

**American College of Radiology  
ACR Appropriateness Criteria®  
Orbital Imaging and Vision Loss-Child**

**Variant: 1 Child. Traumatic visual loss. Suspected orbital injury. Initial imaging.**

Procedure	Appropriateness Category	Peds Relative Radiation Level
CT orbits without IV contrast	Usually Appropriate	⦿⦿⦿
MRI head without IV contrast	May Be Appropriate	○
MRI orbits without IV contrast	May Be Appropriate	○
CT head without IV contrast	May Be Appropriate	⦿⦿⦿
Radiography orbit	Usually Not Appropriate	⦿
MRA head without and with IV contrast	Usually Not Appropriate	○
MRA head without IV contrast	Usually Not Appropriate	○
MRI head without and with IV contrast	Usually Not Appropriate	○
MRI orbits without and with IV contrast	Usually Not Appropriate	○
CT head with IV contrast	Usually Not Appropriate	⦿⦿⦿
CT head without and with IV contrast	Usually Not Appropriate	⦿⦿⦿⦿
CT orbits with IV contrast	Usually Not Appropriate	⦿⦿⦿
CT orbits without and with IV contrast	Usually Not Appropriate	⦿⦿⦿
CTA head with IV contrast	Usually Not Appropriate	⦿⦿⦿⦿

**Variant: 2 Child. Nontraumatic acute vision loss without papilledema. Initial imaging.**

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRI head and orbits without and with IV contrast	Usually Appropriate	○
MRI complete spine without and with IV contrast	May Be Appropriate	○
MRI head and orbits without IV contrast	May Be Appropriate	○
MRI head without and with IV contrast	May Be Appropriate	○
MRI head without IV contrast	May Be Appropriate	○
MRI orbits without and with IV contrast	May Be Appropriate	○
CT head without IV contrast	May Be Appropriate	⦿⦿⦿
MRA head without and with IV contrast	Usually Not Appropriate	○
MRA head without IV contrast	Usually Not Appropriate	○
MRI complete spine without IV contrast	Usually Not Appropriate	○
MRI orbits without IV contrast	Usually Not Appropriate	○
CT head and orbits with IV contrast	Usually Not Appropriate	⦿⦿⦿
CT head and orbits without and with IV contrast	Usually Not Appropriate	⦿⦿⦿⦿
CT head and orbits without IV contrast	Usually Not Appropriate	⦿⦿⦿
CT head with IV contrast	Usually Not Appropriate	⦿⦿⦿
CT head without and with IV contrast	Usually Not Appropriate	⦿⦿⦿⦿
CT orbits with IV contrast	Usually Not Appropriate	⦿⦿⦿
CT orbits without and with IV contrast	Usually Not Appropriate	⦿⦿⦿
CT orbits without IV contrast	Usually Not Appropriate	⦿⦿⦿
CTA head and neck with IV contrast	Usually Not Appropriate	⦿⦿⦿⦿

CTA head with IV contrast	Usually Not Appropriate	☢☢☢☢
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### **Variant: 3 Child with isolated nystagmus. Initial imaging.**

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	○
MRI head and orbits without and with IV contrast	May Be Appropriate	○
MRI head and orbits without IV contrast	May Be Appropriate	○
MRI head without IV contrast	May Be Appropriate	○
MRI orbits without and with IV contrast	May Be Appropriate	○
MRI orbits without IV contrast	May Be Appropriate	○
MRA head without and with IV contrast	Usually Not Appropriate	○
MRA head without IV contrast	Usually Not Appropriate	○
CT head and orbits with IV contrast	Usually Not Appropriate	☢☢☢
CT head and orbits without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT head and orbits without IV contrast	Usually Not Appropriate	☢☢☢
CT head with IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT head without IV contrast	Usually Not Appropriate	☢☢☢
CT orbits with IV contrast	Usually Not Appropriate	☢☢☢
CT orbits without and with IV contrast	Usually Not Appropriate	☢☢☢
CT orbits without IV contrast	Usually Not Appropriate	☢☢☢
CTA head with IV contrast	Usually Not Appropriate	☢☢☢☢

### **Variant: 4 Child. Congenital or developmental abnormality leading to decreased visual acuity or vision loss. No leukocoria. Unilateral or bilateral. Initial imaging.**

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRI head and orbits without IV contrast	Usually Appropriate	○
MRI head and orbits without and with IV contrast	May Be Appropriate (Disagreement)	○
MRI head without IV contrast	May Be Appropriate	○
MRI orbits without IV contrast	May Be Appropriate	○
MRA head without and with IV contrast	Usually Not Appropriate	○
MRA head without IV contrast	Usually Not Appropriate	○
MRI head without and with IV contrast	Usually Not Appropriate	○
MRI orbits without and with IV contrast	Usually Not Appropriate	○
CT head and orbits with IV contrast	Usually Not Appropriate	☢☢☢
CT head and orbits without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT head and orbits without IV contrast	Usually Not Appropriate	☢☢☢
CT head with IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT head without IV contrast	Usually Not Appropriate	☢☢☢
CT orbits with IV contrast	Usually Not Appropriate	☢☢☢
CT orbits without and with IV contrast	Usually Not Appropriate	☢☢☢
CT orbits without IV contrast	Usually Not Appropriate	☢☢☢
CTA head with IV contrast	Usually Not Appropriate	☢☢☢☢

**Variant: 5 Child. Vision loss and suspected optic pathway tumor, with or without a history of neurofibromatosis type 1. Initial imaging.**

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRI head and orbits without and with IV contrast	Usually Appropriate	0
MRI head without and with IV contrast	May Be Appropriate	0
MRI orbits without and with IV contrast	May Be Appropriate (Disagreement)	0
Radiography orbit	Usually Not Appropriate	⚠
MRA head without and with IV contrast	Usually Not Appropriate	0
MRA head without IV contrast	Usually Not Appropriate	0
MRI head and orbits without IV contrast	Usually Not Appropriate	0
MRI head without IV contrast	Usually Not Appropriate	0
MRI orbits without IV contrast	Usually Not Appropriate	0
CT head and orbits with IV contrast	Usually Not Appropriate	⚠⚠⚠
CT head and orbits without and with IV contrast	Usually Not Appropriate	⚠⚠⚠⚠
CT head and orbits without IV contrast	Usually Not Appropriate	⚠⚠⚠
CT head with IV contrast	Usually Not Appropriate	⚠⚠⚠
CT head without and with IV contrast	Usually Not Appropriate	⚠⚠⚠⚠
CT head without IV contrast	Usually Not Appropriate	⚠⚠⚠
CT orbits with IV contrast	Usually Not Appropriate	⚠⚠⚠
CT orbits without and with IV contrast	Usually Not Appropriate	⚠⚠⚠
CT orbits without IV contrast	Usually Not Appropriate	⚠⚠⚠
CTA head with IV contrast	Usually Not Appropriate	⚠⚠⚠⚠

**Variant: 6 Child. Six months of age or older. Papilledema detected on the ophthalmologic examination or signs of raised intracranial pressure. Initial imaging.**

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRI head and orbits without and with IV contrast	Usually Appropriate	0
MRI head and orbits without IV contrast	Usually Appropriate	0
MRI head without and with IV contrast	Usually Appropriate	0
MRI head without IV contrast	May Be Appropriate	0
MRV head with IV contrast	May Be Appropriate	0
MRV head without and with IV contrast	May Be Appropriate	0
MRV head without IV contrast	May Be Appropriate	0
CT head without IV contrast	May Be Appropriate	⚠⚠⚠
CTV head with IV contrast	May Be Appropriate	⚠⚠⚠⚠
MRA head without and with IV contrast	Usually Not Appropriate	0
MRA head without IV contrast	Usually Not Appropriate	0
MRI orbits without and with IV contrast	Usually Not Appropriate	0
MRI orbits without IV contrast	Usually Not Appropriate	0
CT head and orbits with IV contrast	Usually Not Appropriate	⚠⚠⚠
CT head and orbits without and with IV contrast	Usually Not Appropriate	⚠⚠⚠⚠
CT head and orbits without IV contrast	Usually Not Appropriate	⚠⚠⚠
CT head with IV contrast	Usually Not Appropriate	⚠⚠⚠

CT head without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT orbits with IV contrast	Usually Not Appropriate	☢☢☢
CT orbits without and with IV contrast	Usually Not Appropriate	☢☢☢
CT orbits without IV contrast	Usually Not Appropriate	☢☢☢
CTA head with IV contrast	Usually Not Appropriate	☢☢☢☢☢

### **Variant: 7 Child. Suspected orbital or periorbital infection. Initial imaging.**

Procedure	Appropriateness Category	Peds Relative Radiation Level
CT orbits with IV contrast	Usually Appropriate	☢☢☢
MRI head without and with IV contrast	May Be Appropriate	○
MRI orbits without and with IV contrast	May Be Appropriate	○
CT head with IV contrast	May Be Appropriate	☢☢☢
MRA head without and with IV contrast	Usually Not Appropriate	○
MRA head without IV contrast	Usually Not Appropriate	○
MRI head without IV contrast	Usually Not Appropriate	○
MRI orbits without IV contrast	Usually Not Appropriate	○
MRV head with IV contrast	Usually Not Appropriate	○
MRV head without and with IV contrast	Usually Not Appropriate	○
MRV head without IV contrast	Usually Not Appropriate	○
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT head without IV contrast	Usually Not Appropriate	☢☢☢
CT orbits without and with IV contrast	Usually Not Appropriate	☢☢☢
CT orbits without IV contrast	Usually Not Appropriate	☢☢☢
CTA head with IV contrast	Usually Not Appropriate	☢☢☢☢
CTV head with IV contrast	Usually Not Appropriate	☢☢☢☢

### **Variant: 8 Child. Leukocoria or suspected intraocular mass. Initial imaging.**

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRI head and orbits without and with IV contrast	Usually Appropriate	○
MRI orbits without and with IV contrast	Usually Appropriate	○
MRI head without and with IV contrast	May Be Appropriate	○
MRI orbits without IV contrast	May Be Appropriate	○
CT head and orbits with IV contrast	May Be Appropriate	☢☢☢
CT head with IV contrast	May Be Appropriate	☢☢☢
CT orbits with IV contrast	May Be Appropriate	☢☢☢
CT orbits without IV contrast	May Be Appropriate	☢☢☢
MRA head without and with IV contrast	Usually Not Appropriate	○
MRA head without IV contrast	Usually Not Appropriate	○
MRI head and orbits without IV contrast	Usually Not Appropriate	○
MRI head without IV contrast	Usually Not Appropriate	○
CT head and orbits without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT head and orbits without IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT head without IV contrast	Usually Not Appropriate	☢☢☢

CT orbits without and with IV contrast	Usually Not Appropriate	☢☢☢
CTA head with IV contrast	Usually Not Appropriate	☢☢☢☢

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## Summary of Literature Review

### Introduction/Background

Orbital disorders in children consist of varied pathologies affecting orbits, orbital contents, visual pathway, and innervation of the extraocular or intraocular muscles. Clinical manifestations of these disorders may range from ophthalmoplegia, decreased visual acuity, diplopia, nystagmus, exophthalmos/enophthalmos, papilledema, orbital soft tissue swelling, orbital pain, and, in most severe cases, monocular or binocular vision loss. The underlying etiology of these disorders may be traumatic or nontraumatic.

Detailed clinical history along with a thorough clinical examination, including ophthalmologic and neurologic examination, is frequently able to localize the abnormality along the visual pathway. Presumed location of the lesion along with the additional findings like eye pain, swelling, exophthalmos/enophthalmos, erythema, conjunctival vascular dilatation, intraocular pressure, and so on, help in determining if imaging is needed, modality of choice, and extent of coverage (orbits and /or head) [1]. Occasionally clinical signs and symptoms may be nonspecific, and, in these cases, diagnostic imaging studies play key role in depicting the nature and extent of the injury or disease [2].

Localization of the orbital abnormality on imaging is facilitated by a compartmental approach, which helps in the differential diagnosis based on the lesion location within the orbit (globe, extraconal, or intraconal space) or elsewhere in the visual pathway [3].

Traumatic injury in the setting of head injury or nonaccidental injury is discussed separately in ACR Appropriateness Criteria on these topics and is therefore excluded from this review.

### Special Imaging Considerations

Ultrasound (US) is also an important diagnostic tool but is most often performed by the ophthalmologist or emergency physician and therefore not covered in this article.

### Initial Imaging Definition

Initial imaging is defined as imaging at the beginning of the care episode for the medical condition defined by the variant. More than one procedure can be considered usually appropriate in the initial imaging evaluation when:

- There are procedures that are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care)

OR

- There are complementary procedures (ie, more than one procedure is ordered as a set or simultaneously wherein each procedure provides unique clinical information to effectively manage the patient's care).

## **Discussion of Procedures by Variant**

### **Variant 1: Child. Traumatic visual loss. Suspected orbital injury. Initial imaging.**

Traumatic injury is the leading cause of noncongenital monocular vision loss in children [4]. Common signs of orbital injuries include periorbital soft tissue swelling, ecchymosis, hyphema, subconjunctival hemorrhage, diplopia, restriction of ocular movement, or vision loss. The most common causes of pediatric orbital fractures are motor vehicle collision, sports injury, falls, and assault [5]. Data from combined pediatric and adult series show that ocular injuries account for approximately 3% of all visits to the emergency department [6]. Orbital fractures account for 3% to 45% of all pediatric facial fractures [5]. Orbital roof fractures are the most common orbital fractures in children <8 years of age, and orbital floor fractures are most common in older children [7]. Serious ocular injuries in conjunction with orbital fracture are less common in children compared with adults. In one series, serious ocular injury was reported in approximately 20% of children with orbital fractures [7].

Traumatic brain injuries are more commonly associated with orbital fractures in younger children because orbital roof fractures are more common in this age group [7]. Elastic, cancellous bone with resilient periosteum in children leads to trap door orbital fracture, which in turn causes entrapment of the extraocular muscles or other orbital contents [8], leading to restriction of ocular movement and diplopia. Incarceration of the inferior rectus muscle within a trap door fracture may rapidly result in ischemic injury of the muscle and subsequent restrictive strabismus. However, diplopia itself does not imply extraocular muscle entrapment as soft tissue swelling, hematoma, or nerve palsy can also lead to restricted ocular movement [9]. The presence of visual loss or visual field defects suggests direct or indirect injury to the globe or optic nerve.

Open globe injury may be a result of blunt trauma (globe rupture) or penetrating injury (globe laceration). Penetrating injuries may also be associated with intraorbital foreign body. Imaging may be conducted to assess for the intraorbital foreign body, extent of globe damage, and injury to the surrounding structures [10,11].

### **Variant 1: Child. Traumatic visual loss. Suspected orbital injury. Initial imaging.**

#### **A. CT head**

CT of the head may have a complementary role when obtained along with CT orbits in cases in which intracranial abnormality or calvarial fracture is suspected, particularly in children with suspected orbital roof fracture. Outside the setting of concurrent head trauma, there is no relevant literature to support the role for CT head in the initial evaluation of the orbital trauma.

### **Variant 1: Child. Traumatic visual loss. Suspected orbital injury. Initial imaging.**

#### **B. CT Orbits**

CT is considered the most useful imaging modality in evaluating orbital trauma, and it is the most accurate method in detecting fractures [12]. CT may also provide information for the detection of soft tissue injuries (including globe or extraocular muscles), hemorrhage, and presence of an intraocular foreign body. CT was found to be 94.9% sensitive for detection of the intraorbital foreign body, especially metallic or glass foreign bodies [13]. There is no relevant literature to support the role for contrast-enhanced CT orbits in the initial evaluation of the orbital trauma.

**Variant 1: Child. Traumatic visual loss. Suspected orbital injury. Initial imaging.**

**C. CT head without IV contrast**

There is no relevant literature to support the use of CT angiography (CTA) head in the initial evaluation of orbital trauma.

**Variant 1: Child. Traumatic visual loss. Suspected orbital injury. Initial imaging.**

**D. MRA Head**

There is no relevant literature to support the use of MR angiography (MRA) head in the initial evaluation of orbital trauma.

**Variant 1: Child. Traumatic visual loss. Suspected orbital injury. Initial imaging.**

**E. MRI Head**

MRI of the head provides more detailed information about brain parenchyma and other intracranial structures due to its superior soft tissue contrast (versus CT) but remains supplementary to a CT as an initial imaging modality in the setting of orbital trauma. There is no relevant literature to support the role of contrast-enhanced MRI head in the initial evaluation of the orbital trauma.

**Variant 1: Child. Traumatic visual loss. Suspected orbital injury. Initial imaging.**

**F. MRI Orbits**

MRI of the orbits may provide useful detailed information due to its superior soft tissue contrast, particularly in the evaluation of the globe and optic nerves. MRI may also be useful in detecting nonmetallic intraorbital foreign body, especially a wooden foreign body, which may appear hypoattenuating on CT and may be mistaken for air [12]. However, it remains supplementary to a CT scan as an initial imaging modality in orbital trauma. There is no relevant literature to support the role of contrast-enhanced MRI orbits in the initial evaluation of orbital trauma.

**Variant 1: Child. Traumatic visual loss. Suspected orbital injury. Initial imaging.**

**G. Radiography Orbit**

Radiography is insufficient in the detection of the pathology in cases of orbital trauma and has been replaced by CT in current clinical practice [12]. Radiography of the orbits may have a role as a screening modality for the detection of a metallic intraorbital foreign body before performing an MRI scan. However, a CT scan of the orbits is a more sensitive modality for detection of metallic intraocular foreign body and is usually performed prior to MRI in cases of orbital trauma [12,13].

**Variant 2: Child. Nontraumatic acute vision loss without papilledema. Initial imaging.**

Causes of acute nontraumatic vision loss in children may be due to ocular abnormalities or lesions in the neurovisual pathway. Ocular abnormalities involving the cornea, lens, or anterior chamber are best diagnosed by an ophthalmologic examination. Various abnormalities of the neurovisual pathway (including optic nerve, optic chiasm, optic tracts, thalamus, optic radiations, or visual cortex) that may present with acute visual loss or defect are covered in this variant. These may include optic neuritis, which may be secondary to demyelinating disorders (multiple sclerosis,

neuromyelitis optica spectrum disorders), other inflammatory, infectious, or granulomatous disorders, or unknown underlying cause (idiopathic). This variant also includes lesions involving the brain parenchyma in the region of thalamus, optic radiations, or occipital lobes, which may have acute presentation (eg, infarct, hemorrhage, inflammatory, infectious, or demyelination processes). Ocular migraine is a common cause of transient acute monocular vision loss with ipsilateral headache in adolescents. It is diagnosed based on the clinical history and examination. Imaging is performed to exclude other causes of vision loss and headache [14].

Lastly, nonorganic (psychogenic) acute vision loss is not uncommon in school age children. These are diagnosed by exclusion, based on the clinical history, inconsistent clinical examination, and negative imaging studies [15,16].

**Variant 2: Child. Nontraumatic acute vision loss without papilledema. Initial imaging.**

**A. CT head and orbits**

There is no relevant literature to support the use of CT head and orbits in the initial evaluation of children with nontraumatic vision loss.

**Variant 2: Child. Nontraumatic acute vision loss without papilledema. Initial imaging.**

**B. CT head and orbits without and with IV contrast**

CT of the head without intravenous (IV) contrast may be complementary to MRI head for a suspected brain lesion, especially if infarct or hemorrhage in the neurovisual pathway is suspected. There is no relevant literature to support the role for contrast-enhanced CT head in the initial evaluation of children with nontraumatic vision loss.

**Variant 2: Child. Nontraumatic acute vision loss without papilledema. Initial imaging.**

**C. CT Orbits**

There is no relevant literature to support the use of CT orbits in the initial evaluation of children with nontraumatic vision loss.

**Variant 2: Child. Nontraumatic acute vision loss without papilledema. Initial imaging.**

**D. CTA Head and Neck**

There is no relevant literature to support the use of CTA head and neck in the initial evaluation of children with nontraumatic vision loss.

**Variant 2: Child. Nontraumatic acute vision loss without papilledema. Initial imaging.**

**E. CT head without and with IV contrast**

There is no relevant literature to support the use of CTA head in the initial evaluation of children with nontraumatic vision loss.

**Variant 2: Child. Nontraumatic acute vision loss without papilledema. Initial imaging.**

**F. MRA Head**

There is no relevant literature to support the use of MRA head in the initial evaluation of children with nontraumatic vision loss.

**Variant 2: Child. Nontraumatic acute vision loss without papilledema. Initial imaging.**

**G. MRI Head**

MRI of the brain without IV contrast may also be complementary to CT scan for confirmation of acute infarct or intracranial hemorrhage. MRI head with and without IV contrast is obtained to evaluate the brain for intracranial demyelinating lesions, location, and distribution, which helps

determine appropriate differential diagnosis [17,18].

## **Variant 2: Child. Nontraumatic acute vision loss without papilledema. Initial imaging.**

### **H. MRI Head and Orbits**

MRI of the head and orbits with and without IV contrast is the most useful imaging modality for the evaluation of acute nontraumatic vision loss. T1-weighted postcontrast images with fat suppression were reported to identify abnormal enhancement of the optic nerve in 95% of cases of optic neuritis [19]. MRI is also the most useful modality for the evaluation of the lesions involving extraorbital neurovisual pathway and the remainder of the brain parenchyma.

## **Variant 2: Child. Nontraumatic acute vision loss without papilledema. Initial imaging.**

### **I. MRI Orbits**

MRI of the orbits with and without IV contrast is useful for evaluation of globes and optic nerves in cases of acute nontraumatic vision loss. T1-weighted postcontrast images with fat suppression were reported to identify abnormal enhancement of the optic nerve in 95% of cases of optic neuritis [19]. In the setting of acute vision loss, MRI of the orbits alone is inadequate, and it is usually performed along with an MRI of the head because the pathologies leading to visual loss frequently involve extraorbital neurovisual pathway and other locations within the brain parenchyma.

## **Variant 2: Child. Nontraumatic acute vision loss without papilledema. Initial imaging.**

### **J. MRI Complete Spine**

MRI of the spine may help assess the disease burden in the demyelinating disorders and may help differentiate different acquired demyelinating disorders (neuromyelitis optica spectrum disorders versus multiple sclerosis). MRI of the spine as initial imaging may be a useful adjunct to MRI of the brain and orbits but should be used based on the signs and symptoms of the patient. Longitudinally extensive transverse myelitis as seen on MRI spine may be considered a characteristic feature of neuromyelitis optica spectrum disorders, acute demyelinating encephalomyelitis, or anti-myelin oligodendrocyte glycoprotein immunoglobulin antibody disorder. However, it is a much less specific finding in children compared with adults [20]. IV contrast administration is helpful in detecting active demyelinating lesions because enhancing lesions represent inflammatory infiltrates causing leakage of contrast across the blood-brain barrier. Contrast enhancement is typically observable in the first 4 to 6 weeks of lesion formation [21,22].

## **Variant 3: Child with isolated nystagmus. Initial imaging.**

Pediatric nystagmus can be classified as infantile (onset in first 6 months of life) or acquired (later onset) [23]. The most common causes of infantile nystagmus are albinism, retinal disease, low vision, or vision deprivation (congenital cataract) and other retinal disorders [23,24]. These are typically diagnosed with a clinical ophthalmological examination and genetic workup. Infantile nystagmus may also occur in fusion maldevelopment syndrome, which occurs in children with normal ocular development and retinal function.

Acquired nystagmus may be caused by anterior optic pathway lesions (tumors), lesions of the brainstem/cerebellum (structural lesions or space occupying lesions), or various metabolic diseases (leukodystrophies, mitochondrial diseases, etc) [23,25]. Neuroimaging is frequently needed in these cases to exclude above structural lesions, especially in patients with late onset nystagmus, in the presence of concurrent neurological symptoms, with decreased visual acuity, or in the presence of

asymmetric/unilateral or progressive nystagmus [26-28].

Batmanabane et al [29] retrospectively reviewed charts of 148 children who underwent MRI for isolated nystagmus; 23 (15.5%) of these children had abnormal intracranial findings. Most common abnormalities on MRI included abnormal T2 hyperintense signal in white matter (4%), Chiari 1 malformation (3.4%), and optic pathway glioma (2%). This study did not find an association between the time of the onset of nystagmus and abnormal MRI. This study also suggested that administration of IV contrast is not required in all cases with isolated nystagmus and can be considered in children with a suspicious lesion on MRI. Similarly, only 2% of subjects in this study had intraorbital abnormalities that benefitted from dedicated orbital sequences. MRI of the orbits may be considered if the initial screening MRI brain is suspicious for orbital abnormalities [29].

Vestibular nystagmus is also a common form of acquired nystagmus. It may result from the dysfunction of the peripheral (labyrinth, vestibular nerve) or central vestibular pathways (root entry zone of the VIII cranial nerve, brain stem vestibular nuclei to ocular nuclei). The role of imaging is to primarily exclude VIII cranial nerve or brainstem lesion [30].

Spasmus nutans is a rare form of nystagmus that is characterized by a triad of nystagmus, head bobbing, and torticollis. It usually appears at 1 to 3 years of age and abates by 5 to 12 years of age. It usually cannot be easily differentiated from the nystagmus associated with retinal disorders or other lesions caused by anterior visual pathway tumors. Therefore, thorough neuro-ophthalmological and neuroradiological workup with MRI is necessary in these cases [23].

### **Variant 3: Child with isolated nystagmus. Initial imaging.**

#### **A. CT head and orbits**

There is no relevant literature to support the use of CT head and orbits in the initial evaluation of children with isolated nystagmus.

### **Variant 3: Child with isolated nystagmus. Initial imaging.**

#### **B. CT head**

There is no relevant literature to support the use of CT head in the initial evaluation of children with isolated nystagmus.

### **Variant 3: Child with isolated nystagmus. Initial imaging.**

#### **C. CT orbits**

There is no relevant literature to support the use of CT orbits in the initial evaluation of children with isolated nystagmus.

### **Variant 3: Child with isolated nystagmus. Initial imaging.**

#### **D. CTA Head**

There is no relevant literature to support the use of CTA head in the initial evaluation of children with isolated nystagmus.

### **Variant 3: Child with isolated nystagmus. Initial imaging.**

#### **E. MRA Head**

There is no relevant literature to support the use of MRA head in the initial evaluation of children with isolated nystagmus.

### **Variant 3: Child with isolated nystagmus. Initial imaging.**

#### **F. MRI Head and Orbits**

MRI head and orbits without or with IV contrast may be helpful as initial imaging in children with isolated nystagmus [27]. Batmanabane et al [29] retrospectively reviewed charts of 148 children who underwent MRI for isolated nystagmus; 23 (15.5%) of these children had abnormal intracranial findings. Most common abnormalities on MRI included abnormal T2 hyperintense signal in white matter (4%), Chiari 1 malformation (3.4%), and optic pathway glioma (2%).

### **Variant 3: Child with isolated nystagmus. Initial imaging.**

#### **G. MRI Head**

MRI of the head without and with IV contrast may be helpful to evaluate for intracranial abnormalities in children presenting with isolated nystagmus [29]. Batmanabane et al [29] retrospectively reviewed charts of 148 children who underwent MRI for isolated nystagmus; 23 (15.5%) of these children had abnormal intracranial findings. Most common abnormalities on MRI included abnormal T2 hyperintense signal in white matter (4%), Chiari 1 malformation (3.4%), and optic pathway glioma (2%).

### **Variant 3: Child with isolated nystagmus. Initial imaging.**

#### **H. MRI Orbits**

There is no relevant literature to support MRI orbits without or with IV contrast as initial imaging in children with isolated nystagmus. However, it may be considered an adjunct to the MRI of the brain if obtained concurrently. Batmanabane et al [29] retrospectively reviewed charts of 148 children who underwent MRI for isolated nystagmus; 23 (15.5%) of these children had abnormal intracranial findings. Most common abnormalities on MRI included abnormal T2 hyperintense signal in white matter (4%), Chiari 1 malformation (3.4%), and optic pathway glioma (2%).

### **Variant 4: Child. Congenital or developmental abnormality leading to decreased visual acuity or vision loss. No leukocoria. Unilateral or bilateral. Initial imaging.**

Various developmental abnormalities that typically present with decreased acuity or loss of vision primarily include abnormalities of the globes and optic nerves. Abnormalities of the globe may include anophthalmos, microphthalmos, macrophthalmos, anterior segment dysgenesis, or coloboma [2]. These disorders are best evaluated with clinical examination, ophthalmoscopy, and measurement of the size of the globe with ocular US. However, imaging may be helpful in complex abnormalities, which are difficult to delineate by US or for evaluation of the associated syndromes and developmental abnormalities in the brain (eg, coloboma on the setting of Aicardi syndrome) [2].

Optic nerve hypoplasia (ONH) is the most common congenital optic disc anomaly. ONH is the leading cause of visual impairment in children  $\leq 3$  years of age, and ONH is frequently associated with a variety of central nervous system and endocrine abnormalities (absence of the septum pellucidum, corpus callosal hypoplasia, migration anomalies, schizencephaly, gray matter heterotopias, and pituitary abnormalities) [31,32]. Septo-optic dysplasia is characterized by diagnostic triad of ONH, pituitary abnormalities, and midline brain abnormalities (eg, agenesis of the septum pellucidum or callosal hypoplasia). Malformation of the cortical development, particularly schizencephaly, is also a common association of septo-optic dysplasia [31,33].

Children with bilateral ONH usually present in infancy with poor vision and nystagmus, whereas unilateral or asymmetric ONH may be detected later due to strabismus. Indirect ophthalmoscopy is usually sufficient to diagnose severe ONH, but in mild to moderate cases, diagnosis is more challenging. Although imaging is usually obtained for known or suspected ONH to evaluate a child

for associated central nervous system abnormalities, studies have also investigated the usefulness of MRI as a diagnostic modality for ONH [34].

**Variant 4: Child. Congenital or developmental abnormality leading to decreased visual acuity or vision loss. No leukocoria. Unilateral or bilateral. Initial imaging.**

**A. CT head and orbits**

There is no relevant literature to support the use of CT head and orbits in the initial evaluation of children without leukocoria with decreased visual acuity or vision loss due to congenital or developmental abnormality.

**Variant 4: Child. Congenital or developmental abnormality leading to decreased visual acuity or vision loss. No leukocoria. Unilateral or bilateral. Initial imaging.**

**B. CT head**

There is no relevant literature to support the use of CT head in the initial evaluation of children without leukocoria with decreased visual acuity or vision loss due to congenital or developmental abnormality.

**Variant 4: Child. Congenital or developmental abnormality leading to decreased visual acuity or vision loss. No leukocoria. Unilateral or bilateral. Initial imaging.**

**C. CT orbits**

There is no relevant literature to support the use of CT orbits in the initial evaluation of children without leukocoria with decreased visual acuity or vision loss due to congenital or developmental abnormality.

**Variant 4: Child. Congenital or developmental abnormality leading to decreased visual acuity or vision loss. No leukocoria. Unilateral or bilateral. Initial imaging.**

**D. CTA Head**

There is no relevant literature to support the use of CTA head in the initial evaluation of children without leukocoria with decreased visual acuity or vision loss due to congenital or developmental abnormality.

**Variant 4: Child. Congenital or developmental abnormality leading to decreased visual acuity or vision loss. No leukocoria. Unilateral or bilateral. Initial imaging.**

**E. MRA Head**

There is no relevant literature to support the use of MRA head in the initial evaluation of children without leukocoria with decreased visual acuity or vision loss due to congenital or developmental abnormality.

**Variant 4: Child. Congenital or developmental abnormality leading to decreased visual acuity or vision loss. No leukocoria. Unilateral or bilateral. Initial imaging.**

**F. MRI Head and Orbits**

MRI of the head and orbits without IV contrast is the most useful modality for evaluation of the developmental abnormalities of the globes, optic nerves, and associated abnormalities in the brain and pituitary gland [2,32-34]. The sensitivity and specificity of MRI for the detection of pituitary abnormality in the patients with ONH and endocrinopathy are 68% to 96% and 83% to 92%, respectively [32,33]. MRI may also complement fundoscopic examination in the primary diagnosis of ONH by direct measurement of the optic nerve size [34]. There is no relevant literature to support the role for IV contrast in the initial evaluation of children without leukocoria with decreased visual acuity or vision loss due to congenital or developmental abnormality.

**Variant 4: Child. Congenital or developmental abnormality leading to decreased visual acuity or vision loss. No leukocoria. Unilateral or bilateral. Initial imaging.**

#### **G. MRI Head**

There is no relevant literature to support the use of MRI head alone in the initial evaluation of children without leukocoria with decreased visual acuity or vision loss due to congenital or developmental abnormality. MRI of the head is, however, often obtained in conjunction with MRI orbits.

**Variant 4: Child. Congenital or developmental abnormality leading to decreased visual acuity or vision loss. No leukocoria. Unilateral or bilateral. Initial imaging.**

#### **H. MRI Orbits**

MRI of the orbits alone is useful to evaluate for the abnormalities of the globe and orbits, but this is best performed in association with MRI of the brain, to assess the associated developmental abnormalities of the intracranial structures [2,32-34].

**Variant 5: Child. Vision loss and suspected optic pathway tumor, with or without a history of neurofibromatosis type 1. Initial imaging.**

Optic pathway gliomas (OPGs) are the most common tumor of the anterior visual pathway. They comprise 1% of all intracranial tumors and 3% to 5% of all pediatric brain tumors. OPGs in childhood are most commonly pilocytic astrocytoma [35,36]. A large majority of patients diagnosed with OPG have neurofibromatosis type 1 (NF-1). The incidence of NF-1 in patients with OPG varies from 10% to 70% [35,36]. Up to 15% to 20% of patients with NF-1 will have optic nerve glioma. The presence of bilateral OPG is almost pathognomonic for NF-1. OPGs associated with NF-1 are more often low grade tumors, may be multifocal and bilateral, and are usually found within the optic nerve. At least 50% of patients with NF-1-associated OPG have no vision loss. Conversely, sporadic OPG has a more aggressive course than NF-1-associated OPG, with a greater propensity to present symptomatically with a worse visual outcome. [36].

**Variant 5: Child. Vision loss and suspected optic pathway tumor, with or without a history of neurofibromatosis type 1. Initial imaging.**

#### **A. CT head and orbits**

There is no relevant literature to support the use of CT head and orbits in the initial evaluation of children with suspected OPG.

**Variant 5: Child. Vision loss and suspected optic pathway tumor, with or without a history of neurofibromatosis type 1. Initial imaging.**

#### **B. CT head**

There is no relevant literature to support the use of CT head in the initial evaluation of children with suspected OPG.

**Variant 5: Child. Vision loss and suspected optic pathway tumor, with or without a history of neurofibromatosis type 1. Initial imaging.**

#### **C. CT orbits**

There is no relevant literature to support the use of CT head in the initial evaluation of children with suspected OPG.

**Variant 5: Child. Vision loss and suspected optic pathway tumor, with or without a history of neurofibromatosis type 1. Initial imaging.**

#### **D. CTA Head**

There is no relevant literature to support the use of CTA head in the initial evaluation of children with suspected OPG.

**Variant 5: Child. Vision loss and suspected optic pathway tumor, with or without a history of neurofibromatosis type 1. Initial imaging.**

**E. MRA Head**

There is no relevant literature to support the use of MRA head in the initial evaluation of children with suspected OPG.

**Variant 5: Child. Vision loss and suspected optic pathway tumor, with or without a history of neurofibromatosis type 1. Initial imaging.**

**F. MRI Head and Orbits**

MRI of the head and orbit with and without IV contrast is the most useful imaging modality in diagnosis and evaluation of the extent of the symptomatic OPG in patients with or without NF-1 [36,37]. The role of MRI in the early detection of OPG in asymptomatic children with NF-1 is controversial, because there is no evidence it could improve the clinical outcome of patients in reducing the incidence of visual loss [38,39]. The role of IV contrast in the surveillance and follow-up of the optic pathway glioma is not entirely clear, because tumor volume variation is sufficient in most cases to assess tumor progression [38,39].

**Variant 5: Child. Vision loss and suspected optic pathway tumor, with or without a history of neurofibromatosis type 1. Initial imaging.**

**G. MRI Head**

There is no relevant literature to support the use of MRI head alone in the initial evaluation of children with suspected OPG. MRI of the head is, however, often obtained in conjunction with MRI orbits.

**Variant 5: Child. Vision loss and suspected optic pathway tumor, with or without a history of neurofibromatosis type 1. Initial imaging.**

**H. MRI Orbits**

Based on expert consensus, MRI orbits may be considered in cases of isolated OPG confined to the optic nerve and optic chiasm without NF-1.

**Variant 5: Child. Vision loss and suspected optic pathway tumor, with or without a history of neurofibromatosis type 1. Initial imaging.**

**I. Radiography Orbit**

There is no relevant literature to support the use of radiography orbit in the initial evaluation of children with suspected OPG.

**Variant 6: Child. Six months of age or older. Papilledema detected on the ophthalmologic examination or signs of raised intracranial pressure. Initial imaging.**

This variant includes children (older infants or older children) presenting with signs and symptoms of raised intracranial hypertension (papilledema, headache, nausea, vomiting, or transient obscuration of vision) [40]. Increased intracranial pressure may be caused by intracranial structural lesions, like space occupying lesion or hydrocephalus. Idiopathic intracranial hypertension (IIH), or pseudotumor cerebri, is a syndrome defined by elevated intracranial pressure without evidence of a structural lesion or hydrocephalus on neuroimaging and a normal cerebrospinal fluid composition [41]. In postpubertal children, IIH is typically seen in overweight girls; however, in prepubertal children, boys and girls are equally affected [41]. Several secondary causes of

pseudotumor cerebri have been described, including cerebral venous anomalies, intracranial arteriovenous fistulas, medications (tetracyclines, Vitamin A, retinoids, steroids, growth hormone, thyroxine, lithium, etc), and endocrine disorders (Addison disease, hypoparathyroidism, etc) [42]. Diagnostic criteria for pseudotumor cerebri syndrome include papilledema, normal neurological examination (except sixth nerve palsy), normal brain parenchyma on imaging (with absence of mass, hydrocephalus, or abnormal meningeal enhancement), normal cerebrospinal fluid composition, and elevated lumbar puncture opening pressure >280 mm of cerebrospinal fluid in children (or >250 mm cerebrospinal fluid if the child is not sedated and not obese) [42]. In the absence of papilledema and sixth nerve palsy, diagnosis of pseudotumor cerebri can be suggested on neuroimaging based on findings including empty sella, flattening of the posterior aspect of the globes, distention of the perioptic subarachnoid space, and transverse sinus stenosis [42]. This variant includes older infants or older children therefore head ultrasonography is not an optimal imaging option.

**Variant 6: Child. Six months of age or older. Papilledema detected on the ophthalmologic examination or signs of raised intracranial pressure. Initial imaging.**

**A. CT head and orbits**

There is no relevant literature to support the use of CT head and orbits in the initial evaluation of children with papilledema or suspected raised intracranial pressure.

**Variant 6: Child. Six months of age or older. Papilledema detected on the ophthalmologic examination or signs of raised intracranial pressure. Initial imaging.**

**B. CT head**

CT of the head without IV contrast may be a useful and alternative modality to MRI head for the evaluation of intracranial space occupying lesion or hydrocephalus causing raised intracranial pressure and papilledema. There is no relevant literature to support the use of contrast-enhanced CT head in the initial evaluation of children with papilledema or suspected raised intracranial pressure.

**Variant 6: Child. Six months of age or older. Papilledema detected on the ophthalmologic examination or signs of raised intracranial pressure. Initial imaging.**

**C. CT orbits**

There is no relevant literature to support the use of CT orbits in the initial evaluation of children with papilledema or suspected raised intracranial pressure.

**Variant 6: Child. Six months of age or older. Papilledema detected on the ophthalmologic examination or signs of raised intracranial pressure. Initial imaging.**

**D. CTA Head**

There is no relevant literature to support the use of CTA head in the initial evaluation of children with papilledema or suspected raised intracranial pressure.

**Variant 6: Child. Six months of age or older. Papilledema detected on the ophthalmologic examination or signs of raised intracranial pressure. Initial imaging.**

**E. CTV Head**

CT venography (CTV) of the head may be an alternative to MR venography (MRV) for the evaluation of cerebral venous sinuses [42].

**Variant 6: Child. Six months of age or older. Papilledema detected on the ophthalmologic examination or signs of raised intracranial pressure. Initial imaging.**

**F. MRA Head**

There is no relevant literature to support the use of MRA head in the initial evaluation of children with papilledema or suspected raised intracranial pressure.

**Variant 6: Child. Six months of age or older. Papilledema detected on the ophthalmologic examination or signs of raised intracranial pressure. Initial imaging.**

**G. MRI Head and Orbits**

MRI of the head and orbits is the most useful imaging modality for initial evaluation of the child presenting with papilledema and signs of raised intracranial pressure by detection of intracranial space occupying lesion or hydrocephalus. MRI is more useful over CT in children because of the ability of MRI to provide a higher resolution of intracranial and intraorbital structures [42,43]. Various neuroimaging findings in diagnosis of pediatric IIH are best assessed with MRI of the head and orbits. These include posterior globe flattening (56% sensitivity and 100% specificity), intraocular protrusion of the optic nerve (40% sensitivity and 100% specificity), and horizontal tortuosity of the optic nerve (68% sensitivity and 83% specificity). In the patients with IIH, optic nerve sheath was enlarged compared with those in the control group (mean 4.3 versus 3.2 mm), and pituitary gland size is found to be smaller in the patients with IIH compared with those in the control group (mean 3.63 versus 5.05 mm) [43]. Administration of IV contrast can help in characterization of the intracranial space occupying lesion (when detected).

**Variant 6: Child. Six months of age or older. Papilledema detected on the ophthalmologic examination or signs of raised intracranial pressure. Initial imaging.**

**H. MRI Head**

MRI of the head may be useful for initial evaluation of the child presenting with papilledema and signs of raised intracranial pressure by detecting an intracranial space occupying lesion or hydrocephalus. MRI is more useful than CT because of its ability to provide a higher soft tissue contrast resolution of intracranial structures [42,43]. MRI can also aid in the diagnosis of IIH by demonstrating a finding of partially empty sella. However, other imaging findings of IIH are better evaluated on MRI of the orbits. Administration of IV contrast can help in characterization of the intracranial space occupying lesion (when detected).

**Variant 6: Child. Six months of age or older. Papilledema detected on the ophthalmologic examination or signs of raised intracranial pressure. Initial imaging.**

**I. MRI Orbits**

There is no relevant literature to support the use of MRI orbits alone in the initial evaluation of children with papilledema or suspected raised intracranial pressure.

**Variant 6: Child. Six months of age or older. Papilledema detected on the ophthalmologic examination or signs of raised intracranial pressure. Initial imaging.**

**J. MRV Head**

MRV of the head may be useful in demonstrating narrowing of the distal transverse sinuses, which is supportive of the diagnosis of IIH. It may also be helpful in excluding cerebral venous sinus thrombosis, which may be a cause of secondary pseudotumor cerebri, especially in nonobese prepubertal children and individuals at high risk of cerebral venous sinus thrombosis [42,43].

**Variant 7: Child. Suspected orbital or periorbital infection. Initial imaging.**

This variant consists of children presenting with signs of orbital infection. These include preseptal cellulitis (when infection is confined to eyelids and soft tissues anterior to the orbital septum), postseptal cellulitis, and orbital abscess. Preseptal cellulitis is usually caused by percutaneous introduction of the infectious pathogen or secondary to sinusitis or odontogenic in origin.

Postseptal cellulitis is usually secondary to sinusitis (particularly ethmoid sinusitis) [44,45]. Infection of the postseptal space may have various catastrophic complications including raised orbital pressure, retinal artery/superior ophthalmic vein occlusion, optic nerve injury (leading to vision loss), cavernous sinus thrombosis, or empyema [46]. Clinical findings alone may not be specific enough to distinguish preseptal from orbital infections or those with complications. Proptosis and limitation of the extraocular movements are indicators of postseptal inflammation, but these are not very accurate and fail to differentiate postseptal inflammation from abscess [44]. Other risk factors for postseptal inflammation are high neutrophil count, the absence of infectious conjunctivitis, gross periorbital edema, age >3 years, and previous antibiotic therapy [44]. The primary role of imaging is to differentiate preseptal cellulitis from postseptal cellulitis and abscess. It also helps in identifying underlying sinusitis and intracranial complications of orbital infections.

**Variant 7: Child. Suspected orbital or periorbital infection. Initial imaging.**

**A. CT head with IV contrast**

There is no relevant literature to support the use of CT head as the initial evaluation of children with suspected periorbital or orbital infection. CT head with IV contrast may be considered in cases in which intracranial complications of the orbital cellulitis (like subdural empyema or cavernous sinus thrombosis) are suspected [46]. Precontrast imaging is typically not necessary in evaluating these patients because they do not add significant diagnostic information in this scenario.

**Variant 7: Child. Suspected orbital or periorbital infection. Initial imaging.**

**B. CT Orbits**

CT of the orbits with IV contrast is considered the most useful imaging in cases of suspected orbital infection [46]. It aids in differentiation of preseptal cellulitis from postseptal cellulitis and abscess. It is also useful in detection of complications like superior ophthalmic vein/cavernous sinus thrombosis or subdural empyema [46]. Precontrast imaging is typically not necessary in evaluating these patients because they do not add significant diagnostic information in this scenario.

**Variant 7: Child. Suspected orbital or periorbital infection. Initial imaging.**

**C. CTA Head**

There is no relevant literature to support the use of CTA head in the initial evaluation of children with suspected periorbital or orbital infection.

**Variant 7: Child. Suspected orbital or periorbital infection. Initial imaging.**

**D. CTV Head**

There is no relevant literature to support the use of CTV head in the initial evaluation of children with suspected periorbital or orbital infection. It may be useful in evaluation of cases in which cavernous sinus thrombosis is suspected as a complication of orbital cellulitis.

**Variant 7: Child. Suspected orbital or periorbital infection. Initial imaging.**

**E. MRA Head**

There is no relevant literature to support the use of MRA head in the initial evaluation of children with suspected periorbital or orbital infection.

**Variant 7: Child. Suspected orbital or periorbital infection. Initial imaging.**

**F. MRI Head**

MRI of the head with and without IV contrast can be complementary to CT scan and may be considered if a more detailed assessment of intraorbital spread of infection is clinically warranted.

It may be especially considered for cases in which there is clinical or CT-based suspicion for intracranial complications [46].

**Variante 7: Child. Suspected orbital or periorbital infection. Initial imaging.**

**G. MRI Orbits**

MRI of the orbits and head with and without IV contrast can be complementary to CT scan and may be considered if a more detailed assessment of intraorbital spread of infection is clinically warranted. It may be especially considered for cases in which there is clinical or CT-based suspicion for intracranial complications [46].

**Variante 7: Child. Suspected orbital or periorbital infection. Initial imaging.**

**H. MRV Head**

There is no relevant literature to support the use of MRV head in the initial evaluation of children with suspected periorbital or orbital infection. Suspected superior orbital vein or cavernous sinus thrombosis (as a complication of orbital cellulitis) may be better evaluated with MRI of brain and orbits with IV contrast rather than MRV.

**Variante 8: Child. Leukocoria or suspected intraocular mass. Initial imaging.**

Leukocoria is defined as abnormal white reflection from the retina of the eye (compared with normal red reflection) and can be related to abnormalities of the lens, vitreous, or retina. In children, the common causes include retinoblastoma, persistent hyperplastic primary vitreous (PHPV), retinopathy of prematurity, Coats disease, congenital cataract, and larval granulomatosis. Diagnosis of most of these conditions is based on the clinical history, fundoscopic examination, and ocular US performed by the ophthalmologist and may not require additional imaging.

**Variante 8: Child. Leukocoria or suspected intraocular mass. Initial imaging.**

**A. CT head and orbits with IV contrast**

CT head and orbits with IV contrast may be helpful in differentiating various causes of leukocoria and also for the evaluation of the extension of retinoblastoma along optic nerves and intracranially.

**Variante 8: Child. Leukocoria or suspected intraocular mass. Initial imaging.**

**B. CT head**

CT head with IV contrast may be helpful as an adjunct to the orbital imaging for the evaluation of intracranial spread of retinoblastoma.

**Variante 8: Child. Leukocoria or suspected intraocular mass. Initial imaging.**

**C. CT Orbits**

CT of the orbits with IV contrast may be helpful in differentiating various causes of leukocoria and also for the evaluation of the extension of the retinoblastoma along optic nerves.

**Variante 8: Child. Leukocoria or suspected intraocular mass. Initial imaging.**

**D. CTA Head**

There is no relevant literature to support the use of CTA head in the initial evaluation of children with leukocoria or suspected intraocular or orbital mass.

**Variante 8: Child. Leukocoria or suspected intraocular mass. Initial imaging.**

**E. MRA Head**

There is no relevant literature to support the use of MRA head in the initial evaluation of children with leukocoria or suspected intraocular or orbital mass.

## **Variant 8: Child. Leukocoria or suspected intraocular mass. Initial imaging.**

### **F. MRI Head and Orbits**

MRI of the head and orbits with and without IV contrast is the most useful imaging modality in the evaluation of malignant intraocular masses (retinoblastoma) [47-50]. MRI is helpful in evaluation of the intraocular mass/lesion when ocular media is unclear, blocking the clinician's view of the optic disc. In addition, MRI can show retrolaminar optic nerve and choroid-scleral infiltrations, orbital invasion, concurrent intracranial tumor (in the sellar or pineal region), and possible intracranial spread of the tumor [48].

Retinopathy of prematurity and PHPV are associated with microphthalmia compared to normal sized globe in Coats disease or retinoblastoma. Absence of calcification on CT scan is important to differentiate PHPV and Coats disease from retinoblastoma. Postcontrast enhancement of the intraocular mass on MRI also helps to differentiate retinoblastoma from Coats disease, PHPV, or retinopathy of prematurity [2].

## **Variant 8: Child. Leukocoria or suspected intraocular mass. Initial imaging.**

### **G. MRI Head**

MRI head with and without IV contrast may be useful as an adjunct to the orbital imaging for the evaluation of the intracranial extension of retinoblastoma. In situations in which a malignant intraocular or intraorbital mass (retinoblastoma) is suspected, this is best performed in association with the MRI of the orbits [48].

## **Variant 8: Child. Leukocoria or suspected intraocular mass. Initial imaging.**

### **H. MRI Orbits**

MRI of the orbits with and without IV contrast is useful in the initial evaluation of the benign intraorbital masses or other causes of leukocoria [2]. In situations in which a malignant intraocular or intraorbital mass (retinoblastoma) is suspected, this is best performed in association with the MRI of the brain to assess for possible intracranial spread of the tumor [48].

## **Summary of Highlights**

- **Variant 1:** In the setting of traumatic visual loss with suspected orbital injury in a child, CT of the orbits without IV contrast is usually appropriate as initial imaging. CT of the head without IV contrast may have a complementary role in cases in which orbital roof injury or concurrent head trauma is suspected.
- **Variant 2:** In the setting of nontraumatic acute vision loss without papilledema in a child, MRI of the head and orbits with and without IV contrast is usually appropriate as initial imaging.
- **Variant 3:** In the setting of isolated nystagmus in a child when no cause could be determined by clinical ophthalmological examination, MRI of the head with and without IV contrast is usually appropriate as initial imaging. MRI of the orbits with and without IV contrast may be appropriate as an adjunct, if obtained along with MRI of the head.
- **Variant 4:** In the setting of congenital or developmental abnormality leading to decreased visual acuity or vision loss without leukocoria in a child, MRI of the head and orbits without IV contrast is usually appropriate as initial imaging.
- **Variant 5:** In the setting of vision loss and suspected optic pathway tumor with or without the history of NF-1 in a child, MRI of the head and orbits with and without IV contrast is

usually appropriate as initial imaging. MRI orbits with and without IV contrast may be appropriate in cases (without NF-1) in which tumor is confined to optic nerves and optic chiasm.

- **Variante 6:** In the setting of papilledema or signs or raised intracranial pressure in a child >6 months of age, MRI of the head and orbits with and without IV contrast is usually appropriate as initial imaging. Alternatively, MRI of the head and orbits without IV contrast is usually appropriate as initial imaging if intracranial space occupying lesion is not detected. Similarly, MRI of the head with and without IV contrast is usually appropriate as initial imaging if an intracranial space occupying lesion is detected as the cause of raised intracranial pressure. MRV may be complementary to MRI of the head (and orbits) if venous sinus thrombosis or IIH is suspected.
- **Variante 7:** In the setting of suspected orbital or periorbital infection in a child, CT of the orbits with IV contrast is usually appropriate as initial imaging. MRI of the orbits and head with and without IV contrast may be appropriate in the cases in which detailed assessment of the infection spread is required or intracranial extension of the infection is suspected, respectively.
- **Variante 8:** In the setting of leukocoria or suspected intraocular mass in a child, MRI of the head and orbits with and without IV contrast is usually appropriate as initial imaging. Alternatively, MRI of the orbits with and without IV contrast is usually appropriate in cases in which benign intraocular mass or nontumoral causes of leukocoria are suspected.

## Supporting Documents

The evidence table, literature search, and appendix for this topic are available at <https://acsearch.acr.org/list>. The appendix includes the strength of evidence assessment and the final rating round tabulations for each recommendation.

For additional information on the Appropriateness Criteria methodology and other supporting documents, please go to the ACR website at <https://www.acr.org/Clinical-Resources/Clinical-Tools-and-Reference/Appropriateness-Criteria>.

## Appropriateness Category Names and Definitions
















Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients.
May Be Appropriate	4, 5, or 6	The imaging procedure or treatment may be indicated in the specified clinical scenarios as an alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal.
May Be Appropriate (Disagreement)	5	The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel's recommendation. "May be appropriate" is the rating category and a

		rating of 5 is assigned.
Usually Not Appropriate	1, 2, or 3	The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable.

## Relative Radiation Level Information

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, because of both organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared with those specified for adults (see Table below). Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria® [Radiation Dose Assessment Introduction](#) document.

## Relative Radiation Level Designations

Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range
0	0 mSv	0 mSv
	<0.1 mSv	<0.03 mSv
 	0.1-1 mSv	0.03-0.3 mSv
  	1-10 mSv	0.3-3 mSv
   	10-30 mSv	3-10 mSv
    	30-100 mSv	10-30 mSv

\*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (e.g., region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as “Varies.”

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## Disclaimer

The ACR Committee on Appropriateness Criteria and its expert panels have developed criteria for determining appropriate imaging examinations for diagnosis and treatment of specified medical condition(s). These criteria are intended to guide radiologists, radiation oncologists and referring physicians in making decisions regarding radiologic imaging and treatment. Generally, the complexity and severity of a patient's clinical condition should dictate the selection of appropriate imaging procedures or treatments. Only those examinations generally used for evaluation of the patient's condition are ranked. Other imaging studies necessary to evaluate other co-existent diseases or other medical consequences of this condition are not considered in this document. The availability of equipment or personnel may influence the selection of appropriate imaging procedures or treatments. Imaging techniques classified as investigational by the FDA have not been considered in developing these criteria; however, study of new equipment and applications should be encouraged. The ultimate decision regarding the appropriateness of any specific radiologic examination or treatment must be made by the referring physician and radiologist in light of all the circumstances presented in an individual examination.

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