

**American College of Radiology  
ACR Appropriateness Criteria®**

**Clinical Condition:**                      **Orbits, Vision and Visual Loss**

**Variant 1:**                                      **Infant or child with orbital asymmetry, proptosis and visual loss.**

Radiologic Procedure	Rating	Comments	RRL*
MRI head and/or orbit without and with contrast	8		None
MRI head and/or orbit without contrast	7		None
CT head and/or orbit without and with contrast	6		Low
CT head and/or orbit without contrast	5		Low
MRA head and neck	4	If vascular disease suspected.	None
CTA head and neck	2	If vascular disease suspected.	Low
X-ray orbit	1		Min
<b><u>Rating Scale:</u> 1=Least appropriate, 9=Most appropriate</b>			<b>*Relative Radiation Level</b>

**Variant 2:**                                      **Child with slowly progressive visual loss.**

Radiologic Procedure	Rating	Comments	RRL*
MRI head and/or orbit without and with contrast	8		None
MRI head and/or orbit without contrast	7		None
CT head and/or orbit without and with contrast	6		Low
CT head and/or orbit without contrast	5		Low
MRA head and neck	4	If vascular disease suspected.	None
CTA head and neck	2	If vascular disease suspected.	Low
X-ray orbit	1		Min
<b><u>Rating Scale:</u> 1=Least appropriate, 9=Most appropriate</b>			<b>*Relative Radiation Level</b>

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**Clinical Condition:****Orbits, Vision and Visual Loss****Variant 3:****Adult with sudden onset of painless or painful visual loss.**

<b>Radiologic Procedure</b>	<b>Rating</b>	<b>Comments</b>	<b>RRL*</b>
MRI head and/or orbit without and with contrast	8		None
MRI head and/or orbit without contrast	7		None
CT head and/or orbit without and with contrast	6		Low
MRA head and neck	5	If vascular disease suspected.	None
CT head and/or orbit without contrast	5		Low
CTA head and neck	5	If vascular disease suspected.	Low
X-ray orbit	1		Min
<b>Rating Scale:</b> 1=Least appropriate, 9=Most appropriate			<b>*Relative Radiation Level</b>

**Variant 4:****Adult patient with proptosis and/or painful visual loss.**

<b>Radiologic Procedure</b>	<b>Rating</b>	<b>Comments</b>	<b>RRL*</b>
MRI head and/or orbit without and with contrast	8		None
MRI head and/or orbit without contrast	7		None
CT head and/or orbit without and with contrast	6		Low
CT head and/or orbit without contrast	5		Low
MRA head and neck	4	If vascular disease suspected.	None
CTA head and neck	4	If vascular disease suspected.	Low
X-ray orbit	1		Min
<b>Rating Scale:</b> 1=Least appropriate, 9=Most appropriate			<b>*Relative Radiation Level</b>

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**Clinical Condition:****Orbits, Vision and Visual Loss****Variant 5:****Adult patient with uveitis, scleritis, and visual loss.**

<b>Radiologic Procedure</b>	<b>Rating</b>	<b>Comments</b>	<b>RRL*</b>
MRI head and/or orbit without and with contrast	8		None
MRI head and/or orbit without contrast	7		None
CT head and/or orbit without and with contrast	5		Low
CTA head and neck	4	If vascular disease suspected.	Low
MRA head and neck	4	If vascular disease suspected.	None
CT head and/or orbit without contrast	4		Low
X-ray orbit	1		Min
<b>Rating Scale:</b> 1=Least appropriate, 9=Most appropriate			<b>*Relative Radiation Level</b>

**Variant 6:****Adult patient with ophthalmoplegia.**

<b>Radiologic Procedure</b>	<b>Rating</b>	<b>Comments</b>	<b>RRL*</b>
MRI head and/or orbit without and with contrast	9		None
MRI head and/or orbit without contrast	6		None
CT head and/or orbit without and with contrast	6		Low
MRA head and neck	6	If vascular disease suspected.	None
CTA head and neck	6	If vascular disease suspected.	Low
CT head and/or orbit without contrast	5		Low
X-ray orbit	1		Min
<b>Rating Scale:</b> 1=Least appropriate, 9=Most appropriate			<b>*Relative Radiation Level</b>

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**Clinical Condition:****Orbits, Vision and Visual Loss****Variant 7:****Head injury with visual loss.**

<b>Radiologic Procedure</b>	<b>Rating</b>	<b>Comments</b>	<b>RRL*</b>
CT head and/or orbit without contrast	9		Low
MRI head and/or orbit without contrast	7	If MRI safe.	None
MRI head and/or orbit without and with contrast	5	If MRI safe.	None
CTA head and neck	4	If vascular disease suspected.	Low
MRA head and neck	4	If vascular disease suspected.	None
CT head and/or orbit without and with contrast	3		Low
X-ray orbit	2		Min
<b>Rating Scale: 1=Least appropriate, 9=Most appropriate</b>			<b>*Relative Radiation Level</b>

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## ORBITS, VISION AND VISUAL LOSS

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### **Summary of Literature Review**

#### **Orbital Disorders**

Primary diseases of the orbit may present with proptosis, visual disturbances, and/or ophthalmoplegia. These signs and symptoms may occur alone or in combination, and may be accompanied by pain and/or vascular engorgement on the visible surface of the globe.

Proptosis is an abnormal protrusion of the globe from the orbit and may be differentiated from exophthalmos, which is an abnormal prominence of the globe. Clinically, it may not be possible to differentiate these two entities, but once imaging is performed, the differentiation is clear. Proptosis can be classified in one of three anatomic locations. Ocular or bulbar disorders typically cause exophthalmos, unless there is extraocular spread of the mass lesion. Many cases of proptosis are due to primary retrobulbar disorders, including intra-orbital masses and inflammation. Extraorbital neoplasms of the osseous orbital wall, face, paranasal sinuses, nasal cavities, or frontal cranial fossae can also cause proptosis. Similarly, a variety of infectious and inflammatory lesions can cause proptosis, including cellulitis, abscess, mucocele, inflammatory orbital syndrome (IOS) or orbital pseudotumor, sarcoidosis, or Wegener's granulomatosis.

Several types of congenital or developmental lesions may cause proptosis by enlarging intraorbital structures (macrophthalmia, colobomatous cysts) or encroachment on the orbit due to osseous defects (naso-orbital cephaloceles) or osseous overgrowth (developmental fibro-osseous lesions such as fibrous dysplasia).

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Visual loss can occur as a result of damage at any point along the visual pathway extending from the globe to the occipital lobes. Therefore, depending on the specific clinical conditions, imaging may be performed of the orbits, anterior skull base, and/or brain. Visual loss may be seen in infants and children with congenital absence of portions of the eye or visual system as well as septo-optic dysplasia. Intrinsic tumors of any portion of the visual pathway or extrinsic tumors of adjacent structures (eg, sella or suprasellar cistern) may produce visual disturbances. Vascular occlusive diseases, inflammatory disease, and demyelinating disease may produce transient or fixed visual disturbances.

Ophthalmoplegia (abnormally limited eye movement) may be the result of intrinsic abnormalities within the extra-ocular muscles, extrinsic compression of these muscles by orbital masses, or abnormalities of the cranial nerves and brain stem nuclei that innervate these muscles.

Computed tomography (CT) and magnetic resonance imaging (MRI) are complementary diagnostic procedures and may be used together in some circumstances. Dedicated thin section multiplanar orbital imaging is necessary for detecting orbital abnormalities. The intrinsic contrast provided by orbital fat allows for excellent anatomic visualization with either technique. Contrast enhancement is important in assessing most orbital disorders. Because of the absence of radiation and the utility of fat suppressed contrast enhanced images, MRI is the procedure of choice for orbital disorders with the exception of trauma and assessment for foreign bodies.

#### **Optic Nerve and Sheath Disorders**

Primary disorders of the optic nerve and optic nerve sheath typically cause visual disturbances and occasionally proptosis in cases of large neoplasms. The primary neoplasms of the optic nerve include optic nerve tumors (gliomas, astrocytomas, and hamartomas) and meningiomas. There may be a para-optic component of optic nerve tumors that is characteristically seen in patients with neurofibromatosis type I. The para-optic component is due to perineural arachnoidal gliomatosis consisting of a proteinaceous material that seeds and spreads within the optic nerves subarachnoid space and is a portion of the tumor. This process results in optic nerve elongation and resultant kinking of the nerve just posterior to the globe. There is no correlation between the size of the lesion and the extent of visual loss.

Extension of tumors into the optic chiasm, optic tracts, and lateral geniculate bodies of the thalami is more accurately depicted on MRI than on CT. The size and shape of the optic canals are best assessed in the axial

projection, while the size and shape of the optic nerves are best appreciated on coronal and oblique sagittal images. Many optic nerve tumors exhibit fusiform homogeneous enhancement, while the unenhanced portions of optic nerve tumors may represent the sites of arachnoidal gliomatosis.

Meningiomas are the most common optic nerve sheath tumors, arising from the arachnoidal coverings of the optic nerve. Visual loss and optic atrophy are the usual presenting symptoms. Most cases occur in middle-aged or elderly patients, more often in women than men. The tumor may be cuff-like, surrounding the optic nerves, or eccentrically located on one side of the nerve. CT scans will often demonstrate calcifications. Enhancement parallel to the length of the optic nerves with the intact nerve seen within the mass (“tram-tracking”) is seen on both CT and MRI. MRI scans also readily depict the spread into adjacent meninges. Parallel optic cysts may be identified surrounding the optic nerve immediately distal to the meningioma. This process causes trapping of the cerebrospinal fluid in the subarachnoid space and can add to the mass effect and proptosis.

The papilledema associated with pseudotumor cerebri or intracranial mass may be detected on CT or MRI as enlargement of the optic nerve. If severe, there is a reversal of the optic nerve head, with bulging forward into the posterior wall of the globe. While dilatation of the perioptic subarachnoid space is best appreciated on fat suppressed T2 weighted images, reversal of the nerve head may be more readily detected on CT than on MRI because of the chemical shift artifact inherent to the MR studies. There is some correlation between the severity of the visual loss and the detection of enlarged nerves with reversed nerve heads. Patients with more severe visual loss demonstrate more frequent and more severe reversal of the optic nerve head. Prominent perioptic subarachnoid spaces and consequent mild diffuse enlargement of the optic nerves, however, are most often asymptomatic imaging findings unrelated to increased intracranial pressure.

### **Other Orbital Neoplasms**

Primary neoplasms may arise from any constituent orbital tissue. The most common tumors are benign cavernous hemangiomas. These lesions typically present in adulthood with proptosis. They may occur anywhere within the orbit, but they have a predilection for the intraconal space. They are seen as focal round or oval masses, often with vascular calcifications. Complex vascular lesions such as lymphangiomas and capillary hemangiomas are multicompartiment lesions that occur in infancy. They tend to regress with age. Schwannomas arising from branches of V3, V4, V5, and V6 may rarely occur in the orbit. V1 and V2 tumors may extend into the

orbit from the cavernous sinus. Primary benign and malignant lacrimal gland tumors may occur. Metastatic and lymphomatous involvement of the soft tissues of the orbit (without osseous disease) may present as an isolated intraconal mass (eg, breast carcinoma and melanoma) or as involvement of an orbital structure (eg, lymphomatous infiltration of the lacrimal gland).

### **Optic Nerve Neuropathies**

Optic neuritis cannot be reliably detected with CT. It is seen on MRI as (focal or diffuse enlargement of the optic nerve) abnormal signal intensity (hyperintensity on T2 weighted images) and/or enhancement. These features are best appreciated on fat suppressed T2 weighted and contrast enhanced T1 weighted images. Optic neuritis is caused by autoimmune inflammation of the optic nerve, which affects the myelin sheaths with relative preservation of the axons. Optic neuritis is the initial manifestation of multiple sclerosis (MS) in about 20% of cases and may occur at some point in the disease in approximately 50% of cases. Even when MRI scans of the orbit are normal imaging of the brain can reveal foci of demyelination that allow for differentiation of those patients who will subsequently develop MS from those who will not. Thus, MRI is a predictor of MS, as it can help identify a subgroup of patients with optic neuritis whose risk of developing MS appears to be low, and therefore it can help predict the development of MS after optic neuritis.

Isolated visual disturbances may be caused by a variety of optic nerve neuropathies, including radiation, chemotherapy, compressive phenomena, and ischemia. Radiation-induced optic neuropathy (RON) is a rare catastrophic complication of radiation therapy regimens used to treat a variety of neoplasms of the skull base, sella, and parasellar regions. There is typically a latency period of 6 to 36 months following treatment. Clinically, the nerve head may appear normal, but gadolinium-enhanced fat suppressed MR imaging will show patchy, linear, or confluent enhancement along the portions of the optic nerve, chiasm, or optic tract. This correlates precisely with the development of delayed visual loss after radiation therapy. Treatment of RON with corticosteroids may be helpful, although optic atrophy may develop with permanent visual loss.

Compressive optic nerve neuropathies can be caused by a variety of lesions in the orbital apex including IOS (orbital pseudotumor), thyroid ophthalmopathy with muscular hypertrophy and edema causing compression of the intraocular portion of the optic nerve, or other systemic diseases with orbital manifestations such as sarcoidosis or Wegener’s granulomatosis.

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## Vascular Disorders

Compression of the optic nerve may also occur as a result of cavernous carotid fistulae, arteriovenous malformations, or orbital varices. Such vascular anomalies may produce retrograde flow through the ophthalmic vessels with subsequent dilatation of the orbital veins and passive congestion of the orbital tissues. This leads to progressive increase in the intraocular pressure and subsequent decrease in visual acuity, or even blindness. Spontaneous thrombosis of the ophthalmic veins occasionally occurs and may aggravate the mass effect within the apex of the orbit and the compressive neuropathy.

Imaging (MRI or CT) will demonstrate the dilated ophthalmic veins, facial veins and other regional venous structures along with enlargement of the cavernous sinus. Large edematous extraocular muscles and periorbital structures may be identified. Proptosis is typically present. The addition of MR angiography or CT angiography allows for flow assessments along with the static morphologic changes. In some cases, conventional angiography may be required to make the definitive diagnosis, although this is most commonly used in conjunction with therapeutic interventional procedures.

Traumatic optic neuropathy (TON) results in blindness following optic nerve injury. The injury is typically from head trauma without direct trauma to the globe or retina. The clinical presentation of TON may be delayed, as the patients may present with depressed levels of consciousness. Post-traumatic visual loss can be evaluated by investigation of the soft tissue and osseous structures around the optic nerve and chiasm. Thin section CT scans with multiplanar reconstruction are the most useful. Such images provide accurate identification of indirect signs of injury to the optic nerve, such as dehiscence, or bony fragments within the orbit or optic nerve canal, narrowing of the optic canal, or significant bony separations which indicate likely optic nerve injury. MR images have been shown to be more sensitive for detecting optic nerve edema or avulsion. Differential response to steroid therapy or orbital surgical decompressive procedures remains controversial and shows no clear-cut advantages.

## Inflammatory Orbital Syndrome

Inflammatory orbital syndrome (orbital pseudotumor, inflammatory fibromyotendinitis) may appear as an acute or chronic cause of ophthalmoplegia, proptosis, and visual loss that develops as a diffuse infiltrate or focal mass. In the diffuse form, there is inflammatory infiltrate of the orbital fat, extraocular muscles, and adnexal structures, particularly in the orbital apex. The more focal forms of IOS commonly involve the tendinous portions of the extraocular muscles (myositic form), the uveal structures (anterior form), the scleral region (posterior form), or the

lacrimal gland (lacrimal form). All forms of the disease show extensive infiltration that is histologically composed of polyclonal lymphocytes, plasma cells, neutrophils, and macrophages with various amounts of fibrosis. Commonly, the retrobulbar fat has a “dirty” appearance.

The acute form of IOS is typically unilateral, developing over days to weeks, and is characterized by ophthalmoplegia pain, swelling, and tenderness. The chronic form of the disease may occur bilaterally in approximately 10%-15% of cases, develops insidiously over weeks to months, and lacks inflammatory symptoms.

CT and MRI show intraconal or extraconal soft tissue lesions that are diffuse or localized and commonly involve the orbital apices. Occasionally, there may be a well-defined mass lesion that mimics a neoplasm. In virtually all cases, there is prominent enhancement on postcontrast CT or MR scans. In the chronic form of the disease there is increased fibrosis in the lesions, resulting in decreased signal on T2-weighted images.

Many cases of IOS resolve following steroid therapy, but the process may progress to a lymphoproliferative disorder or lymphoma. The differentiation of IOS from lymphoma is based on histopathological examination, with a predominant polyclonal lymphocytic population in the earlier disorder and a monoclonal lymphocytic process in the latter disorder. CT or MRI scans may be used to follow the course of the illness until it resolves, or recurs in the chronic form of the disease.

As previously mentioned, there may be involvement of the optic nerve, resulting in alterations of visual acuity and/or involvement of the ipsilateral cavernous sinus. When there is secondary thrombosis of the sphenoidal veins or cavernous sinus, a painful ophthalmoplegia results, with the presumptive diagnosis of Tolosa-Hunt syndrome. Although usually confined to the orbital soft tissues, IOS can produce bone destruction or extraorbital extension.

A small subset of patients with isolated ocular manifestations of IOS have posterior scleritis. These patients are seen for ocular pain and proptosis within limitation of gaze, scleral effusions, disc edema, and intraocular hemorrhages. Posterior scleritis shows inflammatory signs in the coat of the eye sclera with thickening of the posterior coat of the eye sclera that may be identified as areas of enhancement on CT or MRI. The thickened sclera, enhanced by contrast, presents as a so-called “ring sign.” Sarcoidosis (neurosarcoidosis) and Wegener’s granulomatosis both simulate IOS, lymphoproliferative disorders, or metastatic neoplasms. There are no key clinical differentiating features, nor is the response to corticosteroid therapy an indication of the

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etiology. The definitive diagnosis is made by biopsy and is often essential in order to direct primary treatment.

### **Endocrine Disorders**

Thyroid ophthalmopathy (Graves' disease) may be detected in patients who are hyperoid-thyroid, hypoid-thyroid, or euthyroid. Many (25%-50%) of these cases occur in patients with Graves' hyperthyroidism and occasionally in patients with Hashimoto's thyroiditis. The female-to-male ratio is 4 to 1, and approximately 15% of patients are under age 15. In all age groups, approximately 15% of unilateral orbital proptosis and the majority of bilateral proptosis are secondary to thyroid ophthalmopathy.

The disease presents with eyelid lag, upper lid retraction, diffuse conjunctival edema, and vascular injection at the insertion of the rectus muscles. It is characterized as an immune disorder with both cell-mediated and humoral components. The clinical features are secondary to overproduction of glycosaminoglycans, which are produced by stimulated fibroblasts. The overstimulation of fibroblasts is thought to be centered on the T-cell lymphocytes, which recognize fibroblasts' cross-reactive antigens and release cytokines. The overly produced glycosaminoglycans are deposited into muscle bellies of the extraocular muscles in a perivascular location. This results in endomysial fibrosis secondary to the mucopolysaccharide deposits.

On CT and MRI studies, there is enlargement of one or more of the extraocular rectus muscles. Multiple muscle involvement is much more common than one or two isolated muscle involvement. The disease is bilateral in at least 85% of cases by imaging criteria. The inferior rectus is most commonly and severely involved, followed by the medial superior and lateral rectus muscles. The posterior and middle third of the muscle bellies are most affected, with relative sparing of tendinous insertions. The inherent soft tissue contrast of MR scans provides elegant morphologic information regarding the involvement of the extraocular muscles in patients with thyroid ophthalmopathy. An important role of imaging is demonstrating the relationship of the extraocular muscles to the optic nerve at the orbital apex, and the degree of stretching of the optic nerve due to proptosis, particularly if surgery is contemplated. The ability to measure the T2 signal intensity on MRI helps both in determining which patients may benefit from corticosteroid therapy (those with high T2 values), and/or which patients require combined therapies including cyclosporin (based on a measurable response on serial MR images).

### **Disorders of Size or Shape of the Globe**

A staphyloma represents a diffusely enlarged globe with thin scleral margins resulting from degeneration of the

bulbar coverings. The lesion is thought to be secondary to the normal aging process and is only seen in the elderly population, either unilaterally or bilaterally. Imaging studies will demonstrate the enlarged globe with thin walls and no other lesions. Most often, enlargement is an incidental finding on images that were obtained for unrelated conditions. A diffusely enlarged globe is seen in patients with severe axial myopia, which, unlike a staphyloma, is a heritable condition treated by corrective lenses or keratotomy. Staphyloma is distinguished from coloboma, a congenital lesion where there is a complete defect in the wall of the globe with focal outpouching of the posterior globe at the optic nerve head. Coloboma may be isolated or seen in association with other congenital anomalies of the eye, anterior skull base and/or brain.

### **Retinal, Choroidal and Subhyaloid Detachments**

Serous choroidal detachments result from inflammatory diseases (uveitis, scleritis) or from accidental perforation of the eyeball. As the edema of the choroid increases, fluid accumulates in the subchoroidal potential space. Hemorrhagic choroidal detachments often occur after a contusion, a penetrating injury, or as a complication of intraocular surgery. The differentiation of choroidal effusion from choroidal hemorrhage may be obtained using multiple MRI sequences. There is a crescentic ring-shaped area of increased signal on both T1- and T2-weighted images. With choroidal hemorrhages, the signal intensity varies according to the age of the hemorrhage. In acute hemorrhages, CT may be more specific, showing the increased density of subchoroidal hemorrhage.

Retinal detachments as a complication of systemic diseases such as hypertension or diabetes are fairly common and rarely require imaging. Retinal detachments may also occur with primary ocular neoplasms such as retinoblastomas in children and as uveal malignant melanomas in adults and elderly patients. Ocular sonography may be more accurate in detecting small tumors; however, enhanced MR images are useful in determining the true extent of lesions beyond the ocular structures and also demonstrating associated retinal detachments. CT scanning has specific value in assessing patients with retinoblastoma, since small punctuate calcifications in the contralateral "normal" eye indicate the presence of bilateral disease, altering management and prognosis. Improvement in the differential diagnosis is based on postcontrast T1-weighted images, which are most helpful in detecting uveal melanomas and in differentiating melanomas from subretinal fluid collections. There is enhancement in the case of neoplasms, but not from fluid collections.

The differentiation of an amelanotic melanoma from a hemorrhagic subretinal hemorrhage is based on both the precontrast and postcontrast T1-weighted images. Of note

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are metastatic lesions to the retina or certain inflammatory conditions that cannot be consistently differentiated from primary uveal melanomas. Doppler sonography may help detect vascularity within an intraocular tumor and help differentiate such entities from nonvascular choroidal, subretinal, or subhyaloid effusions, or from hematomas.

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An 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 exams 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.