

# Ophthalmologic aspects of headache in children

Alma Kurent

Community Health Centre Ljubljana, Ljubljana, Slovenia

## ABSTRACT

Headaches are common in children and adolescents and are occasionally due to eye conditions. The most common ophthalmic causes are refractive error, impairment of accommodation or convergence, ocular surface disease, acute glaucoma, optic neuritis, scleritis, uveitis, and orbital conditions.

Headache caused by ocular refractive error is generally symptomatic after prolonged visual tasks. The pain in the eye with accompanying headache, which characteristically gets worse after visual tasks such as prolonged screen use or reading, may be caused by the ocular surface disorder. Headaches that are triggered by dark light should raise suspicions of angle closure glaucoma. Scleral or uveal inflammatory diseases are usually associated with severe, aching, or throbbing ocular pain and photophobia. Examination often reveals redness of the eye. Optic neuritis in children is frequently bilateral, with optic nerve swelling and a very low visual acuity.

Orbital conditions that can cause headaches include preseptal cellulitis, which produces eyelid edema, inflammation, and erythema. Orbital cellulitis also causes restriction and pain in eye movement, exophthalmos, chemosis, reduced visual function, and a relative afferent pupillary defect. Other orbital conditions that can cause headaches are inflammatory diseases, vascular malformations, and orbital masses.

Many other headache causes can present with ocular symptoms, such as elevated intracranial pressure from an intracranial process. Ophthalmologic examination can be helpful in some of these cases. Therefore, cooperation with the neurologist and other specialists is essential.

**KEY WORDS:** eye pain; refractive error; dry eye; glaucoma; uveitis, orbit

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## INTRODUCTION

Children and adolescents with headaches are common patients in the pediatrician's office [1, 2]. The estimated prevalence of headaches over periods between 1 month and a lifetime in children and adolescents is approximately 60% [1]. Roughly 50% of children experience it at one point in their childhood. More than 2% have frequent episodic headaches [3]. Headaches increase in frequency during adolescence. Before puberty, boys are affected more frequently than girls, but after the onset of puberty, headaches occur more frequently in girls [4].

Common causes of childhood headaches are primary headache disorders, e.g. migraine or some

acute viral infection [5, 6]. Red flags requiring further investigation (for example, neuroimaging) for recognition of life-threatening conditions include an abnormal neurological examination or additional symptoms and signs of systemic disease, atypical presentation of headaches, the worst headache that a child had in the life, recent and progressive severe headache or change of headache, headache in very young children, occipital location, headache that awakes a child from sleep or headache that is present in the morning and is accompanied by vomiting or associated to straining [5, 7].

Children with a headache commonly present to a pediatric ophthalmologist because parents or pe-

## CORRESPONDING AUTHOR:

Alma Kurent MD, PhD, Community Health Centre Ljubljana, Metelkova ulica 9, 1000 Ljubljana, Slovenia; e-mail: alma.kurent@gmail.com

diatrics often suspect an underlying ophthalmologic cause of the headache. Headaches in children are occasionally due to eye conditions. The most common ophthalmic causes are refractive error, impairment of accommodation or convergence, ocular surface disease, acute glaucoma, optic neuritis, scleritis, uveitis, and orbital conditions [8–13]. Headache may also be the first sign of a severe disorder, such as elevated intracranial pressure from an intracranial process. Ophthalmologic examination may be helpful in some of these cases [8, 9, 12, 14, 15].

The aim of the paper is to review common ophthalmologic reasons for headaches in children.

## HEADACHE ATTRIBUTED TO DISORDER OF THE EYES

### Refractive error, impairment of accommodation or convergence, strabismus

Headache caused by ocular refractive error(s) is generally symptomatic after prolonged visual tasks [13]. Refractive errors are an uncommon cause of headaches, but visual acuity should be examined, and in children with any eye or visual issues, a full ophthalmologic assessment should be done [14].

In a study [14], headache improved in almost 80% of the patients, regardless of whether they had a change in refractive error. The difference between the two groups of patients with and without the change in refractive error was not statistically significant. On the other hand, in another study [16], with appropriate refractive correction, more than 72% of the patients reported improvement in the headache, and almost 40% had complete resolution.

Intraocular muscular spasms associated with accommodative effort may compensate for uncorrected refractive error. It can result in pain in the eye, headache, and occasionally a convergent form of strabismus. That is the reason why cycloplegic refraction is mandatory. After appropriate refractive error correction, improvement or even resolution of the complaints is expected [12].

Deficient convergence may be a reason for problems in prolonged visual tasks such as reading or screen use. Cover/uncover test can reveal latent or even manifest divergent strabismus. Exercise may improve the convergence deficiency [12].

According to the paper [12], strabismus, either intermittent or manifest, is usually not the reason for the headache. Due to suppression, the bin-

ocular diplopia is absent, so additional muscular effort is not needed to form one picture from images of each eye. According to the International Classification of Headache Disorders, there is little evidence for headaches attributed to heterophoria or heterotopia pending more formal study of the classification [13].

### Ocular surface disease

Chronic pain in the eye that is bilateral and characteristically gets worse after visual tasks such as screen use, doing homework, or reading may point toward a disorder of the ocular surface (dry eye syndrome). Patients usually say they feel ocular discomfort or have 'sand in the eyes'. Patients have paradoxical problems with tearing; they may be photophobic and have conjunctival hyperemia. As in blepharitis, these changes may be accompanied by secretions in the eyelashes and itchy eyelids [12]. Among other prevalent reasons for ocular pain are abrasion of the cornea, corneal foreign body or keratitis caused by infection [8].

These problems can be recognized by the ophthalmological examination. The Schirmer test, the tear film break-up time, and staining of the corneal surface are useful in diagnosing dry eye. Therapy for dry eye includes:

- artificial tears;
- medicines reducing inflammation or occlusion;
- punctal plugs that prolong ocular tear contact.

Infectious keratitis should be evaluated immediately by an ophthalmologist to start necessary antimicrobial or antifungal treatment [8].

### Acute glaucoma

Acute glaucoma is rare in children but should be considered as it can potentially be resolved. Early diagnosis and treatment may prevent long-term ocular damage that may occur because of intermittent angle occlusion. A headache that is worsened or triggered by dark light should raise the suspicion of angle closure glaucoma, as suggested in the case report of a 13-year-old girl with angle closure as a cause of intermittent headache [17].

Angle-closure glaucoma in its fulminated form presents pain in the area of the eye and orbit. Additionally, it is often accompanied by nausea and vomiting. It is characterized by red eye and cloudy cornea. The pupil is mid-dilated and fixed to light due to iris ischemia. In subacute form, the pain is transient, and blurred vision is present. Sometimes, patients report seeing halos

around lights because of the edema of the cornea [9]. The pain associated with angle closure quickly subsides once the intraocular pressure is controlled [8].

### **Uveitis and scleritis**

Scleral and uveal inflammatory diseases are typically associated with ocular pain and headache [12]. Pain is usually very severe, and patients are photophobic. Examination usually reveals redness of the eye. Iris sphincter muscle is in spasm, so the pupil can be small and poorly reactive. Various amounts of inflammatory cells or flares in the eye can be present. The number of inflammatory cells and flares depends on the severity and cause of the uveitis [9]. Pain can vary, but the uveitis can not be excluded when the pain is absent. For instance, in juvenile idiopathic uveitis, pain is not a typical characteristic despite potentially sight-threatening sequelae [8, 18].

There are differences between children and adults that should be considered during evaluation. Uveitis is less common in children than in adults [19]. In children, there is a higher risk of some ocular complications, such as uveitic glaucoma. Other complications, such as amblyopia, also occur in children. Children with uveitis may also have unique dosing requirements. Drug-associated risks, such as growth retardation with systemic corticosteroids, that must be considered may occur in children. Examination and treatment may also be more difficult and challenging with children [18].

The main complaint of patients with scleritis is pain that is reported to be dull and aching and can be severe and constant. Extraocular muscles insert into the sclera, so the pain typically worsens with eye movement. The pain can spread further to other parts, such as the periorbital region, brow, forehead, temple, ear, or jaw, and can sometimes be disproportionate to the clinical findings [20].

The etiology of the inflammation must be determined with diagnostic tests. There are several treatment options including immunosuppressive agents for noninfectious inflammation [8].

### **Optic neuritis**

Optic neuritis in children is far less frequent than in adults [21]. The usual reason for optic neuritis in children is a viral infection. Diagnostic procedures are similar to those in adults. However, what is different in children is that it usually appears bilaterally with optic nerve swelling and very low

visual acuity. Especially when the patient is younger, the prognosis is generally good. The probability of developing multiple sclerosis seems to be very limited if there are no lesions in the white matter in basal magnetic resonance imaging (MRI) [22, 23].

A typical symptom is spontaneous ocular pain triggered by eye movements. It characteristically appears a few days before visual loss in the central part. It affects visual acuity and color vision. A scotoma in the center of the visual field can be seen or generally depressed visual field [12, 24]. Although in children, the pain with eye movements is not a universal presenting symptom, levels of vision loss are variable, and visual field defects include but are not limited to, central or paracentral scotomata [25]. The pain gets worse when the eye is moving. That is believed to arise from the close attachments of the recti muscles to the optic sheath at the orbital apex [26]. Unilateral optic neuritis is suspected if a relative afferent pupillary defect (RAPD) appears. With a fundus exam, an edema of the optic disc can be revealed, or the disc can be normal if the inflammation is located more posteriorly. The pallor of the optic disc indicates chronicity of the process and appears around 4 weeks after the beginning of the symptoms [12].

In the acute phase, the treatment is with intravenous corticosteroids. Immunosuppressive or immunomodulating agents can be later applied depending on the etiology. For infective optic neuritis, specific treatment must be addressed [12]. Treatment tendencies in children consist of monitoring unless the disease is bilateral and accompanied by severe visual acuity impairment. In these cases, there is a possibility of treatment with intravenous corticosteroids [22].

### **Preseptal and orbital cellulitis**

Preseptal cellulitis and orbital cellulitis appear more often in children than in adults. Preseptal cellulitis is not truly an orbital disease. The infection is in the eyelid and soft tissue; the globe and orbit are unaffected. It is more prevalent than the orbital form and usually has a better prognosis [27]. It is very important to differentiate preseptal cellulitis from orbital cellulitis. Preseptal cellulitis involves the superficial soft tissues, and orbital cellulitis is a severe infection in the deep orbit that can be life-threatening [8]. Preseptal cellulitis presents with inflammation involving edema of the eyelids and erythema. Orbital cellulitis is characterized by exophthalmos and chemosis; also, eye movements may be

restricted. Patients report pain when moving their eyes. A reduced visual function and relative afferent pupillary defect can also be present.

Preseptal cellulitis is commonly caused by the extension of infection from the paranasal sinuses. Other very important sources of infection are ocular adnexal or soft face tissue infections. Infection may be caused by trauma, skin infections, eyelid disorders such as chalazia or hordeola, and eyelid or even oral iatrogenic procedures [27]. Contiguous sinusitis is the reason for 90% of orbital cellulitis cases. Other causes include endogenous infections, trauma, or even dental diseases [8].

Most cases of preseptal cellulitis are treated with oral antibiotics, and most cases of orbital cellulitis are managed with intravenous antibiotics. Some abscesses, particularly large ones, require surgical therapy. It was reported that patients under 9 years respond well to medical management more frequently than older patients. However, recent studies suggest that even older children with small or moderate-sized abscesses and normal vision may still respond to medical therapy before proceeding with surgical intervention [28].

### Orbital inflammation

Orbital inflammatory disorders such as idiopathic orbital inflammation or thyroid-related orbitopathy are rare in young children, and they are more common in adolescents [8, 29]. Idiopathic orbital inflammatory disease (OID) presents with characteristics that can be acute, chronic, or even recurrent. Diffuse inflammatory disease is characterized by exophthalmos, hyperemia of the conjunctival tissue, and external swelling. On the other hand, if extraocular muscles are specifically involved, the presenting sign may be limited eye movements and diplopia [8]. Intense eye pain may worsen if the eye globe is retropulsed or with eye movement. Orbital pain is variable and occurs in at least half of the affected individuals. Depending on the involved orbital structures, it is usually associated with other signs and symptoms [30, 31]. Headache or intense pain in the eye may appear even if the external signs of inflammation are not present. OID must be differentiated from other orbital disorders with similar findings, such as orbital cellulitis caused by infection, tumors, and vascular disorders. Inflammation in the posterior sclera is also a form of OID that ultrasonography can visualize. Ocular and orbital assessments are required for the diagnosis. The diagnostic procedure includes targeted laboratory stud-

ies and radiologic imaging. A biopsy may be necessary to rule out neoplasia, as malignancies may show a similar clinical picture. Treatment includes systemic corticosteroids and other immunosuppressant agents [8].

Orbital myositis usually starts suddenly. Eye symptoms include periorbital edema, chemosis, ptosis, diplopia, pain increasing with eye movements, and restriction of eye movements. The diagnosis is based on the orbital imaging. It is most commonly present in middle-aged patients but has also been reported in pediatric patients. Pediatric cases of orbital myositis can develop in association with systemic conditions, e.g., streptococcal pharyngitis [32, 33].

Trochleitis is an inflammation of the superior oblique tendon where it passes through the trochlea. It usually causes swelling of the medial canthus or in the area below the medial brow. Tenderness worsens when the patient tries to look up in adduction [8]. Patients present with dull to severe pain in the superonasal region that worsens with eye movement [10]. The disease is mostly primary or can appear in rheumatoid arthritis, sarcoidosis, and other inflammatory diseases. The options for the treatment of tracheitis include injection of corticosteroids into the trochlear area [11].

### Other orbital conditions

Other orbital conditions that can cause headaches are also vascular malformations and orbital masses. Orbital masses that are rapidly expanding are often painful. The clinical picture often includes proptosis of the eye, edema, reduced visual acuity, and raised intraocular pressure. Some of the vascular disorders in the orbit are capillary hemangiomas, malformations of the venous vascular system, arterial and arteriovenous malformations (AVMs), and venous lymphatic disorders [8, 34].

In children with new onset orbital space-occupying lesions, we should consider rhabdomyosarcoma, which is the most common primary orbital malignancy, and neuroblastoma, which is a common metastasis to the orbit. In children, capillary hemangioma and lymphangioma (lymphatic malformation) are relatively common vascular orbital lesions [35]. Capillary hemangioma is the most common orbital vascular tumor in infants, with an overall incidence of 5.6% [34, 36]. Lesions usually appear at or shortly after birth, rapidly increase in size for 6–12 months, and then gradually involute over the next 5–7 years [34].

## OPHTHALMIC SYMPTOMS AND SIGNS OF OTHER CAUSES OF HEADACHE

Many other causes of headaches also include ocular signs and symptoms. Double vision with periocular pain or headache can result from several reasons, such as lesions in the orbita, cranial neuropathies, and lesions occurring in the cavernous sinus. Anisocoria or other abnormalities in pupillary responses, like in Horner's syndrome, may also be caused by numerous disorders that include headaches. They include aneurysms, carotid dissection, cluster headaches, and others [9]. The most common neoplasm presenting with Horner syndrome is neuroblastoma, and, indeed, an isolated Horner syndrome is the first presenting symptom of neuroblastoma in 2% of cases. However, often, no etiology can be found, and in such instances, the Horner syndrome is labeled idiopathic [37]. Adie's syndrome or ophthalmoplegic migraine may present with painful and dilated pupils that are unreactive.

On the other hand, it can also mean the presence of a life-threatening condition, such as a posterior communicating artery aneurysm. Headache, in combination with transient visual acuity loss, may be caused by common conditions such as classic migraine. But it can also be a symptom of more severe disorders such as ocular hypoperfusion from carotid occlusion or due to idiopathic intracranial hypertension (IIH) [9].

Transient visual disturbances, tinnitus, and double vision due to cranial nerve dysfunction are classic symptoms of IIH. The most common complaints in young children are headaches; they can be irritable, show apathy, or somnolence. They can report dizziness, stiff neck, strabismus, or ataxia. In the pediatric population, it is more common to reveal an underlying disorder that is associated with IIH than in the adult population. Therefore, when IIH is suspected in a child, a detailed exam and evaluation of opening pressure is essential. In adolescence, more girls than boys are affected by the disease. Obesity should also be addressed as it is associated with IIH. On the other hand, in younger children, IIH does not appear as often as in older children, and girls are similarly affected as boys. Also, obesity is not as strongly associated with IIH in younger children as it is in older children [6].

Headache may also be the first sign of a severe disorder, such as elevated intracranial pressure from an intracranial process or hydrocephalus. Ophthalmologic examination can be helpful in some of these cases. However, not all children with

elevated intracranial pressure develop papilledema. Anatomic studies suggest that the subarachnoid space surrounding the optic nerves is a compartment different from the rest of the intracranial subarachnoid space. These separate compartments may be why not all children with elevated intracranial pressure develop papilledema and also why papilledema may persist even in a functional ventriculoperitoneal shunt [25].

Ophthalmic signs such as photophobia or lacrimation occur also in primary non-ophthalmic conditions. Migraine is the most common neurologic disorder causing photophobia. Up to 80% of migraine patients experience photophobia during an attack (38). A typical visual aura in migraine is usually reported as scotomata, blurry or reduced visual acuity that is transient, zig-zag lines, or even more complex visual phenomena [6]. The most common symptom associated with cluster headache is lacrimation (reported by > 90% of patients), but other common ophthalmological symptoms include conjunctival injection, ptosis, eyelid swelling, and light sensitivity [39, 40].

## CONCLUSION

Children with headaches often come for evaluation by an ophthalmologist. That is usually because pediatricians or parents suspect an ophthalmologic disorder as a reason for a headache. The most common ophthalmic causes are refractive error, impairment of accommodation or convergence, ocular surface disease, acute glaucoma, optic neuritis, scleritis, uveitis, and orbital conditions. Signs and symptoms in the ocular region can also appear with several other headache causes, so cooperation with the neurologist and other specialists is essential.

### Conflict of interests

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