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Original Article

# Visual Complications of Pediatric Posterior Fossa Tumors: Analysis of Outcomes

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ABSTRACT

*Objective:* Visual abnormalities are common among children with posterior fossa tumors, resulting from disruption of ocular afferent and efferent systems due to the tumor or surgery. This study describes the visual complications and outcomes associated with these tumors.
*Methods:* Clinical and radiographic data of patients who underwent index surgery for resection of posterior fossa tumor from 2007 to 2016 were reviewed. Descriptive statistics, univariate, and multivariate regression were performed to assess factors contributing to visual acuity and postoperative strabismus.
*Results:* There were 182 patients who underwent posterior fossa craniotomy for neoplasm were included. Ophthalmologic symptoms were the fourth most common presenting complaint; initial ophthalmologic examination was abnormal in 40% of patients. Evaluation of visual acuity demonstrated a good outcome in 88% of patients following treatment. The most common postoperative oculomotor finding was esotropia (29%) which resolved spontaneously in more than half of patients. A good outcome was obtained in all patients who underwent surgery for esotropia. Hypertropia was noted in 14% of the cohort and less than half resolved spontaneously; less than half undergoing strabismus surgery for hypertropia had a good outcome. Multivariate analysis confirmed the association between cerebellar mutism and postoperative esotropia and hypertropia. Clinically significant pathological nystagmus was seen in 8% of the cohort.
*Conclusions:* Our results indicate a good visual outcome in the majority of pediatric patients undergoing resection of posterior fossa tumors. Ophthalmologic complications should be appropriately evaluated and addressed to allow for the best possible vision to survivors of posterior fossa tumors.


## Introduction

Posterior fossa tumors commonly present with ocular signs and symptoms that can compromise the ocular afferent and efferent systems. Direct compression or infiltration of the cerebrospinal fluid pathways can lead to obstructive hydrocephalus and papilledema. Papilledema is a compressive optic neuropathy in which elevated intracranial pressure results in optic nerve head edema. Optic nerve edema is associated with primary afferent or vision dysfunction, which if left untreated or undetected may progress to ganglion cell axon loss, optic nerve atrophy, and permanent vision loss, even despite treatment of hydrocephalus.

Posterior fossa tumors may also disrupt the ocular efferent system, which includes the extraocular cranial nerves and gaze holding centers. Abducens nerve palsy can occur due to injury of the abducens nerve within the subarachnoid space by hydrocephalus and the downward displacement of the brainstem as well as by direct mass effect on the pons. Abducens palsies present clinically with esotropia (inwards eye deviation) and horizontal diplopia. Trochlear nerve palsy resulting from midbrain damage leads to vertical/oblique diplopia, with resultant hypertropia (vertical eye deviation) of the involved eye and anomalous head tilt. Tumors that

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affect the midbrain, cerebellum, and brainstem may disrupt visual fixation and vestibular and/or gaze-stabilization mechanisms, which can lead to acquired nystagmus, skew deviation, and complex gaze palsies. Neurotrophic keratopathy from damage to the trigeminal nerve, or lagophthalmos from damage to the facial nerve, may result in damage to the cornea.

In children, posterior fossa tumors can result in chronic neurological impairment not only at presentation but also iatrogenically with neurosurgical intervention and management. For instance, cerebellar mutism is a postoperative syndrome characterized by diminished or absent speech, ataxia, and hypotonia resulting from resection of a midline posterior fossa tumor.[1] Pediatric posterior fossa tumors may require ophthalmological assessment and intervention; however, the visual complications and outcomes associated with the management of these tumors have received little attention. The purpose of this study is to retrospectively examine a large cohort of pediatric posterior fossa tumor patients and assess the impact on visual and oculomotor outcomes.

## Methods

Approval was obtained from the Institutional Research Board of Baylor College of Medicine for this study and a waiver of individual consent was granted. Patients who underwent index surgery (first surgical intervention for biopsy or resection of the tumor) for a posterior fossa tumor between July, 2007 and December, 2016 were identified from our patient database at Texas Children's Hospital in Houston, TX, USA. Patients were identified using Current Procedural Terminology (CPT) codes 61518, 61520, and 61521 (craniectomy for excision of brain tumor, infratentorial or posterior fossa). Patients were excluded who had previously undergone partial or total resection of the tumor at another institution. Patients were also excluded if pathology did not demonstrate a primary brain tumor. The medical records were abstracted for demographic information, pathology, radiographic presentations, symptomatology, postoperative clinical course, and long-term outcome. Information from pre- and postoperative ophthalmological examination and ophthalmologic interventions was recorded.

Tumor volume was calculated by the Formula Volume = (Length × width × height) × (Pi/6). Extent of resection was determined from postoperative magnetic resonance imaging and was classified as gross-total, near-total (greater than 95% of tumor volume), subtotal, or biopsy. The presence of hydrocephalus was determined by a radiologist based on initial brain imaging. Extent of resection was determined by the radiologist based on a postoperative magnetic resonance imaging. Presence of cerebellar mutism was defined as the absence or reduction of speech postoperatively that may occur with hypotonia, ataxia, or signs of brainstem dysfunction.

Visual acuity at last follow-up was categorized as "good" (20/20 to 20/40 in both eyes), "moderate" (20/40 to 20/200 in at least one eye), or "poor" (less than 20/200 in at least one eye) as previously described.[2] In regards to strabismus, outcome was considered "good" in cases when strabismus resolved spontaneously or when strabismus surgery resulted in realignment of eyes without diplopia; outcome was considered "poor" when diplopia and eye misalignment persisted even following strabismus surgery.

Univariate and multivariate analysis was performed to evaluate the relationship between clinical factors/demographics and visual outcome. This association was assessed using the two-sample *t* test, $\chi^2$ test, and Fisher's exact test, as well as by multivariate logistic regression analysis. Adjusted odds ratio and 95% confidence intervals were calculated. All statistical analyses were conducted using Stata 14.2. *P* value < 0.05 was considered significant.

## Results

We identified 221 patients who had undergone posterior fossa craniotomy for neoplasm. Thirty-nine were excluded who did not meet study criteria (prior tumor resection at outside hospital, pathology which did not confirm neoplasm, tumor location not in posterior fossa). Included in the study were 182 remaining patients. Baseline patient demographics and clinical factors are outlined in Table 1. The most common symptoms reported at initial presentation were nausea or vomiting (74%) followed by headache (69%), ataxia (61%), and ophthalmologic symptoms (27%). Median tumor volume was 37.6 cc (range, 1.4 cc to 197.8 cc). Imaging demonstrated hydrocephalus in 141 patients (83%).

Pathology is outlined in Table 1. Cerebellar mutism was seen postoperatively in 51 patients (28%). Fifty-five patients (30.2%) required treatment for hydrocephalus. Further tumor-directed treatment is outlined in Table 1. Median follow-up after index surgery was 4.0 years.

### *Ophthalmologic course*

Initial preoperative ophthalmologic examination was performed in all 182 patients and revealed papilledema in 53 patients (29%), strabismus in 19 (10%), and nystagmus in 14 (8%) (Table 1). Postoperative ophthalmological findings are summarized in Table 2. Of the total cohort, 88% had good visual acuity, 5% had moderate, and 5% had poor visual acuity. Underlying reasons for moderate or poor vision are outlined in Table 2. Optic atrophy due to prior hydrocephalus or other causes (e.g., chemotherapy or radiation-related) was noted at follow-up in 23 patients (13%); however, this resulted in persistent loss of visual acuity in only four patients (2%). "Moderate" or "poor" visual acuity was more likely in those requiring surgical treatment of hydrocephalus (18%) than those that did not (7%) ($P = 0.02$).

The most common postoperative oculomotor finding was esotropia (55 patients, 30%); this was associated with abduction deficit in 36 patients (20%). The deficit resolved spontaneously in 33 of 55 patients, at a median of three months postoperatively (range, one week to 18 months). Postoperative esotropia was small (0 to 20 prism diopters [PD]) in 30, moderate (21 to 40 PD) in 17, and large (more than 40 PD) in four. Univariate analysis demonstrating the risk factors for esotropia is presented in Table 3, which demonstrated that preoperative esotropia or abduction deficit correlated with esotropia postoperatively, as did the occurrence of cerebellar mutism. Multivariate analysis (Table 4) confirmed association between cerebellar mutism and postoperative esotropia.

Of the 22 patients in whom esotropia did not resolve spontaneously, symptoms were treated with observation in three patients. In the remaining 19 patients, esotropia was clinically significant and/or required treatment. A subgroup analysis was done comparing this subset of 19 patients to the total 55 patients with esotropia (Table 3). Size of esotropia was predictive requirement for further treatment: only 20% of those with small, 53% of those with moderate, and 100% of those with large esotropia required further treatment. Of the 13 patients (7%) who ultimately underwent strabismus surgery, outcome was "good" with eye realignment and resolution of diplopia postoperatively in all 13 patients.

Hypertropia was noted in 26 patients (14%); in 18 patients this was due to trochlear nerve palsy, and in eight due to skew deviation. Hypertropia was small (0 to 4 PD) in five patients, moderate (5 to 10 PD) in seven patients, and large (greater than 10 PD) in 11 patients. In 12 patients, hypertropia resolved spontaneously, at a median of two months; in two patients, hypertropia was minimally symptomatic, and was observed. Univariate analysis demonstrating the risk factors for hypertropia is presented in Table 5, which

**TABLE 1.**
Preoperative Baseline Characteristics

| | | N | % |
|---|---|---|---|
| Total # patients | | 182 | |
| Female | | 80 | 44.0 |
| Median age, years | | 5.3 | |
| Presenting symptoms | Nausea/vomiting | 134 | 73.6 |
| | Headache | 126 | 69.2 |
| | Ataxia/gait instability | 111 | 61.0 |
| | Visual symptoms | 50 | 27.5 |
| | Blurred vision | 26 | 14.3 |
| | Double vision | 15 | 8.2 |
| | Dysconjugate gaze | 11 | 6.0 |
| | Lethargy | 29 | 15.9 |
| | Weakness | 22 | 12.1 |
| Ophthalmological signs at presentation | Papilledema | 53 | 29.1 |
| | Strabismus | 19 | 10.4 |
| | Nystagmus | 14 | 7.7 |
| Symptom duration, weeks (median) | | 4 | |
| Tumor volume, cc (median) | | 37.6 | |
| Cerebral/spinal metastases | | 28 | 15.4 |
| Hydrocephalus | | 141 | 82.9 |
| Pathology | Medulloblastoma | 61 | 33.5 |
| | Juvenilepilocytic astrocytoma | 57 | 31.3 |
| | Anaplastic ependymoma WHO Grade 3 | 22 | 12.1 |
| | Atypical teraroid rhabdoid tumor | 10 | 5.5 |
| | Diffusely infiltrating brainstem tumor | 9 | 4.9 |
| | Ependymoma WHO Grade 2 | 6 | 3.3 |
| | Glioblastoma multiforme | 4 | 2.2 |
| | Exophytic brainstem astrocytoma | 3 | 1.6 |
| | Ganglioglioma | 2 | 1.1 |
| | Cerebellar astrocytoma WHO Grade 2 | 2 | 1.1 |
| | Hemangioblastoma | 2 | 1.1 |
| | Malignant neuroectodermal tumor | 2 | 1.1 |
| | Glioneuronal tumor of 4th ventricle | 1 | 0.5 |
| | Atypical choroid plexus papilloma | 1 | 0.5 |
| Extent of resection | Gross total | 136 | 74.7 |
| | Near total | 19 | 10.4 |
| | Sub total | 20 | 11.0 |
| | Biopsy | 7 | 3.8 |
| Cerebellar mutism | | 51 | 28.0 |
| Hydrocephalus treatment | Ventriculoperitoneal shunt (VPS) | 43 | 23.6 |
| | Endoscopic third ventriculostomy (ETV) | 3 | 1.6 |
| | ETV + VPS | 9 | 4.9 |
| Adjunct treatment | Chemotherapy | 68 | 37.4 |
| | Radiation | 33 | 18.1 |
| | Radiation + chemotherapy | 13 | 7.1 |

Abbreviation:
WHO = World Health Organization

demonstrated that cerebellar mutism was correlated with postoperative hypertropia.

Of the 26 patients with postoperative hypertropia, 12 had deficits that were persistent and symptomatic, requiring surgery in 10 patients. Subgroup analysis was done for these 12 patients (Table 5) which demonstrated no association between persistent hypertropia and clinical factors like size of hypertropia, tumor pathology, cerebellar mutism, or tumor size. Of the 10 patients undergoing surgery, four were orthophoric (eyes correctly aligned) postoperatively and had resolution of diplopia, and six had "poor" outcome with continued hypertropia and diplopia.

Deficits of horizontal gaze occurred postoperatively in eight patients (4%), which included internuclear ophthalmoplegia in four patients. In five patients, the gaze palsy resolved spontaneously. Three patients required strabismus surgery for resultant exotropia; outcome was "good" in all three patients.

Postoperatively 14 patients (8%) developed clinically significant abnormalities of eye movements including upbeat nystagmus (seven patients), horizontal nystagmus (three patients), Brun's nystagmus (two patients), and rotatory nystagmus (two patients).

Visual field deficit was noted in four individuals (2%) with homonymous hemianopsia. In all four patients, tumor was noted to extend superiorly up to the thalamus; three of the four had other signs of long-tract damage such as hemiplegia. Postoperative lagophthalmos due to facial nerve palsy occurred in 11 patients (5%), and unilateral or bilateral neurotrophic keratopathy due to trigeminal nerve palsy in seven patients (4%).

Preoperative and postoperative ophthalmological characteristics are outlined in Table 2.

## Discussion

Brain tumors are the second most common pediatric malignancy, occurring most commonly in the posterior fossa.[3] Maximal safe surgical resection is the mainstay of treatment, which may be followed by craniospinal irradiation and/or chemotherapy depending on tumor pathology. Due to improvement in the treatment of these tumors in the last decade, mortality following tumor treatment is decreasing, resulting in a higher number of long-term survivors. There is increased emphasis on quality of life as a gauge

**TABLE 2.**
Postoperative Ophthalmological Complications

| | N | % |
|---|---|---|
| Visual acuity | | |
| Good | 160 | 89.4 |
| Moderate | 9 | 5.0 |
| Poor | 10 | 5.6 |
| Amblyopia due to strabismus | 4 | |
| Optic atrophy | 4 | |
| Devastated neurological status | 4 | |
| Neurotrophic keratopathy | 3 | |
| Pre-existing visual impairment | 2 | |
| Cataracts | 1 | |
| Nystagmus | 1 | |
| Esotropia | 55 | 30.2 |
| Resolving spontaneously | 33 | |
| Associated with abduction deficit | 36 | |
| Requiring surgery | 13 | |
| Associated with pre-op esotropia | 12 | |
| Small (0-20 PD) | 30 | |
| Moderate (21-40 PD) | 17 | |
| Large (>40 PD) | 4 | |
| Hypertropia | 26 | 14.3 |
| Small (</=10PD) | 12 | |
| Large (>10 PD) | 11 | |
| Resolving spontaneously | 12 | |
| Requiring surgery | 10 | |
| Good outcome | 4 | |
| Poor outcome | 6 | |
| Palsy of horizontal gaze | 8 | 4.4 |
| Resolving spontaneously | 5 | |
| Clinically significant nystagmus | 14 | 7.7 |
| Brun's nystagmus | 2 | |
| Horizontal nystagmus | 3 | |
| Rotatory nystagmus | 2 | |
| Upbeat nystagmus | 7 | |
| Visual field deficit | 5 | 2.7 |
| Lagophthalmos | 11 | 6.0 |
| Neurotrophic keratopathy | 7 | 3.8 |
| Dorsal midbrain syndrome | 4 | 2.2 |
| Horner syndrome | 1 | 0.5 |
| Cranial nerve 3 palsy | 2 | 1.1 |
| Oscillopsia | 2 | 1.1 |
| Ocular tilt reaction | 2 | 1.1 |
| Radiation-induced cataracts | 3 | 1.6 |

Abbreviation:
PD = prism diopters

of successful outcome. Survivors of posterior fossa tumors are known to have deficits of visual and spatial memory,[4] decreased intelligence quotient and academic performance compared with peers, and impairments of motor function and coordination.[5] It is important to promote the best possible vision so that survivors can better cope with these sequelae.

*Clinical presentation*

Visual complaints are common in patients presenting with newly diagnosed posterior fossa tumors. In our series, visual and oculomotor disturbance was the fourth most common symptom at presentation after headache, nausea/vomiting, and gait instability. Numerous patients in this series were initially seen by an optometrist or ophthalmologist for strabismus and either prescribed glasses and/or alternate eye patching. Commonly these patients showed no improvement of their strabismus and eventually developed neurological findings leading to neuroimaging. An associated abduction deficit, progression or lack of resolution of the deficit, optic nerve findings, nystagmus, or associated neurological findings should trigger further evaluation.

*Clinical management*

At our institution, ophthalmology consultation is requested immediately after the diagnosis of posterior fossa tumor, and initial bedside ophthalmologic evaluation occurs preoperatively. Follow-up with a neuro-ophthalmologist is routinely perfromed at four to six weeks postoperatively. If strabismus is noted postoperatively, eye patching may be prescribed to treat symptomatic diplopia and to prevent amblyopia in young patients. Surgery for nonresolving, stable strabismus is only considered six months or more after an initial neurosurgical intervention or any adjunctive therapy (including chemotherapy or radiation) to allow adequate time for cranial nerve palsy recovery and re-establishment of fusion, as many deficits improve spontaneously with resolution of inflammation and swelling.

*Clinical symptomatology*

Preoperative bedside ophthalmologic assessment has limitations due to the acuity of presentation and cooperativity of the child which may particularly hamper assessment of visual acuity. Within these constraints, initial ophthalmologic examination demonstrated an abnormality in 40% of patients, most commonly papilledema. Postoperatively, 37% of patients were noted to have strabismus including abducens palsy, trochlear nerve palsy, oculomotor palsy, or comitant strabismus (eye deviation the same magnitude regardless of gaze position). Strabismus surgery was ultimately required in 14% of the total cohort.

*Visual acuity*

The causes for visual acuity deficit in patients with posterior fossa tumors are diverse. We divided patients into "good" (20/20 to 20/40 in both eyes), "moderate" (20/40 to 20/200 in at least one eye), and "poor" (less than 20/200 in at least one eye) visual acuity as previously described.[6] The majority of our cohort had good visual acuity at the last follow-up visit (88%) (median ophthalmology follow-up, 13 months). This result is similar to a recent study of pediatric posterior fossa patients in whom 72% of patients had good visual acuity at last follow-up.[6] Particularly in young patients, poor eye alignment disrupting binocular vision causes the brain to suppress visual information from the misaligned eye, often permanently. Only four patients (2%) experienced moderate to severe vision loss due to amblyopic strabismus in this series despite the high overall incidence of postoperative strabismus, indicating that prompt treatment of clinically significant eye misalignment may avoid this feared complication. Optic atrophy resulting from hydrocephalus or other causes was rarely a cause for permanent vision loss. Unsurprisingly, patients requiring surgical treatment for hydrocephalus had a poorer visual outcome, suggesting vulnerability of this patient subset even with adequate treatment of hydrocephalus. Other less common causes for impaired vision were neurotrophic keratopathy resulting in corneal scarring, cataracts secondary to radiation, and nystagmus. Our results indicate a good prognosis for visual acuity in most patients with posterior fossa tumors; close attention is required for those with strabismus or hydrocephalus to prevent permanent vision loss.

*Esotropia*

Esotropia (inward eye deviation) is the most common deficit of extraocular movements associated with posterior fossa tumors.[7] Esotropia often occurs with abducens nerve palsy, indicated by a deficit in eye abduction. While the eyes may appear aligned in primary gaze, the esotropia becomes evident on lateral gaze,

**TABLE 3.**
Univariate Analysis of Post-operative Esotropia

| | Post-Op Esotropia (%) n = 56 | No Post-Op Esotropia (%) n = 114 | | Esotropia Did Not Self-Resolve (%) n = 19 | Esotropia Self-Resolved or Asymptomatic (%) n = 36 | |
|---|---|---|---|---|---|---|
| Age | 6.1 years | 6.7 years | $P = 0.428$ | 4.8 years | 6.7 years | $P = 0.071$ |
| Male | 31.4 | 69.2 | $P = 0.842$ | 34.4 | 65.6 | $P = 0.975$ |
| Female | 30.0 | 70.0 | | 34.8 | 65.2 | |
| Pre-op Esotropia/CN6 palsy | 41.6 | 58.3 | **$P = 0.032$** | 42.9 | 57.1 | $P = 0.682$ |
| No pre-op Esotropia/CN6 palsy | 28.8 | 71.2 | | 33.3 | 66.7 | |
| Size of Esotropia | | | | | | |
| Small | | | | 20.0 | 80.0 | **$P = 0.001$** |
| Moderate | | | | 53.0 | 47.0 | |
| Large | | | | 100.0 | 0.0 | |
| Pre-op HCP | 32.6 | 67.4 | $P = 0.596$ | 35.6 | 64.4 | $P = 0.701$ |
| No pre-op HCP | 27.6 | 72.4 | | 25.0 | 75.0 | |
| Post-op HCP | 40.0 | 60.0 | $P = 0.076$ | 45.5 | 54.5 | $P = 0.165$ |
| No post-op HCP | 27.8 | 73.2 | | 27.3 | 72.7 | |
| Brainstem Involvement | 39.6 | 60.4 | $P = 0.097$ | 38.1 | 61.9 | $P = 0.663$ |
| No brainstem Involvement | 27.1 | 72.9 | | 32.4 | 67.6 | |
| Tumor type | | | | | | |
| Medulloblastoma | 34.4 | 65.6 | $P = 0.246$ | 40.0 | 60.0 | $P = 0.852$ |
| JPA | 22.8 | 77.2 | | 38.5 | 61.5 | |
| Ependymoma | 42.9 | 57.1 | | 25.0 | 75.0 | |
| Other | 27.8 | 72.2 | | 30.0 | 70.0 | |
| Tumor volume (mean) | 47.5 cc | 41.9 cc | $P = 0.846$ | 53.6 cc | 44.8 cc | $P = 0.317$ |
| Extent of resection | | | | | | |
| Gross total | 29.4 | 70.6 | $P = 0.767$ | 30.7 | 69.2 | $P = 0.400$ |
| Near total | 36.8 | 63.2 | | 57.1 | 42.9 | |
| Sub total/biopsy | 33.3 | 66.7 | | 33.3 | 66.7 | |
| Cerebellar mutism | 52.9 | 47.1 | **$P = 0.000$** | 37.0 | 63.0 | $P = 0.703$ |
| No cerebellar mutism | 22.1 | 77.9 | | 32.1 | 67.9 | |

Abbrevations:
CN = cranial nerve
HCP = hydrocephalus
JPA = juvenile pilocytic astrocytoma
Bolded values indicate statistically significant *P*-values (defined as $P < 0.05$)

known as incomitant esotropia (the size of the deviation varies with the direction of gaze).[8] Sometimes the abduction deficit resolves over time and leaves the child with residual esotropia that is present even in primary gaze (comitant strabismus: the size of the deviation does not vary with direction of gaze). Of the 56 patients in our series with esotropia, 36 had evidence of a coexisting abduction deficit; the remainder may have had subtle abducens nerve palsy which resulted in a fixed comitant esotropia or developed esotropia due to inability to recover visual fusion. Abducens nerve palsy may result from infiltration or compression of the abducens nucleus in the dorsal pons,[2] or may be related to hydrocephalus due to the sensitivity of this nerve to tractional forces caused by downward displacement of the brainstem. Interestingly, neither the presence of preoperative nor postoperative hydrocephalus correlated to the development of esotropia. In addition, evidence of brainstem involvement with tumor also did not correlate with postoperative esotropia. However, preoperative esotropia was predictive of postoperative esotropia. Cerebellar mutism was also strongly associated with postoperative esotropia.

**TABLE 4.**
Risk of Development of Postoperative Esotropia

| | OR | 95% CI | *P* Value |
|---|---|---|---|
| Pre-op CN6 palsy | 3.26 | 0.90-11.76 | 0.071 |
| No pre-op CN6 palsy | Reference group | | |
| Brainstem involvement | 1.38 | 0.66-2.87 | 0.392 |
| No brainstem involvement | Reference group | | |
| Postoperative hydrocephalus | 1.30 | 0.62-2.72 | 0.487 |
| No postoperative hydrocephalus | Reference group | | |
| Cerebellar mutism | 3.60 | 1.73-7.49 | **0.001** |
| No cerebellar mutism | Reference group | | |

Abbreviations:
CI = confidence interval
CN6 = cranial nerve 6
OR = odds ratio
Bolded value indicates statistically significant *P*-values (defined as $P < 0.05$)

Esotropia can be categorized by the number of PD of eye deviation into small (0 to 20 PD), moderate (21 to 40 PD), and large (greater than 40 PD). Small esotropia often resolves spontaneously, although in some individuals it may decompensate over time and require intervention. Most patients in our series with postoperative esotropia resolved spontaneously within a few months or had small minimally symptomatic esotropia which was observed. We did a subgroup analysis to determine whether there were predictive factors for those who had persistent, clinically significant esotropia. This analysis showed that the degree of postoperative esotropia was predictive of the clinical course of the deficit: most patients with small postoperative esotropia resolved spontaneously (80%), but only 47% of those with moderate esotropia and none with large esotropia resolved spontaneously ($P = 0.001$). Resolution of symptoms was seen in all 13 patients requiring strabismus surgery for esotropia. This suggests an excellent prognosis for patients with postoperative esotropia: most are small and resolve spontaneously within a few months, and those which are persistent and require surgery have a high likelihood of symptom resolution.

### *Hypertropia*

The second most common postoperative deficit of extraocular movements was hypertropia (vertical eye deviation) caused by trochlear nerve palsy and/or skew deviation. The trochlear nerve has a long course and is thin in diameter, rendering it susceptible to injury during surgery; it also arises from the dorsal surface of the

**TABLE 5.**
Univariate Analysis of Postoperative Hypertropia

| | Post-Op Hypertropia (%) n = 26 | No Post-Op Hypertopia (%) n = 156 | | Hypertropia Did Not Self-Resolve (%) n = 12 | Hypertropia Self-Resolved (%) n = 14 | |
|---|---|---|---|---|---|---|
| Age | 7.9 years | 6.3 years | $P = 0.114$ | 8.3 years | 7.4 years | $P = 0.563$ |
| Male | 13.7 | 86.3 | $P = 0.807$ | 50.0 | 50.0 | $P = 0.716$ |
| Female | 15.0 | 85.0 | | 42.9 | 57.1 | |
| Size of hypertropia | | | | | | |
| Small (0-4 PD) | | | | 20.0 | 80.0 | $P = 0.322$ |
| Moderate (5-10 PD) | | | | 57.1 | 42.9 | |
| Large (>10PD) | | | | 63.6 | 36.4 | |
| Midbrain compression | 20.9 | 79.1 | $P = 0.154$ | 55.6 | 44.4 | $P = 0.484$ |
| No midbrain compression | 12.2 | 87.8 | | 41.2 | 58.8 | |
| Tumor type | | | | | | |
| Medulloblastoma | 19.7 | 80.3 | $P = 0.207$ | 41.7 | 58.3 | $P = 0.453$ |
| Juvenile pilocytic astrocytoma | 7.0 | 93.0 | | 25.0 | 75.0 | |
| Ependymoma | 17.9 | 82.1 | | 80.0 | 20.0 | |
| Other | 86.1 | 13.9 | | 40.0 | 60.0 | |
| Tumor volume (mean) | 39.9 cc | 44.2 cc | $P = 0.562$ | 38.2 cc | 41.5 cc | $P = 0.771$ |
| Extent of resection | | | | | | |
| Gross total | 13.2 | 86.8 | $P = .600$ | 38.9 | 61.1 | $P = 0.595$ |
| Near total | 21.0 | 79.0 | | 75.0 | 25.0 | |
| Sub total/biopsy | 14.8 | 85.2 | | 50.0 | 50.0 | |
| Cerebellar mutism | 7.6 | 92.4 | **$P = 0.000$** | 43.8 | 56.2 | $P = 0.756$ |
| No cerebellar mutism | 31.4 | 68.6 | | 50.0 | 50.0 | |

Bolded value indicates statistically significant *P*-values (defined as $P < 0.05$)

brainstem, making it vulnerable during microsurgery in the posterior fossa. Skew deviation is a vertical misalignment of the eyes resulting from a supranuclear lesion in the brainstem, cerebellum, or vestibular system. This syndrome is a result of loss of vestibular input to the interstitial nucleus of Cajal from damage to the medulla, pons, or midbrain. Hypertropia from either trochlear nerve palsy or skew deviation can be distinguished with a thorough sensorimotor examination, noting that trochlear palsy causes a classic pattern of hypertropia of the affected eye, worsening on ipsilateral head tilt, improvement with contralateral head tilt, and excyclotorsion the affected eye. In contrast, skew deviation is a vertical misalignment with associated vestibular disturbance and central neurological dysfunction; it can present with variable strabismus patterns including hypertropia in all gazes (comitant) or hypertropia alternating between each eye relative to head position, and commonly causes incyclotorsion of the eyes.[9]

Hypertropia was noted in 14% of our cohort, but unlike esotropia, less than half of the patients resolved spontaneously. Of the 26 patients with hypertropia, 12 warranted further surgical management. A subgroup analysis was done on these 12 patients. Unlike patients with esotropia, there was no correlation with amount of hypertropia and likelihood of resolution. This finding corroborates our clinical experience that while some patients with hypertropia are minimally symptomatic and adapt to the deficit, others even with a small amount of hypertropia may have debilitating diplopia and are not able to fuse their vision. In addition, of the 10 patients electing to undergo surgical correction of symptomatic diplopia, only four patients were orthophoric (eyes correctly aligned) postoperatively; the remaining had continued symptomatic hypertropia with persistent diplopia despite multiple surgeries. These results indicate a poorer prognosis for patients with postoperative hypertropia than those with esotropia.

### *Gaze stabilization*

The cerebrum, cerebellum, midbrain, pons, and brainstem are integral components in the ocular motor coordination of horizontal and vertical gaze stabilization. Horizontal gaze palsy may result from a disruption to the paramedian pontine reticular formation, medial longitudinal fasciculus, or other gaze pathways in the brainstem and midbrain. Damage to these structures may result in unilateral or bilateral deficits in horizontal gaze. Damage to the medial longitudinal fasciculus can present as internuclear ophthalmoplegia, characterized by ipsilateral adduction palsy with a disassociated nystagmus in the contralateral abducting eye. This was a rare occurrence in our cohort and had a good outcome in all patients requiring surgery for exotropia resulting from horizontal gaze palsy.

Acquired nystagmus is a common preoperative and postoperative finding in those with pediatric posterior fossa tumors. In most, nystagmus is minimally symptomatic or asymptomatic, as in those patients with end-gaze nystagmus—a normal finding in up to 10% of the population. However, in some cases, nystagmus may be an impediment to fusion even when acuity is normal, causing the patient to require more time to process visual information. In our cohort, pathological nystagmus (vertical, horizontal, or torsional) causing impairment of visual fusion was seen in 8% of patients postoperatively. These results are concordant with a study of 72 posterior fossa tumor patients which reported an 8% rate of pathological nystagmus in a similar patient cohort; this study also reported an 11% rate of end-gaze.[10] Vertical nystagmus results from damage to supranuclear and conjugate gaze centers, with disruption to anterior (upbeat) and/or posterior (downbeat) canal projections to the brainstem tegmentum; however, these pathways are not well defined. Vertical nystagmus was noted in 5% of patients, with upbeat being the most common. Collectively, acquired nystagmus, in particular, vertical and upbeat, can be an early sign of posterior fossa disease as well as a prognostic sign for the degree of postoperative recovery.

### *Cerebellar inputs*

Cerebellar mutism is an uncommon but devastating complication of posterior fossa surgery that manifests as an absence or reduction in speech and can include axial hypotonia, ataxia, dysphagia, dysarthria, and other cranial nerve palsies. Ocular motor

deficits may also occur in the context of cerebellar mutism and severe cerebellar damage.[11,12] In this series, postoperative cerebellar mutism was strongly associated with postoperative hypertropia and esotropia, as well as eventual requirement for strabismus surgery. This association may be due to more extensive surgical damage to the posterior fossa and therefore higher likelihood of damage to ocular motor pathways. Damage to the cerebellum itself may also interfere with its interconnections with pontine nuclei and centers controlling conjugate gaze.

Damage to the cerebellum as a result of either tumor or surgery may also disrupt sensory fusion.[13] In our study, seven patients who underwent surgery for strabismus did not regain fusion and had continued diplopia. Of these, three had good alignment following multiple strabismus surgeries, but nevertheless these patients reported continued diplopia indicating lack of sensory fusion. Interestingly, all of these patients had other signs of cerebellar injury (ataxia, scanning speech) which have been persistent on follow-up. The inability to fuse despite adequate alignment may be due to cerebellar injury.[13]

### *Corneal decompensation*

Corneal damage and scarring are serious and treatable consequences of posterior fossa tumors. Neurotrophic keratopathy may result from trigeminal nerve palsy and subsequent corneal anesthesia. Facial nerve palsy resulting in incomplete eye closure, or lagophthalmos, may also predispose to corneal damage. Three patients in our cohort experienced severe deficit in visual acuity due to corneal decompensation despite normal appearance of the optic nerves. Patients should be monitored closely for signs of corneal damage due to exposure or corneal anesthesia; if treated vigilantly with aggressive lubrication, corneal scarring may be avoided.

### *Tumor pathology*

Previous studies have shown certain tumor types, specifically medulloblastoma and ependymoma, correlate with worse visual outcomes, both regarding strabismus and visual acuity. One study of 24 medulloblastoma patients reported a 50% rate of abducens nerve palsy, half of which resolved spontaneously and the remainder persisted (rate in current series: 34%).[7] Their cohort included only two patients with trochlear nerve palsy, both resolving spontaneously. Sixteen percent of patients ultimately required surgical intervention for strabismus (current series: 15%). A recent study of 139 patients with posterior fossa tumors found that patients with ependymoma and medulloblastoma had significantly poorer visual acuity outcome as compared with juvenile pilocytic astrocytoma.[6] In comparison, two studies of survivors of low-grade cerebellar gliomas demonstrated that no patients met criteria for legal blindness (less than 20/200).[14,15] One of these studies reported a rate of strabismus of 18%, though further details were not provided.[15] In our cohort, medulloblastoma and ependymoma were correlated with development of postoperative strabismus ($P = 0.039$). This is likely due to the more aggressive and locally invasive nature of these tumor pathologies; recovery of deficits may also be hampered by adjunctive radiation in these patients.

### *Limitations*

There are several limitations of this retrospective review. Due to the nature of a retrospective case series, there is no case control group, and analyses may have confounding factors. There is likely an information bias regarding the quality of outcomes recorded in postoperative clinic visits; for example, the most severely neurologically impaired individuals may not receive the extent of attention to their ophthalmological issues if their functional prognosis was poor. However, the strength of this study is that it comprehensively summarizes the features of a large pediatric cohort with potential ocular consequences, yielding information that may guide pre- and postoperative counseling of patients with posterior fossa tumors.

### *Conclusions*

Considering these findings, it is imperative to allow for the best possible vision to survivors of posterior fossa tumors, as many of the sequelae are treatable. We recommend that all patients with posterior fossa tumors be evaluated by an ophthalmologist at the time of presentation and be referred to neuro-ophthalmology for further follow-up to assess and treat for postoperative ophthalmologic complications. In particular, untreated strabismus may result in significant impairment in visuomotor skills and quality of life as well as amblyopia in young children.[16] However, timely correction of strabismus can lead to better outcomes in regards to stereopsis[16,17]; esotropia in particular has excellent outcomes following surgical correction in this series. Strabismus surgery should be delayed at least six months following neurosurgical or adjunctive treatment (chemotherapy or radiation) because many deficits resolve spontaneously within a few months.

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