

# CHAPTER 36

## NECK: CYSTS, SINUSES, AND FISTULAS

James O. Adeniran  
Kokila Lakhoo

### Introduction

Lumps in the neck are a common problem in children. Some of the lumps may not be obvious at birth but slowly get bigger and become worrying to the parents. Most of the lumps are asymptomatic, but some can cause respiratory or swallowing difficulties.<sup>1,2</sup> Some lumps become infected and require urgent medical attention. Most sinuses are usually not noticed at birth, but as the child grows, there is persistent discharge from the ostia. Many of these lumps and sinuses are remnants of structures that form the face and neck. Some sinuses are due to chronic infections.

Lumps that appear around the necks of children may be due to various conditions, as listed in Table 36.1. This chapter focuses on cysts, sinuses, and fistulas of the neck, which are remnants of branchial apparatus, and remnants of the thyroid gland. Sinuses due to tuberculosis, human immunodeficiency virus (HIV), and fungi also are discussed. Lymphadenopathy is discussed in Chapter 37, sternomastoid tumours in Chapter 38, thyroid masses in Chapter 40, and lymphangiomas in Chapter 44.

### Branchial Arches, Clefts, and Pouches

#### Embryology and Pathology

Branchial arches appear as four pairs of ridges on the lateral side of the face of the 5-week old embryo (Figures 36.1 and 36.2). The arches bulge into the side walls of the foregut and meet each other in its floor, displacing the heart caudally to establish the neck region of the embryo.<sup>3</sup> The ridges are separated by four pairs of external, ectodermal grooves (branchial clefts) matched internally by four pharyngeal, endodermal pouches. The arches form the skeleton, musculature, and blood vessels of the jaws, palate, larynx, and pharynx, as well as the muscles of the face. As the dorsal ends of each arch approach the hindbrain, these structures are invaded by nerve fibres from the branchial efferent column. The ventral ends of the arches also converge on the pericardium to connect capillaries from the truncus arteriosus.<sup>3</sup> Each arch has mesenchyme, which develops into bone, cartilage, blood vessels, and muscles innervated by the nerve of that arch.

#### First Branchial Arch

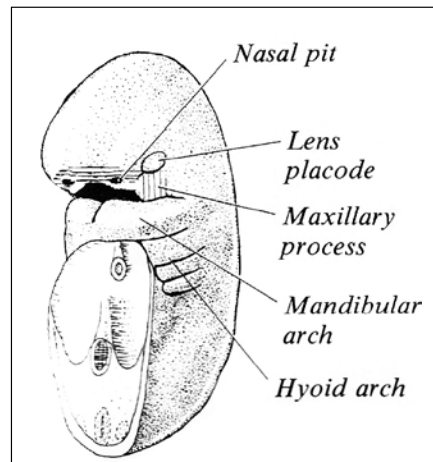
##### Embryology

The first, or mandibular, arch appears on the 22nd gestational day, and by the 6th week fuses in the midline to form the mesenchymal primordium that develops into the anterior two-thirds of the tongue. The core of the arch chondrifies to form Meckel's cartilage, which develops into the malleus and incus bones. The muscles of mastication develop from the first arch mesoderm, all innervated by the motor root of the trigeminal. The first branchial cleft forms the external acoustic meatus.

The first pharyngeal pouch is recognised after the formation of the head fold about the 20th day of embryonic life. The first pair of grooves and pouches persists to form the auditory canal and eustachian tube, which is separated by the tympanic membrane. The first pouch and membrane persist as the pharyngotympanic tube, middle ear cavity, and tympanic membrane.

Table 36.1: Common causes of lumps in the necks of children

Location	Cause
Lateral side of neck	Lymph nodes due to scalp or throat infections, tuberculosis, or lymphomas Cystic hygromas Sternomastoid tumour Teratomas Thyroid masses Branchial cyst—first, second, third, fourth
Midline of neck	Thyroglossal cysts Dermoids Haemangiomas Ectopic thyroid tissue



Source: FitzGerald MJT. Human Embryology: A Regional Approach. Harper & Row Publishers, 1978. Used by permission.

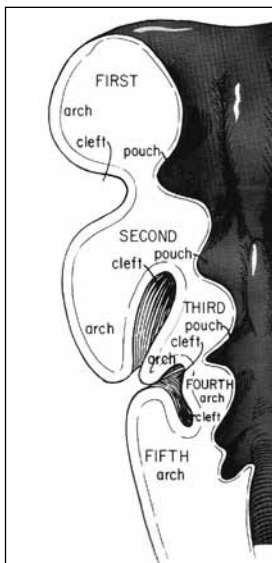
Figure 36.1: Five-week embryo showing the position of branchial arches.

Differentiation of the first branchial arch is shown below:

- **Skin:** skin of lower part of face
- **Bones:** malleus, incus, mandible, maxilla
- **Muscles:** muscles of mastication, floor of the mouth, tensor palati, tensor tympani, anterior belly digastric and mylohyoid
- **Nerve:** mandibular branch of trigeminal nerve
- **Artery:** maxillary
- **Membrane:** mucous membrane of nasopharynx

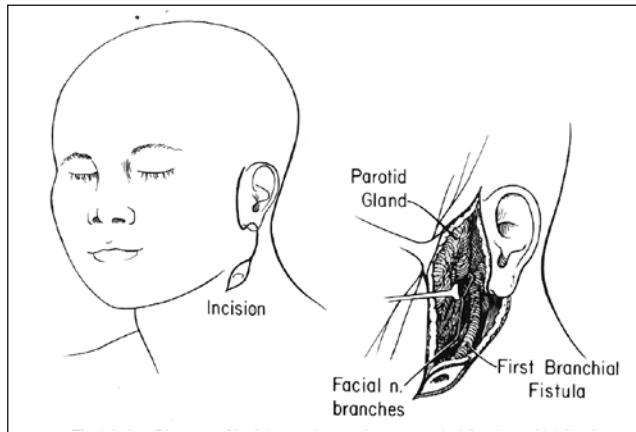
#### Remnants

Abnormal development of the first branchial arch results in cleft lip and palate, pinna deformities, and malformed malleus and incus, which may produce congenital deafness.<sup>3-5</sup>



Source: Welch KJ, Randolph JG, Ravitch MM, O'Neill JA Jr, Rowe MI. *Pediatric Surgery*. Year Book Medical Publishers, 1986. Used by permission.

Figure 36.2: Coronal view of 5-week embryo showing branchial arches.



Source: Welch KJ, Randolph JG, Ravitch MM, O'Neill JA Jr, Rowe MI. *Pediatric Surgery*. Year Book Medical Publishers, 1986. Used by permission.

Figure 36.3: Surgical management of first branchial arch fistula.

### Clinical presentation

True anomalies of the first branchial cleft are uncommon.<sup>5</sup> Skin tabs, pre-auricular cysts, and sinus tracts around the ear are not of branchial cleft origin but occur from abnormal infolding and entrapment of epithelium during the merger of the six hillocks of His that form the external pinna.<sup>4,6</sup>

When they occur, first branchial cleft cysts present as a swelling in front of or behind the pinna. External openings of the fistula lie below the mandible and above the hyoid bone. The tract may pass superficial or deep to the main branches of the facial nerve and through the substance of the parotid gland with the internal opening into the external auditory canal, which forms the source of recurrent infection (Figure 36.3).

### Management

If operation is deemed necessary, a curved incision is made to elevate the pinna and expose the parotid gland. The facial nerve and its trunks are identified and preserved. The tract is then dissected superiorly and then medially to the external auditory canal (see Figure 36.3).

Histologically, the tracts are lined by stratified squamous epithelium with skin appendages. Muscle fibres and cartilages may be seen in the deeper layers.

## Second Branchial Arch

### Embryology

The first two branchial (or hyoid) arches and clefts, and second pharyngeal pouches appear in the 22-day old embryo. The second arch extends down the neck (as the platysma) to overlap the second, third, and fourth branchial clefts, forming a potential space—the cervical sinus of His. The core of the hyoid arches forms a U-shaped cartilage, which forms the upper part of the hyoid bone. The dorsal end forms the stapes and the styloid process of the temporal bone.

Differentiation of the second branchial arch is shown below:

- **Skin:** lateral and anterior part of neck
- **Bones:** stapes, styloid process, upper part of body and lesser cornu of hyoid
- **Muscles:** posterior belly of digastric, muscles of facial expression, stapedius, stylohyoid, platysma
- **Nerve:** facial nerve
- **Artery:** stapedia (remnant of dorsal part of second aortic arch)
- **Membrane:** membrane of oropharynx

The muscular element of the hyoid arch spreads like a fan to form the muscles of facial expression, innervated by the facial nerve (Figure 36.4). Lymphocytes invade the lateral end of the second pharyngeal pouch to form the palatine tonsils

### Remnants

Complete fistulas, external sinuses, and cysts may occur as remnants of the second branchial arch. Although congenital, the tiny openings may not be obvious at birth. Attention is usually drawn to the problem by persistent mucoid drainage and/or recurrent infection. Sinuses present in the first decade of life, and cysts usually in the second decade. The cysts and external openings of the second branchial cleft lie along the anterior border of the sternomastoid muscle at the junction of the upper two-thirds and the lower one-third (Figures 36.4 and 36.5). The tract ascends along the carotid sheath to the level of the hyoid bone, then turns medially between the branches of the carotid artery, behind the posterior belly of the digastric and stylohyoid muscles, and in front of the hypoglossal nerve to end in the tonsilla fossa (see Figure 36.1). Sinuses with external openings at the same site pursue the same course before terminating blindly after variable distances. Secretions in branchial cysts may take a while to accumulate because they are clinically visible or palpable. The cysts may not, therefore, be clinically evident until late in childhood or early adolescence. Bacteria from the oral cavity may contaminate the cysts, leading to an abscess in 25% of cases.<sup>7</sup> The cysts may be bilateral in 10% of the cases.

The cysts contain turbid fluid and therefore do not transilluminate like cystic hygromas. Lymph node enlargement from tuberculosis, tonsillitis, and lymphomas may present with neck masses to form differential diagnosis.

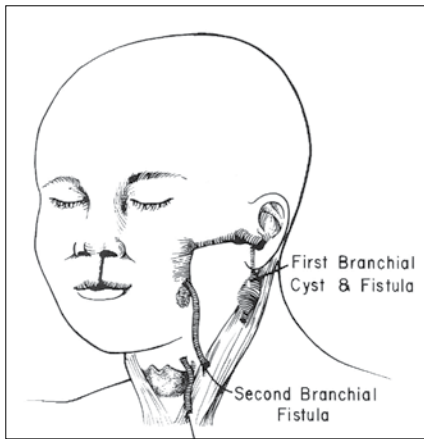
### Investigation

A sinogram may be done to outline the course of the fistula, but it is not a substitute for careful dissection at operation. Fine needle aspiration (FNA) cytology or excision may be needed to confirm the diagnosis, and this must be done aseptically to prevent introducing infection.

### Management

Sclerotherapy has not been developed as a method of management; therefore, operative excision is the treatment of choice. Unless the whole tract is dissected, recurrences may occur, with the possibility of neoplastic degeneration in adult life.<sup>8,9</sup>

Operations to remove the cyst, sinus, and fistula are all approached the same way, with the patient under general anaesthesia with endotracheal intubation.



Source: Welch KJ, Randolph JG, Ravitch MM, O'Neill JA Jr, Rowe MI. *Pediatric Surgery*. Year Book Medical Publishers, 1986. Used by permission.

Figure 36.4: Courses of first, second, and third branchial fistulas.



Figure 36.5: Second branchial cyst in a 4-year-old girl (the cyst had discharged and healed 4 or 5 times).

For the procedures outlined below, a sandbag is placed under the shoulders to extend the neck, and a head-ring is applied to steady the head, similar to the standard draping for a thyroid operation. The head is then turned to the contralateral side as convenient.

#### Branchial cysts

1. In branchial cysts, a transverse incision is made 3–4 cm long over the mass.
2. The skin and platysma are incised and deepened to the edge of the mass.
3. Blunt and sharp dissections are done to identify the tract. It is important to maintain a bloodless field, using mostly diathermy, throughout the dissection and to keep close to the tract. Keeping close to the tract will not only make the tract easily visible, but will prevent inadvertently injuring surrounding structures. It is not necessary to identify all the surrounding structures. If the dissection is very close to the tract, the likelihood of injuring other structures is slim.

#### Sinus

Preparation and draping for surgery is as above.

1. An elliptical incision is made around the opening.
2. The incision is deepened through the skin and platysma and the tract is identified.
3. The operation then proceeds the same way as for removal of a branchial cyst.

#### Fistula

In many cysts and sinuses, the tracts end blindly at various distances but must be pursued as a fistula. Again, preparation and draping for surgery is as above.

1. The fistula is traced along the carotid sheath through the bifurcation of the carotid artery, then medially to the tonsilla fossa.
2. The anaesthetist may assist by inserting a gloved finger into the patient's mouth to push the tongue down.
3. Another skin incision may be necessary, as a step-ladder incision, if the original skin incision is too far down in the neck.

Patients with bilateral fistulas can have both sides operated at the same sitting.

Histologically, the cysts are lined by squamous epithelium, surrounded by muscle fibres and lymphoid tissue, but in 10% of patients, respiratory columnar epithelium may also be present.<sup>4,10</sup>

### Third Branchial Arch

#### Embryology

The three pharyngeal arches and four pharyngeal pouches develop by the 27th day of embryonic life. The pouches are tube-like extensions of the pharynx.<sup>11</sup>

The third arch mesenchyme forms the posterior one-third of the tongue. Its cartilage ossifies to form the lower part of the hyoid bone. Its only muscle, the stylopharyngeus, is supplied by the glossopharyngeal nerve from the nucleus ambiguus.<sup>3</sup> The thymus and inferior parathyroid glands develop from the third pharyngeal pouch.

Differentiation of the third branchial arch is shown below:

- *Skin*: lateral part of neck
- *Bone*: lower part of body and greater horn of hyoid bone
- *Muscle*: superior pharyngeal constrictor
- *Nerve*: glossopharyngeal
- *Artery*: common carotid
- *Membrane*: lower part of pharynx

#### Remnants

Rarely, some cysts may arise from the left side of the neck in close relation to the thyroid gland. The external openings are usually at the anterior border of the clavicular head of the sternomastoid. The tract runs behind the internal carotid artery, the vagus, and the hypoglossal and superior laryngeal nerves, and then turns medially above the spinal accessory nerve and penetrates the thyroid membrane to end in the pyriform sinus.

#### Management

Incision and drainage may be necessary when the cysts are infected. The tract may need excision if there is recurrent infection.

Histologically, thyroid, thymic, and lymphoid tissue and Hassall corpuscles have been seen, which may suggest their origin from lower pharyngeal pouches.<sup>4</sup>

### Fourth to Sixth Branchial Arches

The fourth and sixth arches mingle as they produce the cartilages and ligaments of the larynx, the levator palate, and the intrinsic muscles of the larynx and pharynx, all supplied by the vagus nerve.<sup>3</sup> Part of thymus and the superior parathyroid glands develop from the fourth pharyngeal pouch.

Contributions of the fourth branchial arch are shown below:

- *Skin*: none
- *Cartilage*: thyroid and arytenoids
- *Muscles*: inferior pharyngeal constrictor, cricothyroid, intrinsic laryngeal
- *Nerve*: superior laryngeal branch of vagus
- *Artery*: arch of aorta on left side, first part of subclavian artery on right side
- *Membrane*: hypopharynx

Contributions of the sixth branchial arch are shown below:

- *Skin*: none
- *Cartilage*: cricoid, arytenoids, rings of trachea and bronchi
- *Muscles*: intrinsic muscles of larynx (except cricothyroid, stylopharyngeus, tensor palate)
- *Nerve*: recurrent laryngeal branch of vagus
- *Artery*: pulmonary artery on right side, ductus arteriosus on left side
- *Membrane*: hypopharynx

### Remnants

Anomalies of the fourth branchial pouch very rarely may produce a cyst or a fistula very low in the neck behind the sternomastoid muscle. On the right side, the tract goes behind the subclavian artery; on the left side, it goes under the arch of the aorta. It opens into the cervical oesophagus.

### Midline Cervical Clefts

Midline cervical clefts (Figure 36.6) are due to imperfect midline fusion of the paired branchial arch tissue about the fourth week of embryonic development. These present as raw, weeping areas in the midline of the lower neck. They may have irregular skin tabs or shallow sinuses. Management is usually conservative. If excision is deemed necessary, a Z-plasty may be done to prevent ugly scars or contractures.

### Dermoid Cysts

Dermoid (inclusion) cysts (Figure 36.7) are caused by entrapment of epithelium of branchial arch origin at the time of embryologic midline fusion. Most of the cysts are in the midline, are firm in consistency, and may be attached to overlying skin (Figure 36.7). They do not move with swallowing or protrusion of the tongue. They usually do not have any deep-seated tracts and are easily excised surgically through a transverse collar-stud incision.

### Teratomas

Teratomas are tumours composed of multiple tissues foreign to the anatomical locus.<sup>1,2,12</sup> These tissues cannot have resulted from metaplasia. They develop adjacent to normal anatomical structures or organs and are generally attached by limited vascular pedicles. The most common sites for teratomas are the gonads and the sacrococcygeal areas.<sup>2</sup> These sites embryologically allow deviation of early germinal issue to disorganised complex teratomas. Teratomas are also common in the neck region (Figure 36.8). Most teratomas are benign. They produce secondary symptoms due to their pressure effects on adjacent organs.

### Investigation

Teratomas may be soft to firm in consistency because they may contain cystic areas. They may be confused with cystic hygromas. Neck ultrasound and/or aspiration, under aseptic conditions, may be necessary to confirm the diagnosis.

### Management

Complete excision of the teratoma should be the desired goal. Incomplete excision may lead to recurrence of the mass, recurrent infection, draining sinuses, or the possibility of malignant change.<sup>1,13</sup>

### Thyroglossal Cysts

Thyroglossal cysts are the most common midline masses in children, accounting for 70% of all congenital cervical lesions.<sup>14</sup> They can occur at any age, and one-third become obvious in adult life.

### Embryology

The thyroid gland develops as a diverticulum from the foramen caecum of the tongue, descending in front of the trachea in company with the thymus and the inferior parathyroid glands. It reaches its final position by the 7th week of embryonic life.<sup>14-16</sup> The hyoid bone develops at about the same time, from the second and third arches, and fuses anteriorly so that the thyroglossal duct may pass anterior to, through, or posterior to

the body of the hyoid bone.<sup>3,14</sup> The duct usually spontaneously obliterates, but remnants may be found anywhere from the base of the tongue to the pyramidal lobe of the thyroid gland (Figure 36.9), although 80% are juxtaposed to the hyoid bone.<sup>15</sup>

### Clinical Presentation

Most thyroglossal cysts are clinically evident before the child is 10 years old. Males and females are affected equally. The cysts present as round masses in the midline of the neck that move up and down with swallowing and with protrusion of the tongue because of their connection to the hyoid bone. They are soft to firm in consistency, mobile, and nontender unless infected.



Figure 36.6: Midline cervical cleft.



Figure 36.7: Dermoid cyst.



Figure 36.8: Teratoma.



Figure 36.9: Thyroglossal cyst.



### Differential Diagnosis

Midline dermoids, which occur commonly in the submental triangle, are the commonest differential diagnosis.<sup>16</sup> Ectopic midline thyroid, enlarged pyramidal lobe of thyroid gland, and thyroid adenomas may also present as midline masses.

### Investigations

Neck ultrasonography and thyroid iodine scintigraphy may be needed to confirm the diagnosis and rule out ectopic thyroid tissue.

### Complications

Thyroglossal fistula may result from infection of the thyroglossal cyst or after drainage of a thyroglossal abscess. The fistula is usually in the midline. Sinograms are unnecessary for diagnosis, and sclerotherapy as a means of treatment has not been described.

### Surgical Management

Both thyroglossal cysts and fistulas are surgically approached by the Sistrunk operation described by Walter Sistrunk in 1920.<sup>17</sup> The aim of the operation is to completely excise the duct with the middle part of the body of the hyoid bone and a cuff of the tongue muscles because side branches of the duct may occur within the muscles of the tongue.<sup>16</sup> Complete excision of the duct is essential to prevent recurrence or malignant degeneration.<sup>18</sup>

For the procedures outlined below, the patient is given general anaesthesia with endotracheal intubation. In the supine position, a sandbag is placed under the patient's shoulders to extend the neck, and a head ring is used to steady the head. The neck, lower jaw, and upper part of the chest are cleaned and draped.

#### Thyroglossal cyst

1. A transverse skin incision is made at the midpoint of the mass. The incision is deepened through platysma and subcutaneous tissues to reach the edge of the cyst. With strict haemostasis using diathermy, the tract is easily identified on the upper part of the cyst.
2. Thereafter the procedure follows as for a thyroglossal fistula.

#### Thyroglossal fistula

1. An elliptical skin incision is made around the ostium. This skin incision is deepened through platysma until the tract is identified.
2. Keeping very close to the tract, the tract is dissected between the sternohyoid muscles until the hyoid bone is identified.
3. The body of the hyoid bone is cleared from the sternohyoid muscle inferiorly and the mylohyoid and geniohyoid muscles superiorly with diathermy.
4. An artery forceps is used to separate the body of the hyoid bone from the thyrohyoid membrane inferiorly.
5. The middle portion of the hyoid bone is then excised either with a strong straight scissors or with small bone cutters.
6. The hyoid bone is then held with towel clips and lifted to expose the proximal part of the tract. This is now dissected to the floor of the mouth. The anaesthetist may be asked to depress the tongue with a gloved finger to assist the surgeon to locate the foramen caecum but this manoeuvre is usually not necessary.<sup>16</sup>
7. At the foramen caecum, a small rectangular piece of hyoglossus and genioglossus are excised with the tract.

8. Strict haemostasis is maintained and the wound closed in layers. It is unnecessary to reapproximate the hyoid bone. Drainage is usually unnecessary unless haemostasis is unsatisfactory or there is previous infection. A subcuticular stitch may be used to close the skin.

9. Perioperative antibiotics are justified if previous infection of a cyst has occurred or after operation on thyroglossal fistula.

Histologically, the tract usually contains pseudostratified ciliated columnar respiratory epithelium in 60% of cases and stratified squamous epithelium in the rest.<sup>19</sup> More than 100 carcinomas have been reported from thyroglossal duct remnants. These are either papillary adenocarcinomas or squamous carcinomas.<sup>14,20</sup> Most of these malignancies arise in the substance of the ducts and not from metastasis from the thyroid glands. The tumours are slow-growing and confined to the neck for long periods.<sup>14</sup> Most, including the affected lymph nodes, can be managed surgically with the Sistrunk operation.

### Other Rare Neck Masses

Many rare neck masses appear low in the neck and may have connections to masses in the thoracic cavity.

#### Bronchogenic Cysts

Bronchogenic cysts are attached to the hilum of the lung and may present as low neck masses, where they may compress the trachea, causing stridor.

#### Lymphangiomas

Lymphangiomas involving the posterior mediastinum may extend to the lower part of the neck, generally to the left of the trachea.

#### Thymic Cysts and Mediastinal Tumours

Thymic cysts and mediastinal tumours may rarely present with extensions in the lower part of the neck.

### Evidence-Based Research

Table 36.2 presents a retrospective study that reviews the types of congenital cysts, their management, and problems associated with management.

Table 36.2: Evidence-based research.

<b>Title</b>	Congenital cysts and fistulas of the neck
<b>Authors</b>	Nicollas R, Guelfucci B, Roman S, Triglia JM
<b>Institution</b>	Service d'ORL Pédiatrique, Fédération ORL Hospital de la Timone, Marseille, France
<b>Reference</b>	Intl J Pediatr Otorhinolaryng 2000; 55:117–124
<b>Design</b>	Retrospective study
<b>Aim</b>	To review types of congenital cysts seen, the management given, and problems associated with management.
<b>Period</b>	1984–1999.
<b>Exclusion</b>	Preauricular cysts and cystic hygromas.
<b>Result</b>	Of 191 children with congenital cysts and fistulas, 123 were malformations of the midline, 102 were thyroglossal duct cysts, and 21 were dermoid cysts. Of the 68 malformations of the laterocervical region, 37 were cysts and fistulas of second cleft, 20 were cysts of first cleft, 7 were cysts of fourth pouch, and 4 were thymic cysts.
<b>Problems in management</b>	Diagnosis and management of midline masses are usually straightforward. Misdiagnosis of lateral cysts is common and often leads to inadequate treatment and recurrence.

## Key Summary Points

1. Branchial arches develop as six-paired structures on the side of the neck of the embryo. Many congenital neck lumps and fistulas are remnants of branchial arches.
2. The first arch forms the skin of the lower part of face, malleus, incus, mandible, maxilla, and muscles of mastication. Remnants of the first branchial arch are uncommon.
3. The second arch forms skin of the lateral and anterior part of neck; the stapes, body, and lesser cornu of the hyoid bone; and muscles of facial expression. Cysts (and fistulas) of the second arch appear at the anterior border of the sternomastoid muscle.
4. The third arch and pouch develop into the posterior one-third of the tongue, the lower part of the hyoid bone, the thymus, and inferior parathyroid glands. External openings of remnants of the third arch are usually at the anterior border of the clavicular head of the sternomastoid.
5. The fourth to the sixth arches mingle as they produce the cartilages and ligaments of the larynx and pharynx (all supplied by the vagus nerve), part of the thymus, and the superior parathyroid glands.
6. The thyroid gland develops from a diverticulum at the floor of the mouth. Remnants of the thyroid may be found from the base of the tongue to the pyramidal lobe of the thyroid
7. Thyroglossal cysts are the most common congenital midline cysts. Thyroglossal cysts move with swallowing and protrusion of the tongue.
8. Thyroglossal fistulas are best removed by the Sistrunk operation.
9. Extrapulmonary tuberculosis, fungi, and HIV can affect neck glands to produce discharging sinuses in the neck.

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