Herpes Zoster Oticus / Ramsay Hunt Syndrome Type II

- Also known as Geniculate Herpes

**Ramsay Hunt Syndrome:**

- James Ramsay Hunt (1872–1937): Described three separate neurological syndromes
- **Ramsay Hunt syndrome type I** – also called Ramsay Hunt Cerebellar Syndrome, is a rare form of cerebellar degeneration which involves Dementing Process, Progressive Ataxia, Tremor, Myoclonic Epilepsy (DATE)
- **Ramsay Hunt syndrome type II** is the reactivation syndrome of herpes zoster in the geniculate ganglion. It has variable presentation which may include a lower motor neuron lesion of the facial nerve, deafness, vertigo, and pain.
- **TRIAD of Ramsay Hunt syndrome type II (VIP)**
  - Vesicles in the auditory canal and auricle.
  - Ipsilateral Facial Paralysis
  - Ear Pain
- **Ramsay Hunt syndrome type III** (rare) – an occupationally induced neuropathy of the deep palmar branch of the ulnar nerve. It is also called Hunt's disease or Artisan's palsy

**DEFINITION**

- Herpes zoster Oticus / Ramsay Hunt syndrome II / Geniculate herpetic is defined as a disorder that is caused by the reactivation of pre-existing Varicella Zoster Virus (Chicken Pox Virus) in the geniculate ganglion, a nerve cell bundle, of the facial nerve.
- Ramsay Hunt syndrome accounts for up to 12% of all facial paralyses and generally causes more severe symptoms and has a worse prognosis than Bell palsy

**ETIOLOGY**

- Individuals with decreased cell-mediated immunity resulting from carcinoma, radiation therapy, chemotherapy, or HIV infection are at greater risk for reactivation of latent VZV. Physical stress and emotional stress often are cited as precipitating factors

**SIGNS & SYMPTOMS**

- Sensory root of Facial nerve affected.
- Initially – Severe Otalgia
- Later – Painful, burning Erythematous vesicular rashes/eruptions, blisters and crusts over the pinna, external auditory canal and tympanic membrane. Can also form over Tongue, Tonsils and sometimes hard palate.
- LMN (Lower Motor Neuron) type of Facial Palsy (and its features)
- Vomiting, Giddiness
- Loss of taste in anterior 2/3rd of tongue.
- Hyperacusis, Tinnitus
- Dry mouth (Xerostomia), dry eyes (Xerophthalmia), eye pain, increased lacrimation can occur initially.
- Associated symptoms, such as SNHL and vertigo, are thought to occur as a result of transmission of the virus via direct proximity of cranial nerve (CN) VIII to CN VII at the cerebellopontine angle or via vasa vasorum that travel from CN VII to other nearby cranial nerves.
- Possible involvement of the trigeminal nerve can cause numbness of the face.

**PATHOPHYSIOLOGY**

- RHS type 2 refers to shingles of the geniculate ganglion. After initial infection, varicella zoster virus lies dormant in various nerve cells in the body, where it is kept in check by the patient's immune system. Given the opportunity, for example during an illness that suppresses the immune system, the virus is reactivated and travels to the end of the nerve cell, where it causes the symptoms described above.
- In RHS type 2, the affected ganglion is responsible for the movements of facial muscles, the touch sensation of a part of ear and ear canal, the taste function of the frontal two-thirds of the tongue, and the moisturization of the eyes and the mouth. The syndrome specifically refers to the combination of this entity with weakness of the muscles activated by the facial nerve. In isolation the latter entity would be called Bell's Palsy.
- Like shingles, however, lack of lesions does not definitely exclude the existence of a herpes infection. The virus can be detected, even before the eruption of vesicles, from the skin of the ear.

**COMPLICATIONS**

- Complications of HZ oticus may include the following:
  - Postherpetic neuralgia
  - Residual paralysis
  - Rarely, herpes zoster encephalitis

**PREVENTION**

- This disease is prevented by immunizing against the causal virus, varicella zoster, for example through *Zostavax*, a stronger version of chickenpox vaccine.

**DIAGNOSIS**

- Mostly Clinical
  - Routine Blood picture, Serum Urea Creatinine estimation, Electrolytes etc
  - For immunocompromised patients – IgM and IgA anti VZV antibodies assay
  - If diagnosis of Ramsay Hunt syndrome is not established by physical examination alone, consider a **head CT scan** to investigate other etiologies of facial paralysis.

**TREATMENT**
Until recently, therapy for herpes zoster (HZ) oticus has been generally supportive, including *Warm Compresses*, *Narcotic Analgesics*, and *Antibiotics* for a secondary bacterial infection.

**Antiviral agents**

- Early administration (< 72 h) of *Acyclovir* showed an increased rate of facial nerve function recovery and prevented further nerve degeneration. Also decrease the incidence and severity of postherpetic neuralgia.
- Evidence is accumulating that varicella-zoster virus (VZV) may be responsible for many cases of Bell palsy that go unrecognized because of a lack of cutaneous findings. *Zoster sine herpete* ("zoster without herpes") describes a person who has all of the symptoms of shingles except this characteristic rash.
- Accordingly, the clinician should entertain more liberal use of antivirals such as *Acyclovir*, *Valacyclovir*, and *Famciclovir*.
- Valacyclovir and Famciclovir have been shown to be more effective than acyclovir in reducing risk of pain, with comparable lesion healing and safety profile. Furthermore, patient compliance is likely to be higher with valacyclovir and famciclovir because each has an easier dosing regimen (3 times per day) compared with acyclovir (5 times per day).
- For acyclovir-resistant VZV, IV *Foscarnet* is an appropriate alternative therapy (famciclovir and valacyclovir are not effective against acyclovir-resistant VZV).

**Dosage Of Acyclovir**:  
- **800 mg** orally every 4 hours *(5 times a day)* for 7 to 10 days
- Children OR Severe, immunocompromised host: **10 mg/kg** IBW IV every 8 hours for 7 to 14 days *(TDS)*

**Dosage Of Famcyclovir**:  
- **500 mg** orally every 8 hours for 7 days *(TDS)*

**Dosage of Valcyclovir**:  
- **1 g** orally every 8 hours for 7 days *(TDS)*

**Corticosteroids**

- Systemic corticosteroids are used to relieve acute pain, decrease vertigo, and limit the occurrence of postherpetic neuralgia.
- Prednisolone + Acyclovir treatment more effective than prednisolone alone. (time to healing of rash, time to cessation of acute neuritis, time to return to usual activity and sleep, and time to cessation of analgesics).
- Treatment has shown to achieve complete recovery in a majority of patients if started < 72hrs of facial paralysis, with chances of recovery decreasing as treatment was delayed. Delay of treatment may result in permanent facial nerve paralysis.
- Treatment apparently has NO

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<thead>
<tr>
<th>Drug</th>
<th>Dosage</th>
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<tr>
<td>Acyclovir (Zovirax)†</td>
<td>800 mg orally five times daily for 7 to 10 days; 10 mg per kg IV every 8 hours for 7 to 10 days‡</td>
</tr>
<tr>
<td>Famciclovir (Famvir)†</td>
<td>500 mg orally three times daily for 7 days</td>
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<tr>
<td>Valacyclovir (Valtrex)†</td>
<td>1,000 mg orally three times daily for 7 days</td>
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<tr>
<td>Prednisone (Deltason)</td>
<td>30 mg orally twice daily on days 1 through 7; then 15 mg twice daily on days 8 through 14; then 7.5 mg twice daily on days 15 through 21</td>
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**EFFECT** on the recovery of hearing loss.

- **Diazepam** rarely used for vertigo