

## **Recurrent Respiratory Papillomatosis**

*Dr. Arun Pugalenti, Dr. Sadasivam Suresh Department of Paediatric Respiratory & Sleep Medicine  
Mater Children's Hospital Brisbane, Australia*

**Description:** Respiratory papillomatosis is a rare condition with benign, wart like lesions in the respiratory tract. It has a bimodal distribution with juvenile onset occurring in children and adult onset occurring in the fourth decade of life. The incidence is 4.3 per 100,000 in children. Papilloma occurs in larynx, trachea, bronchi and lung parenchyma. Because of the recurring clinical pattern it is also referred to as recurrent respiratory papillomatosis (RRP).

**Causes:** The human papilloma virus (HPV) is the causative virus that causes the papillomatous lesions. HPV types 6 and 11 cause most of the cases of recurrent respiratory papillomatosis. Type 11 is the more aggressive form with extensive lesions mainly in the laryngeal/tracheal airway. Different periods of recurrence and remission are connected with the existence of the virus in the latent form, which activates periodically. Periods of remission and the course of the disease are difficult to predict.

**Clinical Presentations:** Clinical presentation depends on the site and extent of papillomatosis changes. Following symptoms and signs may be noted: hoarseness, weak cry, stridor, aphonia, dyspnea, recurrent laryngitis, recurrent pneumonia, chronic cough, failure to thrive and sometimes can present with life threatening upper airway obstruction. The lesions are usually localized within the larynx, but spread to other areas of the respiratory tract can occur. Prognosis is worse when there are papillomas in the lower airway as they have an aggressive clinical course.

**Investigations:** Papillomatosis is diagnosed by clinical inspection of the airway and by tissue biopsy of the lesions. The virus signal can be identified in the tissue biopsy, but the intensity does not correlate with severity. Pulmonary function tests [spirometry] are helpful in demonstrating airway obstruction in the flow volume loop. Radiological imaging in the form of CT scan is helpful in delineating severe lesions.

**Treatment:** The goal of the treatment is to relieve airway obstruction, improve voice quality and minimize the risk of recurrence. No single treatment option has been shown to be effective in the eradication of papillomatosis. Total surgical removal of the lesions is not possible in most cases. Carbon dioxide laser is the most widely used surgical tool for removal of the papillomas. It does not prevent the regrowth of the lesions. Other therapies include cryotherapy; direct painting of the lesions with podophyllin, injection of cidofovir into the lesions. Interferon therapy may be used in

particularly aggressive disease, but should not be continued beyond 12 months unless the disease responds.

Most usual course of the disease is for the papillomas to continue to grow locally despite surgical removal and without significant spread. Over time, majority of the cases undergo spontaneous remission. Malignant change to squamous cell carcinoma has been reported in 3% - 5% mainly in adults. A vaccine for prevention of genital HPV infection in young women has recently been introduced onto national immunisation programs. This vaccine protects against HPV types 6, 11, 16, and 18 and may potentially decrease the incidence of RRP.

**References:**

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