Original Article

Optic neuritis in children with poor recovery of vision

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Abstract

We reviewed the records of 10 children with optic neuritis in whom recovery of vision was poor or incomplete. Our cases were otherwise similar to those described in previous studies in that they were always bilateral, often accompanied by a viral prodrome (seven of 10), and usually associated with disc oedema (seven of 10).

Seven of twenty eyes had a final visual acuity of 6/60 or worse and only one patient regained 6/6 vision in either eye. In three patients the best vision in either eye was 6/60 or worse. Recovery of vision was often slow, taking up to six years. Five of 10 patients have developed multiple sclerosis (MS), and one child had acute disseminated encephalomyelitis (ADEM) with optic neuritis.

Optic neuritis in children does not always carry a good prognosis for recovery of vision; however, the failure of vision recovery in a short period of time does not necessarily indicate a poor outcome. Some children with optic neuritis develop MS, which can develop even when optic neuritis follows a viral illness.

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The most important rule regarding the clinical behaviour of optic neuritis in children is that prognosis for rapid recovery of vision is good. This was documented by Kennedy and Carroll in 1960,[1] and every published study since that time has confirmed this report.[2-4] The prediction of a good outcome at the onset of vision impairment helps alleviate the distress of both patients and parents, but in offering an optimistic prediction, we have found ourselves wrong in many instances. Ten children with optic neuritis did not follow the well established rule governing prognosis.

Patient selection

The records of 10 children with optic neuritis who had an unusual or poor outcome from two institutions (University of California San Francisco, Hospital Vargas) were reviewed. Optic neuritis had been diagnosed on the basis of decreased central vision accompanied by other signs of optic nerve dysfunction including colour vision deficit, afferent pupillary defect, or papillitis. None of these children had a systemic disease known to cause optic nerve dysfunction and there was no evidence of inherited optic neuropathies in family histories. No child in this study had macular oedema or neuroretinitis.

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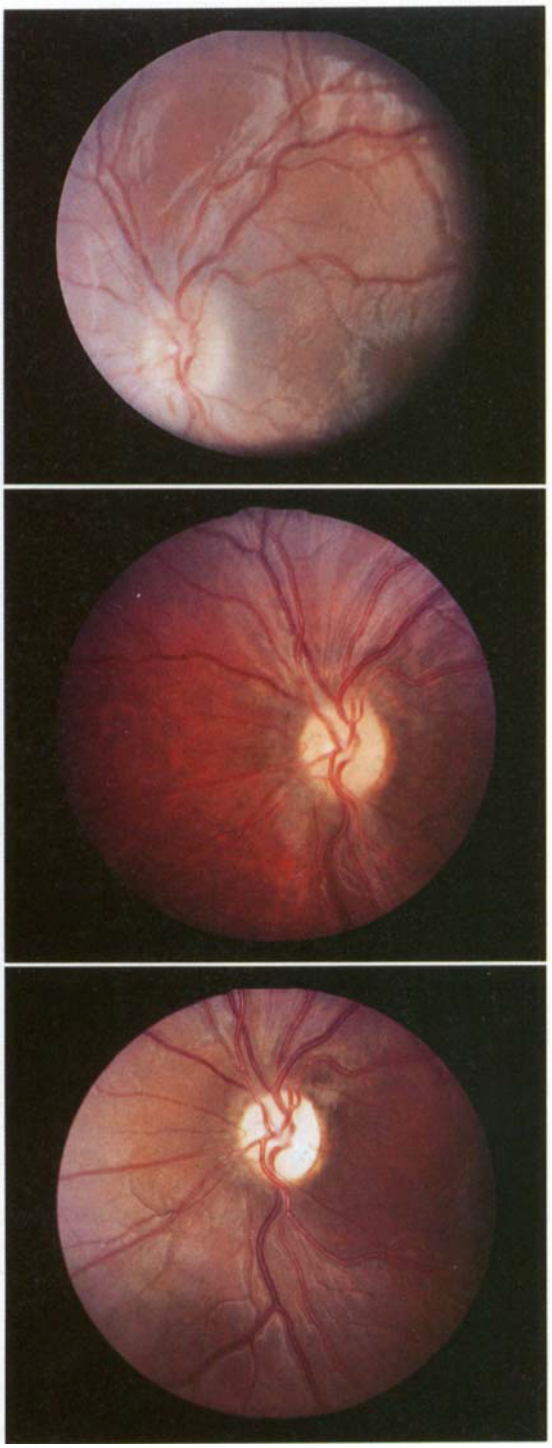

Figure 1 Optic nerve photographs of Patient 10. Note the progression of optic atrophy in photos taken over a three-year period. *Top*, appearance of nerve at disease onset; *middle*, eight months later; *bottom*, three years after onset. All photos are of the left optic nerve.

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Patients were 16 years or younger at the onset of optic neuritis and were followed for at least two years. Information regarding age, prodromal illness, maximal vision loss, treatment, time to recovery, physical findings, final acuity, neuroimaging studies, and development of other neurologic signs and symptoms was obtained from records. In two children (numbers 1 and 4), follow-up information was obtained from referring physicians.

Results

Nine children were female and one was male (Table 1). The average age was 10 years, and six children were under the age of 10.

Seven children had a viral illness preceding the onset of vision failure, and all 10 suffered bilateral optic neuritis. In most cases optic neuritis occurred either simultaneously or within a few days of the onset in the fellow eye. Optic neuritis was considered as bilateral if both eyes were affected within one month. The initial vision loss was usually profound. Four children had no light perception, and in all but two cases, the degree of vision loss was approximately equal in both eyes. Two children showed asymmetric vision loss, each with one eye retaining a visual acuity of 6/7.5 during the initial episode of neuritis. Papillitis was diagnosed ophthalmoscopically in seven children. All 10 children showed progressive optic atrophy (Figure 1). Systemic steroids were administered to seven children and did not appreciably affect speed of recovery or final visual acuity. The steroid regimens varied considerably.

Eight children had an imaging study of the central nervous system, six studies of which were normal. Two patients showed white matter abnormalities and in one, the magnetic resonance imaging (MRI) head scan showed changes compatible with acute disseminated encephalomyelitis (ADEM) (Figure 2).

The period of time for recovery of maximal vision varied, with the shortest interval of three months. Two children had maximal recovery after 18 months, and in one child, the best recorded vision was achieved after six years. On the other hand, one patient had no recovery in one eye after vision fell to no light perception. Two patients showed stepwise and progressive vision failure with no recovery of vision. Follow-up in these patients ranged from 18 months to 20 years.

The final visual acuities in these patients varied.

Table 1. Patient data

Patient number	Age (years)	Sex	Prodrome	Maximum visual loss	Treatment	Scan	Time to recover	Papillitis	Best acuity	Neurological symptoms
1	6	F	Stomach flu	NLP NLP	Steroids	Normal	6 years	Yes	6/12 6/9	No
2	6	F	No	NLP NLP	Steroids	Normal	18 months	Yes	6/9 6/15	No
3	14	F	Conjunc-tivitis	6/60 LP	Steroids	Normal	18 months	Yes	1/60 NLP	Paraesthesiae
4	14	F	Urinary tract infection	NLP NLP	Steroids	None	3-4 months	Yes	6/6 6/6	Ataxia, vertigo Transient
5	16	F	No	6/7.5 LP	None	None	1-2 months episodic	Yes	6/18 6/7.5	No
6	13	F	No	NLP 6/7.5	None	Normal	No recovery	No	NLP 6/7.5	Paralysis lower extremities
7	8	F	Viral illness	NLP NLP	None	White matter abnormality	Progressive	No	1/60 1/60	Hemiparesis, frequency, urgency
8	5	F	Fever	LP LP	Steroids	Normal	7 months	Yes	6/9 6/7.5	Ataxia
9	6	M	Fever, gastro-intestinal upset	LP LP	Steroids	White matter abnormality	6 months	No	6/7.5	Seizure, confusion
10	6	F	Viral illness	LP LP	Steroids	Normal	1 year	Yes	6/60 3/60	No

NLP = no light perception; LP = light perception; F = female; M = male.

One child recovered vision to 6/6 bilaterally, but no other eye in this series regained a visual acuity of 6/6. Seven of 20 eyes had a final acuity recorded at 6/60 or worse, and in three patients the best vision in either eye was only 6/60 or worse. Two eyes in two different patients had no light perception vision. The child with 6/6 vision is now an adult and reports poor colour, contrast and night vision.

Four of the children had no other symptoms or signs of neurologic dysfunction. Patient 3 had paraesthesias 18 months after the optic neuritis, and Patient 4 had an episode of ataxia and vertigo six months after the episode of optic neuritis. Patient 6 developed paralysis of the lower extremities, recurrent optic neuritis, and upper extremity paraesthesias, six years after the episode of optic neuritis. Patient 7 had a left hemiparesis three years later, recurrent bilateral optic neuritis, and frequency and urgency of urination five years later. Patient 8 had an episode of ataxia two years later, and Patient 9 had ADEM with an abnormal MRI head scan (Figure 2).

Discussion

Optic neuritis in children is usually bilateral and ophthalmoscopically anterior; that is, a papillitis.[1-4] The initial vision loss is profound,[5] and recovery of vision is usually rapid and complete, even after a reduction of vision to no light perception.[6] The risk of developing other neurologic signs and symptoms after childhood optic neuritis is small;[1,4] however this notion has been recently contested.[7]

Our patients, referred to UCSF by other physicians, have not followed the rule concerning good visual prognosis, for recovery was generally slow and often incomplete. Three patients are legally blind and only one regained 6/6 vision in both eyes. On the basis of this study, it is difficult to generalise that recovery from poor vision occurs in any notable proportion of cases. Optic neuritis in children is uncommon, and we have accumulated these 10 patients over a period of 20 years. It is possible that other patients with uncomplicated optic neuritis in this geographic region could have been managed elsewhere. Geographic differences in the incidence and severity of optic neuritis may exist as, for example, in the case of multiple sclerosis.[8]

Despite the limitations of this study from an epidemiologic perspective, we wish to emphasise that some children do not see well after an episode of optic neuritis, an observation which has not been emphasised in reports. Despite the optimistic report of Kennedy and Carroll, two of 30 children had no significant return of vision.[1] Meadows in 1969

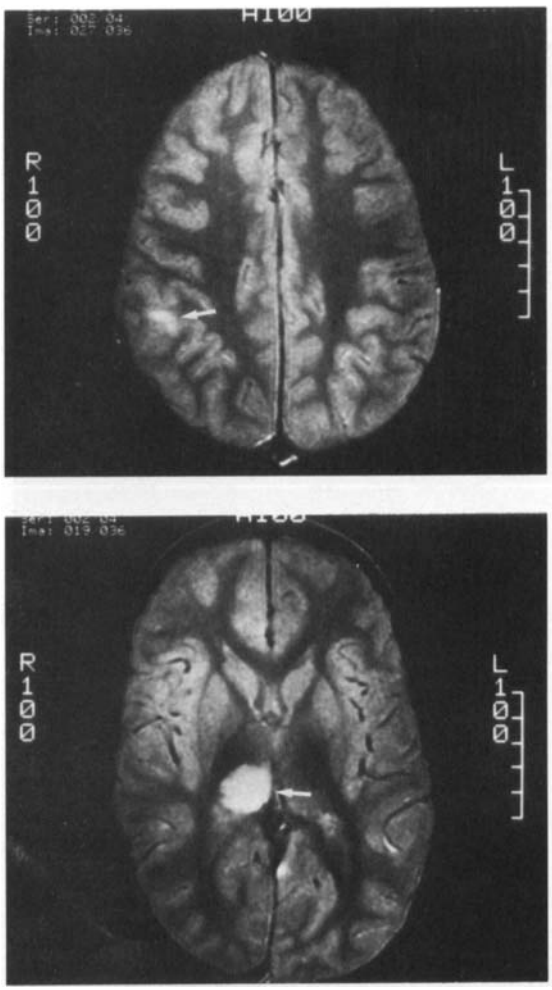

Figure 2 MRI scan of Patient 9 shows multiple white matter lesions (arrows). The lesions occurred acutely and were associated with bilateral optic neuritis. This patient had ADEM.

described 35 children with bilateral optic neuritis, four of whom had a poor visual outcome.[2] In the experience of Haller and Patzold, 18% of eyes were worse than 6/12 at follow-up.[9] In a recent publication, Kriss and associates[4] described two eyes in which recovery of acuity was poor (worse than 6/60), and visual field defects occurred in 35% of patients. Some degree of optic atrophy often occurred in all these studies, even with good return of visual acuity.

Given that some children will not have recovery of vision after optic neuritis, are there clinical features that might help in predicting a guarded prognosis? We examined a variety of patient characteristics including: prodromal febrile illness; maximum vision loss; administration of steroids; neuro-imaging studies; time to recovery; the presence of papillitis; final acuity; and presence of other neurological problems at the time of follow-up.

A prodromal viral illness, contrary to earlier speculations, did not correlate with a good visual or neurological outcome.[10] Four children with a febrile prodromal disease subsequently developed MS.

Two eyes with mild initial visual dysfunction resulted in acuities of 6/18 and 6/7.5. In Patient 5 there were several bouts of optic neuritis. Conversely, there were several eyes with no light perception (NLP) or light perception only (LP) that recovered to 6/15 or better (Patients 1,2,4,5,8 and 9). The degree of initial visual loss was not of prognostic significance in this series.

The use of steroids in childhood postinfectious optic neuritis has been the subject of a recent report,[10] wherein methylprednisolone was administered intravenously, resulting in a 100% rate of rapid and nearly complete recovery of vision. None of our cases received steroids intravenously, but treatment with a short course of high-dose prednisone failed to bring about either a rapid or complete recovery in six children. In one child treated with steroids, vision recovered to 6/6 after three months.

Our study does not support the value of steroid therapy in childhood optic neuritis. It is possible that steroid treatment failure is related to differences in the nature of their administration and/or the dosages employed. It should be noted that our patients had more severe optic neuritis which may be more refractory to steroid treatment. Optic neuritis may also be more responsive to steroids if it occurs in the setting of simultaneous neurological dysfunction, such as ADEM.

One child Patient 9 had a viral illness followed by bilateral optic neuritis and had radiographic evidence of other demyelinating foci, consistent with ADEM (Figure 2). The patient was treated with steroids and recovered vision slowly to 6/15 and 6/7.5. Children with optic neuritis deserve a careful neurological examination, and a neuro-imaging study of the central nervous system is indicated if there are other abnormal neurological findings. Since ADEM generally responds to steroids, their administration must be carefully considered.[11]

Two children had abnormal MRI head scans; it must be noted, however, that most children in this

study were evaluated before magnetic resonance imaging was available. This neuroimaging technique demonstrates optic neuritis and the segment of nerve involved in the disease process. Extensive lesions of the optic pathway, or lesions that involve the optic canal are generally associated with a poor visual prognosis in adult patients.[12] The possibility that MRI scanning could be used to predict outcome in children is certainly provocative and must be investigated further. Children with optic canal lesions, in which compression of the nerve from swelling is more likely, may be better candidates for the administration of steroids.

The time to recovery of maximal visual acuity was often prolonged in our series. Patient 1, diagnosed at six years of age, was followed twice yearly for six years until a best acuity of 6/12 and 6/9 was recorded. Early in the course of her disease, she was referred for low vision services and was unable to see large print. Eventually she received a sighted education. Even with good visual acuity, some visual impairment may be present. Patient 4, now an adult with 6/6 vision, describes poor colour vision and contrast sensitivity and is unable to drive at night, limiting her outdoor activity to hours in which there is direct sunlight.

Seven patients had a papillitis, a finding in agreement with earlier reports in children.[2-4] It is of interest that papillitis does not necessarily indicate anterior optic nerve disease. MRI scanning shows that papillitis can also occur with optic canal inflammatory foci.[12] Papillitis was of no value in predicting visual outcome in our series.

Other neurological signs and symptoms consistent with MS occurred in five children at least six months after optic neuritis.[13] Multiple sclerosis occurs in a small percentage of children[1-4] after optic neuritis. Riikonen and associates found MS in 43% of children with optic neuritis.[7] The risk of MS in an individual case cannot be determined from our small series, but a definite risk exists. In this study poor visual outcome did not necessarily correlate with the development of multiple sclerosis.

Should the general notion that the prognosis of optic neuritis in childhood is favourable be changed? Certainly, on the basis of these patients it should be qualified. For example, some children do not see well after optic neuritis, and those with 6/6 vision may still have some visual impairment. Moreover, optic neuritis can recur. The recovery of vision can be slow; however, the slow rate of vision recovery does not preclude achievement of vision of even 6/9. Multiple sclerosis can occur after optic neuritis in children. Unfortunately, we are unable to identify risk factors that might predict a good visual outcome, nor can we predict which children will develop MS. Newer neuroimaging studies may be of benefit in this regard.

References

1. Kennedy C, Carroll FD. Optic neuritis in children. Arch Ophthalmol 1960;63:747-55.
2. Meadows SP. Retrobulbar and optic neuritis in childhood and adolescence. Trans Ophthalmol Soc UK 1969;89:603-38.
3. Parkin PJ, Hierons R, McDonald WI. Bilateral optic neuritis. A long term follow-up. Brain 1984;107:951-64.
4. Kriss A, Francis DA, Cuendet F, Batchlor JR, McDonald WI. Recovery after optic neuritis in childhood. J Neurol Neurosurg Psychiatr 1988;51:1253-8.
5. Rush JA. Idiopathic optic neuritis of childhood. J Ped Ophthalmol Strab 1981;18:39-41.
6. Taylor D, Cuendet F. Optic neuritis in childhood. In: Hess RF, Plant GT, eds. Optic neuritis. Cambridge: Cambridge University Press, 1986:73-85.
7. Riikonen R, Donner M, Erkkila H. Optic neuritis in children and its relationship to multiple sclerosis. A clinical study of 21 children. Dev Med Child Neurol 1988;30:349-59.
8. Kurtzke JF, Beebe GW, Norman JE, Jr. Epidemiology of multiple sclerosis in U.S. veterans. I. race, sex, and geographic distribution. Neurology 1979;29:1228.
9. Haller P, Patzold U. Die optikusneuritis im kindesalter. Fortschr Neurol Psychiatry 1979;47:209-16.
10. Farris BK, Pickard DJ. Bilateral postinfectious optic neuritis and intravenous steroid therapy in children. Ophthalmology 1990;97:339-45.
11. Adams RD, Victor M. Principles of neurology. New York: McGraw-Hill, 1981:660.
12. Miller DH, Newton MR, van der Poel JC, du Boulay EP, Halliday AM, Kendall BE, *et al.* Magnetic resonance imaging of the optic nerve in optic neuritis. Neurology 1988;38:175-9.
13. Posner CM, Paty DW, Scheinberg L, McDonald WI, Davis FA, Ebers GC, *et al.* New diagnostic criteria for multiple sclerosis: Guidelines for research protocols. Ann Neurol 1983;13:227-31.