

# RESEARCH ARTICLE

## 32 CASES OF PSEUDOTUMOR CEREBRI IN TWO PEDIATRIC HOSPITALS OF TEHRAN, IRAN

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### Abstract:

#### Objective:

Pseudotumor Cerebri (PTC) is a clinical syndrome characterized by increased Intra-Cranial Pressure (ICP) without any evidence of a mass lesion or any obstructive process. The incidence is 0.9-2 per 100.000 people. It is more frequent in adults in the 20-30 years age group, especially obese women, and less common in pediatric age groups; 11-16 years old children may however suffer from PTC, with no difference in the rates of occurrence in either sex.

#### Material & Methods:

In this descriptive retrospective study we reviewed the files of 32 children with diagnosis of PTC admitted during the past 15 years in the neurology ward of the Mofid Children's Hospital (25) and Ali Asghar Children Hospitals (7) between the years 1988 and 2003.

#### Results:

The results of this study revealed that children in the 5-10 years age group, girls in particular, are more vulnerable.

#### Conclusion:

The most frequent complaints that brought these patients to physician included headache, vomiting and strabismus. Common findings of neurological examination were papilledema, abducent nerve palsy and ataxic gait. While all cases recovered with medical treatment, one did need surgical intervention.

Abbreviations; Pseudotumor Cerebri = PTC; Intra Cranial pressure = ICP.

Keywords : PTC , papilledema , ICP.

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### Introduction

Pseudotumor Cerebri (PTC), a clinical syndrome characterized by increased Intra Cranial Pressure (ICP) without any evidence of mass lesion or obstructive process (1), was reported by Quinke in 1893 (2).

The incidence is 0.9-2 per 100.000 people, being more frequent in adults of the 20-30 years age group (especially obese women), and less common in pediatric age groups. However children 11-16 years of age may suffer from PTC, with similar rates of occurrence in either sex (4,5). Considering the lack of literature on PTC in children in our country, this study was conducted with the aim of obtaining better understanding of diagnosis, clinical condition and treatment of PTC in our children .

### Material & Methods

In this descriptive retrospective study we reviewed the files of 32 patients diagnosed with PTC, hospitalized in the neurology department of the Mofid children's hospital (no=25) and the Ali Asghar Children's Hospital (no=7), between the years 1988 and 2003.

### Results

The results of this study revealed that children of the 5-10 year age group (43.5%), girls in particular (56%) are more vulnerable (Table I ).

Table I: Prevalence of 32 PTC patients, according to age and sex.

Age Group	Female		Male		Total	
	NO.	%	NO.	%	NO.	%
0-5	4	12.5	3	10	7	22.5
5-10	8	25	6	18.5	14	43.5
10-15	5	15.5	4	12.5	9	28
15-20	1	3	1	3	2	6
TOTAL	18	56	14	44	32	100

There were more females than males in this study; weight of a third of the patients was under the 5 percentile (33%)(table II).

Table II: Prevalence of sex and weight percentile of 32 PTC patients

Weight Percentile	Female		Male		TOTAL	
	NO.	%	NO.	%	NO.	%
<5	8	24	3	9	11	33
5-10	3	9	3	9	6	18
10-25	2	7	4	13	6	20
25-50	5	16	2	7	7	23
51-75	0	0	1	3	1	3
75-100	0	0	1	3	1	3
TOTAL	18	56	14	44	32	100

The most frequent complaints that brought these patients to physician, consisted of headache (85%), strabismus (67%) and vomiting (56%) (Table III).

Table III: Prevalence of signs in 32 PTC patients.

Sign	Number	Percent
Headache	27	85%
Strabismus	21	67%
Vomiting	18	56%
Ataxia	9	28%
Diplopia	6	19%
Vertigo	6	19%
Mouth deviation	3	9%
Speech Problem	2	6%
Total	32	100%

The most common findings at neurological examination were papilledema (100%), abducent nerve palsy (bilateral - 47%, unilateral - 18%) and ataxia 28% (Table IV).

Table IV: Prevalence of the clinical symptoms 32 PTC patients

Symptom	Number	Percentile
Papilledema	32	100%
Bilateral 6 <sup>th</sup> nerve palsy	15	47%
Unilateral 6 <sup>th</sup> nerve palsy	6	18%
Ataxia	9	28%
Bilateral 7 <sup>th</sup> nerve palsy	2	6%
Unilateral 7 <sup>th</sup> nerve palsy	3	9%
Cerebellar dysfunctions test	3	9%
Hyper reflexia	4	13%
Abnormal plantar reflex	3	9%
Total	32	100%

Etiological factors found to be responsible were infectious diseases (66%); upper respiratory infections(URI), otitis media and urinary tract infections, followed by drugs medications and lipid soluble vitamins. All except one of the PTC cases recovered with medical treatment; this one case required surgical treatment (Table V).

Table V: Types and percentages of various treatments used for 32 PTC patients

Treatment procedure	Number	Percent
Mannitol + corticosteroid + Acetazolamide	5	15
Mannitol +Corticosteroid	11	35
Corticosteroid +Acetazolamide	3	10
Acetazolamide	7	22
Corticosteroid	5	15
Surgical procedure	1	3
Total	32	100

One of the patients, who had recurrence one year after medication was discontinued, suffered from acute lymphocytic leukemia.

### Discussion

Literature reviews show PTC to be a disease with a wide spectrum; at one end of which is an infant with a transient increase of ICP, alleviated by lumbar puncture and at the other more grave conditions with very high ICP and visual complications, which may require surgical procedures (6).The ratio of male to female frequency was 1.3:1, being higher in males, a finding which differs from that of other articles, in which the ratio is almost equal. Whereas the weight of most patients in our study was under the 5 percentile, results of other studies show PTC to be more frequent in adolescents, especially females (4). The etiology of PTC, reported in different researches, are infectious diseases, drugs, metabolic and endocrine diseases (7, 8, 9). Other causes of PTC are mild head trauma, corticosteroid withdrawal, Hypo- and hyper thyroidism, parathyroid disease, addison, Systemic Lupus Erythmatous, carbon dioxide retention secondary to

chronic lung diseases (10, 11). In children under the age of 20 years, thrombosis of one or more of dural sinuses, particularly the lateral, as a complication of otitis media or mastoiditis is probably still one of the most common abnormalites (12, 13).

This syndrome may also occur secondary to hypervitaminosis A, anemia and in contraceptive pile users (14, 15). In our study the most common etiologic findings were URI, otitis media, and then hypervitaminosis A&D and antibiotic usage, e.g. nalidixic acid, nitrofurantoin.

Patients with PTC develop non specific signs of increased ICP, headache-50-75%, nausea and vomiting 35-38%, blurred vision and diplopia 27-35%, cerebral nerve palsy, (especially the 6th )24-47%, and papilledema in the majority of cases (16, 17, 18) .

Cerebro-Spinal Fluid results are generally normal, although mild pleocytosis is not uncommon (19). The signs and symptoms of PTC in this study were very similar to those of others (Table III, IV). All patients of this study had normal cerebrospinal fluid , but 3 of them shown pleocytosis.

Generally imaging studies reveal ventricular system that is midline and normal or mildly reduced in size (4, 20). In this research brain CT scans of most patients were normal although 5 of them shown ventriculomegaly and the ventricular size had reduced in 3.

The treatment of PTC varies according to severity of diseases and degree of visual impairment. It is directed towards the monitoring of ICP to within near normal limits to reduce the danger of permanent visual damage (20-21). The first therapeutic step is to treat the underlying precipitant factors, like vitamin A or tetracycline discontinuation if possible (10-22-23).

41% of patients, in this study used drugs such as vitamin A, Nalidixic Acid and corticosteroids; these were discontinued after diagnosis. All cases of this study recovered with medical treatment except for one who required surgical intervention. One patient had recurrence one year after discontinuation of treatment.

### Conclusion

More attention needs to be paid to clinical training of physicians for diagnosis of PTC in patients with headache and other signs and symptoms of PTC and those with histories of drug and lipid soluble vitamins usage, and diseases that are precipitant factors of pseudotumor cerebri.

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