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PRESSURE

Changes in pressure cause fluid and hairs to move in the inner ear, allowing us to hear and maintain balance.

ARTICLE

066

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Third Window Syndrome: What It Is and How It's Treated

By Yonit Arthur, AuD

WHAT IS THIRD WINDOW SYNDROME?

Third window syndrome describes a group of inner ear disorders that results from a leakage of pressure and/or fluid from the inner ear structures. The most commonly recognized is superior semicircular canal dehiscence (SSCD), but there are other types. These abnormalities can lead to vestibular symptoms such as dizziness, imbalance and difficulty focusing the eyes, as well as hearing symptoms such as hearing loss and sensitivity to loud sounds. Fortunately, this group of disorders is often medically or surgically treatable. Read on to find out more about the symptoms, causes and treatments for this disorder.

WHAT CAUSES THIRD WINDOW SYNDROME?

The Inner Ear is a Pressure Detection System

The inner ear contains two main structures: the cochlea (the hearing organ) and the semicircular canals and otoliths (the vestibular and balance organs). Although the organs look different and have different functions, the way they work is quite similar.

Both the hearing and vestibular organs are fluid-filled tubes, containing small hairs that are sensitive to changes in pressure. When pressure moves the fluid inside these structures, the fluid moves the hairs, which creates electrical signals that the hearing and balance nerves then send to the brain for interpretation.

Sound Is Changes in Pressure

In the hearing system, sound (which is simply changes in pressure in the air) enters the ear through the ear canal, which vibrates the eardrum and bones of the inner ear. This creates pressure changes at the oval window, a flexible cover over the opening of the fluid-filled hearing organ. After the pressure waves travel through the hearing organ, they exit through a second flexible cover at the other end, the round window. The round window acts as a vent that allows pressure out.

Think about these pressure changes like the flow of traffic along a busy

road through an intersection. For traffic to flow through the traffic light, the intersection needs to be clear. If there is a concrete barrier on the far end of the intersection, traffic will build up, and eventually it will back up to the traffic light. The "traffic," or sound pressure, needs to flow freely through the hearing organ to stimulate the hearing nerve.

Movement Creates Different Pressure Waves in Each Ear

In the balance system, movements of your body and head create waves of pressure changes inside the vestibular organs. As you move in one direction, the fluid pushes the hairs in that ear in the "on" direction, which increases its electrical signals to the balance nerve. At the same time, on the opposite side, the hairs are pushed in the "off" direction, which decreases its electrical activity. The brain compares information from both of these organs to figure out your direction and speed. If one organ is giving information that doesn't match the other side, you may feel dizzy.

The Third Window Lets Pressure Leak Out

In both parts of the inner ear, your sense organs depend on a perfect balance of pressure to translate the information they are getting from the outside world into nerve signals. This is similar to a car tire, which has to have the perfect amount of pressure in it for a smooth ride. Third window syndrome occurs when the organs or surrounding tissue are "leaky." That is, the pressure inside them is unstable due to a weakness or hole in the bone or tissues. This means that the organs of hearing and balance end up out of calibration. Think of what happens when you try to drive on a flat tire. The car will wobble, and you'll have difficulty controlling its direction. Similarly, the organs of hearing and balance end up sending confusing messages to the brain, which the brain interprets as sudden changes in hearing or head movement. This results in a variety of symptoms detailed below.

TYPES OF THIRD WINDOW SYNDROME

Third window syndrome can occur when there is any weakness or opening that disrupts pressure in the tissues or bones of the inner ear. That's why there are several potential sites for the issue:

- At blood supply attachment sites to the hearing or balance organs
- At nerve attachment sites to the hearing or balance organs
- In the bones connecting the middle ear to the inner ear
- In the openings connecting the hearing and balance organ to their fluid supply

These can be caused by:

Differences in the shape of the skull and organs
- Genetic tendency toward developing the disorder
- Head trauma
- Surgery
- Major pressure altering events – barotrauma, straining (such as during weightlifting or difficult vaginal delivery)
- Other disorders that disrupt the inner ear, like growths, tumors or infections
- Elevated intracranial pressure

DIAGNOSIS

If your healthcare provider suspects third window syndrome, typically you will see an ear-nose-throat physician for an exam. Then, the following tests may be used to help make a diagnosis:

Hearing Test

An audiologist will measure your responses to sounds of various pitches. People with third window syndrome often have hearing loss that follows a particular pattern involving loss of sensitivity to low-pitched sounds of certain types. In some cases, the hearing can be abnormally sensitive – better than normal hearing thresholds.

Cervical Vestibular-Evoked Myogenic Potentials (cVEMP)

SYMPTOMS OF THIRD WINDOW SYNDROME

As you've learned, there are several potential sites and causes for third window syndrome, which is why the symptoms may vary between people. Some people have just hearing symptoms, others have balance and dizziness symptoms, and still others have both.

Common Vestibular	Dizziness, particularly when hearing loud sounds
	Dizziness with physical strain like lifting something heavy, coughing, sneezing or blowing the nose
	Difficulty keeping a steady gaze when hearing loud sounds, or when experiencing physical strain, coughing, sneezing or blowing the nose
Common Hearing	Hearing your own voice more loudly than normal
	Hearing other internal noises abnormally, such as heartbeat, eye movement, blinking
	Hearing loss with a particular pattern (which can be determined through a hearing test)
	Sensitivity to loud sounds
	Tinnitus, which is the presence of ringing, buzzing or other sounds in the ears that are not from an outside source of sound; some people with third window syndrome have pulsatile tinnitus, which is tinnitus that follows a regular rhythm like a heartbeat
Other	Brain fog or memory issues
	Difficulties knowing where you are in space (feeling disoriented)
	Anxiety and panic attacks

This simple, non-invasive test measures a muscle reflex in response to sound. Sticky sensors (electrodes) will be placed on your neck. While wearing earphones, you will listen to loud pulses or click sounds. At the same time, you will be lifting your head and turning it to one side. The equipment will measure how your muscles change their response when you hear loud sounds, and it is a normal reflex. People with third window syndrome frequently have abnormally low thresholds on this test. That is, their muscles respond to lower-level sounds than normal.

Ocular Vestibular-Evoked Myogenic Potentials (oVEMP)

This test is similar to cVEMP but measures the response from another sensor. However, after you hear the loud sounds, instead of measuring a response in your neck, the response is measured from one of the eye muscles. Again, this is a normal reflex but in third mobile window syndrome, frequently the patients will exhibit an abnormally large response or will respond at high frequencies that normally do not elicit a response.

Electrocochleography (ECoG)

This simple, non-invasive test measures electrical information produced by the hearing organ in response to loud sounds. Typically, a small sensory (electrode) is placed deep into your ear canal. You will then listen to loud click sounds while the equipment records the responses from your hearing organ. People with certain types of third window syndromes have abnormally low responses to this test.

Vestibular Testing

Patients who are suspected of third mobile window syndrome should undergo a full battery of vestibular testing. This defines the patient's underlying vestibular function (the same as a hearing test will define their hearing ability) and

can aid in the diagnosis of third window syndrome. Vestibular tests that may aid in the diagnosis include Tullio testing (evaluation for nystagmus – jerking eye movements – while putting sound in the ear), fistula testing (evaluation for nystagmus or imbalance with pressure applied to the ear drum) and Valsalva testing (evaluation for nystagmus when the patient strains).

CT Imaging

High resolution scans of your skull can also help confirm the diagnosis. However, the diagnosis of third window syndrome is not made using CT scans alone. The scans may miss a weak area in the bone, or they may reveal thin or weak areas in the bone that do not lead to symptoms. The diagnosis is therefore made using multiple criteria and tests.

Diagnosis

The diagnosis of third mobile window syndrome requires three elements: 1.) history and symptoms consistent with third window syndrome, 2.) diagnostic testing consistent with third mobile window syndrome, and 3.) appropriate CT scan identifying a defect. The exception to this is a patient who has conditions 1 and 2 but no identifiable bony defect on CT scan. This group of patients may have either a defect that has not yet been identified or may have a defect of the round or oval window, which will not show up on a CT scan.

TREATMENT

Several options exist for the management of symptoms related to third window syndrome.

Lifestyle Modifications

Some patients are able to tolerate their symptoms and reduce more severe effects by avoiding triggers such as loud sounds and physical strain.

Pressure-Equalization Tubes

Some patients, particularly those who are sensitive to changes in pressure, may get relief from placement of a pressure-equalization (PE) tube in the eardrum of the affected side(s). This is often a quick, in-office procedure that does not require more than local anesthesia. Although this treatment can help some patients, it is very few and it is usually patients who have concomitant eustachian tube dysfunction.

Surgical Correction

Depending on the location of the "third window," surgical repair may be an option. In many cases, the affected area can be plugged or repaired surgically, effectively eliminating the pressure leakage that is causing the problem. Another approach is to reinforce the middle ear windows – while this is much less invasive, it generally has a lower success rate. In many cases, surgery successfully stops the bothersome symptoms. It may, however, result in changes in the function of the inner ear. Some patients need vestibular rehabilitation therapy after the procedure to recover from these changes. It is important to note that surgery can help symptoms related to vertigo, dizziness and autophony (hearing internal sounds too loudly), it is generally not recommended solely for the indication of hearing loss.

CONCLUSIONS

Third window syndrome is a group of disorders that all affect the way the inner ear manages pressure. This happens due to an opening or weakness in the structures of the inner ear or surrounding tissue or bone. When the inner ear has unusual or sudden changes in pressure, that can produce a variety of hearing and balance related symptoms. Because pressure changes with environment (including sound) and physical activity, patients may experience a wide range of symptoms that come and go.

It is important to have a variety of diagnostic tests to determine whether you have third window syndrome. The diagnosis is never made based on a scan alone. Due to its clear physical cause (weakened bone or tissue, or openings in bone or tissue), third window syndrome is one of the few vestibular disorders that has a direct surgical repair as an option for treatment.

ADDENDUM

In March 1998 Lloyd Minor published the first two patients to undergo surgery for Superior Canal Dehiscence, improving their vestibular symptoms. At that time the condition was unknown outside a handful of people and the world of Neurotology believed SCD to be a rare disorder - one that affects less than 200,000 Americans (roughly 0.06%), as defined by the Rare Disease Act of 2002. We now know that this is not the case. The John Hopkins temporal bone histology study of more than 1000 temporal bones demonstrated 0.5% incidence of SCD, and about 1.5% incidence of extreme bony thinning. Cochlear-facial dehiscence is actually more common than SCD, although not recognized or diagnosed nearly as frequently, and when you add their incidence together we have a 1.1% incidence among the general population. This percentage is more than 20 times the upper limit of what defines a rare disease, so TMWD are most definitely not a rare problem. It is most important for everyone to be aware of this. - Dr. Gerard Gianoli

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