

Pediatric Facial Plastic and Reconstructive Surgery

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Chapter 4: Congenital Neck Masses

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This chapter deals with congenital neck masses. By definition, *congenital* means present at birth, and a mass lesion is one that represents an abnormal regional increase in volume. By definition, therefore, congenital neck masses would be exceedingly rare. If, however, we consider congenital to encompass what is present at birth becoming obvious later, then the incidence of congenital neck masses increases dramatically. A number of congenital cervical anomalies will not present as masses, eg, anomalies of the skin.

Congenital masses will be described by tissue types, and anomalies of the branchial arch apparatus will be described collectively. Vascular and lymphatic masses will be discussed in another section of the book and will not be described here; neither will the more frequent benign and malignant true neoplasms.

Embryology of the Neck

Only the briefest outline of the embryology of the neck is possible in this format. The neck develops from the derivatives of the branchial arches. In the human only five arches are represented, I, II, III, IV, and VI - arch V is not detectable in the human. The branchial arch apparatus is first identifiable in the 3.0-mm embryo as tiny, smooth, rounded projections immediately anterior to the notochord in the area between the cephalon and the chest. At this point the embryo consists of two masses, a head and a chest/abdomen. There is no neck.

Each branchial arch consists of a central core of mesoderm that differentiates into a skeletal bar, an artery, nerve, and muscles. The external surfaces are covered by ectoderm and internally they are lined by endoderm. The arches expand ventrally so that by the 5-mm stage they are recognizable as raised cylindrical structures. Between adjacent arches externally are grooves or clefts, and internally similar depressions are called pouches.

Each arch is the precursor of definitive structures as is each cleft and each pouch. The more important derivatives of the arches in the human are listed in Table 1.

Continued growth in a ventral direction brings right and left branchial arches together in the midline where they fuse to form the neck. The lateral surface of the neck is smoothed by a projection from the ectodermal surface of the second arch called the operculum that grows caudally to fuse with a smaller projection from the third arch. There is, therefore, a potential space between the operculum and the lateral surface of the third arch, the cervical sinus of His. Theoretically, a second branchial cleft cyst develops if remnants of the cervical sinus persists. The potential anomalous development of the branchial arch apparatus are listed in Table 2. Only the more common and a few rare anomalies are noted.

The final position of the third pouch has not been definitely decided. Internal sinuses

developing from the lateral pharyngeal wall including the pyriform sinus might be from third or fourth pouches.

Table 1. *Derivatives of the branchial arch apparatus*

Cleft (ectoderm)		Derivative		
1st		Ext aud canal		
2nd		Cervical sinus		
Pouch (endoderm)				
1st		Eustachian tube/middle ear/mastoid cells		
2nd		Tonsil		
3rd		Inf parathyroid, thymus		
4th		Sup parathyroid, ultimobranchial body		
Arch (skeleton)				
1st		Malleus, incus, mandible		
2nd		Stapes, styloid process, stylohyoid lig; sup half body of hyoid bone		
3rd		Inf half body of hyoid bone; gt cornua		
4th and 6th		Laryngeal cartilages		
	Muscle		Nerve	Ligament
1st arch	Temporalis, masseter, pterygoideus, mylohyoid, t tympani, t palatini		VIth Vth	Sphenomandibular
2nd arch	Muscles of face, stapedius, stylohyoid, postdigastric		VIIth	Stylohyoid
3rd arch	Stylopharyngeus, constrictors		XIth IXth	
4th/6th	Laryngeal constrictors		Sup and rec laryngeal	

Table 2. *Developmental anomalies of the branchial arch apparatus*

1st		
Arch	Aplasia/dysplasia malleus, incus, mandible	
Groove	Anomalies of external auditory canal	
Pouch	Rare - diverticulum	
2nd		
Arch	Anomalies stapes and styloid process	
Groove	Branchial cleft cyst	
Pouch	Sinus in tonsil, fistula pharynx to neck	
3rd		
Arch		
Pouch	Cervical thymus, thymic cyst, aplasia parathyroid	
4th		
Arch	Congenital laryngeal stenosis or cleft	
Pouch	Sinus from pyriform sinus, aplasia parathyroid gland.	

Anomalies of the Skin

Congenital masses that arise from the ectoderm are exceedingly rare. There are, however, a number of skin-related problems that are of interest to the surgeon.

Pterygium Colli (Winged Neck)

A pterygium is a fold of skin with a core of connective tissue extending from the mastoid process to the point of the shoulder. Pterygia are found in Turner's syndrome, Noonan's syndrome, the multiple pterygium syndrome, and the leopard syndrome.

Turner's Syndrome

Turner's syndrome is caused by the absence of an X chromosome in a female. It is characterized by a short stature, webbed neck, widely spaced nipples, sexual infantilism, short metacarpals, cubitus valgus, coarctation of the aorta, and midfrequencies sensorineural hearing loss.

Noonan's Syndrome

Noonan's syndrome is characterized by short stature, webbed neck, hypertelorism, and cardiac anomalies. With development the face becomes almost triangular. Noonan's is probably a dominant syndrome. Females are usually fertile, whereas male fertility is variable.

Multiple Pterygium Syndrome

This syndrome consists of pterygia of the neck, antecubital and popliteal areas, fingers, and axillae, as well as a short stature and cleft palate. The multiple pterygium syndrome is probably recessive inherited with a background of consanguinity.

Leopard Syndrome

This is a mnemonic (1) for a syndrome that consists of multiple lentigines, electrocardiographic abnormalities, ocular hypertelorism, pulmonic stenosis, abnormal genitalia, retardation of growth, and sensorineural deafness. Mild pterygium colli are frequently present.

If cosmetically desirable, pterygia are treated by excision and Z-plasties.

Midline Cervical Clefts

These are rare anomalies in which there is an absence of normal tissue in the midline of the neck. Instead, there is a band of fibrous tissue extending from the mandibular symphysis to the sternum. Clefts present in the neonate as a linear raised area covered by thin, pink skin. The cleft tissue grows more slowly than the surrounding normal tissue, and untreated, it tethers the mandible inferiorly. A computed tomography (CT) scan or magnetic resonance imaging (MRI) will identify the depth of the lesion and also will ensure the presence of normal structures such as the thyroid gland. Midline cervical clefts are treated by excision and repair with either Z-plasties or a broken-line closure.

Anomalies of the Musculoskeletal System

Anomalies of the skeleton of the neck occur not infrequently but usually do not present as a mass.

The vertebral column or the clavicle might be affected. Only two skeletal anomalies will be discussed here.

Anomalies of the Vertebral Column

Klippel-Feil Syndrome

This is a condition in which the neck is abnormally shortened because of developmental anomalies of the vertebrae. There might be hemivertebrae or fused or absent vertebral bodies. The cervical vertebral column is frequently represented by a shortened almost solid block of bone with narrow or absent intravertebral discs or fused articular facets. The result is a short or almost absent neck. Rotation of the neck and head is grossly limited or absent, but flexion and extension at the atlanto-occipital joint is normal. The posterior hairline might be at the interscapular level and the chin might almost be resting on the sternum. One-third of these cases have associated hearing problems. The shortness of the vertebral column causes prominence of the trapezius muscles and the clinical impression of a winged neck that should not be confused with a pterygium.

As the vertebral anomalies in this syndrome are variable, difficulty of intubation for general anesthesia varies accordingly. In mild cases intubation might not present a problem in childhood, but in more severe cases, blind nasal intubation or intubation over a fiberoptic endoscope might be necessary.

Anomalies of the Clavicle

The clavicle develops from at least two segments - medial and lateral - that fuse. If fusion fails, a pseudojoint in midclavicle might present as a mass-like lesion. A simple radiologic study identifies the problem. No specific treatment is indicated, but if any type of surgical intervention is planned, then the underlying vascular structures, such as the subclavian artery and vein, must be protected.

Cleidocranial Dysostosis

In this syndrome there is aplasia or dysplasia of the clavicle occasionally with absence of a part or whole of one clavicle. There are also anomalies of the skull and frequently mental retardation and sensorineural hearing loss. The characteristic clinical finding is the ability to bring the shoulders into close apposition in the midline.

Torticollis (Wryneck)

Torticollis is defined as a tethering of the neck to one side with concomitant rotation of the chin to the contralateral side. The usual cause is contraction of the sternocleidomastoid muscle. Other causes might be maldevelopment or inflammatory disease of the cervical spine.

In the so-called congenital torticollis, the sternocleidomastoid muscle frequently develops a firm tumor-like swelling in its lower third. This represents a mass of fibrous tissue and is lily white and gritty in consistency - the white tumor of the newborn. The cause of these tumors is speculative ranging from in vivo venous occlusion to a hamartoma to even the ever-present probability of birth trauma. This condition can be treated by vigorous stretching exercises four or more times daily. If, however, the muscle continues to contract, then surgical section is advisable through a small horizontal incision in the lower neck. This should preferably be done before the first birthday. Muscle lengthening techniques give no better results. Persistence of torticollis results in asymmetry of the face with hypoplasia of the "down side."

The Branchial Arch Apparatus

Anomalies of the First Branchial Cleft

Anomalies of the first branchial cleft are those of the external auditory canal and are classified as aplasia, atresia, stenosis, and duplication anomalies. Here, we are concerned only with duplication anomalies. Work subdivided duplication anomalies into two types:

Type 1 - A skin-lined tract parallels the existing external auditory canal from a blind pouch laterally to the region of the middle ear medially.

Type 2 - Anomalies also consist of a squamous epithelial-lined tract ending in a blind pouch laterally or inferiorly that opens directly into the external auditory canal. Work considered these to be a composite containing ectodermal and mesodermal derivatives. In both cases, the tract becomes filled with keratin and might present because of infection, or as a mass in the upper neck or anywhere in the periauricular area. Clinically, they are diagnosed as recurrent abscesses usually treated by incision and drainage. Alternatively, because of the keratin material, they are frequently called extratemporal bone cholesteatoma.

Duplication anomalies usually require surgical excision. Because both types of anomalies have varying relationships to the facial nerve, the first step in surgery should be identification of the facial nerve in the usual manner and then careful excision of the tract. We recommend a standard parotidectomy incision that includes an ellipse of skin around any external fistula. After the facial nerve is identified, removal of the tract begins from lateral to medial. The tract is dissected through the branches of the facial nerve as necessary, and followed to its deepest point.

Anomalies of the Second Branchial Arch Apparatus

Anomalies of the Second Branchial Groove/Cleft

Persistence of the second branchial groove might present as a sinus, cyst, or fistula.

Second Branchial Sinuses

A sinus is a blind tract open on an epithelial surface. Second branchial cleft sinuses open on the skin, are usually sited along the anterior border of the sternocleidomastoid muscle at the level of the hyoid bone or thyrohyoid membrane and are frequently bilateral. They may drain small quantities of clear fluid probably produced by heterotopic salivary glands, which are sometimes seen in the wall of the sinuses. Tracts at this level are usually about 2 cm long, penetrate the platysma, and end blindly. They are easily excised.

Second cleft sinuses should not be confused with the rare first arch fistula that might open in the same position and track upward and posteriorly toward the external auditory canal.

Second Branchial Cleft Cysts

Cysts of the second branchial cleft might be present at birth, in early childhood, in late teenage or young adulthood, or in the fourth to fifth decade of life. The trigger is usually inflammation or frank infection. The typical position of a second branchial cleft cyst is deep to the sternocleidomastoid muscle at about the level of the hyoid bone, frequently protruding anteriorly beyond the anterior border of the muscle. Their sizes vary considerably, and in the adult might expand rapidly to be many centimeters in diameter. The popular embryological explanation of a branchial cleft cyst is a persistence of the epithelial remnants of the cervical sinus of His. Although this is a simple concept that provides a reasonable explanation, it does not quite fit the embryological pattern as Fraser, in a careful study, showed that the sinus is obliterated from its depths. Although second cleft cysts can be easily diagnosed clinically, it is wiser to confirm the diagnosis by ultrasonography and/or CT scans. As these cysts might be associated with sinus or fistulous tracts that might ascent to open into the region of the tonsil fossa or might descend to the lower neck, these patients should have their mouths and pharynges thoroughly examined.

Branchial cleft cysts are treated by surgical excision via a horizontal incision. Uninfected cysts are easily separated from the surrounding tissues. As the cysts lie lateral to the carotid sheath, they should be carefully dissected, preferably by blunt dissection.

Occasionally there are tracts leading from the deep surface. One tract might run superiorly between the internal and external carotid arteries and might even penetrate the middle constrictor muscle to open internally into the oropharynx close to or through the tonsil fossa. The lower tract might extend inferiorly toward the lower anterior border of the sternocleidomastoid muscle. The surgeon, therefore, must be prepared to follow and excise these tracts. Incomplete removal will result in the potential for recurrent infection or draining sinuses.

Acutely infected cysts are much more difficult to excise because their walls are thicker, surgical landmarks are obscured, and the characteristic lymphoid nodules in the cyst walls might be substantially enlarged. It is better, therefore, if possible, to cool an infected cyst with vigorous antibiotic treatment before attempting excision.

Second Branchial Cleft Fistulae

A fistula of the second branchial groove and cleft consists of an epithelial-lined tract with an internal opening in or adjacent to the pharyngeal tonsil and an external opening along the lower third of the anterior border of the sternocleidomastoid muscle. The tract runs between the external and internal carotid arteries penetrating the middle constrictor muscle superiorly and the platysma inferiorly, and can be outlined by contrast radiography. Branchial fistulae are excised electively if they tend to have recurrent infection or for cosmesis. Usually two ladder-pattern horizontal incisions are necessary, one at the level of the external opening and the second at the level of the hyoid bone. The tract must be traced to the constrictor muscle, and ligated and severed at this level.

Persistent Branchial Fistulae

Incomplete excision of branchial pouch or cleft anomalies usually results in recurrent infections in the neck. Investigation of these cases is not easy. A barium swallow, computed tomography, ultrasonography, and, if there is an external opening, contrast radiography, might all be of value. Frequently, however, the neck needs to be explored surgically using the details of previous surgery as guidelines.

Derivatives of the Third Branchial Pouch

The corresponding final place of the third branchial pouch has not been precisely determined. A possibility might be at the level of the vallecula or thyrohyoid membrane. Similarly, there is some controversy about the position of the fourth pouch, which traditionally has been supposed to be the pyriform sinus. It would seem wise, therefore, to consider third and fourth pouch derivatives together, particularly as the parathyroid glands are derived from both pouches, and the thymus gland from the third pouch. The thymus gland and parathyroid glands are frequently closely related in the developed organism.

Persistence of the third/fourth endodermal epithelial pouches occurs rarely. They occur as a blind sinus tract off the pyriform sinus. The usual method of presentation is a spontaneous infection in the neck in the vicinity of the thyroid gland frequently diagnosed as thyroiditis. If an abscess forms, it is usually incised and drained, only to recur sometime in the future. We have seen cases with seven drainage procedures over a 10-year period. There might also be a persistent fistulous drainage through one of the neck incisions and very rarely a chronic phlegmon in the neck at the level of the thyroid gland. The correct diagnosis can be made by barium study that frequently, but not always, identifies a tract leading from the pyriform sinus. Occasionally, when the barium study is negative, the internal opening can be seen by direct laryngoscopy. If a third/fourth branchial pouch sinus is diagnosed, treatment is by excision via a horizontal collar-type incision, which gives access to the region of the thyroid gland. If possible, the tract should first be injected with methylene blue. The old scar and surrounding tissue is excised, with care being taken not to injure the recurrent laryngeal nerve. The tract should be followed to the pyriform sinus where it is ligated and amputated.

Thymus Gland

Arrest of the thymus glands in the neck is rare. They may present as cystic masses at birth or in adulthood. Less than 80 cases have been reported in the world literature. Graeber et al studied 46 patients with thymic cysts; of these, 7 were in the neck and 36 in the

mediastinum. The neck masses were in the midline, paramedian, or lateral planes. There are, therefore, no distinguishing features for cervical thymic cysts, but there is frequently an associated hypertrophy of the ipsilateral parathyroid gland and consequently a few of the reported cysts have concomitant disorders of calcium metabolism. It is imperative, therefore, that cyst neck masses should be investigated by ultrasonography, CT scans or MRI, and screening of calcium metabolism. The superior mediastinum must also be assessed.

Thyroglossal Duct Cysts

The thyroid gland begins as a diverticulum from the floor of the embryonic pharynx just caudal to the tuberculum impar at a point that will become the foramen cecum. The diverticulum descends to the front of the neck in close relationship to the developing hyoid bone. The thyroid gland is in position pretracheally by the 8th week. The connection between the foramen cecum of the tongue and the thyroid gland forms the thyroglossal duct. The duct usually disappears but might persist anywhere along its length. These persistent areas expand to become thyroglossal duct cysts.

Thyroglossal duct cysts are more frequent in children but might present in adults. The oldest that I have seen is in a 75-year-old man. The trigger for presentation is usually inflammation or frank infection. They are characteristically in the midline of the neck or just lateral to, or just inferior to, or at the level of, the hyoid bone. There is, however, considerable variation in the positions of thyroglossal duct cysts.

Thyroglossal duct cysts are treated by excision. There must be preliminary investigation by radioisotope study, CT scan, or ultrasound to ensure that the mass is not an ectopic thyroid gland. Excision is accomplished by the Sistrunk procedure through a horizontal incision at the level of the hyoid bone. The cyst usually separates easily from the surrounding tissues, and the central 2-cm block of the hyoid bone is cleared of muscle attachments and removed. A block of tissue about 0.5 cm in diameter is then cored from the central hyoid to the foramen cecum and excised at that level. Failure to excise a thyroglossal duct completely will result in a persistent sinus or recurrent infection.

Ectopic Thyroid Gland

During its descent from the foramen cecum, the thyroid gland might come to rest in any point between the foramen cecum and the usual pretracheal position. Ectopic thyroid tissue, therefore, might be present in the substance of the tongue, around the hyoid bone, almost at the level of the cricoid cartilage or in front of the third to fourth tracheal ring, as is usual in the adult. Ectopic thyroid tissue is solid in contrast to a thyroglossal duct cyst. It is usually attached to the tongue and moves upward when the tongue is protruded. Any solid mass in the midline or just off the midline of the anterior neck, where there is a suspicion of ectopic thyroid tissue, should have the following investigations performed. Ultrasonography determines its consistency, and might also show the presence or absence of a normal thyroid gland. In addition, a technetium scan will identify functioning thyroid tissue. It is important

to ensure that ectopic thyroid tissue is not the patient's only functioning thyroid. Ectopic thyroid tissue should be excised only for cosmetic reasons, although there is controversy about whether or not there is an increased incidence of malignant degeneration in these masses.

Teratomas

Teratomas are compound tumors composed of tissue from more than one germ layer. They arise from cells that maintain their embryonic totipotentiality to differentiate into any of the three primary germ layers. Therefore, teratomas might contain skin, muscle, bone, etc. Understandably, teratomas are more frequent in association with the gonads but can occur anywhere on the body. Touran et al reported a teratoma in the neck of a newborn with metastases. Through the time of their report (1989) less than 150 cases of cervical teratomas had been described. Generally, however, cervical teratomas have less tendency to malignancy than those arising elsewhere.

Teratomas, therefore, might present as a mass in the neck or pharynx or any combination thereof, in a neonate, but also might be diagnosed in adulthood. The diagnosis is sometimes aided by the radiologic finding of calcification in the mass. As there is a definite tendency to malignant degeneration, teratomas should be excised as soon as possible. Preliminary investigation with CT scans, MRIs, and even ultrasound might be of value to determine the extent of the lesion and the surgical approaches that might be indicated. After excision, these patients should be carefully followed.