Juvenile Recurrent Respiratory Papillomatosis (JRRP) Overview and General Information for Families, Schools, and Health Care Providers

Mass. Eye and Ear provides family-centered care to children with JRRP. It is one of several treatment centers in the country that use KTP lasers and a Cidofovir as well as an Avastin (bevacizumab) protocol.

Mass. Eye and Ear has established a team approach in which the physicians, nurses, child life specialists, social workers, speech therapists, parents, care givers, etc. work together for the care of the patient. Education is an integral part of the treatment plan in helping patients and family members come to terms with the disease.

What is JRRP and who is at risk?

JRRP stands for Juvenile Recurrent Respiratory Papillomatosis. JRRP is a condition that causes the growth of the growth of viral papilloma (warts) on the surface of the respiratory tract. Papillomata may occur anywhere in the respiratory tract (nose, throat, or lungs) but mostly affect the larynx (voice box). The growths may vary in size and often grow very quickly. Papillomata in the larynx are benign (not cancerous) and can be managed with periodic surgical intervention.

There is no cure for JRRP.

JRRP is caused by Human Papilloma Virus (HPV). This virus is very common and there are more than 100 different strains. Many people have been infected with this virus, even if they do not have symptoms. Different strains can cause different types of illness. HPV is most familiarly associated with cutaneous warts on the hands or feet, genital warts, and cervical cancer. There are two strains of HPV that cause JRRP: 6 and 11, with 11 typically causing more severe disease.

Risk factors include being a firstborn child, children born to mothers younger than 20 years of age, and children born to mothers with active genital warts during pregnancy who deliver vaginally. Parents should check with obstetricians or midwives with questions.
JRRP affects males and females equally, and the average age of diagnosis is 3.8 years of age. However, this does not exclude patients that are younger or older than 3.8 yrs. The incidence in the United States is estimated at 4.3 per 100,000 children and 1.8 per 100,000 adults. Age may be more indicative of prognosis than HPV type: children who develop the disease earlier in life oftenhave worse prognoses.

In children older than 5 years of age who are diagnosed with respiratory papillomatosis, sexual abuse should be considered.

Diagnosis of JRRP

JRRP is a pediatric disease. Patients with JRRP are often initially diagnosed with one or more of the following signs and symptoms:

- Stridor (noisy breathing)
- Chronic cough
- Failure to thrive
- Shortness of breath
- Snoring
- Croup
- Asthma
- Difficulty speaking (Dysphonia)
- Aphonia (loss of voice)
- Hoarseness

It is critically important to recognize the symptoms of JRRP before potential life threatening obstruction develops from the papillomata.

JRRP is diagnosed first by symptoms, and then either by viewing the larynx in the office with a scope or by taking the child to the operating room to inspect the airway while the child is asleep under anesthesia. **The only way to absolutely confirm JRRP is by a biopsy.**

Is JRRP Contagious Peer to Peer?

Children cannot catch JRRP (or HPV) by playing with each other or by sharing utensils. Siblings cannot be infected through regular household contact.

Treatment

The goal in treatment is to clear the airway of papillomata and to prevent or reduce the occurrence of new papillomata. Surgical interventions include a debulking procedure to reduce papillomata, using a potassium titanyl phosphate (HTP) laser, carbon dioxide (C02) laser, microdebrider, or pulse dye laser (PDL).

Your child's physician may also prescribe an antivirial medication, especially for children who require more than four surgical treatments a year.

Prevention

There is currently only one vaccine available in the US to prevent HPV, the virus that causes JRRP. Gardasil9 is a vaccine that provides protection against the four HPV types included in the first Gardasil (6, 11, 16 and 18) as well as five additional types (31, 33, 45, 52, and 58). Types 6 and 11 are most associated with genital warts, as well as JRRP, and the others, especially16 and 18, are associated with cervical cancer. Cervarix, bivalent vaccine containing only types 16 and 18, is no longer available in the US and would not have provided any protection against the HPV types causing JRRP.

Gardasil9 is approved for people 9-26 years of age, is very safe, is typically well-tolerated, and causes a strong immune response providing long-term protection against HPV. The current recommendation is for both boys and girls to receive 2 doses of the vaccine at least 6 months apart starting at age 11. Adolescents and young adults 15 and older, need to receive the

previous standard of 3 required doses. Infants and young children cannot be protected directly, but may be if their mothers were vaccinated as children/young adults prior to HPV infection.

Several additional FAQs from parents addressed to Dr. Madhavan

1. If a mother has child with JRRP, what is the chance her next child will be infected? Is there anything she should do to take precautions? Should she have a caesarian? Anything else?
   First-born children are at greater risk of developing JRRP during delivery, but there is no way to predict the individual risk for a subsequent child as the level/presence of HPV will vary over time. Women should receive all recommended care to make sure that there are no active HPV infections that would raise the risk of transmission to a baby during delivery. They should also discuss with their obstetricians regarding their personal risk factors that might make a caesarian delivery necessary.
2. Until what age is the new vaccination recommended?
   "Catch-up" vaccination is currently recommended for women up to age 26 and for most men up to age 21 (in some cases 26) who did not receive the full HPV series as adolescents. However, the FDA recently approved HPV9 to be given to adults up to age 45 as they might still receive protection from virus types they have not already been exposed to, but may be in the future. The official CDC recommendation (from the Advisory Committee on Immunization Practices) has not yet changed, but may do so in the near future.
3. If a child is already infected with HPV and has JRRP, is there any role for vaccination at this point?
   Absolutely, as there are many types of HPV. Even if a child has JRRP and has type 11, for example, receiving the vaccine would still provide protection in the future against the 8 other types that are included.

Acknowledgements:
Many thanks to Dr. Vandana Madhavan of the Pediatric Infectious Disease division of the Massachusetts General Hospital for Children for her critical read and input of this section regarding the infectious nature of JRRP. Also, please see RRP Foundation for more information.Your child's physician may also prescribe an antiviral medication, especially for children who require more than four surgical treatments a year.