



RAMSAY HUNT SYNDROME

This handout is intended as a general introduction to the topic. As each person is affected differently, speak with your health care professional for individual advice.



Key points

- A shingles outbreak affecting a nerve in the face near one ear.
- Rare – usually affects older people.
- Anyone who has had chickenpox is potentially at risk.
- Seek immediate medical help for sudden facial paralysis, burning pain around the ear, or a painful red blistering facial rash.
- Other symptoms may include vertigo (spinning sensation) along with nausea and vomiting.
- Early diagnosis and aggressive treatment increase the likelihood of full recovery.
- 20% of patients end up with a painful condition affecting nerve fibres and skin.
- 1 in 5 people who have had chickenpox are likely to get shingles – and potentially RHS – without vaccination.
- Vaccination against shingles is recommended for those 50 years and older.

What is Ramsay Hunt syndrome?

Ramsay Hunt syndrome (RHS) is a shingles (herpes zoster) outbreak that affects the facial nerve near one ear. It usually causes varying degrees of one-sided facial paralysis or weakness and hearing loss. Anyone who has had chickenpox (varicella) is potentially at risk of developing RHS.

RHS is sometimes called herpes zoster oticus because of the distinctive chickenpox-like rash that usually appears in and around the ears. Some doctors use this term in reference to only the ear rash, and Ramsay Hunt syndrome for the combined ear rash and facial palsy.

The disorder is named after James Ramsay Hunt, an American neurologist who first described it in 1907.

RHS is most common in older people, typically affecting those over the age of 60. It is a rare disorder. Only about 5 out of every 100,000 people develop RHS each year. By age 85 about 50% of people will develop shingles and be at potential risk for RHS. Men and women are equally affected by RHS. It is responsible for 12% of cases of facial paralysis. RHS is extremely rare in those under 20 years of age.

What are the causes?

RHS is caused by the varicella-zoster virus (VZV), the same virus that causes chickenpox and shingles. Even after the chickenpox infection is over, the virus can lie dormant in the nervous system for years before becoming active again. When the virus re-emerges to cause RHS, it lodges in a collection of fibres and neurons of the facial nerve (called the geniculate ganglion).

Why the virus reactivates in only some people who have had chickenpox is not fully understood. A decrease in cell-mediated immunity may play an

important role. The cell-mediated immune response specializes in recognizing and attacking invaders found within cells in the body. This may explain why someone with an immune system weakened by disease or medications is much more likely to develop shingles than a healthy person.

Stress and depression do not cause RHS, but they may weaken the immune system and increase the risk of developing shingles and potentially RHS. Research suggests that exposure to chronic stress as well as depression may play a role in triggering shingles.

RHS is not a contagious disease; you cannot catch it from someone else. However, the herpes zoster virus found in the blisters of RHS can cause someone who has never had chickenpox, or has not been vaccinated for it, to get chickenpox. This is uncommon; direct contact with fluid from the blisters is needed.

If you have RHS, it is important to wait until scabs form on all blisters before coming into contact with:

- people who have never had chickenpox or been vaccinated for chickenpox
- people with a weak immune system
- babies from birth to one month of age
- pregnant women

What are the symptoms?

In most cases, the main symptoms of RHS are:

- A painful reddish rash with fluid-filled blisters (vesicular rash) in clusters on, in and around one ear. The rash is similar in appearance to chickenpox, though it is more painful than itchy. Blisters may also appear inside the mouth.
- Paralysis or weakness on the same side of the face as the affected ear. This causes the facial muscles to droop.

Usually, pain precedes the rash by 3 to 7 days. In some cases, the pain and rash appear at the same time. Sometimes the rash will appear before the facial paralysis or vice versa. Occasionally no rash

appears, making diagnosis more challenging; this is called zoster sine herpette (ZSH). Usually, only one side of the face is affected (unilateral).

Other symptoms might include:

- deep ear pain (otalgia) on the affected side that progresses from a dull ache to excruciating sensations such as burning, electric shock or sharp jabbing pain; it may progress to the neck
- sensorineural hearing loss and/or ringing in the ear (tinnitus) on the affected side
- sensitivity to sounds (hyperacusis)
- difficulty closing the eye or blinking on the affected side, due to facial weakness on the affected side
- dry eye on the affected side
- vertigo (spinning sensation) accompanied by nausea and vomiting
- difficulty eating, drinking and speaking, as well as a lopsided smile, due to facial weakness on the affected side
- flu-like symptoms, including a high fever
- a change in taste perception, or loss of taste on the affected half of the tongue

The area affected can be extremely painful. Usually, the pain increases with movement or contact with bedding or clothing.

In most cases, the symptoms of RHS are temporary and last for several weeks.

How is it diagnosed?

Seek immediate medical attention if you have sudden facial paralysis, burning pain around the ear, or a painful, red, blistering rash on your face. The diagnosis of RHS is a possibility and prompt treatment increase the likelihood of a full recovery.

A family doctor (GP) can often diagnose RHS based on a physical examination, a detailed patient history and the hallmark symptoms of the disorder. To confirm the diagnosis, your doctor may take fluid from a fresh blister to test for the varicella

zoster virus. If you do not have a rash, your blood may be tested for antibodies.

You may be referred for urgent consultation with a neurologist or an otolaryngologist (ear, nose and throat or ENT doctor).

An MRI (magnetic resonance imaging) of the head might be done to pinpoint areas of inflammation along the facial nerve and to rule out another cause of the symptoms.

The diagnosis of RHS can be difficult because the specific symptoms of the disorder (ear pain, facial paralysis and the distinctive rash) do not always appear at the same time, or no rash may appear. Pain preceding the rash often leads to an initial incorrect diagnosis.

Because the pain of RHS usually starts several days before the blistering rash, an incorrect diagnosis of other conditions can be made. Other causes of similar pain include:

- trigeminal neuralgia
- sinus disease
- glaucoma
- tumours behind the eyeball (retro-orbital tumours)
- inflammatory diseases such as Tolosa-Hunt syndrome
- tumours within the skull (intracranial tumours)

RHS may also be first misdiagnosed as another condition including stroke, Bell's palsy, or acoustic neuroma.

How is it treated and managed?

The treatment challenge of RHS is to give immediate relief of acute pain and symptoms as well as prevent complications, including postherpetic neuralgia. Prompt diagnosis and early aggressive treatment appears to improve outcomes and decrease the likelihood of developing complications. It is generally agreed that starting antiviral treatment within three days of the onset of RHS has the greatest benefits.

Medical treatment

Antiviral medications, including acyclovir or famciclovir, have been shown to shorten the course of RHS. Care must be taken to monitor patients for side effects of these drugs. They can be used in combination with other treatments. The use of antiviral medications in combination with steroids results in better recovery rates than steroids used alone.

Nerve blocks (local anaesthetics and steroids) are generally the treatment of choice to cut pain and prevent postherpetic neuralgia. If nerve blocks are not used immediately and aggressively, especially in elderly patients, may lead to a lifetime of debilitating pain from postherpetic neuralgia.

As the blisters crust over, nerve blocks may be used with a short course of high-dose steroids, like prednisone. This may decrease the likelihood of irreversible nerve damage and postherpetic neuralgia.

Aching pain in the acute phase of a severe case of RHS may be relieved by short-term use of **opioid analgesics**. They are less effective, however, in relieving nerve pain. Care must be taken to monitor patients for side effects of these potent drugs.

Gabapentin, an anticonvulsant drug, may be used to treat nerve pain. It may also help prevent postherpetic neuralgia. Treatment is most effective when used early in the course of RHS. Gabapentin may be used at the same time as nerve blocks and other analgesics.

Carbamazepine, an anticonvulsant drug, may be used in patients with severe nerve pain who do not respond to nerve blocks and gabapentin.

Phenytoin, an anti-epileptic drug, may also be used to treat nerve pain.

Antidepressants may be useful in the initial treatment of RHS if significant sleep disturbance is a problem.

Medications used to suppress vertigo (spinning sensation) on a short-term basis include anti-anxiety

drugs such as diazepam (Valium®), antihistamines and anticholinergics.

Vestibular rehabilitation

RHS can damage the vestibular nerve leading from your inner ear on the affected side to the brain. This leads to dizziness and balance problems. The brain needs to learn to compensate for the loss of balance function. In some patients, this compensation happens naturally, so they do not need vestibular rehabilitation. Other patients, however, may continue to have symptoms. They may benefit from vestibular rehabilitation. A vestibular therapist can help design an appropriate program for you.

Vestibular rehabilitation for RHS might include:

- moving your head while you look at object that is standing still
- exercises to help you balance better while you are standing or walking

It is very important to start the exercises gradually and increase them slowly and steadily. If you try to do too much too soon, your dizziness and imbalance may get worse.

Facial retraining

RHS damages the facial nerve leading from the affected side to your brain. This causes paralysis or drooping on one side of your face. Facial retraining, sometimes called facial neuromuscular reeducation (NMR), uses your brain's ability to change and adapt as a result of experience (neuroplasticity). Active and persistent patient participation is needed to retrain the brain as the nerve regrows. The goals of facial retraining are a more symmetrical face, regained control of the muscles used in facial expression, and increased facial range of movement. A physiotherapist or occupational therapist specializing in facial retraining, also known as can help design an appropriate program for you.

Facial retraining for RHS might include:

- exercises to improve muscular control and decrease unwanted movement

- mime therapy using a mirror to “trick” your brain into thinking that the paralyzed side of your face is moving as you look at the other side in a mirror
- manual stretching and self-massage techniques to decrease muscle tightness

It is important to practice the exercises regularly at home for change and recovery to happen.

Other treatments and home remedies

Other treatments and home remedies used together with primary medical treatments include:

- Applying ice packs to the rash may give temporary relief for some patients.
- Using heat increases pain in most patients, however it is occasionally helpful and may be worth trying if ice packs do not give relief.
- TENS (transcutaneous electrical nerve stimulation) therapy may help a few patients. It uses low-voltage electrical current for pain relief. TENS therapy can be done by a physiotherapist or at home after instruction.

These treatments are reasonable choices for patients who cannot or will not have nerve blocks or take medications.

Aluminum sulphate dissolved in lukewarm water can be applied to soothe and help dry out crusting and weeping blisters.

It is important to keep areas affected by the rash clean. Zinc oxide ointment may be applied to protect the skin during the healing phase when temperature sensitivity is a problem. Covering the rash with loose, non-stick, sterile bandages may be recommended.

Sometimes facial paralysis stops the eye from fully closing. Care must be taken to prevent injuring the cornea (the clear front surface of the eye). Artificial tears and lubricating ointments may be prescribed. Eye protection at night, such as an eye patch, may be recommended.

Complications

Complications of a severe case of RHS may be persistent or permanent and include:

- The most common complication of RHS is postherpetic neuralgia. It affects about 20% of people who get shingles. Postherpetic neuralgia happens when the shingles infection damages nerve fibres. The messages sent by these nerve fibres to the brain become disrupted and exaggerated. Older patients and those who did not receive suitable treatment in the acute state are at greatest risk of developing postherpetic neuralgia.
- Changes in the appearance of the face (disfigurement) from loss of movement; this can have a significant impact on psychological well-being and quality of life.
- Altered ability to taste
- Vision loss resulting from ulcers on the cornea and eye infections
- Nerves that reattach to the wrong structures. This may cause abnormal reactions to movement; for example, smiling may cause the eye to close.
- Facial or eyelid spasms

The symptoms of postherpetic neuralgia range from mild to debilitating. Patients may suffer from constantly burning pain made worse by light touch, movement, anxiety, or temperature change. The pain may be so severe that it interferes with sleep and dominates activities of daily living.

Treatment of postherpetic neuralgia depends on the patient's health condition. It may include antidepressants and pain relief medications applied to the skin (topical analgesics), including lidocaine and capsaicin.

Rarely, the virus may spread to other nerves or the brain and spinal cord. This can cause confusion, drowsiness, headaches, weakness in the arms or legs, and nerve pain. A lumbar puncture (spinal tap)

may be needed to work out if other parts of the nervous system have been infected.

Prevention

Fortunately, vaccination can help prevent reactivation of the herpes zoster virus. About 20% of people that have had chickenpox are likely to get shingles and potentially RHS without the vaccine.

Two vaccines are available in Canada:

Shingrix® is recommended by Canada's National Advisory Committee on Immunization for those 50 years and older who:

- * Have previously received Zostavax II® at least one year ago
- * Have had shingles at least one year ago
- * Are not sure if they had chickenpox in the past

Shingrix®, first used in Canada in 2017, has been shown to reduce the risk of getting shingles by 97% in those over 50 years of age and by 91% in those over 70 years of age. If you do get shingles even after having the vaccination, it is very likely to be a mild case.

Shingrix® is given in two doses, at least 6 months apart. Some provincial or territorial medical plans cover the cost (about \$150 per dose). Some extended health plans may cover part or all of the cost. Shingrix® is available through most pharmacies and a prescription is not needed. Protection lasts for at least four years. As the vaccine is so new, how long protection lasts beyond four years is still unclear. Researchers expect protection will last much longer.

The older Zostavax II® vaccine may be considered for adults 50 years of age or older who cannot get Shingrix® for medical reasons.

Talk with your family doctor before getting either vaccine.

Canadian children have been routinely vaccinated against chickenpox since the early 2000s. This greatly reduces their likelihood of becoming infected with chickenpox and then possibly shingles later on.

What to expect in the future

Chances of full recovery are better if treatment is started early. About 70% of patients have a high rate of complete recovery within a few weeks if treatment is started within three days of developing symptoms. As the blisters heal, the crusts fall off leaving pink scars that gradually become lighter than overall skin tone (hypopigmented). In most cases, the pain also goes away.

Children are more likely than adults to recover completely.

If damage is more severe, full recovery may not happen, even after several months. Some degree of hearing loss and facial paralysis may be permanent. Research suggests that if antiviral drugs are not started until seven days or more of onset, recovery from facial paralysis is only 30%. Physiotherapy may help regain proper use of the facial muscles if facial weakness persists. An injection of Botox® may help those who have trouble closing one eye.

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