DEVELOPMENTAL PROBLEMS ASSOCIATED WITH THE BRANCHIAL REGION

Normal Variants of the Thyroid Diverticulum

These are represented by a pyramidal lobe of thyroid gland and/or a ligament (thyroglossal) extending superiorly from either a pyramidal lobe (if it exists) or the thyroid isthmus up to the hyoid bone. The foramen cecum of the tongue is a normal marker of the site of the thyroid diverticulum.

Thyroglossal (Duct) Cyst

This is the most common anomaly of the head and neck. It is a cyst located beneath the skin of the anterior midline of the neck, usually just below the hyoid bone. It is the second most common cause of a neck mass in children (the most common being lymphadenopathy). A very helpful clue in diagnosing thyroglossal cysts is that they move superiorly when the child protrudes his/her tongue. If an infected cyst ruptures through the skin to drain, it is called a thyroglossal sinus.

Lingual Thyroid

Thyroid tissue may sometimes be left behind embedded in the tongue. This is usually asymptomatic. However, if the mass becomes large, symptoms of dysphagia (difficulty swallowing), dysphonia (difficulty speaking), or dyspnea (difficulty breathing) may arise. The majority of persons with *symptomatic* lingual thyroid tissue have no thyroid gland in the neck. The way to determine if a suspicious mass in the tongue is composed of thyroid tissue is to see if it takes up administered I^{123}.

Lateral Cervical (= Branchial) Cysts, Sinuses, and Fistulae

If some region of the cervical sinus fails to obliterate, a fluid-filled sac persists as a cyst in the side of the neck. This is called a **lateral cervical,** or **branchial, cyst**. If the sac opens onto the skin (probably because the opercular process did not completely fuse with the epipericardial ridge), the resulting condition is called a **lateral cervical,** or **external branchial, sinus**. If the sac opens into the pharyngeal cavity because one of the branchial septa ruptured (usually the second or third), the resulting condition is called an **internal branchial sinus**. If it was the second branchial septum that ruptured, the internal branchial sinus will open through the palatine tonsil. If it was the third branchial septum that ruptured, the sinus will open through the thyrohyoid membrane. If there is a continuous channel from the skin through a remnant of the cervical sinus and a ruptured branchial septum into the pharynx, the resulting condition is called a **branchial fistula**.

Failures of Proper Neural Crest Migration Into Branchial Arches

If the neural crest cells destined to populate the first branchial arch do not adequately do so, major deformations of the maxilla, mandible, and ears result. These are grouped together under the name of "first arch syndromes", or "mandibulofacial dysostoses". Improper migration of neural crest cells into the third and fourth branchial arches produces a condition known as DiGeorge's syndrome. It is characterized by minor deformations of the lower face combined with thymic and parathyroid aplasia (i.e., no thymus and no parathyroids) and problems with aorticopulmonary septation. The absence of a thymus has a very deleterious effect on the development of the immune system. The absence of parathyroids leads to hypocalcemia.

Aberrant Parathyroid Locations

Parathyroids may end their migrations too soon, thereby ending up in odd locations. It is also possible for parathyroids III to continue an association with the thymus beyond the normal time of their separation, with the consequence that the inferior parathyroids are located in the superior mediastinum.

Cleft Tongue

Rarely, the lateral tongue buds fail to fuse, leading to a tongue whose tip is split down the middle.