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 orbits without and with IV contrast	8	CT may be considered the preferred imaging modality when rhinologic or paranasal sinus disease is the suspected etiology for the symptoms and signs.	O
MRI head and orbits without IV contrast	7	CT may be considered the preferred imaging modality when rhinologic or paranasal sinus disease is the suspected etiology for the symptoms and signs.	O
CT head with IV contrast	6	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
CT head without and with IV contrast	6	CT may be considered the preferred imaging modality when rhinologic or paranasal sinus disease is the suspected etiology for the symptoms and signs. Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢☢
CT head without IV contrast	5	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
MRA head and neck without IV contrast	4		O
MRA head and neck without and with IV contrast	4		O
CTA head and neck with IV contrast	2	If vascular disease is suspected.	☢☢☢☢
X-ray orbit	1		☢
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

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

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

Radiologic Procedure	Rating	Comments	RRL*
MRI head and orbits without and with IV contrast	8		O
MRI head and orbits without IV contrast	7		O
CT head with IV contrast	6	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢ ☢ ☢
CT head without and with IV contrast	6	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢ ☢ ☢ ☢
CT head without IV contrast	5	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢ ☢ ☢
MRA head and neck without IV contrast	4		O
MRA head and neck without and with IV contrast	4		O
CTA head and neck with IV contrast	2	If vascular disease is suspected.	☢ ☢ ☢ ☢
X-ray orbit	1		☢
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

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

Variant 3: **Adult with sudden onset of painless or painful visual loss.**

Radiologic Procedure	Rating	Comments	RRL*
MRI head and orbits without and with IV contrast	8	CT may be considered the preferred imaging modality when rhinologic or paranasal sinus disease is the suspected etiology for the symptoms and signs.	O
MRI head and orbits without IV contrast	7	CT may be considered the preferred imaging modality when rhinologic or paranasal sinus disease is the suspected etiology for the symptoms and signs.	O
CT head with IV contrast	6	CT may be considered the preferred imaging modality when rhinologic or paranasal sinus disease is the suspected etiology for the symptoms and signs. Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
CT head without IV contrast	5	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
CT head without and with IV contrast	5	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
CTA head and neck with IV contrast	5	If vascular disease is suspected.	☢☢☢
MRA head and neck without IV contrast	4		O
MRA head and neck without and with IV contrast	4		O
X-ray orbit	1		☢
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

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

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

Radiologic Procedure	Rating	Comments	RRL*
MRI head and orbits without and with IV contrast	8	CT may be considered the preferred imaging modality when rhinologic or paranasal sinus disease is the suspected etiology for the symptoms and signs.	O
MRI head and orbits without IV contrast	7	CT may be considered the preferred imaging modality when rhinologic or paranasal sinus disease is the suspected etiology for the symptoms and signs.	O
CT head with IV contrast	6	CT may be considered the preferred imaging modality when rhinologic or paranasal sinus disease is the suspected etiology for the symptoms and signs. Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢ ☢ ☢
CT head without and with IV contrast	6	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢ ☢ ☢
CT head without IV contrast	5	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢ ☢ ☢
MRA head and neck without IV contrast	4		O
MRA head and neck without and with IV contrast	4		O
CTA head and neck with IV contrast	4	If vascular disease is suspected.	☢ ☢ ☢
X-ray orbit	1		☢
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

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

Variant 5: **Adult patient with uveitis, scleritis, and visual loss.**

Radiologic Procedure	Rating	Comments	RRL*
MRI head and orbits without and with IV contrast	8		O
MRI head and orbits without IV contrast	7		O
CT head with IV contrast	5	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
CT head without and with IV contrast	5	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
CTA head and neck with IV contrast	4	If vascular disease is suspected.	☢☢☢
MRA head and neck without IV contrast	4		O
MRA head and neck without and with IV contrast	4		O
CT head without IV contrast	4	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
X-ray orbit	1		☢
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

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

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

Radiologic Procedure	Rating	Comments	RRL*
MRI head and orbits without and with IV contrast	9		O
MRI head and orbits without IV contrast	6		O
MRA head and neck without IV contrast	6		O
MRA head and neck without and with IV contrast	6		O
CT head with IV contrast	6	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
CT head without and with IV contrast	6	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
CTA head and neck with IV contrast	6	If vascular disease is suspected.	☢☢☢
CT head without IV contrast	5	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
X-ray orbit	1		☢
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

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

Variant 7: **Head injury with visual loss.**

Radiologic Procedure	Rating	Comments	RRL*
CT head without IV contrast	7	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢ ☢ ☢
MRI head and orbits without IV contrast	7	If MRI is safe.	O
MRI head and orbits without and with IV contrast	5	If MRI is safe.	O
CT head with IV contrast	5	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢ ☢ ☢
CT head without and with IV contrast	5	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢ ☢ ☢
CTA head and neck with IV contrast	4	If vascular disease is suspected.	☢ ☢ ☢
MRA head and neck without IV contrast	3		O
MRA head and neck without and with IV contrast	3		O
X-ray orbit	2		☢
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

ORBITS, VISION AND VISUAL LOSS

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Summary of Literature Review

Introduction/Background

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, whereas exophthalmos is an abnormal prominence of the globe. Clinically, it may be impossible to differentiate these two entities without the aid of imaging. Exophthalmos may be caused by primary ocular or bulbar disorders such as macrophthalmia or colobomatous cysts, retrobulbar disorders such as intraorbital masses and inflammation, and extraorbital disorders such as masses or inflammation in the osseous orbital wall, face, paranasal sinuses, nasal cavities, or frontal cranial fossae [1].

Visual loss may be caused by damage at any location along the visual pathway extending from the globe to the occipital lobes. Therefore, the choice of appropriate imaging modality and focus depends on the specific clinical condition, and may include portions 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**.** Imaging of brain lesions that could result in visual loss such as stroke and cerebrovascular disease, demyelination, or tumors are covered in other ACR Appropriateness Criteria® topics.

Ophthalmoplegia (abnormally limited eye movement) may be caused by 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.

Imaging Modalities

Imaging analysis of orbital diseases is facilitated by a compartmental approach that establishes differential diagnoses on the basis of the location of the process within the orbit [2,3]. Computed tomography (CT) and magnetic resonance imaging (MRI) are complementary diagnostic procedures and may be used together in some circumstances [4]. For example, CT is usually used for suspected thyroid ophthalmopathy whereas MRI is preferred for suspected masses [5]. CT is useful in evaluating bony structures, and MRI excels in evaluating soft tissues [6-11]. Dedicated thin-section multiplanar orbital imaging is recommended for detecting orbital abnormalities [12]. 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 its absence of radiation and the utility of fat-suppressed contrast-enhanced images, MRI has emerged as the procedure of choice for orbital disorders, with the exception of trauma and assessment for foreign bodies [10,13-16]. Moreover, specialized surface coils have expanded the utility of MRI [17]. In addition to diagnostic uses for imaging, both CT and MRI are becoming indispensable tools for surgical navigation of the tissues surrounding the orbit such as

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the paranasal sinuses [18,19]. MRI is also an investigative research tool for exploring white matter tracts and functional aspects of the visual system [20-23]. Ultrasound and fluorescein angiography are also important modalities; however, these special procedures are usually performed by the ophthalmologist and are not covered in this article.

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. CT and MRI studies will demonstrate the enlarged globe with thin walls and no other lesions. 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. Hemorrhagic choroidal detachments often occur after a contusion, a penetrating injury, or as a complication of intraocular surgery. MRI may differentiate choroidal effusion from choroidal hemorrhage. 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 in 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 subretinal hemorrhage is based on both the precontrast and postcontrast T1-weighted images. Of note 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.

Disorders of the Optic Nerve Sheath Complex

Primary disorders of the optic nerve sheath complex typically cause visual disturbances and occasionally proptosis. The primary neoplasms of the optic nerve include optic nerve tumors (gliomas, astrocytomas, and hamartomas) and meningiomas. A paraoptic component of optic nerve tumors, consisting of proteinaceous subarachnoid seeding, may be seen in patients with neurofibromatosis type I and contributes to optic nerve elongation and kinking [24]. 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. MRI is best for this differentiation. CT scans best demonstrate calcifications in lesions such as meningiomas. 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.

The papilledema associated with pseudotumor cerebri or intracranial mass may enlarge the optic nerve as detected on CT or MRI. While dilatation of the perioptic subarachnoid space is best appreciated on fat-suppressed T2-weighted images, reversal of the nerve head, manifested by bulging of the posterior portion of the globe, may be more readily detected on CT than on MRI because of the chemical shift artifact inherent to the MR studies. MRI may also monitor optic nerve damage in other disorders such as glaucoma [25].

Optic neuritis is best seen on MRI as focal or diffuse enlargement of the optic nerve, abnormal 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 may be seen with multiple sclerosis (MS). MRI has become an essential study for evaluating patients with suspected MS and supplements other clinical studies [26]. Even when MRI scans of the orbit are normal, imaging of the brain may reveal foci of demyelination.

Radiation-induced optic neuropathy (RON) is best evaluated with gadolinium-enhanced fat-suppressed MRI, which may show patchy, linear, or confluent enhancement along the portions of the optic nerve, chiasm, or optic tract.

Other Orbital Neoplasms

Primary neoplasms may arise from any constituent orbital tissue. The most common tumors are benign cavernous hemangiomas and have a predilection for the intraconal space. They present as focal round or oval masses with distinctive hyperintense appearance on MR images. Vascular calcifications can be detected on CT. Complex vascular lesions such as lymphangiomas and capillary hemangiomas, schwannomas arising from branches of cranial nerves III, IV, V, and VI, primary benign and malignant lacrimal gland tumors, metastases, and lymphomatous involvement of the soft tissues of the orbit (without osseous disease) may also present as isolated masses with or without involvement of adjacent orbital structures. MRI is the preferred imaging modality for evaluation.

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. Imaging (MRI or CT) will demonstrate the dilated ophthalmic veins, facial veins, and other regional venous structures, along with enlargement of the cavernous sinus [27]. Large edematous extraocular muscles and periorbital structures may be identified. The addition of MR angiography or CT angiography allows for flow assessments along with the static morphologic changes [28,29]. In some cases, conventional angiography may be required to make the definitive diagnosis, although it is most commonly used in conjunction with therapeutic interventional procedures.

Traumatic optic neuropathy (TON) and post-traumatic visual loss may be evaluated by investigation of the soft tissue and osseous structures surrounding the optic nerve and chiasm. Thin-section CT scans with multiplanar reconstruction are the most useful [5,30]. 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.

Inflammatory Orbital Syndrome

Inflammatory orbital syndrome (IOS) (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.

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 MRI scans. In the chronic form of the disease there is increased fibrosis in the lesions, resulting in decreased signal on T2-weighted images. 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.

A small subset of patients with isolated ocular manifestations of IOS have posterior scleritis. Posterior scleritis shows inflammatory signs in the coat of the eye (sclera) with thickening of the posterior sclera that may be identified as areas of enhancement on CT or MRI. Sarcoidosis (neurosarcoidosis) and Wegener's granulomatosis both simulate IOS, lymphoproliferative disorders, or metastatic neoplasms.

Endocrine Disorders

Thyroid ophthalmopathy (Graves' disease) may be detected in hyperthyroid, hypothyroid, or euthyroid patients. In all age groups, approximately 15% of unilateral orbital proptoses and the majority of bilateral proptoses are secondary to thyroid ophthalmopathy.

CT is usually the first modality used for evaluation [5]. 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 the involvement of just one or two isolated muscles. 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 MRI 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).

Summary

- Imaging analysis of the orbit is facilitated by a compartmental approach.
- CT and MRI are complementary diagnostic procedures for suspected orbital pathology.
- CT is useful in evaluating bony structures.
- MRI is useful in evaluating soft-tissue structures the globe, optic nerves, and intraconal and extraconal spaces.
- CT angiography, MR angiography, and conventional angiography may be useful in vascular conditions.
- Conventional angiography may be useful in delivering therapeutic intervention.

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, both because of 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 to 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**
O	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 (eg, region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as "Varies".		

Supporting Documents

For additional information on the Appropriateness Criteria methodology and other supporting documents go to www.acr.org/ac.

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