CLINICAL PRACTICE GUIDELINE: Emergency Department

# Seventh Cranial Nerve Palsy

Disclaimer: This Clinical Practice Guideline ('CPG') was written for use in The Royal Victorian Eye and Ear Hospital Emergency Department. It should be used under the guidance of an Ophthalmology or ENT registrar. If clinical advice is required, please contact the Eye and Ear Admitting Officer for assistance: EYE: +61 3 9929 8033; ENT: +61 3 9929 8032. Links to internal Eye and Ear documents cannot be accessed from the website CPG.

See also: Herpes Zoster Ophthalmicus, Temporal Arteritis (TA)

## Description:

Seventh ($7^{th}$) nerve palsy is a dysfunction of the facial nerve (CN7) which supplies the facial muscles, taste to the anterior 2/3 of the tongue, parasympathetic supply to head and neck ganglia. It can be due to either a central or peripheral lesion with partial or complete facial muscle weakness.

### Red Flags:

- Suspect upper motor neuron (UMN), (e.g. stroke) lesion if ipsilateral sparing of forehead movements
- Associated neurologic signs require further investigation
- Suspect temporal arteritis (TA) if associated symptoms and age over 60
- Beware potentially blinding corneal exposure if poor lid closure.

### Aetiology:

Idiopathic:
Bell's palsy – Isolated unilateral lower motor neuron (LMN) lesion. May be related to herpes simplex virus (HSV) activation. Diagnosis of exclusion. See Table 1.

Inflammation of the peripheral nerve:
Ramsay-Hunt syndrome – Acute facial neuropathy from adjacent varicella zoster (HZV) infection with vesicular rash of the ear canal or oropharynx. In the absence of a rash it is termed zoster sine herpete.

Other causes include: otitis media, infectious mastoiditis, parotitis, sarcoidosis, diabetic mononeuropathy, cytomegalovirus, Epstein-Barr virus, HIV seroconversion.

Compression of peripheral nerve:
Trauma (including surgical injury or post-operative swelling), vestibular schwannoma or skull base tumour, facial nerve tumour, cholesteatoma, parotid tumour.

Damage to facial nucleus, fascicle or motor cortex:
Cortical stroke, pontine infarction, demyelinating disease, local infiltration by tumour, TA.

Neuromuscular pathology:
Myasthenia gravis, myotonic dystrophy, Guillain-Barre syndrome.

## How to Assess:

### History:

- Past history of facial weakness, stroke, malignancy
- Recent trauma to head or face, or recent viral infection
- Facial droop/weakness: evolution. Bell's palsy usually comes on quickly (over hours, up to a few days)
- Painful watery eye, blurred vision
- Facial or ear pain
    - Severe pain is suggestive of HZV
    - Mild pain in the face or behind the ear may occur in Bell's palsy
- Hyperacusis and/or altered taste
    - Suggestive of a lesion proximal to the chorda tympani
    - Both are common in Bell's palsy
- Other neurological symptoms
    - Limb weakness/parasthesia
    - Diplopia – consider intracranial tumours causing palsy of cranial nerves III, IV, VI
    - Deafness, tinnitus, vertigo – consider Ramsay-Hunt, rarely cerebellopontine angle tumour (e.g. vestibular schwannoma)
    - Sensory loss – consider cranial nerve V lesion
- Symptoms of TA: headache, jaw claudication scalp tenderness, polymyalgia rheumatica, weight loss, etc. Refer to TA CPG

**Examination:**

- Facial muscle weakness
  - House-Brackmann score (see Table 2)
- Ocular examination
  - Visual acuity, pupils
  - Extra-ocular movements
  - Lids/Cornea
    - Orbicularis function – lid closure
      Document lagophthalmos on gentle and forced lid closure (mm)
    - Levator function
    - Bell's phenomenon (eyeball rolls upward on attempted eyelid closure by patient in normal Bell's phenomenon)
    - Corneal exposure: conjunctival injection, fluorescein staining-punctate epithelial erosions (PEE), epithelial defect
    - Corneal sensation (V1)
  - Temporal artery: non pulsatile, tender – suggestive of TA
- Ear examination
  - Inspect for vesicular rash – suggestive of Ramsay-Hunt syndrome
  - Canal
  - Tympanic membrane and middle ear
- All cranial nerves: including facial sensation (V1, V2, V3)
- Signs of 7$^{th}$ nerve aberrant regeneration (indicates chronicity) - gustatory sweating, co-contraction of ipsilateral facial muscles (synkinesis) or involuntary tearing on the affected side when eating (crocodile tears)
- Upper/lower limb neurological examination

Table 1: Table comparing features typical of Bell's palsy and features that may suggest alternative pathology

| | Features typical in Bell's palsy | Features suggestive of alternative pathology |
|---|---|---|
| **Onset** | Rapid – over hours to 3 days | Insidious onset |
| **Pain** | Mild facial or post-auricular pain only | Headache or severe facial pain |
| **Associated neurology** | Isolated unilateral cranial nerve VII palsy (Note hyperacusis and altered taste commonly occur from chorda tympani involvement) | Bilateral disease, diplopia, paraesthesia, other weaknesses, deafness, tinnitus, vertigo |
| **Systemic features** | Nil | Headache, loss of weight, night sweats, fever, fatigue, rash |
| **Recovery** | Starts to recover over first 3 weeks | No improvement seen within first 4 weeks |

Table 2: House-Brackmann Scale for facial nerve grading system

| Grade | Findings |
|---|---|
| **I** | Normal symmetrical function |
| **II** | Complete eye closure with minimal effort<br>Slight asymmetry of face noticed on maximal effort |
| **III** | Complete eye closure<br>Strong but asymmetrical mouth movement<br>Obvious weakness, but not disfiguring |
| **IV** | Incomplete eye closure<br>Asymmetry of mouth with maximal effort<br>Severe synkinesis, mass movement or spasm |
| **V** | Incomplete eye closure<br>Motion barely perceptible<br>Usually no synkinesis, contracture or spasm present |
| **VI** | No movement detectable and loss of tone |

**Investigation:**

- Swab for viral PCR (including HZV) if rash
- Laboratory investigations for TA, sarcoidosis, neuromuscular etiology if history suggestive
- Consider CT scan if recent history of head or facial trauma, facial mass or surgery
- Consider MRI brain if suspect central lesion. Suggestive clinical features:
    - Gradual onset greater than a few days
    - Sparing of frontalis muscle
    - Any other neurological deficit
- Please remember Bell's palsy is a diagnosis of exclusion

## Acute Management:

Note: all patients must be seen by ophthalmology and ENT

**Systemic:**

- Steroids – Prednisolone 1 mg/kg orally (up to 75 mg) daily for 5 days
    - Indications:
        - Ramsay-Hunt syndrome
        - Bell's palsy
        - Other inflammatory pathologies (in consultation with ENT unit)
    - Steroids should be initiated within 48 - 72 hours of onset of symptoms
- Antivirals – Valacyclovir 1,000 mg TDS for 7 days
    - Indications:
        - Ramsay-Hunt syndrome
        - Bell's palsy (Optional)
          Note: Routine antiviral use for Bell's has been proposed due to possible association with herpes simplex infection, however this is controversial.

**Ocular:**

- Consider for patients with severe facial weakness (House-Brackmann IV-VI)
- All patients with lagophthalmos:
    - Preservative free ocular lubricants (frequency and viscosity based on degree of corneal exposure)
    - Nightly: paraffin based lubricating ointment
    - Consider lid closure with tape/pad. Please note that this can be counterproductive as it is difficult to achieve complete eyelid closure with tape/pad.

Table 3: Corneal exposure risk

| Corneal Exposure Risk | Possible Signs | Suggested Management |
|---|---|---|
| **Mild** | Few PEE<br>Eyes white<br>Complete forced lid closure<br>Minimal lagophthalmos | Regular ocular lubricant drops (6x per day)<br>+/- Nocte lubricant ointment<br>Avoid environments that aggravate dryness |
| **Moderate** | Significant PEE<br>Ocular injection<br>Lagophthalmos 1 – 2 mm | Preservative-free ocular lubricant drops 2-hourly<br>Nocte lubricant ointment<br>Tape lid closed overnight<br>Consider goggles to keep ocular humidity |
| **Severe** | Epithelial defect<br>Widespread PEE<br>Lagophthalmos ≥ 3 mm<br>Reduced corneal sensation<br>Corneal thinning<br>Poor Bell's phenomenon | Preservative-free ocular lubricant drops 1-hourly<br>Nocte lubricant ointment<br>Tape lid closed overnight<br>Consider tarsorrhaphy |

**Dental:**

Indications: any patients with inability to close mouth. Significant oral dryness can result in increased risk of caries.

Prognosis in Bell's palsy:

- Weakness may worsen in the first three weeks
- Most (85%) will start to recover within three weeks. 71% of patients with Bell's palsy have full recovery by 12 months, many of which occur within 3 months. Patients with incomplete paralysis are highly likely to completely recover (94%). Patients with more severe initial weakness (House-Brackmann III – VI) are likely to have ongoing long-term dysfunction.
- Recurrence of Bell's palsy occurs in approximately 7% of patients within 10 years
- Some patients may recover with aberrant facial nerve regeneration

## Follow up:

- Consider referral to Neurology:
    - Atypical features
    - No improvement is observed within first 4 weeks
    - Incomplete resolution of symptoms by 3 months
- Review Acute ENT in 4 weeks (for House-Brackmann III-VI)
- Ophthalmology review (AOS) in 1-2 weeks depending on the degree of lagophthalmos and corneal exposure

**Evidence Table**

| Author(s) | Title | Source | Level of Evidence (I – VII) |
|---|---|---|---|
| | Bell's Palsy | UpToDate 2017 | **I** |
| | Bell's Palsy 2013 | American Academy of Otolaryngology Head and Neck Surgery Clinical Practice Guideline | **I** |
| Pane A, Burdon M, Miller N. Mosby | The Neuro-Ophthalmology Survival Guide | Elsevier | **II** |
| | Electronic Therapeutic Guidelines, Bell's Palsy Therapeutic Guidelines Australia | | **I** |

**The Hierarchy of Evidence**

The Hierarchy of evidence is based on summaries from the National Health and Medical Research Council (2009), the Oxford Centre for Evidence-based Medicine Levels of Evidence (2011) and Melynk and Fineout-Overholt (2011).

I) Evidence obtained from a systematic review of all relevant randomised control trials.

II) Evidence obtained from at least one well designed randomised control trial.

III) Evidence obtained from well-designed controlled trials without randomisation.

IV) Evidence obtained from well-designed cohort studies, case control studies, interrupted time series with a control group, historically controlled studies, interrupted time series without a control group or with case series.

V) Evidence obtained from systematic reviews of descriptive and qualitative studies.

VI) Evidence obtained from single descriptive and qualitative studies.

VII) Expert opinion from clinician, authorities and/or reports of expert committees or based on physiology.

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