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Clinical Observations

Pediatric Optic Neuritis: Does a Prolonged Course of Steroids Reduce Relapses? A Preliminary Study

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ABSTRACT

BACKGROUND: Optic neuritis is an important pediatric disorder causing visual impairment. Because of the absence of pediatric-specific studies, data extrapolated from the adult-based optic neuritis treatment trial are used to guide management of pediatric patients. Recent literature promotes a prolonged course of oral steroids to prevent relapses. However, there are no published data to support this view. Patients who were recently treated in our hospital received a longer course of steroids, relative to those treated several years ago. We hypothesized that a longer course of steroids results in fewer relapses and better final visual acuity. **METHODS:** A retrospective analysis of 26 consecutive patients (age 4.5-19 years) treated for optic neuritis within the past 10 years was conducted. Patients received either a short course (2 weeks) or a prolonged course (more than 2 weeks) of steroids. Some patients were not treated. Mean follow-up was 70 weeks (3 weeks-10 years). Comparisons were made among the groups receiving 2 weeks of steroid treatment (16 of 26 patients) and greater than 2 weeks of steroid treatment (seven of 26 patients) to evaluate relapse rate, eventual visual acuity, and reported side effects. **RESULTS:** There were no significant differences in the relapse rates, reported side effects, and final visual acuity in the two treatment groups. **CONCLUSIONS:** In this cohort, a prolonged course of steroids was not associated with reduced relapse rate, increased side effects, or improved visual outcome. This cohort was small, but the results do not identify any reason to deviate from the common approach of optic neuritis treatment, which is 2 weeks of steroids.

Keywords: optic neuritis, visual impairment, steroid treatment, visual acuity, visual outcome, pediatrics

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Introduction

Optic neuritis (ON) in children characteristically presents with significant visual loss. The reported incidence of pediatric ON is much lower than that of adults.[1] The optic neuritis treatment trial (ONTT) described outcome and treatment strategies of ON in adults, and data from that study provide specific guidelines for treatment.[2] Because of the rarity of pediatric ON, most pediatric clinical data are limited to descriptive studies or retrospective case series. Although pediatric neurologists and neuro-ophthalmologists rely on ONTT in their management of pediatric ON, several pediatric studies suggest that pediatric ON presents and behaves differently than adult ON.[3,4]

The most commonly used treatment of adult-onset ON, although controversial, consists of a course of intravenous (IV) steroids for 3 days, followed by 2 weeks of oral steroids. Treatment with high-dose IV steroids hastens the visual recovery and delays the presentation of multiple sclerosis (MS) within the first 2 years. Traditionally, this same treatment has been used in children. Some authors have suggested that a prolonged course of oral steroids, more than 4-6 weeks, rather than 2 weeks, may reduce relapses in pediatric ON.[1,4-6] However, there are no class I data to

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TABLE 1.
Clinical Characteristics of the Cohort at Presentation

Features	Numbers	Percentage
Age at onset, yr	4.5-19	
Sex (male:female)	6:19	
Preceding illness	11/26	42
Unilateral	14/26	53
Bilateral	12/26	46
Presence of optic disc edema	19/26	73
CSF analysis performed	14/26	53
CSF pleocytosis	6/14	42
Elevated protein	1/14	7
Abnormal MS panel	2/14	14
Abnormal MRI	7/26	26

Abbreviations:
CSF = Cerebrospinal fluid
MRI = Magnetic resonance imaging
MS = Multiple sclerosis

support this approach. In the past decade, there has been a change in the duration of steroid treatment for ON at the University of Iowa Hospitals, which provided us the opportunity to conduct an outcome analysis.

Our purpose was to analyze characteristics of pediatric ON (presenting symptoms, signs, magnetic resonance imaging (MRI) and cerebrospinal fluid analyses, management strategies, visual outcomes, and long-term outcomes) and to compare treatment outcomes of a 2-week steroid course with a longer duration steroid course.

Materials and Methods

After obtaining Institutional Review Board approval, we identified 27 consecutive patients (age 4.5-19 years) diagnosed with a first attack of ON within the past 10 years in the neuro-ophthalmology clinic at the University of Iowa Hospitals. Inclusion criteria were a diagnosis of isolated ON. Exclusion criteria included additional neurological findings at presentation or repeated ON and patients with a secondary cause of optic neuropathy (e.g., Lyme disease, Leber's hereditary optic neuropathy). One patient was excluded because of poor compliance with the recommended treatment.

Presenting symptoms, signs, MRI and cerebrospinal fluid findings, initial visual acuity and the best visual acuity after treatment, relapses, and treatment regimen were retrospectively abstracted (Table 1). Relapses were defined as recurrent ON symptoms occurring more than 2 weeks after the initial onset. Documented follow-up varied from 3 weeks to 10 years with a mean of 70 weeks. Patients with almost complete recovery were monitored as needed, with no further documented follow-up in seven patients. Patients who developed multiple sclerosis or recurrent ON continued to be monitored either in neuro-ophthalmology or neurology clinics.

Relapse rates, eventual visual acuity, and incidence of side effects were analyzed using a chi-square test among two groups, respectively: 2-week steroid treatment (16 of 26 patients) and prolonged steroid treatment (seven of 26 patients). The distribution of the patients in the various groups is illustrated in Figure.

Results

Twenty-seven patients were diagnosed with ON. Twenty-four patients received high-dose IV corticosteroids for 3 days followed by oral steroids. For 17 of these patients, oral steroids were prescribed for 2 weeks and only 16 completed the treatment. One patient did not complete the treatment and was excluded from the analysis, hence, there were a total of 26 patients. For the other seven patients, the duration of oral steroids was longer (3-12 weeks; mean, 6.7 weeks). Three patients received no corticosteroids. Most pediatric patients had severe visual impairment at presentation; in the range of legal blindness in 19 of 26 patients (73%) with either finger counting or light perception. Bilateral disease was present in 46% (12 of 26 patients). Except for one patient, all had good visual outcome, and the final visual acuity was better than 20/40 in 19 patients (73%), regardless of the treatment they received. One patient, who was noncompliant with the steroid treatment, discontinued the treatment after 5 days and presented with worsened visual acuity in the same eye 1 week later. Seven of 26 patients had abnormal white-matter enhancement in T_2 images. Two of these patients recovered completely, two developed recurrent ON, one was subsequently diagnosed with Acute disseminating encephalomyelitis (ADEM), and two patients developed multiple sclerosis (Table 2).

Regarding the final visual acuity, there was no significant difference between the 2-week treatment group and the prolonged treatment group (final visual acuity better than 20/40 in 12 of 16 patients versus five of seven patients; $\chi^2 = 0.03$; $P = 0.50$). Of the 26 patients in the cohort, only

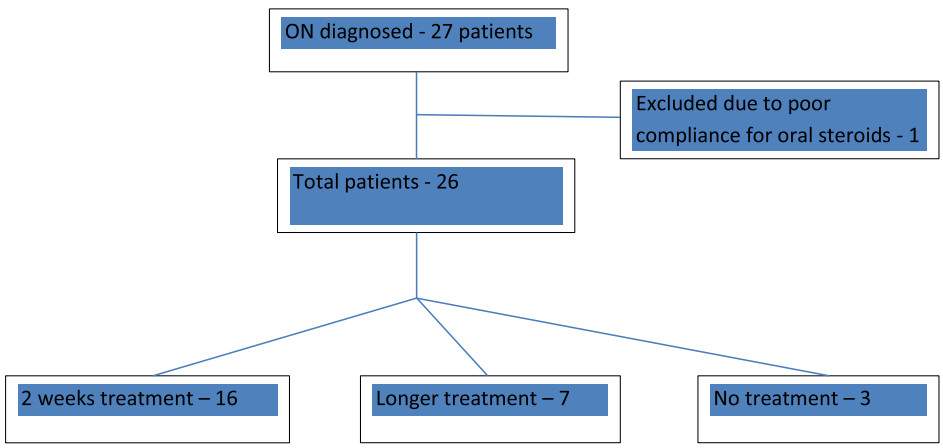

FIGURE.
Distribution of patients in each group. ON, optic neuritis. (Color version of this figure is available in the online edition.)