Three Demonstrations ON CONGENITAL MALFORMATIONS OF PALATE, FACE, AND NECK.

GIVEN AT THE ROYAL COLLEGE OF SURGEONS, ENGLAND.

BY

PROFESSOR ARTHUR KEITH, M.D., F.R.C.S.ENG.,
CONSERVATOR OF THE MUSEUM.

III.—MALDEVELOPMENT OF THE EXTERNAL EAR.

In Figs. 21, 22, and 23 are represented the more common forms of accessory auricular appendages. Fig. 21 is from one of the five examples in the college collection. There is here a very distinct arrest in the closure of the first visceral cleft on the sides of which the external ear is formed. The tragus, anti-tragus, and meatus are pulled towards the chin. The condition was associated with double cleft of the palate and absence of the auditory nerve. In all the three specimens figured the tragus is subdivided, each half projecting as a separate process to a varying degree. A process derived from a subdivision of the tragus is the most common form of auricular appendage. An appeal to comparative anatomy has provided no explanation to the condition. Another very common form of appendix is that shown in Fig. 22, c. It projects from the skin immediately in front of the ascending crus of the helix; when dissected its base is found to be connected with the ascending crus of the helix and to represent the spina helicalis, which is usually buried in the subcutaneous tissues, but here projects on the surface. The pendulous process on the cheek (see Fig. 23) is also a form of auricular appendix. There are three specimens in this museum, and I have seen two others. The cheek

Fig. 21.—Ear of child with cleft palate. A, B, Appendages formed out of the tragus; C, meatus dragged downwards and forwards; D, E, positions of auricular fistulae. Fig. 22.—Ear of an anencephalic fetus, showing A, B, appendages developed from tragus; C, from spine of helix. Fig. 23.—A, B, C, Position of cheek appendages, along the position of a bucco-auricular cleft; D, E, appendages of the tragus; F, position of first visceral cleft (from a photograph in the possession of Dr. William Wright).

TABLE IV.

	A.	B.	C.	Total.
Absence, more or less complete, of external ear	2	2	2	6
Accessory appendages of ear	5	2	0	7
Auricular fistulae	—	3	0	3
Cervical appendages	1	—	2	3
Auricular dermoids	3	1	0	4
Auricular fistula with teeth	—	—	2	2
Malformation of tongue	2	1	1	4
Sublingual dermoids	4	1	0	5
Cysts of cervical sinus	12	6	—	18
Cysts or tumours connected with median thyroids	3	2	—	5
Cysts from lateral aberrant thyroids... ..	1	—	—	1
Congenital tumours of the neck	5	2	—	7

A, Specimens (human) in the Museum of the Royal College of Surgeons.

B, Specimens (human) in the museums of metropolitan medical schools.

C, Specimens from domestic animals, chiefly in the Museum of the Royal College of Surgeons.

appendages occur along the line marked in Fig. 23, a line represented by a fissure in cases such as those published by Mr. J. H. Morgan¹ and Dr. Edginton.² They are pendulous bodies, consisting of a fibrous core covered by skin. In two of our specimens we found a slight cartilaginous rod in the subcutaneous tissue at the point of attachment of the appendix, passing from the base towards the ear, showing that the cheek processes are really detached elements of the ear. As to the meaning of auricular appendages, I am altogether uncertain; all the specimens in museums show that they are associated with grave malformations, such as club-foot, cleft palate, atresia ani, spina bifida; the cause of the one condition is evidently the cause of the others. Auricular appendages, if they occur alone, are signs of an inherited tendency to grave malformation. The fissures which pass from the angle of the mouth to the ears, the processes which occur on the cheek, and the ventral extension of the tragus (Fig. 21) are probably caused, as Ballantyne has inferred, by the occurrence of amniotic adhesions.

Fistulae connected with the external ear, which were first described in this country by Sir James Paget,³ are very imperfectly represented in our museum. There is evidently a relationship between auricular fistulae and auricular appendages, for I noted that one of the commonest sites for their occurrence is in front of the ascending crus of the helix, the point at which a depression or fistula would be formed if a pre-helical appendage became submerged during development (see Figs. 21, 22). The fistula which is found on the ascending helix occurs at the point where two tubercles fuse to form the anterior or mandibular part of the helix. When one remembers that the external ear is formed out of six tubercles which fuse together across the first cleft, one wonders that gaps or fistulae are not of more common occurrence.⁴ The external auditory meatus is a secondary formation of the first cleft. It is at first a solid plug of epithelium which afterwards (ninth week) becomes canalized. A specimen in the college collection shows an arrest in the opening up of the meatus, while the external ear is normally formed. In this case, too, the eighth nerve is absent, and it is observed that malformation of the external ear is frequently attended by a degree of deafness. In the two human specimens in the college museum, with absence or grave arrest of the external ear, there is absence of the auditory nerve.

Dermoids occur in connexion with the first visceral cleft. There are three specimens in the college collection, two of them being retro-auricular and one pre-auricular. I noted two specimens in school museums in which there was a retro-auricular fistula or depression; it is probable that dermoids are formed in connexion with such recesses, which are of uncertain origin.

Cervical Appendages.

The only representative of these abnormal structures in metropolitan museums is a specimen (535) presented to the college collection by Mr. Frederic Eve. He removed it from the neck of a boy, its situation being near the mid-point of the anterior border of the right sterno-mastoid. The lamina of cartilage which supported the process was continuous with a small irregular bar of bone. Mr. Bland-Sutton⁴ states that the cartilage is elastic in nature, and that the appendix is usually associated with a cervical fistula. On the other hand, Rieffel,⁵ who has collected 37 cases, found that a fistula was never associated with

* In 50,000 school children Dr. Francis Warner observed 33 with auricular appendages. In 2,000 people, Urbantschitsch (quoted by Paget) saw 12 with auricular fistulae.

the appendage. In the college collection there are specimens representing the cervical appendage in the pig and goat. It is usually believed that cervical appendages are really auricles developed in connexion with the second cleft. We know that there is a tendency in vertebrates for a segment of the body to assume the characters of an adjoining segment; the last lumbar vertebra, for example, may assume the characters of the first sacral. Here we may have a manifestation on the part of the second cleft to assume to some degree the characters peculiar to the first. There is a difficulty in this assumption which will be dealt with when speaking of cervical fistulae, namely, the second cleft has no separate opening on the neck; it opens in common with the other clefts through the cervical sinus.

Fig. 24.—Ear of ewe showing accessory ostium (Preparation 531, presented by Mr. Bland-Sutton). A, Incisor like tooth; B, ostium distended by rod; C, external auditory meatus.

ear is a small meatus-like opening on the anterior margin of which is implanted a tooth, apparently an incisor. The abnormal opening may lead into the pharynx or end blindly. Although situated so closely to the ear there can be little doubt the opening really represents an accessory mouth. The duplication of the hind gut, and formation of two anal openings is a well known condition; duplication of the fore-gut leading to the formation of a double mouth is also well known. One of the mouths may undergo retrograde changes or imperfect development, and thus come to assume the form seen in these sub-auricular ostia.

Malformation of the Tongue.

Amongst the specimens at my disposal there are four—three of them in the college collection—which show malformation of the tongue. The head of a lamb (No. 186) shows a most interesting anomaly. On one side the lower jaw and all the parts developed in connexion with it are absent, so that the buccal cavity and tongue are fully exposed on the cheek. The basal part of the tongue—the part behind the foramen caecum and V-shaped sulcus—is fully formed. That part is formed out from the second and third arches, but the anterior or mandibular part of the tongue is only formed on the sound side; on the side of the defect the corresponding half is absent. The specimen demonstrates two things: (1) That even the anterior part of the tongue is bilateral in origin; (2) that the anterior part of the tongue is entirely derived from the mandibular arch of the embryo, for in the specimen under discussion the right half of this arch and all the structures derived from it are absent. The two other specimens of lingual malformations are to be found in the pathological series of the college museum, where they are classed as

tumours of the tongue (Nos. A 2271 and A 2271a); they are exactly of the same type; one was obtained from a deformed, the other from an anencephalic child. One is represented in Fig. 25. At first sight it appears as if the anterior part of the tongue were absent, but such is not the case; it is the basal part of the tongue, the part behind the V sulcus and derived from the second and third visceral arches, which is arrested in development. Combined with this there are two other anomalies: (1) The submaxillary gland has been arrested in its development, and forms a clustered glandular mass within the frenum and along the side of the tongue; (2) Meckel's cartilage has become ossified into a chain of bones, the chain ending behind in the malleus. The sublingual gland is normally developed; the sulcus or depression which is formed in the floor of the primitive mouth so as to cut away the submaxillary gland from the tongue has never opened out. There is in these cases a real condition of tongue tie, the frenum being distended by the glandular mass within it. There is a close relation between these specimens and the cases described by Mr. Barling,⁶ Dr. G. W. Griffith,⁷ Dr. Kettner,⁸ and a case I saw at the London Hospital in a girl aged 2 years. In all of these cases the anterior part of the tongue is imperfectly separated from the floor of the mouth, and in two of them the anterior part of the tongue seemed to be divided, the line of division being occupied by a median mass, which I infer contained muscular and glandular tissue representing submaxillary gland. It is also possible that the median mass may represent the primary element described by His as the tuberculum impar, for Kallius⁹ has shown that only a small part of the tongue arises from that element, the major part being formed from lateral elements which grow up from the floor of the mouth and submerge the primitive median element. The fourth specimen on my list (see Table I) is a child in the Museum of St. George's Hospital, which shows a median cleft of the lower lip and jaw, the cleft extending to, and involving, the apex of the tongue.

Fig. 25.—Congenital malformation of the tongue. A, Lower lip; B, gums; C, glandular mass distending frenum and extending backwards in paralingual fold (D); E, buccal part of tongue, developed from first arch; F, tonsil; G, pharyngeal part of the tongue, arrested in growth, developed from second and third arches; H, entrance to larynx and rudimentary epiglottis.

Fig. 26.—Diagram to show the position and connexions of the various embryological remnants in the neck (Dr. S. J. A. Beale). A, Tonsil; B, remnant of tonsillar sac (from second cleft recess); C, carotid body between external and internal carotid arteries; D, stalk of thymus; E, cervical sinus; F, cervical thymus; G, common carotid artery; H, suprahyoid part of median thyroid; I, infrahyoid part of median thyroid, dividing; J, stalk of lateral thyroids; K, K, thymus in thorax.

have spoken of the downgrowth of a ledge of epithelium which occurs in the floor of the mouth on each side of the tongue, and thus separates off the submaxillary glandular area. It seems highly probable

Sublingual Dermoids.

In the mass of glandular tissue lying by the side of the tongue, in the specimen shown in Fig. 25, there is a small cyst with a lining of mucous stratified epithelium, and in the interior of the cyst, debris of epithelial cells. The cyst has all the characters of a dermoid. I

that the cyst alluded to above is a product of the paralingual epithelial downgrowth, and that sublingual dermoids are derived from this downgrowth. There are four specimens of sublingual dermoid cysts in the college museum. Such cysts have been fully described by Mr. Butlin¹⁰ and by Dr. Edginton.¹¹

Embryological Remnants in the Neck.

Before proceeding to describe the various specimens which represent, or are believed to represent, congenital malformations of the neck, it will be well to enumerate, in the light of recent embryological researches by Hammar¹² and of Henry Fox,¹³ the various vestigial structures which may assume a pathological form during the course of development and growth. These elements are shown diagrammatically in Fig. 26. They arise in connexion with the first, second, third, and fourth visceral clefts, the last two being clefts in which gill formations are developed in fishes. From the first cleft arises the median thyroid, the element which forms the isthmus and nearly a half of each lateral lobe of the thyroid in man. The stalk of this element normally disappears, but parts may persist and form tumours or cysts in the line of its formation (Fig. 26). The tonsil is developed at the pharyngeal orifice of the second cleft, but the fundus or outer part of the pocket (see Fig. 26) may persist.¹⁴ In the figure this pocket ought to be shown passing between the lesser and greater horns of the hyoid bone (second and third arches), and ending in the fork between the external and internal carotid arteries. From the third cleft, represented by the thyrohyoid space, arises the thymus and carotid body; the stalk atrophies in man as far down as the lower pole of the thyroid body, but it may persist, as is normally the case in the pig, and is occasionally the case in newly born children.¹⁵ From the fourth cleft (represented in the interior of the pharynx by the pyriform fossa; see Fig. 27) arises the lateral thyroids and the parathyroids. The stalk of the lateral thyroid buds becomes reduced to a fibrous cord which emerges from the larynx behind the inferior cornu of the thyroid cartilage (Fig. 26, r), and forms in man a firm ligamentous bond (the suspensory ligament) which binds the thyroid body to the cricoid cartilage, but in carnivora, as may be seen in specimens prepared by the prosector of this college (Wm. Pearson) form free fibrous cords. Besides these four endodermal derivatives of the visceral clefts—the stalk of the median thyroid, the fundus of the tonsillar clefts, the cervical stalks of the thymus, and the stalks of the lateral thyroids—there is also a fifth element, a derivative of the covering or ectoderm of the embryonic gill area, which may persist and give rise to new formations. That is the cervical sinus (see Fig. 26, s). We see repeated in the human embryo by this sinus, a modification of the gill chambers with which we have long been familiar in the air-breathing fishes—the Dipnoi—and in the more

primitive amphibia. In these vertebrates the gill clefts open on the surface of the neck by a common chamber; there is now no doubt that the cervical sinus of the

human embryo represents that common gill chamber. During the fourth and fifth weeks the endodermal pockets which give rise to the tonsil, thymus, and lateral thyroids abut on, and are attached to, the fundus of the cervical sinus. The lateral thyroid buds are the first to lose their connexion with the sinus, and the tonsillar pockets the last, because their attachment is the most firm and extensive. There is an especially close connexion between the cervical sinus and the carotid body—a derivative of the third cleft. The fundus of the cervical sinus persists in the mammalian embryo long after its orifice on the neck is closed and in the mammalian embryo becomes a temporary thymus-like body; lymphoid tissue develops in its wall. The question naturally arises: Why is the cervical sinus drawn out into a long fistulous passage (Fig. 26), and why is its orifice on the anterior border of the sternomastoid usually near the upper border of the sternum? The answer is to be found in the remarkable change undergone by the neck towards the end of the second month of development. Up to that period, although the cervical part of the spinal column is developed, the chin rests on the breast, the neck being indicated merely by a transverse depression or groove in which the cervical sinus opens on each side (Fig. 28). At the end of the second month and commencement of the third the head becomes extended so that the chin is lifted off the heart and thorax and a neck becomes demarcated. It is not the heart that has receded from the head, but the opposite. If the surface connexion of the cervical sinus has remained unbroken, then the sinus becomes drawn out into a long tubular structure along with the carotid arteries and trachea. The orifice of the sinus usually occurs on the anterior border of the sternomastoid an inch or rather more above the sternum, but under certain circumstances the orifice may be drawn up during extension of the neck to occupy a higher position. Cervical appendages occur at the orifice of the sinus.

Cervical Cysts.

There is not, as far as I am aware, a single specimen in metropolitan museums representing a cervical sinus and fistula *in situ*, but, on the other hand, specimens of excised cervical cysts are numerous, two of which are said to have had a fistulous opening. From their structure and relations, it is evident that the majority of these are derived from the cervical sinus. They are sometimes described as dermoids, sometimes as sebaceous cysts, and sometimes as lymphoid cysts. I

exclude those cysts which contained grumous matter or blood, because such specimens as have been minutely examined appear to be derivatives of the thyroid—cysts formed in aberrant thyroids. Of the eighteen cysts which I regard as derived in part or entirely from

Fig. 27.—Diagram showing the position of the visceral clefts in the human pharynx. A, Eustachian tube (first cleft); B, lateral recess of pharynx; C, tonsil (second cleft); D, position of third cleft, with tonsillar body; E, suprahyoid part of median thyroid stalk; F, retrohyoid mucous glands; G, pyriform fossa (fourth cleft); H, hyoid; I, infrahyoid part of median thyroid stalk; J, right division of median thyroid, forming cyst and fistula; K, left division of median thyroid, forming isthmus.

Fig. 28.—Reconstruction of neck of pig embryo, showing the development of the thyroid, thymus, and cervical sinus (after Henry Fox). A, Mouth; B, groove between mandible and thorax; C, external auditory meatus; D, cervical sinus; E, epidermal pocket from cervical sinus to tonsillar recess; F, epidermal pocket to thymus outgrowth; G, M, thymus outgrowths; H, L, lateral thyroids; I, median thyroid; J, trachea; K, oesophagus; N, epidermal process to tonsillar outgrowth; O, cervical sinus.

the cervical sinus, twelve are in the college museum. A specimen removed from the neck, opposite the thyroid cartilage, by Sir Felix Semon, may be taken as a typical example. A small bar of cartilage is embedded in its wall—such a cartilage as is occasionally found in the tag at the orifice of the cervical sinus—a product of the operculum which forms the outer wall of the sinus. The cyst is lined by columnar epithelium, similar to that which lines the trachea, but not ciliated. Very little lymphoid tissue is to be found in its wall. The contents are of a milky nature, containing epithelial products. The lining epithelium may be stratified and squamous, or columnar with cilia. Usually these cysts have a close connexion with the carotid sheath, especially near the point of bifurcation of the carotid artery into its internal and external branches. The cervical sinus may fuse and be continuous with the tonsillar recess, so that the cyst wall has both an ectodermal and endodermal lining.

Aberrant Thyroid Tumours and Cysts.

Such tumours and cysts are not well represented in metropolitan medical museums. In the college collection there is a fine specimen presented by Mr. Butlin of a double thyroid tumour, one part of which was removed from the tongue above the hyoid bone; the other part lay below the hyoid. Clearly they were derived from remnants of the median thyroid. Mr. Makins¹⁶ recorded the removal of a thyroid tumour from the posterior part of the tongue in the neighbourhood of the foramen caecum, and recently Dr. Perkins¹⁷ and Dr. T. B. Murphy¹⁸ have been able to bring together about 50 cases of intralingual tumours formed of thyroid tissue, most of them in women. Such tumours frequently occur in the *right* submaxillary region;¹⁹ none are recorded from the same region of the left side. Such tumours apparently represent the right division of the median thyroid. The median thyroids occupy a submaxillary position in the frog. There are two specimens of subhyoid cysts in the college collection, but one of these may be connected with a retrohyoid group of mucous glands which open in the larynx, and were first demonstrated to me by Wm. Pearson, prosector to this college (see Fig. 27). The right division of the median thyroid may be also arrested in development, and form a cyst which afterwards establishes a fistulous opening in the neck.²⁰ Schrager²¹ has recorded 15 cases of cervical cysts derived from the lateral thyroids; they occur beneath the lower third of the sterno-mastoids.

Cysts and Tumours connected with the Tonsillar and Thymic Outgrowths.

There are two remarkable specimens in the museum of Guy's Hospital, numbered 499 and 501. In the first specimen, the pharynx of a man suffocated by a foreign body in the pharynx, there is a recess in the position shown in Fig. 27, containing lymphoid tissue—a form of tonsil. The recess represents the inner orifice of the third

cleft. In the second specimen (No. 501) a cystic tumour, occupying the subparotid region, opens at this depression; its position and connexion make it clear that this tumour is connected with the stalk of the thymic outgrowth. There is a similar tumour in the museum of the Westminster Hospital medical school. Dermoid cysts found in the anterior mediastinum and in the lower part of the neck may be derived from epidermal inclusions, carried down by the outgrowing buds of the thymus from the cervical sinus.

Congenital Tumours of the Neck.

Altogether I have seen six specimens of congenital tumour of the neck, four of which are in the college museum. In three of these the tumour is median in position, and has expanded into the neck like a huge goitre (Fig. 29), but in the fourth it has developed to the left side, spreading upwards under the ascending ramus of the jaw, zygoma, and temporal fossa, so as to indent the temporal fossa and compress the brain (Fig. 30), and expand the malar and superior maxilla into thin bony plates which cover the tumour. In none of these specimens is any trace of the thyroid to be found; the tumour is the thyroid. On section these tumours are seen to be multiloculated and solid. On microscopic examination they are found to be composed of compressed cysts lined with columnar epithelium. At the bases of the columnar epithelium a layer of lymphoid tissue usually is found; indeed, a large part of the tumour is lymphoid in nature. In the septa between the spaces are found islands of cartilage. How the thyroid body of the fetus comes to assume such a structure and size is an enigma. The tumours resemble congenital sacral tumours, and in many ways have much in common with the tissues which form gills.

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Fig. 29.—Congenital tumour of neck replacing thyroid gland. The right half of the specimen is represented.

Fig. 30.—Congenital tumour of the neck extending into and filling the left temporal fossa. A, Left cheek in which the malar and superior maxilla are extended as thin bony plates; B, right cheek; C, bony ridge representing temporal crest; E, spinal column and muscles.

UNDER the will of the late Mr. John William Day of Harrogate, Huddersfield Infirmary receives £1,000 for the endowment of a bed, and Harrogate Infirmary a sum of £100.

THE British Association for the Advancement of Science, which this year has gone to Canada, commences its sessions at Winnipeg next Wednesday. The President for the year is Sir Joseph Thomson, F.R.S., Cavendish Professor of Experimental Physics at Cambridge.