

## Clinical Article

## Management of Pseudotumor Cerebri in Children

Roberto Warman, MD

## Abstract

The experience with the management of pseudotumor cerebri at the Miami Children's Hospital in the past decade is presented. Twenty-two patients were identified; nineteen of which are included due to appropriate follow-up and data collection. No gender predominance was found and most cases were idiopathic without recurrences. Permanent visual loss did occur in some children in spite of surgical intervention with lumbar peritoneal shunt and optic nerve sheath fenestration. Pseudotumor cerebri in childhood does not present in the same fashion as in its counterpart adult and close follow-up is required to diagnose appropriately and intervene to prevent visual loss, which fortunately remains a rare occurrence. *Int Pediatr.* 2000;15(3):147-150.

*Key words:* pseudotumor cerebri

## Methods

We reviewed all the records of children 18 years of age or younger with CPT code 348.2 for pseudotumor cerebri from the Ophthalmology Division at Miami Children's Hospital. Twenty-two patients were identified but charts were available on only nineteen. The other three charts only had one evaluation without follow-up prior to 1993 and were not available for review. All cases had neuro-radiologic evaluation either in the form of CT scan or MRI scan and all had lumbar puncture with increased opening pressure. However, not all of the cases had the exact value available. As stated in other reports, the three characteristics required to make a diagnosis of pseudotumor cerebri were: normal or small size ventricles; normal cerebral spinal fluid composition, and increase intracranial pressure more than 200 mm of water by lumbar puncture.

Complete neuro-ophthalmologic examinations were performed in all cases including a visual acuity with best refraction correction with Snellen optotypes and in non-literate children with Allen symbols. Most patients had contrast sensitivity measured with the B-Vat computer, color vision testing with Ishihara plates, and quantitative visual fields either with the Goldmann or the Humphrey visual field system.

Ocular motility was evaluated in particular looking for cranial nerve palsies. Fundus examination was performed with fundus photography whenever possible.

## Results

A total of 19 patients fulfilled the criteria. Follow-up examination ranged from 1 month to 9.5 years (mean 4.5 years).

There were 10 males and 9 females whose ages ranged from 22 months to 18 years of age. All had bilateral papilledema with the exception of one patient 7 years of age with no papilledema, but with a right VI cranial nerve palsy and documented increased intracranial pressure on two occasions. All patients had normal CT scans and seven had normal MRIs. There were two patients with small ventricles. Four patients had bilateral VI cranial nerve palsies and five patients had unilateral VI cranial nerve palsy. Two had esotropia but a VI nerve palsy could not be diagnosed with full abduction present. One patient had an associated VII cranial

## Introduction

Pseudotumor cerebri is a condition characterized by symptoms and signs of increased intracranial pressure without evidence of mass lesion or hydrocephalus. It most frequently occurs in obese women of childbearing age. However, there are many series of children with pseudotumor cerebri.

This disorder affects 0.9 per 100 000 of the general population but 90 of 100 000 obese women. In the adult population it affects preferentially 8 to 1 women over men. In most series of children with pseudotumor cerebri there was no gender predominance and less frequent obesity compared to surveys of adults with tumor cerebri.<sup>1,2</sup>

Permanent visual impairment was infrequently described and a rare occurrence in children. The following study describes the experience of management of pseudotumor cerebri at Miami Children's Hospital in the past decade.

From the Division of Ophthalmology, Miami Children's Hospital, Miami, Florida USA

Address reprint requests to Division of Ophthalmology, Miami Children's Hospital, 3100 SW 62nd Avenue, Miami, Florida 33155-3009, USA (Dr Warman)

Table 1. - Cranial Nerve Deficits at Presentation

Cranial Nerve Palsy	No. of Patients
Bilateral VI cranial nerve palsy	4
Unilateral VI cranial nerve palsy	5
Acute esotropia with full abduction	2
Skew deviation	1
VII cranial nerve palsy	1

nerve palsy and a skew deviation and a right internal ophthalmoplegia (previously published report due to its unusual associations). Symptoms were headache and diplopia and in one case there was tinnitus. One patient had a previous history of retinopathy, which was prematurely treated with laser ablation. (Table 1)

The etiology remained idiopathic in most cases with the following exceptions; one case attributed to minocycline, another to cortisone withdrawal, two cases to chemotherapy for acute myelogenous leukemia (one with methotrexate intrathecally and another one with cys-platinum), one attributed to chorionic gonadotropin treatment and one had an associated ear infection with sphenoid sinusitis. This latter one did not have confirmation by MRI of sinus thrombosis. Only 4 patients had recurrences. All of them were idiopathic and one recovered fully while the other 3 were the only patients with residual visual deficit. The 3 patients that have permanent visual deficit were a three year, eight month old child who had optic atrophy develop after the first episode. The acuity on last examination was finger mimicking at one foot with a 2+ afferent pupillary defect. The patient was from out of the country and the optic atrophy developed at that time.

The second patient was a ten-year-old who developed bilateral optic atrophy in spite of lumbar peritoneal shunts on two occasions and bilateral optic nerve sheath fenestrations. Her final visual acuity was no light perception in the right eye and 20/200 on the left.

The third patient has a craniostomosis, mental retardation and developed pseudotumor cerebri after expansion of sutures requiring bilateral optic sheath fenestrations leaving him with moderate bilateral optic atrophy and ambulating visual function. There is another patient, 17 year old, who had normal visual acuity, contrast sensitivity and color plates. But on two occasions has demonstrated abnormalities on the Humphrey visual field. There is only a two-month short-term follow-up on this patient at the present time.

There were no significant differences in initial visual function, final visual function or optic nerve appearance in younger and older children.

## Discussion

The diagnosis of primary pseudotumor cerebri is usually established when the following modified Dandy criteria is met: (A) signs and symptoms of increased intracranial pressure; (B) absence of localized findings on neurological examination; (C) absence of deformity, displacement or obstruction of the ventricular system in otherwise normal neurodiagnostic studies, except for increased CSF pressure; (D) alert and oriented patient;<sup>4</sup> no other cause of increased intracranial pressure present.<sup>3</sup>

Several studies including one previous report from our own institution<sup>4</sup> have reported cases, particularly in young children, who have other neurological transient abnormalities but who otherwise fulfill the criteria for pseudotumor cerebri including VII cranial nerve palsies and IV cranial nerve palsies and a skew deviation.<sup>5</sup> Also, compared to adults who frequently have multiple recurrences and eventually have permanent visual dysfunction most cases in younger children occur only once and have full recovery.<sup>6,7</sup>

Table 2. - Classification of Pseudotumor Cerebri in Children<sup>3</sup>

1.	Primary pseudotumor cerebri
A.	No recognized cause (idiopathic pseudotumor cerebri or benign intra-cranial hypertension)
2.	Secondary pseudotumor cerebri
A.	Pseudotumor cerebri associated with neurological disease Dural venous sinus thrombosis (associated with otitis media, mastoiditis, or head trauma) Altered CSF composition (meningitis) Arteriovenous malformation draining into a venous sinus Gliomatosis cerebri
B.	Pseudotumor cerebri secondary to systemic disease Malnutrition Systemic lupus erythematosus Polyangiitis overlap syndrome Addison disease Severe anemia (aplastic or iron deficiency)
C.	Pseudotumor cerebri secondary to ingestion or withdrawal of exogenous agents Corticosteroid withdrawal Malnutrition or renutrition Tetracycline or minocycline therapy (used in teenagers to suppress acne) Vitamin A intoxication--often in adolescents who take Vitamin A or the synthetic vitamin A derivative isotretinoin for acne Nalidixic acid (used in the treatment of urinary tract infection and bacillary dysentery) Thyroxine replacement in hypothyroidism Danazol, Danocrine (used for endometriosis or autoimmune hemolytic anemia)
3.	Atypical pseudotumor cerebri
A.	Occult pseudotumor cerebri (no papilledema)
B.	Normal pressure pseudotumor cerebri
C.	Infantile pseudotumor cerebri

Table 2 shows the classification of pseudotumor cerebri in children according to Brodsky, Baker and Hamed. Our study confirms the findings of others that the male/female ratio in prepubescent children is approximately equal. In our study, all our male patients were eleven years or younger and we had three females between the ages of fourteen and eighteen. None of our patients were obese which has also been noted on previous reports of children with pseudotumor cerebri. (Table 3)

Infants and young children may present with irritability and somnolence. Ataxia is also frequently reported. Headaches are the most common complaint. However, it is followed closely by diplopia. Interestingly, ear aches and tinnitus are common in children and in adults. This should always raise the consideration of lateral venous sinus thrombosis although we could not determine it in the one patient that complained of tinnitus in our series. (Table 4)

It was thought that children could tolerate chronic papilledema well and visual loss with pseudotumor cerebri was extremely rare in the pediatric age group. Reports substantiate permanent optic atrophy in the pediatric population and our series supports this finding.<sup>8</sup> In two of our cases the loss of vision evolved over a period of only a few weeks and in one case, in spite of aggressive and urgent intervention. The assessment of progressive visual loss is more difficult in children who do not cooperate for visual field testing and this was probably the cause of the severe optic atrophy, which developed in one of our patients. (Table 5)

The three most commonly recognized causes of childhood pseudotumor cerebri are venous thrombosis, steroid withdrawal and malnutrition associated with refeeding. Only one of our cases could have been attributed to steroid withdrawal. It was the case, which presented with multiple neurological findings. Fortunately, he had full recovery and no recurrences. Otitis media, mastoiditis, and lateral sphenoidal thrombosis in childhood pseudotumor cerebri has long been recognized, however, they have decreased in recent years due to appropriate antibiotic early intervention. One of our patients did have sinusitis as an etiology and responded to appropriate antibiotic therapy. Whether children present with purely isolated idiopathic pseudotumor cerebri or the ones presenting with a presumable causation their presentation, management and final visual outcome are similar.

Most of our patients were treated with Diamox depending on the age, 50 mg/kg/day, most of the time tapering for three to four months. Three patients recovered on no medication and just observation following lumbar puncture. One patient did undergo sphenoid and mastoid surgery and as mentioned before, two patients underwent lumbar peritoneal shunt and two bilateral optic nerve-sheath fenestration. No side effects from the Diamox were documented. (Table 6)

**Table 3. - Age at Presentation**

Age	No. of Patients
One to five years	7
Six to ten years	7
Eleven to fifteen years	3
Sixteen to eighteen years	2

**Table 4. - Symptoms at Presentation\***

Symptom	No. of Patients
Headache	13
Diplopia	9
Irritability	5
Vomiting	3
Tinnitus	1
Decreased vision	1

\*Some children had more than one symptom.

**Table 5. - Pseudotumor Cerebri Disorders**

Disorder	No. of Patients
Minocycline	1
Steroid withdrawal	1
Methotrexate	1
Cys-platinum	1
Chorionic gonadotropin	1
Sinusitis	1

**Table 6. - Treatment of Patients**

Treatment	No. of Patients
Observation	3
Repeat lumbar puncture	1
Diamox	14
Lasix	1
LP Shunt	2
Optic nerve sheath fenestration	2

In children it is imperative to rule-out meningitis or meningoencephalitis and/or inflammatory optic neuritis, besides mass lesions through appropriate neuro-radiologic studies and lumbar puncture.

Our study confirms the finding that spontaneous resolution of pseudotumor cerebri appears to be more common in children than adults. In some children, pseudotumor cerebri resolves after a single lumbar puncture. However, because of the potential for permanent visual loss, children with pseudotumor cerebri should be followed as carefully as adults. Whether the combination of acetazolamide alone or

in combination with furosemide is more effective, it has not been clearly elucidated due to the scarcity of cases.

Current surgical treatment of pseudotumor cerebri is limited to optic nerve sheath fenestration and lumbar peritoneal shunt.<sup>8</sup> Shunt failures are frequent and shunt infections may be life threatening. Lumbar peritoneal shunting will relieve headaches whereas optic nerve sheath fenestration will mostly resolve optic nerve damage and papilledema. It is not exactly clear why unilateral optic nerve sheath fenestration will often eliminate papilledema bilaterally. However, it is also well documented that optic nerve sheath fenestration may close shortly postoperatively. Numerous studies have suggested that optic nerve sheath fenestration is the lowest risk, most effective way to restore or preserve vision in pseudotumor cerebri and has become the surgical treatment of choice, at least in adults. It has also been found in cases where lumbar peritoneal shunting is unsuccessful and we may have shown this in one of our cases in the series. However, optic nerve sheath fenestration only relieves headaches in about 2/3 of patients.

Indication for surgical intervention includes evidence of progressive optic neuropathy based on loss of visual acuity or visual field in spite of maximal medical therapy or worsening papilledema in a child who can not cooperate with the examination and/or shows severe optic neuropathy where the patient can not function normally and would be in jeopardy if further visual loss occurred.

### Conclusion

Pseudotumor cerebri in childhood is mostly idiopathic with resolution either spontaneously or after a short period of tapering dose of acetazolamide and/or with repeated lumbar punctures without recurrences. However, it certainly can

proceed to permanent visual damage in the form of optic atrophy within a short period of time which can be prevented if appropriate surgical intervention either in the form of optic nerve sheath fenestration or lumbar peritoneal shunt is performed at the appropriate time. Children in particular need a close follow-up and evaluation during their course of pseudotumor cerebri to minimize permanent visual loss.

### References

1. Dirge KB, Corbett JJ. Diagnosis and Management of Idiopathic Intracranial Hypertension (Pseudotumor Cerebri). Tusarj, Newman SA, Ed. *Neuro-ophthalmological Disorders diagnosed with work-up and management*. New York, N.Y.: Marcel Dekerd. 1995; 55-64.
2. Scott IV, Siatkowski RM, Eneyni, M. Brodsky, MC, Lam BL. Idiopathic Intracranial Hypertension in Children and Adolescents. *Am Ophthalmology*. 1997; 124:253-255.
3. Brodsky MC, Baker RS, Hamed LM. Swollen Optic Disc in Childhood. In: *Pediatric Neuro-ophthalmology*. New York: Springer - Berlag, Inc.; 1996; 76-87.
4. Warman R. Motility Problems Consistent with Pseudotumor Cerebri in Childhood. *International Pediatrics*. 1990; 5: 266-269.
5. Speer L, Pearlman J, Phillips PH, Cooney M, Repka MX. Fourth Cranial Nerve Palsy in Pediatric Patients with Pseudotumor Cerebri. *Am Ophthalmology*. 1999; 127-237
6. Lessell S. Pediatric Pseudotumor Cerebri (Idiopathic Intracranial Hyper-tension). *Survey of Ophthalmology*. 1992; 37:155-165.
7. Cinciripini GS, Donahue S, Borchert MS. Idiopathic Intracranial Hypertension in Prepubertal Pediatric Patients: Characteristics, Treatment and Outcome. *American Journal of Ophthalmology*. 1999; 127-178-182.
8. Phillips PH, Repka MX, Lambert SR. Pseudotumor Cerebri in Children. *Journal of American Association Pediatric Ophthalmology and Strabismus*. 1998; 2:33-38.

---

© Miami Children's Hospital 2000