



NIDCD Fact Sheet

Recurrent Respiratory Papillomatosis or Laryngeal Papillomatosis

U.S. DEPARTMENT OF HEALTH & HUMAN SERVICES · NATIONAL INSTITUTES OF HEALTH · NATIONAL INSTITUTE ON DEAFNESS AND OTHER COMMUNICATION DISORDERS

What is recurrent respiratory papillomatosis?

Recurrent respiratory papillomatosis (RRP) is a disease in which tumors grow in the air passages leading from the nose and mouth into the lungs (respiratory tract). Although the tumors can grow anywhere in the respiratory tract, their presence in the larynx (voice box) causes the most frequent problems, a condition called laryngeal papillomatosis. The tumors may vary in size and grow very quickly. They often grow back even when removed.

What is the cause of RRP?

RRP is caused by two types of human papilloma virus (HPV), called HPV 6 and HPV 11. There are more than 150 types of HPV and they do not all have the same symptoms.

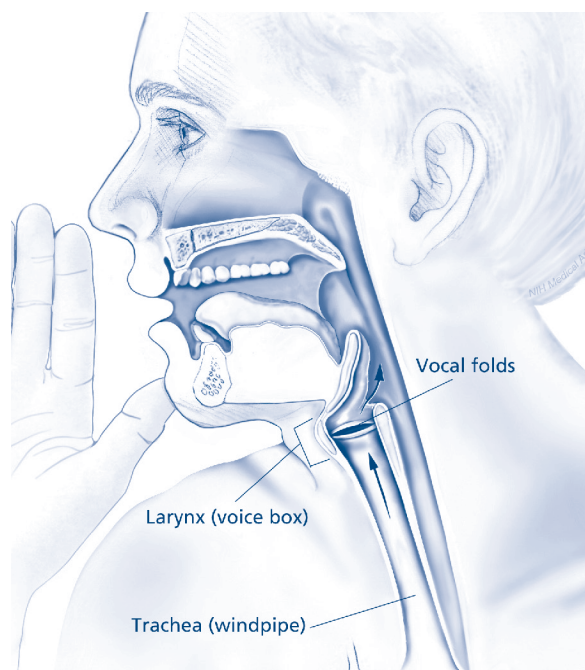
Most people who encounter HPV never develop any illness. However, many HPVs can cause small wart-like, non-cancerous tumors called papillomas. The most common illness caused by HPV 6 and HPV 11 is genital warts. Although scientists are uncertain how people are infected with HPV 6 or HPV 11, the virus is thought to be spread through sexual contact or when a mother with genital warts passes it to her baby during childbirth. HPV 6 and HPV 11 can also cause disease of the uterine cervix and, in rare cases, cervical cancer.

According to the Centers for Disease Control and Prevention, the incidence of RRP is rare. Fewer than 2,000 children get RRP each year.

Who is affected by RRP?

RRP affects adults as well as infants and small children who may have contracted the virus during childbirth. According to the RRP Foundation, there are roughly 20,000 cases in the United States. Among children, the incidence of RRP is approximately 4.3 per 100,000; among adults, it's about 1.8 per 100,000.

Parts of the respiratory tract affected by RRP





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What are the symptoms of RRP?

Normally, voice is produced when air from the lungs is pushed past two side-by-side elastic muscles—called vocal folds or vocal cords—with sufficient pressure to cause them to vibrate (see figure on page 1). When the tumors interfere with the normal vibrations of the vocal folds, it causes hoarseness, which is the most common symptom of RRP. Eventually, the tumors may block the airway passage and cause difficulty breathing.

Because the tumors grow quickly, young children with the disease may find it difficult to breathe when sleeping, or they may experience difficulty swallowing. Adults and children may experience hoarseness, chronic coughing, or breathing problems. The symptoms tend to be more severe in children than in adults; however, some children experience some relief or remission of the disease when they begin puberty. Because of the similarity of the symptoms, RRP is sometimes misdiagnosed as asthma or chronic bronchitis.

How is RRP diagnosed?

Two routine tests for RRP are indirect and direct laryngoscopy. In an indirect laryngoscopy, an otolaryngologist—a doctor who specializes in diseases of the ear, nose, throat, head, and neck—or speech-language pathologist will typically insert a flexible fiberoptic telescope, called an endoscope, into a patient's nose or mouth and then view the larynx on a monitor. Some medical professionals use a video camera attached to a flexible tube to examine the larynx. An older, less common method is for the otolaryngologist to place a small mirror in the back

of the throat and angle the mirror down toward the larynx to inspect it for tumors.

A direct laryngoscopy is conducted in the operating room with the use of general anesthesia. This method allows the otolaryngologist to view the vocal folds and other parts of the larynx under high magnification. This procedure is usually used to minimize discomfort, especially with children, or to enable the doctor to collect tissue samples from the larynx or other parts of the throat to examine them for abnormalities.

How is RRP treated?

There is no cure for RRP. Surgery is the primary method for removing tumors from the larynx or airway. Because traditional surgery can result in problems due to scarring of the larynx tissue, many surgeons are now using laser surgery, which uses an intense laser light as the surgical tool. Carbon dioxide lasers—which pass electricity through a tube containing carbon dioxide and other gases to generate light—are currently the most popular type used for this purpose. In the past 10 years, surgeons have begun using a device called a microdebrider, which uses suction to hold the tumor while a small internal rotary blade removes the growth.

Once the tumors have been removed, they have a tendency to return unpredictably. It is common for patients to require repeat surgery. With some patients, surgery may be required every few weeks in order to keep the breathing passage open, while others may require surgery only once a year. In the most extreme cases where tumor growth is aggressive, a tracheotomy may be performed. A tracheotomy is a surgical procedure in which an incision is made in the front of the patient's neck and a breathing tube (trach tube) is inserted through an opening, called a stoma,

into the trachea (windpipe). Rather than breathing through the nose and mouth, the patient will now breathe through the trach tube. Although the trach tube keeps the breathing passage open, doctors try to remove it as soon as it is feasible.

Some patients may be required to keep a trach tube indefinitely in order to keep the breathing passage open. In addition, because the trach tube re-routes all or some of the exhaled air away from the vocal folds, the patient may find it difficult to speak. With the help of a voice specialist or speech-language pathologist who specializes in voice, the patient can learn how to use his or her voice.

Adjuvant therapies—therapies that are used in addition to surgery—have been used to treat more severe cases of RRP. Drug treatments may include antivirals such as interferon and cidofovir, which block the virus from making copies of itself, and indole-3-carbinol, a cancer-fighting compound found in cruciferous vegetables, such as broccoli and Brussels sprouts. To date, the results of these and other adjuvant therapies have been mixed or not yet fully proven.

What research is being conducted on RRP?

Scientists and clinicians are working to discover more about RRP. While HPV 6 and HPV 11 are known causes, millions of people are exposed to these two viruses without developing the disease. It is not known why some people are more at risk than others or why some cases are much more serious than others.

Researchers funded by the National Institute on Deafness and Other Communication Disorders (NIDCD) are exploring how our genes and immune system may contribute to our risk for contracting the disease as

well as the severity of its symptoms. Researchers are also studying other possible therapies for RRP. In one clinical trial, researchers are investigating whether a common anti-inflammatory drug can inhibit an enzyme that promotes tumor growth in RRP, thus reducing the recurrence of tumors in children and adults with RRP. Researchers are also investigating the effectiveness of a pulsed dye laser, a laser that delivers short intense pulses of light, to determine if it is more effective at preserving the surrounding tissue while removing tumors from vocal folds. In one NIDCD-sponsored clinical investigation, researchers are studying whether a certain dietary supplement can work with the pulsed dye laser to decrease the rate of recurrence of tumors in children with RRP. To read about these and other clinical trials that are recruiting volunteers, go to <http://www.clinicaltrials.gov>.

Where can I get more information?

The NIDCD maintains a directory of organizations that provide information on the normal and disordered processes of hearing, balance, smell, taste, voice, speech, and language. Please see the list of organizations at <http://www.nidcd.nih.gov/directory>.

Use the following keywords to help you search for organizations that can answer questions and provide printed or electronic information on RRP:

- Papillomatosis
- Speech-language pathologists
- Rare disorders
- Laryngology

more →

voice, speech, language

hearing, balance

smell, taste

NIDCD supports and conducts research and research training on the normal and disordered processes of hearing, balance, smell, taste, voice, speech, and language and provides health information, based upon scientific discovery, to the public.

For more information, additional addresses and phone numbers, or a printed list of organizations, contact:

NIDCD Information Clearinghouse

1 Communication Avenue

Bethesda, MD 20892-3456

Toll-free Voice: (800) 241-1044

Toll-free TTY: (800) 241-1055

Fax: (301) 770-8977

E-mail: nidcdinfo@nidcd.nih.gov

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