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CASE REPORT

# Juvenile Laryngeal Papillomatosis

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**Summary** Always ask about hoarseness and quality of voice in a history of any child presenting with cough or asthma-like symptoms. Children presenting with what appears to be an acute onset of hoarseness, without any physical signs of airways obstruction, should be reviewed after two weeks. If there is chronic hoarseness, referral to an ENT specialist should be considered with a view to laryngoscopy. If the child develops clinical signs of acute airway obstruction such as stridor or respiratory distress, prompt paediatric review is indicated. When referring, it is important to emphasise whether or not there is chronic hoarseness in order to differentiate the diagnosis from croup. Juvenile Laryngeal Papillomatosis may present with cough, pneumonia, dysphagia, or stridor, as well as hoarseness. These patients are often misdiagnosed as having asthma or allergies.


## Case history

C.F. was aged three when he joined our general practice in March 1999. He was the eldest of three children. He had always been noted to have a husky voice. Due to this, and a general reluctance to speak, he was referred to a Consultant Paediatrician at the local District General Hospital in November 2000. The Paediatrician noted that he was ''sometimes a bit chesty with a wheeze and tended to snore at night'' but that there was no dyspnoea. The hoarseness was thought to be ''probably benign in origin''.

Apart from his routine vaccinations he had no further contact with the practice until June 2001, when he presented with a history of a pronounced barking cough and stridor at the age of five. He looked well, and the most likely diagnosis was thought to be tracheitis. The attending general practitioner (GP) wondered whether the problem might also have an allergic origin since his eyes had been watering recently. Steam inhalations and an oral antihistamine were prescribed.

Five weeks later his mother brought him to the practice again. She reported that his hoarseness was ongoing, and that he had noisy breathing at night but that this didn't wake him. On examination his chest was clear. A diagnosis of possible asthma was made, and he was prescribed a trial of treatment with a high dose inhaled corticosteroid

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and a bronchodilator. He was referred to the practice asthma clinic to be seen the next week. When he attended the clinic one week later, the asthma nurse reported that C.F. was ''reaching out to grab his inhaler at night-time'', and asked the supervising doctor for permission to change the inhaler device.

Sadly, this was the last occasion that C.F. was seen in our practice. In September 2001 he collapsed at home and his family gave him mouth-to-mouth resuscitation. He was admitted urgently to hospital. On admission, intubation proved to be difficult, but was eventually successful using a very small endotracheal tube. C.F. died three days later on the Intensive Care Unit. Post-mortem examination showed marked narrowing of the larynx in the region of the vocal folds, due to a very large number of papillomas distributed throughout the upper respiratory tract. Histology confirmed these to be papillomas consistent with a diagnosis of Juvenile Laryngeal Papillomatosis.

## Discussion

Juvenile Laryngeal Papillomatosis, sometimes known as Juvenile Respiratory Papillomatosis is a relatively rare condition. Its incidence has been estimated at between 3.84 to 7 cases per million population per year [1,4]. It is caused by the human papilloma virus types 6 and 11, these types being implicated in genital condylomata rather than cervical cancer [2] There is strong circumstantial evidence that the condition is acquired during childbirth. Retrospective studies suggest that around 30% of children who are diagnosed with the condition at a young age have a mother who had genital condylomata at the time of their birth [1]. Juvenile Laryngeal Papillomatosis typically presents in young children — most cases present by the age of four — but presentation can be delayed for some years, occasionally up to the age of 15, since there are rarely any other distinguishing features. There is a slightly higher incidence in first-born children [3].

Hoarseness is the typical initial presentation, together with an abnormal cry. Hoarseness in early childhood is a common problem. In most cases it occurs acutely as a result of infectious croup, and the symptoms prove self-limiting within one to two weeks. When hoarseness persists, however, careful thought should be given to the differential diagnosis. A weak cry since birth suggests a congenital laryngeal problem such as vocal cord palsy, or a web or cyst taking up space in the larynx. Laryngomalacia, the most common cause of stridor in the newborn period, rarely causes hoarseness and this can be a useful distinguishing feature. Causes of later-onset symptoms include: foreign body inhalation in a child who has an abrupt onset of hoarseness after a choking episode; sub-glottic haemangioma in a child with an accompanying strawberry naevus enlarging on the head and neck; and laryngeal thrush in a child who has been using inhaled corticosteroids without a large volume spacer.

Vocal nodules, sometimes called ''screamer's'' or ''singer's'' nodules, are the most common cause of chronic hoarseness in childhood. They tend to occur in the child who talks too loudly, for too long and with too much effort, and can also be caused by chronic coughing and post-nasal drip.

The diagnosis of vocal nodules must be confirmed by laryngoscopy. This is primarily to exclude the important differential diagnosis of Juvenile Laryngeal Papillomatosis. Any associated symptoms suggesting airway obstruction should prompt an urgent ENT referral given the potential for rapid growth of these lesions.

Management of Juvenile Laryngeal Papillomatosis can be difficult, and tends to be surgical. Some authorities report that children may require tracheostomy, although some believe that this should be avoided unless absolutely necessary. Papillomas can be removed surgically by shaving, and treatment with the $CO_2$ laser is widely used. However, many treatments may be required over a prolonged period. Immunotherapy with alpha interferon has been used, but generally has not been found to produce a sustained benefit [3].

With a tragic case such as this there is a need to see where lessons might be learnt. Against a background of very common presentation of upper respiratory symptoms in both general practice and in hospital, a number of points can be identified. Firstly, C.F.'s history of hoarseness was longstanding — he had, according to his mother, always been noted to have a husky voice. Secondly, he also had a history of noisy breathing at night, which, in the context of the prolonged history of hoarseness, could have alerted attending doctors to the possibility of an upper respiratory cause for his symptoms. Thirdly, both symptoms — the hoarseness and the noisy breathing — had persisted over a period of several weeks' observation. Finally, and most importantly, the experienced nurse running the general practice asthma clinic elicited the unusual history of him ''grabbing for his inhaler at night'', but the

significance of this wasn't appreciated at the time.

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