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Evaluation of clinical, radiological characteristics, and treatment outcomes of pediatric pseudotumor cerebri syndrome cases

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MAIN POINTS

- Headache, vomiting, and visual disturbances are the most common symptoms of pediatric PTCS.
- High-dose acetazolamide, initiated early, can prevent long-term visual complications.
- MRI findings such as empty sella and optic nerve sheath distension support diagnosis in the absence of papilledema.
- Secondary PTCS is often associated with medications (e.g., cyclosporine) and vitamin imbalances.
- OCT and visual field testing are critical for monitoring treatment response.

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ABSTRACT

Aim: This study aims to evaluate the clinical, radiological, and treatment outcomes of pediatric patients diagnosed with Pseudotumor Cerebri Syndrome.

Materials and Methods: This retrospective study included 21 children diagnosed with definite or possible Pseudotumor Cerebri Syndrome according to the revised Friedman diagnostic criteria, out of 115 patients evaluated between 2017 and 2019 at a pediatric neurology clinic. Clinical findings, radiological features, treatment responses, and prognosis were analyzed.

Results: The study cohort was predominantly female (85.7%), with a mean age of 12.8 years. The most common presenting symptoms were headache, visual disturbances, and vomiting. A key diagnostic finding was elevated cerebrospinal fluid (CSF) opening pressure (>280 mmH_2O), observed in 67% of patients. Two-thirds of the cases (66.6%) were classified as Primary Pseudotumor Cerebri Syndrome, while the remaining third (33.4%) were secondary, most frequently attributed to drug exposure or vitamin imbalances. The majority of patients showed significant clinical improvement following medical treatment with high-dose acetazolamide. Only a few required adjunctive therapies such as topiramate or corticosteroids, and recurrence was documented in only a single case. Additionally, MRI findings supportive of the diagnosis were present in a subset of patients.

Conclusion: Early diagnosis and treatment of Pseudotumor Cerebri Syndrome in children can prevent permanent visual complications. A multidisciplinary approach, including ophthalmology and pediatric neurology, is essential for accurate diagnosis, effective treatment, and favorable outcomes.

Keywords: Pseudotumor cerebri syndrome, Pediatric neurology, Increased intracranial pressure, Acetazolamide treatment, Papilledema

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INTRODUCTION

Pseudotumor cerebri syndrome (PTCS), also known as idiopathic intracranial hypertension, is a rare but significant condition in children, with an incidence of approximately 0.6–0.71 per 100,000 [1,2]. It is characterized by increased intracranial pressure (ICP) in the absence of structural brain lesions, hydrocephalus, or central nervous system infection, and typically presents with headache, nausea, vomiting, and visual disturbances [3].

The etiology is multifactorial; obesity, rapid weight gain, certain medications (e.g., tetracyclines, corticosteroids, vitamin A derivatives), and systemic diseases are among the risk factors [4–6]. One proposed mechanism is the impaired absorption of cerebrospinal fluid (CSF) at the level of the arachnoid villi [7-9].

In 2013, Friedman et al. [2] proposed revised diagnostic criteria incorporating clinical, radiological, and CSF pressure measurements, which are now widely used in pediatric practice. Early diagnosis is of critical importance, as delayed treatment may lead to irreversible vision loss [10,11].

This study aims to describe the demographic, clinical, radiological, and treatment characteristics of pediatric PTCS pa-

tients followed in a tertiary care center in Turkey and to compare the findings with established diagnostic frameworks, including the revised Friedman criteria.

MATERIALS AND METHODS

This retrospective study was conducted at the Pediatric Neurology Department of Necmettin Erbakan University Meram Medical Faculty between January 2017 and January 2019. Patient records were reviewed based on International Classification of Diseases (ICD) codes H53, H54.0, H47.1, R51, and G93.2 to identify cases potentially related to increased intracranial pressure.

A total of 115 pediatric patients were initially identified. Of these, 21 met the revised Friedman criteria for definite or possible PTCS. Data collected included age, gender, presenting symptoms, imaging findings, etiology, treatment modalities, and clinical outcomes.

The required minimum sample size was calculated using G*Power 3.1 software for a two-tailed Mann–Whitney U test, with an alpha error of 0.05, a desired power of 0.80, and a large effect size (Cohen's d = 0.8) based on previous pediatric PTCS studies. This calculation yielded a minimum sample size requirement of 42 participants (21 per group). In our study, 21 children met the diagnostic criteria for PTCS. Post-hoc power analysis for the comparison of CSF opening pressure between definitive and possible PTCS cases demonstrated a very large effect size (Cohen's d = 2.58) and a power of 0.998, indicating that our sample size was sufficient to detect clinically significant differences in this primary outcome despite the rarity of the condition.

Lumbar punctures (LP) were performed in the lateral decubitus position under sedation by the same pediatric neurologist, and cerebrospinal fluid (CSF) opening pressures were recorded. A CSF opening pressure $\geq$280 mmH_2O was considered elevated [1,12].

Vitamin D deficiency was defined as <20 ng/mL, and insufficiency as 20–30 ng/mL. Normal vitamin A levels were considered 26–49 μg/dL. Papilledema severity was graded as mild (grades 1–2) or severe (grades 3–4).

Neuroimaging findings were evaluated based on six criteria from Friedman's diagnostic framework: empty sella, optic nerve tortuosity and perineural fluid distension, optic disc flattening or bulging, Meckel's cave and cavernous sinus enlargement, transverse sinus stenosis, and enlarged arachnoid villi. All imaging studies were reviewed and interpreted by a single pediatric radiologist.

Treatment responses were assessed through clinical examination, optical coherence tomography (OCT), and visual field testing. Patients were followed at 1, 3, 6, and 12 months after treatment initiation. Recurrence within the first-year post-treatment was also evaluated.

Ethical approval was obtained from the Non-Pharmaceutical and Non-Medical Device Research Ethics Committee of Necmettin Erbakan University Meram Medical Faculty (Approval No: 2019/1657).

Statistical analysis

Statistical analyses were performed using IBM SPSS Statistics for Windows, Version 21 (Armonk, NY: IBM Corp.). Descriptive data are presented as frequencies and percentages. Continuous variables were expressed as median (minimum–maximum) in line with data distribution, and compared between primary and secondary PTCS groups using the Mann–Whitney U test for nonparametric data and the chi-square test (exact method) for categorical variables. A p-value <0.05 was considered statistically significant.

RESULTS

A total of 115 pediatric patients were screened, and 21 were diagnosed with definite or possible Pseudotumor Cerebri Syndrome (PTCS) according to the revised diagnostic criteria by Friedman et al. [2]. Of these, 18 (85.7%) were female and 3 (14.3%) were male, with a mean age of 12.8 years (range: 3–17 years). The most common presenting symptoms were headache, vomiting, and blurred vision. Elevated cerebrospinal fluid (CSF) opening pressure (>280 mmH_2O) was observed in 67% (14/21) of patients.

Fourteen patients (66.7%) were classified as primary PTCS, and seven (33.3%) as secondary PTCS. In the secondary group, drug exposure—particularly cyclosporine and growth hormone therapy—was the leading etiology (43%) [13]. Due to incomplete medical records, obesity status could not be assessed for patients in the primary PTCS group.

Seven patients (33.3%) were categorized as "possible PTCS." The demographic characteristics, clinical findings, and diagnostic classification details are presented in Table 1. In patients without papilledema or with normal CSF pressure, brain MRI findings were evaluated separately. Severe papilledema (grades 3–4) was present in 8 patients (38.1%) Table 2. Radiological findings among "possible PTCS" patients included optic disc flattening/bulging in 5 patients (71%), empty sella in 4 patients (57%), optic nerve tortuosity with perineural fluid accumulation in 2 patients (29%), Meckel's cave and cavernous sinus enlargement in 1 patient (14%), and enlarged arachnoid villi in 1 patient (14%) [14,15]. Transverse sinus stenosis was not observed in any patient, likely due to incomplete MR venography studies. Representative MRI findings of PTCS patients are shown in Figure 1.

Initial treatment consisted of high-dose acetazolamide (30 mg/kg/day in three divided doses). Topiramate was used in patients with acetazolamide intolerance or insufficient response. Of the 12 patients initially treated with acetazolamide, 2 required switching to topiramate due to metabolic acidosis. In 3 patients with elevated CSF pressure and severe papilledema (grades 3–4), corticosteroids were added to acetazolamide. In 4 patients, treatment was limited to removal of

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Table 1. Demographic, etiological, and clinical characteristics of the patients.

Patient	Sex	Age (years)	Complaint	NE	ES	MRI Findings	BOSp (cmH_2O)	Etiology	Additional Findings	Friedman Diagnosis
1	F	9	Headache + Diplopia	N	+	None	35	Primary	-	Definitive
2	F	15	Headache, N/V, VLS	N	+	3*	35	Secondary	Cyclosporine	Definitive
3	F	10	Headache, N/V, VLS	N	+	2,3*	12	Primary	-	Possible
4	F	9	Headache, N/V, VLS	N	+	1*	25	Primary	-	Possible
5	F	11	Headache, N/V, VLS	N	+	None	22	Primary	-	Possible
6	M	8	Headache, N/V, VLS	N	+	1,2*	29	Secondary	Vitamin A deficiency	Definitive
7	F	13	Headache, N/V, VLS	N	+	1,2*	28	Primary	-	Definitive
8	F	14	Headache, N/V, VLS	N	+	1,2*	32	Secondary	Hyperthyroidism	Definitive
9	F	17	Headache, N/V, VLS	N	+	None	35	Secondary	Vitamin B12 deficiency	Definitive
10	F	15	Headache, N/V, VLS	N	+	None	40	Primary	-	Definitive
11	M	16	Headache, N/V, VLS	N	+	1,2*	28	Primary	-	Definitive
12	F	17	Headache, N/V, VLS	N	+	2*	31	Primary	-	Definitive
13	F	17	Headache, N/V, VLS	N	+	1,3*	15	Primary	-	Possible
14	F	16	Headache, N/V, VLS	N	+	None	38	Primary	-	Definitive
15	F	11	Headache, N/V, VLS	N	+	6*	24	Secondary	Growth hormone	Possible
16	F	17	Headache, N/V, VLS	N	+	3,4*	26	Primary	-	Possible
17	M	3	Headache, N/V, VLS	N	+	1-4*	28	Secondary	Growth hormone	Definitive
18	F	15	Headache, N/V, VLS	N	+	4*	31	Secondary	DiGeorge Syndrome	Definitive
19	F	11	Headache, N/V, VLS	N	+	1,2,3*	-	Primary	-	Possible
20	F	17	Headache, N/V, VLS	N	+	None	32	Primary	-	Definitive
21	F	9	Headache, N/V, VLS	N	+	None	29	Primary	-	Definitive

*NE: Neurological Examination; ES: Eye Signs (Papilledema); MRI: Magnetic Resonance Imaging findings; BOSp: Cerebrospinal Fluid Opening Pressure; AF: Additional Findings; N/V: Nausea/Vomiting; VLS: Visual Loss Symptoms.

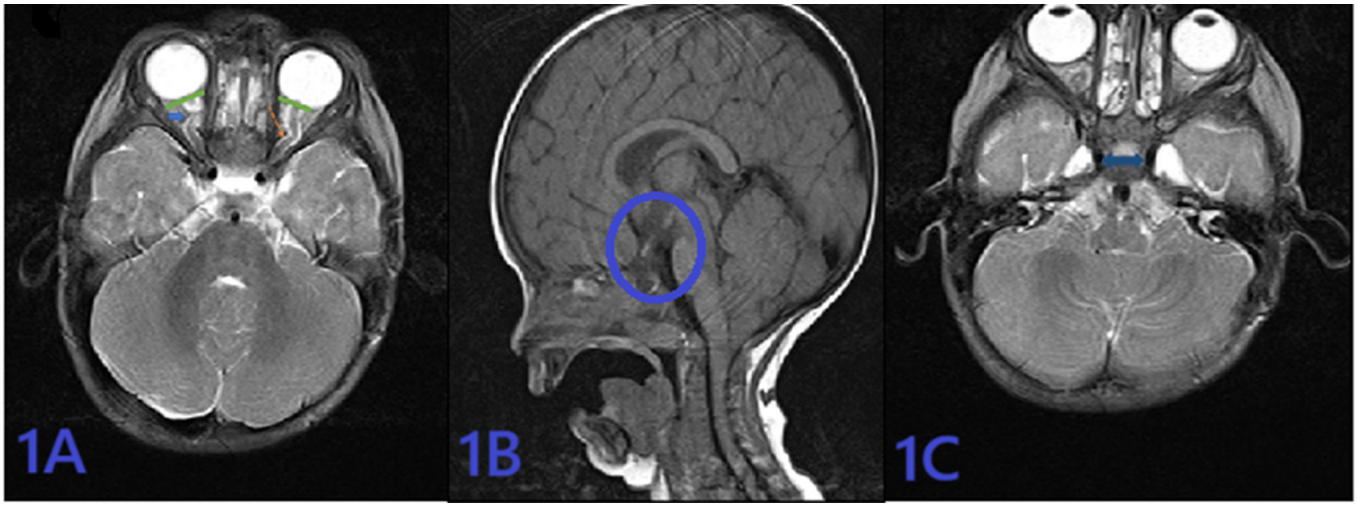

Figure 1. Brain MRI Findings in Patients with PTCS; 1A: Axial T2-weighted images showing bilateral optic nerve tortuosity, increased perineural fluid, and flattening/bulging of the optic disc. 1B: Sagittal T1-weighted images showing an empty sella appearance with the suprasellar cistern extending toward the pituitary. 1C: Axial T2-weighted images demonstrating bilateral enlargement of Meckel's cave spaces.

the underlying etiology (e.g., discontinuation of growth hormone therapy, correction of vitamin deficiencies) [16,17].

Treatment monitoring included clinical evaluation, optical coherence tomography (OCT), and visual field testing. Follow-up visits were scheduled at week 2, and at 1, 3, and 6 months, with a final evaluation at 1 year. Treatment adjustments were made based on clinical status at week 2 and visual field results at month 1. Most patients demonstrated marked improvement within 3 months. Patients with mild papilledema (grades 1–2) had no visual field loss and recovered by month 1, while those with severe papilledema (grades 3–4) improved by month 3.

Only one patient, with secondary PTCS due to cyclosporine toxicity, experienced recurrence within the one-year follow-up period [13]. Patients with mild papilledema and no significant MRI findings responded well to medical treatment alone. Conversely, patients with severe papilledema or notable MRI findings required lumbar puncture and closer follow-up.

No statistically significant differences were found between primary and secondary PTCS groups regarding CSF opening pressure (p=0.80), sex (p=0.18), or age (p=0.30) Table 3. A graphical summary of patient selection, classification, clinical and radiological findings, and treatment outcomes is provided

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Table 2. Characteristics of patients defined according to possible PTCS criteria.

Etiology	Papilledema Grade	CSF Pressure (cmH_2O)	Brain MRI Findings*
Primary PTCS	Grade 3	12	2,3
Primary PTCS	Grade 2	25	1
Primary PTCS	Grade 2	22	-
Primary PTCS	Grade 2	15	1,3
Primary PTCS	-	26	3,4
Primary PTCS	Grade 2	-	1,2,3
Secondary PTCS†	Grade 2	24	6

*MRI findings based on PTCS-specific criteria: (1) Empty sella, (2) Optic nerve tortuosity with increased perineural fluid, (3) Optic disc flattening and bulging, (4) Meckel's cave and cavernous sinus enlargement, (5) Transverse sinus stenosis, (6) Enlarged arachnoid villi. † Secondary PTCS due to growth hormone use.

Table 3. Demographic and Cerebrospinal Fluid (CSF) pressure characteristics of primary and secondary PTCS patients.

Characteristics	Primary PTCS (n=14)	Secondary PTCS (n=7)	p-value
Gender (F/M)	13 / 1	5 / 2	0.18
Age (years)	13(9-17)	15(3-17)	0.30
CSF Pressure (cmH_2O)	26(12-40)	28.5 (15-38)	0.80

in Figure 2.

DISCUSSION

This study provides a comprehensive overview of pediatric Pseudotumor Cerebri Syndrome (PTCS), reaffirming key clinical, radiological, and therapeutic characteristics in agreement with previously published literature. Similar to earlier reports, our data show a predominant female representation (85.7%) and a mean age of 12.8 years, highlighting the susceptibility of adolescent girls to PTCS [1,3,4,17]. Headache, vomiting, and visual disturbances were the most frequent presenting symptoms, consistent with hallmark features of increased intracranial pressure (ICP) described in prior studies [3,9,18].

Two-thirds of our cohort were diagnosed with primary PTCS, and one-third with secondary PTCS, a distribution comparable to earlier epidemiological findings [6,15,19]. Secondary cases were most often associated with cyclosporine use, growth hormone therapy, and vitamin imbalances (particularly excess vitamin A and vitamin D deficiency) [16,13,20]. The only recurrence during follow-up occurred in a patient with cyclosporine-induced PTCS, underlining the necessity of identifying and eliminating underlying etiologies [13].

Papilledema grade emerged as an important prognostic indicator. Severe papilledema (grades 3–4) was linked to more pronounced symptoms, higher rates of visual field defects, and increased need for combination therapy—findings consistent with Tovia et al. and other literature that emphasize the correlation between papilledema severity and treatment intensity [10,21].

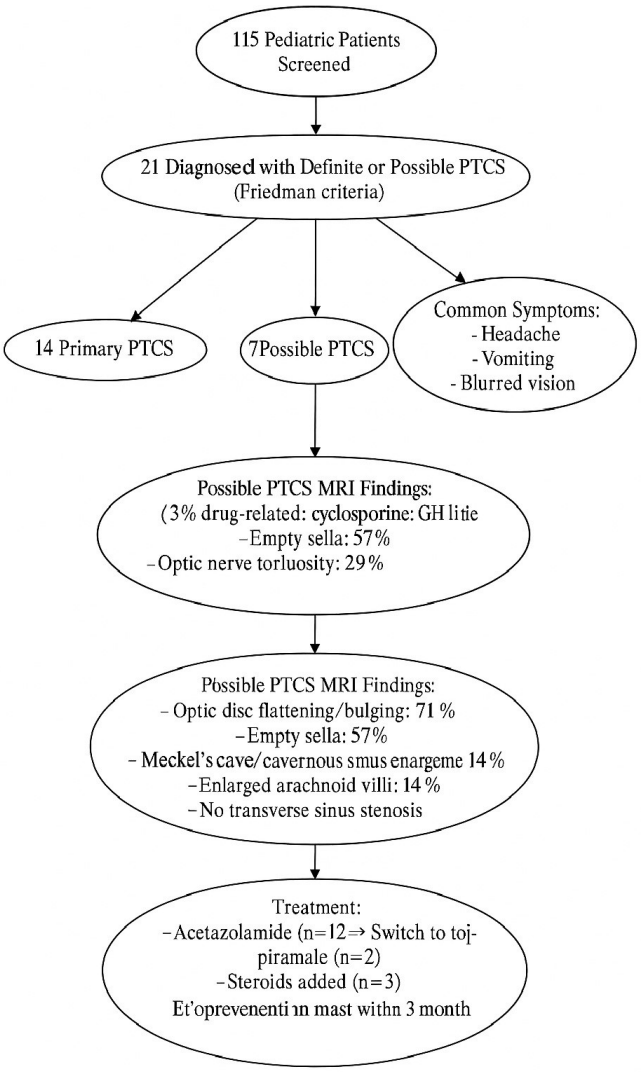

Figure 2. Study Flow Diagram; Flowchart illustrating the study process, from initial screening of 115 pediatric patients to final classification into definite or possible PTCS according to the revised Friedman criteria. The figure summarizes demographic distribution, key presenting symptoms, cerebrospinal fluid (CSF) pressure findings, major radiological signs, and the division into primary and secondary PTCS groups. Treatment modalities (acetazolamide, topiramate, corticosteroids, and etiology-specific interventions) and follow-up outcomes, including recurrence, are also depicted.

Neuroimaging played a pivotal role, especially in "possible PTCS" cases without papilledema. The most common findings—optic disc flattening, empty sella, and optic nerve sheath distension—align with the revised Friedman criteria [2] and were similarly highlighted by Görkem et al. and Maralani et al. [14,15]. Transverse sinus stenosis was absent, likely due to incomplete MR venography, as noted in other cohorts [15,20].

Pharmacological management yielded favorable outcomes. High-dose acetazolamide remained the primary therapy and was effective in most cases, confirming results from Celebisoy et al. [11]. Topiramate and corticosteroids were effective alternatives when acetazolamide was ineffective or poorly tolerated [12]. Only one patient required surgical intervention,

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supporting prior observations that medical therapy is generally sufficient in pediatric PTCS [10].

Continuous ophthalmologic monitoring using optical coherence tomography (OCT) and visual field testing allowed for objective assessment of treatment efficacy and early detection of complications, as emphasized in pediatric PTCS management guidelines [17,21].

Our results align with international findings but also provide region-specific data from a Turkish tertiary center. Comparable demographic and clinical patterns reported by Değerliyurt et al. and Sager et al. support the applicability of the Friedman criteria in diverse populations [20,21].

Strengths of this study include standardized diagnostic protocols, consistent follow-up, and a uniform treatment approach. Limitations involve the retrospective design, single-center scope, relatively small sample size, and incomplete data on obesity and MR venography, which may have restricted further subgroup analysis.

Despite these limitations, our findings contribute to the understanding of pediatric PTCS, underscore the value of Friedman's diagnostic framework, and emphasize the importance of multidisciplinary management involving pediatric neurology, ophthalmology, and radiology to optimize outcomes.

CONCLUSION

This study highlights the clinical and radiological spectrum of pediatric PTCS and supports the diagnostic utility of the revised Friedman criteria. High-dose acetazolamide is confirmed as an effective first-line therapy, with OCT and visual field monitoring essential for preventing long-term visual impairment. Radiological features particularly optic disc bulging and empty sella are useful in diagnosing cases without papilledema. Radiological examples (Figure 1) illustrate the role of MRI in confirming diagnosis. A multidisciplinary approach ensures timely intervention and improved prognosis. Further multicenter, prospective studies are warranted to validate and expand these findings.

Ethics Committee Approval: This study was approved by the Ethics Committee of Necmettin Erbakan University Meram Faculty of Medicine (Approval No: 2019/1657).

Informed Consent: Since the study is a retrospective study, informed consent is not required.

Peer-review: Externally peer-reviewed.

Conflict of Interest: The authors declare that they have no conflict of interest.

Author Contributions: AE: Conseption, Design, Materials, Analysis and Interpretation, Data Collection and Precessing, Literature Review, Writing; DA: Supervision, Analysis and Interpretation, Literature Review, Critical Review; ASG: Supervision, Critical Review; HC: Supervision, Critical Review.

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