

SHINGLES (HERPES ZOSTER)

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Have you ever had chicken pox? If you are over 40 years of age, the answer is probably yes, which means that as you approach your 60's, and possibly before, you are at risk for herpes zoster (shingles, also known simply as "zoster"). As we age, our immune system weakens and we are more and more vulnerable to the infection. Even if you are nowhere near the age of risk, this information is for you and your older relatives and friends who may not be aware that there is a vaccine, Zostavax®, available in your physician's office or at participating pharmacies. Even if older adults do not remember having chicken pox, they should still get vaccinated. This is a really cruel disease!

Awakening of a Really Cruel Dormant Virus

The chicken pox virus, varicella-zoster, does not just "go away" after the disease has run its course. It continues to exist in the body in a dormant state in the posterior spinal ganglia or worst yet, the cranial sensory ganglia. At sometime later in life it can be reactivated, emerge from the ganglion and travel along the nerve and its branches to the skin surface and, if cranial, to the eye, ear, face and mouth supplied by the involved nerve or nerves. In the young person the immune system is usually strong enough to restrict the dormant virus to the ganglia. A weakened immune system may fail to suppress its activation. It is not known for certain if there are triggers for the reactivation of the virus, such as stress, trauma, certain medications or other infections

Let there be no doubt. Zoster is NOT simply a benign rash with associated deep pain; it is an acute neurological disease that should be evaluated on an emergency basis. It can show up in many forms from a self-limited painful rash or occasionally pain without a rash to the most serious involvement of the eye, ear, mouth, throat, facial muscles or spinal cord with muscle weakness.

Ganglion. A ganglion (ganglia, pl.) is a nodule containing a group of spinal nerve cell bodies. Each nerve has a cell body and for the sensory nerves this cell body is located in the spinal ganglia or in the cranial ganglia. A spinal ganglion and its position near the spinal cord are shown in Fig. 1.

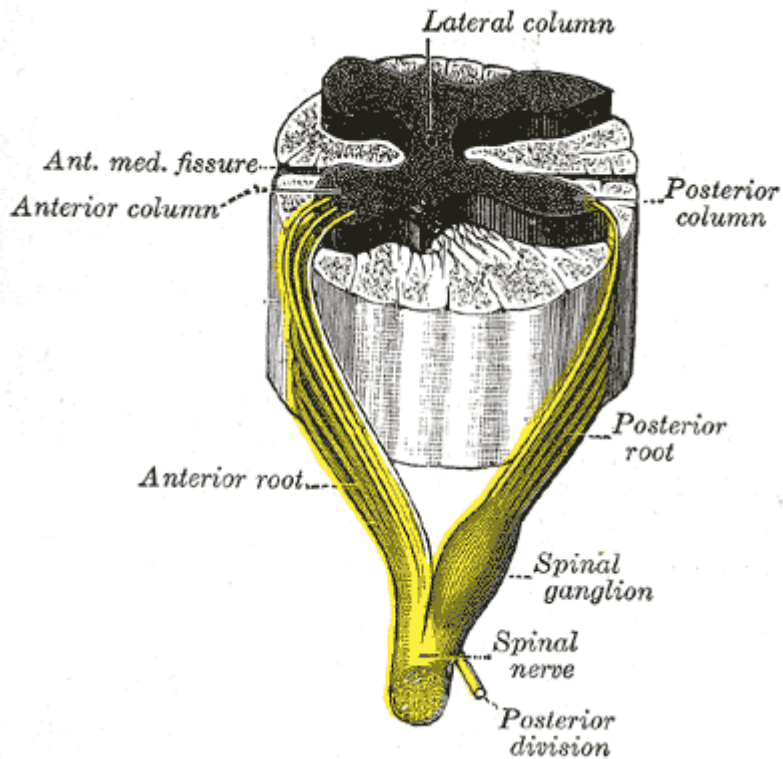


Fig. 1. A spinal ganglion is seen at the end of the posterior root for sensory nerves.
(A Netter drawing.)

Progression. Fig. 2 below, diagrammatically illustrates the progression of the zoster virus from nerve to skin. In this illustration, the position of the "dormant virus" should have been shown located in a spinal ganglion rather than just hanging out in the nerve. That having been said, the diagrammatic illustration of the deep nerve involvement and skin eruption is helpful in understanding the spectrum of this disease. Damage to the nerve may result in pain for months or years after the skin lesions have healed and disappeared. The term used for this pain is *postherpeic neuralgia*.

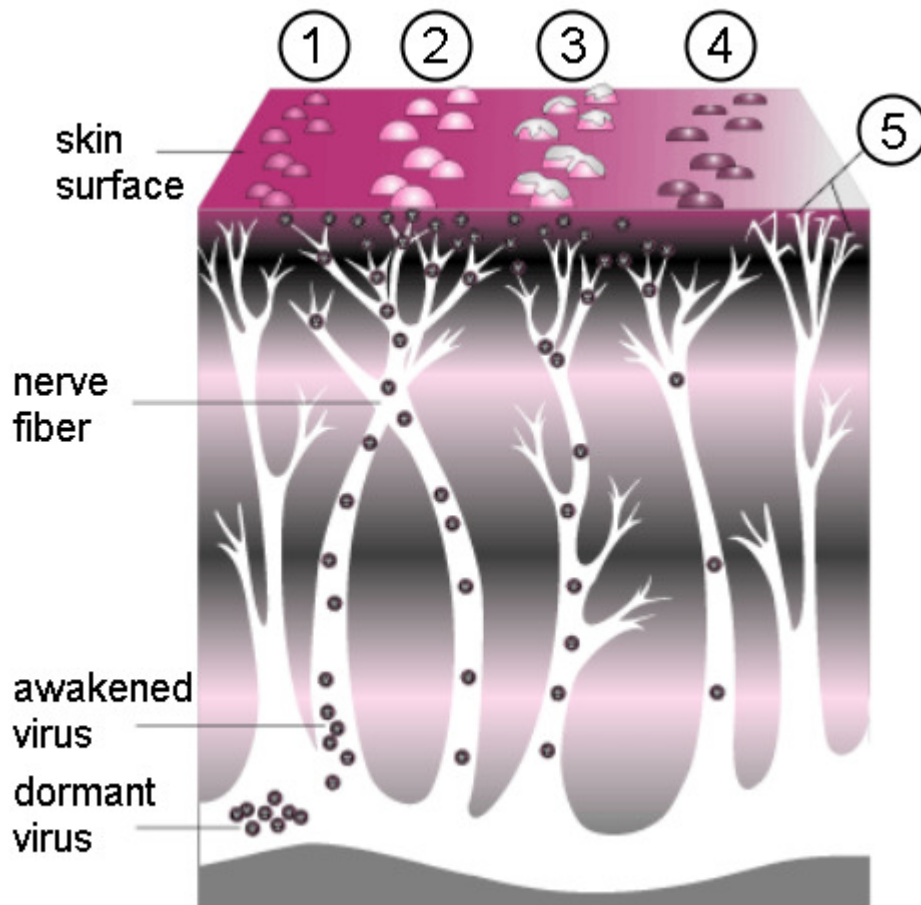


Fig. 2. Progression of herpes zoster. A cluster of small bumps (1) turns into blisters (2). The blisters fill with lymph, break open (3), crust over (4), and finally disappear. Postherpetic neuralgia can sometimes occur due to nerve damage (5). (Illustration and legend From Wikipedia)

Pain. The first sign of this infection is unusual deep pain or burning on one side of the body. This is the ideal time to start an antiviral medication---before the rash appears. However, the pain may not always be recognized for what it is.

First symptom. In my bout with this disease, the initial pain was at a deep spot along the medial border of the scapula. However, I thought it was from my workouts. I was however, aware that it was different, one sided, deep and more persistent than usual. I even mentioned it to Greg, my coach at the time, because it was so different--intense and deep.

A friend had five cranial nerves involved. Her first symptom was a persistent severe ear ache that was not associated with a cold or sore throat. The course of her disease, as you will see, was devastating.

As the nerve involvement becomes complete, the pain associated with zoster is severe, deep, sustained in intensity and unrelenting day and night. There are no peaks and valleys

-- often for more than a month. It is generally more severe in the older person because of the age-related decline in the immune system.

Rash. The first lesion need not necessarily begin at the vertebral column, but may initially appear somewhere within the half circle of the body that the nerve supplies, in particular where that enervation is closer to the surface (the underarm for example).

When a strange rash begins to appear on one side of the body associated with deep intense unrelenting pain on the same side and level on the body, there should be no doubt as to what is happening. See your family care physician immediately for the antiviral prescription and do not delay in getting the prescription filled--hours count. This is a nerve disease that in its least threatening form involves pain and skin lesions. In its more devastating form there is severe pain, lesions and nerve damage to the ear, nasal passages, gag reflex, mouth, and eye. If a specialist needs to be involved, it would therefore be a neurologist or, when the cranial nerves are involved, an ophthalmologist.

Blisters. Red patches on the skin are usually followed by small blisters, as seen below in Fig. 3. At this stage it is beyond the "rash" or "little bumps" and is into the blister stage. The blisters will break and seep, crust over, and disappear. The outbreak on the skin in the area supplied by the nerve, including vesicles in the ear canal with cranial nerve involvement, lasts around two weeks.



Fig. 3. The blisters or herpes zoster.

When the blisters open, there are small ulcers--something like that seen in chicken pox. The ulcers seep and begin to dry and form crusts that fall off in 2 to 3 weeks. Scarring is rare. The rash usually involves a well defined area from the spine around to the front of the belly or chest, as seen in Fig. 4 below.



Fig. 4. *Herpes zoster vesicles showing the rash and blisters. Note that the skin involvement begins at midline; it ends at midline in the back. This is the same path as the infected nerve..*

Unfortunately, zoster may strike through a cranial ganglia and involve the face, eye, mouth, and ear--a far more devastating involvement with risk to the eye, as shown below in Fig. 5. The blisters have opened and scabs are forming. Although zoster is usually seen in people over 60, youth does not always protect except that the pathology may not be as extensive because of a more robust immune system.

NOTE: The "Figure 2" call-out below is permanently attached to the picture and I couldn't delete it. This is really Fig. 5 for this article with thanks to MN Oxman and the University of Calif at San Diego.

FIGURE 2. Case of herpes zoster ophthalmicus



Photo/MN Oxman, University of California, San Diego

Fig. 5. Herpes zoster involving cranial nerves. The ulcers have scabbed over and there appears to be extensive inflammation.

Complications

Postherpetic neuralgia. A common complication of zoster is the continuance of pain in the involved nerve or nerves, known as *postherpetic neuralgia*. This happens when the involved nerves have been damaged by the infection. Pain is chronic, often debilitating, and can last for months or even years.

Herpes zoster ophthalmicus. The viral reactivation at the ophthalmic branch of the fifth cranial nerve is 5 times as common as involvement of the maxillary or mandibular branches. This can result in prolonged or permanent pain, facial scarring, drooping of the eye lid because of nerve damage, and loss of vision in the involved eye with its associated loss of depth perception. The pictures in Figs. 4 and 5 convince one of the seriousness of this viral eruption when it involves the cranial nerves and the necessity for older adults to be vaccinated.

For my friend, there were 5 cranial nerves involved effecting her eye, nose, ear, throat, mouth and jaw. The ear involvement effected her gait. She was unable to chew, smile, or close the eye on the affected side, thus the need for an eye patch and eye drops to avoid corneal damage. The entire side of her face drooped. She also lost her gag reflex and needed to take great care eating. She could not even sip from a straw without drooling. Her pain and discomfort were off the scale---terrible. Eventually she progressed to normal speech. Her ability to eat normally returned, but difficulty with

night vision, balance, and palsy of the right eye lid remain. Her message for those reading this: "If you don't get vaccinated you are foolish."

Incidence

Approximately one in three persons will develop zoster in the United States annually. There is a higher incidence and severity of zoster in older people, especially after age 50. Almost 50% of those who live into their 80s can expect to develop zoster. Regardless of age, people with immune deficiencies such as AIDS, lymphoma, and those who have had bone marrow or kidney transplants are at risk.

Treatment

Antiviral medication mitigates the severity of the disease and its aftermath and that should be your goal and insistence. Although only partially effective, it will at least shorten the time with post-herpetic neuralgia. The medication should be started as soon as possible within 24 hours of feeling pain or burning, and preferably before the blisters appear. The physician will also prescribe a pain medication. In my personal experience, the anticipated long course and unrelenting nature of the pain negated the use of anything strong enough or tolerable enough to make much of a difference. One simply bears the pain.

Taking Care

Scrupulous hygiene is critical. It is important to avoid infection as well as scars. Do not touch the involved skin area. And, of course, avoid scarring by not picking at the scabs. Let them fall off naturally.

The Vaccine

Efficacy. The studies for Zostavax® enrolled approximately 38,000 people throughout the United States who were 60 years of age and older. Approximately half received zostavax® and half received placebo. Study participants were followed for about three years to see if they developed zoster and if they did, how long the postherpetic pain lasted.

At the conclusion of the studies, researchers found the vaccine reduced the occurrence of zoster by about 51%, its effect being highest in people between the ages 60-69. However, its effectiveness declined with increasing age to 41% for the 70-79 age group and 18% for those 80 years of age and older. The zoster vaccine reduced the burden of illness due to herpes zoster among people 60 years of age or older by 61.1 % and reduced the incidence of postherpetic neuralgia by 66.5 %.

For those who were vaccinated with Zostavax®, but still developed zoster, the duration of pain was a bit shorter than for those who received a placebo. Specifically, the pain of those in the zostavax® group lasted on average for 20 days and for those who received

placebo, it lasted for about 22 days. In the Zostavax® study, the severity of the pain did not appear to differ among the two groups.

History. The vaccine (Zostavax®) to help prevent zoster and reduce the impact of its sequelae completed clinical trials and was licensed by the FDA in May 2006. Two years later the Center for Disease Control and Prevention (CDC) and the Advisory committee on Immunization Practices (ACIP) in the U.S. recommended vaccination for those 60 years and older. The vaccine has been successfully used in Japan since 1988, 22 years before it was licensed for use in the U.S.

Already had Shingles?

The recurrence of zoster is common. If you have already had zoster, you are still advised by the CDC to get the vaccination. ZOSTAVAX was shown to boost specific immunity to the varicella-zoster virus.

Who should NOT get the shingles vaccine?

Any of the following situations would preclude vaccination for shingles:

- A previous life-threatening allergic reaction to gelatin or neomycin
- A severe allergy to any component of the vaccine
- A weakened immune system as a result of leukemia, lymphoma, or an other blood or bone cancer
- Infection with HIV/AIDS and a T-cell count below 200
- Being treated with drugs that affect the immune system, including high-dose steroids
- Known or potential pregnancy

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