December

ASSESSMENT PROGRESS: Total Questions: **10** Questions Answered: **10** Correct Answers: **2**

Question 10

A 10-hour-old term male infant born after an uncomplicated pregnancy and vaginal delivery is having significant stertor when he cries. At rest, his stertor is barely audible. Physical examination of the infant reveals a heart rate of 140 beats per minute and respiratory rate of 44 breaths per minute. Upon cursory inspection no abnormalities are noted in the oropharynx; however, when the infant begins to cry, you detect a lesion **(Figures 1** and **2)** in the posterior pharynx.

Figure 1: Two arrows point to lesion. The single arrow points to the tongue (Photograph courtesy of Dr Beste, Children's Hospital of Wisconsin)

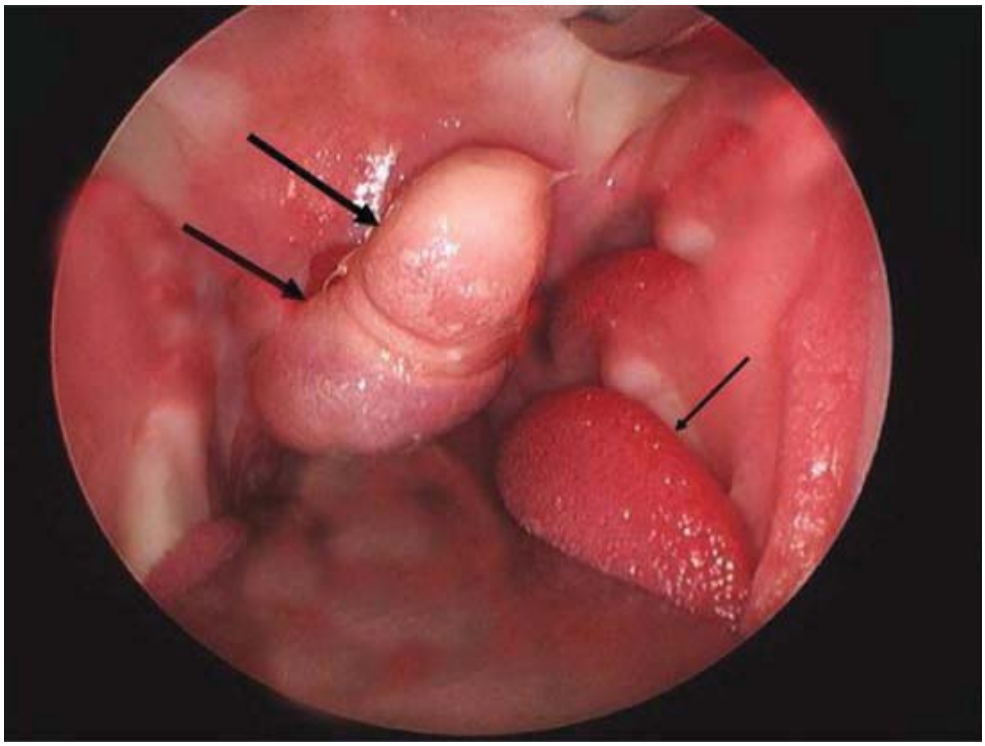
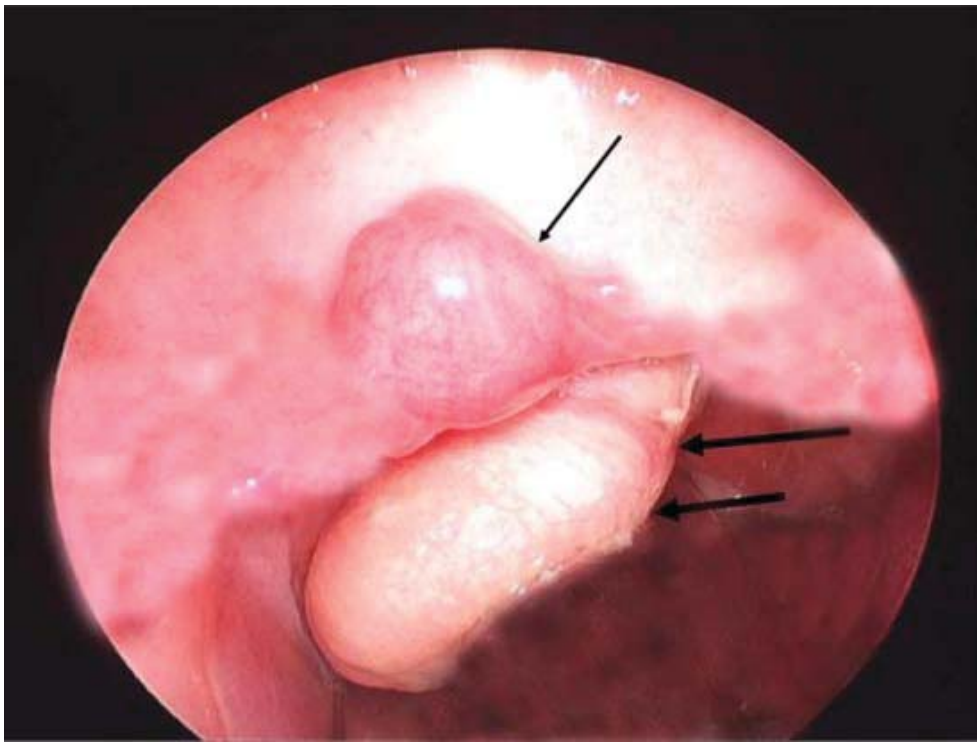


Figure 2: Two arrows point to lesion. The single arrow points to the uvula. (Photograph courtesy of Dr Beste, Children's Hospital of Wisconsin)



Of the following, the lesion MOST likely responsible for this infant's stertor is a(n):

- **A.** epulis
- **B.** first branchial cleft cyst
- **C.** ranula
- **D.** teratoma
- **E.** thyroglossal duct cyst

Incorrect:

Correct Answer: D

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Signs and symptoms of neonates with airway obstruction may differ, depending on the location and severity of the obstruction. Lesions such as a teratoma, which obstruct the airway at the level of the nasopharynx or oropharynx, produce an inspiratory low-pitched sound known as stertor (ie, snoring). If the obstruction is located at the supraglottic level, the airway turbulence produced by the obstruction causes stridor, a higher-pitched inspiratory sound. Obstructions caused by a fixed subglottic laryngeal or cervical tracheal lesion frequently cause stridor during inspiration and expiration. Because intrathoracic pressure rises with exhalation, intrathoracic airway obstructions are commonly associated with expiratory stridor as the airway collapses.

A brief summary of important causes of stertor or stridor in neonates is given in the **Table**.

Table: Common Supraglottic Causes of Noisy Breathing in Neonates*

Site	Congenital Anomaly	Neoplasm	Neuromuscular	Inflammatory
Nose and nasopharynx	Choanal atresia or stenosis	Teratoma		Rhinitis

	Craniofacial anomalies Pyriform aperture stenosis	Encephalocele		Retropharyngeal abscess
Oropharynx or hypopharynx	Macroglossia Lingual thyroid Vallecular cyst Craniofacial abnormalities	Teratoma Hemangioma Lymphangioma	Hypotonia	Retropharyngeal abscess
Supraglottic larynx	Laryngomalacia Laryngocele	Hemangioma Lymphangioma Papilloma		

Feedback

At least 85% of cases of stridor are from abnormalities present at birth. The stertor in the infant in this vignette was caused by a nasopharyngeal teratoma. Teratomas, which can be benign or malignant, are the most common neonatal tumors, accounting for 23.5% of all neonatal tumors. Head and neck teratomas account for 14% of teratomas presenting during the first 2 months after birth. Neonatal oropharyngeal teratomas are rare. Teratomas are classified into four general types:

dermoid, or hairy polyp, the most common type, contains epidermal and/or mesodermal elements

teratoid contains ectoderm, mesoderm, and endoderm, but is poorly differentiated

true teratoma contains all three germ layers that are more organized or differentiated into histologically recognizable tissue

epignathus, a highly differentiated tumor of recognizable organs and limbs

The infant's teratoma was a hairy polyp that contained epidermal (hair follicles) and mesodermal elements. More than 50% of children with this condition are younger than 12 months of age. Hairy polyps normally present as pedunculated tumors of the digestive tract and are the most common congenital tumor of the oropharynx and nasopharynx. Although they are not commonly detected on prenatal ultrasonography, a history of polyhydramnios is often noted at the time of delivery because of impaired ingestion of amniotic fluid in utero. Clinical presentation depends on the size and location of the tumor. In approximately 60% of cases, the teratomas are attached to the lateral wall of the nasopharynx or the superior surface of the soft palate; in the remainder of cases, the site of attachment is most frequently the oropharynx. The infant in the vignette had a lesion that was attached to the superior aspect of the soft palate by a stalk, disappeared into the nasopharynx with exhalation, and became visible in the posterior pharynx with vigorous inhalation.

Hairy polyps usually present as isolated defects that are not associated with a particular congenital syndrome. The pedicle is most often pink and sharply demarcated from the gray or white distal bulk of the malformation. Malignant transformation has not been noted. Computed tomography or magnetic resonance imaging may help to differentiate it from more serious disorders such as encephalocele or meningoencephalocele and help to delineate the extent and origin of lesions before surgical excision.

Epulis, also known as Neumann tumor, is a congenital granular cell tumor that arises from the mucosa of the gingiva. Unlike the lesion in the infant in this vignette, epulis occurs primarily in the mandible, maxilla, or tongue, most commonly from the maxillary alveolar ridge. Epulis is seen only in newborns. It is uniformly benign and treated with simple excision. A smaller lesion may regress spontaneously. Epulis does not damage future dentition.

The branchial apparatus consists of the arches, pouches, grooves, and branchial membranes that contribute to the formation of the head and neck. The first branchial groove is the ectoderm that lies between the first and second branchial arches. The dorsal portion of the groove deepens to produce the external auditory canal. The ventral portion regresses. First branchial cleft anomalies occur when ectodermal cell nests are buried or the ventral portion of the first branchial groove is not obliterated. First branchial cleft anomalies most often present as cystic masses in front of, behind, or below the ear lobe or in the submandibular region. They do not present as pedunculated masses in the posterior pharynx.

A ranula is a rare midline cervical cyst caused by the blockage of a sublingual salivary duct. There are two types of ranulas. Simple ranulas are retention cysts of salivary glands lying in proximity to the lining of the mucous membrane of the oral cavity. They usually present as painless, unilateral, fluctuant, translucent bluish masses that may affect sucking and swallowing. Plunging ranulas are cysts that extend beyond the mucous membrane of the oral cavity down through the mylohyoid muscle into the fascial planes of the neck and can present as masses in the neck without a visible intraoral lesion.

Thyroglossal duct cysts are among the most common congenital neck masses and the most common midline cervical anomaly encountered in children. Thyroglossal duct cysts form as the result of embryologic remnants of the descending thyroglossal duct. Cysts may form anywhere from the base of the tongue to the lower midline of the neck.

The most common presentation of a thyroglossal duct cyst is a painless cystic neck mass in the midline near the hyoid bone; however, rarely thyroglossal cysts may present as oral masses at the base of the tongue that may cause airway obstruction and stridor. Thyroglossal duct cysts do not present in the posterior nasopharynx.

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American Board of Pediatrics Content Specification(s):

17_EENT_mouth_neck: Know the various causes of stridor in the newborn and how to assess severity

17_EENT_mouth_neck: Know the normal development of the nose, mouth, throat, and neck

17_EENT_mouth_neck: Know the clinical manifestations of branchial cleft cysts

17_EENT_mouth_neck: Know the clinical manifestations and approaches to therapy of neck masses in the newborn infant

14_Hematology_Oncology: Know the clinical and laboratory features and management of neonatal teratoma

Complete Assessment