Cranial Nerve X: Vagus Nerve

15

One sentence: The vagus nerve is the longest CN, with the widest anatomical distribution, and it is important for swallowing and autonomic function (Figs. 15.1 and 15.2).

Genetic testing	NCV/EMG Autonomic testing	Laboratory	Imaging	Biopsy
	+		+	

Symptoms

Patients with vagus nerve damage experience swallowing difficulty and hoarseness of voice. "High" vagus nerve lesions are rare and are associated with ear and occipital pain.

Signs

Vagus nerve damage can cause paralysis of the palate, pharynx, and larynx according to the site of the lesion and cause dysphagia. Bilateral lesions can lead to a nasal voice and regurgitation through the nose. The gag reflex can be absent, and the uvula deviates away from the side of the lesion as a failure of palatal elevation occurs.

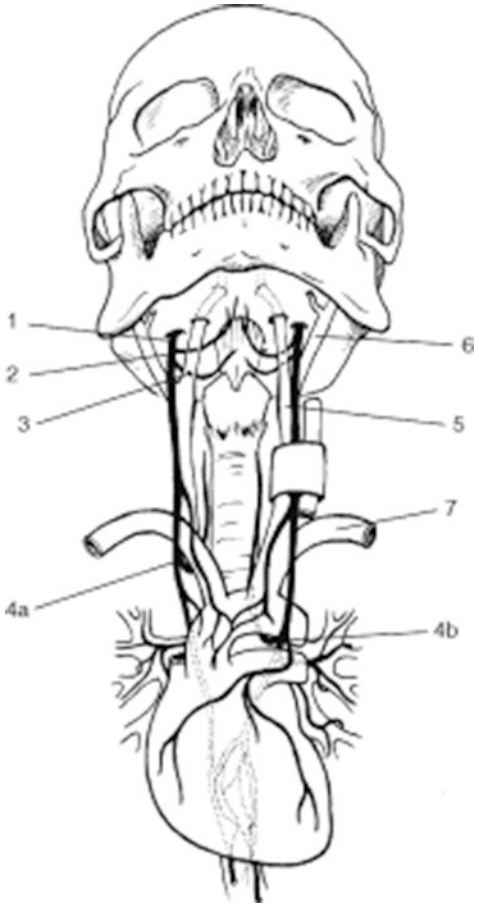

Fig. 15.1 *1* Vagus nerve, *2* pharyngeal branch, *3* internal laryngeal branch, *4a* right recurrent laryngeal nerve (across the subclavian artery), *4b* left recurrent laryngeal nerve (across the arch of the aorta), *5* internal carotid artery, *6* external carotid artery, *7* left subclavian artery

W. Grisold et al., *The Cranial Nerves in Neurology*, https://doi.org/10.1007/978-3-031-43081-7_15

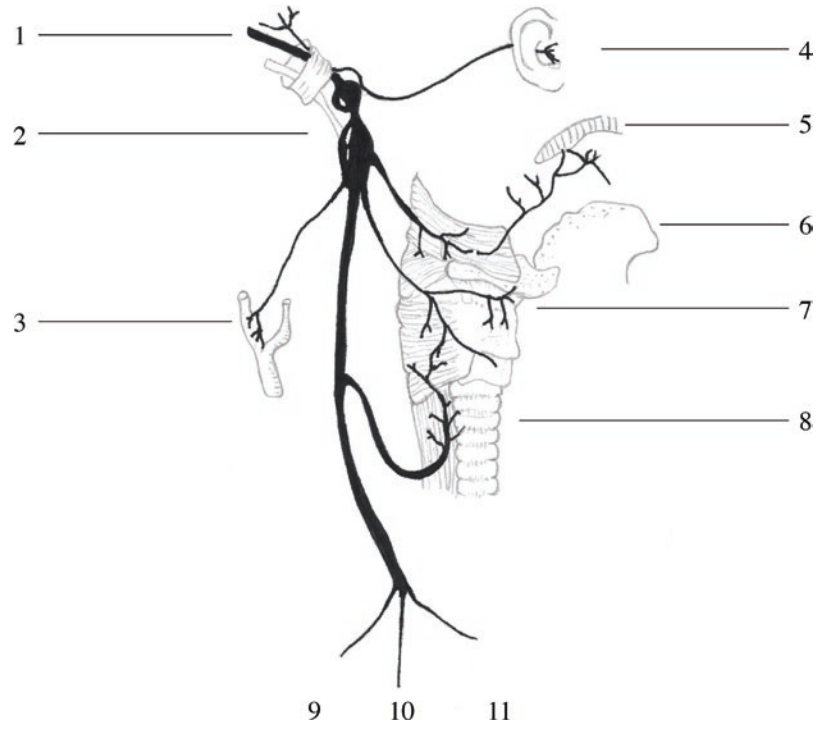

Fig. 15.2 Vagus nerve. *1* Vagus nerve, *2* ganglia, *3* branch to sinus, *4* auricular branch, *5* pharyngeal branch, *6* superior laryngeal nerve, *7* internal laryngeal nerve, *8* recurrent laryngeal nerve, and branches to *9* lung, *10* gastrointestinal tract, and *11* heart

Specific Qualities

Motor: +.

Branchial motor: Pharynx (except stylopharyngeus and tensor veli palatini), larynx, tongue. Striated muscle of soft palate.

Visceral motor: Smooth muscle and glands of pharynx, larynx, thoracic and abdominal viscera.

Sensory: +.

General sensory: Auditory meatus, skin on the back of the ear, external tympanic membrane, pharynx.

Visceral sensory: Larynx, trachea, esophagus, thoracic and abdominal viscera, stretch receptors in the wall of the aortic arch, chemoreceptors in the aortic body.

Autonomic: +.

Special senses: + (taste pharyngeal).

Other:

Location of Lesions

Central:

Supranuclear: Brain motor fibers for the ambiguous nucleus travel from the precentral gyrus via corticobulbar fibers. Impairment jointly with CN IX, XI, and XII.

Only mild symptoms occur in unilateral lesions. Bilateral lesions cause supranuclear palsy.

The dorsal motor nucleus (parasympathetic) receives input from the hypothalamus, olfactory nucleus, and reticular formation.

Brain stem and nuclear: Vascular syndromes, e.g., Avellis and Wallenberg syndrome.

Tumor: Brain stem glioma. Gross total surgical removal of malignant glioma [1].

Infection: diphtheria, herpes zoster encephalitis [2], poliomyelitis.

Intracranial Within the Skull:

The vagus nerve emerges from the medulla oblongata with several rootlets and exits through the jugular foramen, within the same dural sleeve as the accessory nerve.

Two external ganglia, the superior and inferior vagal ganglia, are found along the nerve's course within the jugular fossa of the petrous temporal bone.

Exit of the Skull:

The jugular foramen can be subdivided into three compartments (anterior, anteromedial, and posterolateral). CN IX, CN X, and CN XI are contained in the intermediate compartment. In the anteromedial portion of the jugular foramen, the tympanic branch of the glossopharyngeal nerve (Jacobsen's nerve) is located. The foramen is located behind the carotid canal.

CN X has two ganglia, the superior and the inferior.

The jugular foramen syndrome is well described in metastatic disease.

Vernet syndrome (jugular foramen syndrome): Describes lesions below the jugular foramen, also including CN XII.

Outside of the Skull:

Extracranial pathway: In the neck region, the nerve branches into the meningeal and auricular ramus, the pharyngeal rami, and the superior laryngeal nerve (internal and external rami). The pharyngeal rami innervate all the muscles of the pharynx, except the stylopharyngeus and the tensor veli palatini muscles. The superior laryngeal nerve divides into the internal and external laryngeal nerves. The external laryngeal branch supplies the inferior constrictor muscles. The vocal cords are innervated by the superior laryngeal nerve and the external and internal rami of the inferior laryngeal nerve. The esophageal ramus innervates the striated muscles of the esophagus.

The cervical branches can be asymmetric and are important in vagus nerve stimulation [3]. More distal branches are divided into thoracal and abdominal branches and carry autonomic fibers.

Thoracic branches: Inferior cardiac branches, anterior bronchial branches, posterior bronchial nerve.

Abdominal branches: Gastric, celiac, hepatic nerve.

Combination with Other CNs

Jugular foramen and base of the skull.

Causes and Frequency

Iatrogenic: Surgery of thyroid, neck, and mediastinal tumors, mediastinoscopy, surgery of the trachea and esophagus, thoracotomy, thyroid surgery (recurrent nerve), gastric surgery.

Infectious: Botulism, diphtheria, herpes, meningitis, poliomyelitis, tetanus [4].

Inflammatory/immune-mediated: Dermato- and polymyositis and sarcoid [5].

Neoplastic: Jugular foramen tumor, meningeal carcinomatosis, metastasis (with CN IX involvement), lymphoma [6].

Metabolic: Hyperpotassemia and hypophosphatemia.

Motor neuron disease.

Neuromuscular transmission disorders: Myasthenia gravis, others.

Side effects of vagus nerve stimulation: Voice alterations, vocal cord palsy, throat pain [7].

Surgery: Lung, mediastinum, esophageal cancer, and recurrent nerve in thyroid surgery [8].

Trauma: Fractures that affect the jugular foramen (uncommon). Hyperextension neck injuries are also sometimes associated with injury to these nerves at the cranio-cervical junction [9].

Combat and war: Recurrent laryngeal nerve, unilateral or bilateral, base of the skull injuries.

Toxic: Alcoholic polyneuropathy and thallium.

Tumor (rare): Glomus tumors [10], lymphoma [11], neurofibroma, neurogenic tumors [12], neurilemmoma [13], schwannoma.

Vascular: Medullary infarction, pharyngeal artery embolization (damage of vasa nervorum).

Others: Familial hypertrophic polyneuropathy, myopathies (chronic progressive external ophthalmoplegia, oculopharyngeal muscular dystrophy), polyneuropathies (amyloid – some

types, diphtheria, alcohol). Tardive dyskinesia can involve laryngeal muscles.

Special nerve segments:

"High vagus lesions" involving the meningeal and occipital branch: Swallowing difficulties, hoarseness, local occipital pain, hypersensitivity of the ear [14].

Vagus recurrent nerve:

Focal superior and recurrent laryngeal neuropathies: Peripheral lesions affecting the recurrent laryngeal nerve, with or without involvement of the superior laryngeal nerve, are most common from trauma, surgery, thyroidectomies, carotid endarterectomies, or idiopathic causes.

The laryngeal neuropathy causes inability to forcefully cough, and hoarseness appears. Causes of focal damage of the recurrent laryngeal nerve include diseases of the lungs, tumors in the thoracic cavity (*e.g.*, lung cancer), and heart and lung transplant: [15] Aneurysm of the aortic arch, enlarged lymph nodes, and thyroid surgery. Cervical disc surgery by anterior approach [16]. About 25% of cases are idiopathic [17].

Recurrent laryngeal nerve lesions: Hoarseness is observed in local anesthetic procedures, presumably due to excessive local anesthetic spread.

Multiple CN: [18].

Other entities:

Focal laryngeal dystonia.

The gag reflex can be diminished in patients with schizophrenia, obesity treatment, sexual dysfunction in women after spinal cord injury, and spastic dystonia.

Neuralgia of the laryngeal nerve (rare) [19].

Main Investigations

Diagnosis can be facilitated with ENT examination and vocal cord inspection (with endoscopy), imaging, and video swallowing studies. EMG of the cricothyroid muscle (superior laryngeal nerve) or thyroarytenoid muscle (recurrent nerve) can be done but is uncommon.

Differential diagnosis: Bulbar disorders. Motor neuron diseases, neuromuscular transmission disorders.

Therapy

Treatment depends upon the etiology:

Surgical reinnervation techniques after trauma [20].
Monitoring: Thyroid surgery [21].
Vagus nerve stimulation for intractable hiccup treatment [22].

Prognosis: Prognosis depends upon the etiology.

References

1. Kyoshima K, Sakai K, Goto T, Tanabe A, Sato A, Nagashima H, et al. Gross total surgical removal of malignant glioma from the medulla oblongata: report of two adult cases with reference to surgical anatomy. J Clin Neurosci. 2004;11(1):75–80.
2. Cao DH, Xie YN, Ji Y, Han JZ, Zhu JG. A case of varicella zoster encephalitis with glossopharyngeal and vagus nerve injury as primary manifestation combined with medulla lesion. J Int Med Res. 2019;47(5):2256–61.
3. Hammer N, Glatzner J, Feja C, Kuhne C, Meixensberger J, Planitzer U, et al. Human vagus nerve branching in the cervical region. PLoS One. 2015;10(2):e0118006.
4. Amin MR, Koufman JA. Vagal neuropathy after upper respiratory infection: a viral etiology? Am J Otolaryngol. 2001;22(4):251–6.
5. Vaile JH, Davis P. Isolated unilateral vagus nerve palsy in systemic lupus erythematosus. J Rheumatol. 1998;25(11):2287–8.
6. Mustafa R, Neth BJ, Stitt D. Bilateral Vagus nerve enhancement in a patient with leptomeningeal lymphoplasmacytic lymphoma presenting with intractable hiccups. JAMA Neurol. 2021;78(4):493–4.
7. Verlinden TJ, Rijkers K, Hoogland G, Herrler A. Morphology of the human cervical vagus nerve: implications for vagus nerve stimulation treatment. Acta Neurol Scand. 2016;133(3):173–82.

8. Gunn A, Oyekunle T, Stang M, Kazaure H, Scheri R. Recurrent laryngeal nerve injury after thyroid surgery: an analysis of 11,370 patients. J Surg Res. 2020;255:42–9.
9. Aygun D, Acar E. Isolated unilateral vagus nerve palsy secondary to trauma. Ulus Travma Acil Cerrahi Derg. 2013;19(2):180–2.
10. Netterville JL, Jackson CG, Miller FR, Wanamaker JR, Glasscock ME. Vagal paraganglioma: a review of 46 patients treated during a 20-year period. Arch Otolaryngol Head Neck Surg. 1998;124(10):1133–40.
11. Boasquevisque GS, Guidoni J, Moreira de Souza LA, Gonçalves PG, Andrade CV, Pedras FV, Boasquevisque EM, Boasquevisque ET. Bilateral vagus nerve neurolymphomatosis diagnosed using PET/CT and diffusion-weighted MRI. Clin Nucl Med. 2012;37(9):e225–8.
12. Sugio K, Inoue T, Inoue K, Tateishi M, Ishida T, Sugimachi K. Neurogenic tumors of the mediastinum originated from the vagus nerve. Eur J Surg Oncol. 1995;21(2):214–6.
13. Yumoto E, Nakamura K, Mori T, Yanagihara N. Parapharyngeal vagal neurilemmoma extending to the jugular foramen. J Laryngol Otol. 1996;110(5):485–9.
14. Grisold W, Schwarzmeier J, Frei K, Neumuller G, Breier F. Ti.: "High" vagus nerve lesions in varicella zoster infection. eNeurologicalSci 2021;23:100337
15. Murty GE, Smith MC. Recurrent laryngeal nerve palsy following heart-lung transplantation: three cases of vocal cord augmentation in the acute phase. J Laryngol Otol. 1989;103(10):968–9.
16. Bertalanffy H, Eggert HR. Complications of anterior cervical discectomy without fusion in 450 consecutive patients. Acta Neurochir. 1989;99(1-2):41–50.
17. Reiter R, Hoffmann TK, Rotter N, Pickhard A, Scheithauer MO, Brosch S. Etiology, diagnosis, differential diagnosis and therapy of vocal fold paralysis. Laryngorhinootologie. 2014;93(3):161–73.
18. Erman AB, Kejner AE, Hogikyan ND, Feldman EL. Disorders of cranial nerves IX and X. Semin Neurol. 2009;29(1):85–92.
19. Kandan SR, Khan S, Jeyaretna DS, Lhatoo S, Patel NK, Coakham HB. Neuralgia of the glossopharyngeal and vagal nerves: long-term outcome following surgical treatment and literature review. Br J Neurosurg. 2010;24(4):441–6.
20. Aynehchi BB, McCoul ED, Sundaram K. Systematic review of laryngeal reinnervation techniques. Otolaryngol Head Neck Surg. 2010;143(6):749–59.
21. Abt NB, Puram SV, Kamani D, Modi R, Randolph GW. Neuromonitored thyroid surgery: optimal stimulation based on intraoperative EMG response features. Laryngoscope. 2020;130(12):E970–E5.
22. De Vloo P, Dallapiazza RF, Lee DJ, Zurowski M, Peng PW, Chen R, et al. Long-term relief of intractable hiccups with vagal nerve stimulation. Brain Stimul. 2018;11(6):1385–7.