ +	+ + +		+
6	2	Postuveitis				+		
7	1	Panuveitis	+	+ +				+ +
8	2	Postuveitis			+	+		
9	2	Post. uveitis		+ +	+		+ +	
10	2	Post. uveitis		+	+		+	
11	2	Panuveitis	+	+ +	+ + +			
12	2	Panuveits	+	+	+			
13	2	Post. uveitis		+ +	+			
14	1	Panuveitis	+	+ +	+ +	+ +	+ +	+ +
15	2	Post. uveitis		+ +			+ +	+ +
16	2	Panuveitis	+	+			+ +	+ +

[a]Laterality: 1 = unilateral, 2 = bilateral.

posterior segment involvement, namely, severe vitritis, retinitis, retinal vasculitis, and optic nerve involvement.

Table 4 outlines the systemic treatment. Twelve of thirteen patients in the paediatric group were treated with systemic steroids. Only one patient with mild disease, who was lost to follow-up after 2 years, was treated with topical and periocular steroids only. Eleven patients (85%) were treated with additional immunosuppressive agents and six of them (55%) required multiple drug combinations. Immunosuppressive agents were prescribed to all patients, shortly after the diagnosis was made. Intravitreal triamcinolone injections were used repeatedly in two patients, and one patient received intravitreal bevacizumab injection for neovascularization. Compliance was a major problem in the adolescent patients.

Table 4 Systemic medications used in children and adult patients

Systemic treatment	*Children* (n = *13*)	*Adults* (n = *16*)
Corticosteroids		
Oral	12 (92%)	16 (100%)
Intravenous[a]	5	
Immusuppressive agents		
Total	11 (85%)	16 (100%)
Combinations	6 (55%)	5 (31%)
Methotrexate	8	7
Cyclosporine	5	8
Azathioprine	3	5
Mycophenolate mofetil	1	—
Inifliximab	2	1
Interferon α	1	—
Cyclophosphamide[b]	1	—

[a]Intravenous pulse therapy was given during acute exacerbations.
[b]Cycliphosphamide was given due to systemic indications

Similarly, all adult patients received systemic steroids and immunosuppressive agents. Five patients (31%) required multiple agents. One patient received intravitreal triamcinolone injections.

Mean time of active disease in the paediatric group was 3.3 ± 2.4 years (range, 0.5–8, median 3.0 years) and 4.2 ± 2.9 years in the adult group (range, 0.2–10) (NS). Eleven children (85%) and 13 adults (81%) were on remission for at least 6 months; of them, eight children (73%) and 12 adults (92%) were on remission for 12 months or more. Two children and three adults were still active under treatment at the last visit.

Ocular complications are presented in Table 5. Cataract was the most common anterior segment complication in the paediatric group (five patients, 6 of 22 eyes, 27%). Four of them underwent cataract surgery (one bilaterally). Posterior segment complications occurred in five children (eight eyes, 36%): Early posterior segment complications consisted of active retinitis in the macular

Table 5 Complications in the paediatric and adult patients in our study, compared to previously published data

Complications	*Our study*		*Literature (Tugal-Tutkun* et al)[3,16]	
	Children (n = *22 eyes*)	*Adults* (n = *27 eyes*)	*Children*	*Adults*
Cataract	6 (27%)	7 (26%)	47%	38%
IOP	1 (4%)[b]		11%	14%
Posterior Segment[a]	8 (36%)	13 (48%)		
Early				
Active retinitis in macula	4	—		
Late				
ERM	2[c]	3	45%	17%
CME	—	3		44%
Macular scar	—	1		19%
Macular hole	—	1		
Retinal NV	1[d]	2[c,d]		
RD	1[d,e]	2[c,d]		
Pthysis	1[e]	—		
Optic atrophy	2 (9%)	2 (7%)	39%	24%

IOP = intraocular pressure; ERM = epiretinal membrane; CME = cystoid macular oedema; NV = neovascularization; IVTA = intravitreal triamcinolone acetonide.RD = retinal detachment.
[a]Some of the posterior segment complications occurred simultaneously in the same eyes.
[b]Following IVTA.
[c]Two eyes of the same patient.
[d]Same eye.
[e]Two eyes of same patient who was non-compliant with treatment and inconsistent with follow-up before these complications.

area, and late complications included epiretinal membrane (ERM), retinal neovascularization, and partial optic atrophy. Phthisis and retinal detachment with rubeosis were found in one eye each of the same patient following bilateral vitrectomy with additional bevacizumab injection (this patient was non-compliant with treatment and inconsistent with follow-up before complications) In the adult group, cataract occured in five patients (seven eyes, 26%), of whom three underwent cataract surgery (one bilaterally and one combined with vitrectomy). Posterior segment complications occurred in 10 patients (13 eyes, 48%) and included ERM, cystoid macular oedema, macular scars, macular hole, and retinal neovascularization, causing retinal detachment. Optic atrophy was found in two patients (two eyes).

Figure 1 shows the relation between initial VA upon presentation and VA outcome at the end of the follow-up period. Figure 2 shows the relation between the worst VA during active disease and VA outcome at the end of the follow-up period. Both figures include data for children and adults. In the paediatric group, mean initial VA in the affected eyes ($n = 22$) was 0.6 ± 0.7 logMAR (range, 0–2.2, Snellen equivalent 6/6 hand motion). Fifteen (68%) eyes had further reduced VA during exacerbations, 11 (73%) of which were severely reduced (more than 1 logMAR, Snellen equivalent less than 6/60) (range, 1/20 to NLP, mean 2.0 ± 0.45 logMAR). Of the 15 eyes, 12 (80%) improved after therapy, including 8 of the 11 eyes with a severe reduction. The three non-improving eyes included two eyes of one non-compliant patient and one eye, which developed permanent damage and was diagnosed incidentally. Thus, improvement rate of eyes with reduced VA following prompt treatment was 12 of 13 (92%). The difference in mean VA between eyes with reduced vision to last remission (0.2 ± 0.4 logMAR) was statistically significant ($P < 0.001$); this was also true for the subgroup with severe VA reduction during attacks (lowest mean VA, 1.9 ± 0.4 logMAR; final mean VA,

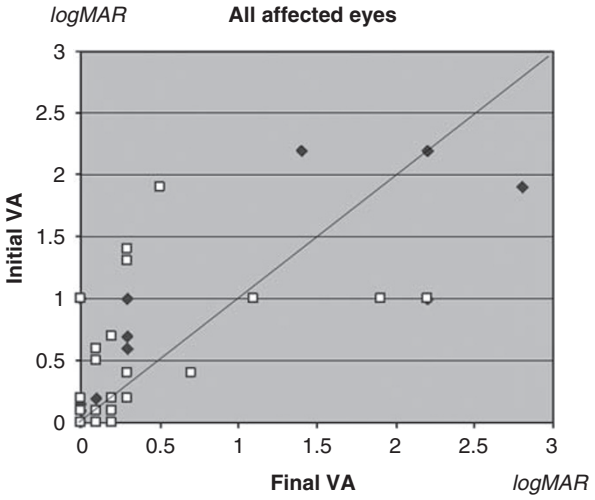

Figure 1 Initial *vs* final VA—children and adults. Initial VAs of the affected eyes are plotted against final VAs. Improved eyes are represented by the spots above the line. Dark diamonds represent data of children's eyes; white rectangles represent data of adult's eyes. Note that most spots are plotted above the line, indicating improvement.

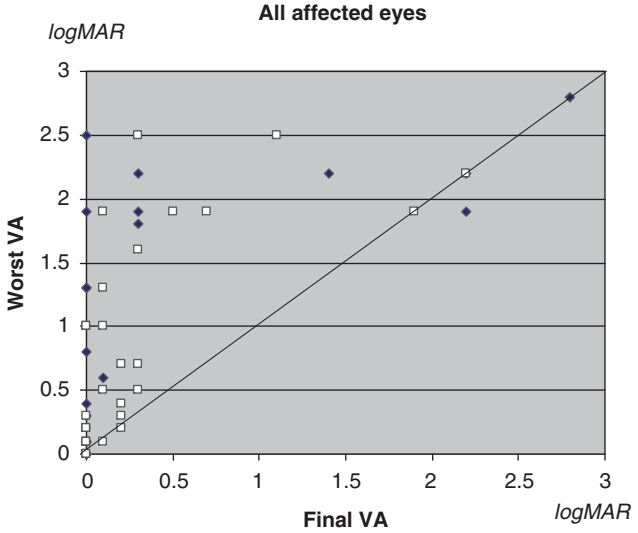

Figure 2 Worst *vs* final VA—children and adults. Worst VAs of affected eyes, during active disease, are plotted against final VAs. Improved eyes are represented by the spots above the line. Dark diamonds represent data of children's eyes; white rectangles represent data of adult's eyes. One spot under the line represents data of both a child and an adult's eye. There are less spots than the number of eyes (children = 22, adults = 29), as some of the spots represent similar data, and thus are overlapping. Note that most spots are plotted above the line, indicating improvement.

0.3 ± 0.45 logMAR, $P < 0.001$). Upon remission, all but one of the improved eyes had a VA of 0.3 logMAR or better (Snellen equivalent 6/12; $P = 0.01$).

A similar pattern was observed in the adult group. Mean initial VA in affected eyes ($n = 29$) was 0.44 ± 0.5 logMAR (range, 0–1.9, Snellen equivalent 6/6 counting fingers). VA was further reduced in 14 patients (25 eyes; mean 0.88 ± 0.8 logMAR, Snellen equivalent 6/7.5 light perception). Following treatment, an improvement was noted in all 25 eyes, including all nine eyes with severe reductions (mean 1.8 ± 0.6 logMAR, Snellen equivalent range, 6/60—no light perception). Mean final VA was 0.2 ± 0.3 logMAR (Snellen equivalent range, 6/6–1/12; $P = 0.001$). Twenty-two (88%) eyes with reduced VA during attacks achieved a VA of 0.3 logMAR or better (Snellen equivalent, 6/12). The final mean VA of the nine eyes with the most severe reduction was 0.35 ± 0.55 logMAR, which was significantly better than their lowest VA ($P < 0.001$).

Overall, 4 of the 22 affected eyes in the paediatric group (18%) sustained irreversible damage and low VA. As mentioned before, one patient was poorly treated because of non-compliance and inconsistent follow-up. Excluding this patient, 2 of the 20 affected eyes in the paediatric group (10%), who presented with severe visual loss, showed either partial or no response to treatment, thus sustaining irreversible damage and VA worse than 1 logMAR. Similar findings were present in three eyes of two adult patients (12%). One of these eyes was severely affected and improved significantly, still achieving final VA of worse than 1 logMAR, and two eyes developed irreversible damage before treatment.

Discussion

This study compared the clinical course and visual outcome of chronic relapsing uveitis in BD between children (age 16 years or less) and adults treated in the same departments during the same period. To the best of our knowledge, this is the first study to follow longitudinally the severe fluctuating clinical course of uveitis in JBD, providing quantitative analysis of the visual changes, and compare them to adult patients. Most previously reported clinical series evaluated the systemic disease, thus only mentioning ocular involvement.[9–11] Only few studies have been specifically directed at childhood-onset uveitis in BD.[2,3,12] The clinical study published by Tugal-Tutkun *et al*[3] reported 36 patients over 27 years. The rates of ocular findings were similar to our study. We shared the same aggressive treatment strategy, which included systemic steroids and early immunosuppression. However, we limited our study to patients diagnosed within the last 10 years, to decrease variability in management, and compare children and adults with similar disease duration. In addition, though our study was performed in tertiary centres, our uveitis clinics serve also as secondary centres, thus patients are referred mostly during the early stages of the disease.

Owing to the lack of a definitive diagnostic test, the diagnosis of BD is determined according to the clinical criteria, using various sets of criteria.[6,7] Some of the studies of JBD included only patients who met the full set of criteria,[4,10] whereas others also included patients with incomplete disease,[1–3,13] with the rate of definite diagnosis ranging from 10–80%. The rates of definite diagnosis in our study are comparable to other clinical series.

The male/female ratio in our paediatric group was 1.6:1 in accordance with an earlier study of Israeli patients,[4] as opposed to the male predominance in the adult group (3:1), as reported also in early study in Israeli patients,[14] but not in the more recent one.[4] The reported ratio among childhood-onset uveitis in BD in a large series in Turkey was 2.3:1.[3] Ethnic variability may account for the discrepancy between countries, as well as the difference between children and adults in our study, with different proportions of Jewish and Arab patients.

Pivetti-Pezzi *et al*[2] found that ocular disease was a presenting sign more often in children than in adults. In our study, a trend was noted towards having more children than adults diagnosed when the ocular disease appeared. However, this was not the first sign of the disease. The most common first sign was oral aphthae, confirming the earlier reports,[3] although in most cases, the ulcers did not prompt patients to seek medical attention. We usually made the diagnosis after the patients presented with ocular involvement, on the basis of the clinical ocular findings combined with the history, and supported by HLA typing, with positive results in 75% of the paediatric group compared with 80% of the adult group. An earlier study of JBD in Israel reported positive HLA B5 in 58% of complete JBD, and 94% in the incomplete group.[11] Our rates were comparable to recently published data regarding adult Israel patients.[15]

The clinical course in most of the children in our series was remarkable for hyperacute abrupt exacerbations of chronic disease. Severe inflammation accompanied by profoundly reduced VA occurred in some of the patients within 24–48 h. We have shown the dramatic fluctuations in VA between active disease and recovery. As visual outcome in BD is determined by the residual damage to the retina and optic nerve after recovery from these exacerbations, treatment was initiated in all patients upon diagnosis and consisted of corticosteroids and immunosuppressive agents; sometimes multiple drug combinations. Aggressive treatment is the standard

strategy for Behçet's uveitis, with studies over the years reporting improved visual outcome.[16]

The improvement rate in children treated aggressively shortly after the diagnosis and who were compliant with the treatment was high, even in the eyes with most severely reduced VA. Visual improvement was clinically significant, with most patients achieving useful vision. Total rate of visual impairment (including the noncompliant patients) was 18%. The rate in the treated patients represents a more favourable outcome than the previously reported rate of 23% of severe deterioration of vision in uveitis in childhood-onset BD,[3] as well as the 23 and 20% reported rate in recently published large multicentered studies of Behçet's patients.[17,18] Complications in BD occur either as a result of direct tissue inflammation, such as retinitis or optic nerve involvement, leading to residual anatomic and functional damages, or indirectly from cataract, elevated intraocular pressure, cystoid macular oedema, and epiretinal membrane. The rates of complications in both the paediatric and adult groups in our study were lower than the previously published data[3,16] (Table 5), except for posterior segment complications in the adult group, which were similar to the literature. It may be possible that the favourable visual outcome in our study, as well as the lower prevalence of complications result from shorter disease duration and follow-up. However, even though our study groups are small, which makes it hard to draw firm conclusions, it may be that early referrals, as well as the recent period (1997–2007) in which our patients were treated and the short interval between diagnosis and aggressive treatment in most cases, may account for these results. One of the consistent findings in the literature was a long interval (up to 9 years) between initial appearance of the clinical signs and diagnosis.[1,13,19] Thus, it is crucial to identify the clinical course, make the diagnosis, and initiate aggressive treatment even before the disease is fully expressed. The retrospective nature of this study and the small number of cases precluded conclusions regarding the preferred choice of drugs.

Earlier studies have shown variable trends in the severity of disease in children and adults relative to different clinical parameters. In an early report by Yazici *et al*,[20] male sex and younger age at presentation were associated with more severe disease. Krause *et al*[4] found a lower severity index in children than in adults. We noted a similar pattern of ocular disease in our paediatric and adult groups with differences related to the course of the disease (fluctuating in children) and the degree of irreversible anatomic damage (more in adults). It is possible that ethnic and genetic factors, as well as compliance with treatment, affect these disease patterns. In our study, poor compliance to medications was not uncommon among the teenagers with recurrent attacks.

In conclusion, Behçet's uveitis in children is a chronic relapsing disease of explosive nature. The course is usually characterized by acute exacerbations causing severe visual loss. In both children and adults, visual function may be restored by timely aggressive immunosuppressive therapy. In this study, we followed the course of the disease and found favourable outcome. As there is often a long interval between the appearance of uveitis and the expression of the full systemic disease, recognizing the pattern of ocular disease that follows this course, even in the absence of systemic signs, is important and should prompt clinicians to consider the diagnosis of BD. Early diagnosis will lead to earlier treatment. The severe course of the disease justifies the administration of aggressive systemic therapy, which appears to preserve visual function.

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