WHAT IS JUVENILE – ONSET RECURRENT RESPIRATORY PAPILLOMATOSIS (JORRP)?

JoRRP is a rare disease of the respiratory tract. JoRRP is a condition that causes the growth of viral papillomas (warts) on the surface of the respiratory tract. When the papillomas grow inside the voice box they result in hoarseness but more importantly, they can restrict breathing. They can grow rapidly and can obstruct or block the airway causing breathing difficulties. There are two strains of Human Papilloma Virus (HPV) that cause JoRRP; genotypes 6 and 11.

Papillomatosis is found typically in the larynx, it can also be found in the nasopharynx, tracheobronchial tree or even in the lungs but this is less common. The course of the disease is unpredictable; it can vary from mild to severe. The symptoms of an infant can include hoarse or weak cry progressing to no cry. A child may have a hoarse voice progressing to no voice. Some children experience minor symptoms and spontaneous and complete remission at puberty, while others require multiple surgeries throughout childhood.

WHAT CAUSES JORRP?

It has been estimated that HPV virus may be present in the genital tract of up to 25% of all women of child-bearing age. Many newborns are likely exposed to HPV, however few of them develop JoRRP and the severity of the disease varies from one child to another. HPV infection in children occurs most often in birth, during passage through the birth canal, it can also occur prior to birth, through the placenta. Risk factors include; being the firstborn child, children born to young mothers, and children born to mothers with active genital warts during pregnancy.

The incidence and prevalence of JoRRP is estimated at 0.24 to 4.3 births per 100,000. (IJPO 2014)

Photo of larynx with papilomas
HOW IS IT DIAGNOSED AND TREATED?

When JoRRP is suspected, the ENT surgeon performs a laryngoscopy to look into the airway while the child is under general anaesthetic. If papillomas are seen, the genotype is determined from a biopsy taken at this time. It will take 3-4 months for the results. After initial diagnosis, a treatment plan is arranged for the child and this is discussed with you.

To manage this condition, the papillomas need to be removed by the ENT surgeon. This may be necessary every 4-6 weeks, depending on the severity of the disease. Papillomas can vary in size and grow very quickly, they grow back even when removed. Treatment is designed to keep the airway clear, to improve voice quality and to avoid damage to the vocal cords. The current treatment is surgery, the aim of the surgery is to remove most of the papillomas and protect the larynx and vocal cords.

Additional therapies have been used alongside surgeries. To date the results of these have been mixed or not yet fully proven, the risks of these therapies outweigh the benefits.

HPV VACCINES

There is now a vaccine against HPV genotype 6,11,16 and 18. This will protect children from developing JoRRP in the future, if their mothers were vaccinated as children / young adults.

CANCER

The HPV genotype 16 and 18 can cause cervical cancer in adults. These types that cause cancer, are called high-risk. HPV 6 and 11 are extremely low-risk for cancer.

THE SURGERY

The procedure is called a micro laryngoscopy and bronchoscopy (MLB). This is done as a day procedure. Your child will need to arrive to the hospital fasting, you will receive fasting guidelines from the hospital prior to this. They will be admitted to the day ward here in the hospital, on the morning of the procedure. From here, your child will go to theatre and have an anaesthetic; while your child is asleep under anaesthetic, the surgeon passes a scope with a camera into their airway so that the vocal cords are in view. The surgeon then removes the growths. The reasons for this are; to protect and ensure patency of the airway, to preserve the voice and to avoid damage to the vocal cords.

WHAT HAPPENS AFTER THE SURGERY?

After the surgery and some time on the recovery ward, your child will return to the ward to wake up fully from the anaesthetic. If they are feeling well and have taken something to eat and drink, they can go home the same day.

You will be given the contact details of the ENT department and airway management clinical nurse specialist. If you have any questions, you can contact them.

If your child develops any breathing problems, you must bring them to the Emergency department of your nearest hospital.

WHAT IS THE OUTLOOK FOR CHILDREN WITH JORRP?

Unfortunately JoRRP has no cure. Although surgery is the treatment of choice, it does not prevent the growths from recurring. The management of the condition requires repeated removal of the papillomas sometimes as often as every four to six weeks.

WHEN WILL IT DISAPPEAR?

It is usual for the disease to go into remission at puberty, a small percentage will continue to have it into adulthood.
IS JORRP CONTAGIOUS?
No it is not contagious, children cannot catch JoRRP or HPV from each other.

NEXT PREGNANCY?
A study was conducted to see if there was sufficient reason to perform caesarean section in preventing transmission of recurrent respiratory papillomatosis. The findings showed a very limited role in prevention.

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<thead>
<tr>
<th>TELEPHONE NUMBERS</th>
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<tbody>
<tr>
<td>Surgical Day Unit (SDU)</td>
<td>01 428 2616</td>
</tr>
<tr>
<td>Medical Day Unit (MDU)</td>
<td>01 409 6525</td>
</tr>
<tr>
<td>Clinical Nurse Specialist - Airway Management Team</td>
<td>01 409 6100 - bleep 8662</td>
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<tr>
<td>Emergency Department</td>
<td>01 409 6326</td>
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Remember: Ask your nurse if you are unsure about anything about your child’s care.

Additional instructions

Developed by Roisin Mullan Clinical Nurse Specialist and John Russell Paediatric ENT Surgeon

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