# Clinical Features and Outcome of Childhood Optic Neuritis at Queen Sirikit National Institute of Child Health

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***Objective:*** *To study the clinical features, treatment, outcome and factors affecting the outcome of optic neuritis in children.*
***Material and Method:*** *Children under 16 years of age diagnosed with optic neuritis (ON) at Queen Sirikit National Institute of Child Health over an 11-year period were reviewed. Demographic data, clinical characteristics, treatment and the outcome were analyzed.*
***Results:*** *Thirty-one patients fulfilled the inclusion criteria. Mean age of onset was 9.2 years. Female to male ratio was 1.8:1. All cases had vision loss, bilateral 74.2% and monocular 25.8 %, including decreased color vision 35% disc edema 54.8% and ocular pain 38.7%. The mean duration of follow-up was 20.38 months. The final diagnosis of these patients were 2 multiple sclerosis (MS), 2 neuromyelitis optica (NMO), 3 acute disseminated encephalomyelitis (ADEM) and 24 or isolated optic neuritis (ION). There was no statistically significant difference in gender, age of onset, number of ocular involvement, severity of visual acuity impairment, presented preceding infection, associated symptoms, CSF pleocytosis, high protein in CSF, abnormal brain or eye MRI. 93.3% of patients had clinical improvement, including 70% complete recovery visual acuity of both eyes. The statistically significance was that complete remission in females was more than in males.*
***Conclusion:*** *Childhood optic neuritis had bilateral vision loss and complete recovery. Females with optic neuritis had statistical significance with complete recovery, more than males.*

***Keywords:*** *Isolated optic neuritis, Multiple sclerosis, Clinical feature, Outcome*

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Optic neuritis (ON) is an inflammatory disorder of optic nerves, usually present with decreased visual acuity, periorbital pain and decreased color vision[1]. In children, optic neuritis can present as the initial symptoms of central nervous demyelinating diseases such as multiple sclerosis (MS), acute disseminated encephalomyelitis (ADEM), neuromyelitis optica (NMO) or isolated optic neuritis (ION). The incidence in children is geographically different and more studies are needed[2,3]. There are a few reports in the natural history of childhood optic neuritis and associated factors on the outcome.

Then objective of the present study was to analyze the clinical features, treatment, outcome and factors that associated with the outcome of childhood optic neuritis.

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## Material and Method

The retrospective study was done in patients with ON at QSNICH between 1st January 1999 and 31st December 2009. The define criteria of ON was acute or subacute visual loss and one or more of the following: relative afferent papillary defect in the affected eye, visual field defect or scotoma, impaired color vision or optic disc edema, in the patient's age at onset less than 16 years. All patients were diagnosed and treated at the Department of Neurology and Ophthalmology. Epidemiologic data, clinical features, treatment, outcome and the associated factors were analyzed with SPSS program. The Chi-square test was used to determine the relationship between the outcome and the possible associated factors. The diagnosis of MS, NMO was defined by McDonald criteria.

## Results

Thirty-one patients fulfilled the criteria of ON. The mean age at onset was 9.21 years. Range was 4-15 years. Female to male ratio was 1.8: 1. The mean duration of follow up was 20.38 months. None of the patients had underlying diseases or received vaccine before the visual loss and no familial history of multiple sclerosis. Nine (29%) patients had preceding infection prior to the vision loss, 4 upper respiratory tract infections, 2 gastrointestinal tract infections and 3 non-specific febrile illnesses. The initial symptom included 23 (74.2%) vision loss, 4 (12.9%) ocular pain, 3 (9.7%) limb weakness and 1 (3.2%) headache (Table 1).

All patients had visual loss which varied from blurred vision to total blindness, 74.2% bilateral and 25.8 % unilateral. The degree of the vision loss was defined as follows: mild low vision (VA < 20/70-20/200) in 20.96%, severe low vision (VA < 20/200-20/400) in 6.45% and blindness (< 20/400, finger count, hand movement, light perception and no light perception) in 45.15% (Table 2).

Brain and orbits with gadolinium contrast MRI was performed in 25 patients, showing abnormal in 19 (76%) patients. MRI of the orbits showed enhancement of the affected optic nerves in 13 (52%) patients and brain MRI showed white matter lesions on T2-weighted in 11 (44%) patients. Five patients had both abnormality of optic nerves and brain.

Lumbar puncture and cerebrospinal fluid examination performed in 14 patients were mild pleocytosis (7-15 cell/$mm^3$) in 3 patients and high CSF protein level (59 mg/dl) in 1 patient.

Six patients developed recurrent ON. Three of them aged 7.9, 9 and 7.5 years developed recurrent optic neuritis without other neurological deficit 1.5-3 years after the first attack, two patients were treated with methylprednisolone 1,200 mg/$m^2$/day for 3 day, one patient treated with dexamethazone 1 mg/kg/day for 3 days, followed with prednisolone 1 mg/kg/day for 11 days. Three of recurrent ON aged 5.8, 10 and 14 years developed recurrent ON and multifocal corticospinal tract deficit. By McDonald criteria 2 patients were MS and the other was NMO.

The final diagnosis of these patients was 2 MS (6.7%), 1 NMO (3.3%), 3 ADEM (10%) and 24 ION (80%). Comparison of isolated optic neuritis and the others (ADEM, MS, NMO), there was no statistically significant difference in gender, age of onset, number of ocular involvement, severity of visual acuity impairment, the presentation of preceding infection, CSF pleocytosis, high protein in CSF, abnormal MRI (Table 1).

Follow-up information was available in 30 cases that were in outcome analysis. 54.8% of patients had treatment within 14 days after the onset of visual loss. The treatment included 83.9% pulse methylprednisolone, 12.9% dexamethasone and 3.2% hydrocortisone. Most patients (93.3%) had clinical improvement, complete remission of normal visual acuity both eyes was found in 70%, no significant clinical improvement in 2 patients. There was statistical significance that females had more complete recovery of vision than males (Table1, 3).

## Discussion

Optic neuritis in children differs from the adult optic neuritis in clinical features. There were several studies of childhood optic neuritis reported that the mean age of onset 7-11 year, mostly female predominant, prodomal illness in 24-66%, bilateral eye in 37-86% and normal visual acuity outcome in 58-89% (Table 4)[1,2,4-12]. The present study supported these findings by mean age of onset 9.2 years, female predominant (female: male 1.8: 1), bilateral ocular involvement (74.2%), complete visual acuity recover (70%).

The recurrent optic neuritis was not uncommon. The present study revealed 20% of patients had recurrent ON which is similar to Visudhiphun P. report[6] that four of 22 patients (18%) had recurrent optic neuritis, 2 ION, 2 MS. The underlying of optic neuritis in the present study was MS only 2 patients (6.5%). No risk factors predict the diagnosis of MS was demonstrated. These was different from the studies which the subsequent developed MS 23-50%[5,9,11,13]. The risk factors for subsequent development of MS were an abnormal brain MRI, female, bilateral sequential or recurrent ON, older age at on set, oligoclonal bands in CSF and elevated immunoglobulin G index[1,10,14,15]. The studies which had high risk for development of MS and demonstrated the risk factors had the long follow-up time for 6-15 years. Whereas the present study had mean follow-up time 20.38 months which may be too short follow-up time for developed MS. However, the present study was similar to Visudhiphun P[6] and Hwang JM[7] reports that only 9% and 4.3% of ON in Thai children and Korean children consequently developed MS. The lower incidence of MS after ON in Thai and Korean children may be the difference in the race and genetic of Asian and Caucasian populations.

The outcome of the patients with optic neuritis

in the present study was good in visual acuity but the sequelae impairments of color and stereoscopic vision were similar as in previous studies[6-8,11,13,16-19]. Factors associated with a good prognosis in children were younger age, bilateral disease, and a normal MRI. The present study revealed that females had more complete recovery of vision than males which may be another good prognosis.

**Table 1.** Comparison of clinical features and outcomes in patient with ION and others (multiple sclerosis, acute disseminated encephalomyelitis, neuromyelitis optica)

| Characteristic | ION (24) | Others (7) | Total (n) | p-value |
|---|---|---|---|---|
| Sex | | | | 0.68 |
| Male | 8 | 3 | 24 | |
| Female | 16 | 4 | 7 | |
| Age (year) | | | | |
| < 6 | 3 | 3 | 6 | 0.11 |
| 6-12 | 16 | 2 | 18 | |
| > 12 | 5 | 2 | 7 | |
| Preceding infection | 6 | 3 | 9 | 0.19 |
| Initial symptoms | | | | |
| Decrease visual acuity | 20 | 3 | | |
| Ocular pain | 3 | 1 | | |
| Weakness | 0 | 3 | | |
| Headache | 1 | 0 | | |
| Clinical features | | | | |
| Decrease visual acuity | | | | 0.66 |
| Unilateral | 6 | 1 | 7 | |
| Bilateral | 18 | 6 | 24 | |
| Ocular pain | 9 | 3 | 12 | 0.56 |
| Headache | 11 | 3 | 14 | 0.62 |
| Vomiting | 5 | 1 | 6 | 1.0 |
| Fever | 2 | 2 | 4 | 0.2 |
| Optic disc edema | 14 | 3 | 17 | 0.38 |
| CSF Abnormal protein | 0 | 1 | 1 | 0.36 |
| CSF pleocytosis | 1 | 2 | 3 | 0.51 |
| MRI brain and orbit | | | | |
| Normal | 5 | 1 | 6 | 0.36 |
| Abnormal | 13 | 6 | 19 | |
| Eye | 12 | 1 | 13 | |
| Brain | 5 | 6 | 11 | |

**Table 2.** Degree of visual loss of the children with optic neuritis before treatment and after treatment

| Visual acuity | Before treatment (n = 31) | | After treatmen (n = 30) | |
|---|---|---|---|---|
| | Right eye | Left eye | Right eye | Left eye |
| 20/20-20/70 | 8 | 9 | 26 | 23 |
| < 20/70-20/200 | 7 | 6 | 2 | 5 |
| < 20/100-20/400 | 2 | 2 | 1 | 1 |
| < 20/400-Finger count | 4 | 6 | 1 | 1 |
| Hand movement | 3 | 2 | 0 | 0 |
| Light perception | 3 | 3 | 0 | 0 |
| No light perception | 4 | 3 | 0 | 0 |

**Table 3.** Factors and outcome with complete remission, normal visual acuity both eyes

| Characteristic | VA Complete remission | | p-value |
|---|---|---|---|
| | yes | no | |
| Sex | | | |
| Male | 4 | 7 | 0.004 |
| Female | 17 | 2 | |
| Age (year) | | | |
| < 6 | 4 | 2 | 1.0 |
| 6-12 | 12 | 5 | |
| > 12 | 5 | 2 | |
| Preceding infection | | | |
| No | 13 | 8 | 0.21 |
| Yes | 8 | 1 | |
| Decrease visual acuity | | | |
| Unilateral | 5 | 2 | 1.0 |
| Bilateral | 16 | 7 | |
| Degree of visual loss | | | |
| < Finger counte (number of effected eyes) | 19 | 8 | 0.69 |
| ≥ finger count | 17 | 15 | |
| Optic disc | | | |
| Normal | 11 | 2 | 0.23 |
| edema | 10 | 7 | |
| CSF Abnormal protein | | | |
| No | 10 | 3 | 1.0 |
| Yes | 1 | 0 | |
| CSF pleocytosis | | | |
| No | 9 | 2 | 1.0 |
| Yes | 2 | 1 | |
| MRI brain | | | |
| Normal | 5 | 1 | 0.62 |
| Abnormal | 12 | 7 | |
| Treatment | | | |
| Before 7 days | 9 | 2 | 0.42 |
| After 7 days | 12 | 7 | |
| medication | | | |
| Pulse Methylprednisolone | 19 | 6 | 0.22 |
| Dexamethasone | 1 | 3 | |
| Hydrocortisone | 1 | 0 | |

**Table 4.** Epidemiologic data, clinical features, outcome, developed MS of previous study in childhood ON

| | n | Age range (yr) | Mean age (yr) | Female /Male | FH of MS | Prodomal illness (%) | Bilateral eye (%) | Ocular pain | Abnormal MRIBrain (%) | Outcome: normal visualacuity (%) | Mean follow-up | Developed MS |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Kennedy & Carrol | 30 | 4-16 | 9.5 | 19/11 | - | 43.33 | | 14/30 | | - | 8 yr | 8/30 (26%) |
| Riikonen | 21 | 4-14 | 9.9 | 16/5 | 3/21 | 23.8 | 62 | | - | - | 6 yr | 9/21(42.8%) |
| Visudhiphan | 22 | 1-12 | 7.1 | 12/10 | | 27.3 | 86 | | - | 78 | | 2/22 (9%) |
| Lucchinetti | 94 | 2-16 | 11 | 58/36 | - | 41.5 | 42 | 35/94 | - | - | 22 yr | |
| Morales | 13 | 4-15 | 9.8 | 6/7 | - | 66 | 66 | | 33% | 58 | 17.5 mo | 4/15(26.6%) |
| Lana-Peixoto | 27 | 3-16 | 10.9 | 12/15 | - | 37.0 | 37 | 10/27 | 3/6 | 70 | 13 mo | 1/27 (3.7%) |
| Mizota | 41 | 2.5-15 | 9.4 | 23/18 | - | 39.0 | 49 | | - | 89 | 10.7 yr | 13/41(31.7%) |
| Hwang JM | 23 | 3-15 | 8.9 | 13/10 | - | 39.1 | 87 | | 4% | 79 | 14 mo | 1/23 (4.3%) |
| Chirapapaisan | 31 | 2-12 | 7.9 | 20/11 | - | 48.4 | 64 | 9/31 | 4/14 | 77 | 2.7 yr | 2/31 (6.4%) |
| This study | 31 | 4-15 | 9.2 | 20/11 | 0 | 29.0 | 74.2 | 4/31 | 11/25 | 70 | 20.4 mo | 2/30 (6.6%) |

## Potential conflicts of interest

None.

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## อาการทางคลินิกและผลการรักษาโรคประสาทตาอักเสบ (optic neuritis) ในเด็กในสถาบันสุขภาพเด็กแห่งชาติมหาราชินี

*สมจิต ศรีอุดมขจร, เก๋ พงศ์วัชราภรณ์*

***วัตถุประสงค์:*** *ศึกษาอาการทางคลินิก ผลการรักษาและปัจจัยที่มีผลต่อการรักษา optic neuritis ในผู้ป่วยเด็ก*
***วัสดุและวิธีการ:*** *ผู้ป่วยเด็กอายุน้อยกว่า 16 ปีที่วินิจฉัยว่าเป็น optic neuritis ตั้งแต่วันที่ 1 มกราคม พ.ศ. 2542 ถึง 31 ธันวาคม พ.ศ. 2552 โดยนำข้อมูลอาการทางคลินิก การรักษา ผลการรักษาและปัจจัยที่อาจมีผลต่อการรักษามาวิเคราะห์ หาความสำคัญทางสถิติ*
***ผลการศึกษา:*** *พบผู้ป่วย 31 ราย เป็น optic neuritis อายุเฉลี่ย 9.21 ปี อัตราส่วนเพศหญิงต่อชาย 1.8:1 อาการมองเห็นลดลงเป็นที่ตาทั้งสองข้างร้อยละ 74.2 ตาข้างเดียวร้อยละ 25.8 ความผิดปกติในการมองเห็นสีร้อยละ 35 ตรวจพบประสาทตาบวมร้อยละ 54.8 อาการปวดตาร้อยละ 38.7 ระยะเวลาการตรวจติดตามอาการเฉลี่ย 20.38 เดือน ได้การวินิจฉัยเป็น MS 2 ราย, NMO 2 ราย, ADEM 3 ราย ION 24 ราย เปรียบเทียบระหว่างกลุ่ม ION และกลุ่มอื่น ไม่พบความแตกต่างอย่างมีนัยสำคัญทางสถิติของ เพศ อายุที่มีอาการ จำนวนดวงตาที่มีอาการ ความรุนแรงของการสูญเสียการมองเห็น การเจ็บป่วยก่อนมีอาการสูญเสียการมองเห็น อาการร่วมอื่น จำนวนเม็ดเลือดขาวและโปรตีนในน้ำไขสันหลัง ความผิดปกติของสมองและตาจาก MRI ในผู้ป่วยทั้งสองกลุ่ม เมื่อได้รับการรักษาผู้ป่วยร้อยละ 93.3 อาการดีขึ้น ร้อยละ 70 มีสายตากลับมาเป็นปกติทั้งสองข้าง และพบว่าสายตาหายเป็นปกติใน เพศหญิงมากกว่าเพศชาย อย่างมีนัยสำคัญทางสถิติ*
***สรุป :*** *ประสาทตาอักเสบในเด็กมักมีอาการทั้งสองข้าง และมีพยากรณ์โรคดี การสูญเสียของสายตาเพศหญิงจะหายเป็นปกติมากกว่าเพศชาย อย่างมีนัยสำคัญทางสถิติ*