

# Vogt-Koyanagi-Harada disease in children

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## Abstract

**Purpose** To identify prognostic factors for outcome in children with Vogt-Koyanagi-Harada (VKH) disease.

**Methods** All children 16 years and younger with acute uveitis associated with VKH disease treated between 1999 and 2006 were reviewed.

**Results** Twenty-three children (46 eyes) were identified; 20 (87%) girls and three (13%) boys with a mean age at presentation of  $12.5 \pm 2.4$  years. Mean follow-up period was  $48.6 \pm 30.8$  months. Visual acuity of 20/40 or better was achieved in 38 (82.6%) eyes. Eleven eyes developed at least one complication, including cataract in eight eyes, glaucoma in eight eyes, subretinal neovascular membranes in two eyes, and subretinal fibrosis in one eye.

Disease recurred during follow-up in 18 eyes. Development of complications was negatively associated with final visual acuity of 20/20 ( $P = 0.0317$ ). Shorter interval between symptoms and treatment was a predictor of final visual acuity of 20/20 (odds ratio = 10.4; 95% confidence interval = 1.61–67.3).

Recurrence of inflammation was significantly associated with development of complications ( $P = 0.003$ ), worse visual acuity ( $P = 0.022$ ) and presence of posterior synechiae of the iris at presentation ( $P = 0.0083$ ), longer interval between symptoms and treatment ( $P = 0.013$ ), initial treatment with intravenous corticosteroids ( $P = 0.0012$ ), and rapid tapering of corticosteroids ( $P = 0.0063$ ).

**Conclusion** Visual prognosis of VKH in children is generally favourable. Clinical findings at presentation, development of complications, interval between symptoms and treatment, recurrence of inflammation, use of intravenous corticosteroids, and method of tapering of systemic corticosteroids were significant prognostic factors.

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## Introduction

Vogt-Koyanagi-Harada (VKH) disease is a chronic, bilateral, granulomatous panuveitis, and exudative retinal detachment associated with poliosis, vitiligo, alopecia, and central nervous system and auditory signs.<sup>1</sup> The exact cause of VKH disease remains unknown, but evidence suggests that it involves a T-lymphocyte-mediated autoimmune process directed against one or more antigens found on or associated with melanocytes. Several studies demonstrated that tyrosinase family proteins are the antigens specific to VKH disease<sup>2–5</sup> and that VKH disease is characterized by a T helper type 1 cell-mediated immune response.<sup>6,7</sup>

Vision-threatening complications have been clearly recognized to occur in the chronic, recurrent phase of VKH disease, namely cataract, glaucoma, subretinal neovascular membranes, and subretinal fibrosis. The occurrence of these complications is known to be associated with a worse visual outcome.<sup>1,8,9</sup> The principles of therapy of VKH disease are to suppress the initial intraocular inflammation with early and aggressive use of systemic corticosteroids, followed by slow tapering. Such treatment may shorten the duration of the disease, may prevent progression into the chronic stage, and may reduce the incidence of extraocular manifestations as well.<sup>1,10</sup>

VKH disease predominately occurs in women in their third to fourth decades of life at onset of the illness.<sup>1</sup> However, VKH disease can occur in the paediatric population, but the prognosis in this subpopulation appears to be quite variable.<sup>8,11–13</sup> It is possible that differences in interval between onset of uveitis and starting treatment, type of treatment, and/or duration of treatment could have played a role in the

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differences in outcomes reported by previous studies.<sup>8,11–13</sup> To determine prognostic factors for final visual acuity, development of complications, and recurrence of inflammation, we analysed 23 children with VKH disease who were followed from disease onset and treated with systemic corticosteroids.

### Patients and methods

We retrospectively reviewed the clinical charts of all children 16 years and younger diagnosed with acute uveitis associated with VKH disease seen at King Khaled Eye Specialist Hospital and King Abdulaziz University Hospital, Riyadh, Saudi Arabia between 1999 and 2006. All patients had typical ocular findings compatible with uveitis associated with VKH disease in the acute stage, and other uveitis conditions had been excluded by medical history, laboratory examination, and ancillary testing. Children who presented with chronic recurrent uveitis associated with VKH disease were excluded. Diagnosis of VKH disease was based on the revised criteria described recently.<sup>14</sup> The following laboratory and ancillary tests were performed: complete blood count with differential, erythrocyte sedimentation rate, blood sugar, blood chemistry, urinalysis, chest X-ray, tuberculin skin test, Venereal Disease Research Laboratory test, fluorescent treponemal antibody absorption test, intravenous fluorescein angiography, and posterior segment ultrasound. In addition, patients were examined with optical coherence tomography (OCT3 Stratus; Carl Zeiss Ophthalmic Systems, Humphrey Division, Dublin, CA, USA) after its availability in our institutes (Figure 1).

Charts were reviewed for demographic data (age and gender), initial and final visual acuities, results of slit-lamp examination, results of dilated fundus examination, results of fluorescein angiograms, duration from onset of symptoms to institution of corticosteroid therapy, details of corticosteroid therapy (type, tapering, and duration), ocular complications, number and location of recurrent episodes, extraocular manifestations (headache, tinnitus, neck stiffness, hearing loss, alopecia, poliosis, or vitiligo), and duration of follow-up.

### Statistical analysis

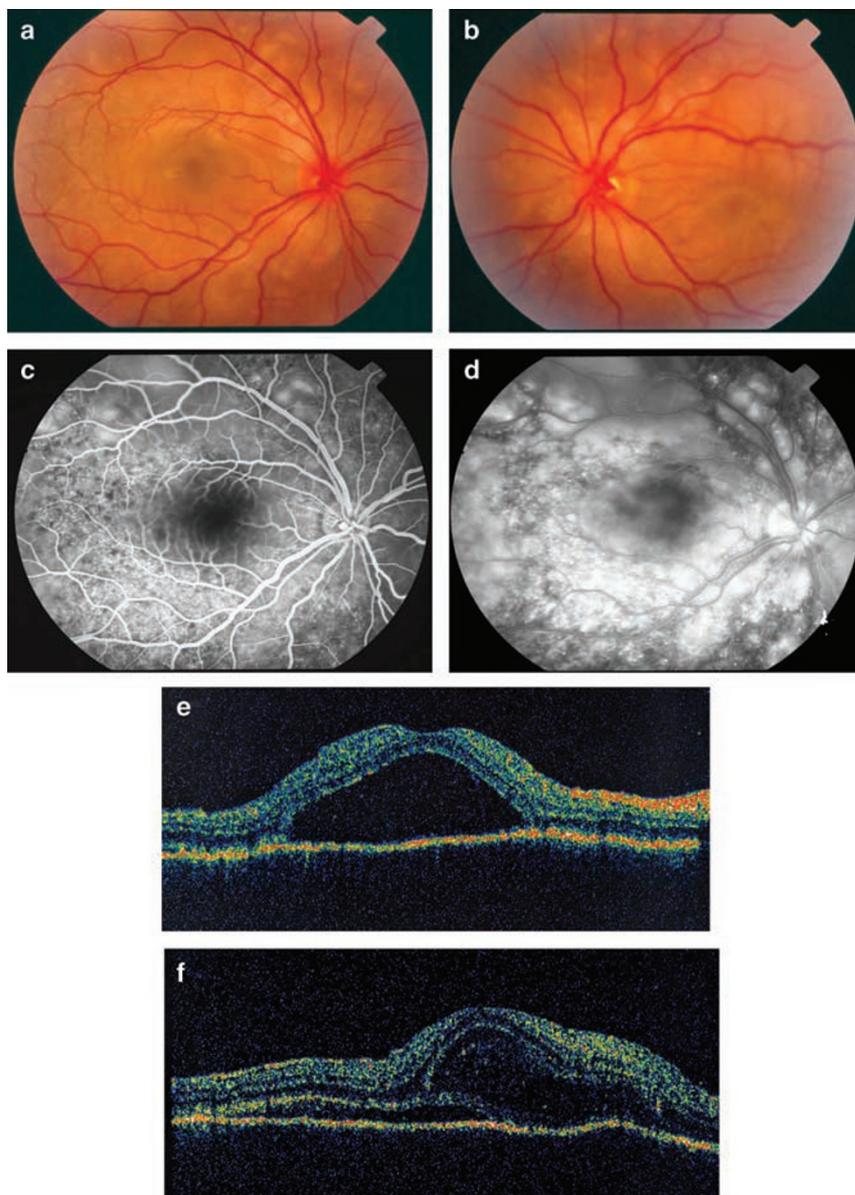
The association between two categorical variables was investigated using either the  $\chi^2$  test or Fisher's exact test as appropriate. Proportions from the same sample were compared using Student's *t*-test. Stepwise logistic regression analysis was conducted using program LR from the BMDP statistical software. For the  $\chi^2$  test and Fisher's exact test, a *P*-value less than 0.05 indicated statistical significance.

### Results

A total of 23 children (46 eyes) with VKH disease were identified. Twenty (87%) were girls and three (13%) were boys. The age at presentation ranged from 5 to 16 years with a mean of  $12.5 \pm 2.4$  years, and a median of 13.2 years.

The chief presenting complaint was diminution of vision. Associated prodromal symptoms included headache in 14 (60.9%) patients, tinnitus in six (26.1%) patients, and hearing impairment in three (13%) patients. Slit-lamp examination at presentation revealed anterior uveitis manifested as aqueous cellular reaction and flare in all cases. The anterior chamber reaction was graded as more than 2+ in 24 (52.2%) eyes.<sup>15</sup> Iris nodules were observed in four (8.7%) eyes, and posterior synechiae of the iris were observed in 14 (30.4%) eyes. Posterior synechiae of the iris at presentation were observed in none of the 22 (0%) eyes that had a shorter interval of 2 weeks or less between the onset of symptoms and starting treatment and in 14 of the 24 (58.3%) eyes that had a longer interval of more than 2 weeks between the onset of symptoms and starting treatment. The difference between the two percentages was significant ( $P < 0.001$ ). Fundus examination showed hyperaemia and swelling of the disc, and exudative retinal detachment in all eyes. Fundus fluorescein angiography was performed in all cases. Multiple pinpoint hyperfluorescent leaks at the level of the retinal pigment epithelium, staining of the disc, and late accumulation of fluorescein in the subretinal exudates were observed in our patients (Figure 1).

The interval between the onset of symptoms and starting treatment ranged from 0.4 to 52 weeks with a mean of  $6.7 \pm 11.0$  weeks, and a median of 2.9 weeks. All patients were treated with systemic corticosteroids. In 13 (56.5%) patients, the regimen given was intravenous methylprednisolone 15–30 mg/kg of body weight per day for 3 days followed by oral prednisone (1 mg/kg of body weight per day). Ten (43.5%) patients were treated with oral prednisone alone. Anterior segment inflammation was treated with topical corticosteroids and cycloplegic agents. Tapering of oral corticosteroid therapy was slow in 17 (73.9%) patients and rapid in five (21.7%) patients. Data were missing in one patient. Rapid tapering was defined as the use of a high-dose oral prednisone (1 mg/kg of body weight per day) for less than 2 weeks or tapering to the dose of 10 mg per day within a 2-month period or less. The duration of systemic corticosteroid therapy ranged from 3 to 73 months with a mean of  $26.2 \pm 20.0$  months and a median of 21 months. Ten (43.5%) patients required immunomodulatory therapy in addition to corticosteroids as follows: cyclosporine in eight patients, cyclosporine and



**Figure 1** A 12-year-old girl with VKH disease. (a) Right eye; (b) left eye. Note the multifocal exudative retinal detachments and the hyperaemic optic discs. Fluorescein angiography of the right eye shows typical features of VKH disease, including multiple pinpoint hyperfluorescence at the level of the retinal pigment epithelium (c) and late pooling of dye in the areas of exudative retinal detachment (d). Optical coherence tomography of the right eye (e) and left eye (f) shows the presence of exudative retinal detachments.

azathioprine in one patient, and azathioprine in one patient. The use of intravenous corticosteroids in relation to other variables is shown in Table 1. Intravenous corticosteroids were used in 11 of 13 (84.6%) eyes that had visual acuity of 20/200 or worse at presentation and in 15 of 33 (45.5%) eyes that had visual acuity of better than 20/200 at presentation. The difference between the two percentages was significant ( $P = 0.037$ ). In addition, intravenous corticosteroids were used in 16 of 34 (47.1%) eyes that had slow tapering of oral corticosteroid therapy and in 10 of 10 (100%) eyes that had rapid tapering of

oral corticosteroid therapy. The difference between the two percentages was significant ( $P = 0.0021$ ).

Furthermore, intravenous corticosteroids were used in 71.4% of eyes with posterior synechiae of the iris at presentation compared to 50% of eyes presenting without posterior synechiae of the iris. The difference between the two percentages did not attain statistical significance.

The follow-up period ranged from 3 to 88 months with a mean of  $48.6 \pm 30.8$  months and a median of 43 months. The distribution of initial and final visual acuity is

**Table 1** Use of intravenous corticosteroids in relation to other variables

Variable	Use of intravenous corticosteroids (%)	P-value
<b>Initial visual acuity</b>		
≤20/200	11/13 (84.6)	0.037*
>20/200	15/33 (45.5)	
<b>Anterior chamber reaction</b>		
≤2+	12/22 (54.5)	0.515
>2+	14/24 (58.3)	
<b>Posterior synechiae of the iris at presentation</b>		
No	16/32 (50)	0.153
Yes	10/14 (71.4)	
<b>Interval between symptoms and treatment (weeks)</b>		
≤2	12/22 (54.5)	0.515
>2	14/24 (58.3)	
<b>Duration of systemic corticosteroid therapy<sup>a</sup> (years)</b>		
≤1	8/14 (57.1)	0.5563
>1	18/30 (60)	
<b>Tapering of systemic corticosteroid therapy<sup>a</sup></b>		
Slow	16/34 (47.1)	0.0021*
Rapid	10/10 (100)	
<b>Immunomodulatory therapy</b>		
No	14/26 (53.8)	0.907
Yes	12/20 (60)	
<b>Extraocular manifestations<sup>b</sup></b>		
No	8/12 (66.7)	0.583
Yes	14/22 (63.6)	

\*Statistically significant at 5% level of significance.

<sup>a</sup>Data were missing for one child.

<sup>b</sup>Data were available for 17 children.

illustrated in Table 2. Of the 46 eyes, 38 (82.6%) achieved visual acuity of 20/40 or better. The frequencies above the left to right diagonal line represent eyes that had improvement in visual acuity, those below the line experienced worsened vision, and those along the diagonal line had no change in visual acuity. Therefore, 36 (78.3%) eyes had improved vision, four (8.7%) eyes had worsened vision, and there was no change in vision in six (13.0%) eyes. Eighteen (39.1%) eyes had visual acuity of 20/100 or worse at presentation and only two (4.3%) eyes had final visual acuity of 20/100 or worse. The difference between the two percentages was statistically significant ( $P < 0.001$ ). In addition, only seven (15.2%) eyes had visual acuity of 20/20 at presentation compared with 31 (67.4%) eyes that had final visual acuity of 20/20. The difference between the two percentages was statistically significant ( $P < 0.001$ ). Causes for final visual acuity of less than 20/40 are shown in Table 3.

**Table 2** Relationship between initial visual acuity and final visual acuity for 46 eyes

Final visual acuity	Initial visual acuity					Total
	Counting fingers	20/200–20/100	20/80–20/50	20/40–20/30	20/20	
20/20	6	6	6	8	5	31
20/30–20/40	0	1	4	1	1	7
20/50–20/80	5	0	0	0	1	6
20/100–20/200	0	0	1	1	0	2
Counting fingers	0	0	0	0	0	0
Total	11	7	11	10	7	46

**Table 3** Causes for final visual acuity of less than 20/40

Cause	Number of eyes
Cataract and glaucoma	4
Cataract and subretinal neovascular membrane	1
Glaucoma and subretinal neovascular membrane	1
Subretinal fibrosis	1
Unknown	1

Depigmentation of the fundus was evident in all eyes resulting in the characteristic appearance known as 'sunset glow' fundus. In addition, multiple small, yellow, well-circumscribed areas of chorioretinal atrophy developed in the midperiphery of the fundus in most of the eyes.

Overall, 11 (23.9%) eyes developed at least one complication. The ocular complications encountered were cataract in eight (17.4%) eyes, glaucoma that necessitated either medical or surgical intervention in eight (17.4%) eyes, subretinal neovascular membranes in two (4.3%) eyes, and subretinal fibrosis in one (2.2%) eye. There were six eyes that developed both cataract and glaucoma. Cataract extraction was performed on two eyes, and six eyes required surgical procedures for control of glaucoma. Disease recurred during the follow-up in 18 (39.1%) eyes. Of these 18 eyes, recurrence manifested as anterior uveitis in 15 (83.3%) eyes, exudative retinal detachment in two (11.1%) eyes, and combined anterior uveitis and exudative retinal detachment in one (5.5%) eye.

Of the 17 patients from whom data were available, six (35.3%) developed vitiligo, three (17.6%) developed poliosis, and two (11.8%) developed alopecia. Of these 17 patients, sensory hearing loss was documented in four (23.5%) patients after full audiologic assessment. Overall, 11 (64.7%) patients developed at least one of these extraocular manifestations.

Univariate analysis demonstrated a significant association between final visual acuity of 20/20 and

**Table 4** Factors affecting final visual acuity

Variable	Final visual acuity of 20/20 (%)	P-value
<i>Initial visual acuity</i>		
≤20/200	8/13 (61.5)	0.8554
>20/200	23/33 (69.7)	
<i>Anterior chamber reaction</i>		
≤2+	12/22 (54.5)	0.143
>2+	19/24 (79.2)	
<i>Posterior synechiae of the iris at presentation</i>		
No	22/32 (68.8)	0.9644
Yes	9/14 (64.3)	
<i>Interval between symptoms and treatment (weeks)</i>		
≤2	18/22 (81.8)	0.0923
>2	13/24 (54.2)	
<i>Intravenous corticosteroids</i>		
No	14/20 (70)	0.989
Yes	17/26 (65.4)	
<i>Duration of systemic corticosteroid therapy<sup>a</sup> (year)</i>		
≤1	10/14 (71.4)	0.8523
>1	19/30 (63.3)	
<i>Tapering of systemic corticosteroid therapy<sup>a</sup></i>		
Slow	24/34 (70.6)	0.2022
Rapid	5/10 (50)	
<i>Immunomodulatory therapy</i>		
No	21/26 (80.8)	0.0588
Yes	10/20(50)	
<i>Development of complications</i>		
No	27/35 (77.1)	0.0317*
Yes	4/11 (36.4)	
<i>Recurrence</i>		
No	22/28 (78.6)	0.0900
Yes	9/18 (50)	
<i>Extraocular manifestations<sup>b</sup></i>		
No	10/12 (83.3)	0.1925
Yes	12/22 (54.5)	

\*Statistically significant at 5% level of significance.

<sup>a</sup>Data were missing for one child.

<sup>b</sup>Data were available for 17 children.

absence of all complications ( $P = 0.0317$ ) (Table 4). When logistic regression analysis was performed, a shorter interval of 2 weeks or less between the onset of symptoms and starting treatment was a predictor of final visual acuity of 20/20 (odds ratio = 10.4; 95% confidence interval = 1.61–67.3). On univariate analysis, the development of at least one complication was significantly associated with recurrence of inflammation ( $P = 0.003$ ) (Table 5). Recurrence of inflammation was significantly associated with initial visual acuity of

**Table 5** Factors affecting development of complications

Variable	Development of complications	P-value
<i>Initial visual acuity</i>		
≤20/200	6/13 (46.2)	0.0664
>20/200	5/33 (15.2)	
<i>Anterior chamber reaction</i>		
≥2+	7/22 (31.8)	0.3912
>2+	7/24 (18.2)	
<i>Posterior synechiae of the iris at presentation</i>		
No	6/32 (18.8)	0.3867
Yes	5/14 (35.7)	
<i>Interval between symptoms and treatment (weeks)</i>		
≤2	4/22 (18.2)	0.5985
>2	7/24 (29.2)	
<i>Intravenous corticosteroids</i>		
No	4/20 (20)	0.8438
Yes	7/26 (26.9)	
<i>Duration of systemic corticosteroid therapy<sup>a</sup> (year)</i>		
≤1	2/14 (14.3)	0.4548
>1	9/30 (30)	
<i>Tapering of systemic corticosteroid therapy<sup>a</sup></i>		
Slow	8/34 (23.5)	0.4840
Rapid	3/10 (30)	
<i>Immunomodulatory therapy</i>		
No	4/26 (15.4)	0.1159
Yes	7/20 (35)	
<i>Recurrence</i>		
No	2/28 (7.1)	0.0030*
Yes	9/18 (50)	
<i>Extraocular manifestations<sup>b</sup></i>		
No	1/12 (8.3)	0.0826
Yes	8/22 (36.4)	

\*Statistically significant at 5% level of significance.

<sup>a</sup>Data were missing for one child.

<sup>b</sup>Data were available for 17 children.

20/200 or worse ( $P = 0.0220$ ), a longer interval of more than 2 weeks between the onset of symptoms and starting treatment ( $P = 0.013$ ), presence of posterior synechiae of the iris at presentation ( $P = 0.0083$ ), initial treatment with intravenous corticosteroids ( $P = 0.0012$ ), rapid tapering of systemic corticosteroids ( $P = 0.0063$ ), and the development of at least one complication ( $P = 0.0015$ ) (Table 6).

## Discussion

In the current study of a homogenous group of 23 children 16 years of age and younger who had uveitis

**Table 6** Factors associated with recurrence of inflammation

Variable	Recurrence (%)	P-value
<i>Initial visual acuity</i>		
≤20/200	9/13 (69.2)	0.0220*
>20/200	9/33 (27.3)	
<i>Anterior chamber reaction</i>		
≤2+	8/22 (36.4)	0.9476
>2+	10/24 (41.7)	
<i>Posterior synechiae of the iris at presentation</i>		
No	8/32 (25)	0.0083*
Yes	10/14 (71.4)	
<i>Interval between symptoms and treatment (weeks)</i>		
≤2	4/22 (18.2)	0.0130*
>2	14/24 (58.3)	
<i>Intravenous corticosteroids</i>		
No	2/20 (10)	0.0012*
Yes	16/26 (61.5)	
<i>Duration of systemic corticosteroid therapy<sup>a</sup> (year)</i>		
≤1	4/14 (28.6)	0.4191
>1	14/30 (46.7)	
<i>Tapering of systemic corticosteroid therapy<sup>a</sup></i>		
Slow	10/34 (29.4)	0.0063*
Rapid	8/10 (80)	
<i>Immunomodulatory therapy</i>		
No	8/26 (30.8)	0.3077
Yes	10/20 (50)	
<i>Development of complications</i>		
No	9/35 (25.7)	0.0015*
Yes	9/11 (81.8)	
<i>Extraocular manifestations<sup>b</sup></i>		
No	4/12 (33.3)	0.4095
Yes	12/22 (54.5)	

\*Statistically significant at 5% level of significance.

<sup>a</sup>Data were missing for one child.

<sup>b</sup>Data were available for 17 children.

associated with VKH disease in the acute stage, 82.6% of the eyes achieved visual acuity of 20/40 or better by final follow-up. Previous reports detailed visual outcome in children with VKH, with apparently contradictory results.<sup>8,11–13</sup> Rathinam *et al*<sup>12</sup> reported that of the 20 eyes from children 16 years of age or younger with VKH disease in the literature with final visual acuity data, 15 (75%) achieved 20/40 or better, whereas five (25%) achieved 20/200 or worse. In the study by Read *et al*,<sup>8</sup> there were seven patients who were 16 years of age or younger. Of the 13 eyes from these patients with final visual acuity data, 11 (85%) achieved 20/40 or better, whereas two (15%) achieved 20/50 or worse. Thus, these data suggest that children with VKH disease generally

do well. These data are contradictory to other reports in which a worse prognosis was found for younger patients. Tabbara *et al*<sup>11</sup> reviewed 97 patients diagnosed with VKH, 13 of whom were 14 years of age or younger. They found that eight (61%) of these patients had final visual acuities of 20/200 or worse despite medical and surgical therapy. This was significantly worse than the adults, 22 (26%) of whom had 20/200 or worse final visual acuity. The authors stated that the course of VKH disease in children tended to be more aggressive than in adults. Soheilian *et al*<sup>13</sup> reviewed 10 patients with paediatric VKH disease-associated panuveitis (onset of disease at age 14 years or younger). Final visual acuity was 20/40 or better in only 30% of the eyes, which is in agreement with Tabbara *et al*<sup>11</sup> study to some extent. Similarly, Ohno *et al*<sup>9</sup> reported that the younger age at disease onset, the worse the final visual acuity. The differences in outcomes reported by these studies could be related to differences in interval between onset of uveitis and starting treatment, type of treatment, and/or duration of treatment. In our study, we included only children with uveitis associated with VKH disease in the acute stage. In Soheilian *et al*<sup>13</sup> study, only one child presented with the acute phase of inflammation with bilateral exudative retinal detachment, which responded well to high-dose oral corticosteroids alone. Nine of their patients presented with chronic bilateral panuveitis and had sequelae of long-standing inflammation and complications that compromised vision. In the current study, there was a significant association between a shorter interval between the onset of symptoms and starting treatment and final visual acuity of 20/20. In addition, there was a significant association between a longer interval between the onset of symptoms and starting treatment and recurrence of inflammation.

In this study at least one complication occurred in 23.9% of the eyes. Cataract and glaucoma were the most common, occurring in 17.4% of eyes each. Subretinal neovascular membranes occurred in 4.3% of eyes. Rathinam *et al*<sup>12</sup> reported three children with VKH disease and reviewed the literature in relation to paediatric VKH. Of the 36 eyes, nine (25%) developed cataract and eight (22.2%) developed glaucoma. These figures are consistent with our results. In contrast, Tabbara *et al*<sup>11</sup> and Soheilian *et al*<sup>13</sup> separately found that VKH disease in children tended to be more aggressive than in adults. Tabbara *et al*<sup>11</sup> reported cataract in 61.5%, glaucoma in 46%, and subretinal neovascular membranes in 54% of children with VKH disease. Similarly, Soheilian *et al*<sup>13</sup> reported the development of cataract in 55%, glaucoma in 40%, and subretinal neovascular membranes in 70% of the eyes of children with VKH disease. In the current study, the development of ocular complications was significantly associated with

a worse final visual acuity. Similarly, previous studies showed a significant association between poor final visual acuity and greater number of complications.<sup>8,9</sup>

In the present study, 18 (39.1%) eyes developed recurrent inflammation during the follow-up period. The presence of recurrent inflammation was significantly associated with the development of complications including cataract, glaucoma, subretinal neovascular membranes, and subretinal fibrosis. Similarly, previous studies demonstrated that chronic recurrent anterior segment inflammation was significantly associated with poor final visual acuity and the development of complications.<sup>8,16–18</sup> Our data also demonstrated that initial ocular findings including a worse initial visual acuity and the presence of posterior synechiae of the iris at presentation were significantly associated with recurrent inflammation. Our analysis demonstrated that the presence of posterior synechiae of the iris at presentation was significantly associated with a longer interval between the onset of symptoms and starting treatment. Therefore, the presence of posterior synechiae of the iris at presentation was a reflection of delayed start of treatment. These results are consistent with those of Keino *et al*<sup>18</sup> who demonstrated that the presence of signs of more severe disease at onset including presence of severe anterior uveitis, peripheral anterior synechiae, and posterior synechiae of the iris was significantly associated with chronic ocular inflammation. These data may be interpreted as showing that more severe disease at onset is associated with a greater risk to develop recurrent inflammation.

Early and aggressive high-dose systemic corticosteroid therapy has become the mainstay therapy of VKH disease. Patients with VKH disease adequately treated with corticosteroids have a favourable visual prognosis.<sup>1,9,10,16</sup> This treatment may prevent the progression of VKH disease, lessen its duration, prevent severe ocular complications, and prevent systemic diseases such as ear, skin, or hair lesions.<sup>1,10</sup> The current study has identified a positive association between final visual acuity of 20/20 and a shorter interval between the onset of symptoms and starting treatment. In addition, the current study demonstrated a significant association between recurrent inflammation and a longer interval between the onset of symptoms and starting systemic corticosteroid therapy, and rapid tapering of systemic corticosteroid therapy. Similarly, Rubsamen and Gass<sup>16</sup> found that almost all recurrences within 6 months of presentation were associated with too rapid or early decrease in steroid doses. These observations strongly suggest that it is critical to start adequate corticosteroid therapy as soon as possible at onset for suppression of ocular and systemic inflammation of VKH disease. Surprisingly, our analysis demonstrated a significant

association between recurrent inflammation and the use of intravenous corticosteroids. One possible explanation is that there was a tendency to use intravenous corticosteroids in those patients who had more severe disease at presentation. In fact, intravenous corticosteroids were used more frequently in eyes that had a worse initial visual acuity and posterior synechiae of the iris at presentation. In addition, rapid tapering of systemic corticosteroid therapy was performed more frequently in patients who used intravenous corticosteroids.

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