

# Neurologic Syndromes of the Head and Neck

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## KEYWORDS

• Facial nerve paralysis • Trigeminal neuralgia • Herpes zoster

## KEY POINTS

- Malignant (necrotizing) otitis externa can cause facial paralysis in addition to ear pain. Ear pain from necrotizing otitis externa is described as severe, deep, constant pain. If facial nerve paralysis is present, eye care with artificial tears in the day time, lubricating ointment at night time, and taping the eyelid shut at night should be done to protect against corneal abrasion and ulceration.
- Herpes zoster (HZ) can occur along the distribution of any nerve after reactivation of varicella zoster, including the cranial nerves.
- Treatment of HZ oticus consists of combined antiviral and corticosteroid therapy. About 75% of patients of HZ oticus will recover from facial paralysis provided treatment is begun within the first 72 hours of onset of symptoms. Early diagnosis is therefore critical.
- Bell's palsy is a term for *idiopathic* facial nerve paralysis, although it is now widely believed that reactivation of Herpes Simplex Virus type 1 (HSV-1) accounts for most Bell's palsy cases. Some cases of Bell's palsy may be attributed to ischemia and microvascular disease from diabetes mellitus, analogous to diabetic mononeuropathy. Bell's palsy is believed to account for 60% to 75% of all cases of unilateral facial paralysis.
- Facial paralysis in children is different than in adults in that up to 70% of cases have identifiable secondary causes compared to 25% to 40% in adults. Except in areas endemic for Lyme disease, the most common cause of facial paralysis in children is acute otitis media.
- A patient experiencing a trigeminal neuralgia paroxysm typically freezes in place with hands slowly rising to the area of pain on the face but not touching it. He or she then grimaces with an involuntary spasm of the facial muscles referred to as tic douloureux and then either remains in this position or cries out in pain.

Although patients with Bell's palsy and trigeminal neuralgia occasionally present to primary care physicians, most of the syndromes in this article are somewhat rare in primary care practice. It is important to recognize signs and symptoms of these syndromes so that appropriate management is carried out. Patients may experience sensory (pain, neuralgia, altered taste, hypesthesia, altered hearing, etc.) and/or motor

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(paresis/paralysis) symptoms of the head and neck due to disorders of the cranial and cervical nerves. The syndromes and symptoms discussed in this article include ear pain, sinus pain, herpes zoster (HZ) oticus (Ramsay Hunt), HZ ophthalmicus, facial nerve paralysis in adults and children, superior laryngeal neuralgia, trigeminal neuralgia, glossopharyngeal neuralgia, nervus intermedius (geniculate) neuralgia, and Raeder paratrigeminal syndrome.

## EAR PAIN

The ear is innervated by sensory fibers from cranial nerves V, VII, IX, and X, as well as the second and third cervical nerves.<sup>1</sup> When ear pain is caused by intrinsic ear disease, such as otitis media or otitis external, the otologic examination is usually abnormal. Common causes of ear pain with a normal ear examination include neuralgias and referred pain from conditions such as temporomandibular joint disorders, pharyngitis, dental disease, cervical spine arthritis, and neuralgias.

There are many causes of ear pain.<sup>1</sup> **Table 1** lists causes of ear pain and their distinguishing characteristics. A focused history, physical examination, and diagnostic testing will help differentiate these causes (**Box 1**).

Malignant (necrotizing) otitis externa can cause facial paralysis in addition to ear pain.<sup>2</sup> Ear pain from necrotizing otitis externa is described as severe, deep, constant pain. If facial nerve paralysis is present, eye care with artificial tears in the day time, lubricating ointment at night time, and taping the eyelid shut at night should be done to protect against corneal abrasion and ulceration.

Patients with ear pain not explained by otologic findings who have a significant history of tobacco or alcohol use and are older than 55 years of age should be considered for nasolaryngoscopy and enhanced MRI of the head and neck to look for tumors of the nose, nasopharynx, oral cavity, oropharynx, infratemporal fossa, and neck.<sup>1</sup> Other significant risk factors raising the suspicion of tumor include dysphagia, weight loss, radiation exposure, and hoarseness.

Patients with temporal (giant cell) arteritis typically experience temporal pain and tenderness that may occasionally radiate to the ear. They also commonly experience malaise, weight loss, fever, anorexia, and visual loss. Most patients with temporal arteritis have elevated erythrocyte sedimentation rates, and may or may not have a positive biopsy of the temporal artery.

## SINUS PAIN

Sinus pain can be confused with facial pain from other sources, such as neuralgia. Acute purulent rhinosinusitis can cause both local and referred pain. These are the common patterns of pain referral:

- Maxillary sinusitis causes pain and tenderness over the cheek. Pain tends to be constant and burning in nature with zygomatic and dental tenderness.<sup>3</sup>
- Frontal sinusitis causes frontal (forehead) pain and tenderness.<sup>4</sup>
- Sphenoid and ethmoidal sinusitis causes pain behind and between the eyes.<sup>4</sup>

Pain from rhinosinusitis is typically exacerbated when the patient bends forward and relieved as soon as the infected material drains from the sinus.<sup>4</sup>

## HERPES ZOSTER

HZ can occur along the distribution of any nerve after reactivation of varicella zoster, including the cranial nerves. The elderly and patients who have diminished

<b>Table 1 Differential diagnosis of ear pain</b>	
<b>Diagnosis</b>	<b>Distinguishing Characteristics</b>
Otitis media	Abnormal examination of tympanic membrane
Otitis externa	Pain elicited by traction on ear; swelling and erythema of external auditory canal with debris
Foreign body	Visible on otoscopic examination
Barotrauma	TM hemorrhage and/or serous or hemorrhagic middle ear fluid; pain onset of airplane descent or while scuba diving
TMJ syndrome	Tender TMJ; crepitus of clicking with opening and closing mouth
Dental causes	Caries, abscesses, gingivitis, teeth tender to percussion
Pharyngitis	Erythema of throat, swelling, exudate, sore throat
Necrotizing (malignant) otitis externa	Associated with osteitis of the base of the skull, granulation tissue on floor of external auditory canal, may cause facial nerve palsy, older and immunocompromised patients, especially diabetics
HZ oticus	Vesicular rash on auricle, facial nerve paralysis, hearing loss, vertigo
Cellulitis/perichondritis	Preceding insect bite, piercing, scratch usually involving lobe
Relapsing polychondritis	Recurrent swelling and redness of auricle; a systemic disease involving cartilage but spares the earlobes (where there is no cartilage); often involves both ears
Trauma	Blunt or sharp, lacerations, frostbite
Mastoiditis	Recent or concurrent otitis media; pain behind ear
Tumors or cysts in auricle or canal	History of smoking, alcohol use, age >50, hoarseness, dysphagia, weight loss
Neuralgias	Brief attacks of pain that is intense, episodic, electric shocklike
Temporal arteritis	Older age, jaw claudication, vision loss, high ESR
Oral aphthous ulcers	Localized pain in mouth with sores
Cervical adenopathy	URI, tender cervical nodes
Eagle syndrome (elongated styloid)	Deep, pain exacerbated by swallowing or chewing
Sinusitis	Nasal congestion; tender over involved sinuses
Carotidynia	Dysphagia and throat tenderness; tender carotid artery; more common in women with migraine
Thyroiditis	Pain in thyroid with tenderness and referral to ear
Salivary gland disorders	Pain in front of ear, prominent and tender parotid glands
GERD	Acid reflux, heartburn; referral to ear
Angina pectoris	Cardiac risk factors
Thoracic aneurysms	Older men usually, chest or back pain
Psychogenic	History of depression or anxiety
Ramsay Hunt syndrome (HZ oticus)	HZ of cranial nerves VII and VIII
Cholesteatoma	May be painful, but more likely associated with a sense of fullness of the ear
Tumors in the nose, nasopharynx, oropharynx, oral cavity, infratemporal fossa, neck, chest	May cause referred pain to the ear

*Abbreviations:* ESR, erythrocyte sedimentation rate; GERD, gastroesophageal reflux disease; TMJ, temporomandibular joint; URI, upper respiratory tract infection.

*Adapted from* Ely J, Hansen M, Clark E. Diagnosis of ear pain. *Am Fam Physician* 2008;77(5):621–8; with permission.

**Box 1****Key history, physical testing, and diagnostic testing to differentiate causes of ear pain**

## History

- What is the location of pain? (Ask the patient to point to the pain with their finger).
- Does the pain radiate? If so, where?
- What aggravates the pain? (eg, chewing, swallowing, coughing)?
- Are the associated symptoms? (otologic or systemic)
- Are there risk factors for tumors (eg, age >50, tobacco and/or alcohol use)?
- Do symptoms favor a primary disorder of the ear (discharge, tinnitus, hearing loss, vertigo)?
- Is there a history of significant allergy or recent upper respiratory infection (eustachian tube dysfunction)?
- Has there been recent air travel, scuba diving history, or other causes of barotrauma?

## Physical Examination

- Examine the auricle. Tenderness with traction on the auricle indicates otitis externa.
- Perform a thorough otoscopic examination (foreign body in ear canal, erythema of temporomandibular joint or ear canal)?
- Palpate temporomandibular joint for tenderness and crepitus as patient opens and closes mouth.
- Palpation of the head, face, and neck. Frontal, maxillary, sphenoid, and ethmoid sinusitis can cause referred pain.
- Inspect the nose and oropharynx.
- Inspect the gingiva and percuss the teeth with a tongue depressor to detect dental disease.
- Examine the cranial nerves.

## Diagnostic tests

- Assessment of hearing.
- Pneumatic otoscopy or tympanometry to test mobility of tympanic membrane.
- Nasaloryngoscopy and enhanced magnetic resonance imaging (MRI) of the head and neck if patient is at high risk for tumor (MRI is best for soft tissue involvement).
- Computed tomography (CT) with contrast if MRI result is positive or if history of trauma to temporal bone (to look for bone involvement).

cell-mediated immunity (HIV, malignancy, immunosuppressive therapy, chemotherapy, corticosteroid use, and emotional stress) are more likely to develop HZ.<sup>5</sup> Typically, patients developing HZ have a prodrome of 2 to 3 days with varying degrees of pain. Descriptions of the quality of pain may include “burning, shooting, stabbing, or throbbing.” After the prodrome, most patients develop a dermatomal rash that is initially erythematous, then papular, then vesicular. These lesions usually become pustular about 1 week after the rash begins. Late appearance of new vesicles beyond 1 week of rash onset often indicates an immunodeficiency syndrome.<sup>5</sup> Adjacent dermatomes may occasionally be involved. Disseminated HZ involving multiple dermatomes is more commonly seen in immunocompromised patients. Approximately 5% of patients with HZ will have a second episode; most of these patients are immunocompromised.<sup>5</sup> Post-herpetic neuralgia (PHN) is a common complication from HZ. PHN is defined as at least 90 continuous days of documented pain. The incidence of

PHN increases with age.<sup>6</sup> Sometimes pain can be evoked by stimuli that normally would not cause pain, such as light touch with a cotton swab. This is known as allodynia and is somewhat common in acute HZ pain, as well as PHN.

The treatment of HZ is with antiviral drugs. Acyclovir (800 mg 5 times a day for 7 days), valacyclovir (1000 mg 3 times a day for 7 days), or famciclovir (500 mg 3 times a day for 7 days) should be prescribed within 48 to 72 hours of onset of the rash.<sup>5</sup> Antiviral drugs do not, unfortunately, prevent or cure chronic pain from HZ. They also do not eradicate the virus. Antivirals are effective against viral replication, thus reducing the duration of viral shedding and new lesion formation. Antiviral drugs have been shown to decrease the duration and severity of acute pain of HZ.<sup>5</sup> By reduction of acute neural damage, antiviral drugs are believed to significantly reduce the duration of pain in PHN.<sup>5</sup> Corticosteroids given in the acute stages of HZ do not seem to prevent chronic pain of PHN, but they have been demonstrated to significantly reduce acute pain from HZ.<sup>5</sup> HZ vaccine should not be used in the treatment of HZ. Acute and chronic pain from HZ can be treated with tricyclic antidepressants, venlafaxine, duloxetine, opioids, tramadol, gabapentin, pregabalin, nerve blocks, and intravenous VZV hyperimmune globulin.<sup>5</sup>

### HERPES ZOSTER OPHTHALMICUS

Patients with HZ ophthalmicus (HZO) typically have a prodrome of severe headache, malaise, and fever, with unilateral pain (or hypesthesia) in the affected eye and on the forehead and top of the head.<sup>7</sup> The distribution of HZO is the V1 (ophthalmic) distribution of the trigeminal nerve. Along with vesicular eruption of the dermatome, patients commonly have conjunctivitis, episcleritis, and drooping of the eyelid. When the rash spreads down the nose, indicating nasociliary branch involvement, viral keratitis occurs (Hutchinson sign).<sup>8</sup> Keratitis is typically a very painful condition that may appear as a corneal abrasion or as a branching (“dendritic”) pattern on fluorescein staining of the cornea. HZO keratitis is a potentially sight-threatening condition. Patients with HZO should be referred to ophthalmologists urgently. Approximately two-thirds of patients with HZO are estimated to develop keratitis.<sup>8</sup>

### HERPES ZOSTER OTICUS: RAMSAY HUNT SYNDROME

The incidence of HZ oticus (HZ otics) is 5 per 100,000 per year, mostly in patients older than 60 years of age.<sup>9,10</sup> **Box 2** lists the clinical characteristics of HZ oticus.

HZ oticus was first termed “HZ cephalicus” by James Ramsay Hunt,<sup>11</sup> who described varicella zoster virus (VZV) involvement of cranial nerves V, VII, VIII, IX, and X. HZ oticus is caused by reactivation of latent VZV in the geniculate ganglion and affects both cranial nerves VII and VIII due to their close proximity to one another.<sup>5</sup> Approximately 8% of patients with HZ oticus do not have vesicles, a condition known as “sine herpette.”<sup>12</sup> Mucocutaneous vesicles may also occur in the oral mucosa or on the tongue.

Patients with facial nerve paralysis from HZ oticus tend to have a worse prognosis for recovery of facial muscle function than patients with Bell’s palsy.<sup>12</sup> Recovery of facial function from HZ oticus is often incomplete, as is recovery of hearing and problems with balance. When patients have facial nerve paralysis with concomitant severe pain or sensorineural hearing loss, a diagnosis of HZ oticus should be considered instead of Bell’s palsy.<sup>11</sup> Prediction for recovery of facial paralysis often cannot be made until 3 weeks after the onset of facial palsy. Electroneuronographic (ENG) studies of facial nerve function performed between days 10 and 14 for HZ oticus have been recommended by some experts, but may not routinely provide accurate information on the prognosis or recovery rate from facial paralysis.<sup>13</sup>

**Box 2****Clinical characteristics of HZ oticus**

- Vesicles of the auricle or ear canal with ear pain
- Facial paralysis
- Tinnitus
- Sensorineural hearing loss
- Hyperacusis/dysacusis
- Vertigo
- Dysgeusia

*Data from Adour KK. Otolological complications of Herpes Zoster. Ann Neurol 1994;35:S62–4.*

Treatment of HZ oticus consists of combined antiviral and corticosteroid therapy. About 75% of patients of HZ oticus will recover from facial paralysis provided treatment is begun within the first 72 hours of onset of symptoms.<sup>5,9</sup> Early diagnosis is therefore critical. Antiviral treatment options include acyclovir (800 mg 5 times daily for 7–10 days), valacyclovir (1000 mg 3 times a day for 7–10 days), or famcyclovir (500 mg 3 times a day for 7–10 days).<sup>9</sup> One example of a common corticosteroid regimen is prednisone, 60 mg/d, for 14 days, then taper over 7 days.<sup>14</sup> Treatment with corticosteroids is believed to relieve acute pain, reduce vertigo, and speed healing of skin lesions.<sup>14</sup>

### BELL'S PALSY AND OTHER CAUSES OF FACIAL PARALYSIS

The incidence of Bell's palsy is 20 to 30 cases per 100,000<sup>15</sup> per year. The highest incidence is in the 70s age group.<sup>16</sup> The gender distribution is equal,<sup>15</sup> and Bell's palsy is more common in diabetic and pregnant women.<sup>17</sup>

Bell's palsy is a term for *idiopathic* facial nerve paralysis, whereas it is now widely believed that reactivation of herpes simplex virus type 1 (HSV-1) accounts for most Bell's palsy cases.<sup>18</sup> Some cases of Bell's palsy may be attributed to ischemia and microvascular disease from diabetes mellitus, analogous to diabetic mononeuropathy.<sup>16</sup> Bell's palsy is believed to account for 60% to 75% of all cases of unilateral facial paralysis.<sup>16</sup> **Box 3** outlines the clinical presentation of Bell's palsy. Other causes of facial paralysis are numerous and are listed in **Table 2**.

The facial nerve (cranial nerve VII) supplies innervation to all the muscles of facial expression, parasympathetic fibers to the lacrimal and salivary glands, and sensory fibers for taste to the anterior two-thirds of the tongue.

Stroke and Bell's palsy are the 2 most common causes of *abrupt onset* of facial paralysis.<sup>16</sup> One study noted that most Bell's palsy patients first noticed their paralysis on awakening in the morning.<sup>15</sup> Patients first notice that they cannot close the affected eye and that water spills from the mouth when they try to drink. The study theorized that ischemia of facial nerve increases at night (causing edema of cranial nerve VII, entrapment of cranial nerve VII in the bony facial nerve canal, and reactivation of HSV-1 in the geniculate ganglion). Ischemic brain and heart disease incidence also appear to be increased at night.<sup>15</sup>

A familial tendency in Bell's palsy has been reported in 2.4% to 28.6% of cases.<sup>19</sup> Patients with familial Bell's palsy have *recurrences* of facial paralysis more often<sup>20</sup> and tend to have Bell's palsy at younger ages than other patients. A viral cause is not

**Box 3****The clinical presentation of Bell's palsy**

- Weakness or complete paralysis of all the muscles on one side of the face
- Disappearance of facial creases, nasolabial folds, and forehead furrows
- Drooping of the corner of the mouth
- Failure of the eyelids to close
- Eye rolls upward on attempt to close the eyelid (Bell's phenomenon)
- Eye irritation resulting from dryness to exposure and reduction of tear production
- Food and saliva may pool in the affected side of the mouth and may spill out from the corner of the mouth
- Preservation of facial sensation (although patients may have a perception of numbness from paralysis)
- Hyperacusis from paralysis of stapedius muscles (paralysis results in no dampening of vibration of the ossicles, and sounds are abnormally loud on the affected side); there is no hearing loss from Bell's palsy<sup>16</sup>

implicated in familial Bell's palsy. Proposed causes of familial Bell's palsy include an inherited anatomic abnormality of the facial canal, vascular risk factors (eg, diabetes mellitus, hypertension), and other immunogenetic factors. Familial Bell's palsy appears to have an autosomal dominant pattern of inheritance.<sup>19</sup>

Progression of facial paralysis that is *slow or gradual in onset*, progressive in nature, and without evidence of spontaneous recovery should raise the suspicion of tumor as the cause of facial paralysis. Benign and malignant neoplasms are estimated to account for about 5% of all cases of peripheral facial nerve paralysis, with parotid carcinomas the most common.<sup>21</sup>

Four historical findings appear to be important in recommending surgical exploration of the facial nerve for tumor involvement:<sup>22</sup>

**Table 2**  
**Etiologies of facial paralysis**

<b>Category</b>	<b>Examples</b>
Idiopathic	Bell's palsy
Congenital	Traumatic birth, Moebius syndrome, trisomies 13 and 18
Trauma	Basal skull fracture, penetrating injury to middle ear, barotrauma
Neurologic	Amyloidosis, multiple sclerosis
Metabolic	Pregnancy, diabetes mellitus, hypertension, hyperthyroidism
Neoplastic	Parotid tumors, cholesteatoma, glomus tumor, cranial nerve tumors, metastatic carcinomas, neurofibromatosis
Vascular	Anomalous or ectatic vessels, carotid artery aneurysm
Infection	Otitis externa, otitis media, mastoiditis, Lyme disease, numerous other infections
Iatrogenic	Mandibular block anesthesia, intranasal influenza vaccine, parotid surgery, mastoid surgery, dental procedures, post-immunization, post-T&A
Autoimmune	Temporal arteritis, Thrombotic thrombocytopenic purpura, Periarthritis nodosa, Guillain-Barré, multiple sclerosis, sarcoidosis

- Progressive and prolonged pattern of paralysis without recovery
- History of pain
- Involvement of other cranial nerves
- History of regional skin cancer

Imaging consisting of enhanced MRI and contrast-enhanced CT of the temporal bone, brain, neck, and parotid gland should be done when tumor is suspected. About 85% of patients with Bell's palsy have some degree of spontaneous recovery of facial function by 3 weeks.<sup>17</sup> One author has recommended that imaging should be considered if there is no spontaneous recovery of facial paralysis by 12 weeks and tumor is suspected.<sup>21</sup>

Evaluation of the patient with facial paralysis is summarized in **Box 4**.<sup>17</sup>

Patients with older age, hypertension, impairment of taste, pain other than in the ear, and complete facial weakness have a poorer prognosis for recovery of facial nerve function.<sup>16</sup> Corticosteroids are usually given for Bell's palsy. A common regimen is prednisone, 60 mg/d, tapering over 10 days.<sup>17</sup> Another proposed regimen is prednisone, 1 mg/kg daily, for 7 days, starting within 2 to 14 days after onset of symptoms.<sup>16</sup> One study showed significant improvement in the cure rate in diabetics when they were given intravenous prednisolone, 200 mg/d, for 2 days tapered to 70 mg by day 7.<sup>25</sup>

Antiviral drugs, given with corticosteroids, are believed to cause a slightly higher recovery rate than from corticosteroids alone.<sup>16</sup> Antiviral drug regimens studied include acyclovir, 400 mg 5 times per day, for 7 days and valacyclovir, 1 gm 3 times a day, for 7 days. No benefit has been demonstrated if treatment with antivirals is delayed more than 4 days after the onset of symptoms.

Surgical decompression of the facial nerve may be considered for patients who have persistent loss of function (ie, >90% loss on ENG at 2 weeks). This degree of degeneration of the facial nerve appears to be irreversible after 2 to 3 weeks. Patients may have several complications from facial nerve decompression, including permanent unilateral deafness.<sup>16</sup> The role of surgical decompression remains controversial.<sup>26</sup>

Referral to a neurologist should be carried out if facial paralysis is bilateral or not improving within 2 to 3 weeks after onset.<sup>16</sup>

Because patients with facial paralysis are usually unable to close the affected eye, they are at risk for irritation and dryness of the globe, leading to corneal abrasion and ulceration. To minimize this risk, patients should be instructed to use artificial tears several times during the day, use lubricating ointment in the eye at bedtime, and tape the eyelid shut at night.

## FACIAL PARALYSIS IN CHILDREN

Facial paralysis in children is different than in adults in that up to 70% of cases have identifiable secondary causes compared to 25% to 40% in adults.<sup>27</sup> Except in areas endemic for Lyme disease, the most common cause of facial paralysis in children is acute otitis media. Paralysis of facial muscles usually begins 5 to 8 days after the onset of signs and symptoms of acute otitis media.

Lyme disease is more common as a cause of facial paralysis in children in endemic areas. Facial nerve paralysis from Lyme disease may be unilateral or bilateral and can last up to 2 months. Painless and nontender facial swelling and erythema often develop before the onset of facial paralysis.<sup>28</sup> **Box 5** outlines the evaluation of a child with facial paralysis, and **Box 6** outlines the management of the child with facial nerve palsy.

Facial paralysis in a newborn has a 78% to 90% association with birth trauma (forceps delivery), birth weight greater than 3500 g, or prematurity. Fortunately, traumatic lesions are much more likely to recover.<sup>30</sup>



**Box 4****History, physical examination, laboratory, imaging, and electrodiagnostic testing in the evaluation of the patient with facial paralysis**

## History:

- Abrupt onset? (favors Bell's Palsy)
- Gradual onset over weeks? (favors neoplasm)
- History of tick exposure, rash, or arthralgias? (Lyme disease)
- History of ear and facial pain, hearing loss, and taste disturbance with facial paralysis? (HZ oticus)
- Recent history of acute otitis media (more gradual onset of paralysis with fever), otitis externa, or mastoiditis?
- Bilateral facial weakness? (favors polyneuropathies, eg, Guillain-Barré or sarcoidosis)
- History of other central nervous system lesions? (eg, multiple sclerosis, stroke, tumor, myasthenia gravis, and often have other systemic neurologic findings)
- Family history of facial paralysis? (consider familial facial paralysis)
- History of recent trauma to ear or skull?
- History of regional skin cancer?
- History of diabetes mellitus?
- History of pregnancy or recent pregnancy, especially with eclampsia?
- History of facial, dental, or parotid surgery?
- Recent infections? (varicella, mumps, mononucleosis, influenza, cat scratch, HIV, many others)
- History of thalidomide, ethylene glycol ingestion, alcoholism, arsenic intoxication, carbon monoxide poisoning?
- History of autoimmune disease? (temporal arteritis, sarcoidosis, multiple sclerosis, periarteritis nodosa, thrombotic thrombocytic purpura)
- History of recent intranasal influenza vaccine?

Physical examination<sup>17</sup>:

- Inspection of ear canal (vesicles, granulation tissue, edema)
- Inspection of tympanic membrane (cholesteatoma, effusion)
- Evaluation of peripheral nerve function of the extremities
- Evaluation of other cranial nerves
- Palpation of parotid glands (tumor)
- Ask the patient—"Close your eyes"—to test upper facial muscles. Failure to close the ipsilateral eye indicates a peripheral lesion of cranial VII. Central lesions of cranial VII occur in the pons of the contralateral hemisphere or above, and the patient has intact upper facial muscles bilaterally and can close both eyes.<sup>16</sup>
- Ask the patient—"Show me your teeth"—to test the lower facial muscles. Denervation of the risorius muscle is indicated by drooping of one side of the mouth.<sup>16</sup>
- Hearing screen (audiometry, tuning fork, or finger rub test) for hearing loss or hyperacusis
- Taste test. Permanent taste loss occurs in patients with lesions proximal to the geniculate ganglion.
- Inspection of orbit for dryness and irritation. Exposure of the orbit from failure to close the eye.

Laboratory evaluation:

- Lyme titer (immunoglobulin [Ig]G, IgM), especially if recently in an endemic area or history of recent tick exposure
  - Serologic testing is often not sensitive enough in the first 2 weeks, and patients should be treated based on clinical suspicion. Patients who remain symptomatic for 6 to 8 weeks and are seronegative more likely have another diagnosis.<sup>23</sup>

Imaging in patients with facial nerve paralysis:

- Indications for enhanced MRI and CT with contrast of temporal bone, brain, neck, and parotid glands
  - No improvement in facial paralysis in 3 to 12 weeks
  - Onset of facial paralysis is gradual in onset (over weeks)
  - Bilateral facial weakness (multiple sclerosis, myasthenia gravis, or other central lesions)
  - Surgical decompression of the facial nerve is under consideration
  - Suspicion of tumor

Electroneuronography:

- ENG decline in facial muscle function is typically seen from days 4 to 10 from onset of facial paralysis.
- ENG is probably most useful when performed within 2 weeks after a complete loss of voluntary facial function<sup>16</sup> when surgical decompression is considered.
- Among patients who have 90% or more degeneration with the first 3 weeks, only 50% have good recovery of facial function.<sup>24</sup>

*Adapted from Tiemstra J, Khatkhate N. Bell's palsy: diagnosis and management. Am Fam Physician 2007;76:997–1004.*

The prognosis for recovery is based on House-Brackmann criteria (**Table 3**).<sup>31</sup> House-Brackmann criteria apply to both children and adults. The prognosis of a House-Brackmann grade of I or II is considered good, III or IV is moderate, and V or VI is poor. The prognosis is good if some recovery of facial nerve function is seen in the first 21 days after onset.

## CRANIAL NERVE NEURALGIAS

Before discussing specific cranial neuralgias, the pain from neuralgia will be described. Neuralgia is a form of neuropathic pain characterized by<sup>32</sup>

- Paroxysmal, brief (seconds to a few minutes), electric shocklike or lightninglike pain that follows a specific nerve distribution.
- No objective neurologic deficits in the distribution of the affected nerve.
- Attacks of pain provoked by nonpainful stimulation (allodynia) of trigger points or zones.
- A refractory period (without symptoms) following attacks; the duration of the refractory period shortens as the disease progresses.
- Attacks of pain tend to be stereotyped.

## SUPERIOR LARYNGEAL NEURALGIA (BRANCH OF CRANIAL NERVE X)

Superior laryngeal neuralgia is rare, and the gender distribution ratio is thought to be equal.<sup>3</sup> The superior laryngeal nerve is a branch of the vagus nerve (CN X) that runs

**Box 5****Evaluation of a child with facial nerve palsy**

## History

- Are there associated symptoms or acute problems? (eg, acute otitis media)
- Is there any systemic disease? (eg, HIV, diabetes, immunosuppression)
- Is there a history of tick bites?
- What is the birth history? (forceps delivery, weight >3500 g, prematurity)
- Is there a family history? (ie, familial Bell's palsy)
- Are any other congenital abnormalities present? (Moebius syndrome)

## Physical examination

- Careful examination of facial musculature (closing eyes, elevating eyebrows, frowning, smiling, showing the teeth, puckering lips, observation while crying in infants)
- Examination of ear, mastoid region, parotid glands, and neck
- Complete cranial nerve examination
- Otological examination: external ear, external auditory canal, pneumatic otoscopy, tuning fork tests
- Audiometry
- Tympanometry

## Imaging studies

- Probably indicated in most cases of acute facial nerve palsy in children
  - Temporal bone CT with contrast if there is a history of demonstrable mass, chronic otitis media, acute mastoiditis, previous mastoid surgery, or suspected fracture of temporal bone
  - MRI with and without contrast of brainstem, temporal bone, and parotid gland

## Laboratory

- Lyme (IgM, IgG) in endemic areas or history of recent travel to endemic areas
- Cerebrospinal fluid (CSF) examination for elevated white blood cell count and protein level and CSF antibodies (IgG, IgM) to *B. Burgdorferi*

**Box 6****Treatment of a child with facial nerve palsy**

- When idiopathic, spontaneous recovery of facial nerve function in children is 80% to 90% at 6 months and nearly 100% at 1 year<sup>29</sup>
- Children may be treated with antiviral drugs and corticosteroids, but the evidence is not as compelling as in the treatment of adults with Bell's palsy
- Eye care to prevent corneal abrasion and ulceration
- Acute otitis media: myringotomy with or without tube and second- or third-generation cephalosporin or amoxicillin-clavulanic acid
- Lyme disease: 30-day regimen of oral doxycycline (children 8 years old and older)
- HZ oticus: prednisone, 2 mg/kg/d for 5 days, followed by 5-day taper plus acyclovir or valacyclovir, 20 mg/kg 3 times a day, for 7 days<sup>29</sup>

<b>Grade</b>	<b>Characteristics</b>
I. Normal	Normal function in all areas
II. Mild dysfunction	Slight weakness noticeable on close inspection
May have slight synkinesis	Normal symmetry and tone at rest Forehead: moderate to good function Eye: complete closure with minimal effort Mouth: slight asymmetry
III. Moderate dysfunction	Obvious but not disfiguring difference between the 2 sides Noticeable but not severe synkinesis, contracture, or hemifacial spasm Normal symmetry and tone at rest Forehead: slight to moderate movement Eye: complete closure with effort Mouth: slightly weak with maximum effort
IV. Moderately severe dysfunction	Obvious weakness and/or disfiguring asymmetry Normal symmetry and tone at rest Forehead: no motion Eye: incomplete closure Mouth: asymmetric with maximal effort
V. Severe dysfunction	Only barely perceptible motion Asymmetry at rest Forehead: no motion Eye: incomplete closure Mouth: slight movement
VI. Total paralysis	No movement

Data from House JW, Brackmann DE. Facial nerve grading system. *Otolaryngol Head Neck Surg* 1985;93(2):146-7.

adjacent to the carotid bifurcation and innervates the cricothyroid muscle of the larynx.<sup>33</sup> Superior laryngeal neuralgia usually presents as attacks of lancinating pain lasting seconds to minutes, radiating from the side of the thyroid cartilage or pyriform sinus to the angle of the jaw and occasionally to the ear, more commonly on the left side.<sup>3</sup> A trigger zone is present superolateral to the thyroid cartilage. The painful paroxysms are provoked by swallowing, straining the voice, yawning, turning the head, stretching the neck, coughing, sneezing, speaking, or blowing the nose. During attacks, patients often describe an irresistible urge to swallow. Hoarseness and muteness may occur during the paroxysms.<sup>33</sup>

The cause of superior laryngeal neuralgia includes a precedent viral infection such as influenza; scarring from surgical procedures such as tonsillectomy and carotid endarterectomy, which leads to nerve compression; and chronic repetitive micro-trauma to larynx from singing, talking, or swallowing over a long period of time. The differential diagnosis of superior laryngeal neuralgia includes glossopharyngeal neuralgia, nervus intermedius neuralgia and carotidynia, a pain syndrome in which the carotid artery is tender due to migraine disorder or dissection of the carotid. Differentiating superior laryngeal neuralgia from carcinoma of the neck may require laryngoscopy, electromyography and CT of the larynx and laryngeal muscles.

Nerve block of the superior laryngeal nerve may be diagnostic and usually results in temporary relief. Carbamazepine may be helpful. Neurectomy is usually curative.

## TRIGEMINAL NEURALGIA

The incidence of trigeminal neuralgia is 4.7 per 100,000 per year in men and 7.2 per 100,000 per year in women. The age of onset is usually more than 50 years of age.<sup>34</sup> About 1% to 2% of patients with multiple sclerosis will have trigeminal neuralgia.<sup>35</sup>

The pain of trigeminal neuralgia is extreme, intense, sharp, shooting, and electric shocklike. It is felt in the skin or buccal mucosa. Attacks of pain are stereotyped<sup>35</sup> and can be triggered by light mechanical contact (eg, light touch, shaving, washing, chewing, contact with cold air) from a trigger zone. Trigger zones are usually very small and located on the face, nose, and lips. The duration of pain is usually a few seconds, but may last up to 1 to 2 minutes. The frequency of pain attacks is quite variable. The pain is unilateral in about 95% of cases.<sup>3,34</sup> No clinically evident neurologic deficit can be found. Attacks are rare during sleep. The V2 (maxillary) branch of the trigeminal nerve is more commonly affected than the V3 (mandibular) branch, which, in turn, is more commonly affected than the V1 (ophthalmic) branch.

A patient experiencing a trigeminal neuralgia paroxysm typically freezes in place with hands slowly rising to the area of pain on the face but not touching it. He or she then grimaces with an involuntary spasm of the facial muscles referred to as *tic douloureux*, and then either remains in this position or cries out in pain.<sup>34</sup>

Although a definitive cause of trigeminal neuralgia is not known, vascular loops of aberrant vessels causing compression of the trigeminal nerve appears to be an important cause. Demyelination of the trigeminal nerve is a frequent finding on pathology specimens. Secondary causes of trigeminal neuralgia are numerous, including multiple sclerosis, basilar artery aneurysm, tumor, and brainstem infarction.<sup>34</sup>

MRI of the brain is useful to look for plaques of multiple sclerosis, tumors, and vascular compression.<sup>35</sup> MR angiography (MRA) may also be useful to look for aberrant vascular loops.<sup>34</sup>

The differential diagnosis of trigeminal neuralgia is lengthy. Other causes of similar pain are listed with their distinguishing characteristics (**Table 4**).<sup>35</sup>

Carbamazepine is the drug of choice for trigeminal neuralgia. The usual starting dose is 100 mg 1 to 2 times a day, increasing by 100 to 200 mg every 3 days until pain relief is obtained. Most patients reach a maintenance dosage between 400 and 800 mg, although some patients need much more.<sup>34</sup> Patients often complain of drowsiness when starting on carbamazepine, but this usually lessens after several days.

Baclofen may be substituted for, or added to, carbamazepine. It may be used if the patient experiences significant adverse effects to carbamazepine (especially hematologic).

Other medications or treatments that may be useful include clonazepam, valproic acid, lamotrigine, oxcarbazepine, topiramate, botulinum toxin, and acupuncture.<sup>34</sup>

Neurosurgery is an option for patients who have not had satisfactory responses to medications.

## GLOSSOPHARYNGEAL NEURALGIA

Glossopharyngeal neuralgia is very rare. The incidence is 0.5 per 100,000 per year. It is usually seen in patients aged 60 years and older.<sup>3</sup>

The characteristic clinical presentation of glossopharyngeal neuralgia is paroxysms of severe, stabbing pain in the oropharynx radiating upward toward the ear. The pain can be felt deep in the throat or ear.<sup>3</sup> The duration of attacks usually range from several seconds to 1 minute. The frequency of attacks is usually several times to several dozen times daily.<sup>34</sup> Paroxysms may be triggered by swallowing, chewing, coughing, sneezing, speaking, yawning, certain tastes, and touching the neck or external auditory canal. Obviously this can interfere with eating.<sup>3</sup> The trigger zone

<b>Diagnosis</b>	<b>Distinguishing Characteristics</b>
Cluster HA	Pain lasts longer; orbital or supraorbital; autonomic symptoms
Dental pain	Localized; sensitive to hot and cold foods; abnormal oral examination
Giant cell arteritis	Persistent pain, jaw claudication
Glossopharyngeal neuralgia	Pain in tongue, mouth, or throat brought on by swallowing, talking, or chewing
Intracranial tumors	Other neurologic signs possible
Migraine	Longer-lasting pain, family history, photophobia, phonophobia
Multiple sclerosis	Eye symptoms, other neurologic symptoms
Otitis media	Pain localized to ear; abnormal ear examination
Paroxysmal hemicrania	Pain in forehead or eye; autonomic symptoms; responds to indomethacin
Post-herpetic neuralgia	Continuous pain, tingling, history of zoster; usually in ophthalmic division of V
Sinusitis	Persistent pain; associated nasal symptoms
SUNCT	Ocular or periocular, autonomic symptoms
TMJ syndrome	Persistent pain; localized tenderness, jaw abnormalities
Trigeminal neuropathy	Persistent pain; associated sensory loss

*Abbreviations:* SUNCT, shorter lasting unilateral neuralgiform, conjunctival injection, and tearing; TMJ, temporomandibular joint.

*Adapted from* Krafft R. Trigeminal neuralgia. *Am Fam Physician* 2008;77(9):1291–6; with permission.

for glossopharyngeal neuralgia may be in the preauricular or post-auricular area, neck, throat (tonsil or tonsillar fossa), and external auditory canal. Up to 2% of patients have syncope (from bradycardia or asystole) during an attack.<sup>34</sup>

Glossopharyngeal neuralgia may be confused with trigeminal neuralgia limited to the mandibular division (V3).<sup>3</sup>

The cause of glossopharyngeal neuralgia includes various anatomic abnormalities affecting the glossopharyngeal nerve including tumor, Chiari I malformation, infarction, neurovascular compression by the posterior inferior cerebellar artery, anterior inferior cerebellar artery, or other vessels and compression of cranial nerve IX by a calcified or elongated styloid process (Eagle syndrome).<sup>36,37</sup> Vertebral artery dissection can also cause glossopharyngeal neuralgia.

Application of local anesthetic to trigger point gives relief and may serve as a diagnostic aid.<sup>34</sup> The physical examination is usually normal, except for findings of a trigger point. A plain film may help identify an elongated styloid process.<sup>37</sup> An enhanced MRI may identify vascular compression or tumor.<sup>36</sup>

Treatment of glossopharyngeal neuralgia includes the use of carbamazepine and other anticonvulsants, as in the treatment of trigeminal neuralgia. Baclofen and gabapentin may also be helpful. If the cause proves to be from vascular compression of the glossopharyngeal nerve on MRI, vascular decompression may be indicated.

### **NERVUS INTERMEDIUS (GENICULATE) NEURALGIA**

The nervus intermedius is a small branch of the facial nerve (VII) that innervates the inner ear, middle ear, mastoid cells, eustachian tube, and part of the pinna of

the ear.<sup>4</sup> Neuralgia of the nervus intermedius is very rare, with an estimated incidence of 0.03 per 100,000 per year. It is seen most commonly in patients older than 50 years of age,<sup>3</sup> and middle-aged women seem to be affected more than men.<sup>4</sup>

The clinical characteristics of nervus intermedius neuralgia include paroxysms of intense, lightninglike, or burning pain limited to the depths of the ear.<sup>38,39</sup> The pain is occasionally located deeply in the face behind the orbits and posterior nasal cavity. Painful paroxysms typically last from a few seconds to not more than a few minutes. The frequency of attacks is quite variable. A trigger zone may be located on the posterior wall of the external auditory canal.<sup>38</sup> Patients may have a sense of bitter taste in their mouth during an attack.

Nervus intermedius neuralgia may be difficult to differentiate from glossopharyngeal neuralgia confined to the ear. Contrast-enhanced imaging of the brain focusing on the internal auditory meatus and MRA looking for ectatic or aberrant vessels, which may compress the nerve, may help in localization of the cause.

Treatment of nervus intermedius neuralgia includes carbamazepine and other anti-convulsants used in the treatment of trigeminal neuralgia. Neurosurgery may be necessary for patients not responding to medications.<sup>38</sup>

### RAEDER PARATRIGEMINAL SYNDROME

Raeder paratrigeminal syndrome is a rare neuralgia of the trigeminal nerve. The onset is usually in the middle-aged and there is a male preponderance.<sup>3</sup> Clinical characteristics include decreased sensation in the distribution of the trigeminal nerve, most commonly the ophthalmic division.<sup>40</sup> Weakness of the muscles innervated by the trigeminal nerve result in problems with mastication and swallowing. The neuralgic pain of Raeder paratrigeminal neuralgia is usually deep and boring and is not excruciating or continuously severe.<sup>3</sup> Patients also demonstrate unilateral oculosympathetic paresis, usually miosis and ptosis, similar to Horner syndrome. Unlike Horner syndrome, facial sweating is preserved.

Other cranial nerves may be involved (II, III, IV, V, VI).<sup>3</sup> The cause of Raeder paratrigeminal neuralgia may result from trauma or middle cranial fossa tumor. Treatment includes surgery if a mass lesion is present. Otherwise, analgesics may control the discomfort.

### SUMMARY

Discussion of localized cranial nerve neurologic syndromes becomes complicated due to the many types of peripheral nerve fibers (ie, general and special, visceral and somatic, afferent and efferent) and specialized functions of each cranial nerve. The tortuous courses of cranial nerves through foramina of the skull demonstrate vulnerability of cranial nerves to compression by adjacent anatomic structures. Cranial nerves may be involved in many disease processes. Because most of the cranial nerve neuralgias are relatively rare in primary care, sorting through the differential diagnosis can be confusing and difficult. It is important to recognize situations that require urgent attention so patients will receive optimal care.

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