

# Epiglottic Hypoplasia: A Natural Model for Supraglottic Laryngoplasty

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## ABSTRACT

Non-syndromic hypoplasia of the epiglottis presenting without symptoms at middle age is an extremely rare entity. We report a 50 years female who presented with oral ulcers and incidentally was found to have an asymptomatic rudimentary epiglottis and thickened bilateral aryepiglottic folds forming an inverted funnel shaped supraglottis protecting the airway from any aspiration. This case demonstrated a functional model of natural supraglottic laryngoplasty which can be used in treating intractable aspiration, similar to the Steam boat modification of Biller's technique of supraglottic laryngoplasty. This natural example can be used to design or scrutinize the existing models of supraglottic laryngoplasty in the treatment of intractable aspiration.

**Key Words:** *Epiglottic hypoplasia. Agenesis. Congenital. Epiglottis. Anomalies. Supraglottic laryngoplasty.*

## INTRODUCTION

Hypoplasia of the epiglottis can result from an event during the organogenesis leading to the arrest of development of the supraglottis at different stages of development accounting to some presenting early with aspiration and others not until adulthood.

The laryngeal development is first noted at 33 days' of gestation with the appearance of the mesenchymal-arytenoid swellings just adjacent to the cranial end of the laryngotracheal tube. The arytenoid swellings continue to grow cranially and to differentiate into the arytenoid and corniculate cartilages and the primitive aryepiglottic folds. The hypobranchial eminence gives rise to the epiglottic and cuneiform cartilages, completing the supraglottic structure. Failure of epiglottic development is thought to result from defects involving both the third and fourth branchial arches during the period of organogenesis through the 8th week of intrauterine life. This type of laryngeal anomaly usually presents in infancy or early childhood and commonly occurs in association with congenital anomalies of other organ system.<sup>1</sup>

To authors' knowledge, only three cases of hypoplasia of the epiglottis in the middle age have been reported in literature so far, only one of them was non-syndromic.

## CASE REPORT

A 50 years old female presented to the ENT Outpatient Department with a history of recurrent oral ulcers. She did not give any history suggestive of aspiration like coughing, choking or recurrent chest infections nor did she complain of dysphagia, hoarseness of voice or shortness of breath. She was an asthmatic and had undergone endoscopic sinus surgery for benign nasal polyps 15 years prior to that. The only regular medication she was taking was bronchodilating inhalers.

On general physical examination, she was a well nourished, afebrile adult female with normal vital signs. Examination of the oral cavity revealed multiple aphthous ulcers on the oral mucosa. Flexible rhinolaryngoscopy showed a hypoplastic epiglottis and thickened bilateral aryepiglottic folds forming a circumferential ring of thickened mucosa joining the arytenoids posteriorly (Figure 1). The position of the larynx was normal and vocal cord movement with phonation and respiration was normal. The rest of the head and neck examination were normal.

The laboratory tests included full blood count, urea and electrolytes, erythrocyte sedimentation rate, auto-immune antibody screen and sarcoidosis screening

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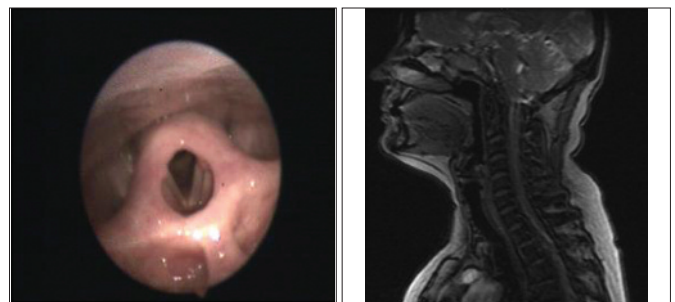
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**Figure 1:** Endoscopic view of the larynx demonstrating the hypoplastic epiglottis.

**Figure 2:** MRI scan showing a normal position of the larynx and a hypoplastic epiglottis.

which were all normal. MRI scan of neck demonstrates a rudimentary epiglottis (Figure 2) with an inverted funnel like configuration of the supraglottic region with the opening angulating posteriorly. A barium swallow examination was performed to assess swallow and to check for any aspiration which was entirely normal. The patient had no obvious abnormalities of the other organ systems. The patient's oral ulcers cleared spontaneously on further review. She had no further evaluation or treatment and was discharged.

## DISCUSSION

Being the first line of defense against aspiration the epiglottis plays a very important and a definitive role in protecting the airway during swallowing. Embryologically, the larynx develops in the floor of the cranial part of the foregut during the 3rd or 4th week of development.<sup>5</sup> The epiglottis is clearly demarcated from the tongue during the 48th day of life. Defects of intrauterine growth at any time before the epiglottis is separated from the tongue are thought to result in epiglottic anomalies, ranging from complete absence, to hypoplasia, to bifid epiglottis.

Hypoplasia of the epiglottis is a result of arrest of the development of the epiglottis in utero during the first 8 weeks of gestation, the extent of development determining the nature of presentation of these cases.<sup>6</sup>

Congenital anomalies of the larynx include atresia, webs and stenosis. They usually present in infancy and early childhood with respiratory and feeding problems. They are often incompatible with life unless the condition is recognized at once at birth and immediate steps are taken to establish an airway.

Hypoplasia of the epiglottis is very rarely found in middle age. To our knowledge, only 3 cases, including syndromic and non-syndromic, in middle age were reported in English literature.<sup>2-4</sup> Roh reported a case of congenital laryngeal anomaly detected in a middle-aged man who had symptoms of laryngopharyngeal reflex.<sup>7</sup> Hong and Yang reported a case of syndromic absence of the epiglottis associated with absent false vocal folds and hypoplastic maxilla.<sup>8</sup> The patient had hoarseness and aspiration. Ali and Snow described a severe congenital laryngeal hypoplasia in a 45 years old man.<sup>2</sup> The patient had no epiglottis, absent aryepiglottic folds, rudimentary vestibular folds, enlarged arytenoids, absent thyroid laminae and grossly thickened cricoid cartilage. He presented only with hoarseness of voice with no history of stridor, dysphagia, aspiration or respiratory infections.

The patient presented at the age of 50, with an incidental finding of epiglottic hypoplasia and a band like thickening of the aryepiglottic folds forming a circumferential

hood-like thickening of the mucosa over the laryngeal inlet with normal and mobile vocal cords and no other congenital anomalies were noted. Despite this laryngeal anomaly, the patient had no symptoms of hoarseness, choking or aspiration and was entirely asymptomatic. The only co-incidental symptom was of oral ulceration which cleared spontaneously on follow-up.

The protective function of the larynx depends on its functional integrity and this could be affected by various factors. The surgical options in the treatment of intractable aspiration should be simple, reliable and not compromising voice or respiration and should be reversible, so far only Biller's technique of tube supraglottic laryngoplasty for the management of intractable aspiration preserving the phonatory function of the larynx satisfies these criteria.<sup>3</sup>

In conclusion, non-syndromic hypoplasia of the epiglottis presenting without symptoms at middle age is an extremely rare entity. Intractable aspiration leading to lethal complications is usually a common outcome of absence of epiglottis or epiglottic anomalies. This shows that the phase in which the development arrests is very important in shaping the symptoms and the outcome of the disease process. This also highlights the adaptive efficiency of the human body to retain the respiratory and phonatory functions. In doing so it has given us a functioning model which could be used in treating intractable aspiration provided the vocal function is normal.

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