

RRP

Recurrent Respiratory Papillomatosis



RRP is caused by a virus that infects tens of millions of people.



A publication by



International
RRP ISA Center

Information



International
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“The single most important thing that you can do if you or your family has been touched by RRP is to educate yourself.”

What is RRP?

RECURRENT RESPIRATORY PAPILLOMATOSIS (RRP) is a disease of the respiratory tract caused by the Human Papillomavirus (HPV). It is sometimes called Laryngeal Papillomatosis. It causes tumor-like lesions to grow on the larynx and, in some cases, in the trachea and lungs. They usually cause voice difficulties, including hoarseness and vocal fatigue. Left untreated, the lesions may grow and can cause suffocation and death. [The arrows point to multiple papilloma growths on the larynx caused by a viral infection. Permission to reproduce photo at right courtesy of the University of Pittsburgh Voice Center.]



RRP is NOT the same as vocal cord polyps or nodules. The latter are easily removed, rarely come back, and tend not to cause long term voice problems once they are removed. RRP can cause years of hoarseness or worse.

Few people have ever heard of RRP and the media has paid it scant attention. Although the HPV virus that causes Recurrent Respiratory Papillomatosis is widespread (the CDC estimates that tens of millions of people in the United States are infected with HPV), the prevalence of RRP is very low. As of 2003, it was estimated that there were only 15-25,000 people in the United States with this disease. Some researchers (Steinberg) have estimated the incidence of RRP to be only 10 people out of a million.

One does not catch RRP from someone else who has it. It does not show up as an opportunistic infection even among patients diagnosed with advanced AIDS, which suggests that most people couldn't contract it even if they tried. Genetic

factors and impaired immune responses at the *cellular level of the respiratory tract* appear to play a key role in determining who is susceptible to contracting this disease.

The prevalence of RRP is spread fairly evenly between children and adults. Even after repeated surgical excisions, the lesions often recur. Infants and young children sometimes have to undergo biweekly surgery just to keep their airway open. Some children have undergone hundreds of surgeries under general anesthesia.

Children contract the disease from mothers who have genital HPV (can be genital warts or asymptomatic HPV). Babies are infected as they pass through the birth canal. It is therefore imperative that OB/GYN and pediatric physicians be astute in diagnosing and helping to prevent this disease. Unfortunately, however, an infant's first diagnosis of RRP is all-too-frequently given in the emergency room, since he or she can no longer breathe. The ER visit might have been preventable with early diagnosis and care. The epidemiology of *adult RRP* is less well-understood, but theories have been advanced.

However challenging the disease is in itself, the story does not end there. Treatment-induced injuries (*iatrogenic injuries*) caused by over-aggressive surgery often lead to vocal cord scarring and a lifetime of hoarseness. While there is no cure for this disease, there are prevention and treatment strategies, some of which appear to work better than others. Physicians and patients alike need to be aware of the pros and cons of these various strategies.

Society often responds to RRP patients with fear, and schools and employers have been known to discriminate against them.

The disease may flare up even after a 20-year hiatus, and the effects of RRP can extend over decades. Clinical depression and a sense of stigma, hopelessness and isolation are not unusual in patients and their families.

While the human cost of RRP is devastating, the economic cost is staggering. In the United States, the lifetime cost for RRP can run into hundreds of

thousands of dollars. Single mothers are especially hard-hit because they have to choose whether to stay home and care for a child with a life-threatening disease or work several jobs just in order to pay their rent and cover their medical bills.

In developing countries, treatment is often marginal to non-existent. Untreated, this disease is as deadly as cervical cancer, which is also caused by HPV. To say the disease is a heartbreaker is an understatement.



International RRP ISA Center

The International RRP ISA Center, is a non-profit, charitable foundation. Contributions are tax-deductible under section 501(c)(3) of the Internal Revenue Code (Federal ID 91-2156850).

We have been providing information, support and advocacy (ISA) to the RRP community since 1998. It is not our intention to offer medical advice or establish a doctor-patient relationship with our constituents. We hope that our extensive resources will be helpful to patients and health care professionals and also support the relationships that RRP patients establish with their physicians of choice.

We have an internationally-based Board of Directors that includes several health care professionals including an otolaryngologist (M.D.), working parents of children who have RRP, adults with RRP, and individuals living in developing countries. We also have a world-class RRP scientific advisory panel.

Hundreds of patients, families and health care professionals have completed RRP ISA's online survey. We count all of these individuals as members of our organization.

Goals & Activities

- To educate RRP patients and families so that they can make more informed treatment decisions.
- To create an empowering and supportive community network for those afflicted with RRP.
- To improve treatment of RRP and eventually find a cure.
- To make distributions to RRP-related organizations and to RRP patients and families who cannot afford medically necessary treatment.
- To sponsor educational forums that will provide treatment and research-related information to physicians, patients and families.
- To advocate on behalf of patients whose insurance coverage has denied benefits for RRP.
- To advocate with healthcare professionals, when appropriate, on behalf of individual patients.
- To develop and maintain a powerful relational database that will help us offer more personal service and enhance our understanding of the demographics, epidemiology and impact of RRP on patients and their families.
- To participate as an engaged member of the RRP Task Force, as first constituted by the Centers for Disease Control.
- To sponsor research on ways of treating and curing RRP. (In spring of 2004, for example, we funded a "Long Peptide HPV Therapeutic Vaccine" Research Study through the Institute for Medical Research at North Shore – Long Island).
- To educate the public and raise media awareness of RRP.

Website

Our website is a resource through which health care professionals, patients and families may become better informed, network and exchange information. It was established in 1998 and includes sections on

- Treatment Trends
- Research Trends
- RRP ISA Database and Online Survey
- RRP ISA Data Analysis Reports
- Advocacy/Self-Advocacy
- Frequently Asked Questions (FAQ)
- RRP-Related Resources & Links
- Library (featuring hundreds of abstracts and articles)
- Message Boards
- RRP Chat Room
- PowerPoint Presentations & In-service Training
- Guestbook

For information on these topics, please visit

www.rrpwebsite.org

You can participate online...

Patients and families are encouraged to complete RRP ISA's *Online Patient Survey* to add to our understanding of RRP.

Healthcare providers are encouraged to complete RRP ISA's *Online Healthcare Provider Survey*, which we will use as a referral source for patients.

Unfortunately, many children are first diagnosed with RRP in the Emergency Room. OB-GYN and Family Care physicians urgently need to know more about RRP.

"You are an excellent resource for patients, families, and health care professionals. Your motivation to continually discover better treatments is infectious (no pun intended), and you seem to distill the latest research and reports with minimal bias. As a busy otolaryngologist who sees patients with some relatively uncommon diseases that don't get much NIH or popular press support or coverage, I have appreciated having resources such as yours to which to refer my patients."

—Lisa A. Orloff, M.D.

Professor of Otolaryngology, UCSF School of Medicine

"There were several situations in which informed decision-making required immediate access to information. This would have been impossible without RRP ISA. Crisis counseling, guidance, and case management resources were made available quickly and in a most caring manner. RRP ISA even talked with and met with my physicians. We are most fortunate to have such dedication, compassion and competence in providing information, support and advocacy to patients and their families."

—Kathy B.

A RRP patient who converted to laryngeal cancer. This patient may be contacted through RRP ISA.

Partial Listing of Website Articles

- HPV Vaccine Updates
- HPV Implicated in Laryngeal & Lung Cancer
- Artemisinin & HAMLET in Treating HPV Infections
- RRP Focus Session PowerPoint Presentations
- Celebrex Multicenter RRP Study Announcement
- AAO Celebrex PowerPoint Presentation by Mark Shikowitz, MD
- Effect of Laryngeal Acid-Reflux on RRP
- On RRP, Mitomycin C, Cidofovir and Vocal Cord Scarring by Steven Gray, MD
- Risk Factors for Juvenile Onset RRP (JORRP)
- RRP Task Force Practice Guidelines for Children

Partial Listing of Treatment-Related Issues

- Surgery Using Cold Steel
- Surgery Using the Laser
- Surgery Using the Surgical Microdebrider
- Indole-3-Carbinol & Diindolylmethane (DIM)
- Interferon(s)
- Cidofovir (HPMPC)
- HspE7
- MMR & Mumps Vaccine
- GERD & Laryngeal Reflux Disease

Partial Listing of Frequently Asked Questions (FAQ)

- How is RRP Diagnosed?
- Who Gets RRP?
- Is RRP Contagious?
- I Have Genital Warts. Is There a Risk to My Unborn Baby?
- What Course Does RRP Usually Take?
- What Can I Do about Burnout and Depression?
- Is There Anything that is Known to Make RRP Worse?
- How Can RRP Patients and Families Get in Touch with One Another?

Future Plans

As of December 2004, these are a few of the projects that RRP ISA is working on:

- Development of a study of the long-term psychosocial effects of RRP on children.
- Writing grants for innovative research projects such as the “Long-Peptide HPV Therapeutic Vaccine” research study.
- Organizing local RRP focus sessions for patients, physicians and researchers. They will be modeled after the 2004 RRP Focus Session that RRP ISA co-sponsored in New York City.

The 2004 RRP Focus Session agenda included presentations by:

- (1) *Robert Bastian, MD, on “Two Methods of Cidofovir Injection in the Office Procedure Room”*
- (2) *Bettie Steinberg, PhD, on “Host Immune Response in RRP”*
- (3) *Tom Broker, PhD, on “Creating Greater RRP/HPV Awareness & Comments on HPV Vaccines”*
- (4) *Mark Shikowitz, MD, on “Celebrex and Cox-2 Inhibitors”*
- (5) *Gracelia Andrei, PhD, on “Cidofovir Mechanisms in Suppressing Papilloma”*
- (6) *Nigel Pashley, MD, on “Mumps and MMR Vaccine for RRP”*
- (7) *Brian Wiatrak, MD, on “Pediatric RRP Issues”*

- Development of audio-visual materials that will help new patients and families learn about RRP and will provide in-service training to healthcare professionals.
- Data development to expand our knowledge of the epidemiology, demographics and psychosocial impact of RRP.
- Expanding our capacity to offer coaching, counseling, advocacy and case management to RRP patients and families.

How to Help

Grant funding has been drastically reduced in today's economic environment. We must depend on donations to make it possible to present at medical conventions, maintain our website and database, support patients and families, and offer funding to assist in treatment and research.*

You may make donations by credit card through our website or mail us a check using the form on next page. You may also make a **United Way** contribution through monthly employer deductions, indicating that you want these tax-deductible donations to be sent to the **International RRP ISA Center**. It is important that your designation show our name and **Federal ID (91-2156850)**.** For further information, please visit www.rrpwebsite.org/supporter.htm.

Contact Information:

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** Please note that RRP ISA has its own budget and is entirely distinct from any other organization that may be affiliated with the RRP community.*

*** We are registered with the Washington State Secretary of State's Charity Division. For more information, call 1-800-332-GIVE.*

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Donation Form

The **International RRP ISA Center** is a non-profit, charitable foundation. Contributions are tax-deductible under section 501(c)(3) of the Internal Revenue Code (Federal ID 91-2156850).

YES, I want to support the work of RRP ISA. I am sending a donation of:

- \$25 \$50 \$75
 \$100 \$250 \$500
 Other (\$ _____).

I pledge \$ _____ a month / quarter / semi-annually (circle one).

I am sending a check made payable to the **International RRP ISA Center**.

Please charge my: Visa MasterCard
 AMEX Other _____

Card Number _____

Expiration Date _____

Signature _____

Date _____

Your Name _____

Patient's Name _____

Address _____

City _____ State _____ Zip _____

Country (if not USA) _____

Phone (Day) _____ (Evening) _____

Email _____

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