

Pharyngeal arches
Tongue development and malformations
Salivary glands development

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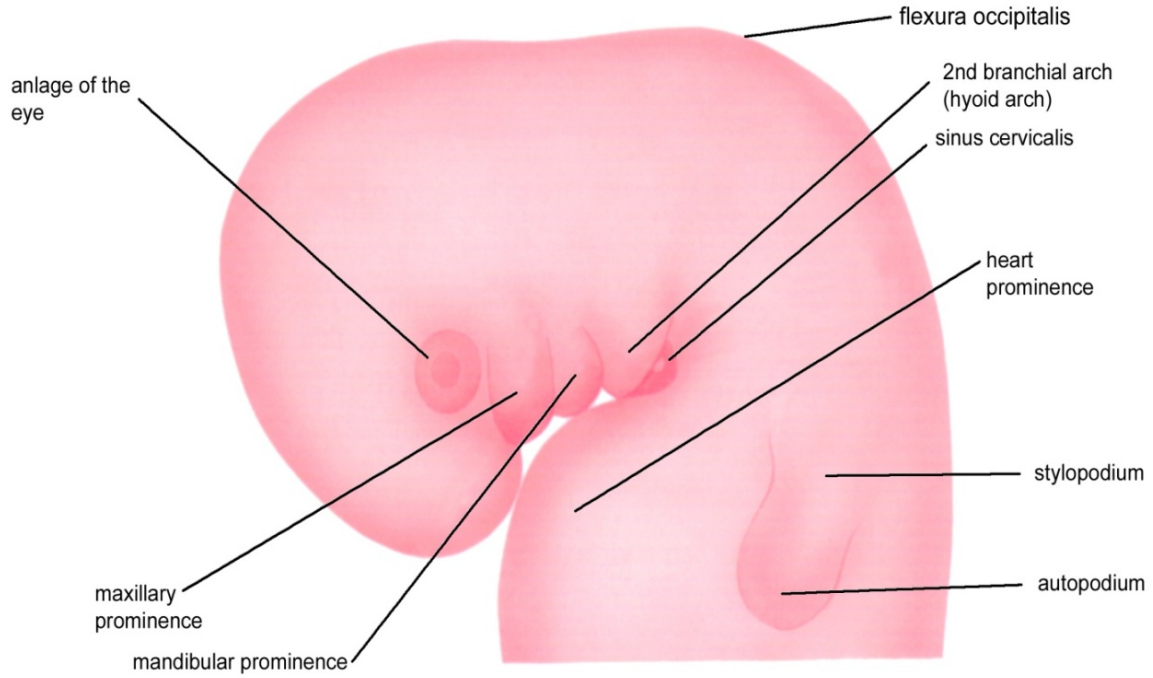
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Pharyngeal arches

Phylogenetically conserved organ, serves as a carrier for gills (which work as a respiratory organ)

First appears in sharks, around the pharyngeal gut

In vertebrates, transforms and forms the basis of important organs - branchiogenic organs



Pharyngeal arches

The pharyngeal apparatus starts to develop in human embryos in the neck region behind the frontal (frontonasal) prominence in the second half of the 4th week

Pharyngeal arches

Pharyngeal pouches (entodermal)

Pharyngeal clefts (grooves) (ectodermal)

Membranae obturantes

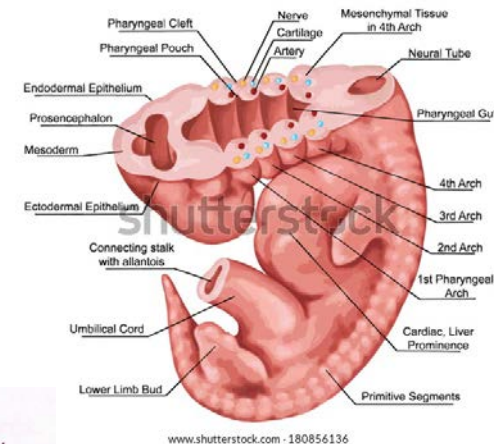
All structures are paired

6

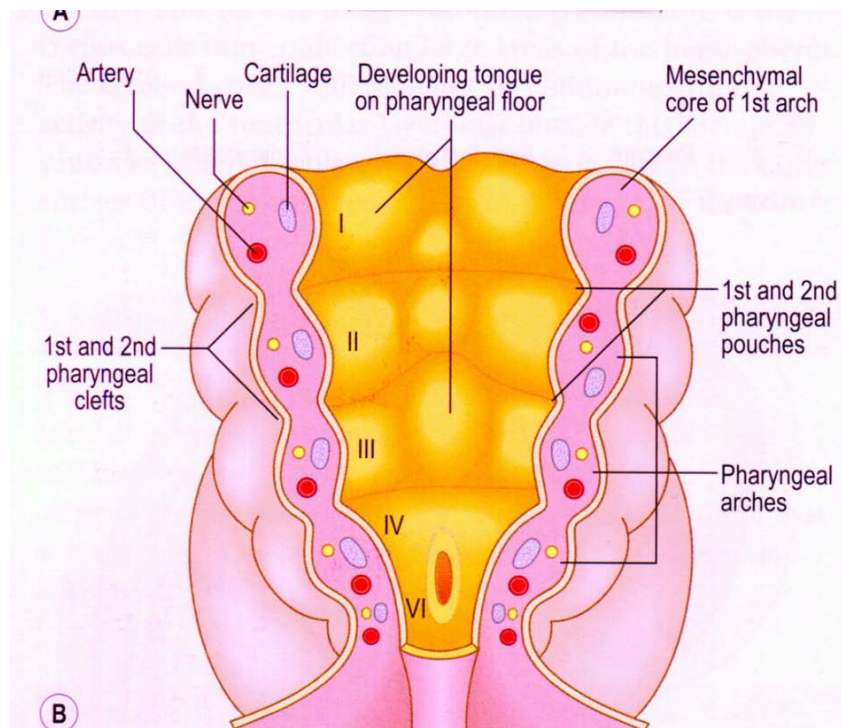
5

4

4



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B

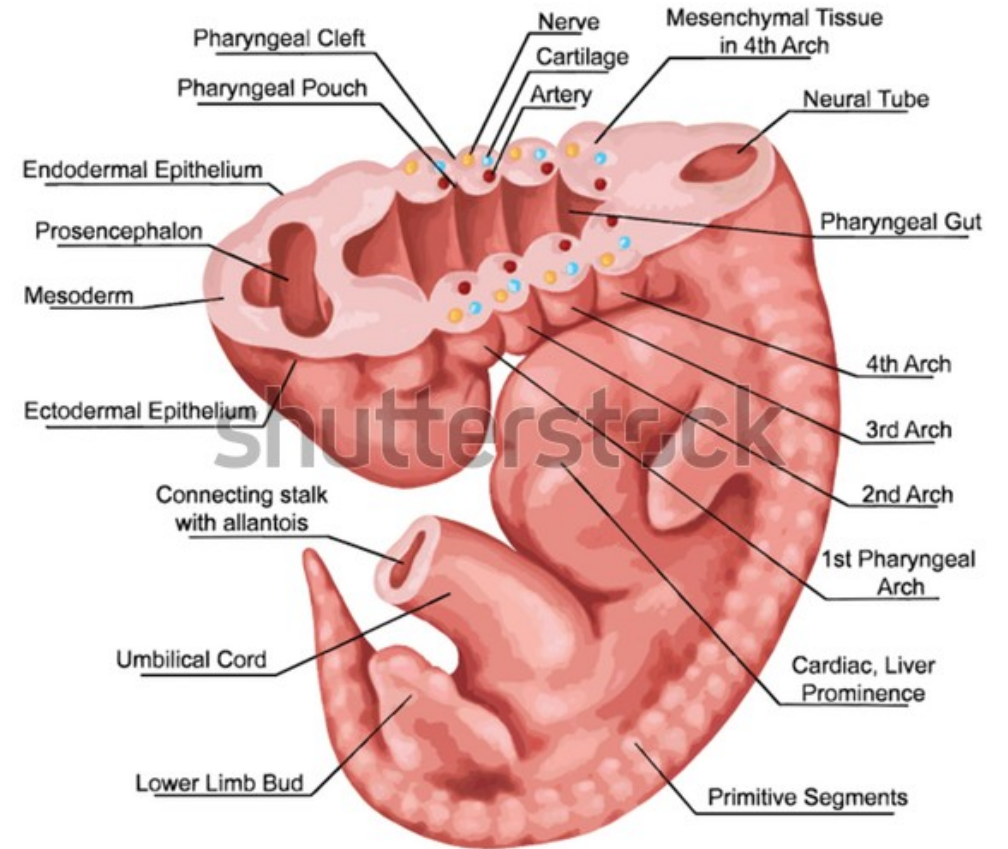
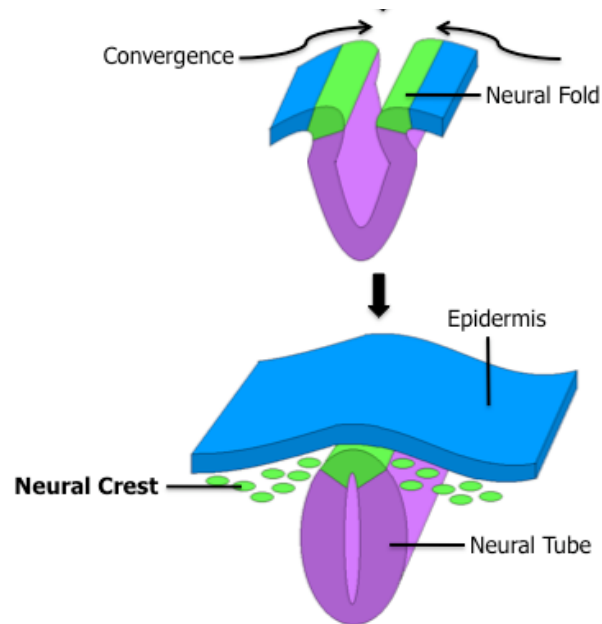
Derivates of pharyngeal folds	Arch number	Aortic arch	Cranial nerve	Examples of branchiomeric muscles	Skeletal derivates	Derivates of pharyngeal pouch
external auditory meatus	I mandibular	maxillary artery	V trigeminal	muscles of mastication etc.	malleus, incus spheno-mandibular lig. Meckel cart.	I middle ear auditory tube
	II hyoid	hyoid, stapedia artery	VII facial	muscles of facial expression etc.	stapes, styl. proc., stylohyoid lig., part of hyoid cart.	II supra-tonsillar fossa
neck	III	internal carotid artery	IX glosso-pharyng.	m. stylopharyngeus	parts of hyoid cart.	III thymus, parathyr. gland
	IV	right subclavian artery, aorta	X vagus	pharyngeal and laryngeal musculature	laryngeal cart.	IV thymus parathyr. gland ultimobranch. body

Pharyngeal (branchial) arches (6)

The first four - cause a obvious segmented structure of the neck (5th and 6th are rudimentary)

Cells of the **mesencephalic and rhombencephalic part of neural crest migrate** into the paraaxial mesoderm of the first cervical somites and contribute to formation on arches and subsequently organs

The formation of pharyngeal arches is controlled by the endoderm of the pharyngeal arches



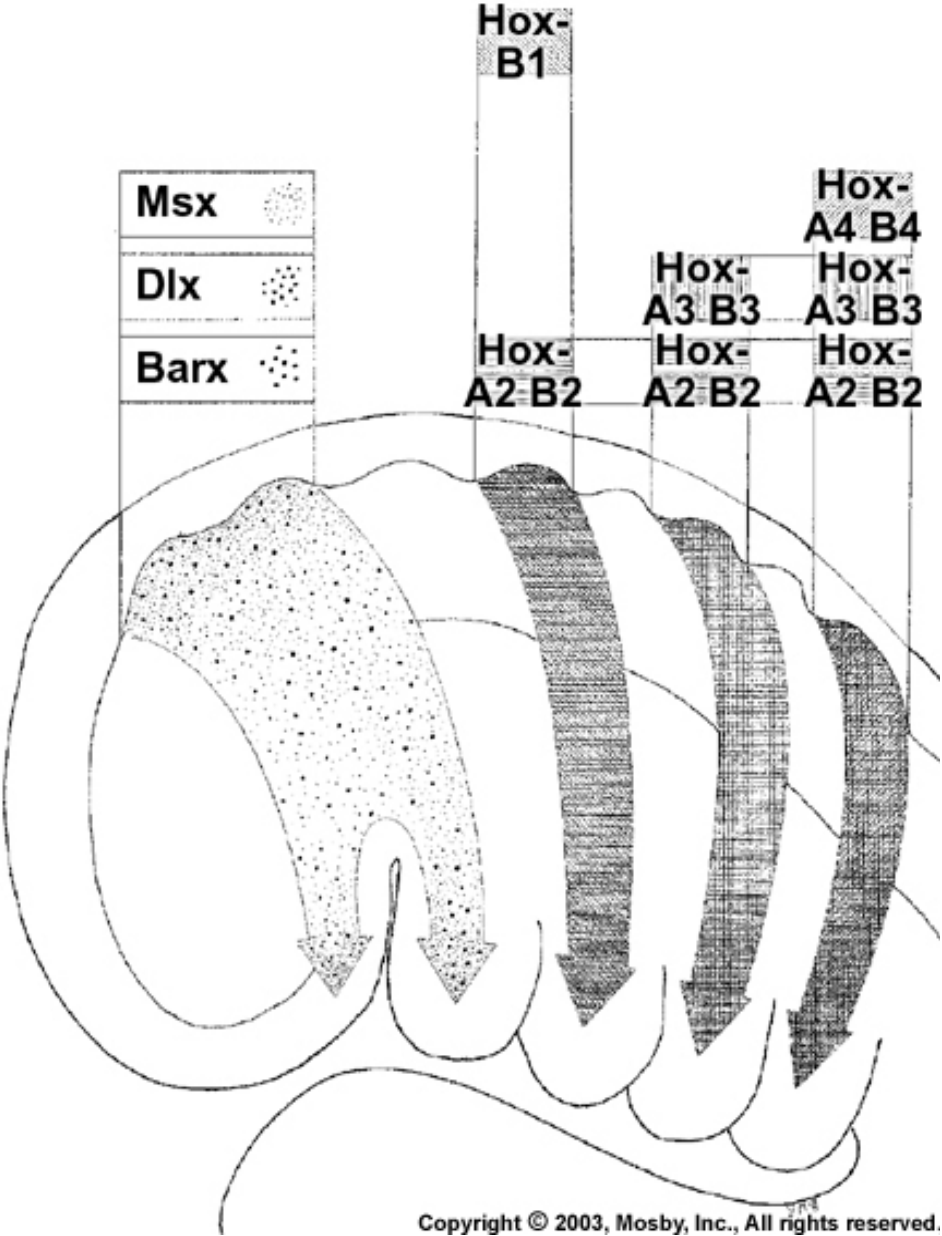
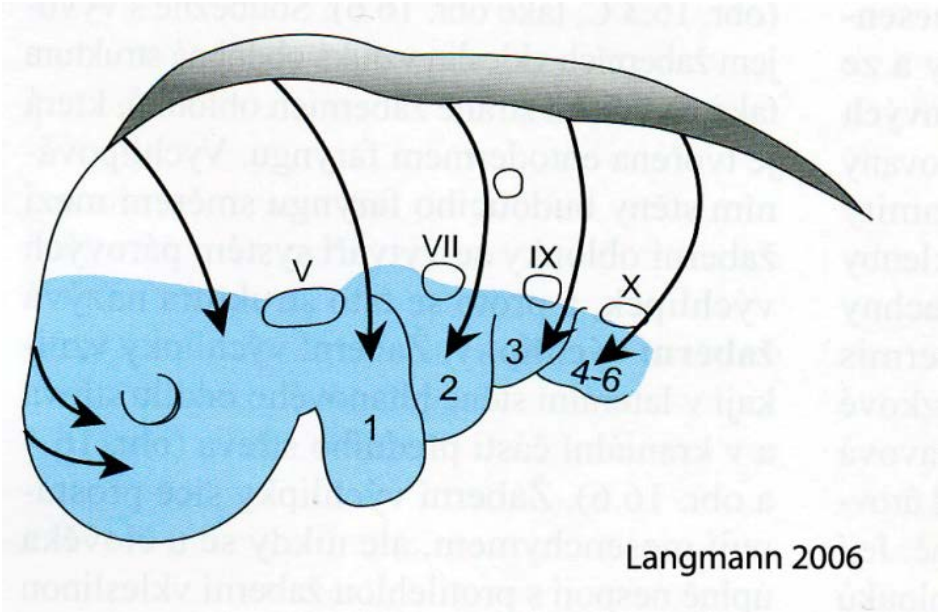
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Ectomesenchymal derivatives: ligaments, cartilages, bones

Paraaxial mesoderm derivatives: muscles of pharyngeal arches and branchial arteries

Migration of neural crest (ectomesenchyme) in several migratory pathways

Controlled by **Hox genes** which regulate expression of transcription factors with effector function

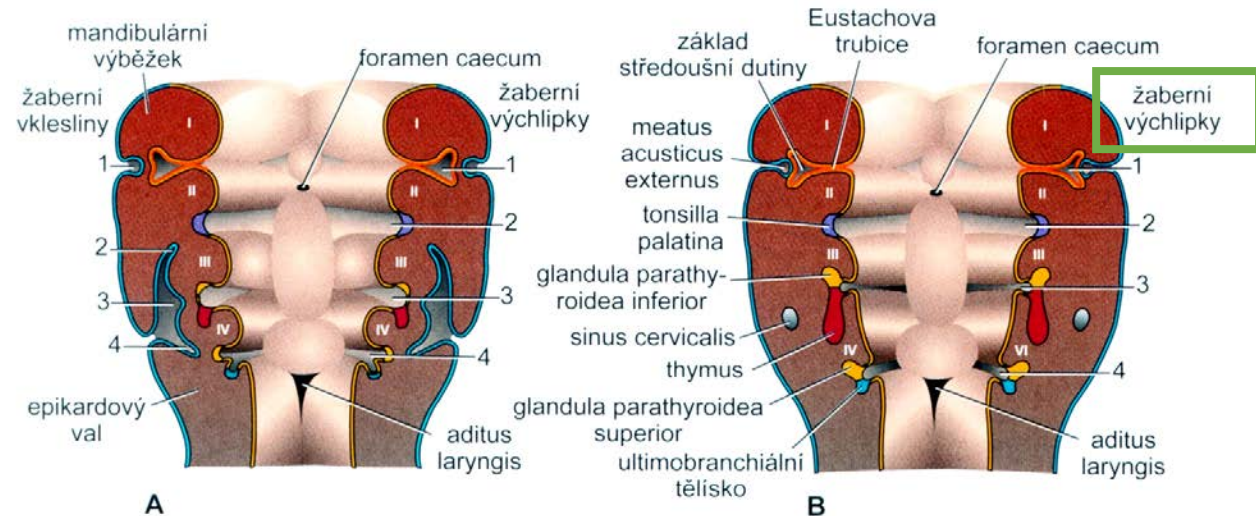
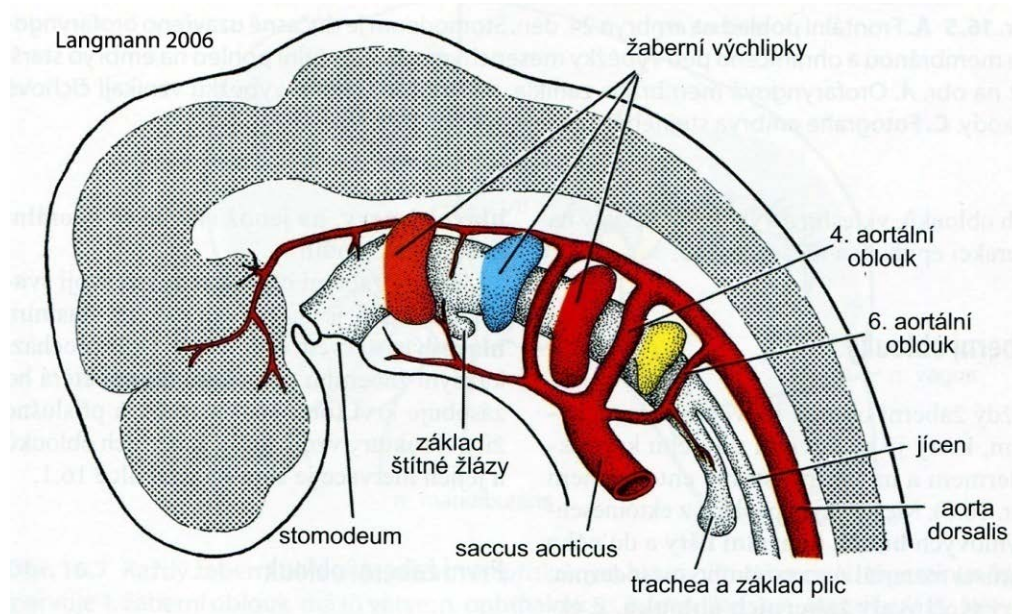
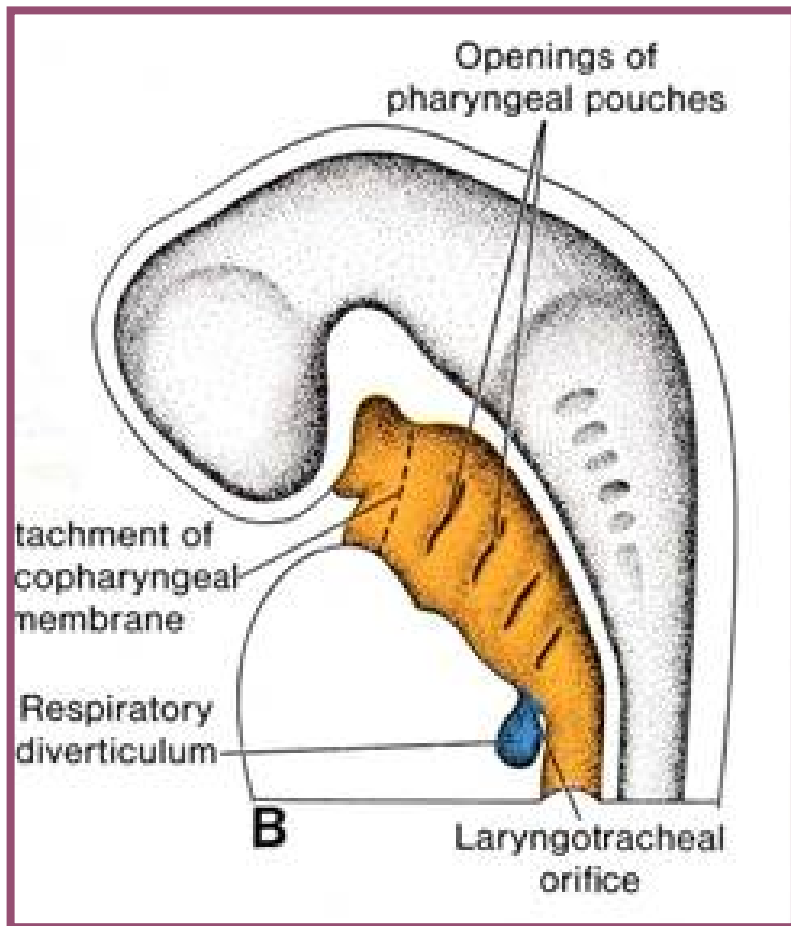


Pharyngeal pouches - 5

The first starts to develop on the stage of 5 somites

The 5th is rudimentary and develops as a part of the fourth pouch at end of the 1st month

Endodermal origin

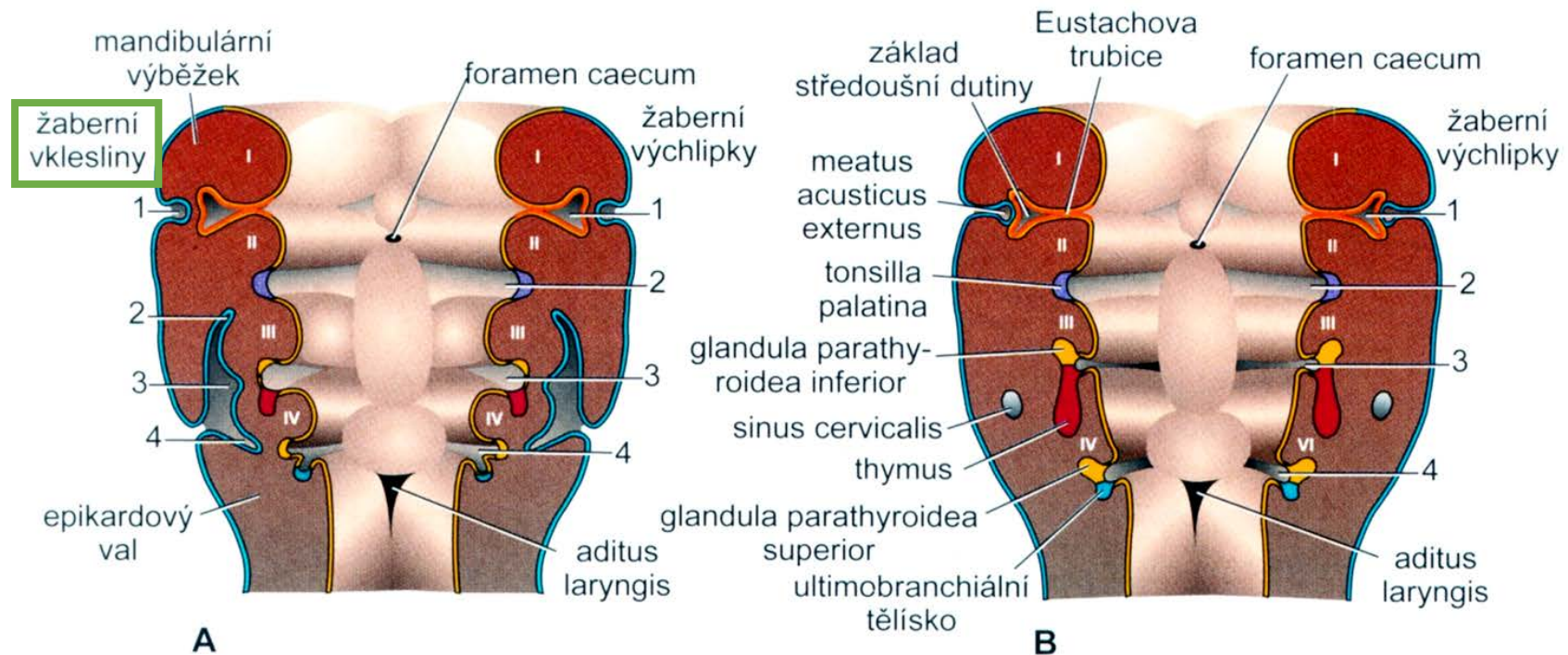


Pharyngeal clefts - 4

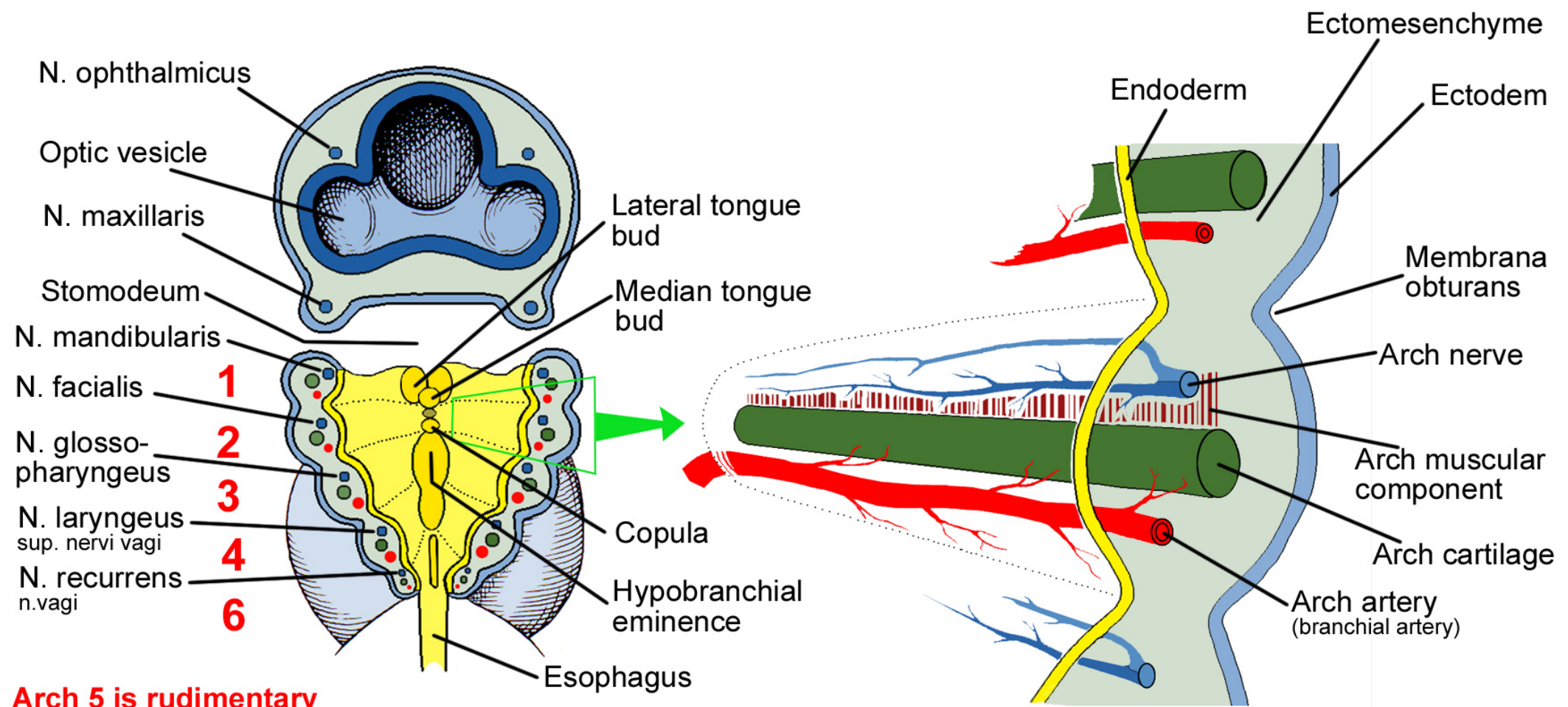
They have the form of shallow grooves
ectoderm origin

Membranae obturantes - 4

Two-layer membranes that separate each ectoderm and entoderm groove (physiologically do not perforate in humans)



Frontal section through apparatus and branchial arch components



Arch 5 is rudimentary

A

In each arch is:

- Cartilage**
- Skeletal muscle basis (mezoderm)**
- Arch artery**
- Branchial arch nerve**

B

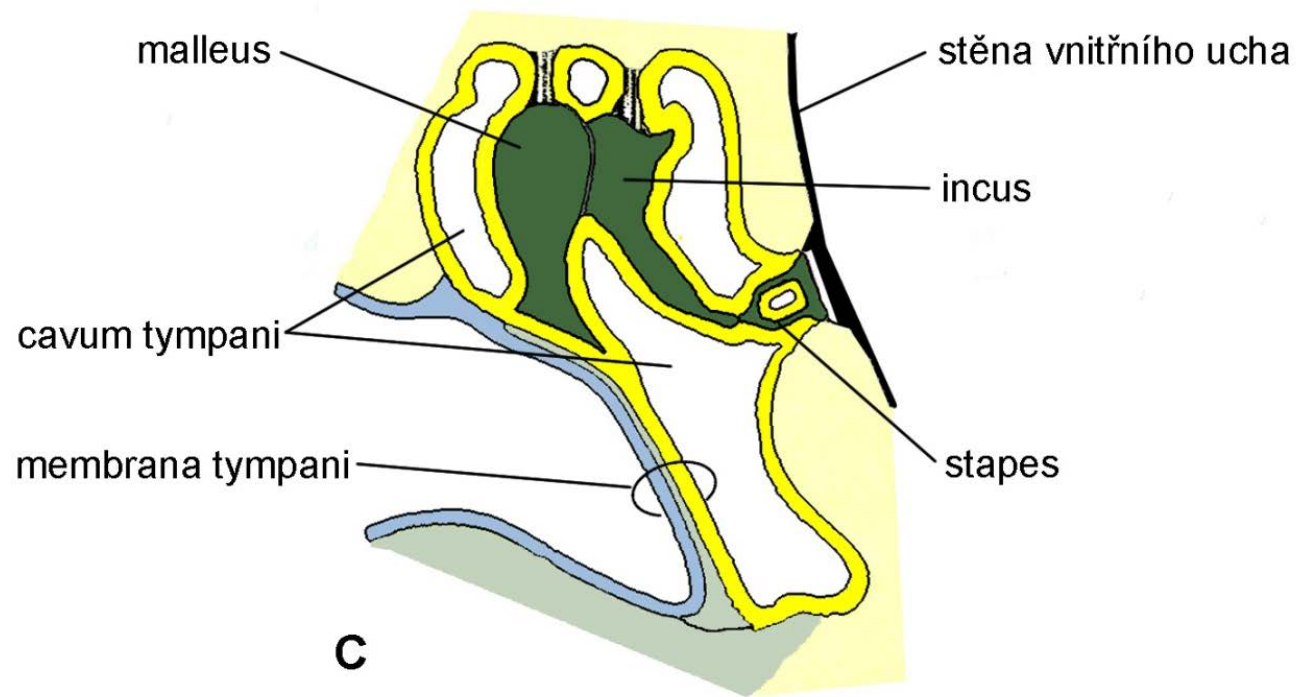
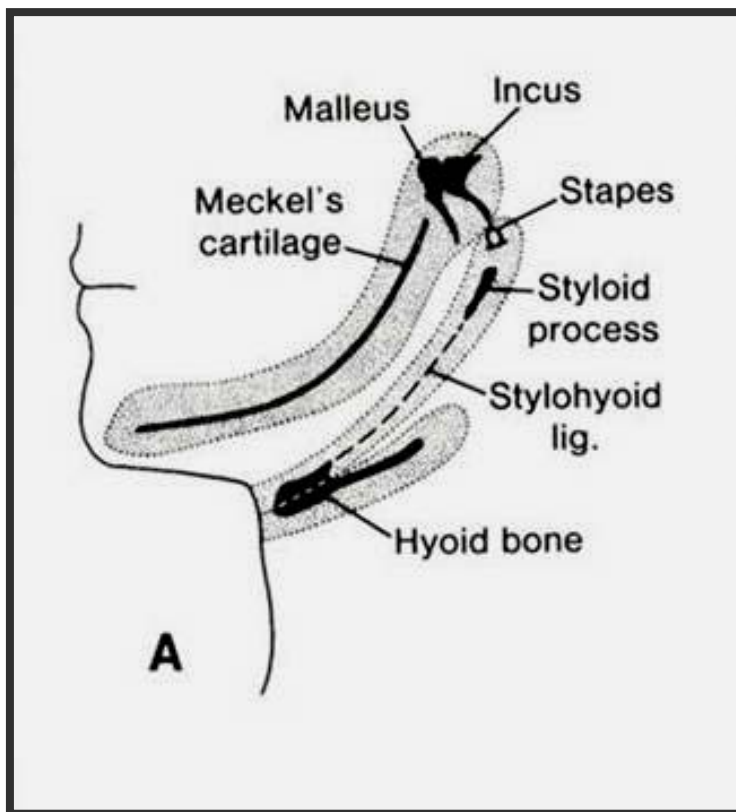
1. Pharyngeal arch (mandibular)

arch cartilage (Meckel's cartilage) - malleus, incus, lig. mallei ant., sphenomandibulare lig.

muscles of mastication, mylohyoid and anterior belly of digastric, tensor tympani, tensor veli palatini

the 1st aortic arch - disappears (a small portion may persist and form maxillary artery)

the 1st branchial nerve - trigeminal



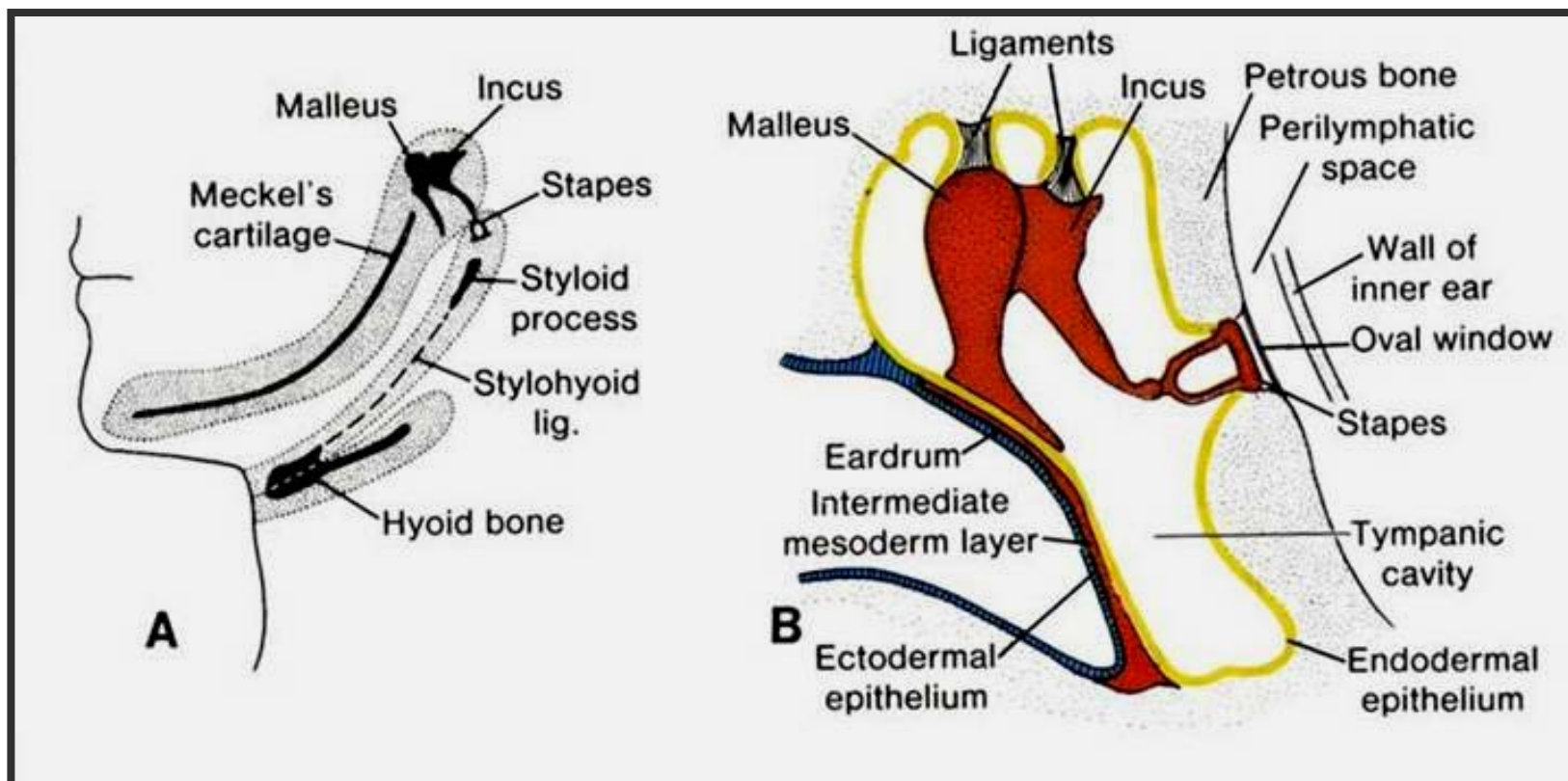
2. Pharyngeal arch (hyoid):

arch cartilage (Reichert's cartilage) - stapes, styloid process, lesser cornu of hyoid, upper part of body of the hyoid bone

muscles of facial expressions, stapedial and stylohyoid muscle, posterior belly of digastric

the 2nd aortic arch - disappears (small portions of this arch contributes to the hyoid and stapedial arteries)

the 2nd branchial nerve - facial



3. Pharyngeal arch

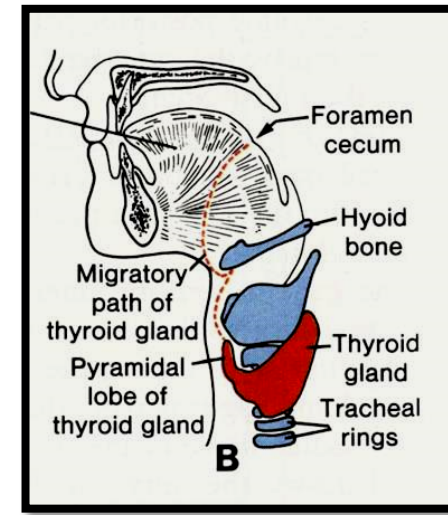
arch cartilage - greater cornu of hyoid, lower part of body of the hyoid cartilage

stylopharyngeus muscle

the 3rd aortic arch - has the same fate on the right and left sides and forms the first part

of the internal carotid artery

the 3rd branchial nerve - glossopharyngeal



Derivatives of pharyngeal arches

4. - 6. Pharyngeal arch

arch cartilages - laryngeal cartilages and tracheal rings

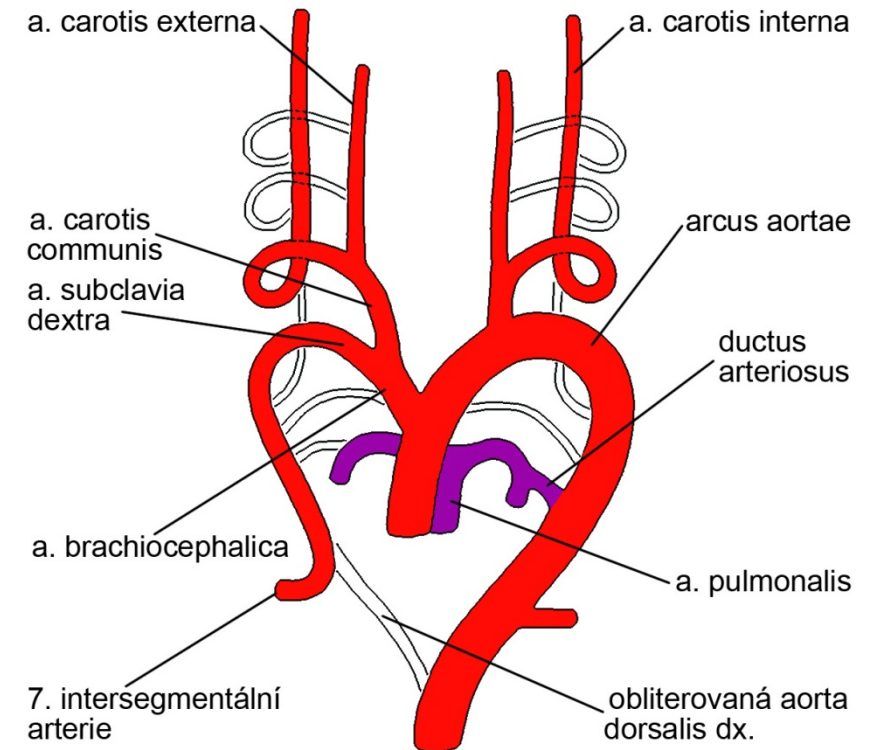
cricothyroid, levator veli palatini, constrictors of pharynx, intrinsic muscle of larynx

the 4th aortic arch - gives rise on left: a part of the aortic arch between left common carotid a left subclavian arteries; on the right: the proximal segment of the subclavian artery

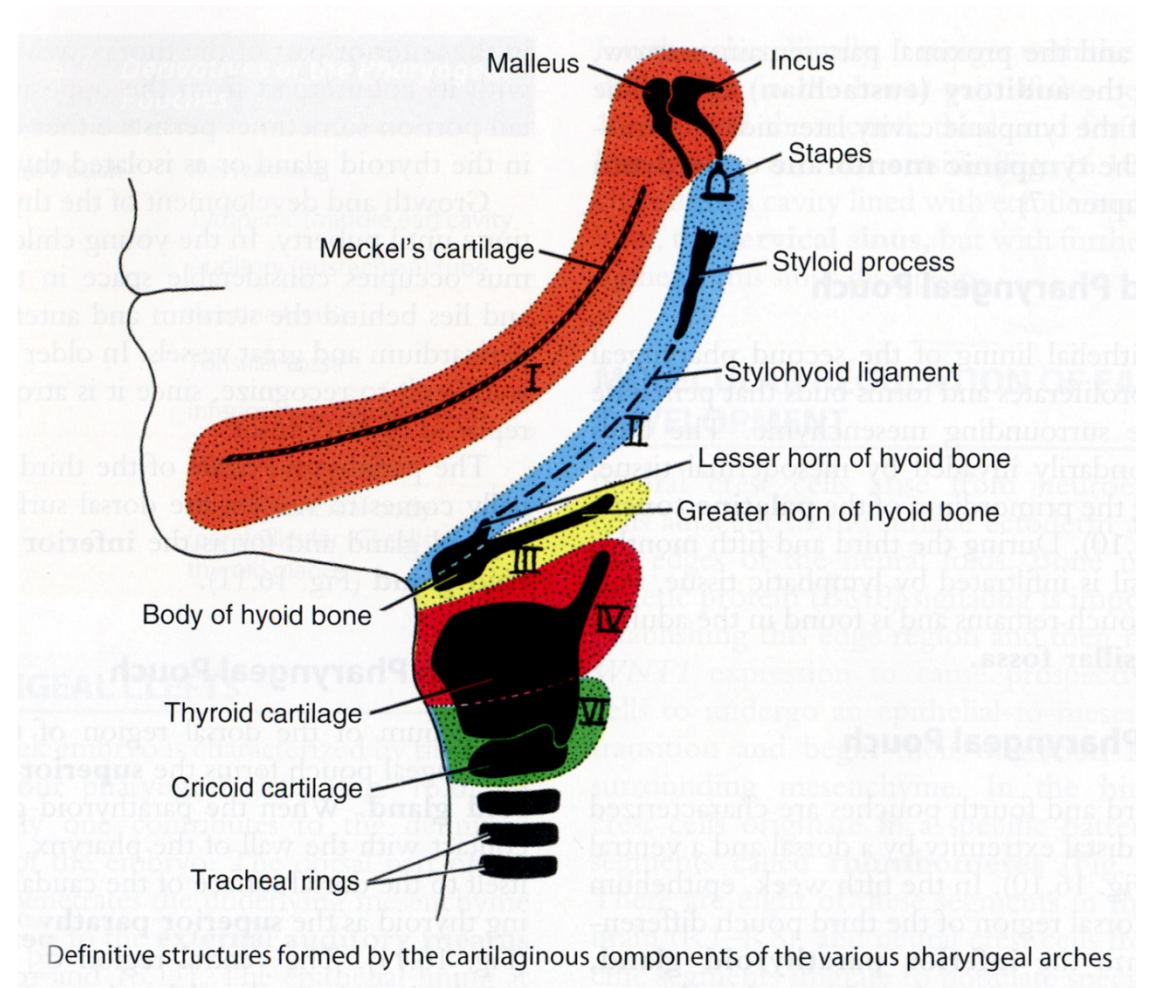
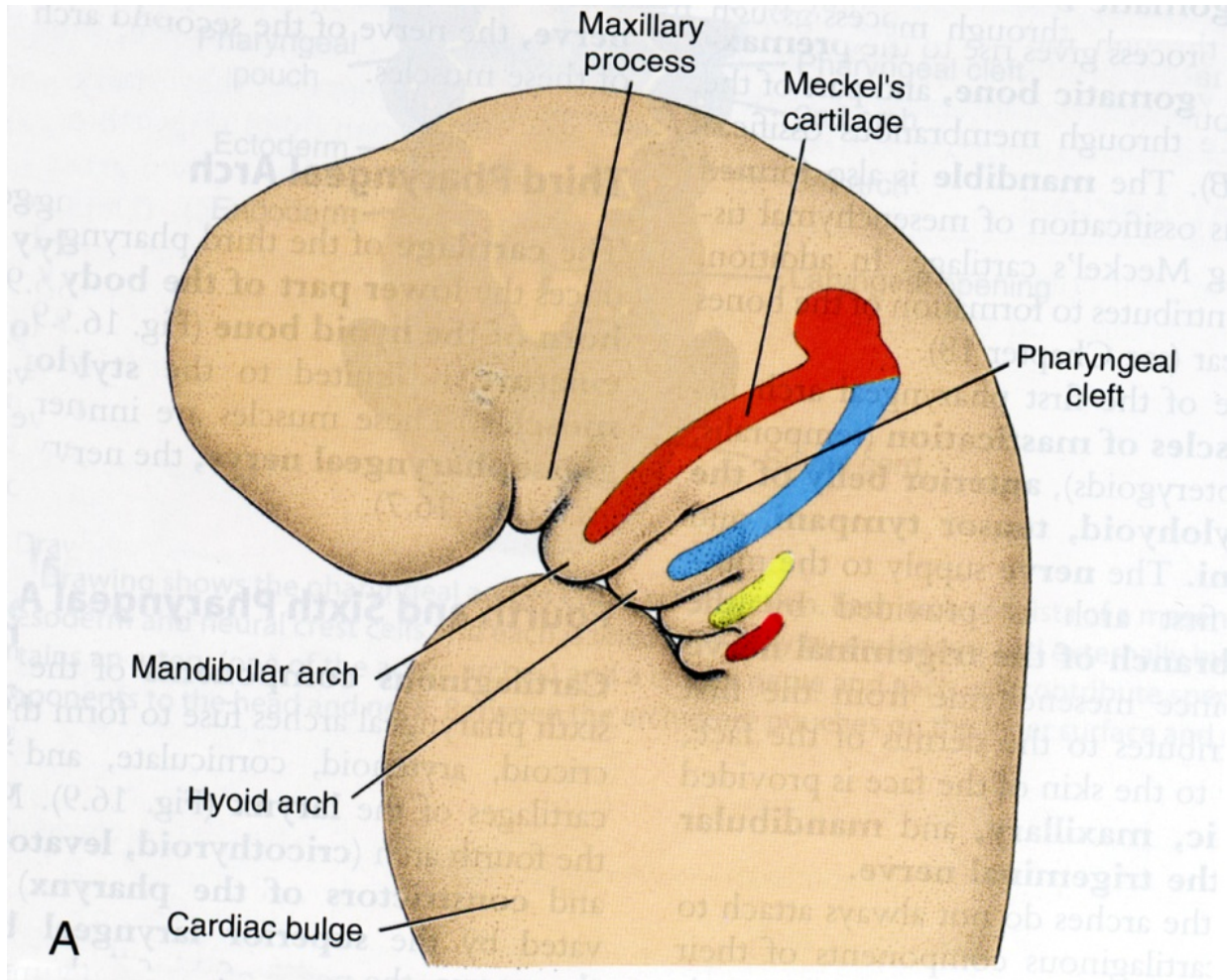
the 5th aortic arch - transient and obliterates

the 6th aortic arch - transformed into the pulmonary artery (their branches)

branchial nerves - vagus nerve /superior laryngeal, branch of vagus (from the 4th), recurrent laryngeal branch of vagus (from the 6th)



Transformation of cartilages of pharyngeal arches (summary)



Transformations of aortic arches (summary)

the 1st aortic arch – **disappears** (a small portion can persist to form short piece of the maxillary artery)

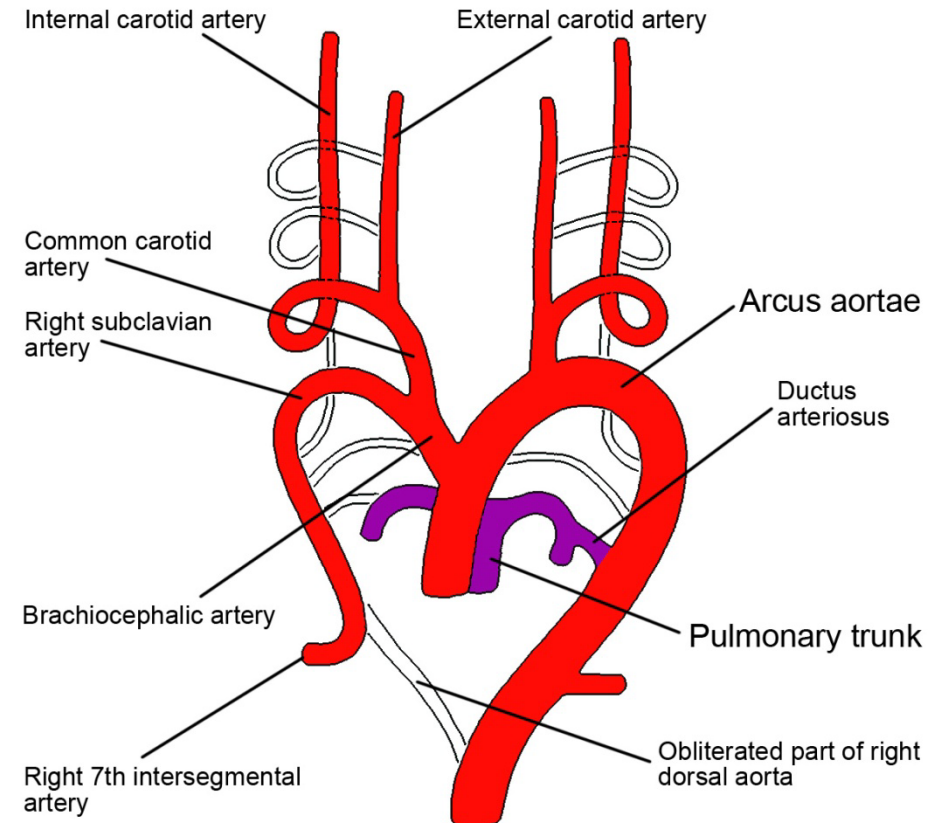
the 2nd aortic arch – **disappears** (small portions of this arch contributes to the hyoid and stapedia arteries)

the 3rd aortic arch – has the same development on the right and left side, it gives rise to the initial portion of **the internal carotid artery**

(the continuation of its trunk is formed by the cranial portion of the dorsal aorta + primitive internal carotid)

the external carotid derives from
the cranial portion of the ventral aorta

the common carotid corresponds to a
portion of the ventral aorta between
exits of the third and fourth arches

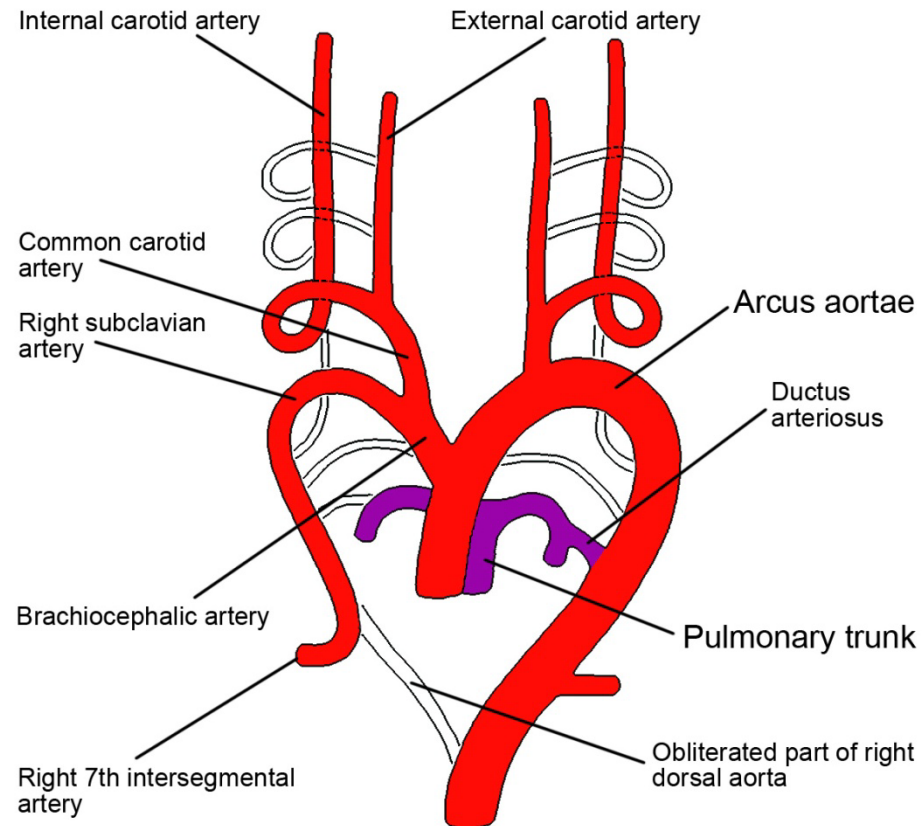


the 4th aortic arch - has ultimate fate different on the right and left side:

Left: forms **part of the arch of the aorta** between left common carotid and left subclavian artery

Right: forms the proximal segment of the **right subclavian artery**

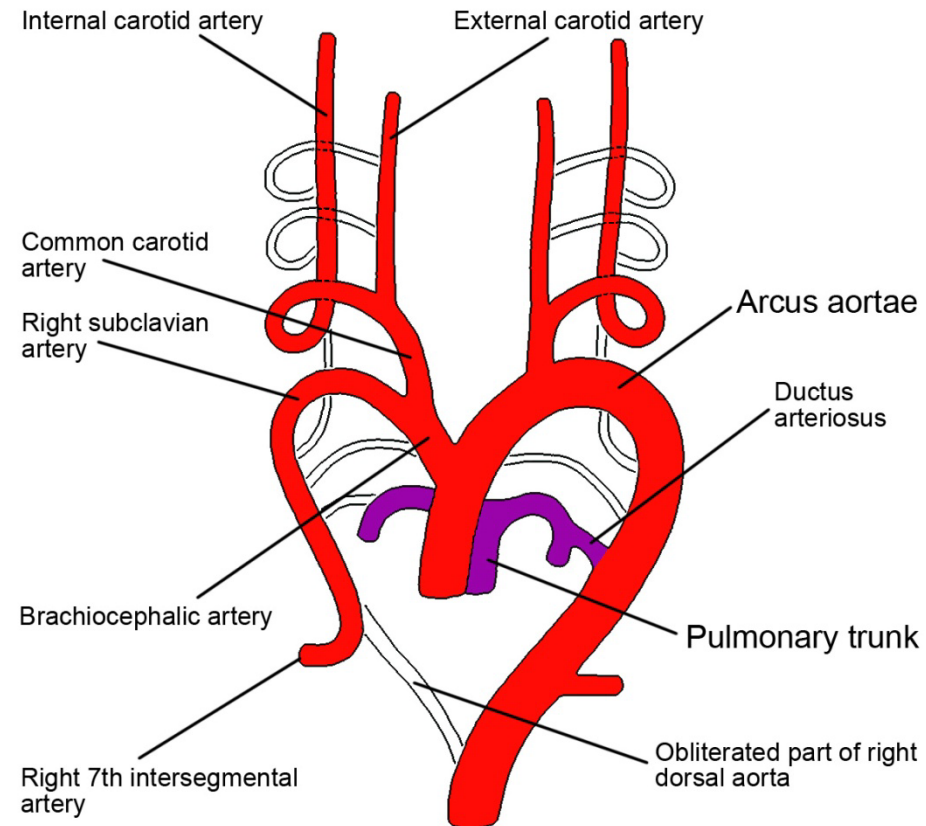
the 5th aortic arch - is transient and soon obliterates

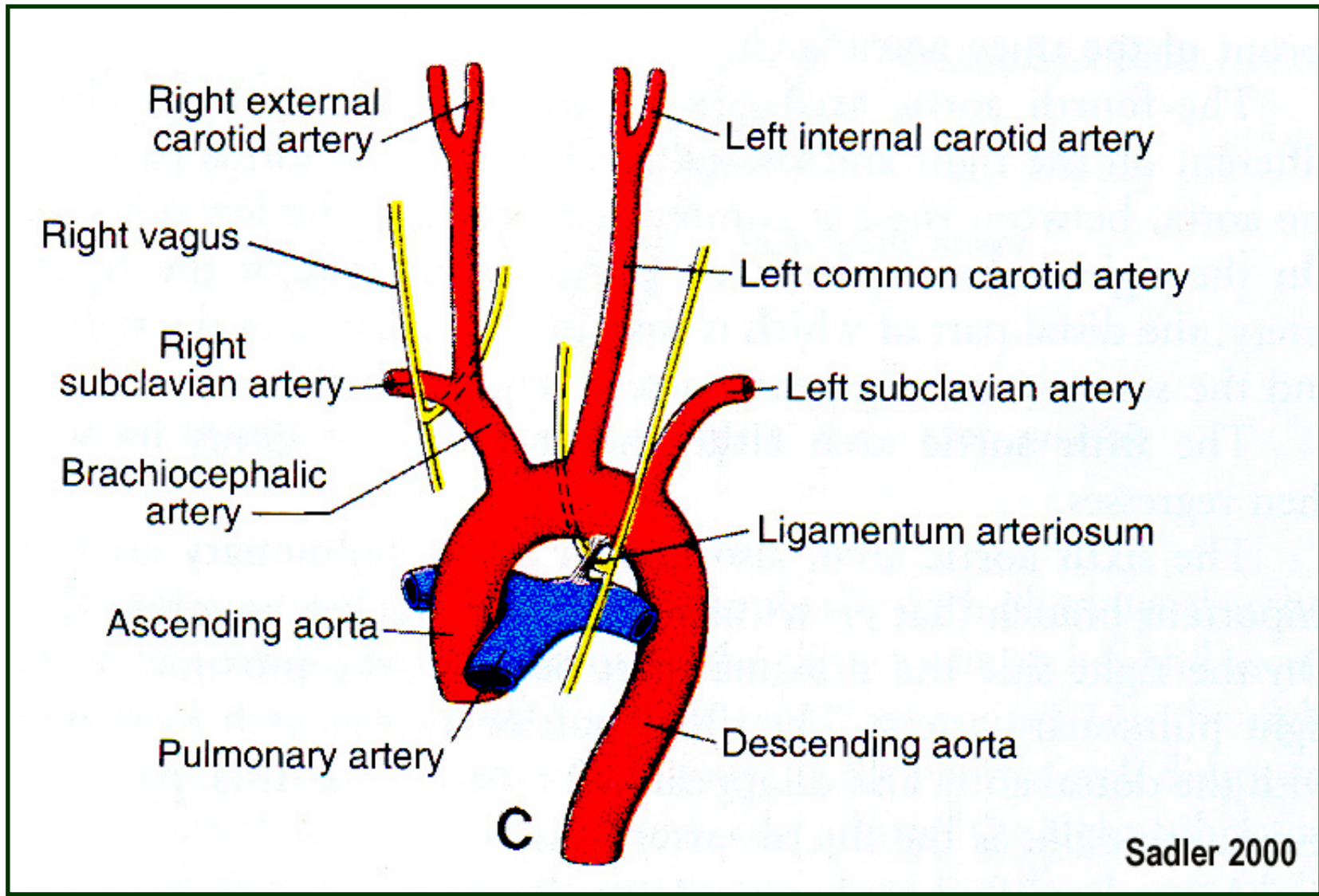


the 6th aortic arch - pulmonary arch - the proximal part transforms into the right branch of the pulmonary artery and the distal part disappears

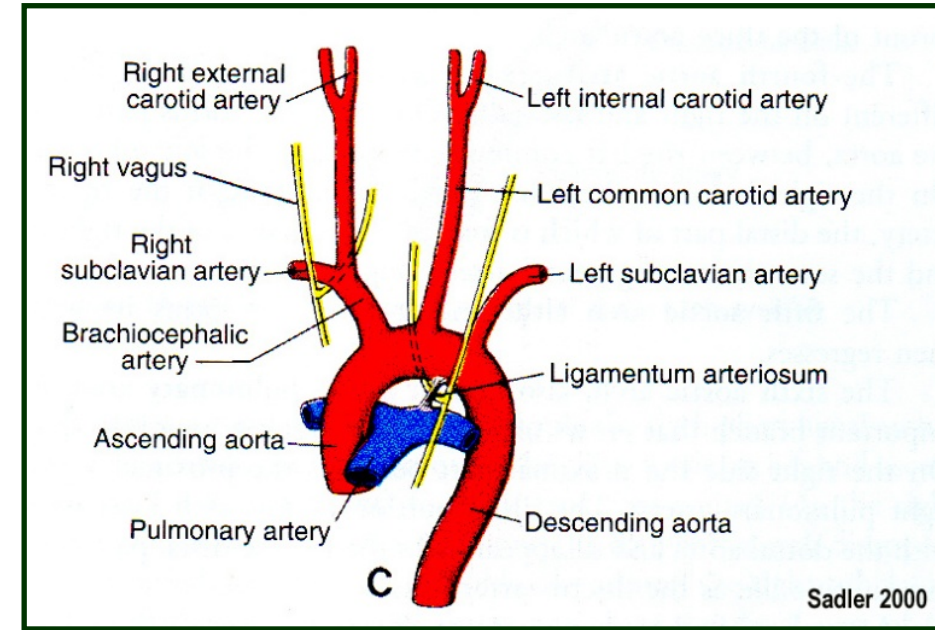
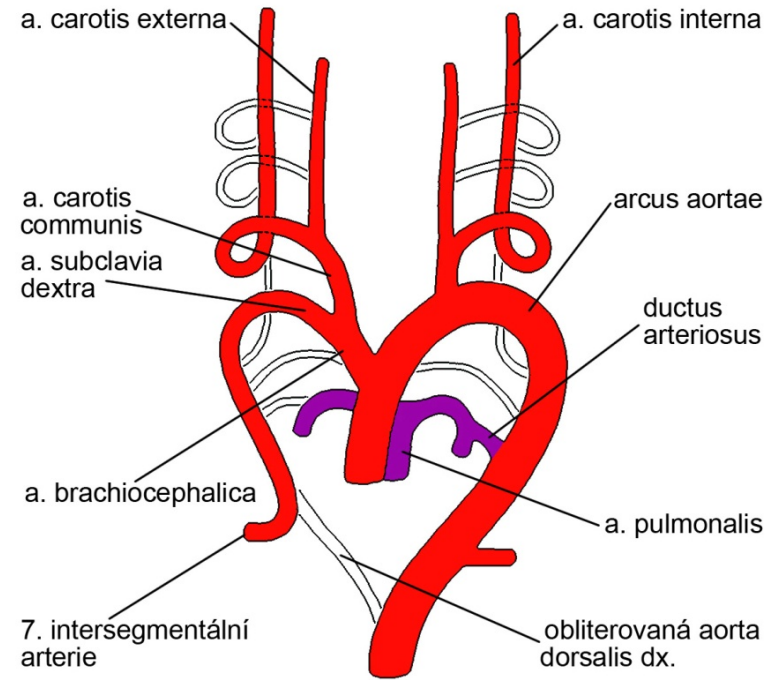
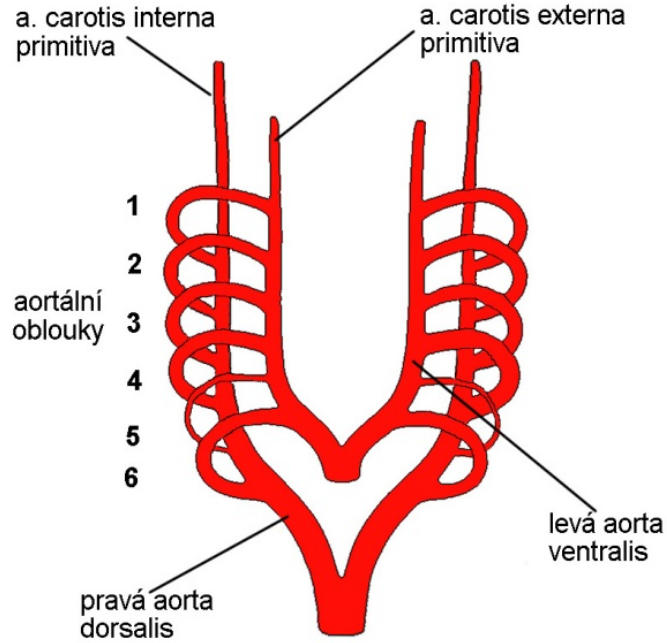
On the left side, the distal part persists as the **ductus arteriosus** during intrauterine life

The proximal part gives rise to the left branch of the pulmonary artery





Summary



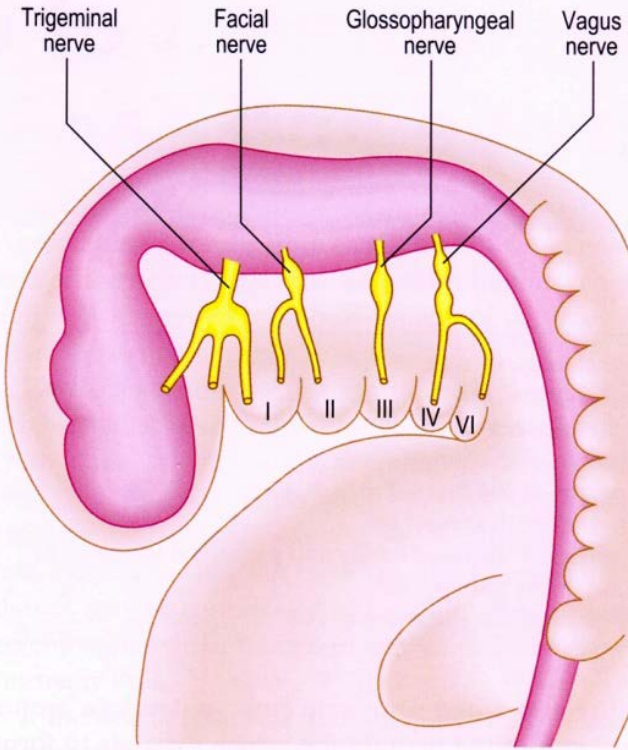


Fig. 11.3 Sagittal section of a 5-week-old embryo indicating the cranial nerves supplying the pharyngeal arches.

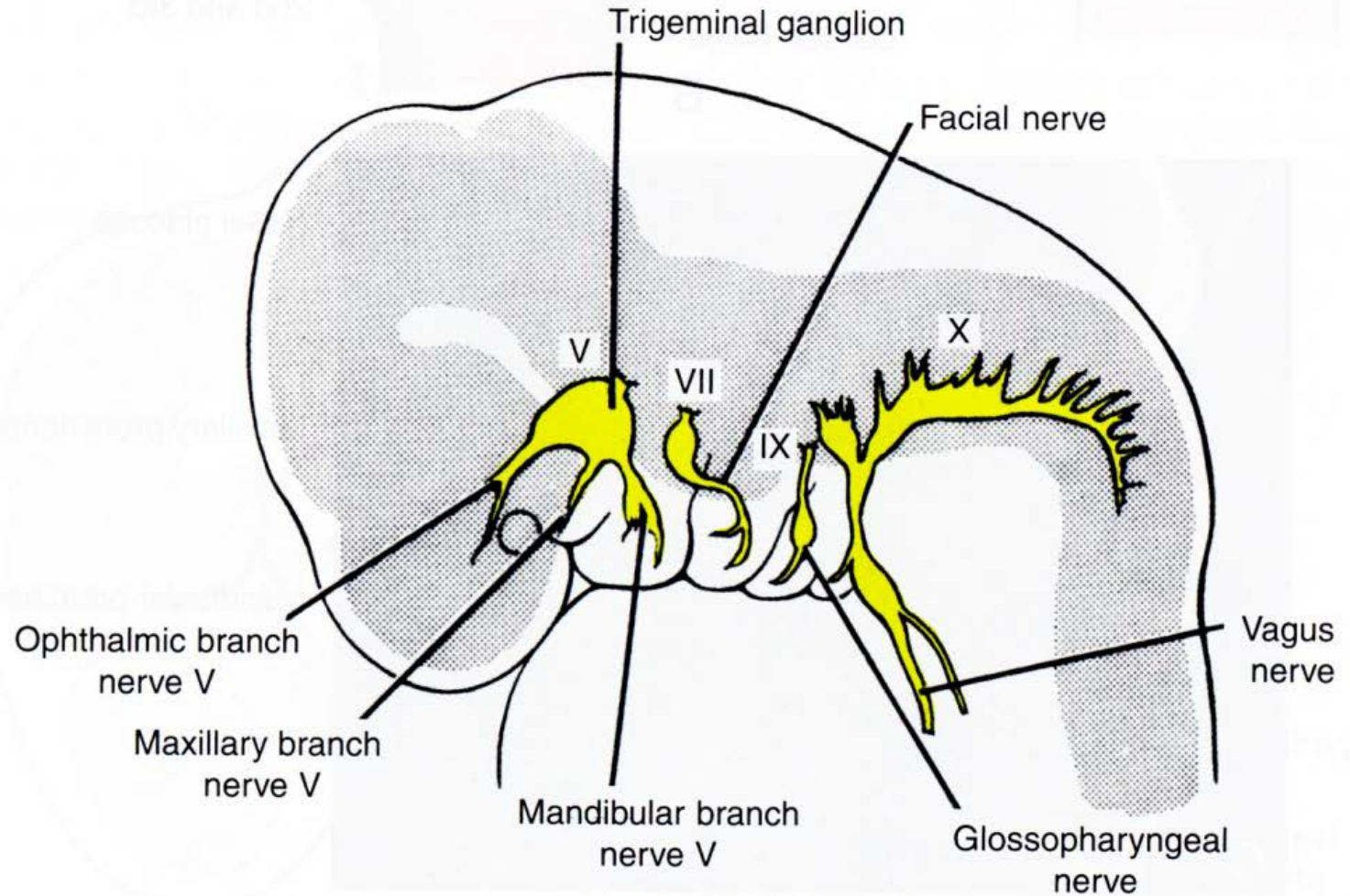
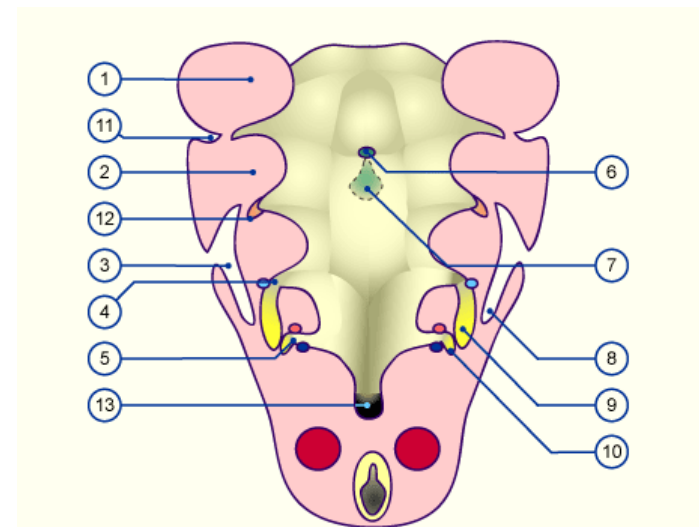
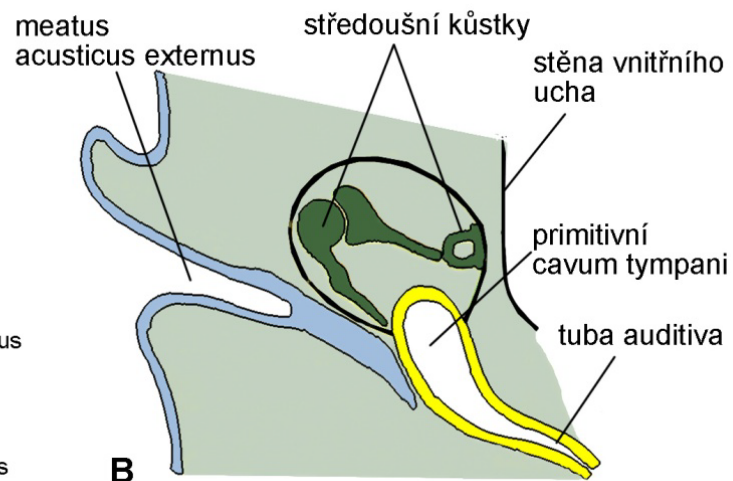
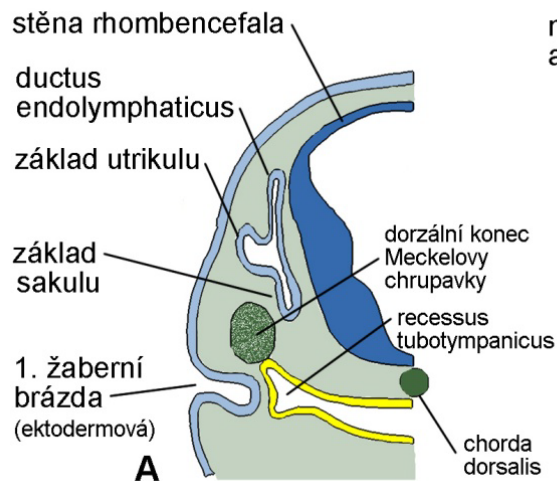
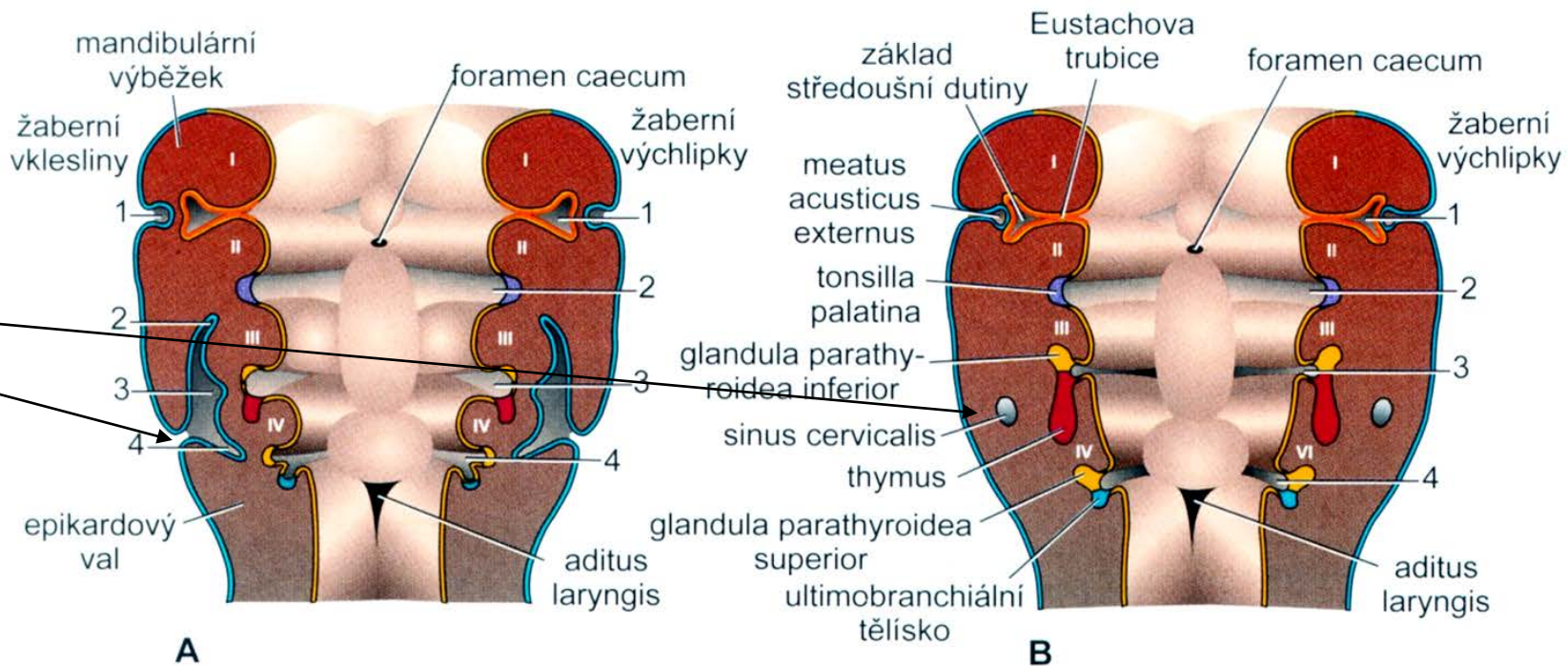


Figure 16.7 Each pharyngeal arch is supplied by its own cranial nerve. The trigeminal nerve supplying the first pharyngeal arch has three branches: the ophthalmic, maxillary, and mandibular. The nerve of the second arch is the facial nerve; that of the third is the glossopharyngeal nerve. The musculature of the fourth arch is supplied by the superior laryngeal branch of the vagus nerve, and that of the sixth arch, by the recurrent branch of the vagus nerve.

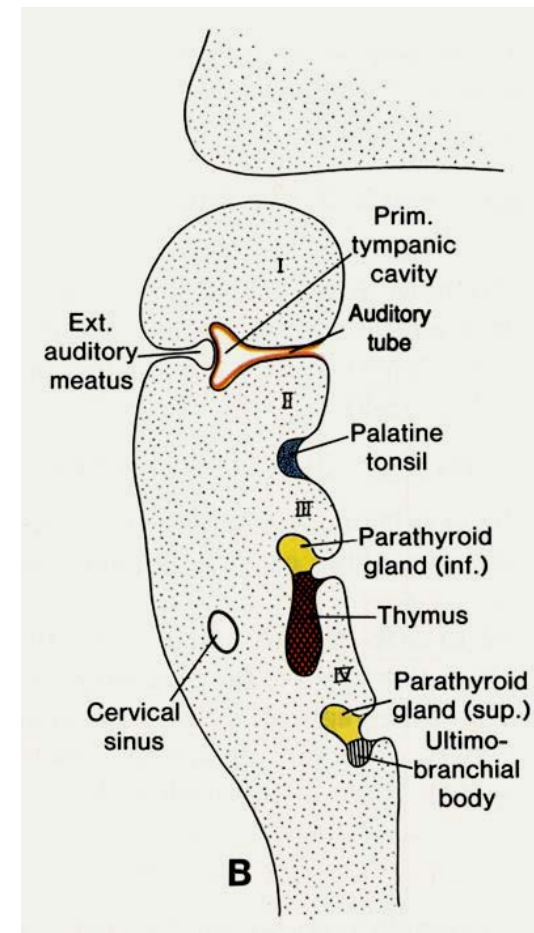
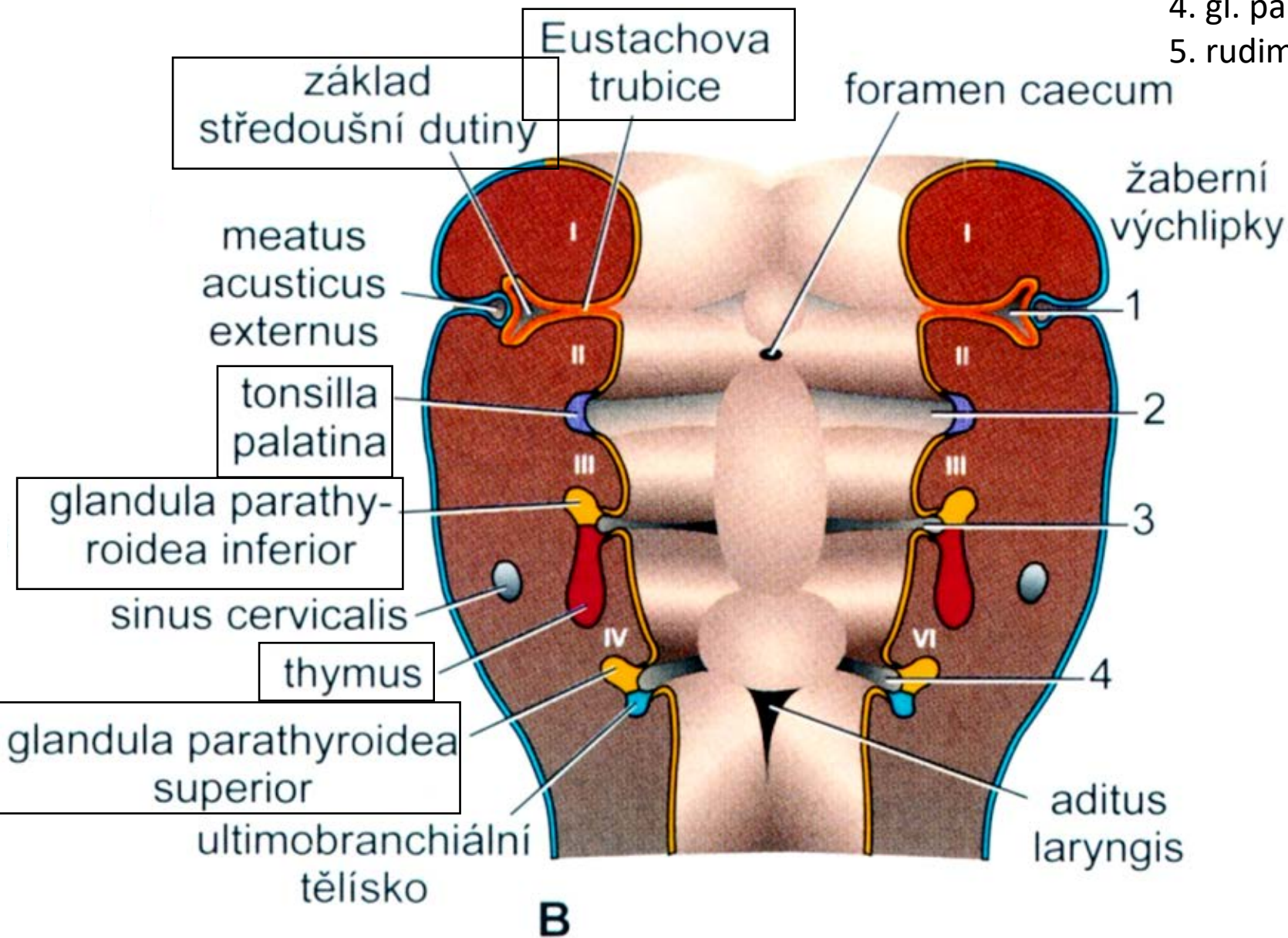
Pharyngeal clefts (ectodermal)

- Functional future has only the
1. pouch
 2. - 4. obliterates and form a sinus cervicalis



Derivatives of pharyngeal pouches

1. Cavum tympani and Eustachian tube
2. Fossa tonsilaris
3. Epithelial reticulum of thymus and gl. parathyr. inf.
4. gl. parathyroidea sup. + ultimobranchial body
5. rudimentary



Defects caused by maldifferentiation of the pharyngeal apparatus

1. **Branchial (cervical) cysts**
2. **Branchial (cervical) fistulae**
3. Branchial (cervical) vestiges (rudiments of branchial arches)
4. Preauricular cysts a fistulae
5. **Syndrome of the 1. branchial arch**
6. **DiGeorge syndrome**
7. Ectopia of thymus

Branchial cysts (lateral neck cysts)

Origin from persisting sinus cervicalis, positioned under angulus mandibulae

Subcutaneously or deep around the pharynx (possibly larynx)

When a cyst ruptures, communication occurs with the body surface or pharynx

Lined with stratified squamous epithelium

They may contain a liquid content with cholesterol crystals

Usually clinically not important



Figure 1 Branchial cleft cyst in the neck

Branchial fistula (lateral cervical fistula)

Abnormal communication of the pharyngeal cavity with the body surface

They arise when the membranae obturantes obliterate

Between 2. pouch and cleft

(fossa tonsillaris - sternocleidomastoideus muscle)

Between 3. pouch and cleft

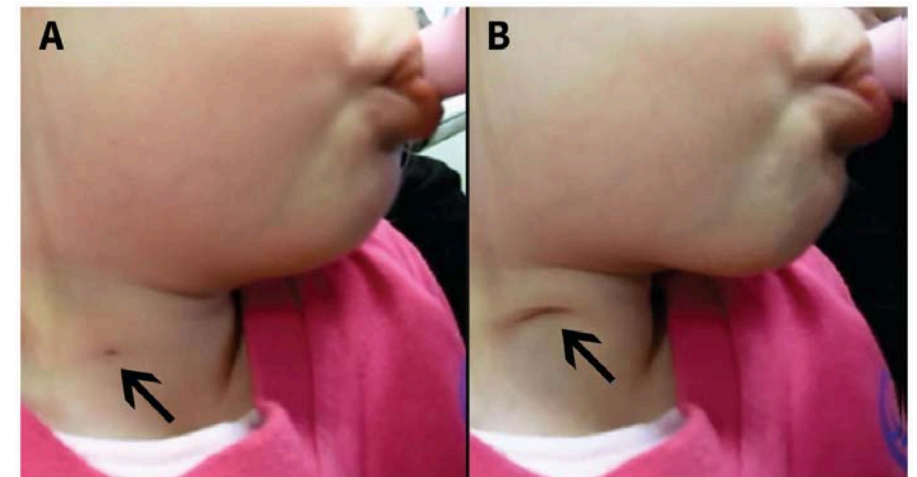
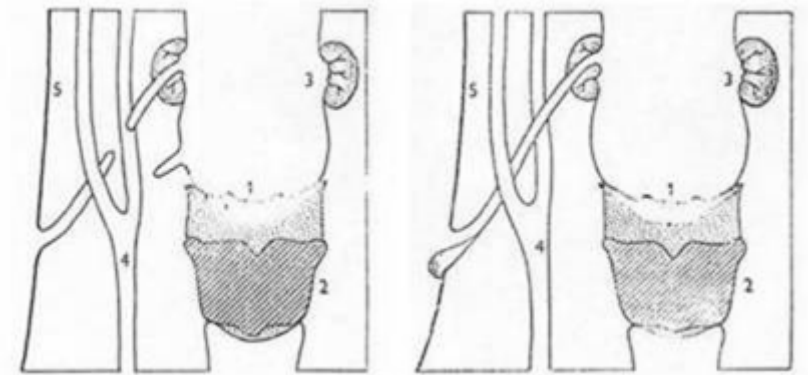
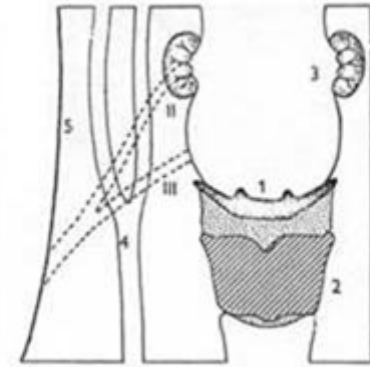
(tongue - art. sternoclavicularis)

Complete

at the outlet on the skin

Incomplete

external, internal



Branchial vestiges (rudiments of branchial arches)

Residues of some components of the pharyngeal arches, usually cartilage.

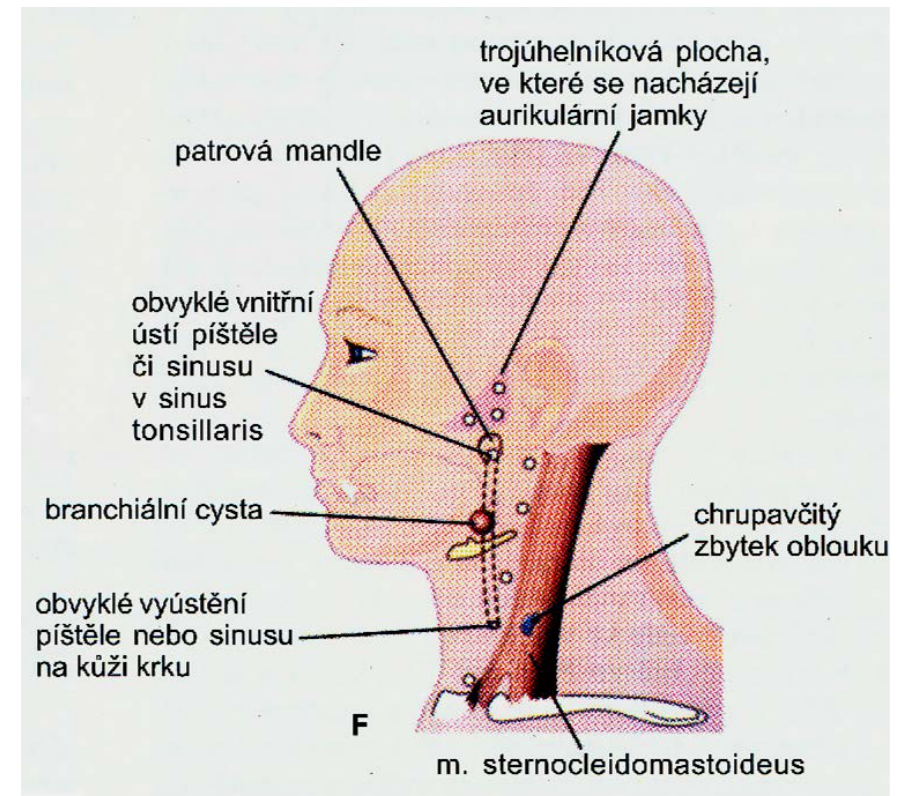
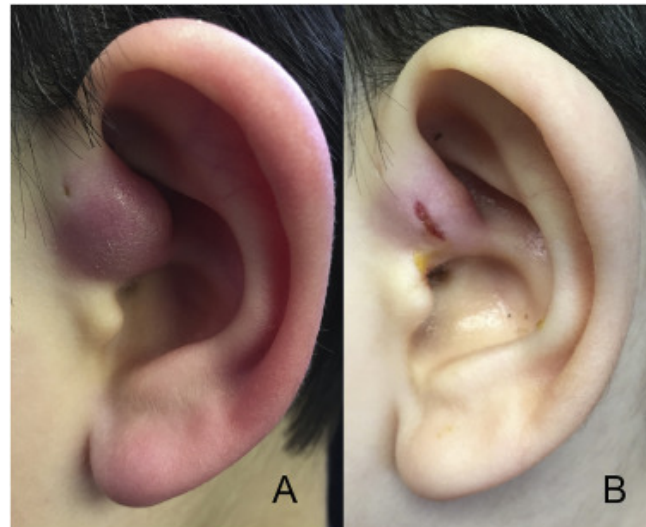
Occurrence: in the subcutaneous ligament of the neck above the lower 1/3 m.sternocleidomastoid

Rare

Preauricular cysts and fistulae

Small grooves, pits or cysts in skin in triangular area anteriorly to the pinna (auricle)

Origin: by persistence of sulci separating auricular hillocks



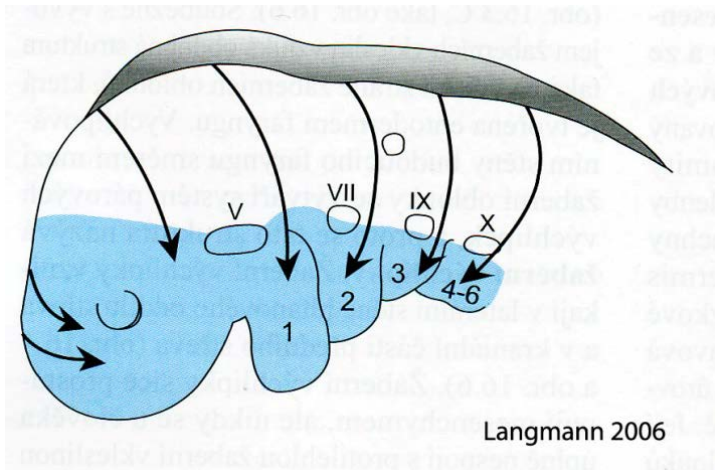
The First pharyngeal arch syndrome

Complex malformation of the skeleton of the face (both jaws, palate), eye and ear, **caused by delay or non-migration of crista neuralis** into the 1st pharyngeal arch

Types:

1) **Treacher-Collins syndrome** - dysostosis mandibulofacialis – autosomal dominant hereditary malformation

anatomically: hypoplasia to aplasia of zygomatic bones, hypoplasia of the upper and lower jaw, macrostomy, gothic floor, hypoplastic and sparse teeth, malocclusion - the face shows a characteristic physiognomy



2) Pierre-Robin syndrom

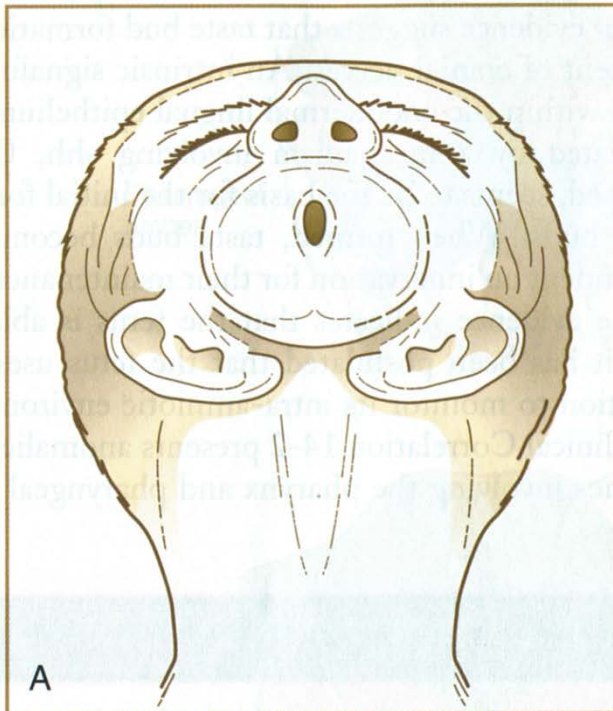
Hypoplasia of the mandible, gothic floor or posterior cleft palate, glossoptosis, ear defects

Autosomal recessive inheritance, X chromosome - linked

The intellect of individuals is not affected

Symptoms: due to the shortened base of the oral cavity, individuals after birth have difficulty feeding and breathing (stridor - caused by a disproportion between the lower jaw and the tongue)

Agnathia



DiGeorge syndrome

Incorrect development of the 1st pharyngeal arch. **Caused by improper migration of neural crest cells.**

Anatomically: hypoplasia of the mandible, shortened philtrum - nasal hypoplasia, congenital aplasia of the thymus and parathyroid glands, hypoplasia of the thyroid gland, defects of the heart and large vessels (right aortic arch), external ear defects

Clinically: hypoparathyroidism (hypocalcemic seizures), absence of cellular immunity, manifestations of heart defect

Incidence 1: 50 000

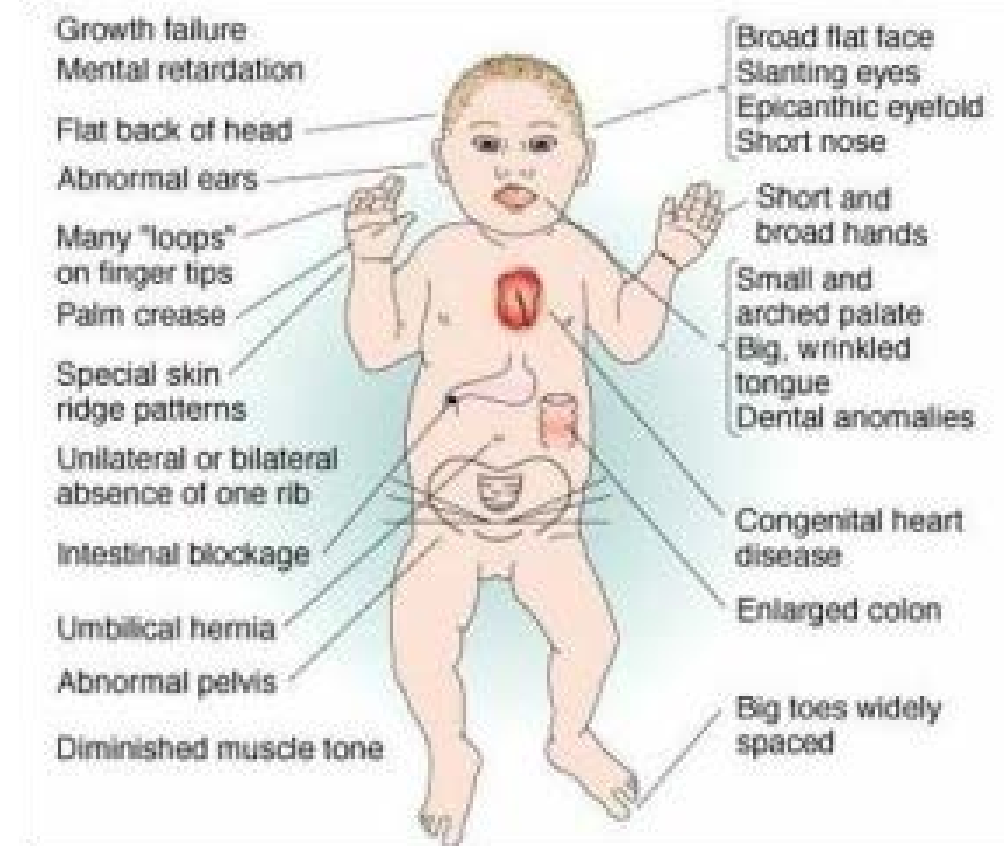
Etiology: Most frequently deletion on chromosome 22 - (22q11)

Thymus ectopia

Ectopia = correctly developed organ/structure in incorrect place

When thymus fails to descend: Cervical thymus - near the lower pair of parathyroid glands

Accessory thymus



Tongue development

The development of the tongue begins in the **5th week** at the interface of the stomodeum and the beginning of the primitive pharynx

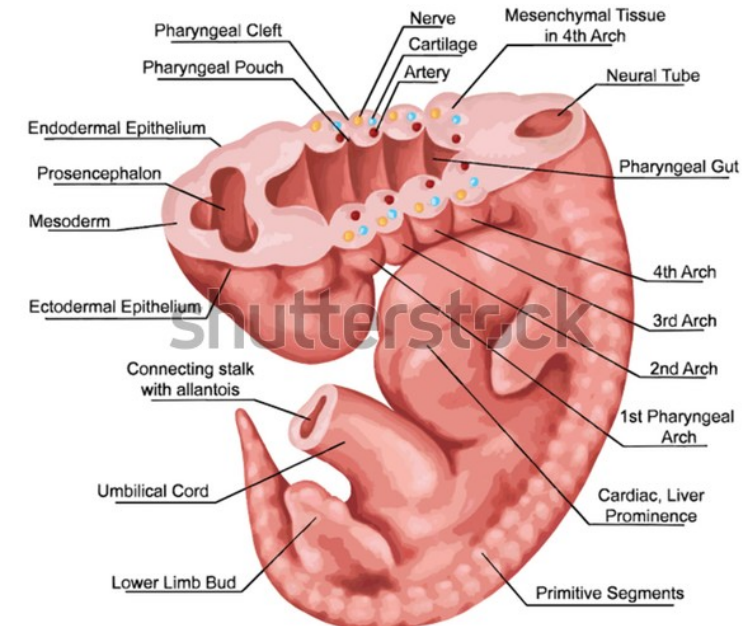
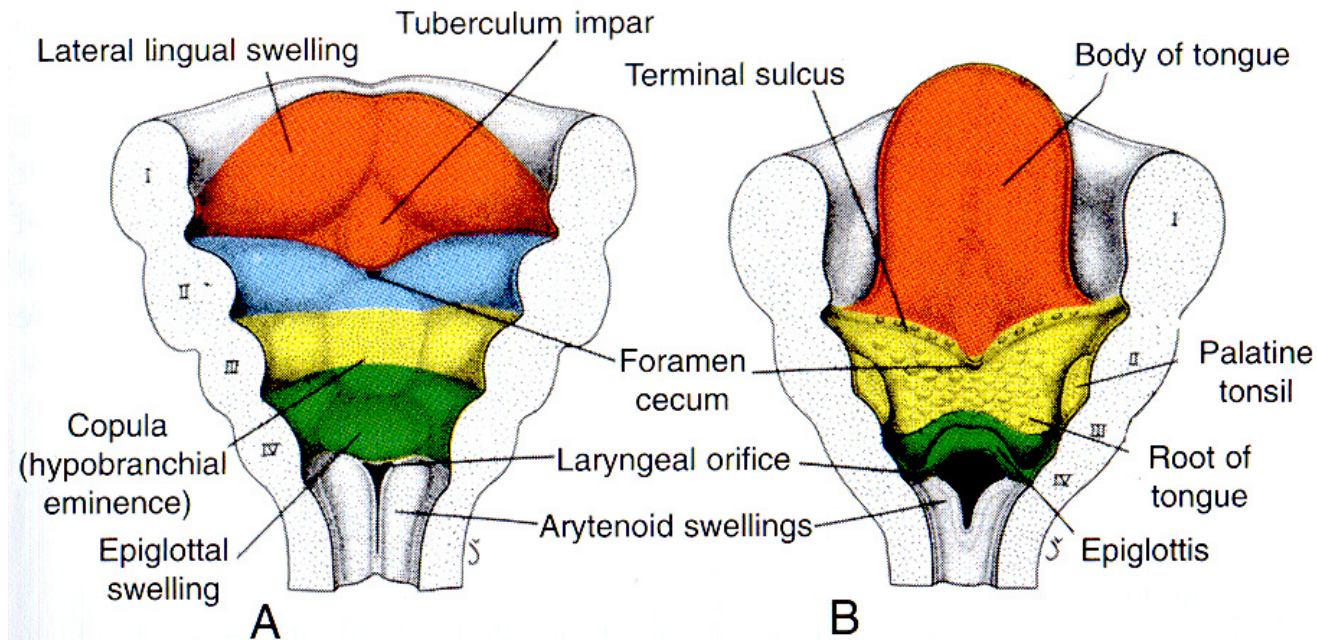
Anterior 2/3 of the tongue	Apex and corpus linguae	Formed from the mandibular process of the 1st pharyngeal arch
Posterior 1/3 of the tongue	Radix linguae	Formed from the 3rd and 4th pharyngeal arch

Apex and corpus

On the mandibular prominence are 3 mesenchymal protrusions covered with **ectoderm**:

Paired **tuberculum linguale laterale** (dx et sin) - distal lingual protrusion

Middle unpaired **tuberculum impar** (tuberculum linguale mediale) - middle tongue protrusion - more caudally



Radix linguae

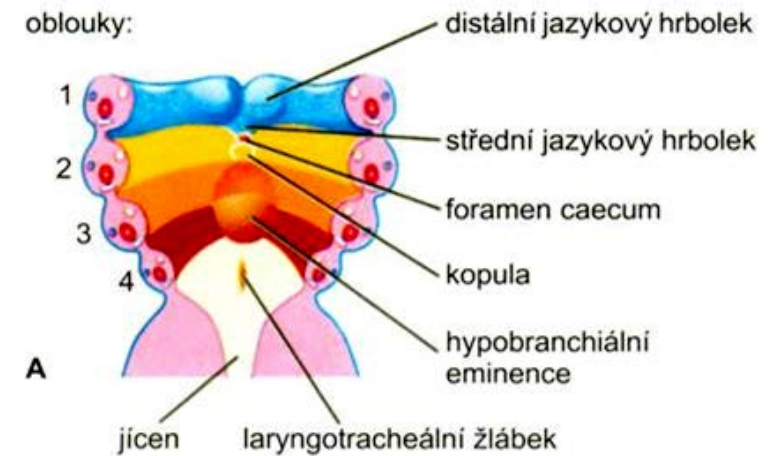
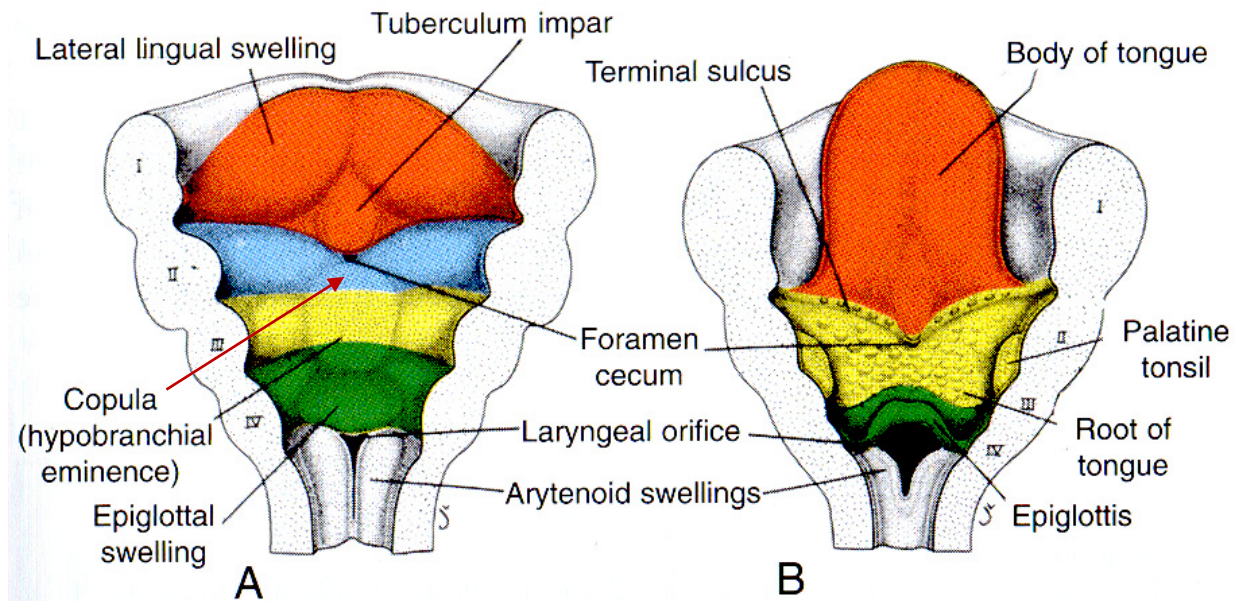
2 foundations:

copula - fused ectomezenchyme of the ventral ends of the hyoid arch

eminentia hypobranchialis - formed by fusion of ventral ends of 3rd and 4th pharyngeal arch

both the copula and the hypobranchial eminence are covered by the **endoderm**

Endoderm between the tuberculum impar and the dome very intensively proliferates and grows caudally, its luminization creates a ductus thyreoglossus (see thyroid gland)



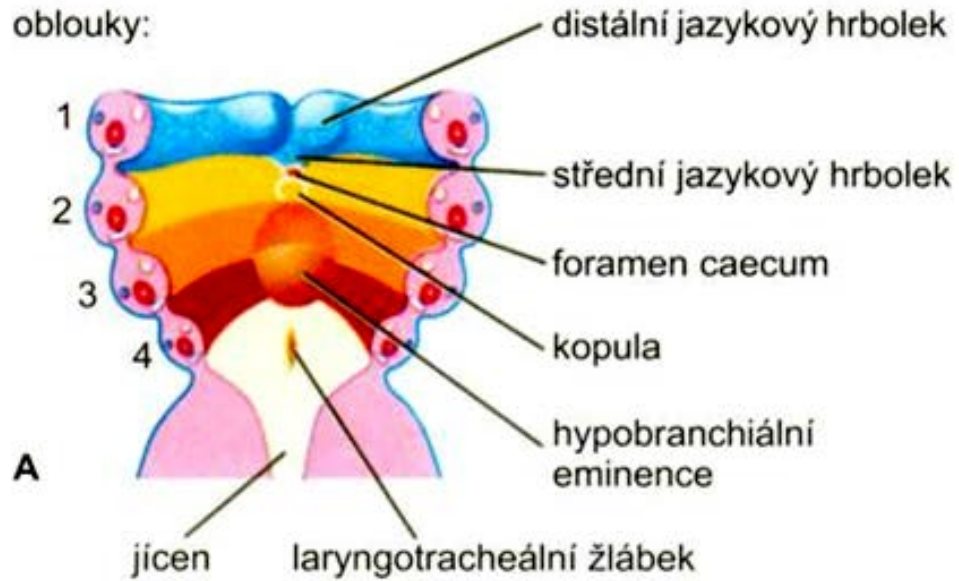
During the **6th week**, the protrusions begin to fuse together

Lateral protrusions enwrap the unpaired **tuberculum impar** - a uniform apex and corpus linguae is formed

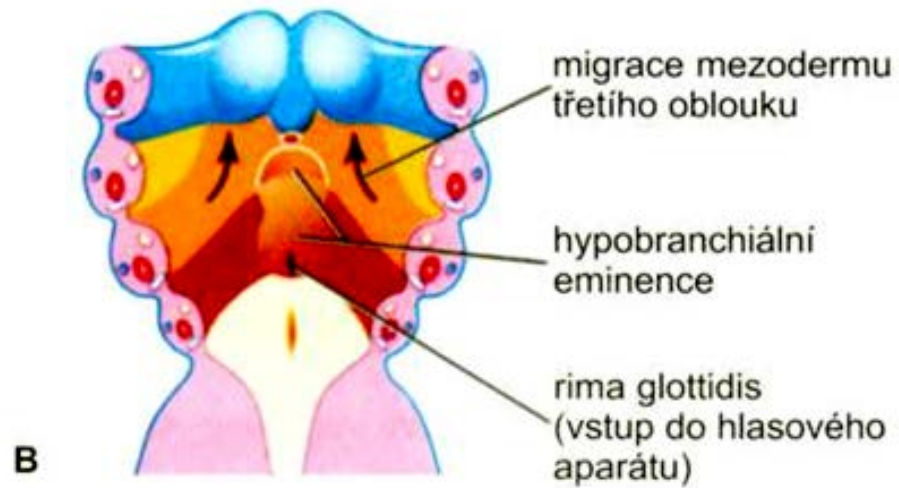
In definitive proportions, it resembles the original symmetrical origin of the tip and body of the tongue **sulcus medianus linguae** (+septum linguae)

Only a small part of the body near the root of the tongue comes from the tuberculum impar)

oblouky:



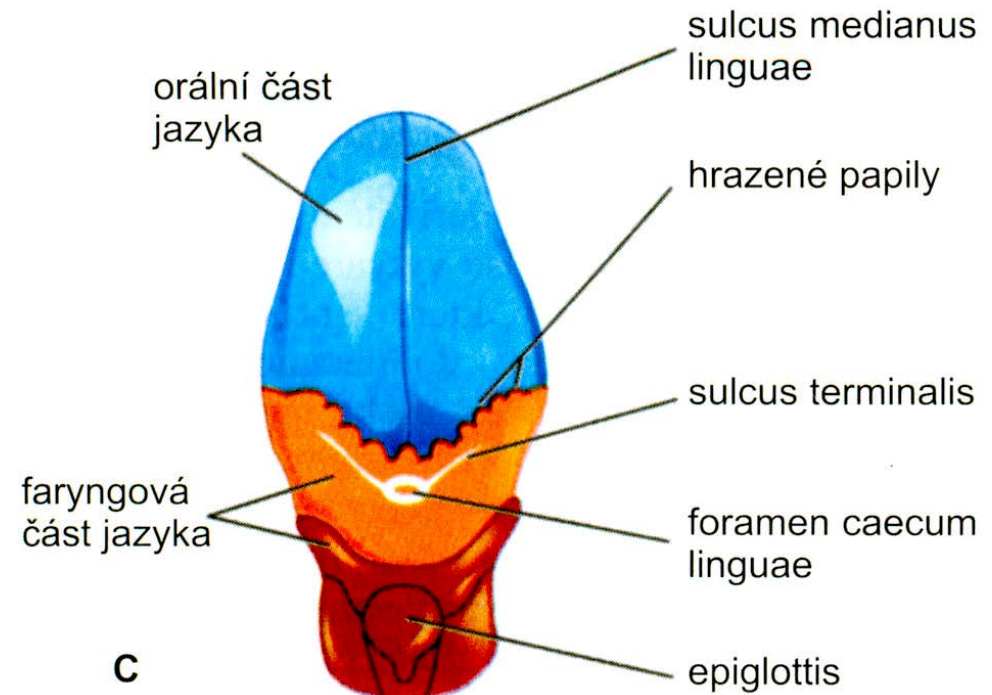
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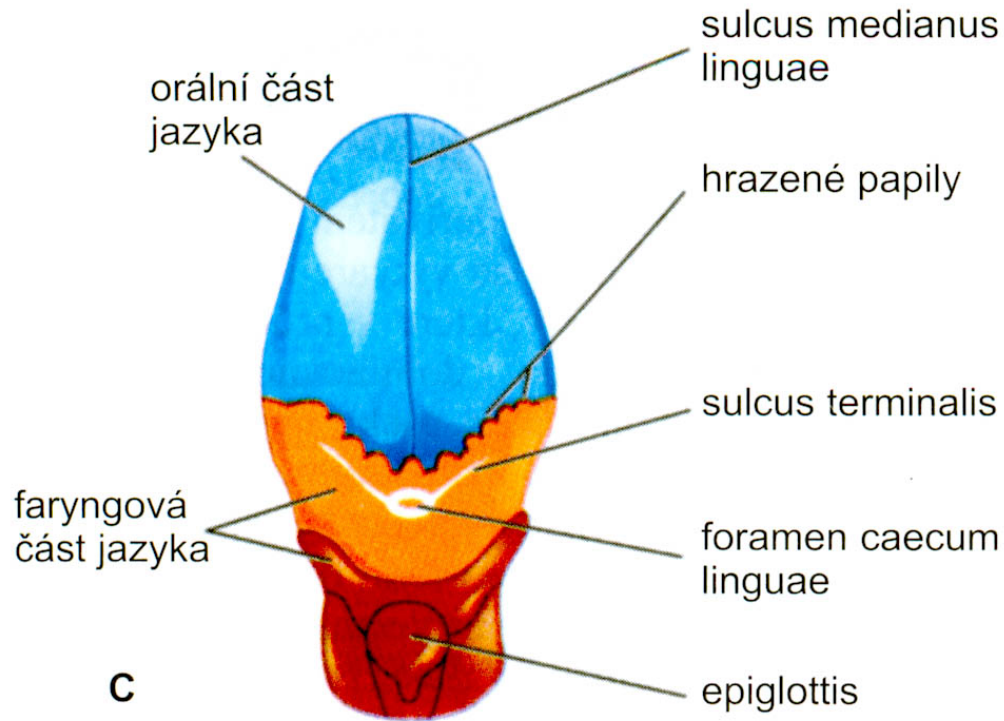
B

The hypobranchial process merges with copula and moves forward - approaching the base of the corpus with which it merges

Radix - Pharyngeal part of the tongue







C



The fusion line is visible until adulthood as a shallow "V" - shaped groove - **Sulcus terminalis**

At the top of the "V" is a short channel: **Foramen caecum**, remnant of the proximal end of the **ductus thyreoglossus**

Deriváty faryngových oblouků obsažené v jazyku

- | | |
|--|--|
|  1. faryngový oblouk
(CN V – ramus mandibularis) |  2. faryngový oblouk
(CN VII – chorda tympani) |
|  3. faryngový oblouk
(CN IX – glossopharyngeus) |  4. faryngový oblouk
(CN X – vagus) |

Tongue development

The ectoderm and entoderm of the common base of the tongue differentiate into stratified squamous epithelium, taste bud cells, and secretory compartments and ducts of the tongue glands

From ectomesenchyme of fused protrusions, the ligament of the tongue, blood and lymph vessels develop, incl. lymphatic tissue of the root of the tongue

Muscles of the tongue come from the occipital myotoms, which move to its base and merge together.

During the fusion of myotomes, their motor nerves also merge (segmental arrangement) - the **hypoglossus nerve** is formed

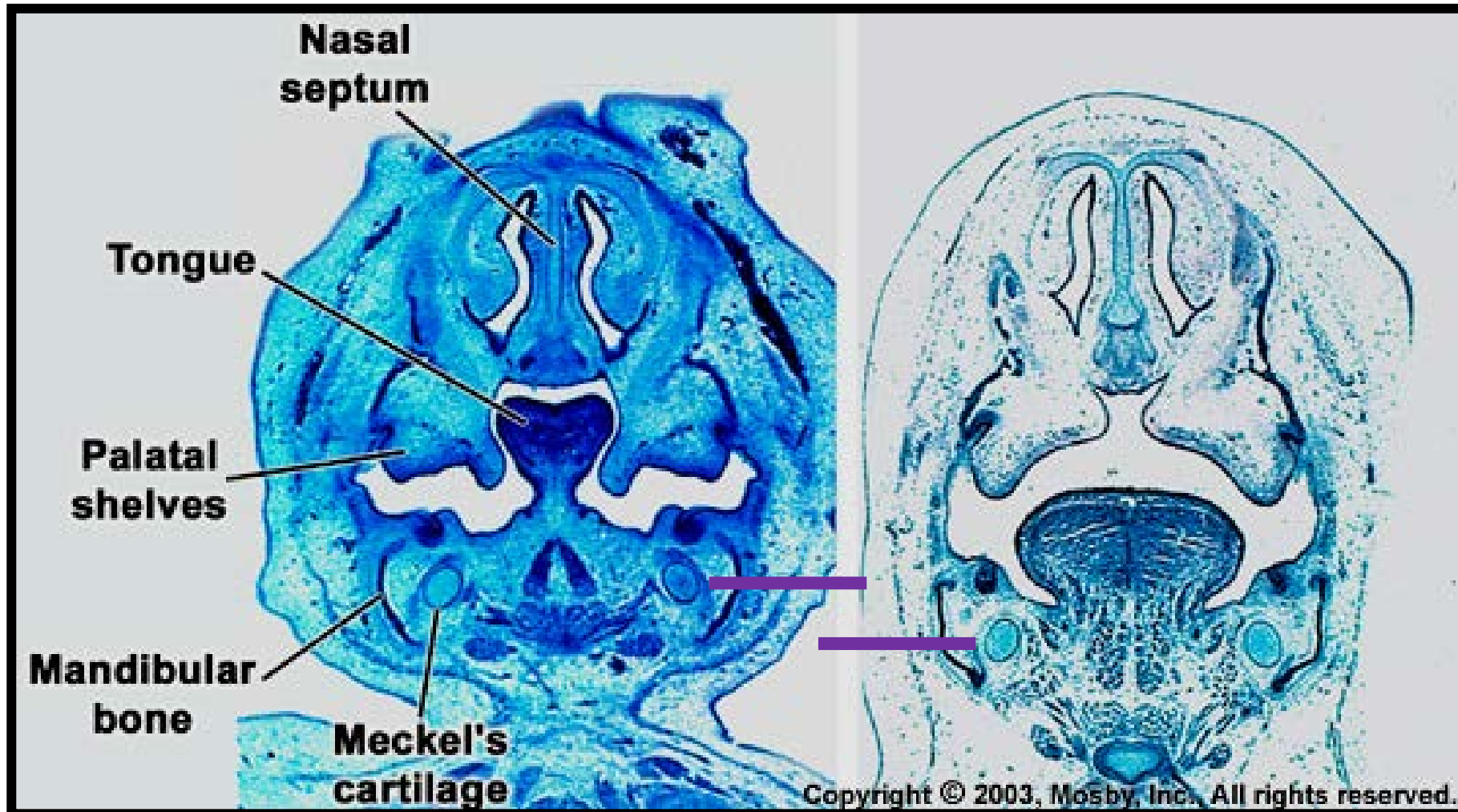
Development of tongue papillae - in the 8th week – firstly papillae vallatae, foliatae (near the branches of the n. IX.), fungiformes (branches of the n. Lingualis), filiformes (the 11th-12th week)

Taste buds - weeks 11-13

Sensitive innervation:
Apex and corpus - trigeminal nerve (n. mandibularis)
Radix - n. Glossopharyngeus

Innervation of taste buds:

- Taste buds in papillae fungiformes - **n. facialis** - chorda tympani
- Taste buds in papillae foliatae and circumvallatae - **n. glossopharyngeus**
- Taste buds in another location (radix linguae, isthmus faucium) - **n. vagus**



At birth: the tongue occupies the oral cavity

Postnatally: the root of the tongue descends into the pharynx – process finished at the 4th year of life

Overview of tongue development defects

Ankyloglossia (lingua accreta) - short frenulum, limited mobility of the tip of the tongue, it is not possible to stick out the tongue (difficulty breastfeeding), 1: 300 births. The frenulum usually lengthens spontaneously (surgery is not needed)

Congenital lingual cysts and fistulas - persistence of ductus thyreoglossus – clinically usually non important, causes problems only when enlarged (discomfort in the pharynx or dysphagia)

Macroglossia - a rare, abnormally large tongue (associated with some syndromes, e.g. Down sy.)

Microglossia - a rare, abnormally small tongue (mostly associated with micrognathia; microglossia in combination with limb defects - Hanhart's syndrome)

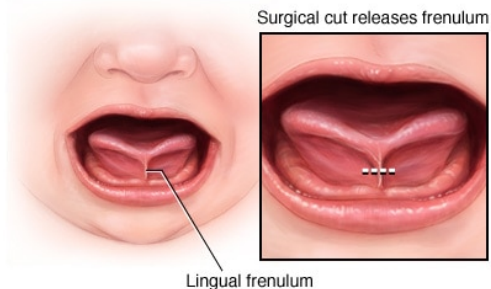
Glossoptosis - displacement of the tongue dorsally. Pushes on the epiglottis, narrowing of the pharynx.

Lingua bifida (lingua fissa, glossoschisis) - a very rare anomaly, incomplete fusion of the tubercula lingualia lateralia

complete cleft - including the tip of the tongue (associated with the cleft of the lower lip and jaw)

partial cleft - deep longitudinal groove (groove) in the body of the tongue

Aglossia – tongue not developed



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Development of salivary glands

Salivary glands as derivatives of the lining of the stomodea or other structures: the oral side of the palate, the tip (ectoderm) and the root of tongue and the oral base (entoderm)

ectoderm: small salivary glands of lips and face, palate, gl. apicis lingue and parotid gland

entoderm: Weber's and Ebner's glands of the tongue, gl. submandibularis and gl. sublingualis

They all develop in a similar way:

From the epithelium (ecto- or entoderm) at the site of the future gland(s): cells begin to proliferate against adjacent ectomezenchyme

They lengthen and branch - the basis for the glandular duct system is created, the last 6th generation form **terminal branches**

