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# Subglottic Laryngeal Atresia

## —Asssociated with Tracheoesophageal Fistula—

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A case of congenital laryngeal atresia of infraglottic type in a newborn infant is described for its rarity and importance of early diagnosis.

This case was associated with tracheoesophageal fistula, and died in 5 minutes of life due to asphxia. Failure to intubate in the neonate should raise the possibility of laryngeal atresia and emergency management should be provided.

The lung tissue was normally developing despite the complete obstruction of the outlet of the air and possible amniotic fluid passage.

Congenital laryngeal atresia is a rare congenital anomaly, usually with fatal outcome. Although early diagnosis and emergency surgical intervention could be life-saving it is always difficult to make the diagnosis quick enough. The other problem in this anomaly is the assoication of othe major congenital anomalies which probably themselves are incompatible with life.

In Korea, we have reported a case of subglottic type of laryngeal atresia which was also the victim of neonatal death becauses of failure of early diagnosis[1].

We report this case for two reasons. One is to remind the necessity of early diagnosis. Two is to report the associated tracheoesophageal fistula and normally developed lung tissue in laryngeal atresia.

## CASE REPORT

A premature male infant was delivered by cesarean section after 29 weeks of pregnancy because of hydramnios, premature rupture of membrane and breech presentation. Apgar scores were 1 at 1 minute. Because of severe respiratory difficulty, intubation was tried without success. The baby died of asphyxia at 5 minutes after birth.

On postmortem examination, the larynx was completely obstructed at the subglottic level. The epiglottis and glottis showed no abnormality. Middle sagittal section of the larynx revealed a dome shaped cricoid cartilage completely obstructing the lumen in the subglottic area and the upper end of the trachea(Fig.1,2,3). Histologically the atretic portion of the larynx was composed of irregular cartilage islands, muscle and connective tissue that were irregularly arranged. Glandular component was scattered in them. In spite of multipe serial sections of the larynx, no pharyngotracheal duct was identified. The lower end of the larynx consisted of submucous glands and connective tissue (Fig. 2). The upper esophageal segment ended as a blind pouch at the level of the larynx and the lower esophageal segment started entirely from the middle portion of the trachea and was normally continued to the stomach. A small thymic nodule was seen in the thyroid. The lungs were normal in development and specifically there was no abnormal accumulation of fluid. No squames, however, could be seen in the air

**Fig. 1.** Mid-sagittal sections of laryngc tracheal structures of subglottic laryngeal atresia in this case (Left) and normal larynx of 28 weeks of gestational age (Right). The larynx is completely obstructed by irregularly arranged cartilage island, dome-shaped cricoid cartilage and connective tissue.

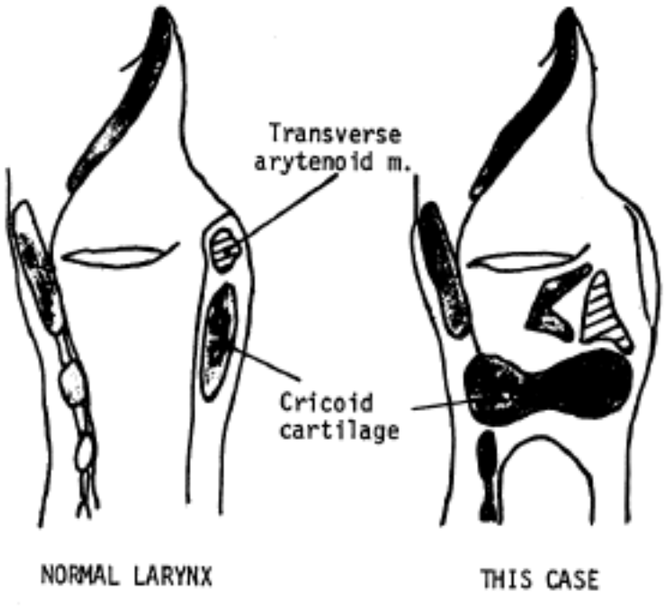


**Fig. 2.** Schematic drawing of mid-sagittal section of the larynx in this case and age matched control in the picture shown Fig. 1.

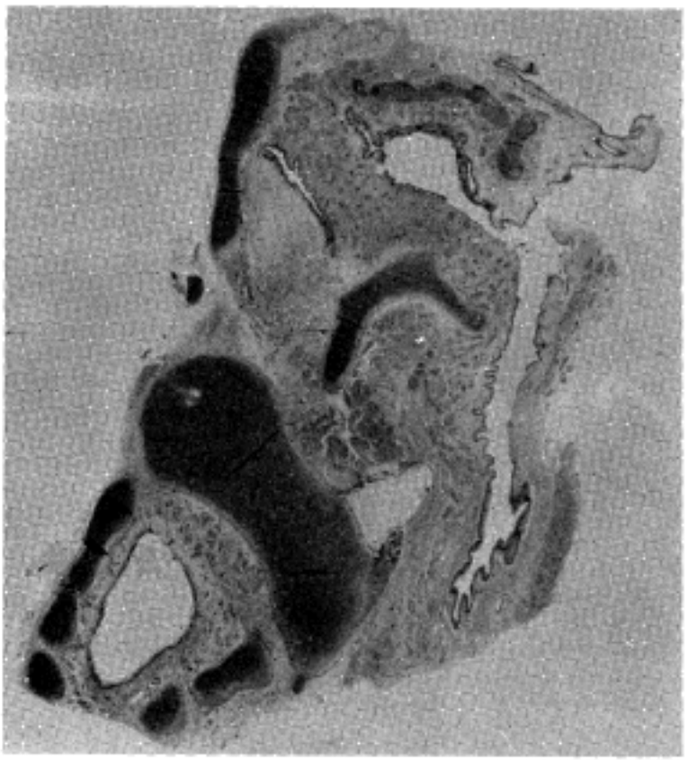

**Fig. 3.** Section from Fig. 1. (Left)
The subglottic region and upper most portion of the trachea is completely obstructed by dome-shaped cricoid cartilage, irregularly arranged cartilage island, muscle and connective tissue in which glandular component is irregularly arranged. (H&E, x1)

## DISCUSSION

The larynx develops in two parts, the infraglottic as a median sagittal cleft in the floor of the pharynx at the upper end of the pulmonary outgrowth, and the supraglottic (transverse part) as a modification of the floor of the pharynx above the sagittal cleft. The vocal cord lie, approximately at the boundary between the two parts. During the third or fourth week the sagittal cleft is almost completely closed by overgrowth of the lateral masses (fifth branchial arches) at the sides. The epithelium fuses, levaing a small channel at the back (ductus pharyngotrachealis). With the development of cricoid cartilage in the lateral masses, the cleft reopens at the eighth-ninth week to form the infraglottic part of the larynx. The failure of reopening is thought to be the mechanism of glottic or infraglottic atresia[5-10]. And in these situations solid dome-like deformity of cricoid contilage is invariably demonstrated as also seen in two cases.

There has been some debate in the literature on

the lung findings in laryngeal atresia. Since Snyder and Rosenfeld submitted evidence that a tidal flow of amniotic fluid into the developing lungs was neccessary for their normal development, it would be reasonable to assume certain types of developmental anomalies could result from it[1]. However, as seen in our previous case (Suh et al) the alveoli were overdistended by serous fluid despite the complete obstruction of larynx except for a minute pharyngotracheal duct. In this case there was not even pharyngotracheal duct. Therefore the lung development does not appear to be directly related to the patency of the larynx. Rankin and Mendelsohn claimed that retained secretions lead to dilatation of the alveoli and so assist the development of the lungs[6,12]. We have not noticed "precipitated protein" in alveoli as in a case of Potter and Bohlender[13]. Associated anomalies in laryngeal atresia include gastrointestinal, genitourinary, skelectal and cardiovascular systems[2]. This case was assocated with type C tracheoesophageal fistula. Lieberman et al reported a similar case who had sublottic laryngeal atresia associated with a tracheoesophageal fistula. But this case managed to survive until 3 years of age by doing emergency tracheostomy in neonatal period[8].

## 참 고 문 헌


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## =국문초록=

### 기관식도루를 수반한 선천성 후두 폐쇄증

—성대하형—


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**강 구·지 제 근**



신생아에서 발견된 성대하형 선천성 후두폐쇄증의 부검증례를 기술하였다.

본 예는 우리나라에서 선천성 후두폐쇄증의 2번째 부검예이며 첫번째에 이어 이번에도 신생아기에 진단되지 못한채 질식사하였다.

본 예는 동반된 기형으로 기관식도루를 가지고 있었으며 현미경적으로 폐는 정상이었다.