

Eye Findings in Patients with Juvenile Dermatomyositis

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ABSTRACT. Objective. Reports of eye involvement in juvenile dermatomyositis (JDM), including significant retinopathy with visual loss, have led some to recommend routine formal ophthalmologic assessments for all patients with JDM at diagnosis. Our objective was to document the frequency and spectrum of eye involvement in patients followed in a single clinic caring for children with JDM.

Methods. A chart review was conducted of formal ophthalmologic consultation notes for patients with JDM followed at the Hospital for Sick Children between 1981 and 2002.

Results. Ophthalmologic assessments were found for 82 of 108 patients with JDM. The mean age at diagnosis of JDM was 7.0 years and 68.3% were female. Forty-five patients (55.6%) had abnormal eye examinations. Lid manifestations, found in 37 patients (45.7%), were the most common abnormality. Fourteen patients (17.1%) had corticosteroid-induced cataracts. Two patients had retinal abnormalities; one had a small retinal hemorrhage, the other an incidental chorioretinal scar. Neither had impairment of vision. No patient had uveitis.

Conclusion. Eyelid and lens abnormalities are common in patients with JDM, while retinopathy is rare. As lid lesions and cataracts are easily detected by non-ophthalmologists, and retinal lesions are rare, we feel that JDM patients without visual symptoms do not require routine formal ophthalmologic assessment for disease manifestations. (J Rheumatol 2005;32:1986–91)

Key Indexing Terms:

JUVENILE DERMATOMYOSITIS RETINOPATHY CATARACTS HELIOTROPE

Juvenile dermatomyositis (JDM) is a systemic inflammatory vasculopathy in which the predominant clinical manifestations occur in skin and muscle. Less common manifestations include arthritis, interstitial pneumonitis, and visceral vasculopathy¹. Eye involvement in both adult DM and JDM has been described. Most common manifestations are lid changes, including heliotrope rash²⁻⁴, scaly erythema⁵, and edema^{6,7}, either as sole manifestations or in combination with other findings. Avascular areas within the conjunctiva, possibly the result of previous vasculopathy⁸, as well as uveitis, episcleritis, and glaucoma have also been described⁹⁻¹¹. Of particular concern are reports of retinopathy, both symptomatic and asymptomatic. Cotton wool spots, retinal hemorrhages, and retinal edema have all been

described in adult patients with DM¹¹⁻¹³, in some cases producing permanent visual impairment^{11,13}. Similar findings, including resultant permanent visual loss, have been reported in children with JDM^{2,4-6,9,14,15}. Indeed, it has been postulated that children may be at particular risk for this complication given the prominent vasculopathy seen in JDM⁴.

Concern about missing asymptomatic retinopathy has led some clinicians to recommend routine examination by an ophthalmologist for all patients with DM^{3,12}. Presently, there are few comprehensive data regarding the frequency of eye involvement, and in particular retinopathy, in JDM. Such data would allow an evidence based approach to the allocation of clinical resources in detecting significant eye involvement in this disease. We investigated the frequency and spectrum of eye involvement in a large cohort of patients followed in a single specialist clinic for children with JDM.

MATERIALS AND METHODS

The JDM clinic at Hospital for Sick Children in Toronto was established in 1991 as a multidisciplinary specialist clinic caring for children with idiopathic inflammatory myopathies, of which JDM is the most common. Children followed in the JDM clinic undergo standardized physical assessments at each visit. Data regarding demographic details, diagnosis (based on the criteria of Bohan and Peter¹⁶), and predefined clinical and laboratory characteristics, recorded for each patient during followup, are stored in an electronic database. Until recently, the majority of patients were routinely referred for ophthalmologic assessment around the time of diagnosis and, in many cases, during followup.

The protocol for this study was approved by the hospital Research

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Ethics Board. All patients with a confirmed diagnosis of JDM followed by the Division of Rheumatology were eligible for inclusion, including those followed prior to the establishment of the JDM clinic. Eligible patients were identified using the JDM database kept by the Division of Rheumatology. Patients were included in the study if they had undergone at least one documented assessment by a pediatric ophthalmologist at the hospital. Evidence of this was sought for all eligible patients by a search of the hospital record and separate charts kept by the Department of Ophthalmology. All patients had standardized eye examinations, including examination of the eyelids and other adnexal structures and pharmacologic dilation of the pupils for funduscopy. The majority of eye examinations were conducted by one pediatric ophthalmologist with a special interest in the ocular manifestations of rheumatic diseases (AVL). For patients who had had more than one eye assessment, abnormalities were noted cumulatively; if an abnormality was noted at any visit then it was considered as present for that patient. Relevant demographic, clinical, and laboratory features were recorded for each included patient from the established JDM database.

RESULTS

Between 1981 and 2002 a total of 108 patients with the diagnosis of JDM were followed at Sick Children's Hospital. Eighty-two patients (76%) had at least one documented ophthalmologic assessment in their chart, of whom 59 (72%) had been assessed after the establishment of the JDM clinic. The demographic and clinical features of the 82 patients with documented ophthalmologic assessments and of the 26 patients for whom an assessment could not be found are compared in Table 1. The majority of the latter group were either seen prior to the establishment of the clinic, when eye assessments were not routine, or had been seen only once in the JDM clinic for a second opinion. Patients included in this study were younger than those not included, and were more likely to have evidence of vasculitis as defined by ulcerative skin lesions at the onset of their disease. However, in all other respects the disease features of the 2 groups were similar.

A total of 170 eye examinations were documented for the 82 patients. In one patient, no comment could be found with respect to the lids at either of 2 eye examinations at which all other findings were normal. The median number of

examinations per patient was one (range 1–9). The median time from diagnosis to the first eye examination was 2.4 months (range 0–148 mo). Thirty-six (44.4%) of the 81 patients for whom complete data at all visits were available had normal eye examinations at every visit.

Thirty-seven of the 81 patients with complete eyelid data at every visit (45.7%) were found to have abnormalities of the lids on at least one visit. Table 2 details the lid abnormalities identified. Of the 31 patients noted to have heliotrope of the lids, 10 had a "purple line" variant in which, rather than diffuse skin involvement of the lids, there was a supraciliary line of purple discoloration, usually containing telangiectatic vessels without hemorrhage or edema, on the upper lid (Figure 1). In 2 patients this was seen prior to the development of typical heliotrope rash (Figure 2), and in 2 other patients it was seen either concurrent with or following heliotrope rash. In 6 patients it was seen as the only lid abnormality. Six (7.4%) patients had lid scarring, which typically appeared as a pox-like noninflamed atrophic well circumscribed round lesion, about 0.5–1 mm in diameter, usually on the medial half of the upper lid (Figure 3).

Lens abnormalities were noted in 15/82 patients (18.3%). Of these patients, 14 were felt to have acquired steroid induced posterior subcapsular cataract (PSC), most com-

Table 2. Eyelid findings documented in 81 JDM patients. Patients with different findings at sequential visits may be included in more than one category.

Abnormality	No. of patients (% of total*)
Heliotrope	31 (38)
Scarring	6 (7.4)
Blepharitis	4 (4.9)
Other**	3 (3.7)

* Total calculated using 81 patients as denominator as no information recorded regarding lids for one patient. ** One each of scaling, papule, and edema.

Table 1. Comparison of demographic and selected clinical features of JDM patients with (included) and without (excluded) ophthalmology consultation. Unless otherwise indicated, all comparisons were made using Fisher's exact tests.

	Patients with Eye Data, n = 82*	Patients without Eye Data, n = 26*	p
Mean age at diagnosis, yrs	7.0 (0.9–17.8)	8.7 (1.9–15)	0.03**
Male:female	1:2.3	1:2.2	1.00
Mean length of followup, yrs	5.2 (0–17.8)	3.7 (0–13.2)	0.07**
Heliotrope at onset (%)	69/81 (85.2)	21/26 (84.6)	0.55
Malar rash at onset (%)	30/63 (47.6)	5/18 (27.8)	0.18
Abnormal nailfolds (%)	76/81 (93.8)	21/26 (80.8)	0.06
Vasculitis at onset (%)	18/81 (22.2)	1/26 (3.8)	0.04
Livedo at any time (%)	2/63 (3.2)	0/18 (0.0)	1.0
Calcinosis at any time (%)	22/81 (27.2)	9/26 (34.6)	0.47

* Where given, denominators indicate the number of patients with adequate information. ** Two-sample t test.



Figure 1. Purple line variant of eyelid heliotrope. Telangiectatic vessels are visible with associated supraciliary purple discoloration of the eyelid. This may precede or follow a more typical heliotrope rash.



Figure 2. Typical heliotrope eyelid rash with more diffuse purple discoloration of the upper eyelid, often associated with eyelid edema.

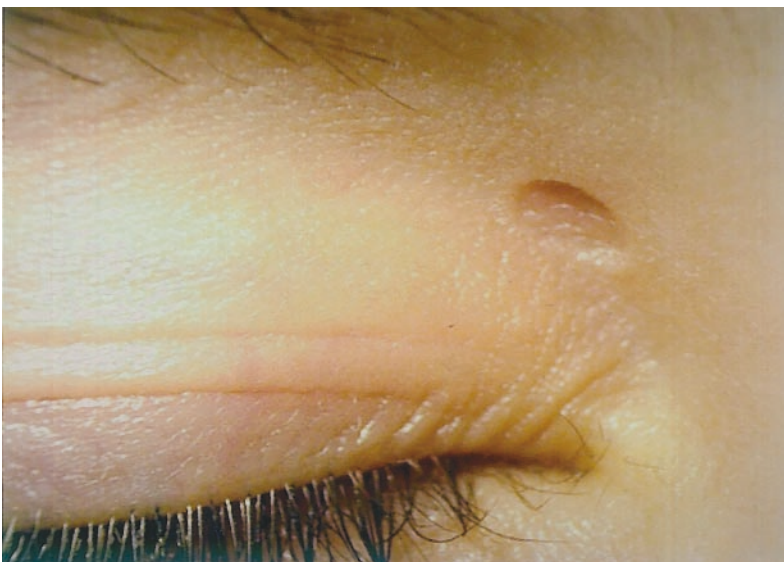


Figure 3. Typical "pox-like" lid scar in a patient with JDM.

monly bilateral, and one was felt to have congenital PSC. One child with acquired PSC was also felt to have a congenital anterior subcapsular cataract. Accurate cumulative corticosteroid data were available from the electronic database for 7 of the 15 (46.7%) patients with cataracts and 15 of 67 (22.4%) without cataracts. The median duration of corticosteroid therapy and median cumulative corticosteroid dose at either the time of the first documented cataract (cataract group) or last eye visit (non-cataract group) was 331 versus 283 days and 8918 mg versus 7876 mg for the cataract and non-cataract groups, respectively. These differences were not statistically significant.

Two children were found to have abnormalities on retinal examination. One child had an incidental, nonspecific chorioretinal scar and the other a single small dot hemorrhage with no other associated retinal change. Neither child had any disturbance of their vision.

No child was found to have uveitis. Tonometry was performed in 12 children and was abnormally elevated in one. This child was felt to have corticosteroid induced glaucoma, which resolved as her corticosteroid doses were tapered.

Figure 4 illustrates the distribution and interrelationship of eye findings across the study group.

DISCUSSION

Eye findings have been described as part of the spectrum of disease involvement in both adults and children with DM since early in the last century. Most of these descriptions have been case reports and have concentrated on retinal findings, emphasizing the permanent visual impairment that may result from retinopathy in this disease. However, in large series documenting the prevalence of various clinical findings in JDM, retinal findings have been rare^{1,17,18}. In the absence of clear prevalence data, determining time- and cost-effective strategies to ensure that rare but important

manifestations of uncommon diseases are detected can be difficult. Our objective was to determine prevalence data for all aspects of eye disease in a tertiary clinic population of patients with JDM in order to provide an evidence base for decisions regarding ophthalmic screening.

The most common eye findings in our study were related to changes in the skin of the lids. This is not surprising, given the frequency with which heliotrope rash occurs in JDM. In a previous description of the clinical features of the JDM clinic population at Sick Children's Hospital, heliotrope was seen in 83% of patients at diagnosis¹. If anything, the percentage of patients in this study with lid abnormalities (46%) is smaller than might be expected. This is probably because many patients had their first eye examination several months after they were diagnosed with JDM; the median time to first eye examination after diagnosis was 2.4 months. In the interim these patients had started therapy that would be expected to reduce their skin findings.

The stereotypical heliotrope rash of JDM usually presents as a diffuse purple-red erythema of the lids, with or without swelling, which often extends onto the malar region. It is thought to represent active underlying inflammatory vasculopathy. We observed a variant heliotrope rash in 10 patients in whom a supraciliary purple line on the upper lid, often containing telangiectatic vessels without hemorrhage or edema, was the only sign of discoloration. We were not able to detect any particular clinical significance of this variant.

A well circumscribed atrophic lid scar was seen in 7.4% of the study group. This finding has previously been described and is felt to relate to vasculopathy within the skin causing ulceration and subsequent scars^{1,18}. On this basis we postulated that canthal lesions may represent a marker for more severe disease. This notion is perhaps supported by the fact that 5 of the 6 patients with this finding also had heliotrope; 2 of 6 had cataracts and one of them was the only patient in the study population to have had a disease related retinal finding. However, the numbers involved are too small to draw any firm conclusions.

After lid abnormalities, ocular complications of therapy were the next most common finding in our study. Posterior subcapsular cataracts are a recognized complication of longterm corticosteroid therapy. The risk of their development is felt to relate to both the dose and duration of corticosteroid treatment^{19,20}. Although there were insufficient data to allow comparisons of corticosteroid therapy between children who developed cataracts and those who did not, it is notable that for those who did develop them, cataracts were identified at a median duration of corticosteroid therapy of less than 11 months.

Tonometry was performed infrequently in our study population. In part this reflects the impracticality of such measurements in very young patients (the median age at diagnosis in our study population was 6.1 years); however, it also

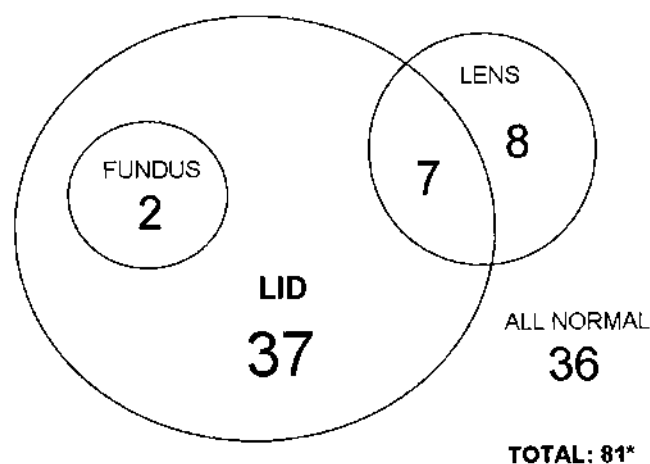


Figure 4. The relationship of eye findings in the study group patients with JDM. *Total = 81 as one patient had no documentation of lid findings at either of 2 visits, all other findings normal.

reflects that glaucoma has not been described in JDM and only rarely in adult DM²¹. Only one of the screened patients was found to have elevated intraocular pressure. This was not associated with changes in visual acuity and resolved with reduction in corticosteroid dose. The association between chronic low dose oral corticosteroid use and glaucoma in children is not consistent^{20,22,23}, and at this time there are no official guidelines regarding referral of children undergoing chronic oral corticosteroid therapy for glaucoma screening. Our current practice is to recommend twice yearly screening by an ophthalmologist or optometrist for children using corticosteroids continuously for more than 6 months.

Retinal findings in our study population were rare and not clinically significant. Of the 2 patients with retinal abnormalities, one had a probable developmental defect (chorioretinal scar) unrelated to JDM. The other had a small dot hemorrhage without evidence of cotton wool spots and with no disturbance in vision. The low prevalence of disease related retinopathy in this study (1.2%; 95% confidence interval 0–3.6%), conducted in a tertiary referral clinic population, is perhaps surprising given the reports in the literature and accompanying recommendations. While it is possible that some of the JDM patients not included in this study had a retinopathy we did not detect, the probability of them having clinically significant retinal lesions is very small; if anything, on the basis of a significantly greater prevalence of vasculitis, the patients included in this study might be expected to have been more likely to have retinopathy than those who were not.

A limitation that needs to be taken into account when interpreting our results is the retrospective design of this study. A potential problem associated with this type of study, particularly when examining the prevalence of disease features that may develop over time, is non-uniform initial assessment and followup intervals. Although the majority of patients had ophthalmologic assessments early in their disease course, less than half had them on the actual day of diagnosis or prior to starting therapy. Similarly, followup eye examinations were not necessarily done at times of disease flare. Therefore it is possible that retinal lesions were missed in patients in this study — that they resolved prior to their eye assessment on the therapy required for their extraocular disease. Prevalence data for retinopathy in JDM would most accurately be obtained by a prospective study involving eye assessments in a large number of patients before starting or escalating therapy, at diagnosis and times of disease flare. However, given the excellent visual outcome in our patients, we feel that the probability of this study underestimating clinically significant retinal lesions (i.e., those severe enough to warrant therapy escalation, irrespective of extraocular disease considerations) is low.

One of the principal justifications for routine formal ophthalmologic screening in JDM patients without visual symp-

toms is the desire to detect and treat retinopathy at an early stage in order to minimize the chances of visual loss. This assumes that patients with JDM may have an asymptomatic retinopathy whose treatment requirements are greater than that which would be indicated for their extraocular disease alone. This situation might arise, for example, if significant, but asymptomatic, retinal lesions were to occur in the context of disease that was otherwise in remission or very mild. However, based on our experience and review of the pediatric literature, this does not occur. In every case of JDM-associated retinopathy reported in the English literature, the retinopathy was found at the time of the patient's initial disease presentation when extraocular manifestations predominated^{2,4-6,9,14,15}. In these reports all patients who went on to have permanent visual impairment had visual symptoms at the time of presentation^{4,5,15}. In the single reported case of an asymptomatic retinal lesion at the time of diagnosis, the lesion resolved with the therapy required for treatment of the patient's skin and muscle disease alone². Unfortunately, there are no details of visual symptoms or acuity in 2 of the 3 cases reported by Bruce⁶ and Nutt¹⁴. In the third case, vision was described as being "dim" and the child recovered and the retinopathy resolved without apparent specific therapy⁶. Although no comment can be made regarding visual symptoms in these cases, it is clear that the patients' systemic disease would have required treatment with vigorous immunosuppression regardless of their retinal findings.

Eye findings in JDM are common and mostly relate to changes in the skin of the eyelids. The most common eyelid finding is heliotrope rash. Less common, but potentially a marker of more severe disease, are canthal scars. Ocular complications of corticosteroid therapy in the form of PSC are also common and can be found in patients treated with corticosteroids for less than a year. In our experience, retinal lesions are rare. As lid changes and PSC are easily detected by non-ophthalmologists, and sight-threatening retinal lesions rare and never asymptomatic, we feel that JDM patients without visual symptoms who are not at risk of corticosteroid induced glaucoma do not require routine formal ophthalmologic assessment.

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