American College of Radiology
ACR Appropriateness Criteria®
Cranial Neuropathy

Variant 1: **Anosmia and abnormalities of the sense of smell. (Olfactory nerve, CN I.)**

Radiologic Procedure	Rating	Comments	RRL*
MRI head without and with IV contrast	8	This procedure is performed in conjunction with MRI of the orbit, face, and neck.	O
MRI orbit face neck without and with IV contrast	8	This procedure is performed in conjunction with MRI of the head.	O
MRI head without IV contrast	6		O
MRI orbit face neck without IV contrast	6		O
CT maxillofacial with IV contrast	6		☢☢
CT head with IV contrast	5		☢☢☢
CT head without IV contrast	5		☢☢☢
CT head without and with IV contrast	5		☢☢☢
CT maxillofacial without IV contrast	5		☢☢
CT maxillofacial without and with IV contrast	4		☢☢☢
FDG-PET/CT whole body	2		☢☢☢☢
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

Variant 2: **Weakness or paralysis of the mastication muscles. Sensory abnormalities of the head and neck. Trigeminal neuralgia. (Trigeminal nerve, CN V.)**

Radiologic Procedure	Rating	Comments	RRL*
MRI head without and with IV contrast	8	This procedure is performed in conjunction with MRI of the orbit, face, and neck.	O
MRI orbit face neck without and with IV contrast	8	This procedure is performed in conjunction with MRI of the head.	O
MRI head without IV contrast	6		O
MRI orbit face neck without IV contrast	6		O
MRA head without IV contrast	6		O
CTA head with IV contrast	5		☢☢☢
CT head with IV contrast	5		☢☢☢
CT maxillofacial with IV contrast	5		☢☢
CT maxillofacial without IV contrast	5		☢☢
CT head without IV contrast	4		☢☢☢
CT head without and with IV contrast	4		☢☢☢
CT neck with IV contrast	4		☢☢☢
CT neck without IV contrast	4	Contrast-enhanced imaging is preferred.	☢☢☢
CT maxillofacial without and with IV contrast	4		☢☢☢
CT neck without and with IV contrast	3		☢☢☢
FDG-PET/CT whole body	2		☢☢☢☢
US neck	2		O
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

Variant 3: **Weakness or paralysis of facial expression. Hemifacial spasm. Bell palsy. (Facial nerve, CN VII.)**

Radiologic Procedure	Rating	Comments	RRL*
MRI orbit face neck without and with IV contrast	9	This procedure is performed in conjunction with MRI of the head.	O
MRI head without and with IV contrast	8	This procedure is performed in conjunction with MRI of the orbit, face, and neck.	O
MRI head without IV contrast	5		O
MRI orbit face neck without IV contrast	5		O
CT head with IV contrast	5		☢☢☢
CT head without IV contrast	5		☢☢☢
CT head without and with IV contrast	4		☢☢☢
CT neck with IV contrast	4		☢☢☢
CT neck without IV contrast	3		☢☢☢
CT neck without and with IV contrast	3		☢☢☢
FDG-PET/CT whole body	2		☢☢☢☢
US neck	2		O
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

Variant 4: **Palate weakness. Oropharyngeal pain. (Glossopharyngeal nerve, CN IX.)**

Radiologic Procedure	Rating	Comments	RRL*
MRI head without and with IV contrast	8	This procedure is performed in conjunction with MRI of the orbit, face, and neck.	O
MRI orbit face neck without and with IV contrast	8	This procedure is performed in conjunction with MRI of the head.	O
MRI head without IV contrast	6		O
MRI orbit face neck without IV contrast	6		O
CT neck with IV contrast	6		☢☢☢
CT head with IV contrast	5		☢☢☢
CT head without IV contrast	5	Contrast-enhanced imaging is preferred.	☢☢☢
CT neck without IV contrast	5	Contrast-enhanced imaging is preferred.	☢☢☢
CT head without and with IV contrast	4		☢☢☢
CT neck without and with IV contrast	4		☢☢☢
FDG-PET/CT whole body	2		☢☢☢☢
US neck	2		O

Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate** ***Relative Radiation Level**

Variant 5: **Vocal cord paralysis. (Vagal nerve, CN X.)**

Radiologic Procedure	Rating	Comments	RRL*
MRI orbit face neck without and with IV contrast	8	This procedure is performed in conjunction with MRI of the head. CT of the neck is an alternative examination and in some instances can be complementary.	O
CT neck with IV contrast	8	MRI of the orbit, face, and neck can be an alternative examination and in some instances can be complementary.	☢☢☢
MRI head without and with IV contrast	7	This procedure is performed in conjunction with MRI of the orbit, face, and neck. CT of the neck can be useful to assess the extracranial course of CN X.	O
MRI head without IV contrast	6		O
MRI orbit face neck without IV contrast	6		O
CT chest with IV contrast	6		☢☢☢
MRI chest without and with IV contrast	5		O
CT head with IV contrast	5		☢☢☢
CT neck without IV contrast	5	Contrast-enhanced imaging is preferred.	☢☢☢
CT chest without IV contrast	5		☢☢☢
MRI chest without IV contrast	4		O
CT head without IV contrast	4		☢☢☢
CT head without and with IV contrast	4		☢☢☢
CT neck without and with IV contrast	4		☢☢☢
X-ray chest	4		☢
FDG-PET/CT whole body	4	This procedure is not a first-line examination.	☢☢☢☢
US neck	4		O
CT chest without and with IV contrast	3		☢☢☢
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

Variant 6: **Weakness or paralysis of the sternocleidomastoid and trapezius muscles. (Accessory nerve, CN XI.)**

Radiologic Procedure	Rating	Comments	RRL*
MRI head without and with IV contrast	8	This procedure is performed in conjunction with MRI of the orbit, face, and neck.	O
MRI orbit face neck without and with IV contrast	8	This procedure is performed in conjunction with MRI of the head. CT neck imaging can be complementary.	O
MRI orbit face neck without IV contrast	7	Contrast-enhanced imaging is preferred.	O
CT neck with IV contrast	7	MRI of the orbit, face, and neck can be an alternative examination and in some instances can be complementary.	☢☢☢
MRI head without IV contrast	6		O
CT head with IV contrast	6		☢☢☢
CT head without IV contrast	5		☢☢☢
CT head without and with IV contrast	5		☢☢☢
CT neck without IV contrast	5		☢☢☢
CT neck without and with IV contrast	5	The panel did not agree on a recommendation.	☢☢☢
FDG-PET/CT whole body	3		☢☢☢☢
US neck	2		O
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

Variant 7: **Weakness or paralysis of the tongue. (Hypoglossal nerve, CN XII.)**

Radiologic Procedure	Rating	Comments	RRL*
MRI head without and with IV contrast	8	This procedure is performed in conjunction with MRI of the orbit, face, and neck.	O
MRI orbit face neck without and with IV contrast	8	This procedure is performed in conjunction with MRI of the head.	O
CT neck with IV contrast	7	MRI of the orbit, face, and neck is preferred, but in some instances CT neck can be complementary.	☢☢☢
MRI head without IV contrast	6		O
MRI orbit face neck without IV contrast	6		O
CT head with IV contrast	5		☢☢☢
CT head without IV contrast	5		☢☢☢
CT head without and with IV contrast	4		☢☢☢
CT neck without IV contrast	4		☢☢☢
CT neck without and with IV contrast	4		☢☢☢
FDG-PET/CT whole body	2		☢☢☢☢
US neck	2		O
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

Variant 8: **Perineural spread of tumor. (Most commonly trigeminal nerve [CN V], facial nerve [CN VII].)**

Radiologic Procedure	Rating	Comments	RRL*
MRI head without and with IV contrast	9	This procedure is performed in conjunction with MRI of the orbit, face, and neck.	O
MRI orbit face neck without and with IV contrast	9	This procedure is performed in conjunction with MRI of the head.	O
MRI orbit face neck without IV contrast	7	Addition of contrast-enhanced imaging is preferred.	O
CT neck with IV contrast	6		☢☢☢
MRI head without IV contrast	5		O
CT head with IV contrast	5		☢☢☢
CT head without IV contrast	5		☢☢☢
CT neck without IV contrast	5		☢☢☢
CT head without and with IV contrast	4		☢☢☢
CT neck without and with IV contrast	4		☢☢☢
FDG-PET/CT whole body	4	This procedure is not a first-line examination.	☢☢☢☢
US neck	2		O
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			***Relative Radiation Level**

CRANIAL NEUROPATHY

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Summary of Literature Review

Introduction/Background

The cranial nerves arise from nuclei within the brain and brainstem and supply sensory and motor innervation to the head and neck region, whereas the spinal nerves arise from the spinal cord and supply the rest of the body. As a group, the cranial nerves have both sensory and motor components similar to those of the spinal nerves. Individually the cranial nerves may be purely sensory or purely motor or a mixture of both sensory and motor. Functions of the cranial nerves may be divided into 3 sensory and 3 motor categories. The sensory group includes visceral sensory, which supplies sensory input from the internal organs; general sensory, which supplies tactile, pain, temperature, and other sensations; and special sensory, which includes the special senses of smell, vision, taste, hearing, and balance. Of the 3 motor functions, somatic motor innervates muscles that develop from the body somites; branchial motor innervates muscles derived from the branchial arches; and visceral motor innervates the viscera, glands, and smooth muscle [1-3].

Cranial nerves emerge in an orderly fashion from the rostral portion of the embryologically developing neural tube, which will subsequently mature to form the brain and brainstem. Anatomically, the 12 pairs of cranial nerves are designated by numbers and are organized most rostral to most caudal in descending order. The cranial nerves include the olfactory (cranial nerve [CN] I), optic (CN II), oculomotor (CN III), trochlear (CN IV), trigeminal (CN V), abducens (CN VI), facial (CN VII), vestibulocochlear (CN VIII), glossopharyngeal (CN IX), vagus (CN X), spinal accessory (CN XI), and hypoglossal (CN XII) nerves. The olfactory (CN I) and optic (CN II) nerves are actually tracts formed from the telencephalon and diencephalon, respectively, and are not considered true nerves [1]. The optic (CN II), oculomotor (CN III), trochlear (CN IV), and abducens (CN VI) nerves are considered functionally to be part of the visual and extraocular motor system and have been discussed in the ACR Appropriateness Criteria® "Orbits, Vision and Visual Loss" [4]. Also, the vestibulocochlear nerve (CN VIII) has been reviewed in the ACR Appropriateness Criteria® "Hearing Loss and/or Vertigo" [5]. Therefore, this discussion will focus on CN I, CN V, CN VII, CN IX, CN X, CN XI, and CN XII.

In approaching cranial neuropathy, several concepts should be emphasized:

1. Because of the complex anatomic structures within the brain and brainstem and because the cranial nerves may take long, circuitous routes to their destinations, a detailed knowledge of cranial nerve anatomy is essential for proper clinical localization of potential lesions and for appropriate application of specific imaging protocols.

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2. Because some individual nerve fibers, such as the autonomic nerves, may travel with several different cranial nerves from their nuclei of origin to their ultimate destinations, loss of a specific function may indicate involvement of potentially more than 1 cranial nerve.
3. Because of the close proximity of many cranial nerve nuclei and of many exiting sites of the nerves themselves, some mass lesions may involve multiple cranial nerves.

Special Imaging Considerations

In the evaluation of cranial neuropathy complete evaluation of the nerves from their brainstem nuclei to their ‘‘end organs’’ must be performed. The pathology can be located in the nucleus, cisternae, and skull base segments of the cranial nerves. For cranial nerve VII, the lesion can also be located in the parotid. For cranial nerves IX, X, XI, and XII, the lesion can also be located in the neck. Evaluation of the upper chest is necessary for complete evaluation of the CN X (recurrent laryngeal) course. This can be accomplished by extending the neck scan into the mid thorax (aortic pulmonary window) or dedicated chest computed tomography (CT). Patients presenting with otalgia may require evaluation of CN V, VII, IX, and X and upper cervical nerves C2 and C3 since any of these nerves may be the source for the otalgia [6]. The use of intravenous contrast is imperative for the evaluation of cranial neuropathy with magnetic resonance imaging (MRI). Neck CT also requires the utilization of contrast when evaluating pathology affecting the neck. Dual-phase CT before and after administration of contrast is rarely necessary. This should be avoided because of the extra radiation exposure and minimal added benefits.

The primary plane of study for head and neck evaluation of cranial neuropathy is usually the axial plane. Additional orthogonal planes are required depending upon the course of the various nerves. Coronal and sagittal reconstructions are typically performed on CT. In order to obtain high-resolution orthogonal reconstructions, the axial plane is acquired with thin sections, typically <1 mm. On MRI, orthogonal reconstructions are typically performed on the postcontrast T1-weighted images; however, they can also be obtained on T2-weighted images. Thin-section MRI images are required to evaluate the cisternal segment and should be performed.

High-field-strength magnets (1.5T–3.0T) are preferred to low-field-strength units because of achievable signal to noise ratios, gradient strength, and spatial resolution [7]. A phased-array head coil suffices for most examinations; specialized surface coils may supplement examinations of peripherally located nerves.

Fundamental techniques include T1-weighted, T2-weighted, and enhanced T1-weighted imaging sequences. The unenhanced T1-weighted sequence remains an excellent baseline technique for anatomical evaluation because of the natural contrast provided by neck and skull base fat. Specialized versions of sequences may be available on scanners depending on manufacturer options. For example, various 3-D and heavily T2-weighted sequences—such as constructive interference in steady state, 3-D–balanced fast field echo, 3-D–driven equilibrium radio frequency reset pulse, 3-D fast spin echo, fast imaging using steady-state acquisition, and 3-D fast spin-echo extended echo-train acquisition — may provide excellent spatial resolution of the cisternal segments of some of the cranial nerves, but they must be used judiciously because of potentially misleading artifacts [7-17]. Enhanced fat-suppression T1-weighted techniques may emphasize abnormally enhancing lesions and nerves but may potentially mask subtle pathology if the suppression is nonuniform. Additional sequences, such as diffusion-weighted imaging, may be added to evaluate specific pathologies, such as infarctions, or specific lesions, such as epidermoids, that may affect cranial nerve function. Slice thickness should be calculated for optimal spatial resolution without introducing partial-volume effect. Because cranial nerve examinations tend to be lengthy, strategies such as parallel imaging may improve patient compliance and image quality [7].

Discussion of Procedures by Variant

Variant 1: Anosmia and abnormalities of the sense of smell. (Olfactory nerve, CN I.)

Abnormalities of the special sense of smell are mediated by the olfactory nerve (CN I) and can be grouped into clinical categories. Quantitative disturbances imply diminished or enhanced sense of smell (anosmia, hyposmia, or hyperosmia). Qualitative disturbances involve distortions of the sense of smell (dysosmia). Discrimination disturbances involve an inability to differentiate among various smells. Hallucinations or delusions in the sense of smell may also occur. The latter may be caused by temporal lobe dysfunction (see the ACR Appropriateness Criteria® “Seizures and Epilepsy” [18]) or by degenerative or psychiatric disease. Taste, mediated by the facial (CN VII) and glossopharyngeal (CN IX) nerves, may also be affected by pathology involving the olfactory nerve (CN I).

Most patients with olfactory complaints do not require imaging. Chronic tobacco use, upper respiratory infections, and inflammatory conditions most commonly affect the sense of smell [19]. More serious conditions affecting the olfactory nerve include trauma (the olfactory nerve is the nerve most commonly disrupted by trauma); cribriform plate tumors such as invasive squamous cell carcinomas of the paranasal sinuses, meningiomas, and esthesioneuroblastomas; inflammatory lesions such as sarcoidosis and granulomatosis with polyangiitis (formerly known as Wegener granulomatosis); and congenital conditions such as cephaloceles and Kallmann syndrome [8,20-22]. Recent investigations have focused on olfactory bulb volume as an indicator of olfactory dysfunction and even a marker for such disorders as early Parkinson disease and depression [20,23-26].

Magnetic resonance imaging and computed tomography
MRI is the mainstay for examining the olfactory apparatus, although CT remains useful when evaluating fractures, paranasal sinus inflammatory disease, and bony anatomy [20,27,28]. Imaging protocols should cover the major anatomic divisions of the olfactory nerve and pathway, including the olfactory epithelium, which is located in the upper nasal cavity; the olfactory neurons and bulbs, located in the cribriform plate and inferior frontal lobes; and the olfactory pathways, which travel in portions of the temporal and frontal lobes [7].

FDG-PET/CT and other imaging modalities
Efforts using functional MRI, single-photon emission CT, and fluorine-18-2-fluoro-2-deoxy-D-glucose positron emission tomography (FDG-PET) in studying olfactory dysfunction remain largely investigative and are not generally used in routine evaluations [29-33].

Variant 2: Weakness or paralysis of the mastication muscles. Sensory abnormalities of the head and neck. Trigeminal neuralgia. (Trigeminal nerve, CN V.)

The trigeminal nerve (CN V) provides general sensation to large portions of the head and neck and branchial motor innervation to the muscles of mastication [3]. It is the largest cranial nerve and is divided into 3 main divisions, known as the ophthalmic (V1) , maxillary (V2), and mandibular (V3) branches [8]. Symptoms largely depend on the involved segment and division [34]. Abnormalities of the nerve may manifest as sensory disturbances, such as trigeminal neuralgia (*tic douloureux*), facial numbness, or motor abnormalities such as weakness when chewing food.

The trigeminal nerve (CN V) is the nerve of the first branchial arch and may be involved in congenital conditions such as Goldenhar-Gorlin syndrome [34]. Intra-axial and extra-axial processes may affect the brainstem trigeminal nuclei and nerve root entry and exit zones. Conditions localized to the brainstem portion of the trigeminal nerve (CN V) include vascular lesions (such as compressing vascular loops, aneurysms, vertebrobasilar dolichoectasia, and infarctions), inflammatory and infectious conditions (such as meningitis, encephalitis, sarcoidosis, and multiple sclerosis), and tumors (such as gliomas, lymphomas, metastases, and meningiomas) [35-38]. The cisternal portion of the nerve may be especially vulnerable to compression from adjacent vascular loops, causing trigeminal neuralgia [38,39]. Tumors, vascular lesions, and inflammatory processes may also affect the branches of the nerve as they traverse the Meckel cave, the pterygopalatine fossa, the orbit, the skull base, and the masticator space [34,40].

Magnetic resonance imaging and computed tomography
MRI is the preferred modality for investigating the trigeminal nerve (CN V) [7,34]. CT is very useful for evaluating the skull base and neural foramina. Three-dimensional and heavily T2-weighted magnetic resonance (MR) sequences and MR and CT angiography are helpful noninvasive methods for reviewing the anatomy of potentially compressing vascular loops [41-56]. Patients may benefit from MRI studies performed in a high-strength magnet (3T), given the higher anatomic resolution [57,58]. With the growing popularity of radiosurgery, such as gamma knife procedures, and radiofrequency thermocoagulation in the treatment of trigeminal neuralgia, both CT and MRI have become indispensable planning and follow-up tools, although imaging may not reliably predict outcome [35,59-73]. Because of the complex branching patterns of the nerve, multiple imaging planes are essential [34].

Ultrasound
US is not routinely used in the initial evaluation of the trigeminal nerve.

FDG-PET/CT
PET is not routinely used in the initial evaluation of the trigeminal nerve.

Advanced imaging modalities
Advanced imaging applications, such as fractional anisotropy derived from diffusion tensor imaging and virtual endoscopy, are promising future directions in investigating trigeminal neuralgia [74-76].

Variant 3: Weakness or paralysis of facial expression. Hemifacial spasm. Bell palsy. (Facial nerve, CN VII.)

The facial nerve (CN VII) is one of the most complex cranial nerves and contains branchial motor (innervation to the muscles of facial expression), visceral motor (parasympathetic innervation to most of the glands of the head), general sensory (surface innervations to a small portion of the external ear and tympanic membrane), and special sensory (taste to the anterior two-thirds of the tongue) functions [3]. It is the one of the most commonly paralyzed nerves in the body, and most of the clinical attention it receives focuses on its role in facial expression [8]. Tinnitus, conductive and sensorineural hearing loss, and hemifacial spasm may also signal a lesion involving the facial nerve [15].

The intracranial course of the facial nerve includes pontine, cisternal, and intratemporal segments [7]. Within the pons, the facial nuclei can be affected by intra-axial conditions such as infarction, vascular malformations, tumors, and multiple sclerosis [77]. As the nerve exits the brainstem and courses through the temporal bone, it may be affected by facial and vestibular schwannomas, meningiomas, vascular lesions, inflammation, cholesteatomas, paragangliomas, trauma, and intrinsic bone tumors [15,78]. The extracranial segment of the facial nerve courses through the parotid gland and may be affected by parotid tumors and inflammation and conditions of the neighboring anatomic spaces and skull base such as carcinomas, sarcomas, trauma, and inflammatory disease.

Magnetic resonance imaging
MRI is the mainstay of evaluating both intracranial and extracranial portions of the facial nerve [15,79-83]. Facial paralysis in the form of Bell palsy is one of the most common syndromes confronting the otolaryngologist. In general, Bell palsy patients need not be imaged unless the symptoms are atypical or persist for >2 months [15]. When imaging is considered, MRI is the method of choice [15,82,84]. Enhancement may be seen in the canalicular, labyrinthine, geniculate, tympanic, and mastoid portions of the nerve in neuritis, although geniculate, tympanic, and mastoid portions may enhance normally [15,85-89]. MRI may also be useful in establishing prognosis [88,90-94]; however, there is 1 current study with a small cohort of patients that shows no association between the degree of enhancement and the clinical severity of facial nerve palsy in the early stage, stating that predicting the prognosis is difficult [95].

Computed tomography
CT provides useful information regarding temporal bone fractures and trauma, presurgical osseous anatomy, nerve involvement with inflammatory middle ear disease, foraminal expansion, patterns of bone erosion, and intrinsic bone tumor matrices [83,84,96-98]. In patients with risk of contrast allergy and contrast-induced nephropathy, a noncontrast CT may be sufficient if patients cannot undergo MRI. A dedicated temporal bone CT with thin sections should be obtained instead of a head CT to evaluate the course of CN VII.

Ultrasound
US is not routinely used in the initial evaluation of the facial nerve.

FDG-PET/CT
PET is not routinely used in the initial evaluation of the facial nerve.

Variant 4: Palate weakness. Oropharyngeal pain. (Glossopharyngeal nerve, CN IX.)

The glossopharyngeal nerve (CN IX) arises in the medulla and is responsible for branchial motor innervation to the stylopharyngeus muscle, which elevates the palate, and visceral motor parasympathetic innervation to the parotid gland [3]. Visceral sensory innervation to the carotid sinus plays a role in regulating circulation and general and special sensory functions that supply sensation and taste to the posterior tongue. The nerve exits the jugular foramen in close proximity to the vagus (CN X) and the spinal accessory (CN XI) nerves [3,7,77,99]. Therefore, isolated syndromes involving the glossopharyngeal nerve are rare. Intra-axial lesions include gliomas, lymphomas, metastases, vascular malformations, infarctions, and inflammatory abnormalities. Multiple sclerosis may also affect the medulla adjacent to the cranial nerve nuclei. Leptomeningeal metastases, granulomatous disease, and even tortuous or aneurysmal dilatation of vessels may affect the nerve as it enters the cistern. Lesions in the region of the posterior skull base and jugular foramen, such as metastases, schwannomas, paragangliomas, and meningiomas, usually also involve the other lower cranial nerves [99]. Tonsillar pain syndromes, palate

weakness, and loss of gag reflex accompanied by loss of taste and sensation in the posterior pharynx may signal a glossopharyngeal nerve lesion [99].

Magnetic resonance imaging and computed tomography
As with the other cranial nerves, MRI of CN IX is the preferred modality for investigating possible lesions such as masses or vascular compression, with CT providing information on the bony integrity of the foramina [7,100-103]. Imaging protocols should focus on the posterior skull base and upper neck.

Ultrasound
US is not routinely used in the initial evaluation of the glossopharyngeal nerve.

FDG-PET/CT
PET is not routinely used in the initial evaluation of the glossopharyngeal nerve.

Variant 5: Vocal cord paralysis. (Vagal nerve, CN X.)

The vagus nerve (CN X) supplies visceral sensation to the pharynx, larynx, and viscera and general sensation to the ear. Branchial motor branches innervate muscles of the pharynx and larynx, whereas visceral motor branches play a predominant role in parasympathetic supply to the thorax and abdomen [3,7,99]. The vagus nerve boasts the longest course in the body of any cranial nerve and is therefore vulnerable to a wide range of pathologies occurring throughout its trajectory from the posterior fossa and skull base to the neck, thorax, and abdomen [8]. Intracranial processes such as meningiomas, schwannomas, metastases, granulomatous disease, ischemia, vascular conditions, and infection may affect the vagal nuclei and the nerve as it exits the medulla. Paragangliomas, schwannomas, and metastases involving the skull base may affect the nerve and the neighboring glossopharyngeal nerve (CN IX) by infiltration of fibers or by compression. Within the neck, trauma may also affect the vagus nerve, in addition to masses, vascular lesions, thyroid conditions, infection, or inflammation [104]. Viral neuropathy may be one of the most common causes of idiopathic vagal palsies [104].

One of the most troubling symptoms of vagus dysfunction is vocal cord paralysis. Because lesions anywhere in the long course of the nerve may potentially cause paralysis, the imaging protocol must visualize the full extent of the nerve from the skull base to the mid chest [99].

Magnetic resonance imaging, computed tomography, and radiography
With its rapid scanning time and availability, CT provides an excellent means of examining the lower course of the nerve [105]. Moreover, thoracic causes of paralysis, such as lung cancer, tuberculosis, and thoracic aortic aneurysm, are common [106]. Although chest radiographs may detect many of these causes, chest CT is more sensitive, especially for lesions concealed in the aortopulmonary window [106,107]. This can also be accomplished by extending the neck CT scanning to the mid thorax. For imaging of the upper course of the nerve including the skull base, MRI is preferred [103,108,109]. For the mid neck and larynx, CT and MRI complement one another [110,111]. For example, CT may differentiate traumatic arytenoid dislocation from neurogenic paralysis [112]. Rapid multislice CT scanning, including functional 3-D applications, also allows the patient to perform phonation and breathing maneuvers during imaging to augment diagnosis [113-117]. In patients with risk of contrast allergy and contrast-induced nephropathy, a noncontrast CT may be sufficient if patients cannot undergo MRI.

Ultrasound
US may also have a role in imaging of the neck [104,105]. It may be useful for assessing lesions such as tumors or lymphadenopathy that have caused CN X neuropathy. It can be utilized in cases of neck lesions as a problem-solving technique. It can also be used as a technique to guide biopsies of lesions in the neck.

FDG-PET/CT
PET imaging used for evaluating head and neck malignancy may yield false-positive findings in the larynx for patients with vocal cord paralysis or unrecognized physiological asymmetry [118,119]. It may be useful as a problem-solving technique following initial cross-sectional imaging in patients with a known primary malignancy. PET/CT may also be superior to cross-sectional imaging for both localization and determination of response to therapy [120-124].

Radiographs
Chest radiographs can be utilized as a screening tool if chest CT or chest MRI is unavailable or contraindicated. It can reveal lesions in the lung apex or mediastinum that may cause CN X deficits.

Variant 6: Weakness or paralysis of the sternocleidomastoid and trapezius muscles. (Accessory nerve, CN XI.)

The spinal accessory nerve (CN XI) supplies the sternocleidomastoid muscle and the upper portion of the trapezius muscle as its sole branchial motor function [3,7,99]. Palsy is clinically manifested by weakness and atrophy of these muscles and may be accompanied by evidence of involvement of the glossopharyngeal (CN IX) and vagus (CN X) nerves in combined syndromes [99]. Loss of volume and fatty infiltration of the sternocleidomastoid and trapezius muscles may be noted on imaging.

Magnetic resonance imaging and computed tomography

CT and MRI are complementary in diagnosing conditions such as posterior fossa and skull base infarctions, vascular lesions, Chiari malformations, paragangliomas, schwannomas, meningiomas, and metastases or in recognizing nerve involvement from prior neck surgeries [99,103,104]. In patients with risk of contrast allergy and contrast-induced nephropathy, a noncontrast CT may be sufficient if patients cannot undergo MRI. Protocol with thin-section MRI should be performed to evaluate the cranial nerves.

Ultrasound

US is not routinely used in the initial evaluation of the accessory nerve.

FDG-PET/CT

PET is not routinely used in the initial evaluation of the accessory nerve.

Variant 7: Weakness or paralysis of the tongue. (Hypoglossal nerve, CN XII.)

The hypoglossal nerve (CN XII) supplies somatic motor innervation to the intrinsic and extrinsic muscles of the tongue, except the palatoglossus muscle [3,7,99]. Palsy of this nerve is recognized by dysarthria and deviation of the tongue to the affected side on protrusion. Atrophy and fatty infiltration of the tongue may be noted on imaging. Lesions of the posterior fossa, skull base, upper neck, and floor of the mouth may affect the hypoglossal nerve. They include infarctions, meningiomas, schwannomas, paragangliomas, carcinomas, metastases, subarachnoid hemorrhage, Chiari malformations, basilar invagination, and fractures [99].

Magnetic resonance imaging and computed tomography

As with the other lower cranial nerves, MRI is the preferred modality for CN XII, and CT provides complementary information on the integrity of the bony structures and foramina [125]. Evaluation of the entire course of the nerve is required, which includes evaluation of the nucleus in the brainstem medulla and the nerve in the cisternal segment and high carotid space. This is preferably obtained with a neck MRI that covers the entire nerve pathway. In patients with risk of contrast allergy and contrast-induced nephropathy, a noncontrast CT may be sufficient if patients cannot undergo MRI.

Ultrasound

US is not routinely used in the initial evaluation of the hypoglossal nerve.

FDG-PET/CT

PET is not routinely used in the initial evaluation of the hypoglossal nerve.

Variant 8: Perineural spread of tumor. (Most commonly trigeminal nerve [CN V], facial nerve [CN VII].)

Because of the complex anatomy of the head and neck and the close proximity of several cranial nerves, many clinical presentations of cranial neuropathy involve multiple nerves. As in syndromes of combined neuropathy of the upper cranial nerves, such as those related to vision and the extraocular muscles (which are covered in other Appropriateness Criteria), syndromes involving the lower cranial nerves are also grouped primarily by the proximity of the involved cranial nerves. For example, Gradenigo syndrome involves CNs V and VI as they travel in the vicinity of the petrous apex, whereas Vernet syndrome involves CNs IX, X, and XI as they travel within the jugular foramen. Collet-Sicard syndrome involves CNs IX, X , XI, and XII related to lesions just below the skull base or large lesions affecting both the jugular foramen and the hypoglossal canal. Imaging protocols should be tailored to evaluate the suspected region of anatomy when the syndrome is identified by the clinician.

A difficult problem for the surgeon is the perineural spread of head and neck malignancy. The trigeminal (CN V) and facial (CN VII) are the most common nerves involved; however, any cranial nerve traveling in the vicinity of a malignancy may become involved [126]. Perineural spread of tumor along the facial nerve may evade even the most meticulous imaging [127]. Subtle clues such as nerve enhancement, nerve enlargement, foraminal expansion, or muscle volume loss may indicate cranial nerve involvement with tumor [128]. For example,

asymmetry of facial musculature may be useful in detecting perineural tumor spread along the facial nerve or predicting return of function after nerve grafting [91,128].

Magnetic resonance imaging and computed tomography
MRI has emerged as the preferred imaging method for evaluating the perineural spread of tumor, although CT may be very useful for visualizing the neural foramina [126,129-134]. In patients with risk of contrast allergy and contrast-induced nephropathy, a noncontrast CT may be sufficient if patients cannot undergo MRI. Protocol with thin-section MRI should be performed to evaluate the cranial nerves.

FDG-PET/CT
PET imaging may also be helpful [133,135]. It may be useful as a problem-solving technique following initial cross-sectional imaging in patients with a known primary malignancy. PET/CT may also be superior to cross-sectional imaging for both localization and determination of response to therapy [120-124].

Ultrasound
US is not routinely used in the initial evaluation of the perineural spread of tumor.

Summary of Recommendations

- Pathology affecting the olfactory nerve is best evaluated with contrast-enhanced MRI. The protocol should be tailored to the anterior cranial fossa. CT may be useful in the evaluation of sinus inflammatory disease and trauma.
- Contrast-enhanced MRI is the primary technique to evaluate trigeminal neuralgia. MR angiography can be used to evaluate vascular compression.
- Facial nerve neuropathy is best evaluated with a dedicated contrast-enhanced MRI of the orbit, face, and neck, tailored to the temporal bone and parotid area.
- In the evaluation of cranial nerve IX symptoms, a focused contrast-enhanced MRI tailored to the posterior fossa is the study of choice.
- Cranial nerve X paralysis is well evaluated with either contrast-enhanced MRI or contrast-enhanced neck CT. Pathology in the posterior fossa will be better demonstrated with MRI. The complete evaluation of the nerve requires imaging the upper chest to the level of the anteroposterior window.
- In the evaluation of cranial nerve XI symptoms, a focused contrast-enhanced MRI tailored to the posterior fossa is the study of choice. Contrast-enhanced CT of the neck is complementary to the skull base imaging.
- Evaluation of lesions affecting cranial nerve XII is best done with a contrast-enhanced MRI tailored to the posterior fossa. The evaluation of the neck can also be done with contrast-enhanced neck CT.
- Perineural tumor spread most commonly affects CNs V and VII. Evaluation is best done with contrast-enhanced MRI tailored to the skull base.

Summary of Evidence

Of the 136 references cited in the *ACR Appropriateness Criteria® Cranial Neuropathy* document, 2 are categorized as therapeutic references, including 1 good-quality study. Additionally, 134 references are categorized as diagnostic references, including 3 well-designed studies, 19 good-quality studies, and 46 quality studies that may have design limitations. There are 67 references that may not be useful as primary evidence.

The 136 references cited in the *ACR Appropriateness Criteria® Cranial Neuropathy* document were published from 1998 to 2017.

Although there are references that report on studies with design limitations, 23 well-designed or good-quality studies provide good evidence.

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 [136].

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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