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# CRANIAL NERVES IX, X, XI, AND XII

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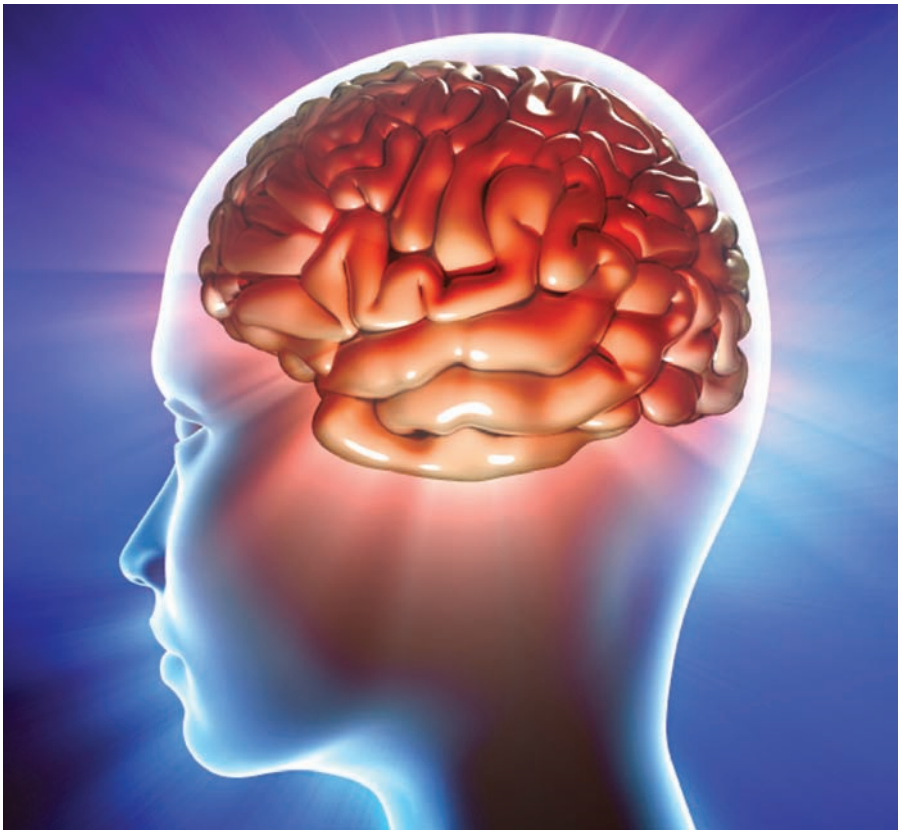
In this series, Drs. Sanders and Gillig explain how aspects of the neurological examination can aid in differential diagnosis of some common (and some uncommon) disorders seen in psychiatric practice.

## ABSTRACT

This article concludes the series on cranial nerves, with review of the final four (IX–XII). To summarize briefly, the most important and common syndrome caused by a disorder of the glossopharyngeal nerve (craniel nerve IX) is glossopharyngeal neuralgia. Also, swallowing function occasionally is compromised in a rare but disabling form of tardive dyskinesia called tardive dystonia, because the upper motor portion of the glossopharyngel nerve projects to the basal ganglia and can be affected by lesions in the basal ganglia. Vagus nerve funtion (craniel nerve X) can be compromised in schizophrenia, bulimia, obesity, and major depression. A cervical lesion to the nerve roots of the spinal accessory nerve (craniel nerve XI) can cause a cervical dystonia, which sometimes is misdiagnosed as a dyskinesia related to neuroleptic use. Finally, unilateral hypoglossal (craniel nerve XII) nerve palsy is one of the most common mononeuropathies caused by brain metastases. Supranuclear lesions of cranial nerve XII are involved in pseudobulbar palsy and ALS, and



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lower motor neuron lesions of cranial nerve XII can also be present in bulbar palsy and in ALS patients who also have lower motor neuron involvement. This article reviews these and other syndromes related to cranial nerves IX through XII that might be seen by psychiatry.

## INTRODUCTION

This article concludes the cranial nerves article series with a review of the final four (IX–XII). Cranial nerves (CN) IX through XI all leave the skull together through the jugular foramen. CN IX and X are so closely related that it is difficult to test them separately at the bedside. We added CN XII to this discussion because in some movement disorders and other neurological disorders, this cranial nerve is impacted along with IX, X, and/or XI. Many of us have not necessarily spent much time reflecting on the significance of these four cranial nerves within the field of psychiatry. However, in addition to other less familiar disorders, these four cranial nerves and their signs can be involved in some types of chronic pain, movement disorders, epilepsy, syncope, depression, schizophrenia, dementia, and eating disorders.

## CN IX: GLOSSOPHARYNGEAL

**Anatomy.** CN IX is innervated bilaterally and has sensory, parasympathetic, and motor components.[1–5] The sensory division receives general sensory fibers from the tonsils, pharynx, middle ear, and the posterior one-third of the tongue, as well as taste fibers from the posterior third of the tongue. The sensory division of the nerve projects to the solitary nucleus (taste).

Parasympathetic visceral fibers to the CN-IX arise in the carotid bodies of the neck. The nerve supplies parasympathetic fibers to the parotid gland that originate in the inferior salivary nucleus.

The cell bodies of the neurons forming the lower motor neuron portion of the CN IX, which innervates the stylopharynegeus muscle, are located in the nucleus ambiguous of the medulla. The glossopharygeal nerve has a communicating branch to the vagus nerve (CN X). Lesions to the CN IX are often accompanied by lesions to the CN X.

**Testing.** The sensory function of the nerve can be tested by evaluating taste (sweetness) on the posterior one-third of the tongue. Tactile sense in the pharynx is normally rolled into the gag reflex, but can be tested using light touch and direct inquiry about problems with aspiration.[4,5]

Lesions in the motor portion may be reflected in some difficulty swallowing, although the gag reflex itself may be normal.[6]

**Glossopharyngeal neuralgia and "swallow syncope."** The most important and common syndrome caused by a disorder of the CN IX is glossopharyngeal neuralgia. Glossopharyngeal neuralgia can be associated with fainting in 1 to 2 percent of affected people due to its reflex association with CN X. In glossopharyngeal neuralgia, the nerve dysfunction induces a reflex syndrome of pain, bradycardia (or actual stoppage of the heart temporarily), and syncope.[5,7,8] The same mechanism likely accounts for *swallow syncope*, where consciousness is lost during or immediately after a swallow even though there is no pain.

Glossopharyngeal neuralgia is a rare disorder. It usually begins after age 40 and occurs more often in men. Often, its cause is unknown, but sometimes glossopharyngeal neuralgia results from an abnormally positioned artery that compresses the CN IX near where it exits the brain stem.[3] The cause is rarely a tumor in the brain or neck or by dissection of the carotid artery. There are case reports of a schwannoma of the nerve itself,[9,10] which may also account for symptoms. Occasionally glossopharyngeal neuralgia is caused by a lesion associated with the onset of multiple sclerosis.[1] There is also a pharyngeal-facial variant of Guillian-Barre syndrome involving CN IX, the glossopharyngeal, and the post-infections syndrome of varicella zoster.[12,13]

Attacks of glossopharyngeal neuralgia are brief and occur intermittently, but they cause excruciating pain similar to trigeminal neuralgia. Attacks may be triggered by a particular action, such as chewing, swallowing, talking, coughing, or sneezing. The pain is localized to the base of the tongue, pharynx or larynx, tonsillar areas, and one ear. The pain may last several seconds to a few minutes and usually affects only one side of the throat and tongue. In 1 to 2 percent of people, the heartbeat is affected. It slows so much that it stops temporarily, causing fainting, as occurs in swallow syncope, but there is associated pain.

**Swallowing.** The CN IX is involved in a patient's ability to swallow because it innervates the stylopharyngeus muscle, which elevates the larynx and pulls it forward during the pharyngeal stage of the swallow. The gag reflex does not test this motor function of the glossopharygeal nerve. It is possible to swallow and not aspirate despite paralysis of the CN IX, but a recent fluoroscopic and endoscopic study of swallowing function in animals (horses) revealed a trend toward more associated tongue pushes and greater swallowing time when glossopharyngeal function was blocked.[14] Patients with developmental disabilities or strokes should be assessed for swallowing difficulties, which include an assessment of the functioning of the glossopharyngel nerve, because problems with this CN may subtly interfere with their ability to eat or take oral medication.

**Tardive dyskinesia.** Swallowing function occasionally is compromised in a rare but disabling form of tardive dyskinesia called tardive dystonia. In this condition, the upper motor portion of the glossopharyngel nerve projects to the basal ganglia and can be affected by lesions in the basal ganglia.[15]

**Hiccups.** Psychiatrists are sometimes consulted by patients suffering emotional distress as a result of chronic, intractable hiccups. The hiccup reflex is a coordinated motor activity that causes a brief, strong, inspiratory movement accompanied by glottic adduction of the stylopharyngeus muscle. It is useful to be aware that a CN IX block has sometimes provided relief of chronic hiccups.[16,17] This is because the hiccup reflex likely is mediated by the motor branch of CN IX.[18]

## CN X: VAGUS

**Anatomy.** Most of the fibers of CN X are sensory or parasympathetic fibers from the viscera (projecting to solitary nucleus) and parasympathetic nerves to internal organs such as the heart (originating in the nucleus ambiguous or dorsal motor nucleus). The CN X also has a general sensory division and three motor divisions.[19]

The three motor branches of CN X have bilateral upper motor neuron innervation, and consist of 1) the pharyngeal branch, which supplies the muscles of the soft palate and pharynx; 2) the superior laryngeal nerve, which supplies the inferior pharyngeal constrictor and cricothyroid muscles of the larynx, which are responsible for pitch adjustments of the voice; and 3) the recurrent laryngeal branches, which innervate all of the other intrinsic muscles of the larynx. The left side of the recurrent laryngeal branch is longer because it loops around the aorta and can be damaged during trauma surgery of the chest (e.g., surgery on a dissection of the ascending aorta, when the blood vessel is moved or retracted). Damage to the recurrent laryngeal nerve can result in temporary (up to one year) or permanent hoarseness.

The sensory division of the CN X receives information from the mucous membranes lining the larynx and the vocal cords, and the stretch receptors in the muscle spindles of the larynx.

**Testing.** The patient with a lesion of the laryngeal portion of CN X complains of a hoarse voice, difficulty in swallowing (dysphagia), and choking when drinking fluid. There is also loss of the gag reflex, which is the common clinical test for CN X motor function during the office exam (CN IX carries the afferent limb of the reflex). The uvula deviates away from the side of the lesion and there is failure of palate elevation.[4,5]

**Schizophrenia.** Impaired gag reflex has been found consistently in schizophrenia. Runeberg[20] found it reduced in 29 percent of patients with schizophrenia and absent in 24 percent. Craig et al[21] found it impaired in 31 percent of unmedicated patients with schizophrenia. This is somewhat contrary to the finding that vomiting is fairly common in at least chronic schizophrenia.[22]

**Bulimia.** Patients with bulimia nervosa lack the normal sensation of satiety when they eat.[23] The sensation of satiety is mainly a function of the CN X. Similarly, patients with bulimia nervosa have a higher pain threshold.[24] Pain is modulated by input from the CN X and can be affected by CN X stimulation. It has been proposed that a destabilization of the positive vagovagal reflex, which is found in patients with bulimia nervosa and affects satiety, may also be amenable to vagal stimulation and be a potential avenue for treatment.[23,24]

**Obesity treatment.** A six-month, open-label trial involving three medical centers in Australia, Mexico, and Norway using CN X blocking (VBLOC) therapy, which is similar to vagal nerve stimulation but only used during the day, has resulted in 31 participants with obesity losing an average of nearly 15 percent of their excess weight over a six-month period. A one-year, 300-participant, double-blind Phase II trial has begun.[25–27]

**Vagotomy (cutting of the CN X).** Vagotomy is currently being researched as a less invasive alternative weight loss procedure to gastric bypass surgery. The procedure curbs the feeling of hunger and is sometimes performed in conjunction with putting a band on the patient's stomach, resulting in average weight loss of 43 percent of body weight at six months with diet and exercise.[27]

**Sexual dysfunction in women after spinal cord injury.** Studies have shown that women who have complete transection of the spinal cord can experience orgasms through the CN X, which can travel from the uterus, cervix, and probably the vagina to the brain via the CN X sensory pathways.[28,29]

**Major depression.** There is considerable evidence from both animal and human neurochemical and neuroimaging studies that CN X influences limbic and higher cortical brain regions that are implicated in mood disorders. This provides a rationale for vagal nerve stimulation in the treatment of major depression.[30,31] At this time, the acute and long-term efficacy of vagal nerve stimulation for the treatment of psychiatric disorders is still under debate. The exact mechanism of action of vagal nerve stimulation in these circumstances is still not well-understood. In addition, although vagal nerve stimulation is well-tolerated, the optimal dosing strategies have not been determined, and we, as of yet, do not have clear predictors of who will respond to the treatment (as is also true for other current treatments of major depression).

**Tardive dyskinesia.** There is a syndrome of tardive dyskinesia that involves the muscles of the larynx due to involvement of a motor division of CN X, the vagus. This is usually referred to as tardive dysphonia and is characterized by a combination of hoarseness and pitch change when the patient attempts to speak.

## CN XI: SPINAL ACCESSORY

**Anatomy.** Although CN XI is included in the CNs, the cell bodies of this nerve actually reside in the spinal cord. They probably are

continuous with the nucleus ambiguous of the medulla, however. CN XI emerges from the jugular foramen with CNs IX and X, but it does so by first ascending into the skull via the foramen magnum, and then exiting the central nervous system via the jugular foramen. This unusual anatomical configuration makes CN XI at risk for injury in fractures of the cervical vertebrae or other spinal cord injuries. CN XI innervates the sternocleidomastoid muscle and the trapezius muscle.[5,6]

**Testing.** CN XI may be damaged from surgery or trauma involving the neck, such as lymph node biopsy or even whiplash, or other cervical cord lesions (such as syringomyelia). Symptoms of damage to CN XI include shoulder pain, scapular winging, and weakness or atrophy of the trapezius muscle.

**Cervical dystonia (torticollis).** A cervical lesion to the nerve roots of CN XI can result in cervical dystonia or torticollis. Patients who are receiving antipsychotic medications and develop cervical dystonia should be evaluated for lower motor neuron nerve root compression of CN XI at the cervical level in a differential diagnosis, which also involves the more obvious and common cause of cervical dystonia—central involvement of the motor pathways at the level of the basal ganglia resulting in tardive dystonia.[32]

## CN XII: HYPOGLOSSAL NERVE

Like CN XI, CN XII is also almost entirely a motor nerve. CN XII supplies the tongue muscles.

**Anatomy.** The nuclei of CN XII is more dorsal and medial in the medulla than the other CNs in the medulla. Supranuclear innervation is mainly from the contralateral cortex and descends in the corticobulbar tract. The fibers leave the medulla and pass through the hypoglossal canal. CN XII leaves the skull through the hypoglossal foramen, which is different than the previous three CNs. The CN XI is somewhat bilaterally innervated at the upper motor neuron level, but with more input from the contralateral hemisphere. Unilateral upper motor neuron lesions result in mild contralateral weakness of the tongue. Interruption of the nerve at the lower motor neuron on one side causes complete paralysis of that side of the tongue. The tongue will curve slightly to the healthy side when lying inactive and deviate to the affected side upon protrusion because it has lost the strength to resist the push to that side. The denervated side becomes wrinkled and atrophied. CN XII involvement is seen with many of the peripheral lesions that affect the spinal accessory nerve because they are anatomically close at that level.[33]

**Metastases.** Unilateral 12th nerve palsy (CN XII) is one of the more common cranial mononeuropathies caused from brain metastases.

**Pseudobulbar palsy.** Bilateral supranuclear lesions of CN XII that are seen in pseudobulbar palsy produce moderate-to-severe inability of the tongue to function.

**Amyotrophic lateral sclerosis (ALS).** As one might expect, bulbar onset of ALS is more likely to affect the tongue than is limb onset. ALS has features of both upper and lower motor neuron damage. Some patients will also display upper motor neuron involvement of the hypoglossal nerve, and if this is the sole manifestation it is referred to as pseudobulbar palsy. A smaller group of ALS patients will present initially with lower motor neuron damage, with primary bulbar involvement. Such patients will show evidence of tongue weakness, atrophy, and fasciculations. This is referred to as bulbar palsy.

## SUMMARY

Many of us have not necessarily spent much time reflecting on the significance of cranial nerves IX, X, XI, XII within the field of psychiatry (and perhaps before reading this article, no time at all!). However, in addition to other less familiar disorders, these four cranial nerves and their signs can be involved in a number of conditions that are commonly seen by the Psychiatrist, and also a few rare ones that now might occur to us in a differential diagnosis.

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