American College of Radiology ACR Appropriateness Criteria[®] Orbits, Vision and Visual Loss

Variant 1: Traumatic visual defect. Suspect orbital injury. Initial imaging.			
Procedure	Appropriateness Category	RRL	
CT orbits without IV contrast	Usually Appropriate	\$ \$ \$	
CT head without IV contrast	Usually Appropriate	\$ \$ \$	
MRI head without IV contrast	May Be Appropriate	0	
MRI orbits without IV contrast	May Be Appropriate	0	
CT orbits with IV contrast	May Be Appropriate (Disagreement)	\$ \$ \$	
CTA head and neck with IV contrast	May Be Appropriate	\$ \$ \$	
MRI head without and with IV contrast	May Be Appropriate	0	
MRI orbits without and with IV contrast	May Be Appropriate (Disagreement)	0	
MRA head and neck without and with IV contrast	May Be Appropriate	0	
MRA head and neck without IV contrast	May Be Appropriate	0	
Arteriography cervicocerebral	Usually Not Appropriate	* * *	
CT head with IV contrast	Usually Not Appropriate	\$P \$P \$P	
CT head without and with IV contrast	Usually Not Appropriate	\$P \$P \$P	
CT orbits without and with IV contrast	Usually Not Appropriate	\$P \$P \$P	
X-ray orbit	Usually Not Appropriate	\$	

Traumatic visual defect. Suspect orbital injury. Initial imaging.

<u>Variant 2:</u>

Nontraumatic orbital asymmetry, exophthalmos, or enophthalmos. Initial imaging.

Procedure	Appropriateness Category	RRL
MRI orbits without and with IV contrast	Usually Appropriate	0
CT orbits with IV contrast	Usually Appropriate	\$ \$ \$
CT orbits without IV contrast	May Be Appropriate	\$ \$ \$
CTA head and neck with IV contrast	May Be Appropriate	\$ \$ \$
MRA head and neck without and with IV contrast	May Be Appropriate	0
MRI head without and with IV contrast	May Be Appropriate	Ο
MRI orbits without IV contrast	May Be Appropriate	О
MRA head and neck without IV contrast	May Be Appropriate (Disagreement)	0
MRI head without IV contrast	May Be Appropriate	0
Arteriography cervicocerebral	May Be Appropriate	* * *
CT head with IV contrast	May Be Appropriate	* * *
CT head without IV contrast	May Be Appropriate	* *
CT head without and with IV contrast	Usually Not Appropriate	\$P \$P \$P
CT orbits without and with IV contrast	Usually Not Appropriate	\$ \$ \$
X-ray orbit	Usually Not Appropriate	•

<u>Variant 3:</u>

Suspected orbital cellulitis, uveitis, or scleritis. Initial imaging.

Procedure	Appropriateness Category	RRL
CT orbits with IV contrast	Usually Appropriate	\$ \$ \$
MRI orbits without and with IV contrast	Usually Appropriate	0
CT orbits without IV contrast	May Be Appropriate	\$ \$ \$
MRI head without and with IV contrast	May Be Appropriate	0
MRI orbits without IV contrast	May Be Appropriate	0
CTA head and neck with IV contrast	May Be Appropriate	\$ \$ \$
MRA head and neck without and with IV contrast	May Be Appropriate	0
MRI head without IV contrast	May Be Appropriate	0
CT head with IV contrast	May Be Appropriate	\$ \$ \$
MRA head and neck without IV contrast	May Be Appropriate	0
Arteriography cervicocerebral	Usually Not Appropriate	\$ \$ \$
CT head without IV contrast	Usually Not Appropriate	\$ \$ \$
CT orbits without and with IV contrast	Usually Not Appropriate	\$ \$ \$
CT head without and with IV contrast	Usually Not Appropriate	\$ \$ \$
X-ray orbit	Usually Not Appropriate	•

<u>Variant 4:</u>

Procedure	Appropriateness Category	RRL
MRI orbits without and with IV contrast	Usually Appropriate	0
MRI head without and with IV contrast	Usually Appropriate	0
MRI orbits without IV contrast	Usually Appropriate	0
MRI head without IV contrast	May Be Appropriate	0
CT orbits 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 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 and neck with IV contrast	Usually Not Appropriate	\$ \$ \$
MRA head and neck without and with IV contrast	Usually Not Appropriate	0
MRA head and neck without IV contrast	Usually Not Appropriate	0
Arteriography cervicocerebral	Usually Not Appropriate	* * *
X-ray orbit	Usually Not Appropriate	•

<u>Variant 5:</u>

Visual loss. Etiology identified on ophthalmologic examination or laboratory tests.

Procedure	Appropriateness Category	RRL
MRI orbits without IV contrast	Usually Not Appropriate	0
CT head without IV contrast	Usually Not Appropriate	\$ \$ \$
CT orbits without IV contrast	Usually Not Appropriate	\$ \$ \$
MRI head without IV contrast	Usually Not Appropriate	0
MRI orbits without and with IV contrast	Usually Not Appropriate	0
Arteriography cervicocerebral	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 and neck with IV contrast	Usually Not Appropriate	\$ \$ \$
MRA head and neck without and with IV contrast	Usually Not Appropriate	0
MRA head and neck without IV contrast	Usually Not Appropriate	0
MRI head without and with IV contrast	Usually Not Appropriate	0
X-ray orbit	Usually Not Appropriate	•

<u>Variant 6:</u>

Procedure	Appropriateness Category	RRL
MRI orbits without and with IV contrast	Usually Appropriate	0
CT orbits with IV contrast	Usually Appropriate	\$ \$ \$
MRI orbits without IV contrast	Usually Appropriate	0
CT orbits without IV contrast	May Be Appropriate	\$ \$ \$
MRI head without and with IV contrast	May Be Appropriate	0
CT head with IV contrast	May Be Appropriate	\$ \$ \$
MRI head without IV contrast	May Be Appropriate	0
CT head without IV contrast	May Be Appropriate	\$ \$ \$
CTA head and neck with IV contrast	May Be Appropriate	\$P \$P \$P
MRA head and neck without and with IV contrast	May Be Appropriate	0
MRA head and neck without IV contrast	May Be Appropriate	О
Arteriography cervicocerebral	Usually Not Appropriate	* * *
CT head without and with IV contrast	Usually Not Appropriate	* * *
CT orbits without and with IV contrast	Usually Not Appropriate	* * *
X-ray orbit	Usually Not Appropriate	Ð

<u>Variant 7:</u>

Procedure	Appropriateness Category	RRL
MRI head without and with IV contrast	Usually Appropriate	0
MRI head without IV contrast	Usually Appropriate	0
CT head with IV contrast	May Be Appropriate	* * *
CT head without and with IV contrast	May Be Appropriate	\$ \$ \$
CT head without IV contrast	May Be Appropriate	* * *
CTA head and neck with IV contrast	May Be Appropriate	* * *
MRA head and neck without and with IV contrast	May Be Appropriate	0
CT venography head with IV contrast	May Be Appropriate	* * *
MR venography head without and with IV contrast	May Be Appropriate	0
MR venography head without IV contrast	May Be Appropriate	0
MRA head and neck without IV contrast	May Be Appropriate	О
CT orbits with IV contrast	Usually Not Appropriate	* * *
CT orbits without IV contrast	Usually Not Appropriate	* * *
MRI orbits without and with IV contrast	Usually Not Appropriate	0
MRI orbits without IV contrast	Usually Not Appropriate	0
Arteriography cervicocerebral	Usually Not Appropriate	\$ \$ \$
CT orbits without and with IV contrast	Usually Not Appropriate	\$P \$P \$P
X-ray orbit	Usually Not Appropriate	\$

<u>Variant 8:</u>

Procedure	Appropriateness Category	RRL
MRI head without and with IV contrast	Usually Appropriate	0
MRI orbits without and with IV contrast	Usually Appropriate	0
CT orbits with IV contrast	Usually Appropriate	\$ \$ \$
MRI orbits without IV contrast	Usually Appropriate	0
CT orbits without IV contrast	May Be Appropriate	\$ \$ \$
CTA head and neck with IV contrast	May Be Appropriate	\$ \$ \$
MRA head and neck without and with IV contrast	May Be Appropriate	0
MRA head and neck without IV contrast	May Be Appropriate	О
MRI head without IV contrast	May Be Appropriate	0
CT head with IV contrast	May Be Appropriate	\$ \$ \$
CT head without IV contrast	May Be Appropriate	\$ \$ \$
Arteriography cervicocerebral	Usually Not Appropriate	\$ \$ \$
CT head without and with IV contrast	Usually Not Appropriate	\$ \$ \$
CT orbits without and with IV contrast	Usually Not Appropriate	\$ \$ \$
X-ray orbit	Usually Not Appropriate	•

ORBITS, VISION AND VISUAL LOSS

Expert Panel on Neurologic Imaging: Tabassum A. Kennedy, MD^a; Amanda S. Corey, MD^b; Bruno Policeni, MD^c; Vikas Agarwal, MD^d; Judah Burns, MD^e; H. Benjamin Harvey, MD, JD^f; Jenny Hoang, MBBS^g; Christopher H. Hunt, MD^h; Amy F. Juliano, MDⁱ; William Mack, MD^j; Gul Moonis, MD^k; Gregory J. A. Murad, MD^l; Jeffrey S. Pannell, MD^m; Matthew S. Parsons, MDⁿ; William J. Powers, MD^o; Jason W. Schroeder, MD^{p*}; Gavin Setzen, MD^q; Matthew T. Whitehead, MD^r; Julie Bykowski, MD.^s

Summary of Literature Review

Introduction/Background

A thorough neurologic and ophthalmologic examination can often accurately localize a defect along the visual pathway. Combined with factors such as the age of the patient, the time-course for onset, degree of visual loss at presentation, the presence of eye pain, and visible exophthalmos or enophthalmos determine if imaging is needed, the choice of imaging modality, coverage to include the orbit, anterior skull base and/ or brain, and contrast phase [1].

Disease along the visual pathway may be intrinsically related to the globe, optic nerve, optic chiasm, optic tracts, optic radiations, or primary visual cortex or related to extrinsic mass effect from adjacent pathology upon these structures. Primary diseases of the orbit may present with proptosis, visual disturbance, and/or ophthalmoplegia. These signs and symptoms may occur alone or in combination and may be accompanied by pain or other features including vascular congestion or erythema. The differential diagnosis in adults and subsequent appropriate imaging tests are dependent on the pattern and whether visual loss is monocular or binocular [2].

It is important to note the overlap of visual loss and other conditions addressed by independent ACR Appropriateness Criteria. Acute ischemic or hemorrhagic stroke should be emergently excluded in the setting of sudden onset, painless visual loss, and is extensively reviewed in the ACR Appropriateness Criteria[®] "<u>Cerebrovascular Disease</u>" [3]. The ACR Appropriateness Criteria[®] "<u>Headache</u>" [4] addresses the need for immediate evaluation in the setting of papilledema [1], as well as imaging of suspected giant-cell arteritis and posterior reversible encephalopathy, which may have associated visual symptoms.

Imaging analysis of orbital diseases is facilitated by a compartmental approach that establishes a differential diagnosis on the basis of lesion location along the visual pathway [5]. Computed tomography (CT) and magnetic resonance imaging (MRI) are often complementary when assessing visual loss [6,7]. The inherent contrast provided by orbital fat allows for excellent anatomic definition with either technique. Ultrasound (US) and fluorescein angiography are also important diagnostic tools; however, these unique procedures are most often performed by the ophthalmologist and are beyond the scope of this article.

Discussion of Procedures by Variant

Variant 1: Traumatic visual defect. Suspect orbital injury. Initial imaging.

Patients with traumatic orbital injury may have injuries that are isolated to the orbit or have intracranial manifestations, depending on the mechanism and severity of injury. In the United States, orbital trauma accounts for approximately 3% of visits to the emergency department [8]. Orbital injury should be suspected if periorbital soft-tissue swelling, hyphema, vision loss, or extraocular restriction is present.

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The American College of Radiology seeks and encourages collaboration with other organizations on the development of the ACR Appropriateness Criteria through society representation on expert panels. Participation by representatives from collaborating societies on the expert panel does not necessarily imply individual or society endorsement of the final document.

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СТ

Traumatic optic neuropathy and post-traumatic visual loss are typically evaluated with noncontrast thin-section orbital CT imaging with multiplanar reconstructions [7,9-13]. Contrast is typically not needed in the trauma setting. Orbital CT is superior at identifying the integrity of the osseous orbit and skull base and is useful in identifying fractures, displaced fracture fragments, as well as narrowing of the optic canal. Associated soft-tissue findings include intraorbital foreign bodies, hematomas, and extraocular muscle injury and are readily seen with noncontrast CT imaging of the orbits [14]. If orbital fractures are identified, orbital CT imaging can be useful at identifying the position of the extraocular muscles relative to the fracture defect as well as the overall size of the fracture as a predictor for the development of enophthalmos, which may be useful in surgical planning [15]. In patients with more severe mechanisms of injury, a CT of the head without contrast may also be indicated to assess for intracranial injury. Please refer to the ACR Appropriateness Criteria[®] "Head Trauma" [16]. Precontrast and postcontrast head CT or orbit CT imaging is usually not appropriate.

MRI

MRI of the orbits without contrast may be complementary in assessing traumatic optic neuropathy. MRI has been shown to be more sensitive for detecting optic nerve edema or avulsion than CT [17]. In patients with optic nerve injury initially suspected on CT, or in patients with unexplained visual loss following facial trauma, MR of the orbits without contrast may be helpful in assessing the integrity of the optic nerve. MRI of the brain without contrast may also provide additional findings related to intracranial hemorrhage in the setting of traumatic brain injury and in assessment of traumatic cranial nerve injury. Please refer to the ACR Appropriateness Criteria[®] "<u>Head Trauma</u>" [16] for additional recommendations. However, if there is a suspicion for a metallic foreign body in the orbit, an MRI is contraindicated. Contrast is typically not needed in the setting of trauma.

CTA, MRA, Arteriography

There is a role for angiography in assessing vascular injury in the setting of trauma; however, this should be assessed in the larger context related to the overall extent of traumatic facial and intracranial injury and is not typically indicated as the initial imaging test for orbital trauma. Please refer to the ACR Appropriateness Criteria[®] "<u>Head Trauma</u>" [16]. CT angiography (CTA), MR angiography (MRA), or catheter-based digital subtraction angiography (DSA) may all be performed to evaluate for traumatic vascular injury; however, CTA in the trauma setting is often the preferred study for assessment of vascular injury.

Radiography

Orbital or skull radiographs are insufficient to detect pathology in patients presenting with trauma and have primarily been supplanted by CT.

Variant 2: Nontraumatic orbital asymmetry, exophthalmos, or enophthalmos. Initial imaging.

Orbital asymmetry is a broad term that refers to the external appearance of the orbit. This asymmetry may be related to globe position, with one eye perceived or measured as more proptotic compared to the other, or one eye perceived as sunken or retracted compared to the other. Normal range measurements vary and are dependent on the race of the individual [18,19].

Bilateral exophthalmos may indicate an underlying systemic or diffuse condition, such as thyroid eye disease. Unilateral or asymmetric proptosis is concerning for an underlying mass or pathologic process intrinsic to the globe, optic nerve, extraocular muscles, lacrimal glands, or adjacent soft-tissue structures, posterior to the orbit, within the adjacent skull base or cavernous sinus. Vascular malformations may result in proptosis in adults, and occur anywhere within the orbit. Carotid-cavernous fistula (CCF) may present with proptosis with orbital congestion and chemosis in the setting of an anterior-draining CCF or diplopia and pain in the posterior-draining CCFs [20].

Enophthalmos, or posterior displacement of the globe, may be caused by a development condition resulting in an absent globe (anophthalmia) or small globe (microphthalmia) by traumatic injury to the bony orbit, silent sinus syndrome, processes that result in atrophy of the extraocular muscles, or by a desmoplastic neoplastic/inflammatory process [9,21-24].

If the asymmetry is associated with a white pupillary reflex (leukocoria), the primary concern is an abnormality localized to the globe. Although leukocoria is a term often used in the pediatric population, this term is not limited to children. Any condition that prevents passage of light through the globe may cause leukocoria, including tumors, developmental processes, and infection [25,26]. Initial evaluation in a patient with leukocoria consists of

a thorough assessment by an ophthalmologist. Many of the aforementioned conditions can be diagnosed based on the patient's clinical history, ophthalmologic investigation including ophthalmoscopy, and ophthalmology-directed US and may not require additional imaging.

Patients with disconjugate gaze between the two eyes may also present with orbital asymmetry. These patients may present with diplopia or double vision and is further discussed in Variant 8.

MRI

In patients with proptosis or if a mass lesion is suspected within the globe, optic nerve, within the adjacent orbital soft tissues, or within the adjacent skull base, an MRI of the orbits without and with contrast is the optimal imaging modality used to localize and characterize the primary lesion [27-30].

MRI has improved soft tissue characterization, and diffusion-weighted imaging may be particularly useful in situations where lymphoma is a consideration [31]. Although contrast is preferred, an MRI of the orbits without contrast may be appropriate if contrast cannot be given. An MRI of the head without and with contrast may also be added to assess the extent of intracranial disease and to evaluate for distant intracranial metastasis. A CT of the orbits is complementary in assessing orbital lesion characteristics and the extent of disease in this clinical presentation [7].

Orbital inflammatory conditions including thyroid eye disease, IgG4-related disease, and idiopathic orbital inflammatory syndrome may all present with unilateral or bilateral proptosis as a clinical manifestation. Like other orbital conditions, patients with these conditions may be imaged with CT or MRI, and these modalities provide overlapping information related to disease extent. Currently, there is no consensus on the optimal imaging modality to assess patients presenting with idiopathic orbital inflammatory disease or IgG4-related orbital disease. If intracranial extension is suspected, an MRI of the head is the preferred next step in assessment.

СТ

For assessment of orbital asymmetry, CT of the orbits with contrast is complementary to MRI [7]. In the setting of thyroid eye disease, CT provides useful information about orbital, muscle, and fat volumes and osseous anatomy, particularly when orbital decompression is a surgical consideration [7,32-34]. Orbital inflammatory conditions including thyroid eye disease, IgG4-related disease, and idiopathic orbital inflammatory disease may all present with unilateral or bilateral proptosis as a clinical manifestation. Like other orbital conditions, patients with these conditions may be imaged with CT or MRI, and these modalities provide overlapping information related to disease extent. Currently there is no consensus on the optimal imaging modality to assess patients presenting with idiopathic orbital inflammatory disease or IgG4-related orbital disease. CT of the head with contrast may also be added to assess the extent of intracranial disease, particularly if MRI is not available or contraindicated. Precontrast and postcontrast imaging is typically not necessary in evaluating these patients as the precontrast images do not add significant diagnostic information in this scenario.

CTA, MRA, Arteriography

Vascular structures in and around the orbit may be imaged with CTA, MRA, or catheter-based DSA. Similar to conventional CT and MRI, CTA and MRA may provide complementary information. CTA is performed following the injection of intravascular iodinated contrast and is typically imaged in the arterial phase. MRA can be performed without contrast using the time-of-flight technique or with contrast with an added benefit of producing time-resolved information.

Angiographic imaging is helpful in evaluating adults with a suspected vascular anomaly to define high- or lowflow vascular components, vascular supply, and drainage. This may be achieved with MRA, CTA, or DSA. MRA is the preferred method for evaluating these lesions because of the improved soft-tissue lesion characterization with this modality, superior anatomic localization, and the ability to perform time-resolved techniques. If the differential consideration is vascular mass versus malformation, flow characterization may be achieved with timeresolved MRA [35,36].

If a CCF is suspected, noninvasive vascular imaging with MRA and CTA are often indicated for diagnosis confirmation and pretreatment planning. When MRA or CTA is combined with anatomic MRI or CT, the secondary findings associated with CCF, including proptosis, vascular congestion within the orbit, extraocular muscle enlargement, and enlarged superior ophthalmic veins, can be easily identified. DSA is performed for more detailed assessment and intervention in patients with confirmed CCFs or in patients with a high index of suspicion for CCFs not confirmed on noninvasive imaging [37]. Although DSA is considered the gold standard for imaging

evaluation and treatment and may be appropriate in the acute assessment of acute visual loss related to suspected CCF, it is relatively invasive and carries its own procedural risks and is typically not performed as the initial test. In addition, DSA lacks in the ability to provide regional soft-tissue information seen with cross-sectional imaging that may assist in making the diagnosis. In a retrospective comparative analysis between CTA, MRA, and DSA, CTA was shown to be as useful as DSA for CCF detection in a cohort of 53 patients [20]. MRA was slightly less successful but still determined as good by demonstrating CCFs in approximately 80% of cases [20].

Radiography

Orbital or skull radiographs are insufficient to detect pathology in patients presenting with proptosis and have primarily been supplanted by CT.

Variant 3: Suspected orbital cellulitis, uveitis, or scleritis. Initial imaging.

Patients presenting with symptoms and signs of orbital cellulitis (postseptal cellulitis) are often referred for imaging to assess for complications including intraorbital abscess, intracranial involvement, and vascular compromise. The source of this infection is often from the adjacent paranasal sinuses and may be viral, bacterial, or fungal [38].

Idiopathic orbital inflammatory syndrome (IOIS). IgG4-related orbital disease. and other inflammatory/granulomatous processes are potential clinical and imaging mimics for orbital cellulitis. IOIS, previously known as orbital pseudotumor, may present with signs and symptoms that mimic infection and is a diagnosis of exclusion. IgG4-related orbital disease is a relatively recently described inflammatory condition that may account for a significant percentage of patients that have been previously described as idiopathic [39-41]. Manifestations include evelid or periocular swelling, lacrimal gland enlargement, extraocular muscle involvement, intraorbital mass, proptosis, and cranial nerve V involvement.

СТ

CT of the orbits with contrast is often the initial imaging modality in the emergent setting for suspected infection [2,7,42,43]. CT is superior to MRI for foreign body assessment, calcification detection, and osseous evaluation [43]. CT can be used in conjunction with the Chandler criteria to evaluate for the presence of bone erosion and subperiosteal abscess, which may require surgical intervention [38,44-46]. Imaging findings may show bone erosion on CT, opacification of a neighboring infected sinus, and/or intraorbital extension of inflammatory disease [47]. In patients who cannot receive contrast, a noncontrast orbit CT may still add useful information. Precontrast and postcontrast imaging is typically not necessary in evaluating these patients as the precontrast images do not add significant diagnostic information in this scenario.

Currently there is no consensus on the optimal imaging modality to assess patients presenting with IOIS or IgG4related orbital disease. Orbital CT and MRI are often complementary in their roles. Signs of inflammation may be detected with CT or MRI, which show intraconal or extraconal soft-tissue lesions that are diffuse or localized and commonly involve the orbital apex [43,48]. These findings may be initially seen on CT and subsequently further evaluated with MRI for improved soft-tissue characterization.

MRI

Orbital MRI is complementary to CT in evaluating intraorbital spread of infection. An MRI of the orbits without and with contrast should be considered if a more detailed assessment of intraorbital spread of infection is clinically warranted. In patients with suspected intracranial extension or complications, an MRI of the brain with high-resolution images to include the cavernous sinuses [2,42,43,46,47] provides greater soft-tissue resolution than CT. A high index of suspicion and low threshold for MRI is needed if invasive fungal infection is of concern in an immunocompromised patient [42,46,47] because of the morbidity of the disease. Although contrast is preferred, in patients who cannot receive contrast, a noncontrast orbital MRI may provide useful information.

Orbital MRI is complementary to orbital CT in evaluating patients for IOIS, IgG4-related orbital disease, or other inflammatory/granulomatous disease. Currently there is little evidence to support one modality's superiority to others in evaluating this patient population [39-41]. A hallmark of IOIS in its chronic form is fibrosis, which results in decreased signal on T2-weighted MRI sequences [48]. A small subset of patients with isolated ocular manifestation of IOIS had posterior scleritis, with inflammatory enhancement of the sclera on postcontrast imaging.

CTA, MRA, Arteriography

CTA or MRA may be added to routine CT or MRI scans if there is a suspicion for vascular invasion including cavernous sinus thrombosis, particularly in the setting of fungal disease. MRA may be performed without and/or with contrast. In the setting of cavernous sinus thrombosis, a contrast-enhanced MRA may provide additional information not provided by a traditional noncontrast MRA examination. There is a limited role for DSA in evaluating patients with orbital infection.

Radiography

Orbital radiographs are insufficient to detect orbital cellulitis. Radiographs have largely been supplanted by CT when imaging is necessary [49].

Variant 4: Suspected optic neuritis. Initial imaging.

Optic neuritis is defined as an acute inflammatory condition of the optic nerve, and can be unilateral or bilateral. It often presents with painful visual loss [50] but can also be painless. The primary differential consideration includes multiple sclerosis, neuromyelitis optica, neuromyelitis optica spectrum, or other infectious/granulomatous conditions. Although optic neuritis can be idiopathic, it is often seen as the initial manifestation of multiple sclerosis.

MRI

In patients presenting with a clinical suspicion for optic neuritis, both MRI of the orbits and MRI of the head without and with contrast are the primary imaging studies for initial assessment. This serves two primary purposes. The first purpose is to evaluate for abnormal enhancement and signal changes within the optic nerve, and the second is to evaluate the brain for associated intracranial demyelinating lesions, as the latter is a strong predictor of the subsequent development of multiple sclerosis [50-52]. MRI is incorporated into the revised McDonald criteria and MAGNIMS consensus guidelines [53] for diagnosing multiple sclerosis, which is characterized by establishing dissemination of lesions in space and time [51]. Neuromyelitis optica is a demyelinating condition that typically affects the optic nerves and spinal cord and is best assessed with MRI [54]. Serum and cerebrospinal fluid laboratory tests may also be useful in differentiating between these two entities [51,55].

СТ

Although an imaging test of the brain may be indicated prior to lumbar puncture in patients with optic disc edema to exclude a space occupying mass, CT imaging of the head is typically not indicated specifically for the evaluation of a patient with optic neuritis.

CTA, MRA, Arteriography

Angiography is not routinely used in the initial evaluation of optic neuritis.

Radiography

Orbital or skull radiographs are insufficient to detect pathology in patients presenting with clinical concern for vision loss.

Variant 5: Visual loss. Etiology identified on ophthalmologic examination or laboratory tests.

Excluding stroke and ischemic attack, transient visual loss can be due to a range of processes, including cataracts, glaucoma, retinal or choroidal detachments, vitreous or anterior segment hemorrhage, drusen, hypercoagulability syndromes, primary vasospasm, blepharospasm, and metabolic derangements such as those seen with glucose imbalance. These are most often diagnosed with dedicated ophthalmologic evaluation and laboratory results. A complete ophthalmologic evaluation is needed to diagnose these conditions and cross-sectional imaging is usually not necessary in cases where glaucoma, cataract, or macular degeneration are identified.

Please refer to the ACR Appropriateness Criteria[®] "Headache" [4] regarding evaluation in the setting of disc edema.

MRI

MRI is not routinely used in the evaluation of non-neoplastic ocular processes. In patients with glaucoma, a primary cause of irreversible blindness, there has been interest in applying advanced MRI techniques to earlier detection of this disease process; however, additional research is needed to validate the utility of these advanced techniques [56].

СТ

CT is not routinely used in the evaluation of nontraumatic, noninfectious, or non-neoplastic ocular processes.

CTA, MRA, Arteriography

CTA, MRA, and DSA are not first-line tests in this scenario.

Radiography

Orbital or skull radiographs are insufficient to detect pathology in patients presenting with vision loss.

Variant 6: Visual loss. Intraocular mass, optic nerve, or pre-chiasm symptoms. Initial imaging.

Monocular visual loss may be due to an intraocular mass, such as melanoma [57], or may involve the intraorbital, intracanalicular, or pre-chiasm segments of the optic nerve. This includes lesions intrinsic to the nerve, such as an optic nerve glioma, or extrinsic to the nerve resulting in mass effect, such as an optic nerve sheath meningioma. The differential diagnosis varies based on the age of the patient.

MRI

MRI provides excellent soft-tissue resolution of structures within the orbit, including the globe, muscles, tendons, nerves, and vascular structures. MRI of the orbits without and with contrast is the preferred modality in evaluating soft-tissue pathology within and around the orbit, particularly in mass characterization, optic nerve pathology, and assessing disease within the globe and orbit [1,7,58]. If contrast cannot be given, a noncontrast orbit MR may still provide useful information. If there is a significant intracranial component, additional MRI of the brain without and with contrast may be indicated to evaluate for intracranial spread of disease.

СТ

CT is superior to MRI for foreign body assessment, calcification detection, and osseous evaluation [43]. Orbital CT is complementary to MRI in evaluating patients with ocular, orbital, and skull base neoplasms. In patients presenting with clinical suspicion for an intraorbital mass lesion, orbital CT with contrast may be complementary to MRI in providing additional information about adjacent bone involvement, including bone erosion, sclerosis, or periosteal reaction that may not be readily seen with MRI [2,7].

If contrast cannot be given, a noncontrast orbit CT may still add useful information. CT imaging of the head with contrast may also be appropriate if more extensive skull or skull-base involvement is suspected. Precontrast and postcontrast orbital imaging is typically not necessary in evaluating these patients as the precontrast images do not add significant diagnostic information in this scenario.

CTA, MRA, Arteriography

CTA or MRA are complementary and may be added to routine CT or MRI scans if there is a suspicion for an intraorbital vascular lesion [35,36]. MRA without and with contrast may be preferred over CTA if time-resolved information is needed in lesion characterization. DSA is not a first-line test in this scenario.

Radiography

Orbital or skull radiographs are insufficient to detect pathology in patients presenting with monocular vision loss.

Variant 7: Nonischemic visual loss. Chiasm or post-chiasm symptoms. Initial imaging.

If a patient presents with a junctional scotoma or bitemporal hemianopia, a parasellar lesion is suspected, with mass effect on the optic chiasm affecting the crossing temporal fibers. Lesions may arise from the pituitary gland, hypothalamus, or adjacent dura, and accompanying endocrine abnormalities may also be present.

In patients presenting with a homonymous defect, a post-chiasm lesion involving the optic tracts, lateral geniculate nucleus, optic radiations, or primary visual cortex in the occipital lobe is suspected. It is important to remember that patients presenting with acute onset of visual loss with post-chiasm symptoms may be presenting with deficits related to an anterior or posterior circulation arterial stroke, intracranial hemorrhage, or venous sinus thrombosis. Please refer to the ACR Appropriateness Criteria[®] "Cerebrovascular Disease" [3] for imaging in this context.

Slowly progressive binocular visual defect findings suggest an intracranial or skull-base abnormality, including primary neoplasms and metastatic lesions. Mass effect from other intracranial pathology, including abscess, multiple sclerosis, or vascular lesions such as arteriovenous malformations and cerebral aneurysms, may also present with a similar visual field deficit.

MRI

Patients with a junctional scotoma or bitemporal visual defect are best assessed with an MRI of the brain without and with contrast, which includes the thin-slice profile needed to evaluate the pituitary gland and any suprasellar mass effect [43]. Detailed assessment of the optic chiasm and its relationship to an underlying mass are easily seen with an MRI of the brain without and with contrast. If contrast cannot be given, an MRI of the brain without contrast may be appropriate.

Patients presenting with a homonymous hemianopia or quadrantanopia defect are best assessed with an MRI of the brain without and with contrast [2,7,59-61]. Because the defect is most likely in a post-chiasm location, additional smaller field-of-view images of the orbit are typically not necessary.

СТ

For lesion characterization in and around the sella, CT of the head may be complementary to MRI and add additional information on the characteristics of the lesion, including the presence of calcification such as in a craniopharyngioma [7,59,62]. In patients with post-chiasm symptoms, an MRI of the brain is typically preferred over CT, particularly in a subacute, slowly progressive presentation. In the acute setting, a noncontrast head CT is reasonable for initial imaging. If a patient is unable or unwilling to have MRI, then a CT of the head without and with contrast may be an appropriate alternative.

CTA, MRA, Arteriography

It is important to remember that patients presenting with acute onset of visual loss with post-chiasm symptoms may be presenting with deficits related to an anterior or posterior circulation arterial stroke, intracranial hemorrhage, or venous sinus thrombosis. Please refer to the ACR Appropriateness Criteria[®] "<u>Cerebrovascular</u> <u>Disease</u>" [3] for imaging in this context. If a cerebral aneurysm or vascular malformation is identified on conventional diagnostic imaging, MRA, CTA, and/or DSA may provide additional information regarding aneurysm characterization as well as arterial supply, arteriovenous shunting, and venous drainage related to an arteriovenous malformation. If a vascular mass/malformation is a differential consideration, vascular flow characterization may be achieved with time-resolved MRA or DSA [35,36]. Please refer to the ACR Appropriateness Criteria[®] "<u>Cerebrovascular Disease</u>" [3] for imaging guidelines in this context.

If a mass such as a meningioma is identified in close proximity to the sagittal sinus, additional MRV or CTV imaging may be indicated to assess the integrity of the dural venous sinus. Postcontrast MRV and CTV are complementary in their utility in evaluating the dural venous sinuses. Noncontrast MRV may be performed in the event that contrast cannot be administered.

Radiography

Orbital or skull radiographs are insufficient to detect pathology in patients presenting with vision loss.

Variant 8: Ophthalmoplegia or diplopia. Initial imaging.

Ophthalmoplegia is paralysis of one or more extraocular muscles. This may be caused by impaired motility of the muscles, disrupted nerve conduction along the neuromuscular junction, or from denervation of the affected cranial nerve or brainstem nucleus. Ophthalmoplegia may also be related to granulomatous, inflammatory, neoplastic, and traumatic abnormalities that primarily affect the extraocular muscles. Traumatic orbital injury is covered in Variant 1 of this document.

A patient presenting with diplopia or disconjugate gaze may have an abnormality that involves the globe; the extraocular muscles; neuromuscular junction; cranial nerves III, IV and/or VI; or their respective fascicles, nuclei or connecting tracts within the brain stem. A broad differential including developmental, neoplastic, granulomatous, infectious, inflammatory, demyelinating, vascular, and traumatic causes can be considered in patients with diplopia. This broad differential can be narrowed when one considers the age of the patient, the onset of symptoms, and the presence of associated findings. The pattern of involvement can usually lead to the anatomical localization of the offending lesion. It is important to remember that patients presenting with acute onset of diplopia may be presenting with deficits related to a posterior circulation stroke. Please refer to the ACR Appropriateness Criteria[®] "<u>Cerebrovascular Disease</u>" [3] for imaging in this context. In the setting of intracranial traumatic injury, please refer to the ACR Appropriateness Criteria[®] "<u>Head Trauma</u>" [16].

Patients with isolated cranial nerve III palsies can be divided into pupil-involving or pupil-sparing, suggesting vascular compression versus vasculopathic etiologies, respectively. Isolated cranial nerve IV palsies are most often caused by trauma [63] and rarely nerve sheath tumors. Isolated cranial nerve VI palsies may be caused by

lesions within the prepontine cistern, skull base, cavernous sinus, or sella. Isolated cranial nerve VI palsies may also be seen in the setting of increased intracranial pressure without direct compression of the nerve [64]. Multiple ipsilateral cranial nerve palsies that affect cranial nerves III, IV, and VI suggest a lesion at the cavernous sinus or orbital apex [65] and can occur with pathology in the basilar subarachnoid space, as seen in infectious meningitis (TB, fungal, Lyme disease) or noninfectious causes (sarcoid, neoplasm, perineural, or leptomeningeal tumor spread). In patients with internuclear ophthalmoplegia, a brain-stem lesion affecting the medial longitudinal fasciculus should be suspected. A demyelinating plaque in the setting of multiple sclerosis is a primary consideration in younger patients and stroke in older patients presenting with an acute internuclear ophthalmoplegia [66]. Other likely considerations include tumor, hemorrhage, and infection [66].

MRI

MRI of the orbits without and with contrast is preferred [67] if ophthalmoplegia is felt to be related to a primary disease process within the orbit affecting the extraocular muscles or if there is history of trauma, enophthalmos, proptosis, orbital inflammation, or chemosis. An MRI of the orbits with the globes imaged during different gaze positions may aid in identifying a potential muscular slip or pulley abnormality [68].

If the disease process is felt to involve the brain stem, brain, or cisternal segments of the cranial nerves, an MRI of the head without and with contrast including additional small field-of-view high-resolution T2-weighted images of the cranial nerves is the preferred imaging modality to evaluate for an underlying abnormality of the brain, brain stem, and cranial nerves [1,2,43]. This dedicated MRI of the cranial nerves primarily focuses on the nuclear, cisternal, and skull-base cranial nerve segments and can be centered upon cranial nerves III–IV, including the cavernous sinuses. For example, patients with isolated pupil-sparing third-nerve palsies, which primarily involve the oculomotor fibers, vasculopathic considerations are the primary differential consideration and are best evaluated with an MRI examination of the head with special attention to the cranial nerves.

СТ

In patients with ophthalmoplegia or diplopia with associated secondary signs of proptosis, orbital inflammation, or trauma, a dedicated orbit CT is typically indicated to evaluate the extraocular muscles. Contrast is often indicated in the setting of orbital inflammation assessment but not indicated in the acute traumatic setting, as specified in Variant 1. CT is superior to MRI for foreign body assessment, calcification detection, and osseous evaluation [43]. Although CT imaging of the orbits is preferred, CT imaging of the head may be appropriate if an intracranial abnormality is suspected. Precontrast and postcontrast imaging of the orbits is typically not necessary in evaluating these patients as the precontrast images do not add significant diagnostic information in this scenario.

CTA, MRA, Arteriography

Isolated, pupil-involving third-nerve palsy suggests external compression of the parasympathetic nerves that surround the oculomotor fibers in the third-nerve fascicles. As the primary consideration is vascular compression from an adjacent aneurysm, vascular imaging either with CTA or MRA is indicated [69]. This assessment is not performed in isolation but rather as a complement to anatomic cross-sectional imaging. Please refer to the ACR Appropriateness Criteria[®] "Cerebrovascular Disease" [3] for imaging in this context. There is a limited role for DSA in the initial evaluation of patients with diplopia. However, if an aneurysm is detected in cross-sectional evaluation, DSA may be indicated for further assessment and treatment.

Radiography

Orbital or skull radiographs are insufficient to detect pathology in patients presenting with proptosis and have primarily been supplanted by CT.

Summary of Recommendations

- Orbital trauma is best assessed with a noncontrast orbit CT and/or noncontrast CT of the head, which are often complementary.
- Orbital asymmetry, exophthalmos, or enophthalmos can be evaluated with contrast-enhanced orbit CT or contrast-enhanced orbit MRI, which are often complementary.
- Contrast-enhanced CT or contrast-enhanced MRI are both appropriate in evaluating orbital cellulitis, uveitis, or scleritis, with CT often performed first during the initial assessment.
- Optic neuritis is best assessed with a contrast-enhanced MRI of the orbits and contrast-enhanced MRI of the head, which are often performed in conjunction with one another.

- There is typically no role for imaging in patients with visual loss due to abnormalities such as cataracts, macular degeneration, or glaucoma.
- Evaluation of visual loss localized to the orbit or disease process involving the pre-chiasmatic optic nerve is best assessed with targeted contrast-enhanced MRI of the orbits or contrast-enhanced CT of the orbits, which are complementary.
- Evaluation of visual loss involving the chiasm or post-chiasm is best assessed with a contrast-enhanced MRI of the brain. Although contrast is preferred, an MRI of the brain without contrast may also be appropriate if contrast cannot be given.
- Diplopia or opthalmoplegia can be evaluated with contrast-enhanced MRI of the head, contrast-enhanced MRI of the orbits, contrast-enhanced CT of the orbits, or noncontrast MRI of the orbits, which are complementary in their roles. Whether to focus the assessment on the orbits and/or head will depend on suspected anatomic localization and differential diagnosis related to the patient's specific clinical presentation.

Summary of Evidence

Of the 70 references cited in the *ACR Appropriateness Criteria*[®] *Orbits, Vision and Visual Loss* document, 5 are categorized as therapeutic references. Additionally, 64 references are categorized as diagnostic references including 1 well-designed study, 3 good-quality studies, and 16 quality studies that may have design limitations. There are 49 references that may not be useful as primary evidence. There is 1 reference that is a meta-analysis study.

The 70 references cited in the ACR Appropriateness Criteria[®] Orbits, Vision and Visual Loss document were published from 1965 to 2017.

Although there are references that report on studies with design limitations, 4 well-designed or good-quality studies provide good evidence.

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.

Appropriateness Category Names and Definitions

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 [70].

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 (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 <u>www.acr.org/ac</u>.

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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.