

Headache Academy 2025

Other Cranial Neuralgias & Facial Pain Syndromes

Dr Tim Yates

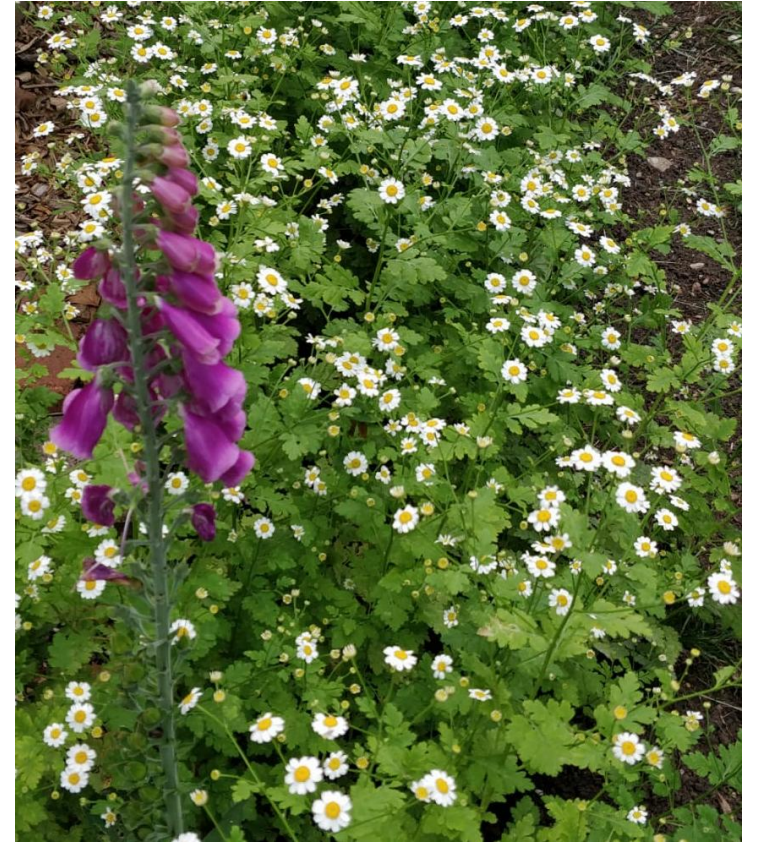
National Hospital for Neurology & Neurosurgery

London, UK



Declarations

- Pfizer – speaker fees
- Abbvie – educational support



To cover

- Familiar conditions
- Neuralgic v neuropathic
- Less familiar conditions

Consider migrainous orofacial pain

- Common
- Very often forehead/retro-orbital
- Less often V2 and/or V3 ($\approx 9\%$), rarely isolated facial pain ($< 1\%$) [Yoon, Cephalalgia 2010]
- Episodic or chronic
- Usually plenty of clues

Consider familiar neurological conditions

- Trigeminal neuralgia ICHD 13.1.1
 - V2 & V3 > V1, neuralgiform, refractory period
- SUNA ICHD 3.3
 - V1 > V2 & V3, short duration, neuralgiform, autonomic features
- Paroxysmal hemicrania ICHD 3.2
 - Peri-orbital, intermediate duration, autonomic features
- Cluster headache ICHD 3.1
 - Peri-orbital, longer lasting, autonomic features
 - Can be lower face [Bahra, Acta Neurol Scand 2004]
- Painful optic neuritis ICHD 13.6
 - Objective evidence of the condition (clinical, imaging, electrophysiological etc)
 - Worse on eye movement

Consider other familiar conditions

- Headache attributed to acute angle-closure glaucoma (ICHD 11.3.1)
- Sinusitis
 - Avoid unless prominent features of infection / no better diagnosis [Landmark trial]
- Headache attributed to temporomandibular disorder (ICHD 11.7)
 - Group of disorders, joint, myofascial or both
 - Pain can be widely distributed, aching and shooting, usually asymmetrical
 - Common, $\approx 1/15$ aged 20-40 [Slade, J Pain 2013]
 - Comorbid with other pain and mood disorders
 - More common in hypermobility
 - Ask: bruxism, clicking/locking, TMJ tenderness, exacerbations with chewing
 - Appliance, physio, CBT, treat comorbidities, TCA, Botox, surgery

Consider less familiar conditions

A thick, wavy orange line that spans the width of the slide, positioned below the main text.

Neuralgic v neuropathic

Neuralgic

- “an affection of one or more nerves causing pain which is usually of an intermittent but frequently intense character” – Chaussier, 1801
- Paroxysms main feature
- Electric shock-like / shooting
- Often triggered
- R_x: more anti-epileptics, baclofen

Neuropathic

- Continuous, more than recurrent
- Paroxysms not main feature
- Burning, aching, squeezing, tingling > shooting
- Sensory change: numbness, allodynia, hyperaesthesia
- “Post-herpetic neuralgia” is usually more neuropathic
- R_x: more gabapentinoids, TCAs, SNRIs

Diagnostic complications

- Reliance on history to make a diagnosis
- More than 1 type of pain may co-exist
 - eg migraine + trigeminal neuralgia + temporomandibular joint disorder
- Local pathology can mimic primary syndromes
 - Inflammation, infection, neoplasm
- Conditions may evolve
 - Trigeminal neuralgia into “TN with background pain” then into painful trigeminal neuropathy



Blind men and the Elephant, Katsushika Hokusai 1760-1849

Cranial neuralgias and painful neuropathies

- Trigeminal
- Glossopharyngeal
- Superior laryngeal
- Nervus intermedius
- Occipital
- Persistent idiopathic facial pain
- Neck tongue syndrome
- Central neuropathic pain
- Periorbital syndromes
- Oral/dental conditions

Painful trigeminal neuropathy – ICHD 13.1.3

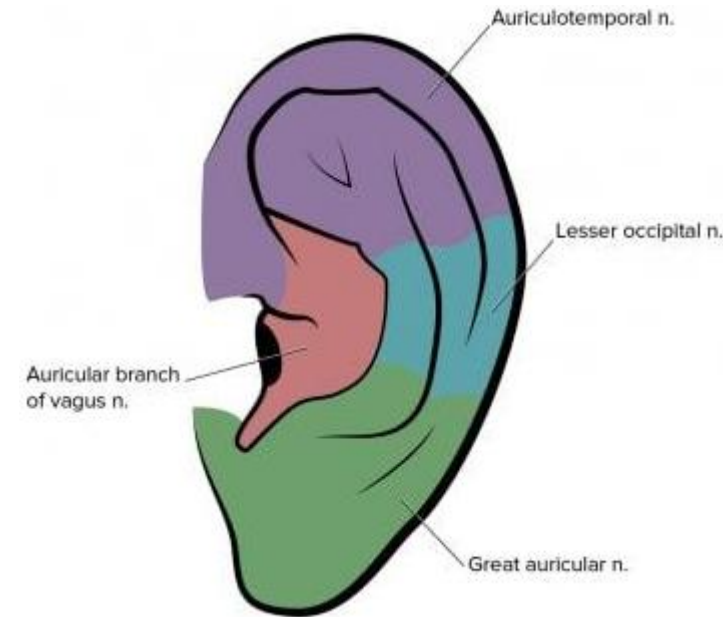
- Neuropathic symptoms in ≥ 1 branch of trigeminal nerve
- Requires positive/negative sensory symptoms/signs
- Significant reduction in QoL
- Look for causes
 - Causes of trigeminal neuralgia
 - Treatment of trigeminal neuralgia – decompressive, ablative
 - Viral infection, herpetic/post-herpetic
 - Traumatic, if within 6 months of event
 - Anything, including fractures, dental work, surgical/ablative treatments of neuralgia, sinus surgery, tonsillectomy
 - Inflammatory eg MS
 - Vasculitic/connective tissue (very often bilateral)
 - Neoplastic/CLIPPERS
 - Classify as idiopathic after extensive search

Persistent idiopathic facial pain – ICHD 13.12

- Previously “atypical facial pain”
- Can be similar to painful trigeminal neuropathy
- Neuropathic symptoms, poorly localised, often triggered by stress
- Often unilateral, spreading bilaterally, occasionally beyond the face
- If mainly teeth, “atypical odontalgia”/“persistent idiopathic dental pain”
- >2 hours a day for >3 months
- **Usually unconvincing sensory symptoms/signs**
- Frequent significant psychosocial comorbidities
- Exclude other causes, including dental

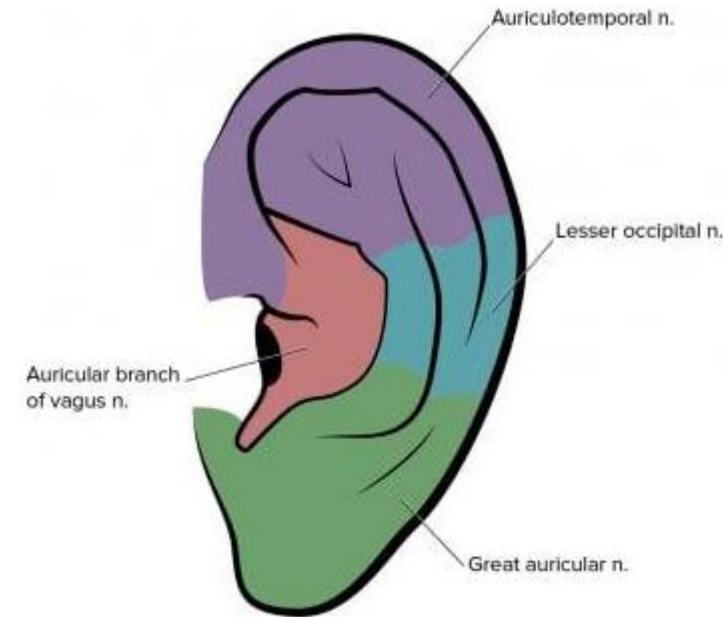
Glossopharyngeal neuralgia – ICHD 13.2.1

- Much less common than trigeminal or occipital neuralgia, $\approx 0.2-0.7/100,000$
- Neuralgia in distribution of sensory branches of CN IX, and auricular and pharyngeal branches of CN X
 - Ipsilateral oropharynx, palatine tonsil
 - Posterior $\frac{1}{3}$ tongue
 - Part of concha and back of pinna
- Often felt beneath angle of jaw, radiates to face and shoulder
- Triggered by swallowing, talking, coughing
- 2-120 seconds, severe, can lead to weight loss
- Rarely vagal symptoms, even asystole (ie associated collapse)
- **Expect no sensory signs / no change in gag reflex**
- May relapse and remit like trigeminal neuralgia
- Can coexist with trigeminal neuralgia



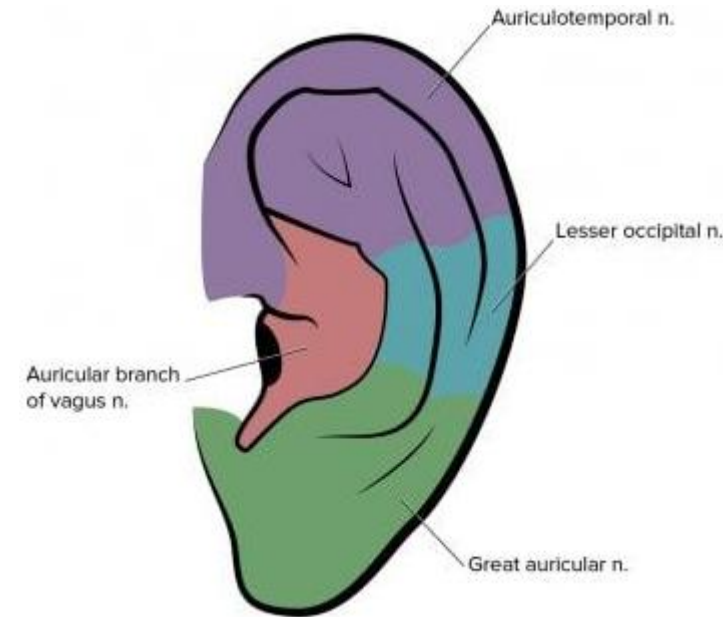
Glossopharyngeal neuralgia – ICHD 13.2.1

- Topical lidocaine abolishing symptoms suggestive of diagnosis
- Classical, idiopathic or secondary
- Look for causes
- Investigations
 - Screen for vasculitic/CTD/infective
 - MRI brain: vascular compression from posterior inferior cerebellar artery > anterior inferior cerebellar artery > vertebral artery; SOL/inflammatory lesions
 - MRI soft tissues of neck: for secondary cause of neuralgic pain in tongue, pharynx, parapharyngeal region, neck
 - CT neck: elongated styloid process ie Eagle syndrome
 - Ambulatory ECG where necessary
- Predictors of poor prognosis: bilateral pain, severe attacks, idiopathic aetiology [Zheng, J Neurosurg 2022]



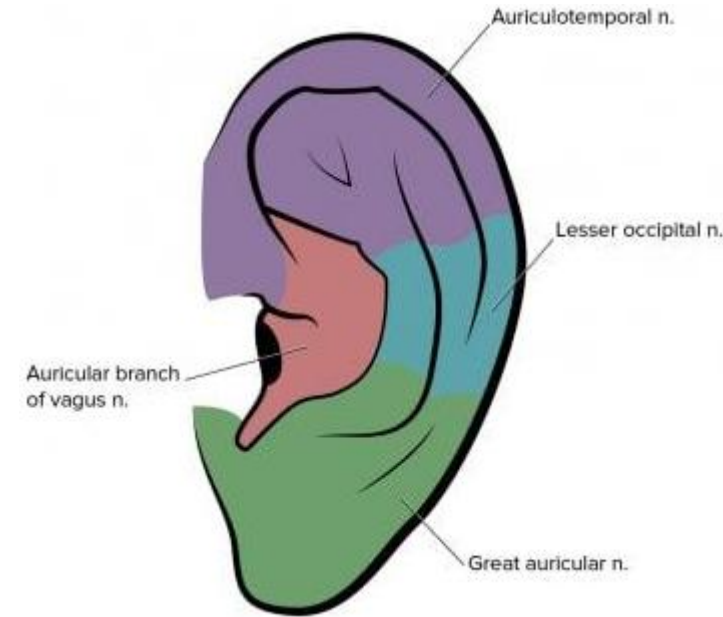
Glossopharyngeal neuralgia – differentials

- Confused with trigeminal neuralgia
- Auricular GPN difficult to distinguish from nervus intermedius neuralgia
- Superior laryngeal neuralgia with swallowing but around vocal cords
- Pain from
 - Acid reflux
 - Oral tumours or infections
 - Referred pain from a carotid body tumour



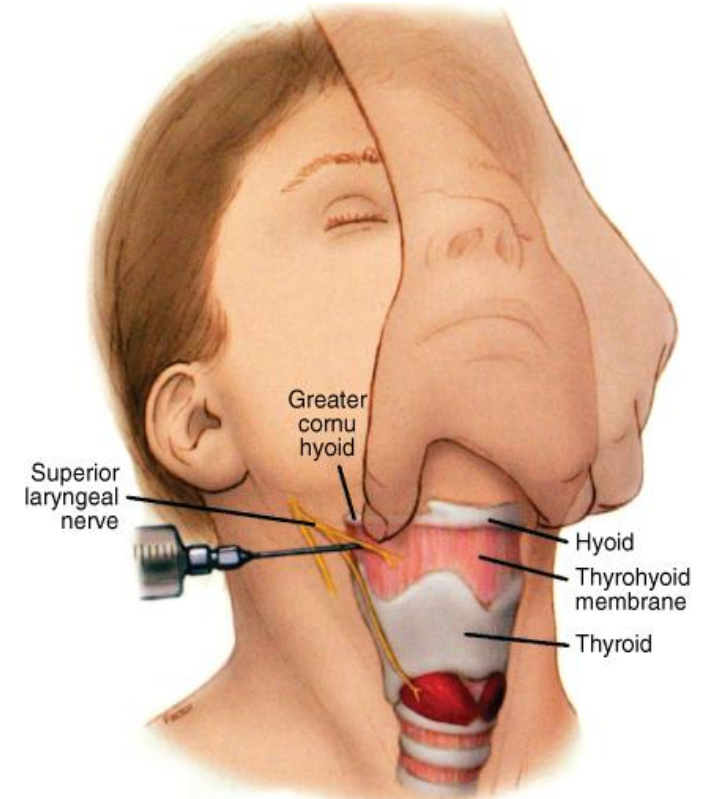
Glossopharyngeal neuropathy – ICHD 13.2.2

- Continuous/nearly continuous pain in same distribution
- Main differences from the neuralgia
 - Brief paroxysms may be superimposed, but are not the predominant pain
 - Sensory deficits may be present in the ipsilateral posterior part of the tongue and tonsillar fossa, and the gag reflex may be weak or missing



Superior laryngeal neuralgia

- Differential of glossopharyngeal neuralgia
- Rare
- Dull aching pain in throat radiating to ear with neuralgic exacerbations
- Triggers: talking, swallow, yawn, cough
- Presence of trigger point diagnostic
- Associated with surgery, inflammation, laryngeal diverticulum

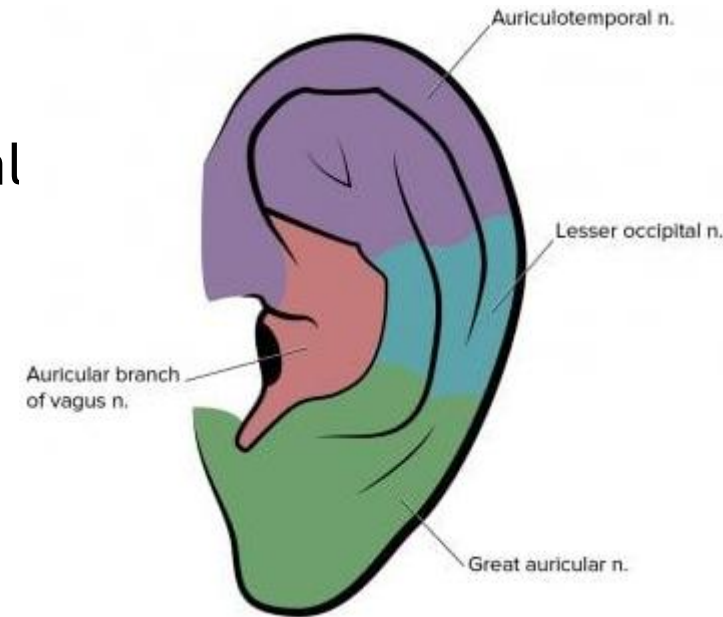


Source: Hung OR, Murphy MF: *Management of the Difficult and Failed Airway*, 2nd Edition: www.accessanesthesiology.com

Copyright © The McGraw-Hill Companies, Inc. All rights reserved.

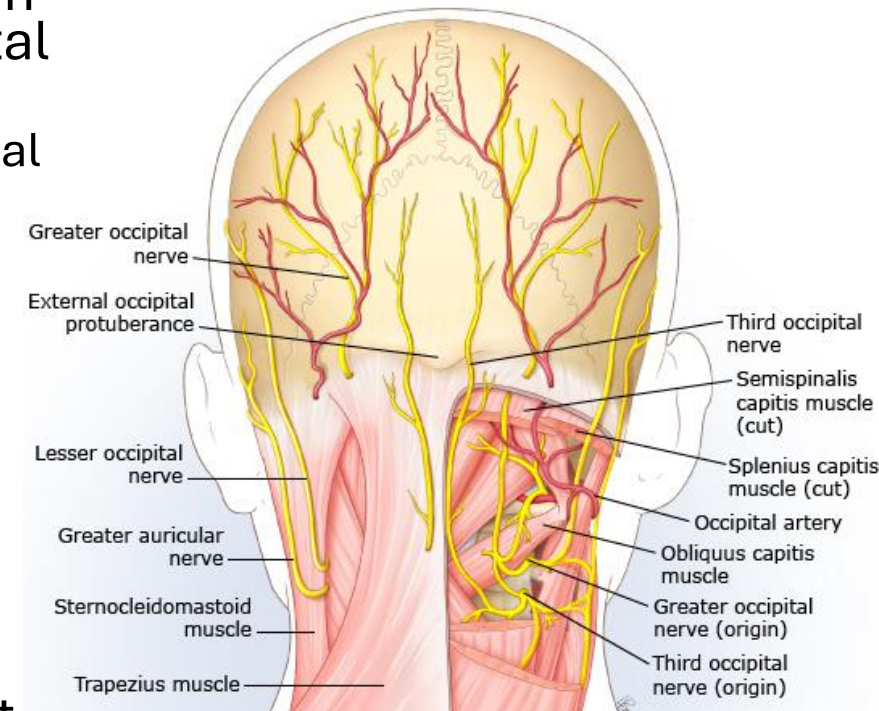
Nervus intermedius neuralgia – ICHD 13.3.1

- Rare
- Episodes of neuralgic pain felt deeply in the auditory canal
- Can affect mastoid process, soft palate, temple or the angle of the jaw
- May be associated with salivation or lacrimation
- Seconds to minutes
- Usually triggered by stimulus affecting same area
- Innervation of external ear complex and overlapping: trigeminal (auriculotemporal), facial (nervus intermedius), glossopharyngeal, vagus
- Investigate as per principles of other neuralgias, looking for vascular contact, tumour, inflammation, history of infection, etc
- Diagnose neuropathic form based on character of pain



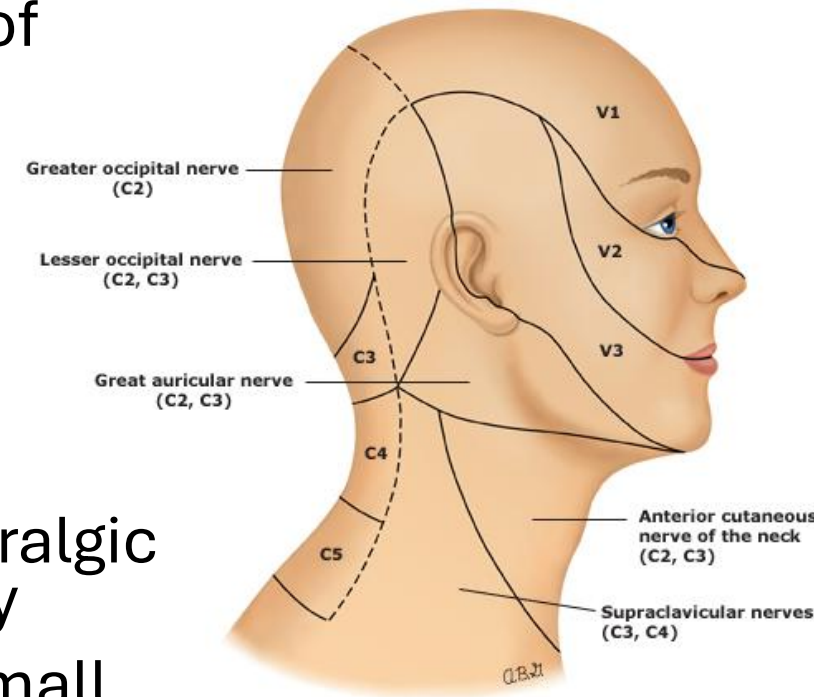
Occipital neuralgia – ICHD 13.4

- Rarer than some clinicians think
- Episodes of severe neuralgic pain at the posterior scalp, in the distributions of the greater, lesser and/or third occipital nerves, usually unilateral
 - Rarely forehead/orbital area via trigeminocervical interneuronal connections in the trigeminal spinal nuclei
- Commonly allodynia in affected area and trigger pain emergence of greater occipital nerve
- Often accompanied by diminished sensation or dysaesthesia in area
- Seconds to minutes
- Pain should be eased at least temporarily by local anaesthetic block of the affected nerves eg a GON/LON
- Causes: cervical pathology, scalp myofascial entrapment
- Consider whether pain may arise from disorder of atlantoaxial/upper zygapophyseal joints or tender trigger points in neck muscles or their insertions



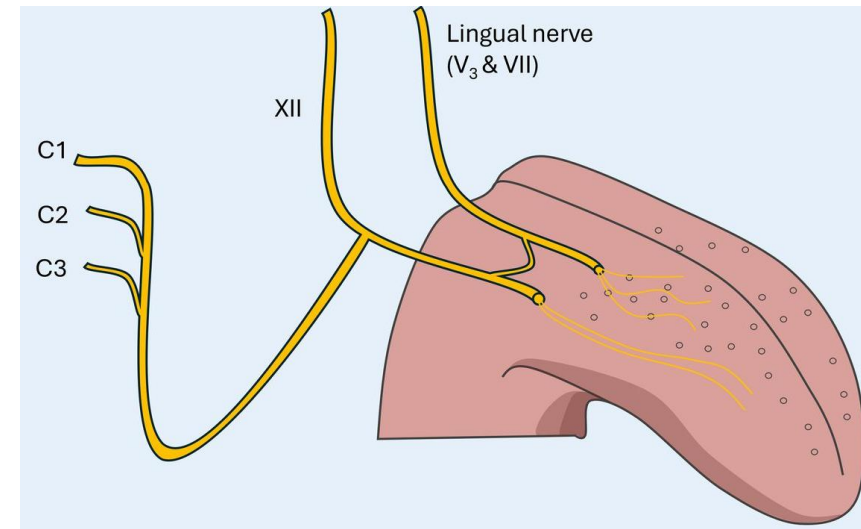
Occipital neuralgia – ICHD 13.4

- Response to anaesthetic block eg GON/LON part of diagnostic criteria
 - Does not confirm diagnosis
 - Periodic blocks often helpful
 - Cervical joint blocks can be used in some
- Use agents for neuralgic/neuropathic pain [Tepper, Continuum 2018]
- Botox might be an option with longer benefit than blocks [Kapural, Pain Pract 2007], might only improve neuralgic component [Taylor, Headache 2008] but small series only
- Pulsed radiofrequency, better initial response in small series, benefit lost over 3-6 months
- Surgical decompression, encouraging small series
- ONS – limited data



Neck tongue syndrome – ICHD 13.5

- Sharp, unilateral upper neck and/or occipital pain with ipsilateral dysaesthesia of the tongue
- Seconds to 1 minute
- Triggered by fast, sudden axial rotation of the head to either side
- Associations: upper limb radicular symptoms, jaw pain, anterolateral neck pain, sensation of choking, ear pressure, dysarthria, nasal congestion, sialorrhea, lingual pseudoathetosis, lingual paralysis, and retroauricular numbness
- Onset often in early teenage
- Joint abnormality irritates upper spinal roots (esp C2), sensory and motor connections to tongue
- Differentials: cervical/vertebral artery pathology, migraine, ligamentous insufficiency, and Chiari



Central neuropathic pain – ICHD 13.13

- Unilateral or bilateral craniocervical pain of central origin, with variable presentation and with or without sensory changes. Depending on the cause, it may be constant or remitting and relapsing.
- Involvement of ascending projections of the trigeminal nuclei, cervical spinothalamic pathways, areas involved in cortical processing eg thalamus, lateral medulla

Periorbital syndromes

- Headache attributed to ischaemic ocular motor nerve palsy ICHD 13.7 – “pain with microvascular IIIrd”
 - Continuous unilateral frontal/periorbital with symptoms/signs of ischaemic paresis of one or more of the ipsilateral IIIrd, IVth, VIth cranial nerves
- Tolosa-Hunt syndrome ICHD 13.8
 - As ICHD 13.7, but evidence of granulomatous disease; usually resolving with steroids
- Paratrigeminal oculosympathetic (Raeder’s) syndrome ICHD 13.9 - “classic painful IIIrd”
 - Constant V1 pain, ipsilateral to Horner’s, aggravated by eye movement
- Recurrent painful ophthalmoplegic neuropathy ICHD 13.10 - “ophthalmoplegic migraine”
 - Recurrent attacks of paresis of 1+ ocular nerves (commonly IIIrd) with ipsilateral headache without structural pathology except thickening of the affected nerve
- Trochlear headache ICHD 11.3.4
 - Not uncommon, unilateral, usually worse with supraduction; inflammation of the trochlea and/or sheath of superior oblique muscle
- Headache attributed to ocular inflammatory disorder 11.3.3
 - eg iritis, uveitis, scleritis or conjunctivitis

Oral/dental conditions

- Burning mouth syndrome (ICHD 13.11)
 - Intraoral burning or dysaesthetic sensation, apparently superficial; variable in intensity
 - Usually bilateral; most common site is tip of tongue
 - Daily >2 hours/day over >3 months
 - Worse with hot/cold, spicy food, stress
 - Subjective dryness of the mouth, dysaesthesia and altered taste reported
 - Normal intraoral appearances and sensation
 - Exclude eg Candidiasis, lichen planus, causes of sicca (inc Sjögren), medication induced, anaemia, diabetes, deficiencies: zinc, B12, folic acid
 - Menopausal females over-represented
 - Some studies show comorbid psychosocial and psychiatric disorders
 - Possibly centrally mediated
- Headache attributed to disorder of the teeth (ICHD 11.6)
 - Alternate pain loci: pulpal, periodontal, gingival, oral mucosal, salivary gland, jaw bone
 - Consult specialist dentist or maxfax clinician

Summary

- Ensure you consider the common and familiar disorders
- Bear in mind the rare
- Diagnose according to the distribution, character and associated features
- Differentiate the neuralgic & neuropathic
- Investigate for secondary causes
- Select treatment, escalating for the refractory
- Low bar to involve ophthalmologists, dentists, maxfax, spinal surgeons to help with diagnosis
- Monitor the condition over time

