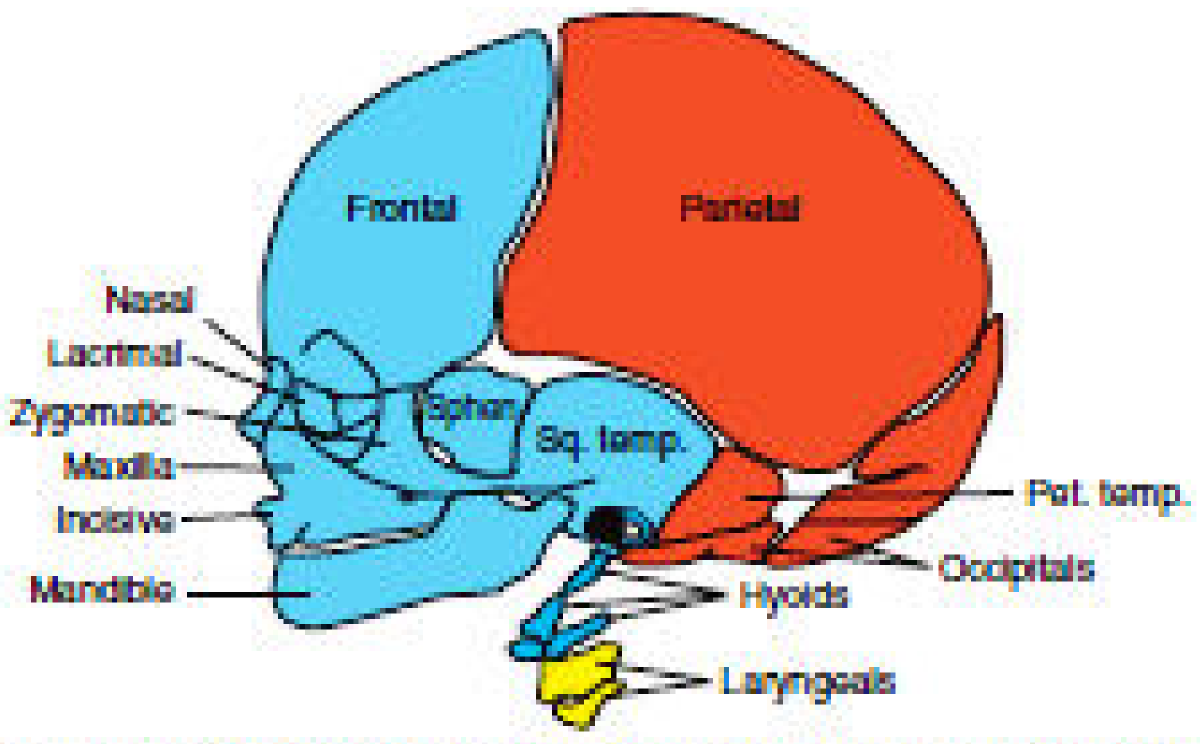
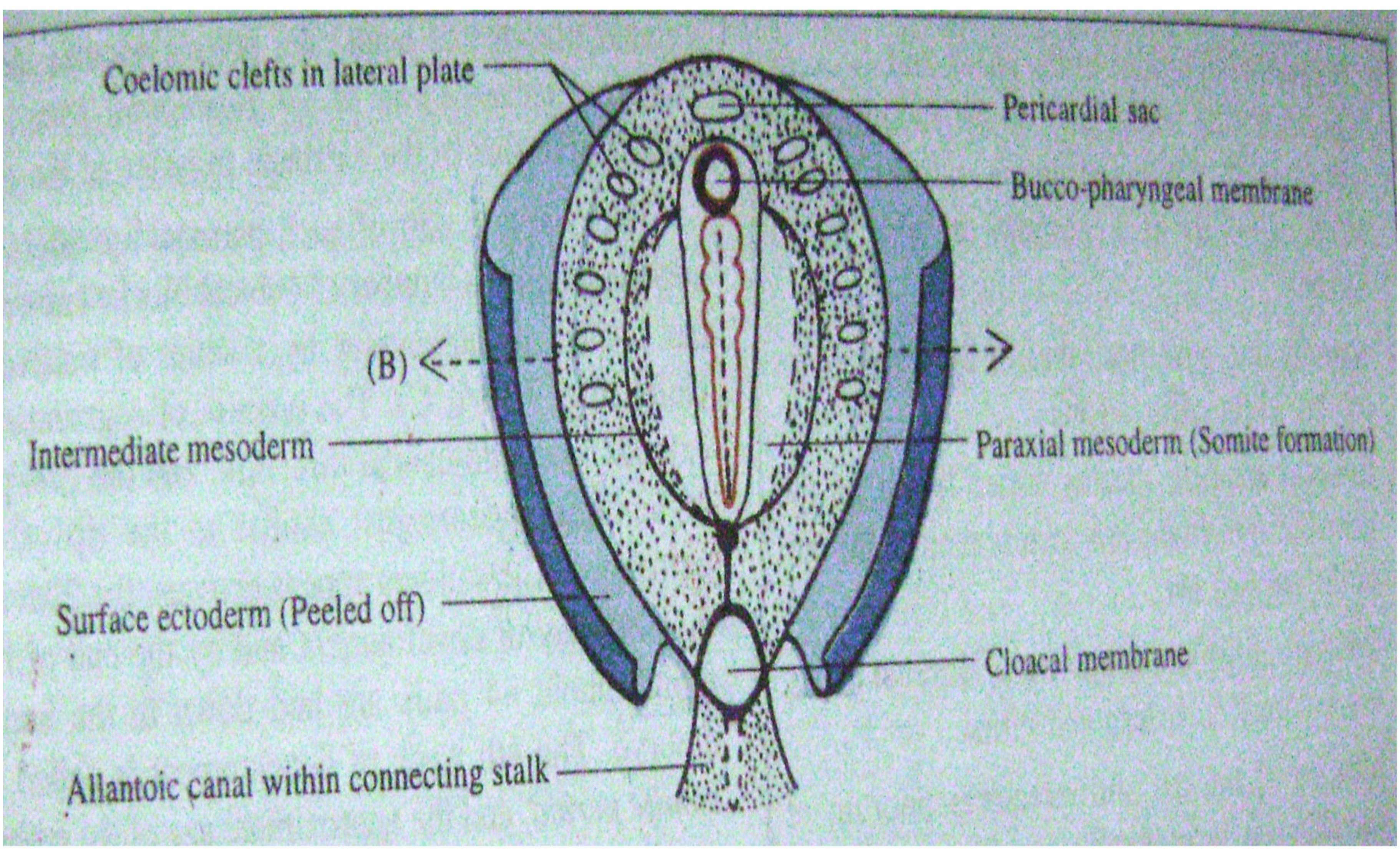
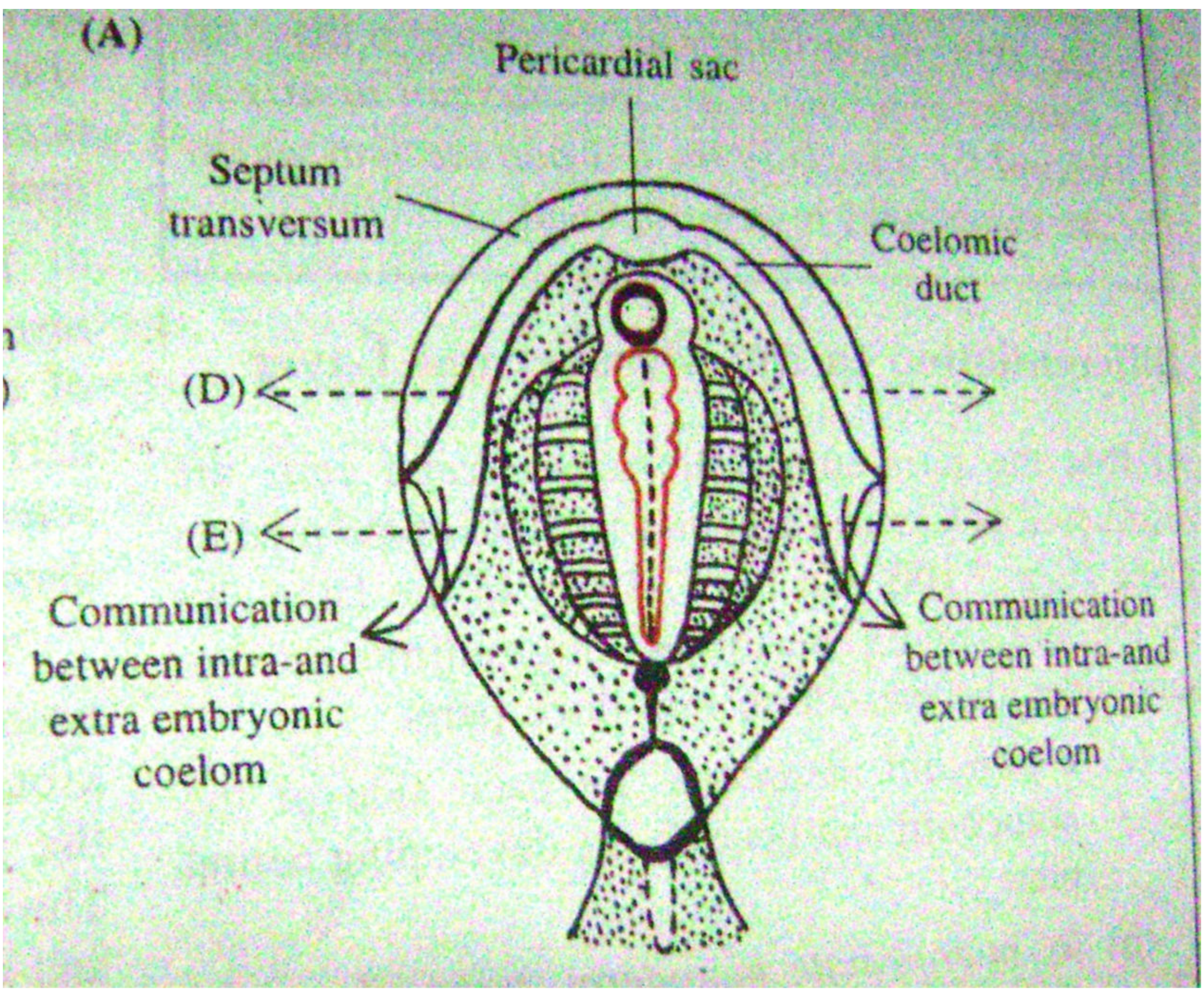
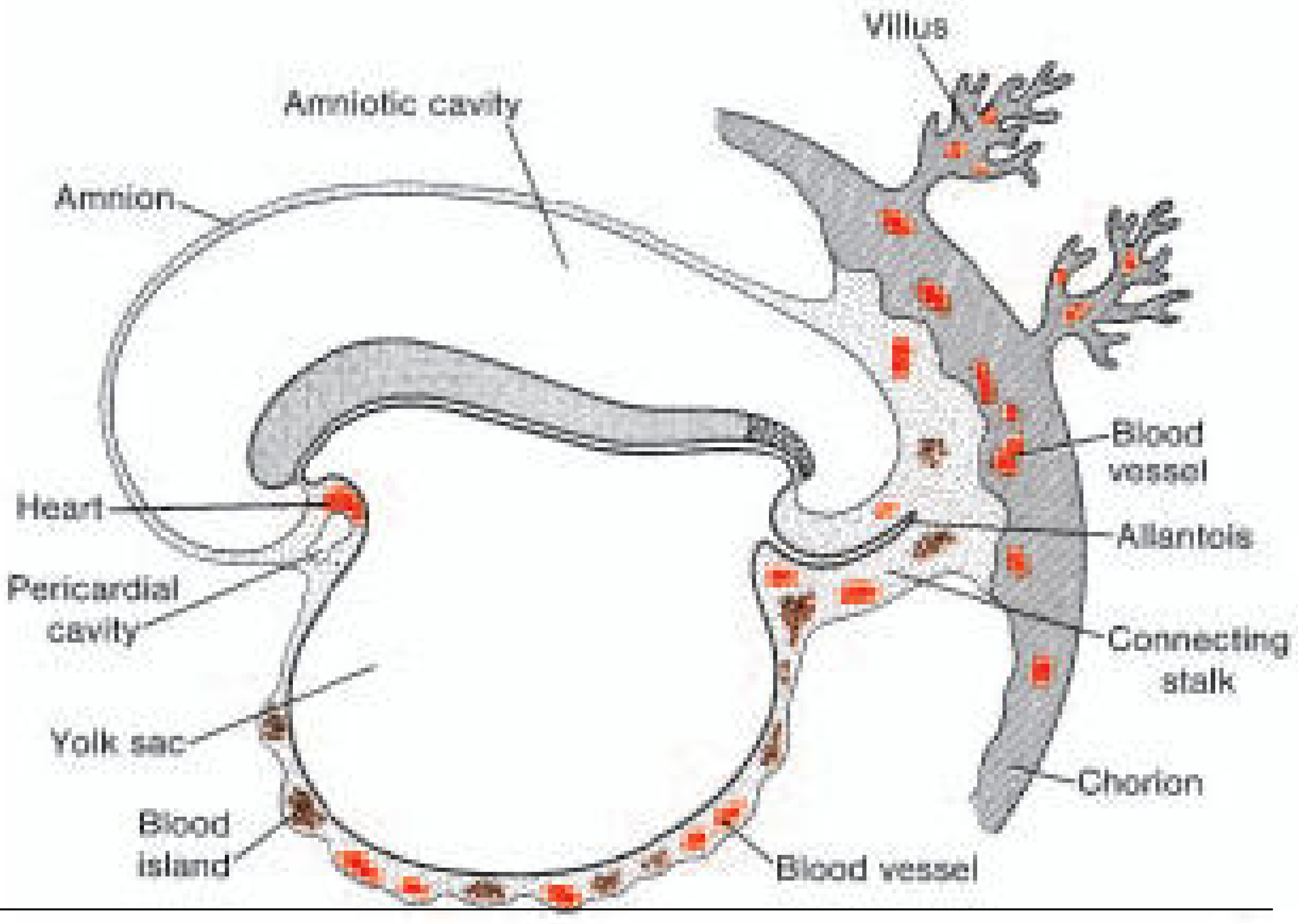
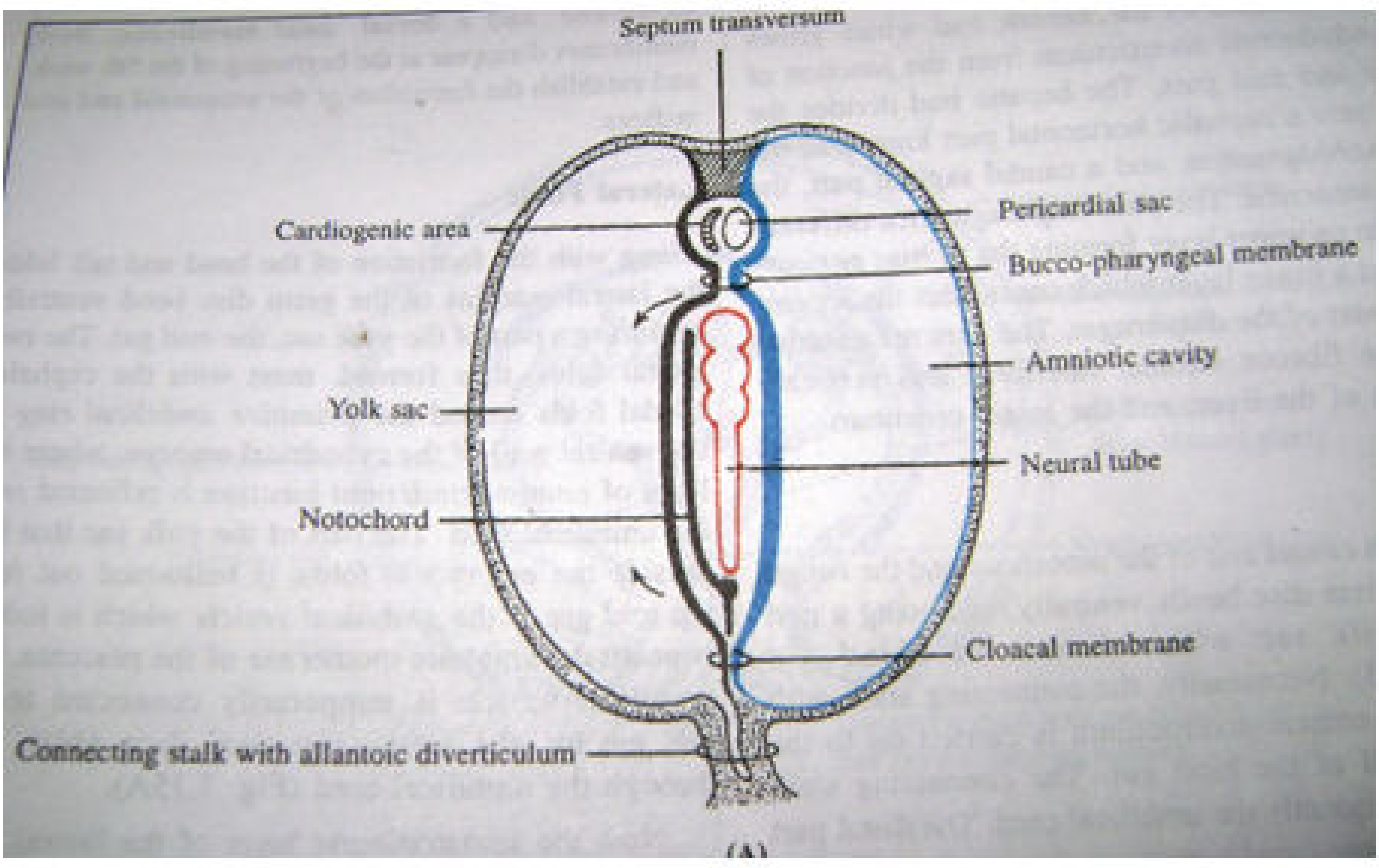


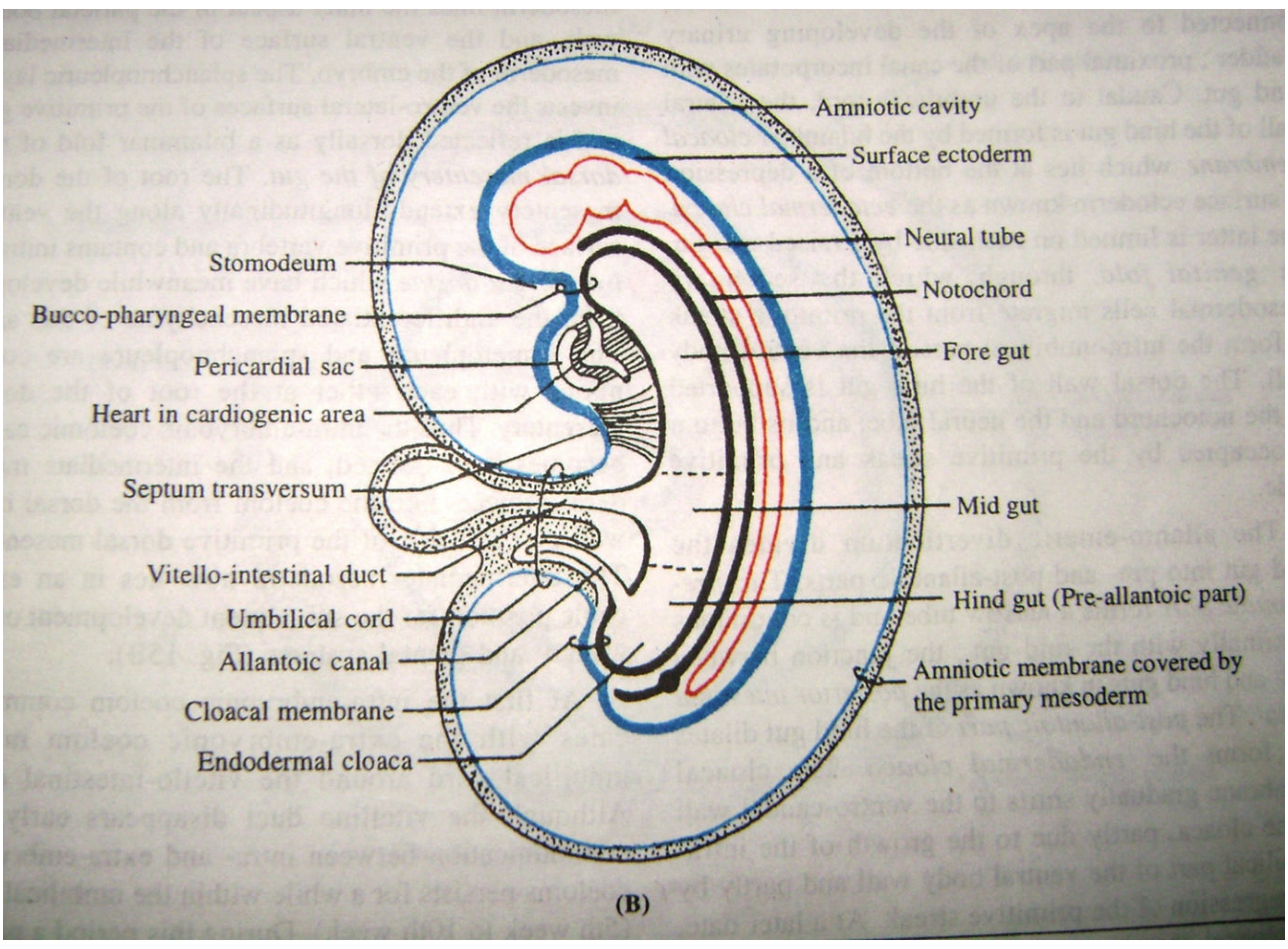
Figure 17.1 Skeletal structures of the head and face. Mesenchyme for these structures is derived from neural crest (blue), lateral plate mesoderm (yellow), and paraxial mesoderm (somites and somitomeres) (red).

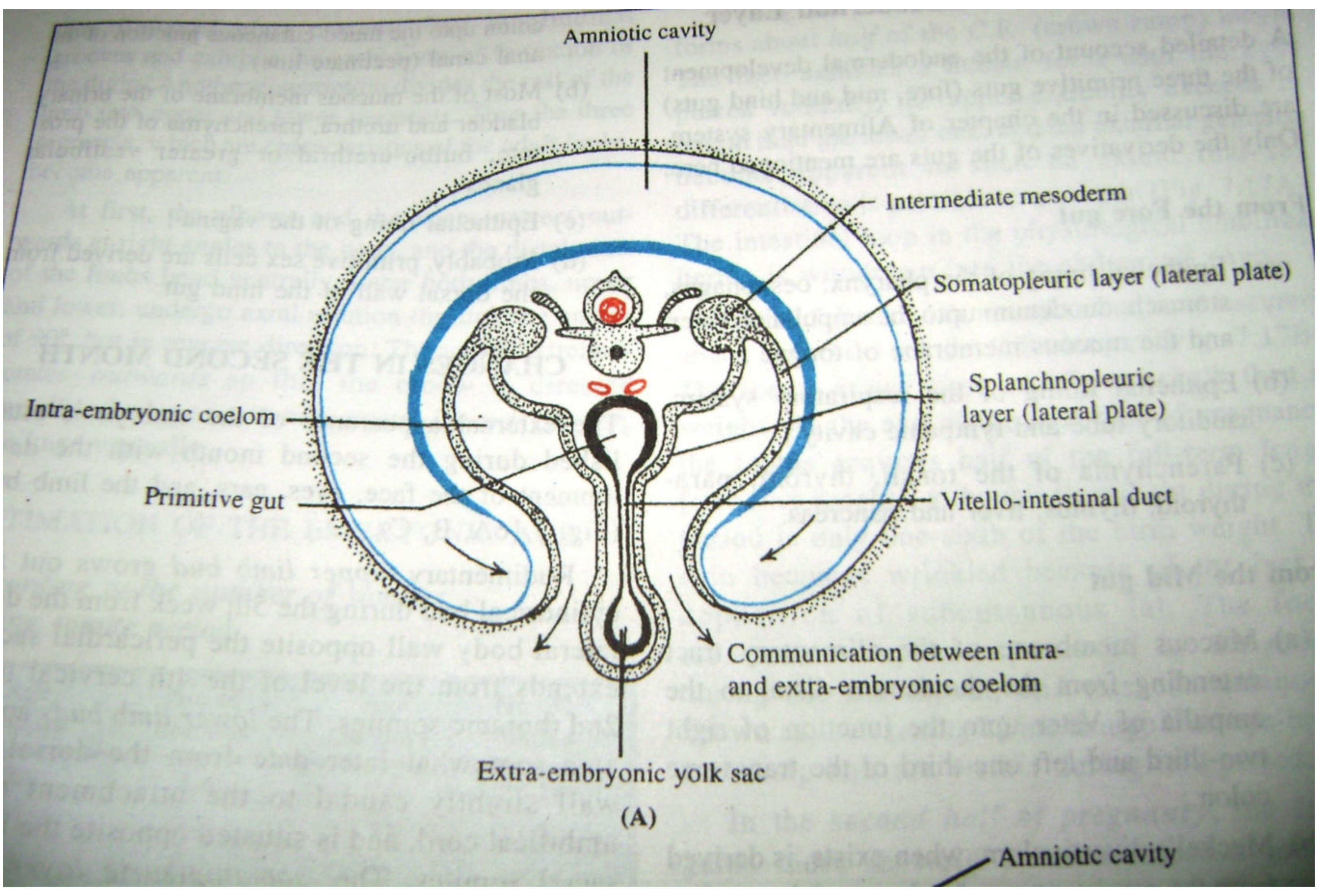












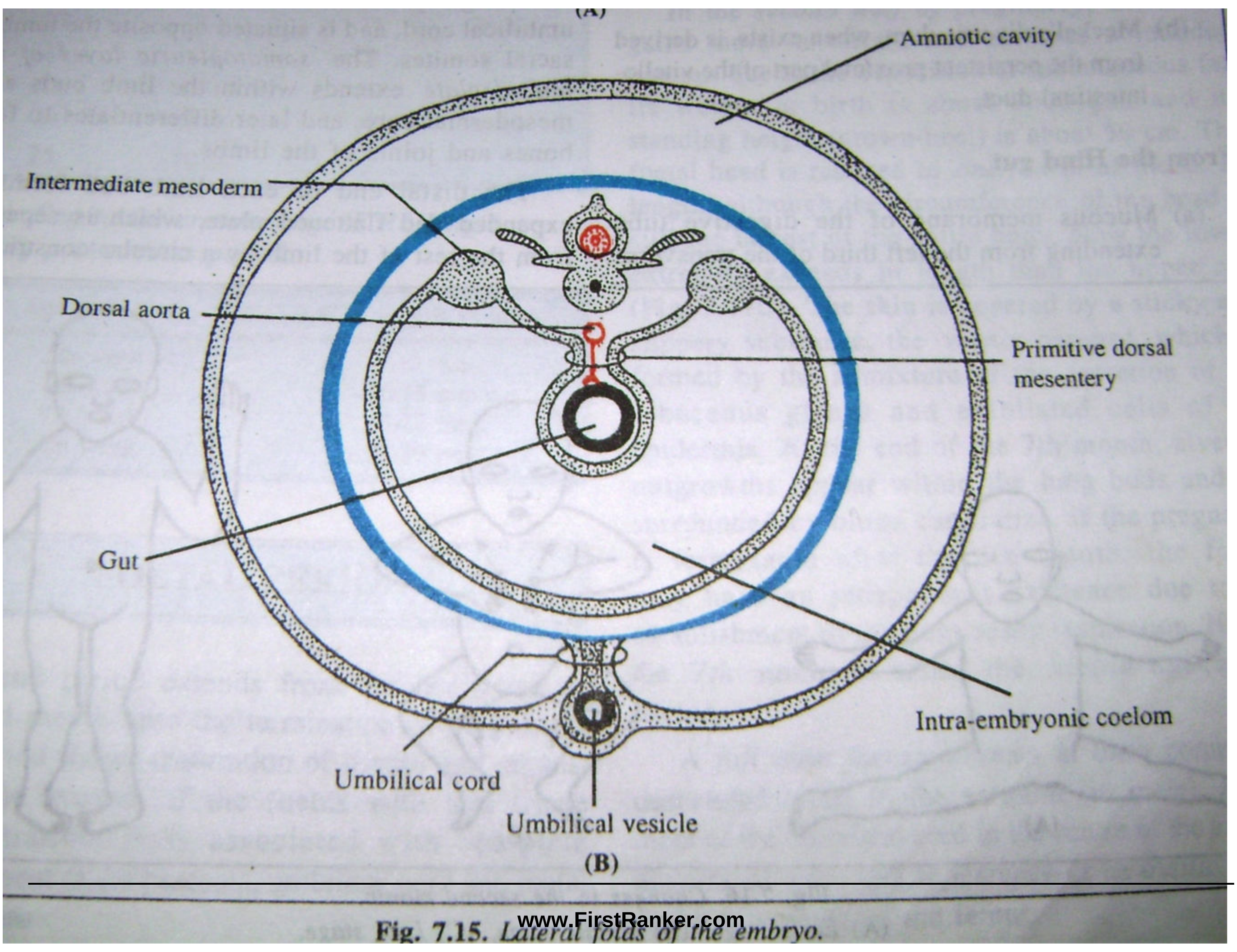
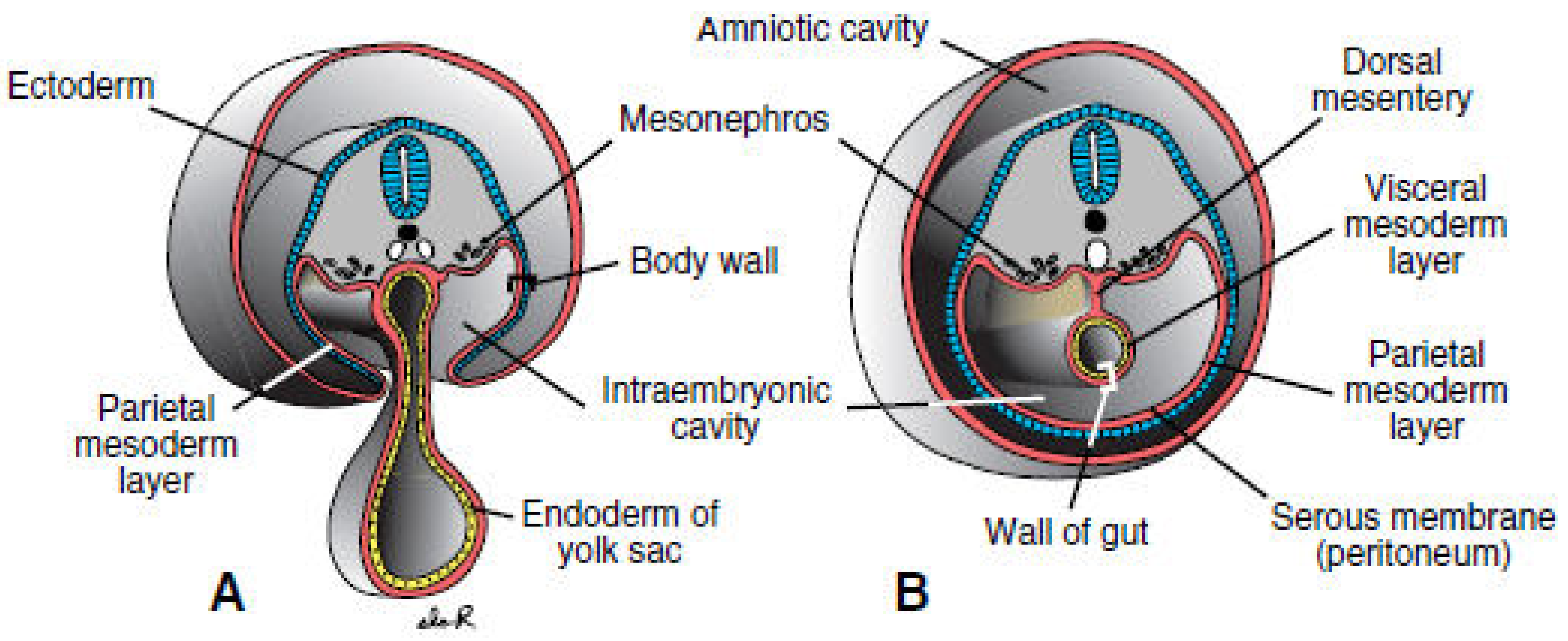
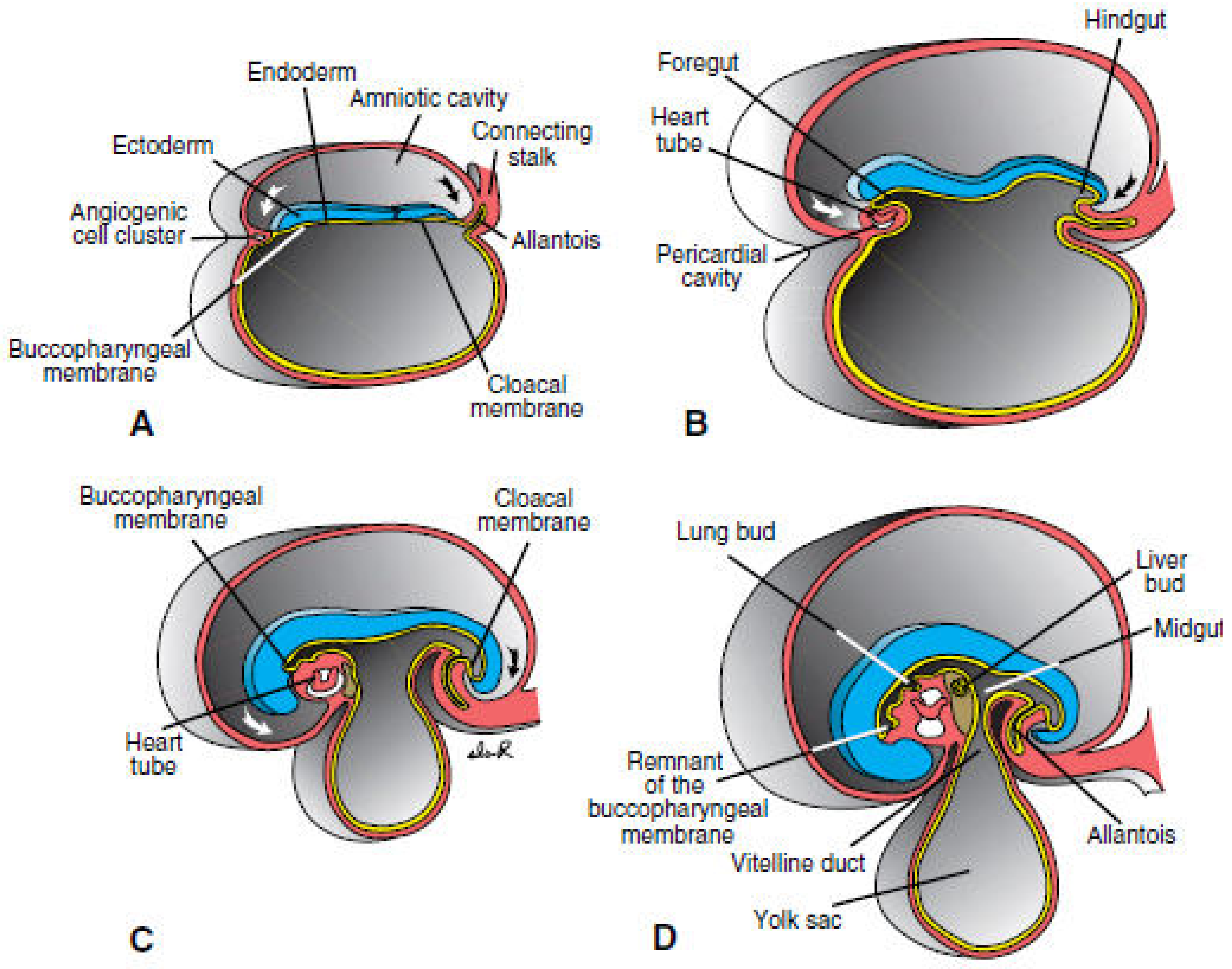
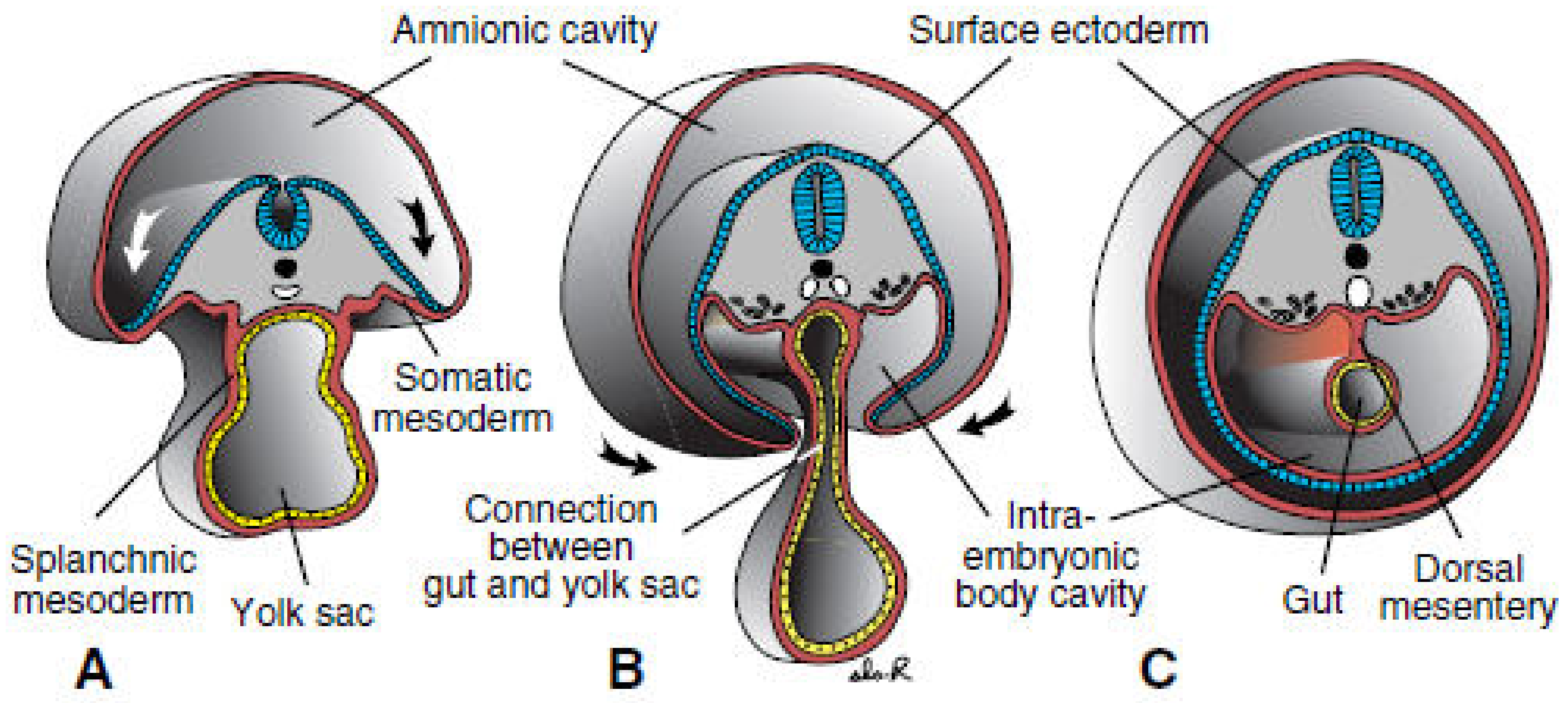


Fig. 7.15. Lateral folds of the embryo.







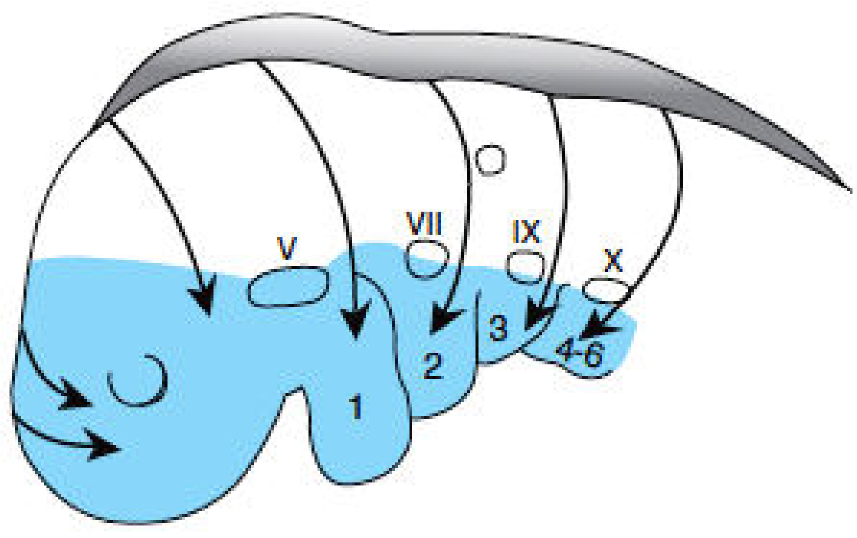


Figure 17.2 Migration pathways of neural crest cells from forebrain, midbrain, and hindbrain regions into their final locations (*shaded areas*) in the pharyngeal arches and face. Regions of ectodermal thickenings (placodes), which will assist crest cells in formation of the fifth (*V*), seventh (*VII*), ninth (*IX*), and tenth (*X*) cranial sensory ganglia, are also illustrated.

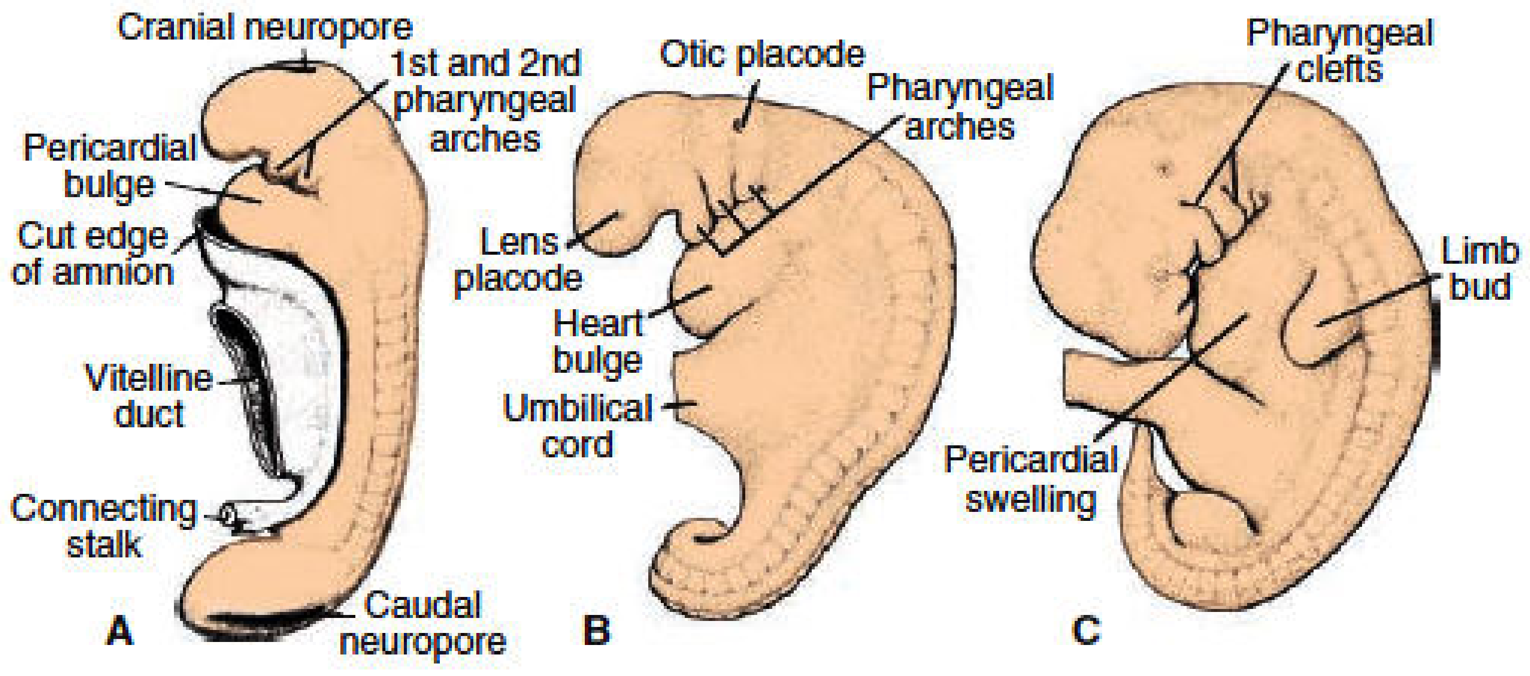


Figure 17.3 Development of the pharyngeal arches. **A.** 25 days. **B.** 28 days. **C.** 5 weeks.

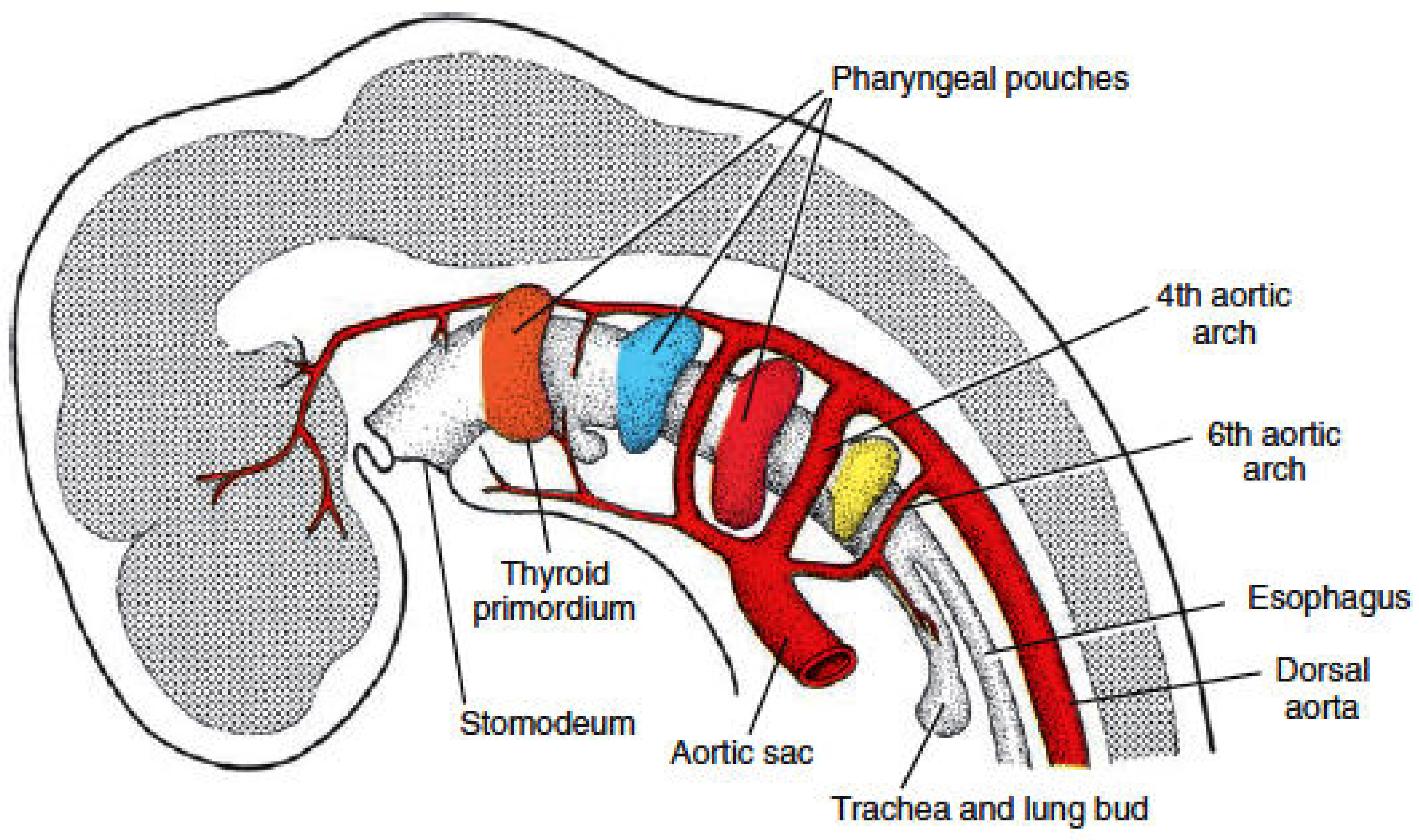


Figure 17.4 Pharyngeal pouches as outpocketings of the foregut and the primordium of the thyroid gland and aortic arches.

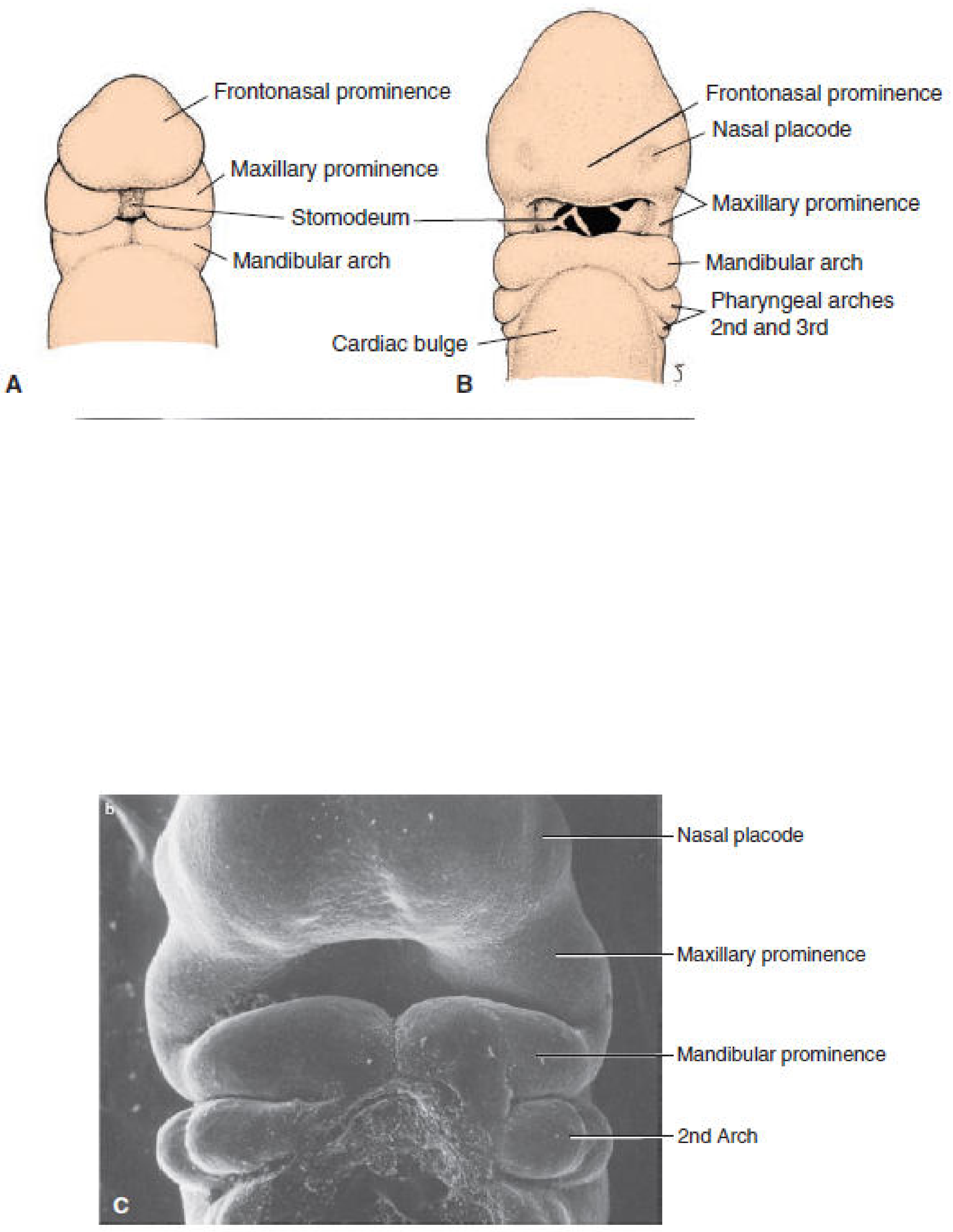
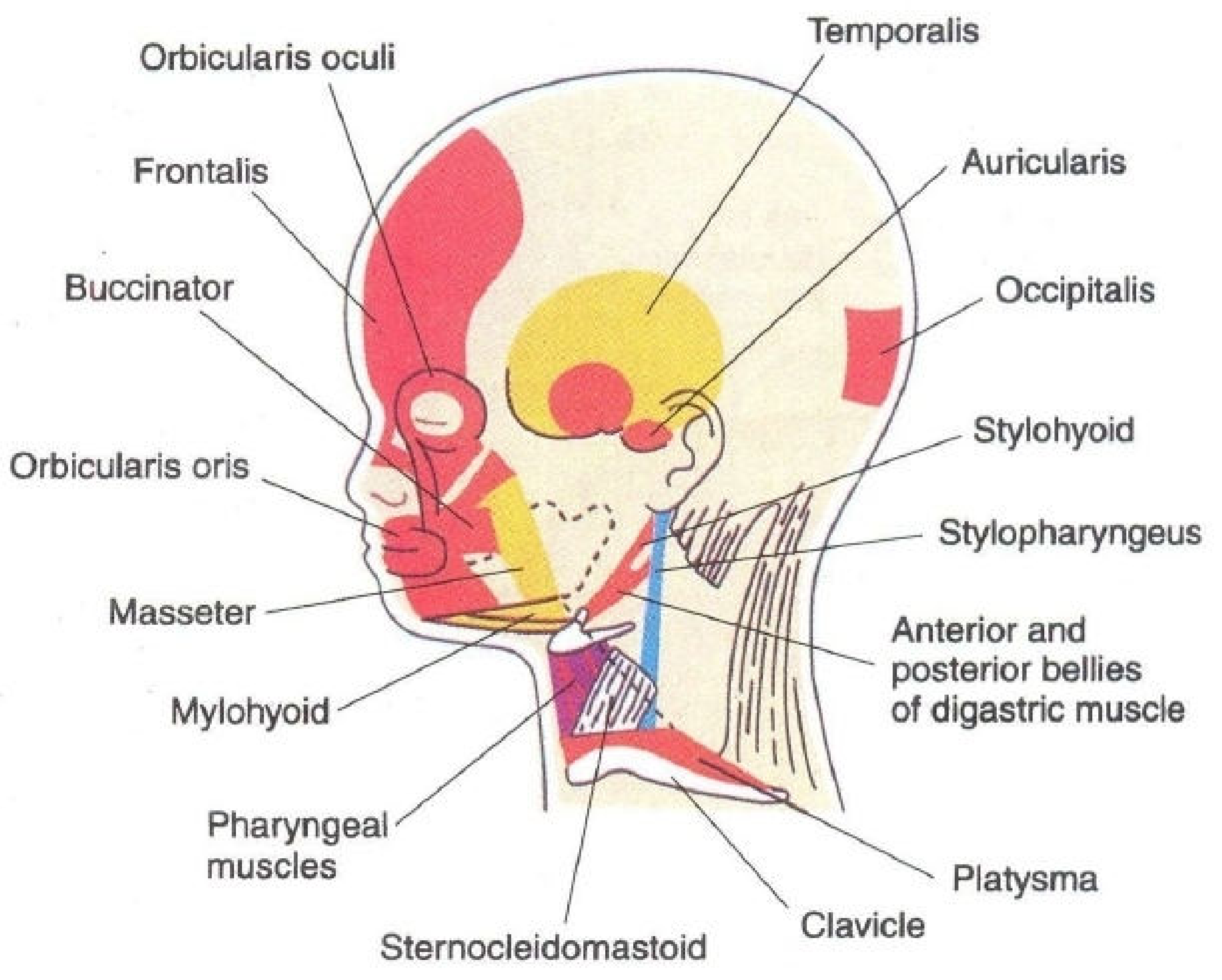


Figure 17.5 **A.** Frontal view of an embryo of approximately 24 days. The stomodeum, temporarily closed by the oropharyngeal membrane, is surrounded by five mesenchymal prominences. **B.** Frontal view of a slightly older embryo showing rupture of the oropharyngeal membrane and formation of the nasal placodes on the frontonasal prominence. **C.** Scanning electron micrograph of a human embryo similar to that shown in **B.**

TABLE 17.1 *Derivatives of the Pharyngeal Arches and Their Innervation*

Pharyngeal Arch	Nerve	Muscles	Skeleton
1. Mandibular (maxillary and mandibular processes)	V. Trigeminal: maxillary and mandibular divisions	Mastication (temporal; masseter, medial, lateral pterygoids); mylohyoid, anterior belly of digastric, tensor palatine, tensor tympani	Premaxilla, maxilla, zygomatic bone, part of temporal bone, Meckel's cartilage, mandible malleus, incus, anterior ligament of malleus, sphenomandibular ligament
2. Hyoid	VII. Facial	Facial expression (buccinator, auricularis, frontalis, platysma, orbicularis oris, orbicularis oculi) posterior belly of digastric, stylohyoid, stapedius	Stapes, styloid process, stylohyoid ligament, lesser horn and upper portion of body of hyoid bone
3.	IX. Glossopharyngeal	Stylopharyngeus	Greater horn and lower portion of body of hyoid bone
4–6	X. Vagus • Superior laryngeal branch (nerve to fourth arch) • Recurrent laryngeal branch (nerve to sixth arch)	Cricothyroid, levator palatine, constrictors of pharynx Intrinsic muscles of larynx	Laryngeal cartilages (thyroid, cricoid, arytenoid, corniculate, cuneiform)

which of the following are associated with the 2nd pharyngeal arch?
a) the malleus bone
b) facial nerve
c) glossopharyngeal nerve
d) the lower portion of the hyoid bone



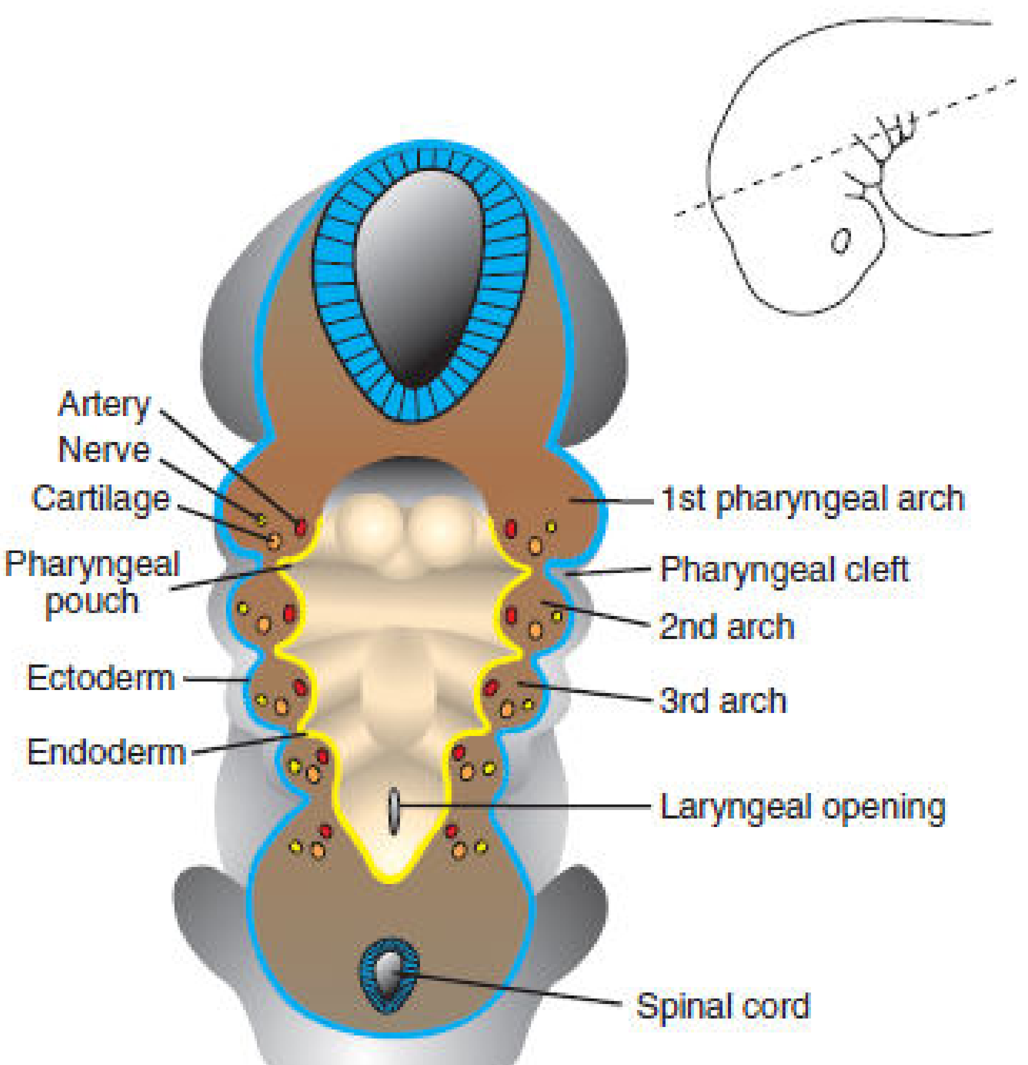


Figure 17.6 Drawing shows the pharyngeal arches cut in cross section. Each arch consists of a mesenchymal core derived from mesoderm and neural crest cells and each is lined internally by endoderm and externally by ectoderm. Each arch also contains an artery (one of the aortic arches) and a cranial nerve and each will contribute specific skeletal and muscular components to the head and neck. Between the arches are pouches on the inner surface and clefts externally.

. Branchial cysts or lateral cysts:
a) are found along the anterior border of the sternocleidomastoid muscle
b) are formed from a rupture of the membrane between pharyngeal pouches and branchial clefts
c) are remnants of the thyroglossal duct
d) are found in front of the ear

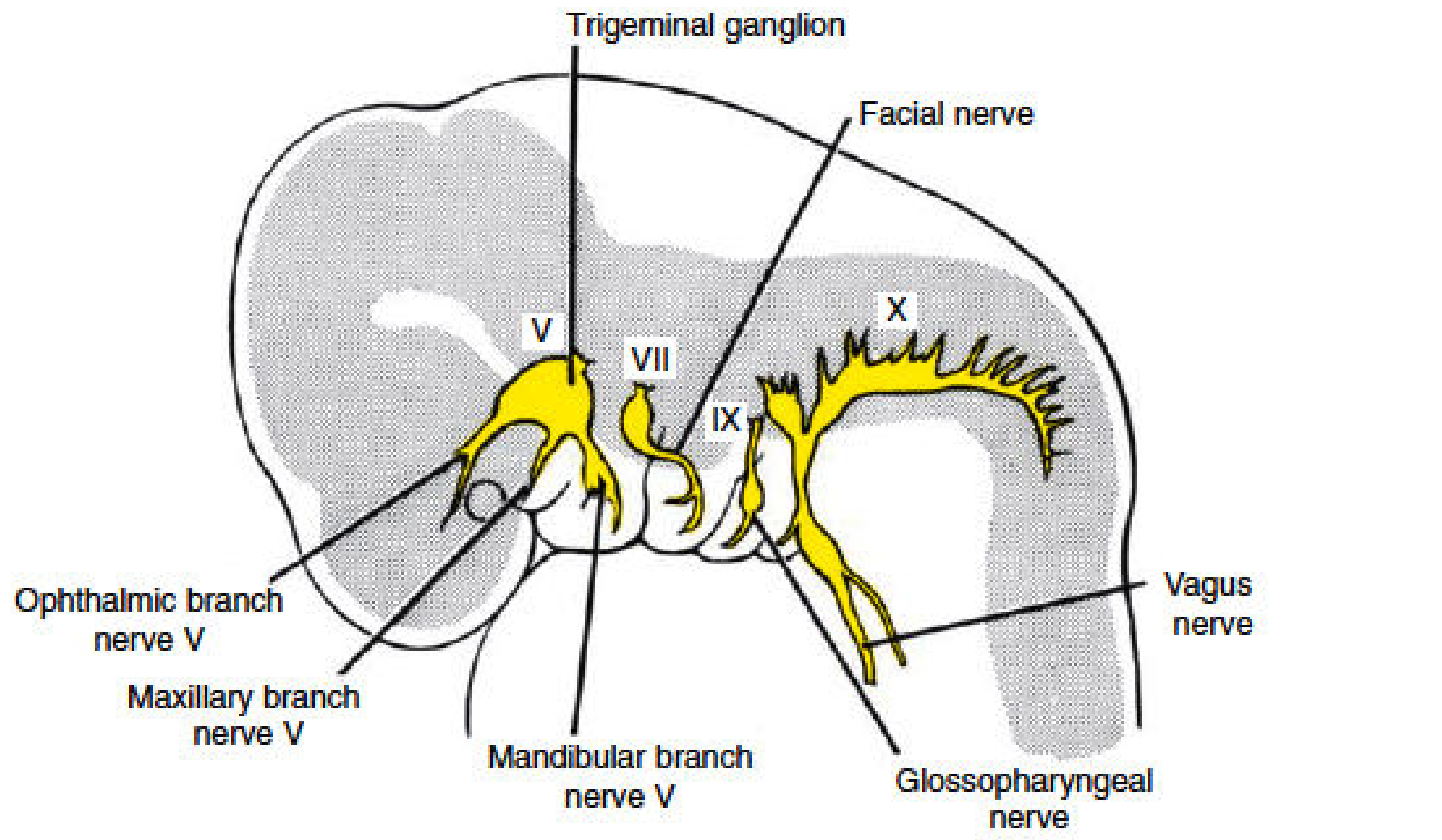


Figure 17.7 Each pharyngeal arch is supplied by its own cranial nerve. The trigeminal nerve supplying the first pharyngeal arch has three branches: the ophthalmic, maxillary, and mandibular. The nerve of the second arch is the facial nerve; that of the third is the glossopharyngeal nerve. The musculature 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.

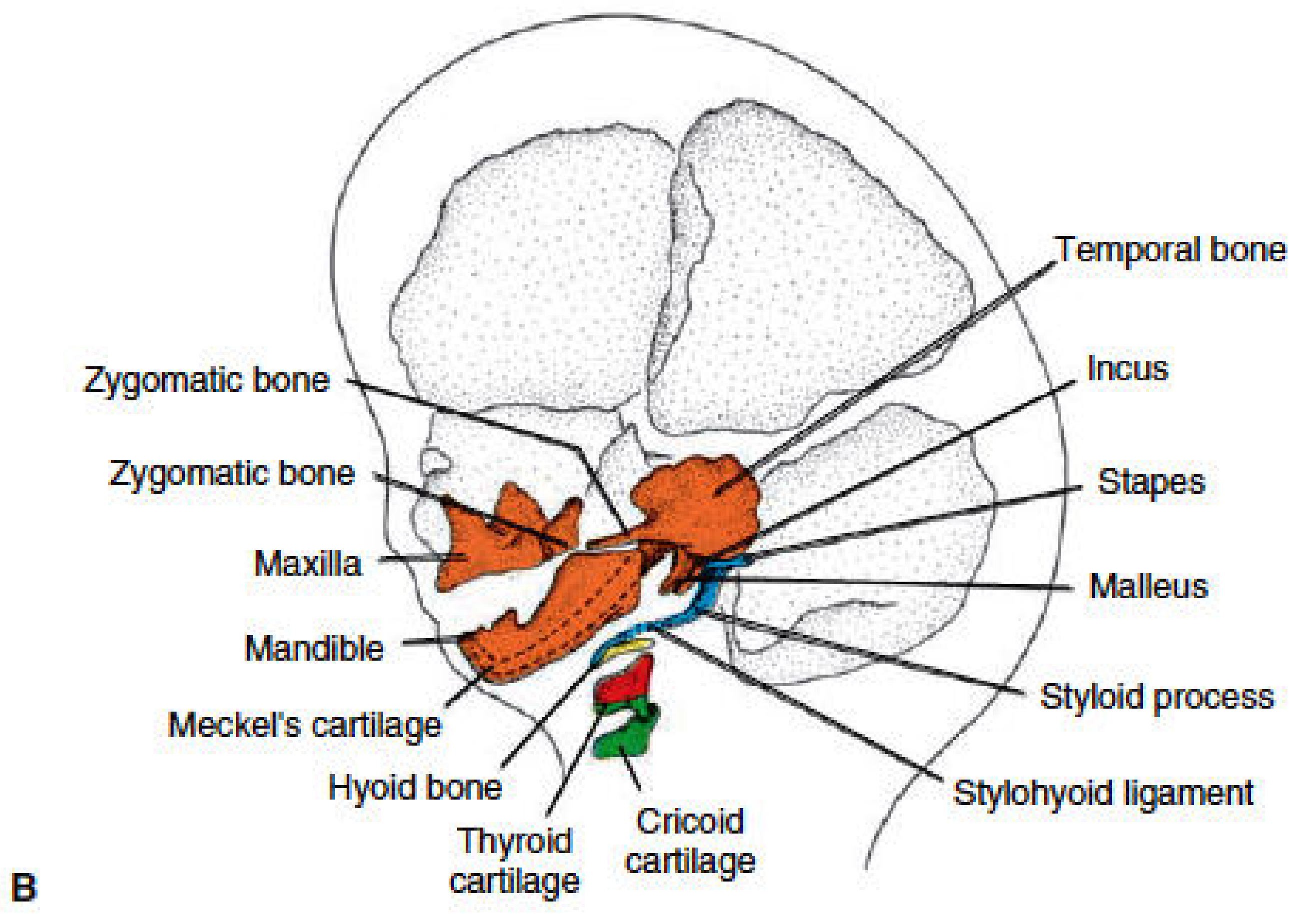
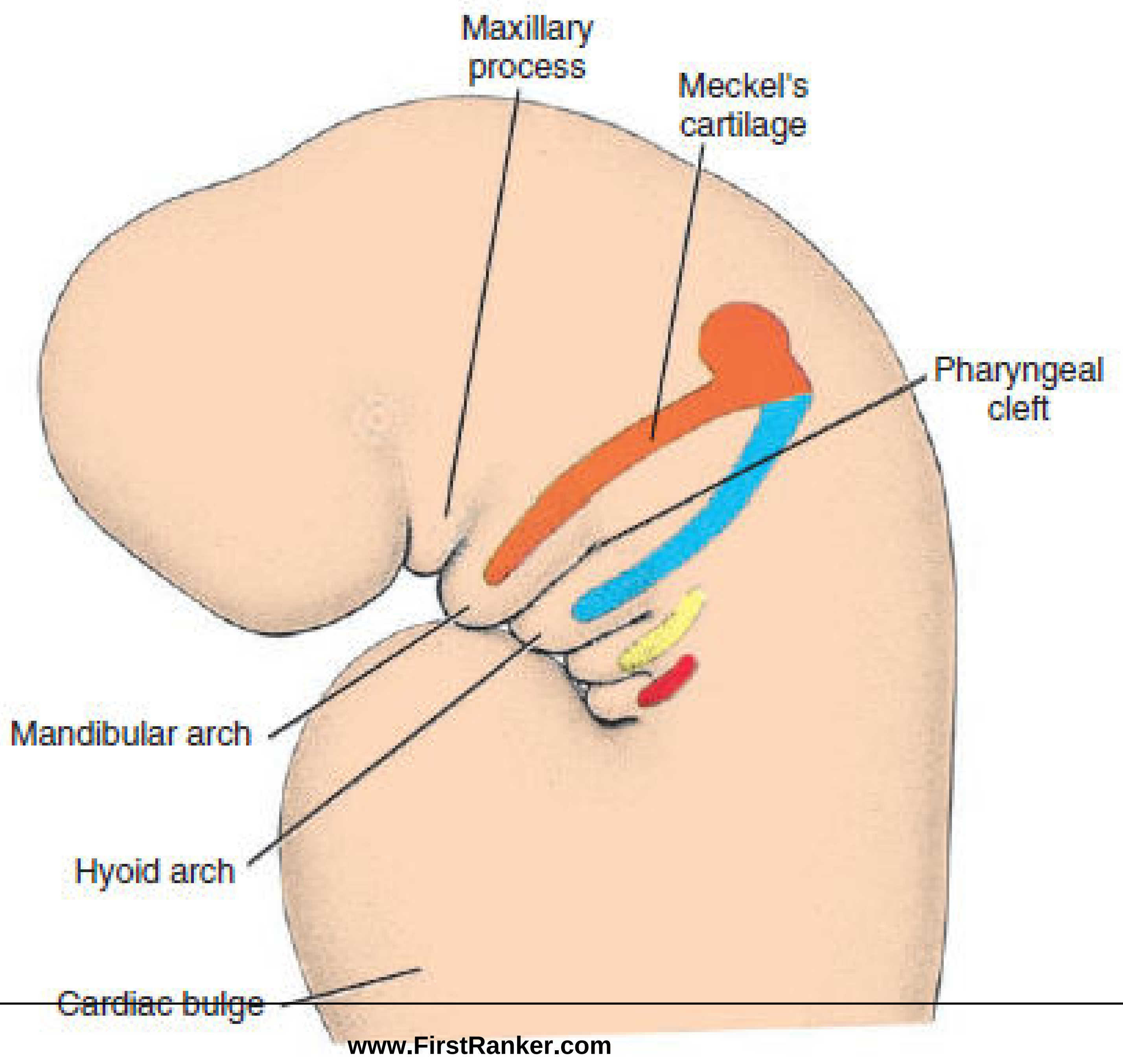


Figure 17.8 A. Lateral view of the head and neck region demonstrating the cartilages of the pharyngeal arches participating in formation of the bones of the face and neck. **B.** Various components of the pharyngeal arches later in development. Some of the components ossify; others disappear or become ligamentous. The maxillary process and Meckel's cartilage are replaced by the maxilla and mandible, respectively, which develop by membranous ossification.



A

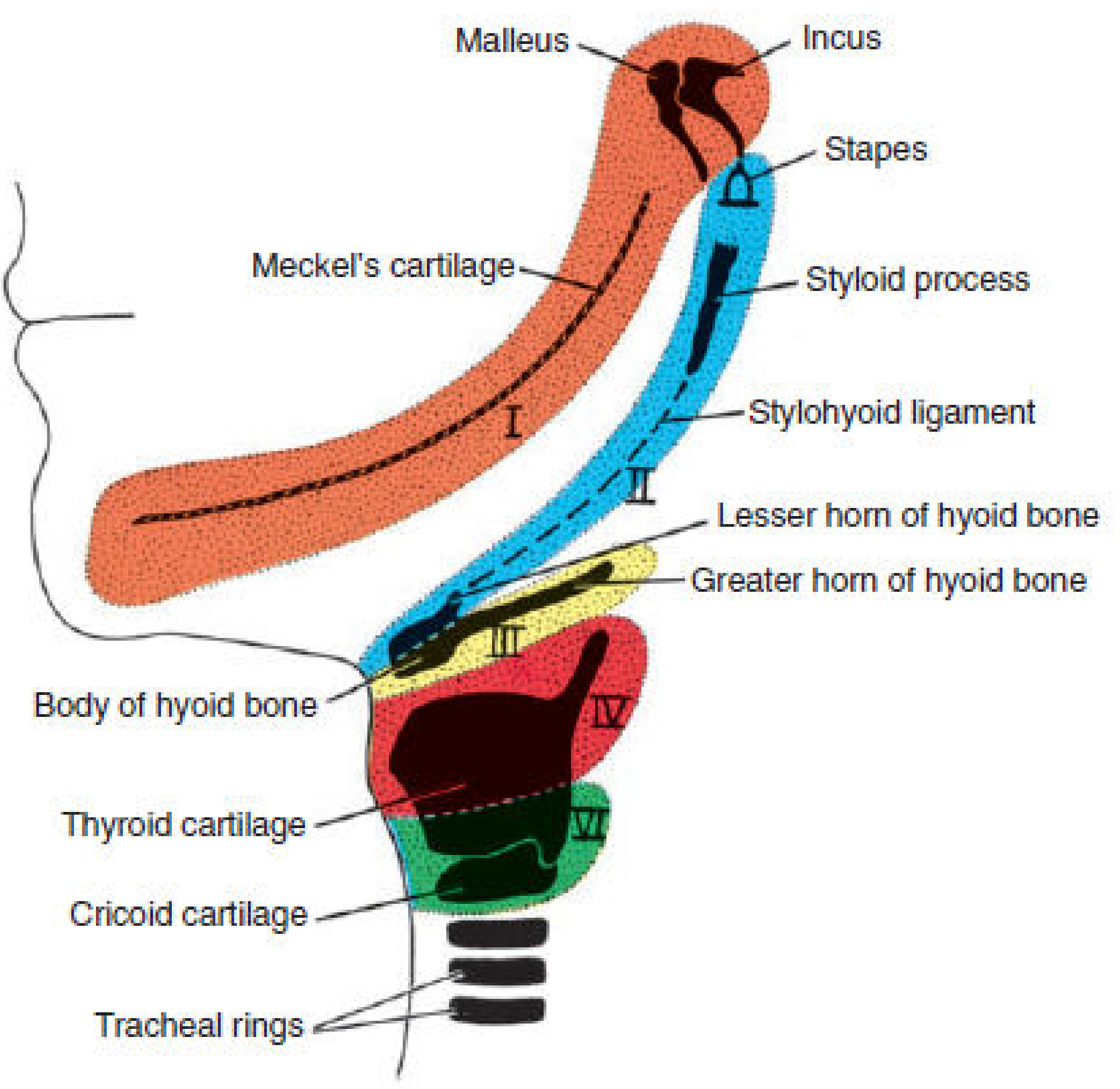


Figure 17.9 Definitive structures formed by the cartilaginous components of the various pharyngeal arches.

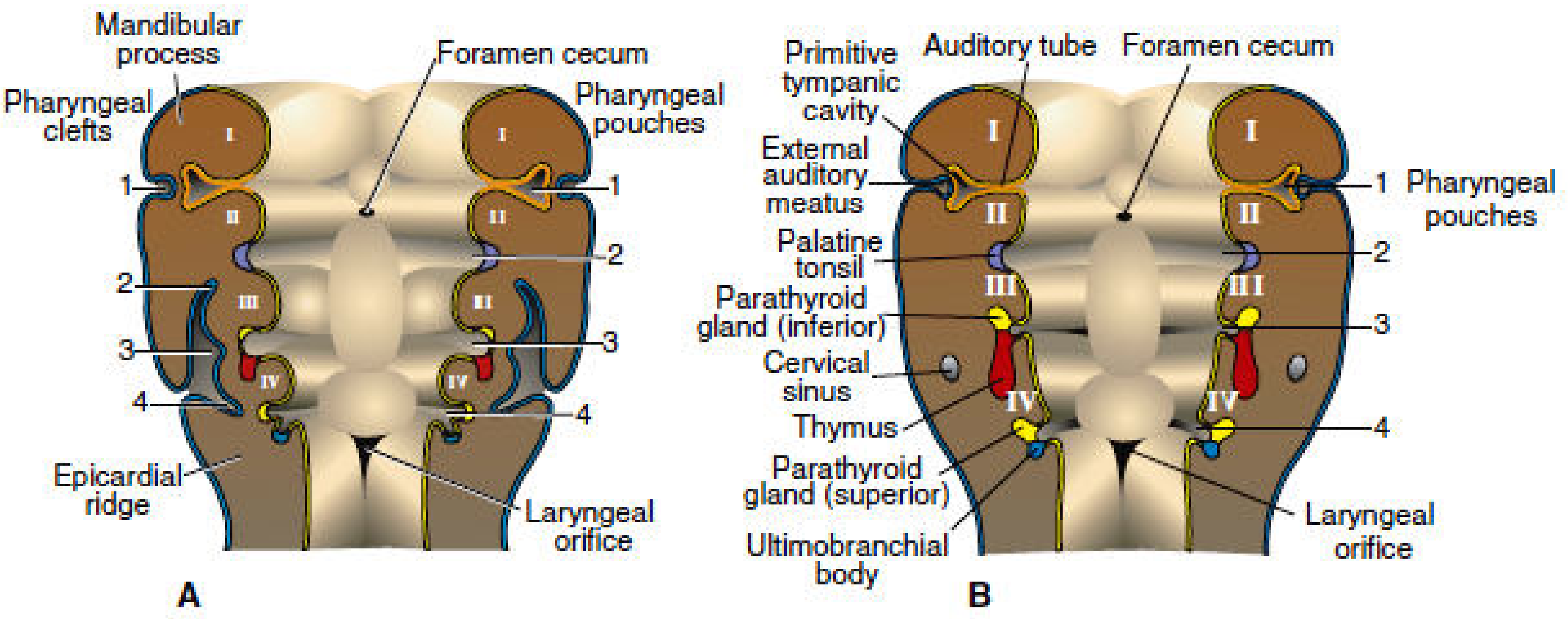


Figure 17.10 **A.** Development of the pharyngeal clefts and pouches. The second arch grows over the third and fourth arches, burying the second, third, and fourth pharyngeal clefts. **B.** Remnants of the second, third, and fourth pharyngeal clefts form the cervical sinus, which is normally obliterated. Note the structures formed by the various pharyngeal pouches.

TABLE 17.2 Derivatives of the Pharyngeal Pouches

Pharyngeal Pouch	Derivatives
1	Tympanic (middle ear) cavity
	Auditory (eustachian) tube
2	Palatine tonsils
	Tonsillar fossa
3	Inferior parathyroid gland
	Thymus
4	Superior parathyroid gland ultimobranchial body (parafollicular [C] cells of the thyroid gland)

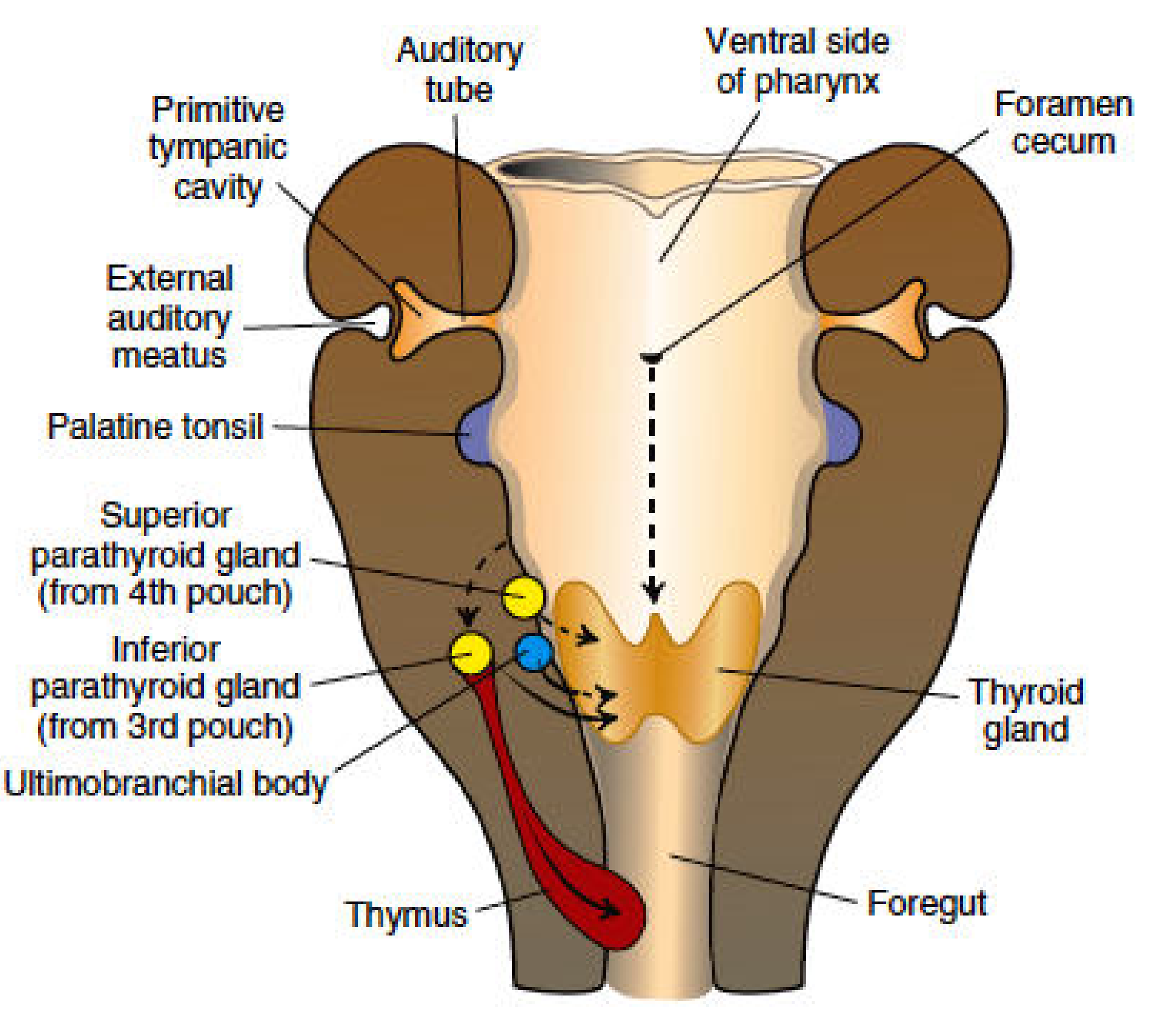


Figure 17.11 Migration of the thymus, parathyroid glands, and ultimobranchial body. The thyroid gland originates in the midline at the level of the foramen cecum and descends to the level of the first tracheal rings.

Which of the following structure is NOT part of the first branchial arch?
a) malleus
b) mandibular process
c) sphenomandibular ligament
d) stylohyoid ligament

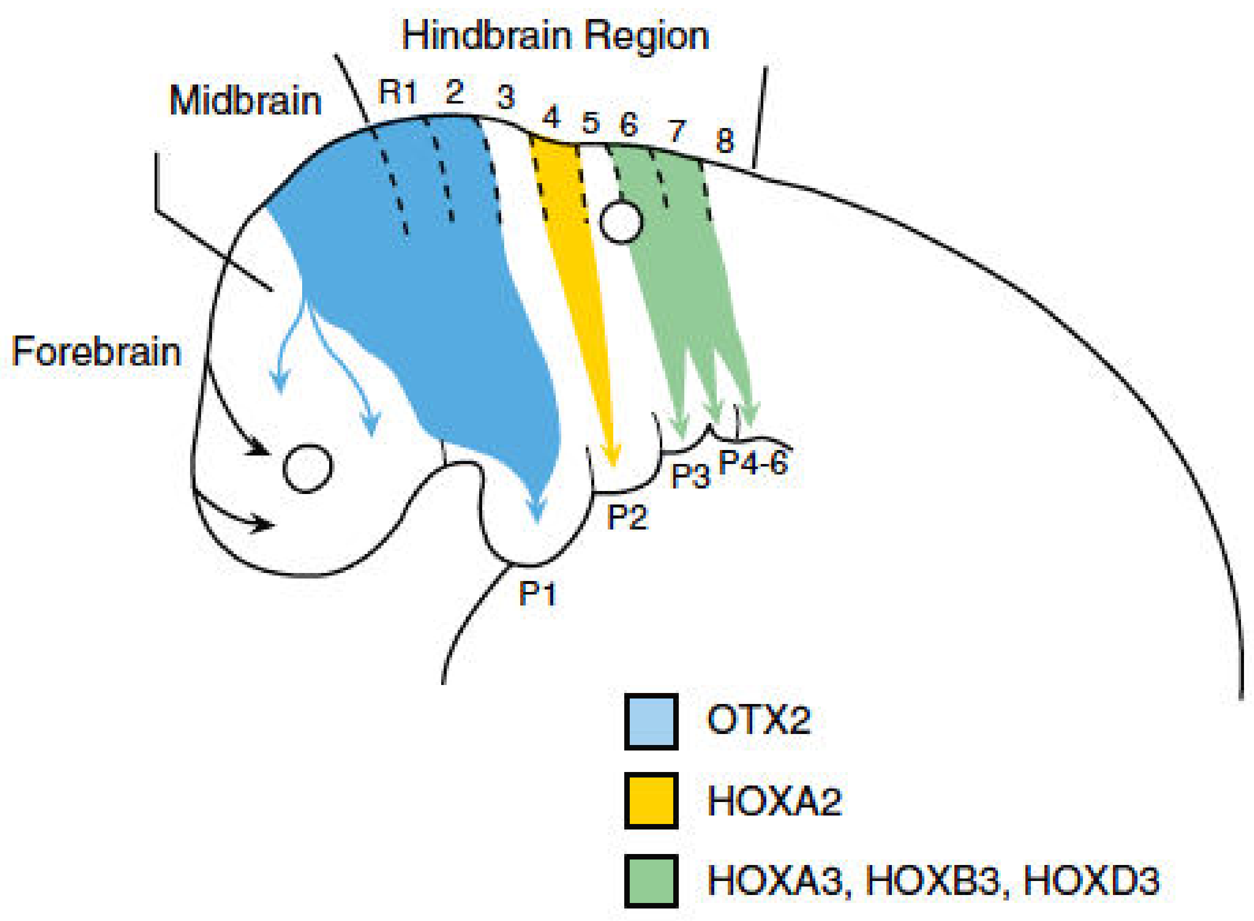


Figure 17.12 Drawing showing the pathways of neural crest cell migration from the cranial neural folds into the face and pharyngeal arches. From the hindbrain region, crest cells migrate from segments called *rhombomeres*. Rhombomeres express a specific pattern of *HOX* genes (the midbrain and rhombomeres 1 and 2 express the homeodomain-containing transcription factor *OTX2*; see also Fig. 18.31), and neural crest cells carry these expression patterns into the pharyngeal arches. Also, notice that there are three streams of crest cells and that rhombomeres 3 and 5 do not contribute many (if any) cells to these streams. The three streams are important because they provide guidance cues for cranial nerves growing back from their ganglia to establish connections in the hindbrain (see also Fig. 18.40).

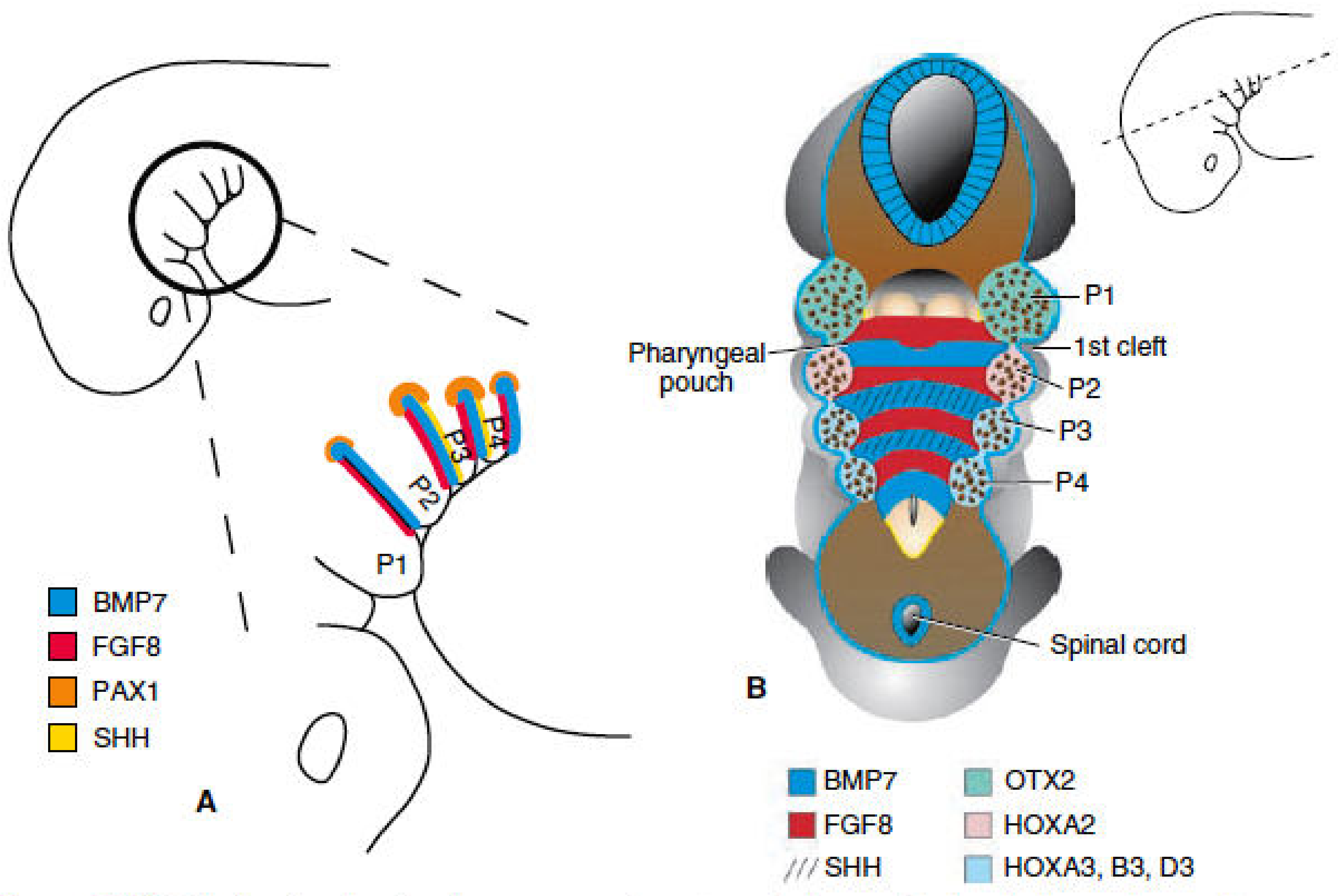


Figure 17.13 A,B. Drawings showing the gene expression patterns in pharyngeal arch endoderm and mesenchyme. Endoderm is responsible for patterning the skeletal derivatives of the arches, but the response of the mesenchyme to these signals is dictated by the genes that the mesenchyme expresses. Gene expression in the endoderm of the pouches shows a specific pattern: *FGF8* is expressed in the anterior region of each pouch with *BMP7* expressed in the posterior region; *SHH* is expressed in the posterior region of pouches 2 and 3, while *PAX1* is expressed in the dorsal-most area of each pouch. **A,B.** Mesenchymal expression patterns are established by neural crest cells that migrate into the arches and carry the genetic code from their rhombomeres of origin (or also from the midbrain in the case of the first arch) to the arches **B** (see also Figs. 17.12 and 18.31).

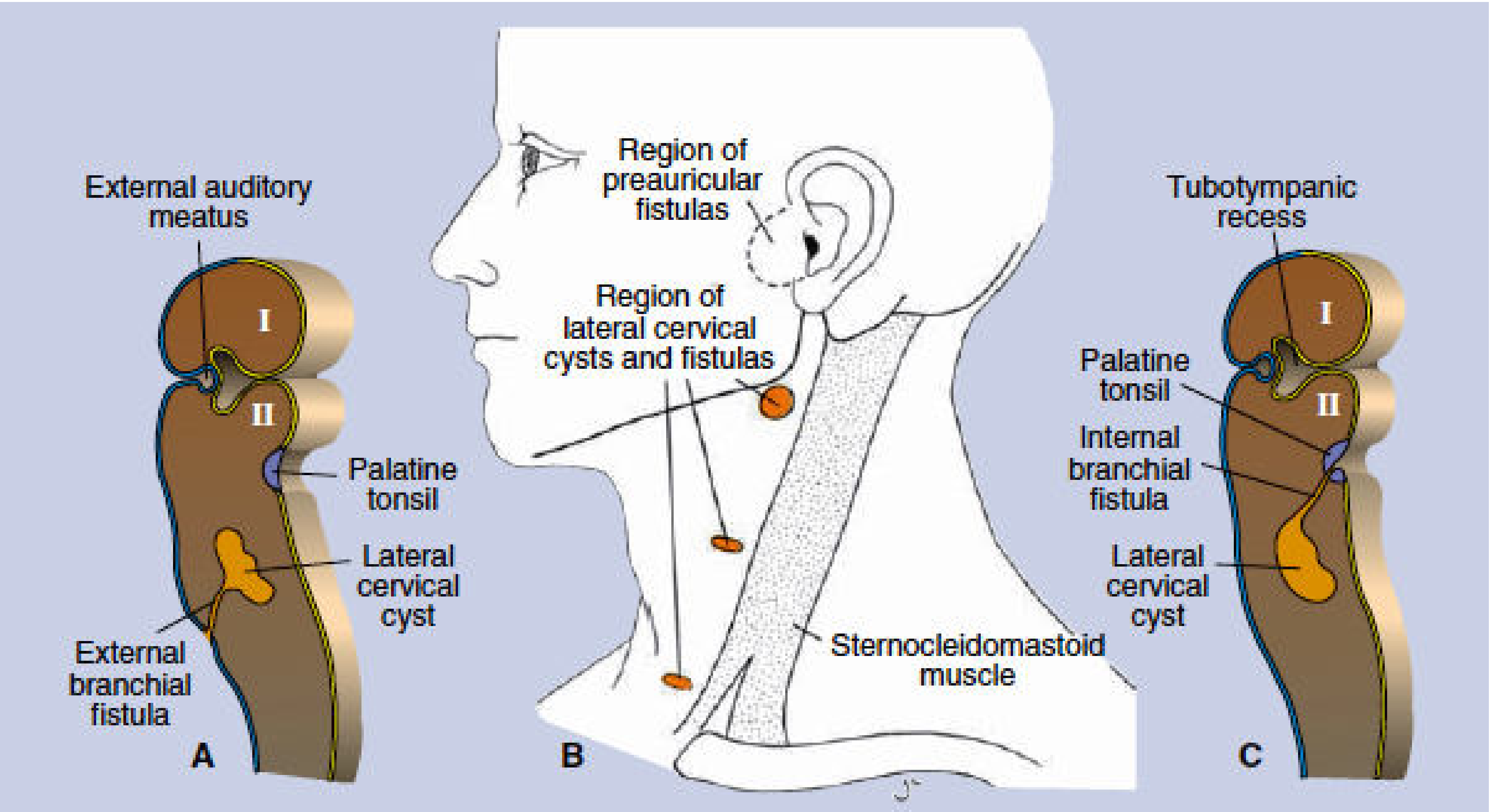


Figure 17.14 A. Lateral cervical cyst opening at the side of the neck by way of a fistula. **B.** Lateral cervical cysts and fistulas in front of the sternocleidomastoid muscle. Note also the region of preauricular fistulas. **C.** A lateral cervical cyst opening into the pharynx at the level of the palatine tonsil.

Figure 17.15 Patient with a lateral cervical cyst. These cysts are always on the lateral side of the neck in front of the sternocleidomastoid muscle. They commonly lie under the angle of the mandible and do not enlarge until later in life.

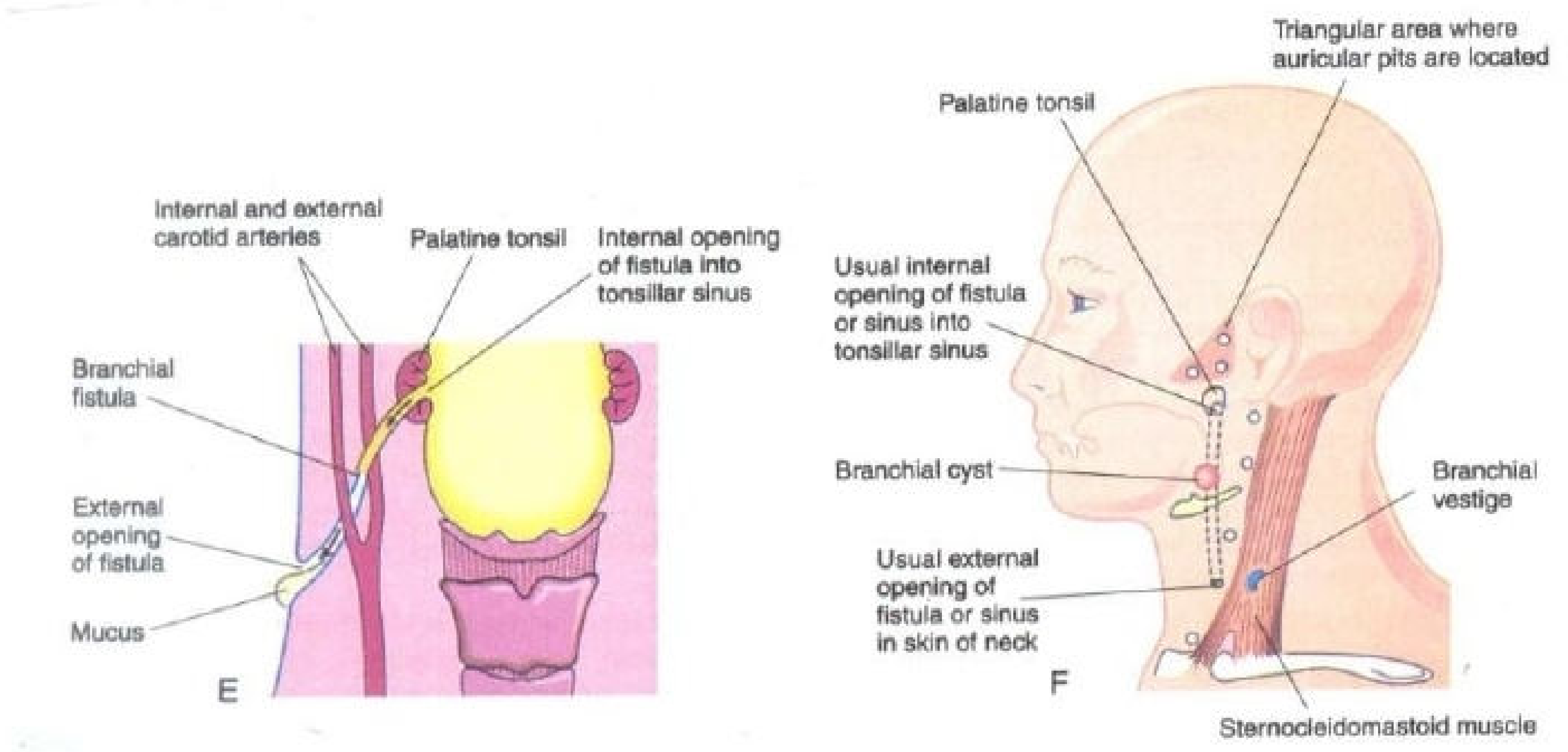


Figure 17.16 Patients with craniofacial defects thought to arise from insults to neural crest cells. **A.** Robin sequence. Note the very small mandible (micrognathia) that keeps the tongue from "dropping" out of the way of the palatal shelves resulting in cleft palate. **B,C.** Examples of 22q11.2 deletion syndrome: DiGeorge syndrome. **B.** Note the small mouth, nearly smooth philtrum, micrognathia, prominent nasal bridge, and posteriorly rotated ears; Velo-cardio-facial syndrome. **C.** This patient shows mild facial dysmorphology, including mild malar hypoplasia, micrognathia, prominent upper lip, and large ears. **D.** Hemifacial microsomia (oculoauriculovertebral spectrum, or Goldenhar syndrome). Note the abnormal ear with skin tags and the small chin.

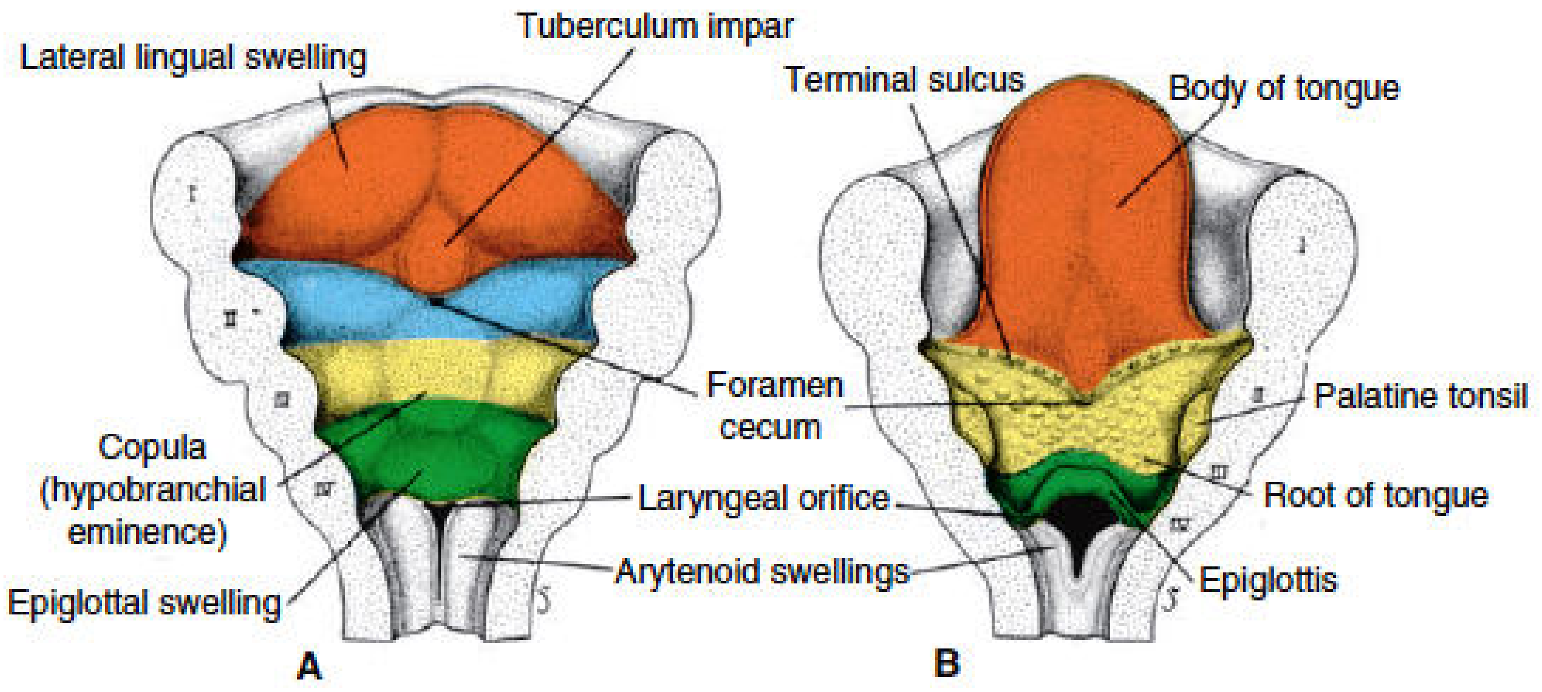
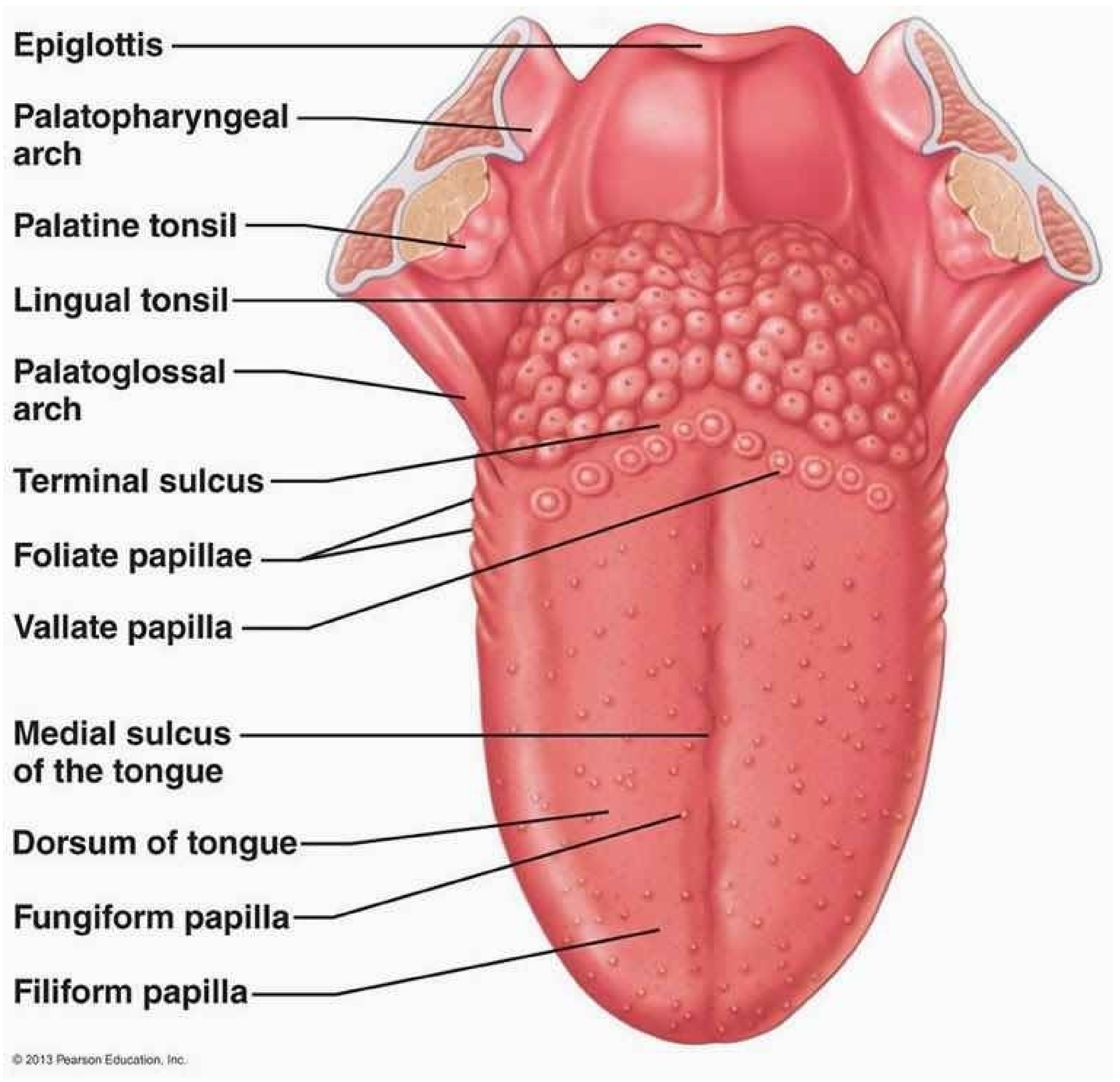
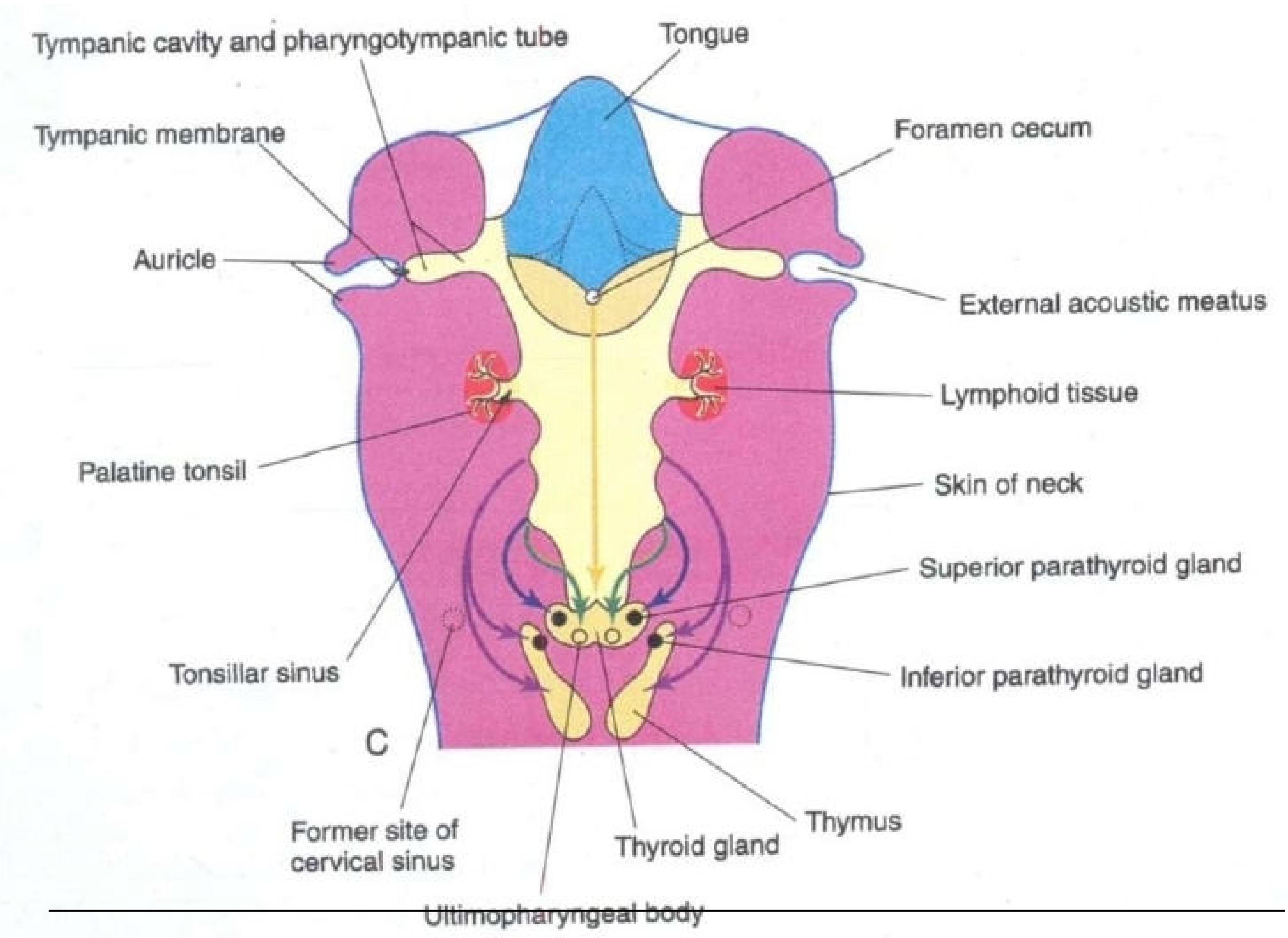


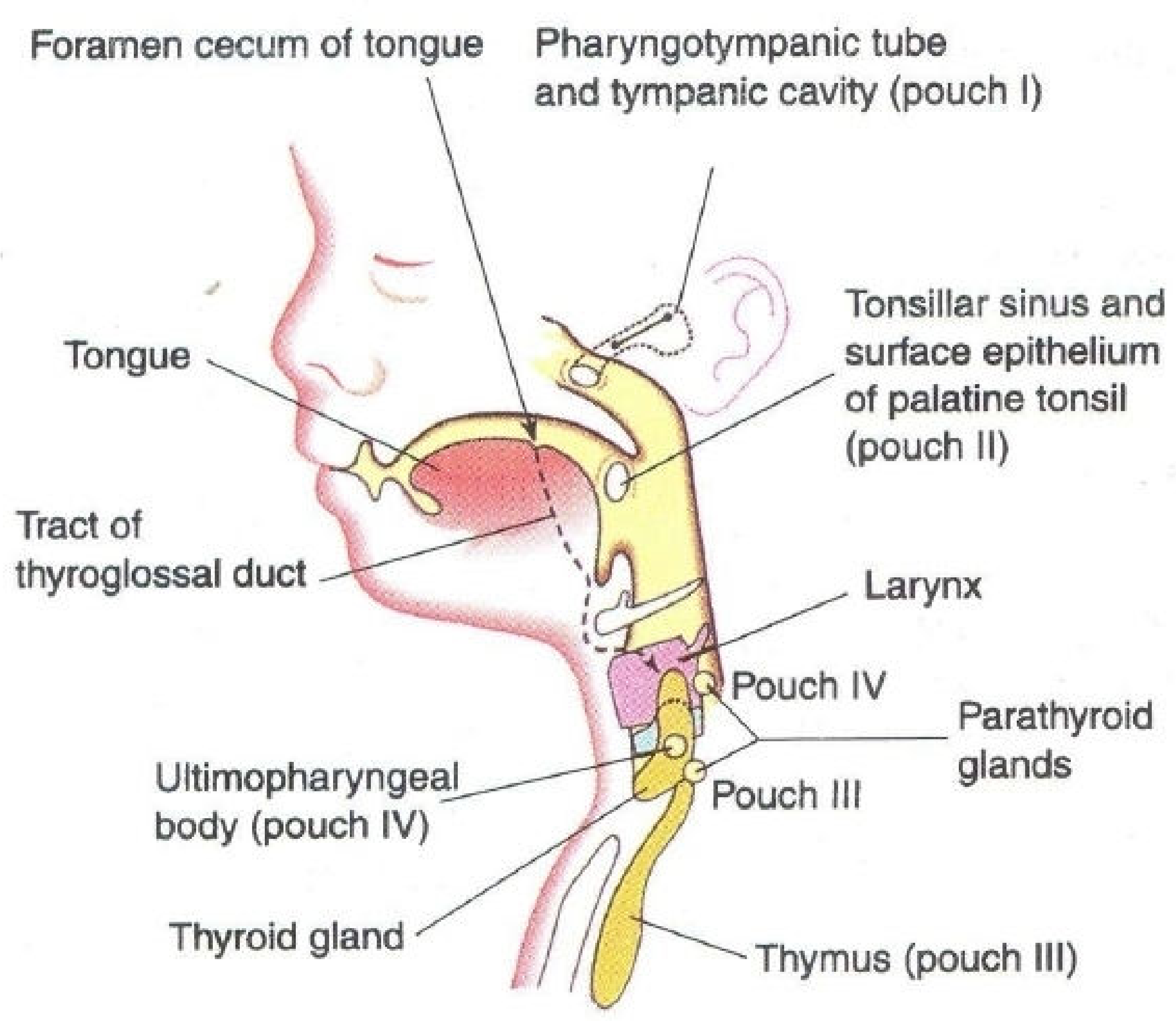
Figure 17.17 Ventral portion of the pharyngeal arches seen from above showing development of the tongue. I to IV, the cut pharyngeal arches. **A.** 5 weeks (~6 mm). **B.** 5 months. Note the foramen cecum, site of origin of the thyroid primordium.

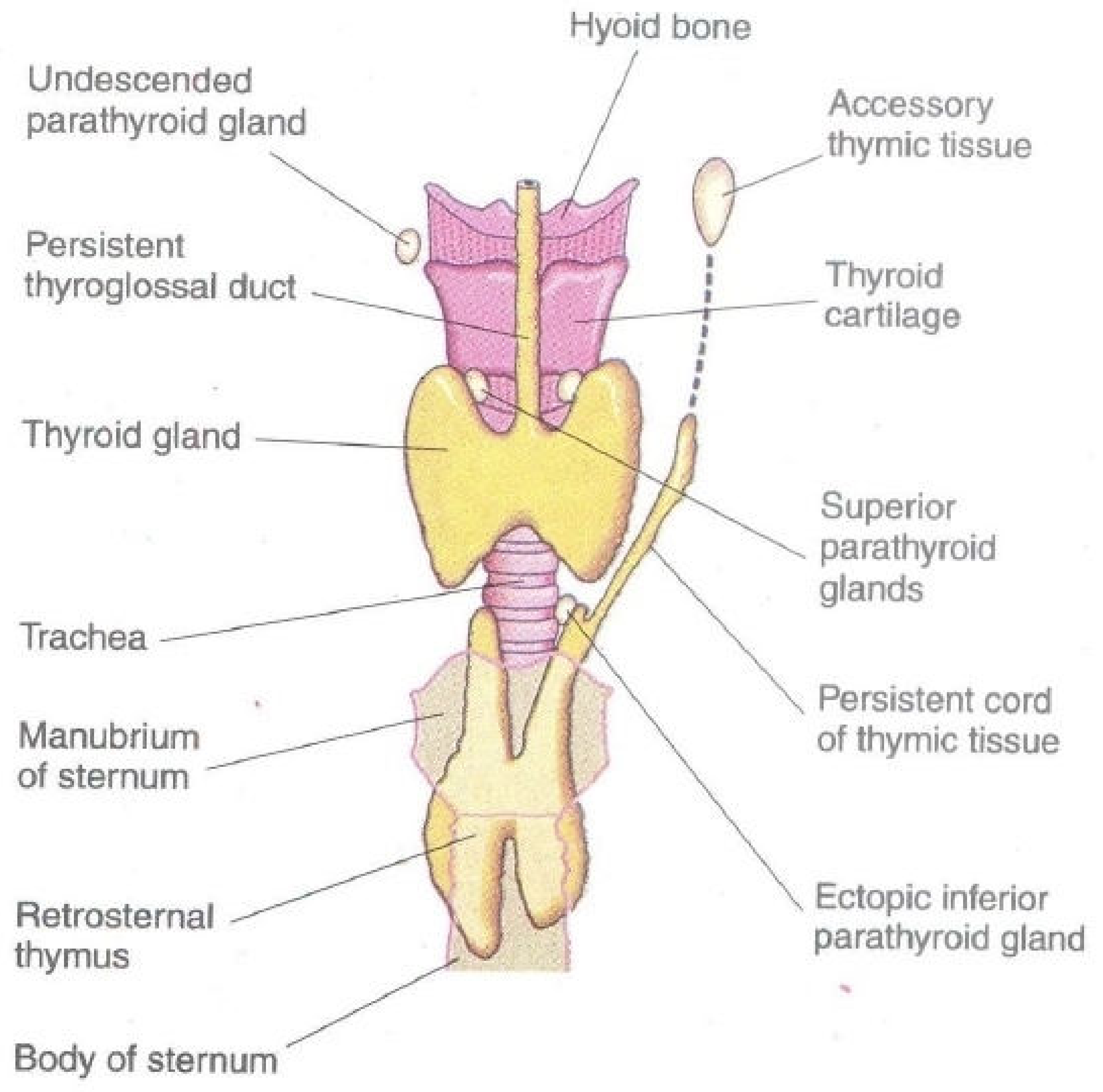
The hypobranchial eminence contributes to formation of:
a) anterior portion of the tongue
b) Posterior portion of the tongue
c) musculature of the tongue
d) Epiglottis

Myoblasts form the occipital myotomes believed to give rise to the muscle of the:
a) eye
b) face
c) ear
d) tongue









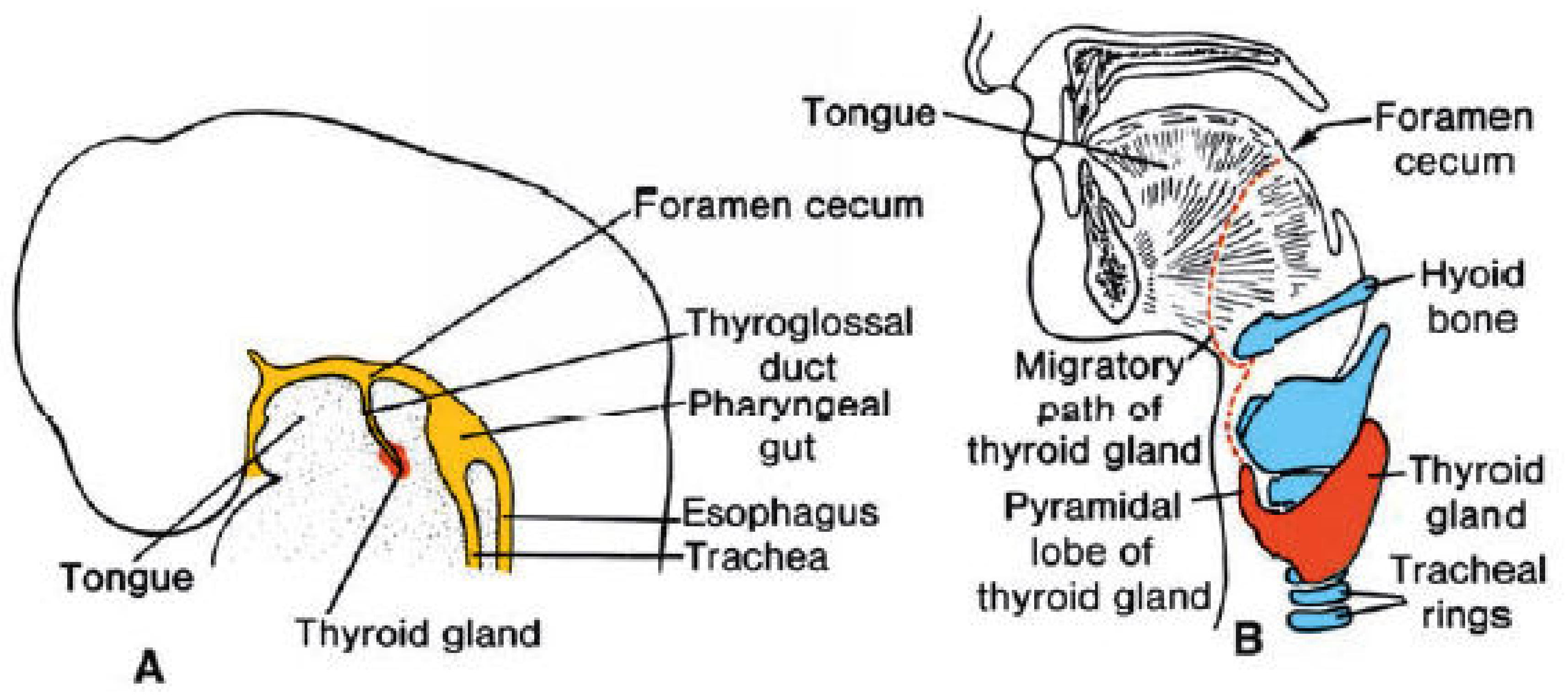


Figure 17.18 **A.** The thyroid primordium arises as an epithelial diverticulum in the midline of the pharynx immediately caudal to the tuberculum impar. **B.** Position of the thyroid gland in the adult. *Broken line*, the path of migration.

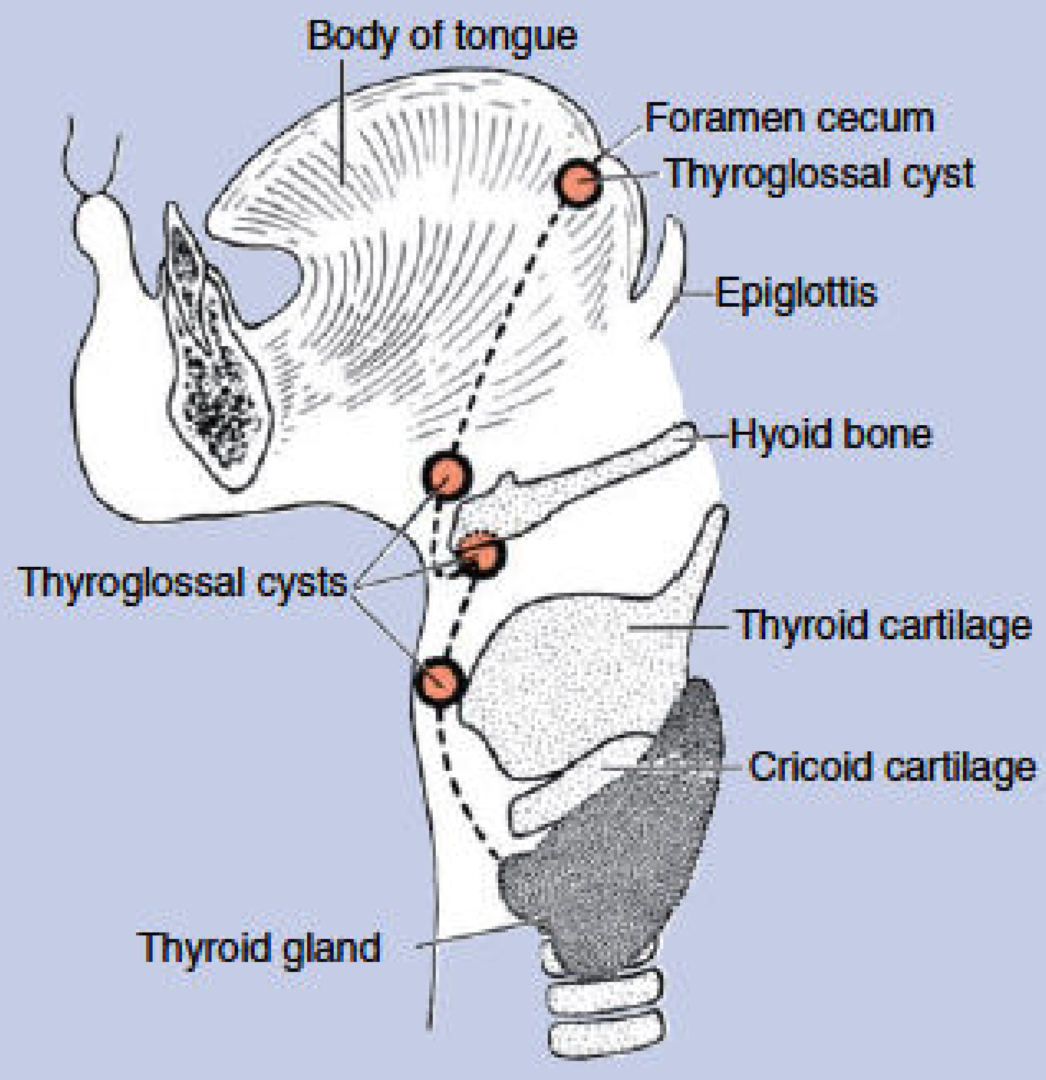


Figure 17.19 Thyroglossal cysts. These cysts, most frequently found in the hyoid region, are always close to the midline.

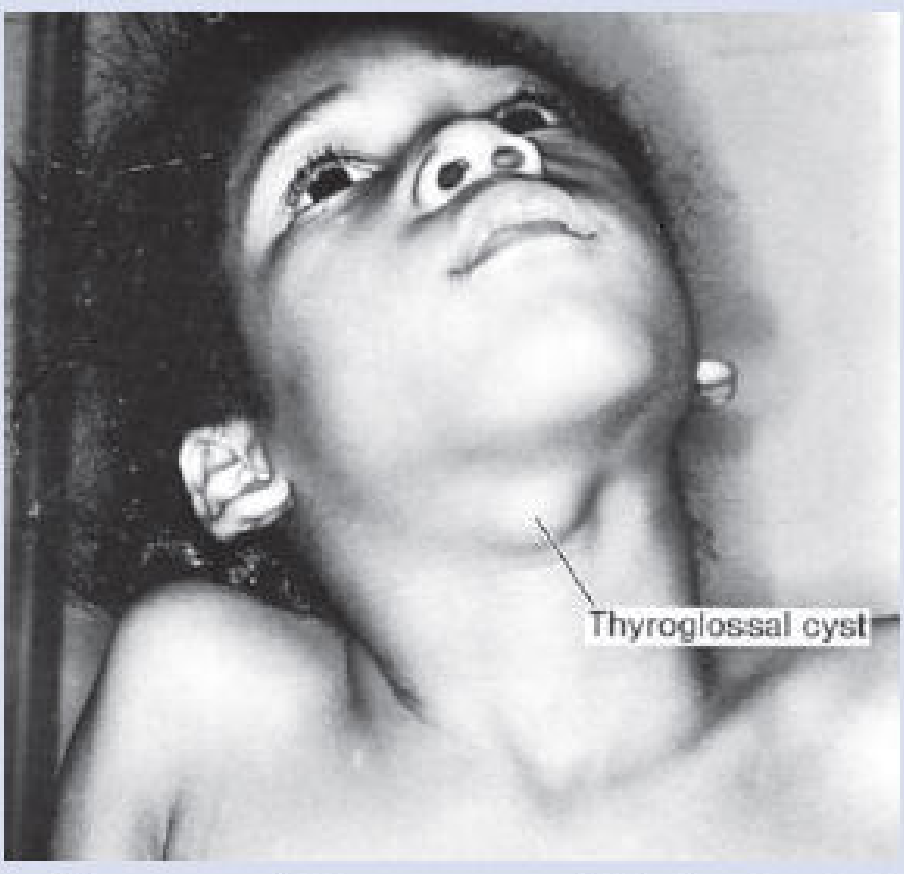
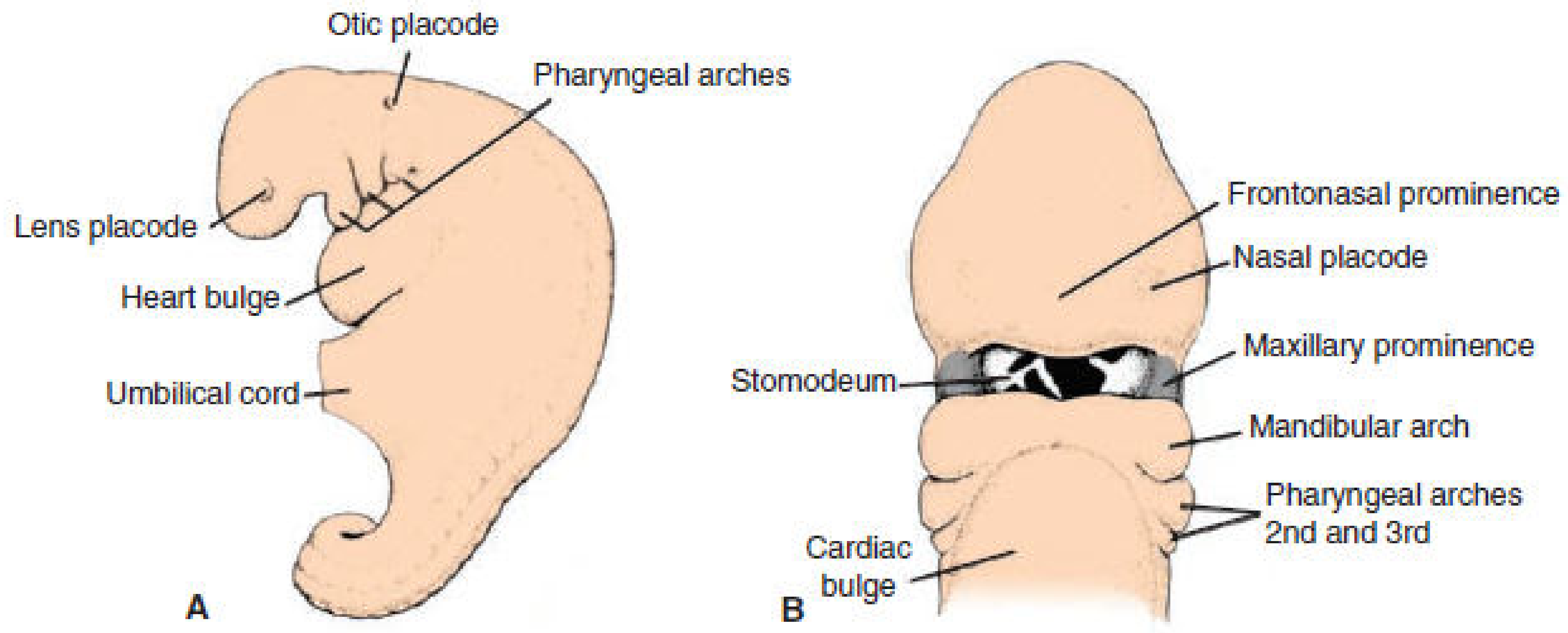


Figure 17.20 Thyroglossal cyst. These cysts, which are remnants of the thyroglossal duct, may be anywhere along the migration pathway of the thyroid gland. They are commonly found behind the arch of the hyoid bone. An important diagnostic characteristic is their midline location.

The most common site of a thyroglossal cyst is the
a) Dorsal aspect of neck
b) Anterior border of sternocleidomastoid muscle
c) Superior mediastinum
d) Midline close to the hyoid bone



DEVELOPMENT OF FACE

Figure 17.29 A. Lateral Cleft lip (compare to Fig. 17.28C). B. Bilateral cleft lip (compare to Fig. 17.28D). C. Cleft palate (compare to Fig. 17.28E). D. Midline cleft lip

MACROSTOMIA

Bilateral cleft of lip & palate
(fig.1)

cleft of the hard & soft palates
(fig.2)

Unilateral cleft of lip
(fig.3)

cleft of lip & palate

(fig.4)

Bifid uvula

(fig.5)

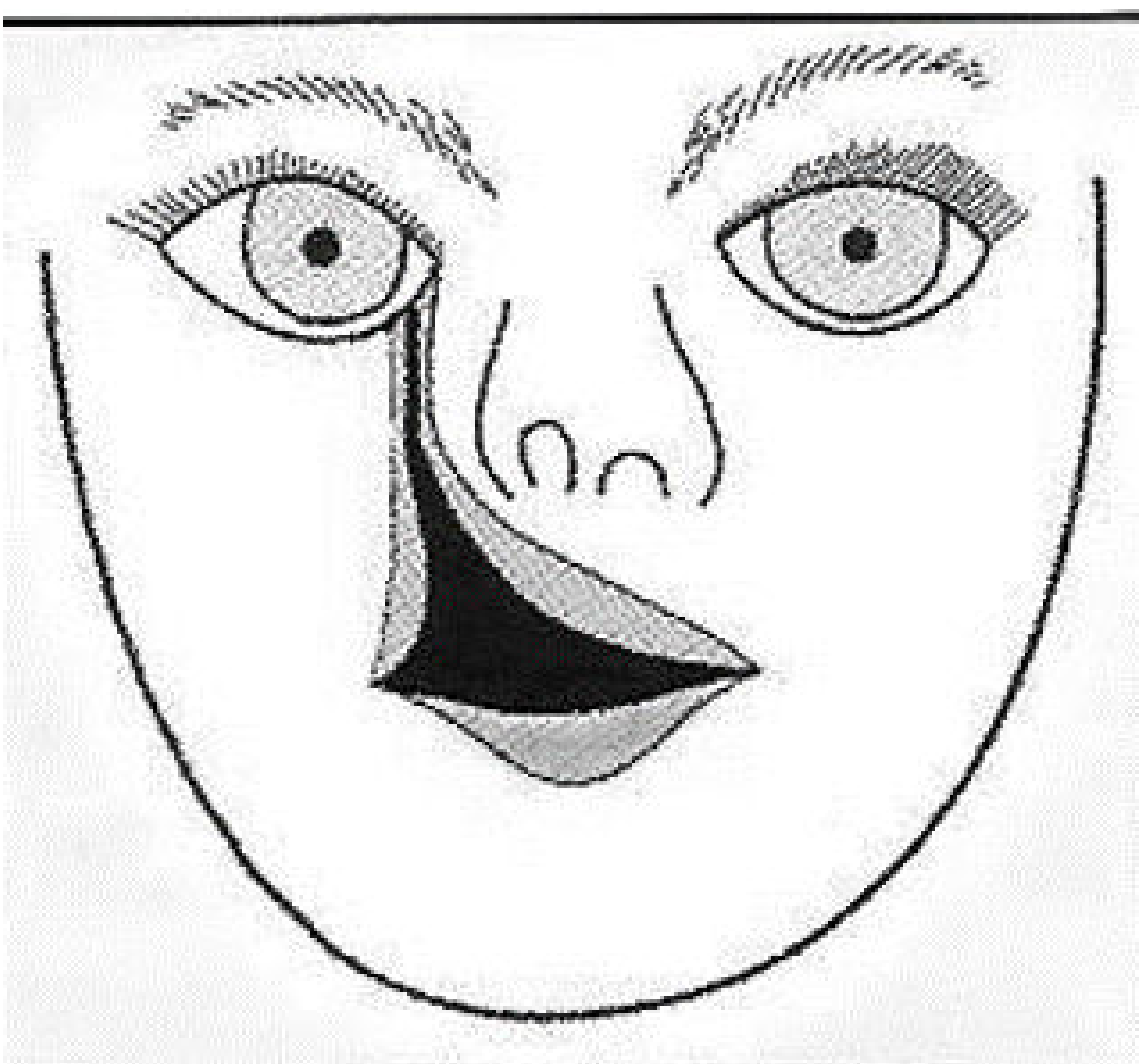


Figure 17.21 **A.** Lateral view of an embryo at the end of the fourth week, showing position of the pharyngeal arches. **B.** Frontal view of a 4.5-week embryo showing the mandibular and maxillary prominences. The nasal placodes are visible on either side of the frontonasal prominence. **C.** Scanning electron micrograph of a human embryo at a stage similar to that of **B.**

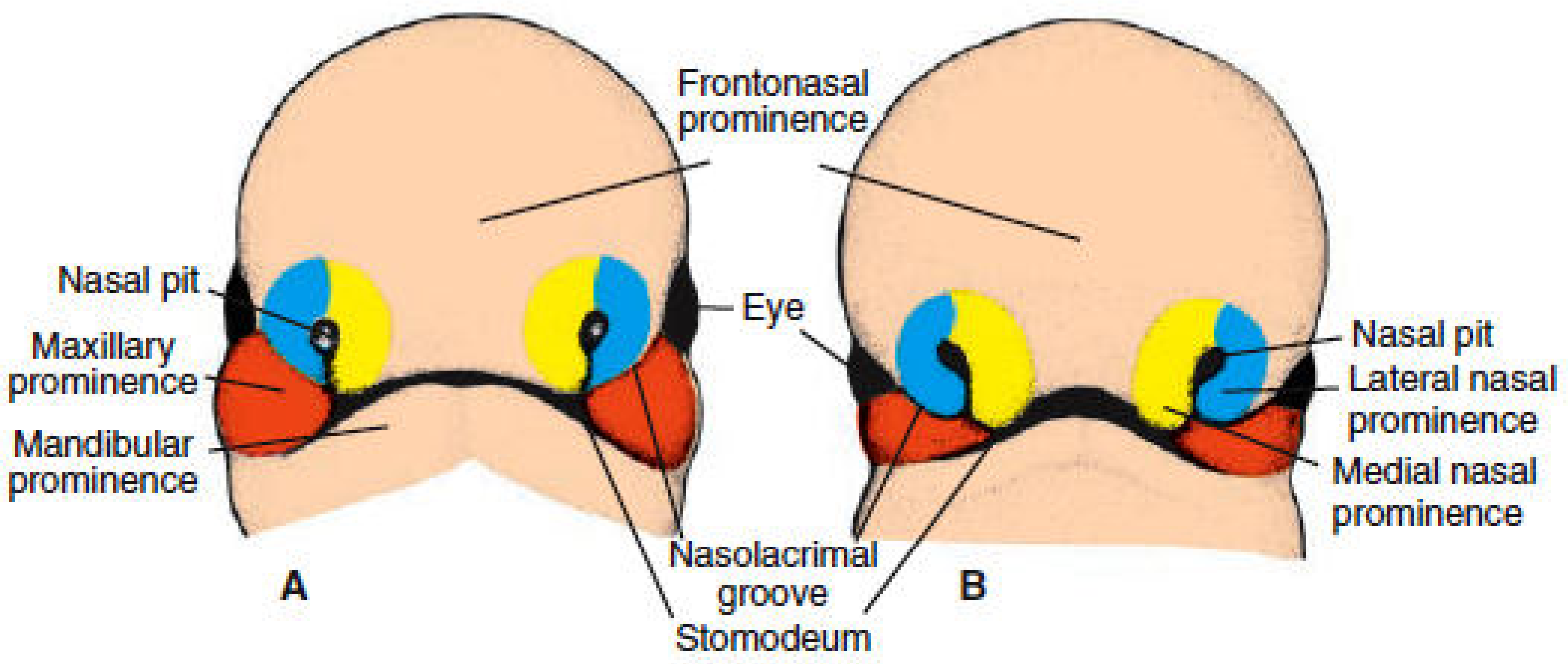
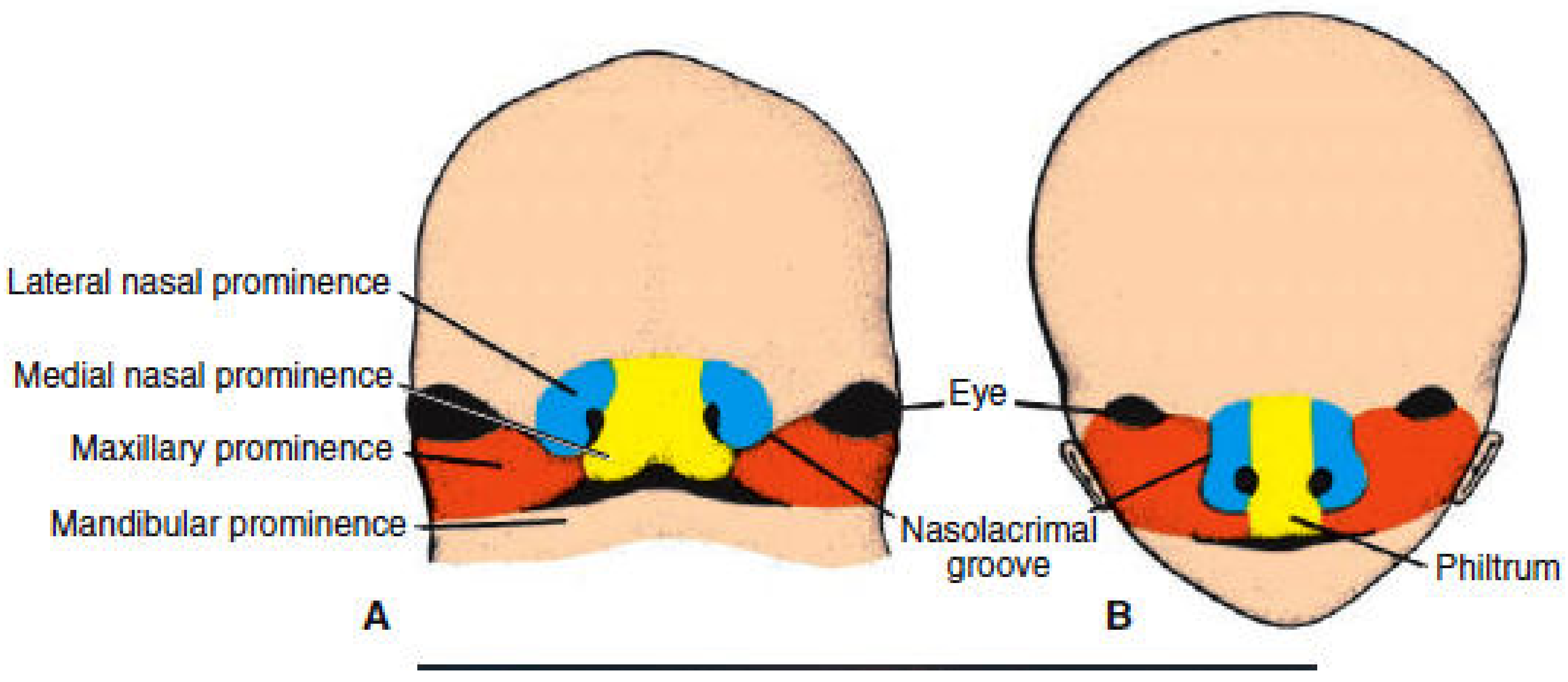


Figure 17.22 Frontal aspect of the face. **A.** 5-week embryo. **B.** 6-week embryo. The nasal prominences are gradually separated from the maxillary prominence by deep furrows.



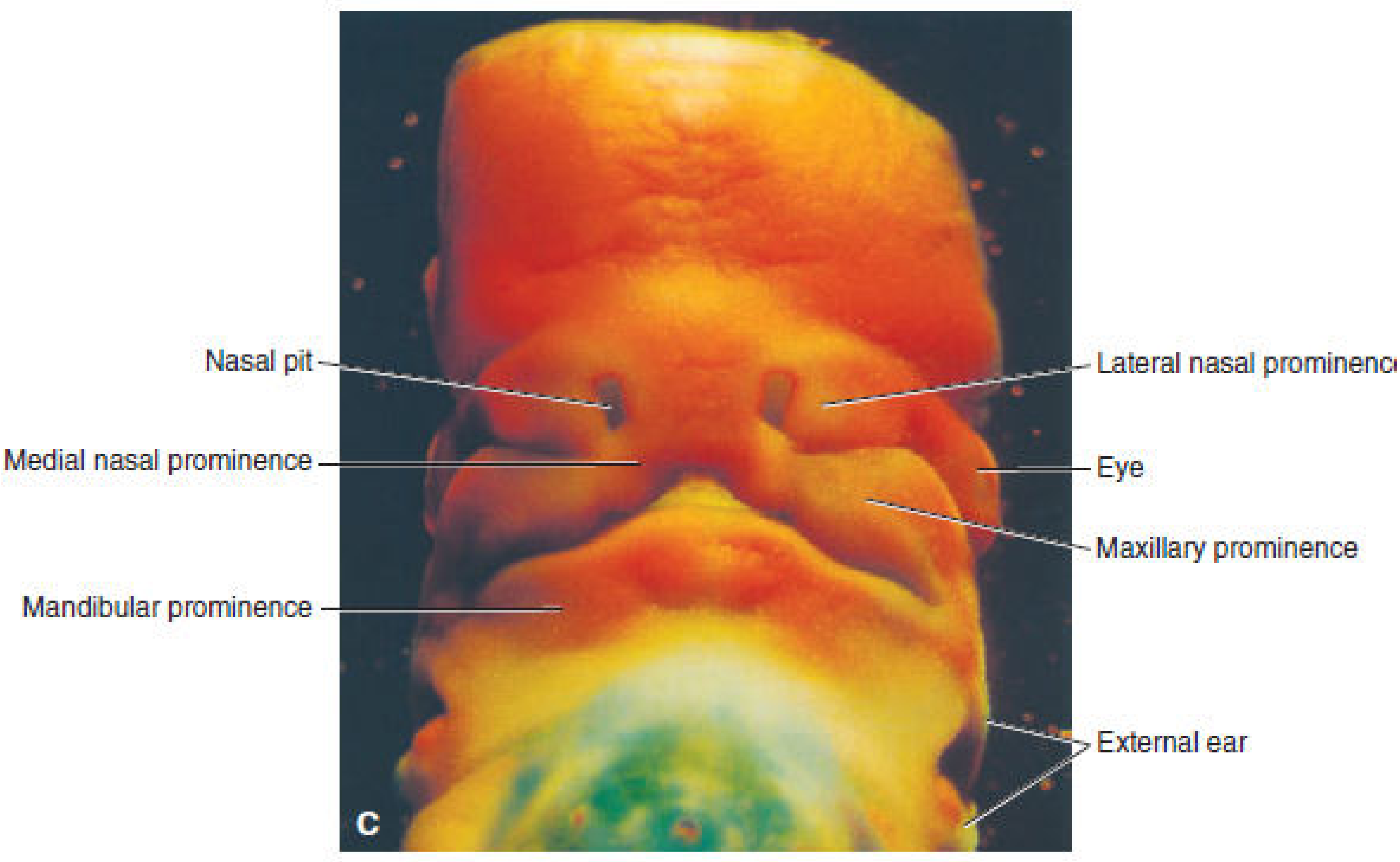


Figure 17.23 Frontal aspect of the face. **A.** 7-week embryo. Maxillary prominences have fused with the medial nasal prominences. **B.** 10-week embryo. **C.** Photograph of a human embryo at a stage similar to that in **A.**

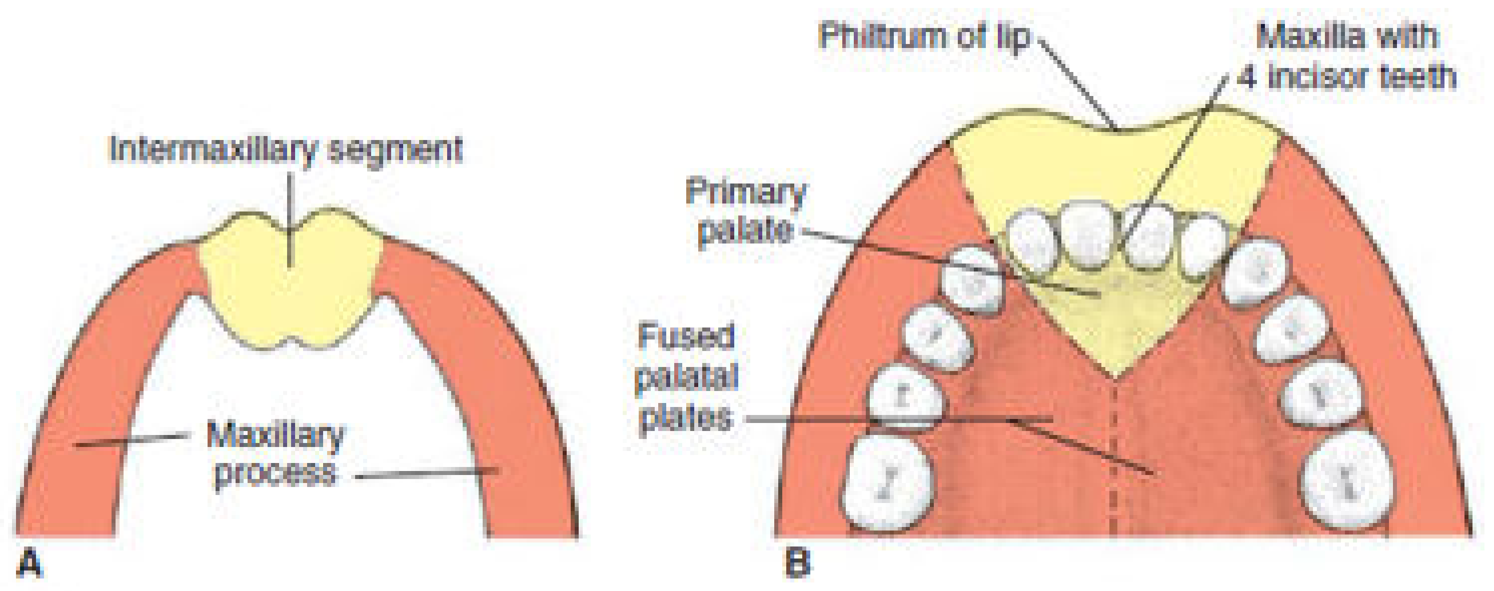


Figure 17.24 **A.** Intermaxillary segment and maxillary processes. **B.** The intermaxillary segment giving rise to the philtrum of the upper lip, the median part of the maxillary bone with its four incisor teeth, and the triangular primary palate.

The intermaxillary segment forms via the fusion of the
a) Maxillary prominences
b) Mandibular prominences
c) Medial nasal prominences
d) Lateral nasal prominence

Fronto-nasal Process

- **Nasal placode:** rounded thickening in the lower lateral parts of the fronto-nasal process.
- **Nasal pits & swellings:** invagination of placode will form nasal pits which are surrounded by medial & lateral nasal prominences (folds).
- **Intermaxillary segment:** from fused medial nasal prominences . It forms intermaxillary segments that form middle part of nose, filtrum, part of upper jaw that carries upper 4 incisors and primary palate.

Maxillary Process

- It is separated from side of fronto-nasal process by naso-lacrimal groove, inside which a cord of ectodermal cells is formed then becomes canalized to form naso-lacrimal duct. Its upper end forms lacrimal sac.
- Maxillary process fuses with fronto-nasal process covering naso-lacrimal duct.
- It forms lower boundary of nasal pit.
- It is separated from other maxillary process by intermaxillary segment.

-It also fuses partially with mandibular process to form the cheek.
-Palatine process is formed as inward projection of maxillary process to form secondary palate which divides stomodeum into upper nasal and lower oral cavity.
So maxillary process forms upper part of cheek, upper lip except filtrum, upper jaw except part that carries upper incisors and most of hard palate.

Mandibular Process

- It forms lower part of cheek, whole lower lip and lower jaw and floor of mouth.
- **Development of palate:**
- 1- **Primary palate:** from intermaxillary segment of fronto-nasal process.
- 2- **Secondary palate:** from palatine processes of maxillary processes that form most of hard palate and soft palate.
- -Hard palate receives downward growth of nasal septum.

TABLE 17.3 Structures Contributing to Formation of the Face

Prominence	Structures Formed
Frontonasal[a]	Forehead, bridge of nose, and medial and lateral nasal prominences
Maxillary	Cheeks, lateral portion of upper lip
Medial nasal	Philtrum of upper lip, crest, and tip of nose
Lateral nasal	Alae of nose
Mandibular	Lower lip

[a]The frontonasal prominence is a single unpaired structure; the other prominences are paired.

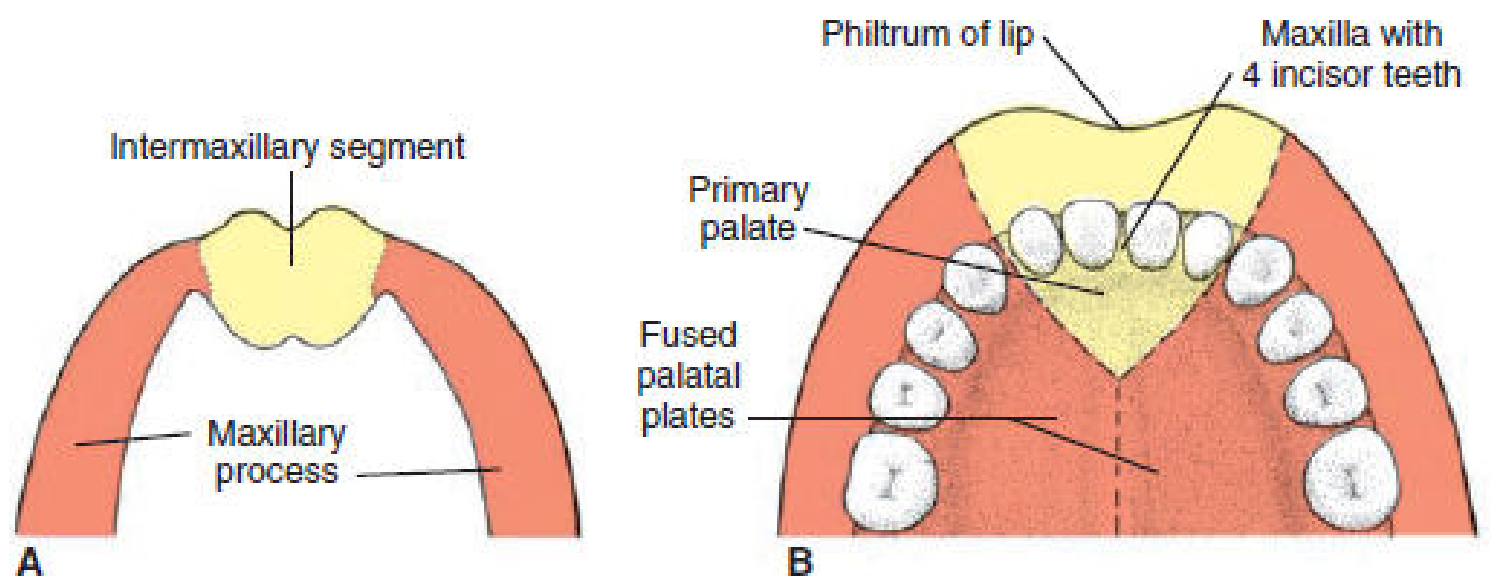


Figure 17.24 A. Intermaxillary segment and maxillary processes. B. The intermaxillary segment giving rise to the philtrum of the upper lip, the median part of the maxillary bone with its four incisor teeth, and the triangular primary palate.

. The nasolacrimal groove separates the:
a) mandibular and maxillary swelling
b) lateral nasal swelling and maxillary swelling
c) medial nasal swelling and maxillary swelling
d) first and second branchial swelling

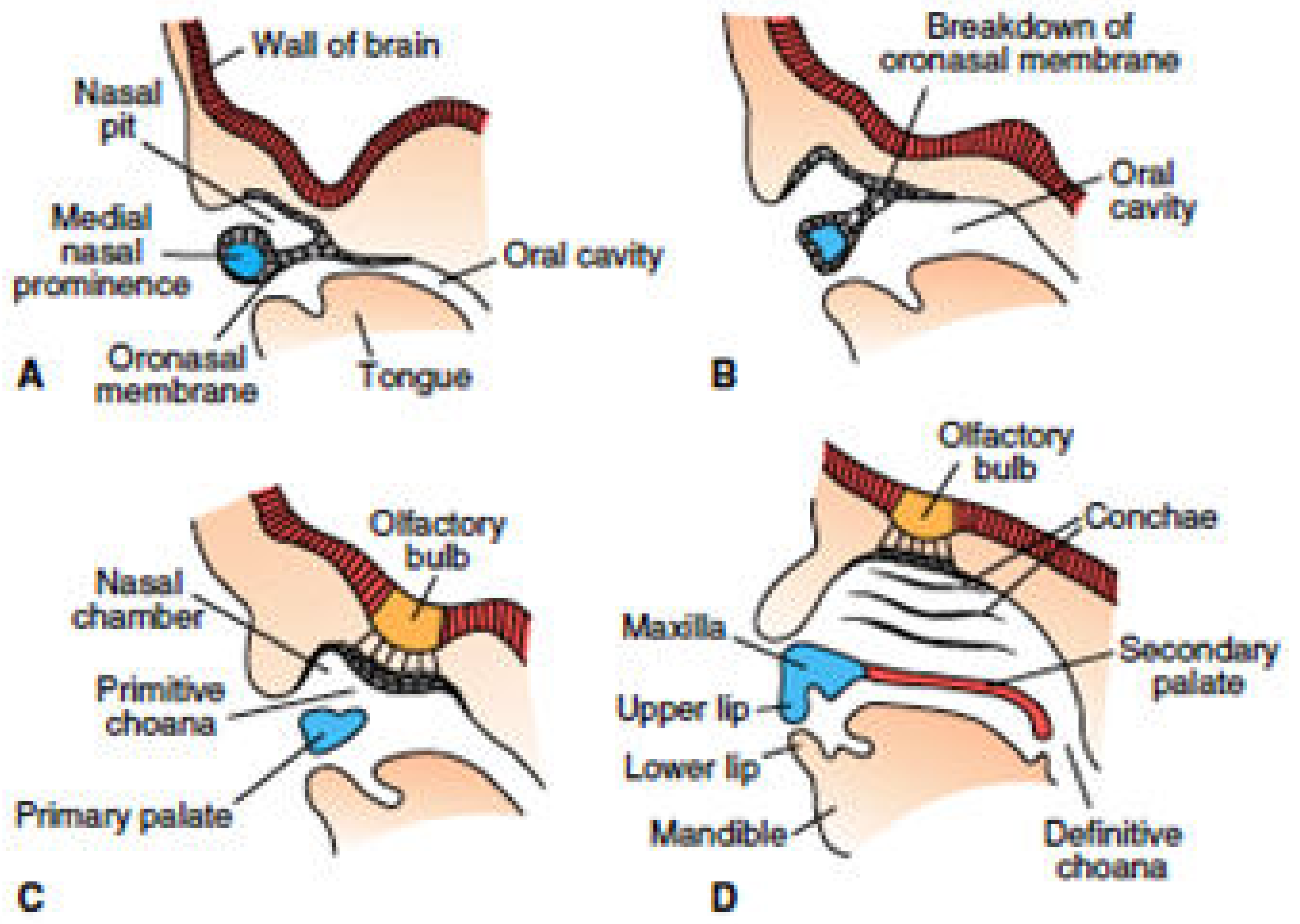
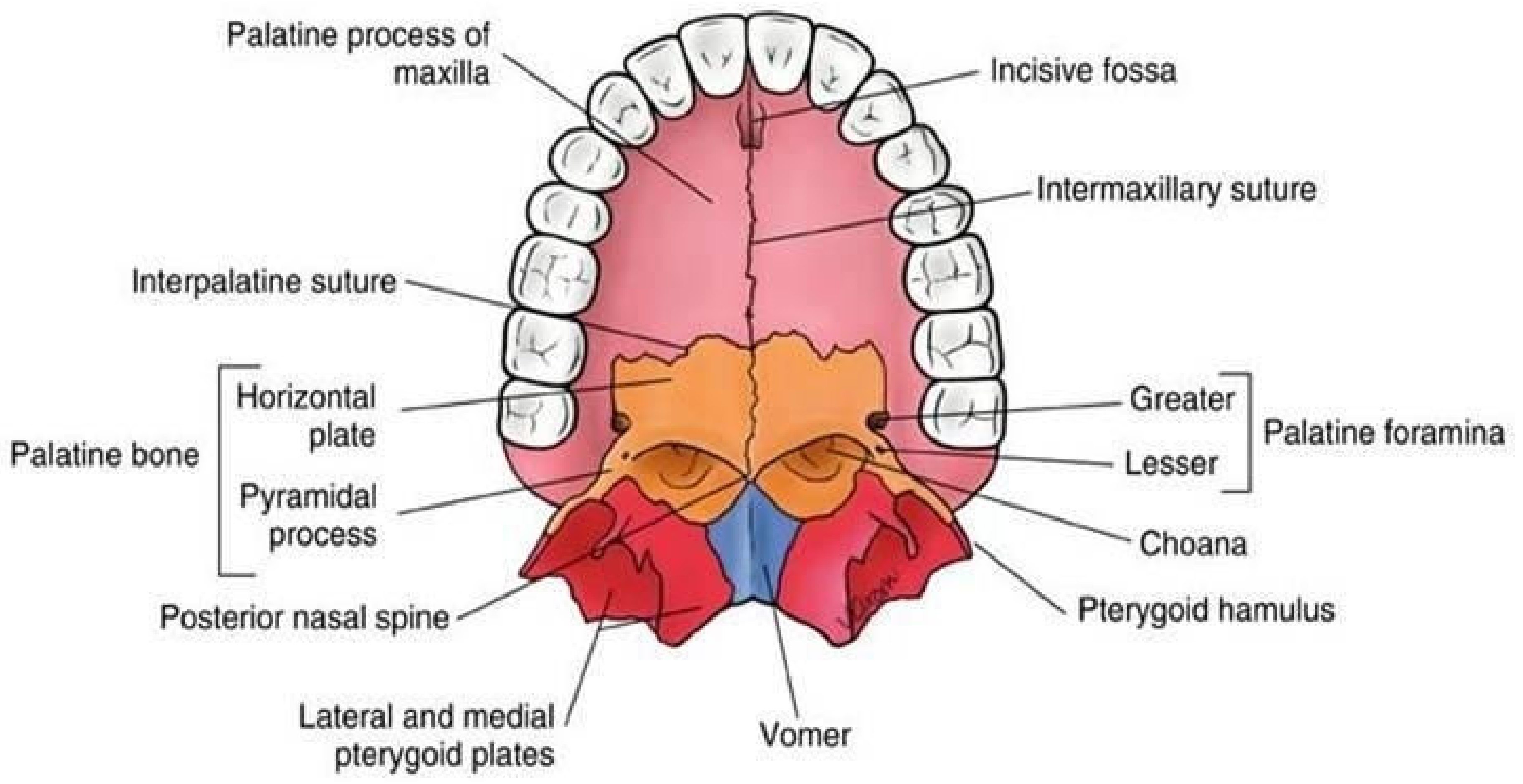
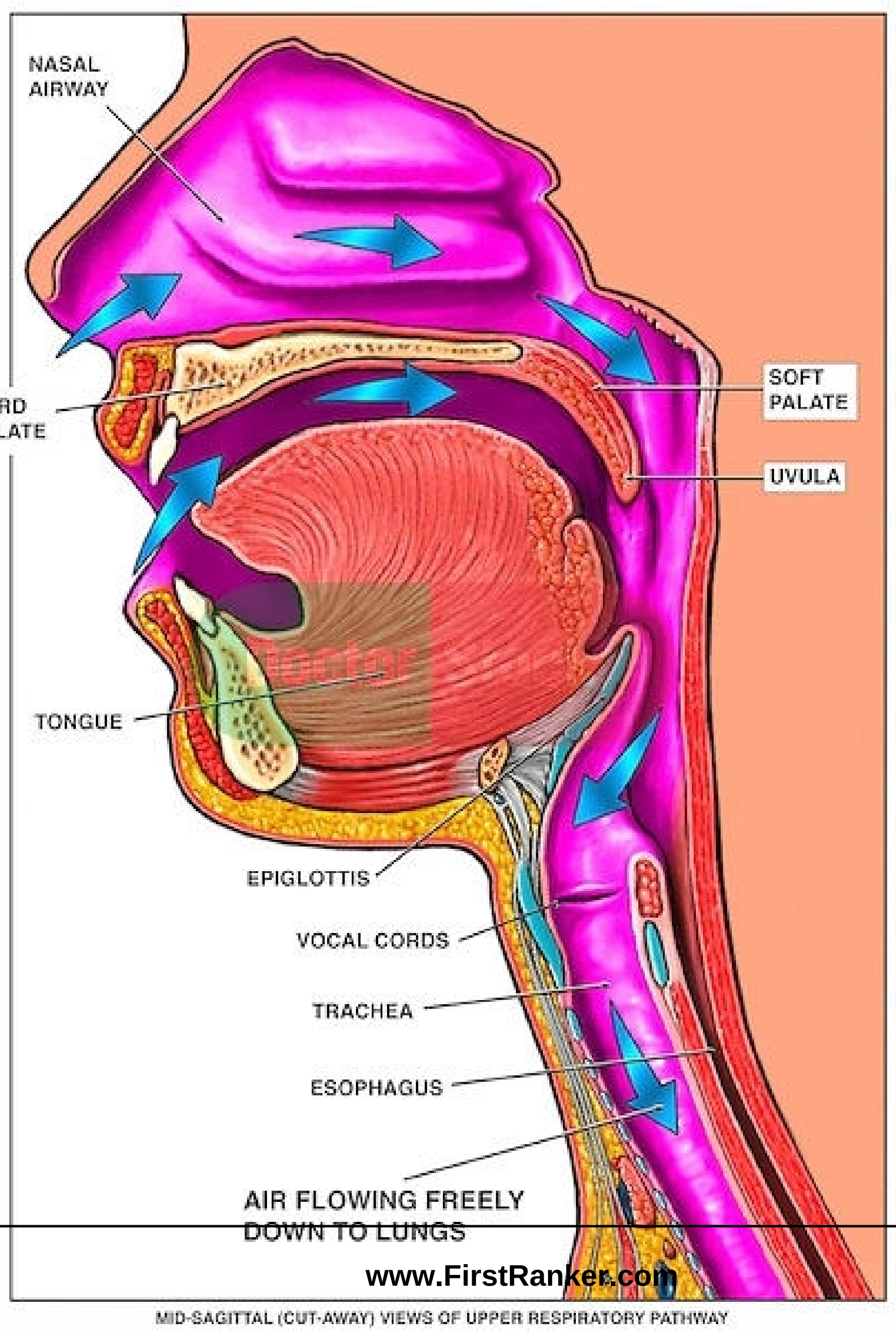


Figure 17.31 **A.** Sagittal section through the nasal pit and lower rim of the medial nasal prominence of a 6-week embryo. The primitive nasal cavity is separated from the oral cavity by the oronasal membrane. **B.** Similar section as in **A** showing the oronasal membrane breaking down. **C.** A 7-week embryo with a primitive nasal cavity in open connection with the oral cavity. **D.** Sagittal section through the face of a 9-week embryo showing separation of the definitive nasal and oral cavities by the primary and secondary palate. Definitive choanae are at the junction of the oral cavity and the pharynx.





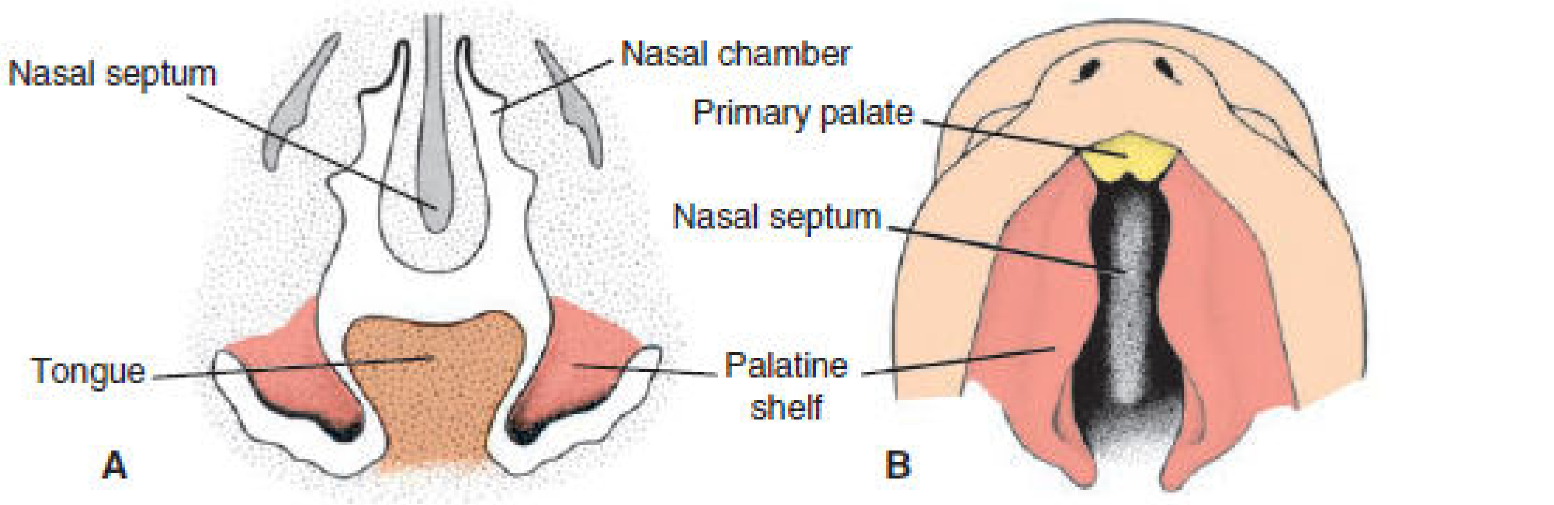


Figure 17.25 **A.** Frontal section through the head of a 6.5-week embryo. The palatine shelves are in the vertical position on each side of the tongue. **B.** Ventral view of the palatine shelves after removal of the lower jaw and the tongue. Note the clefts between the primary triangular palate and the palatine shelves, which are still vertical.

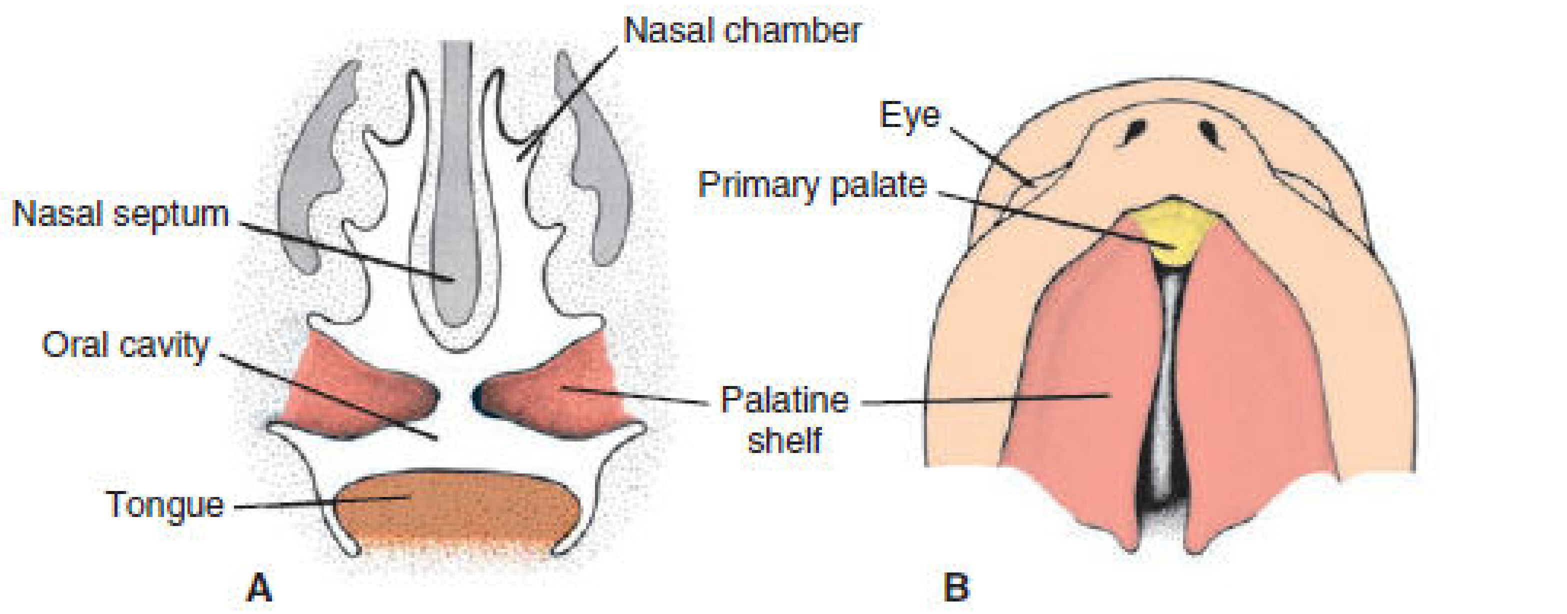


Figure 17.26 **A.** Frontal section through the head of a 7.5-week embryo. The tongue has moved downward, and the palatine shelves have reached a horizontal position. **B.** Ventral view of the palatine shelves after removal of the lower jaw and tongue. The shelves are horizontal. Note the nasal septum.

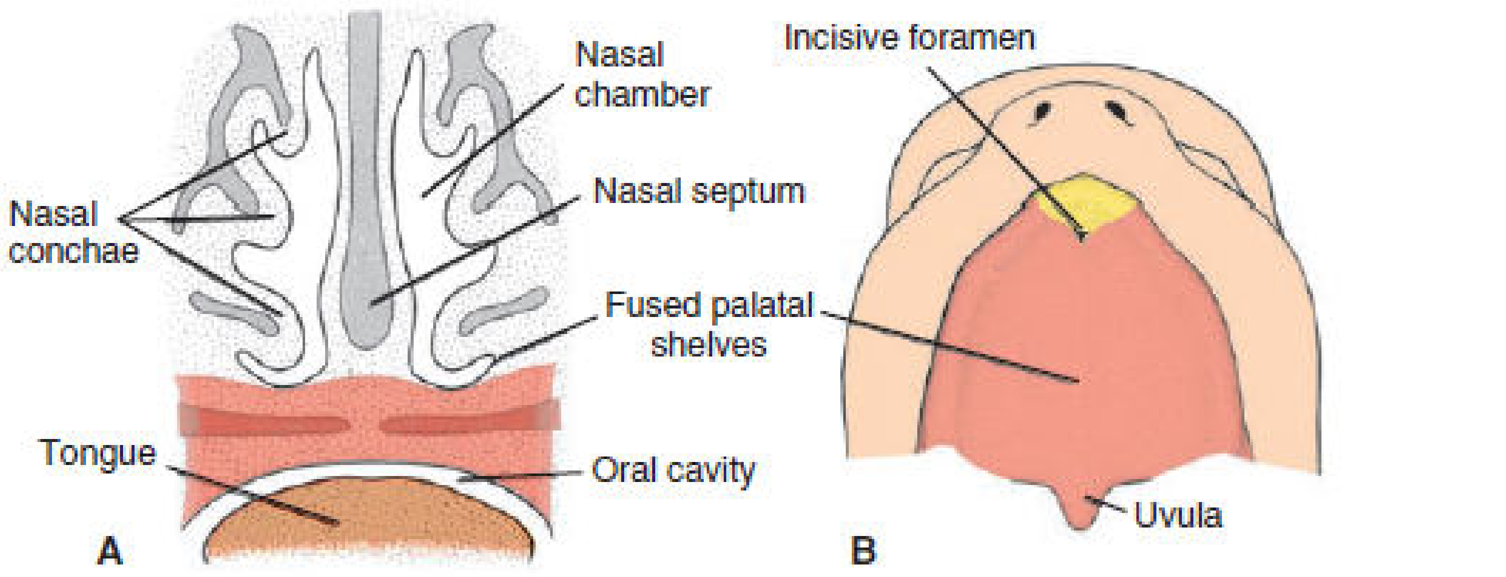


Figure 17.27 **A.** Frontal section through the head of a 10-week embryo. The two palatine shelves have fused with each other and with the nasal septum. **B.** Ventral view of the palate. The incisive foramen forms the midline between the primary and secondary palate.

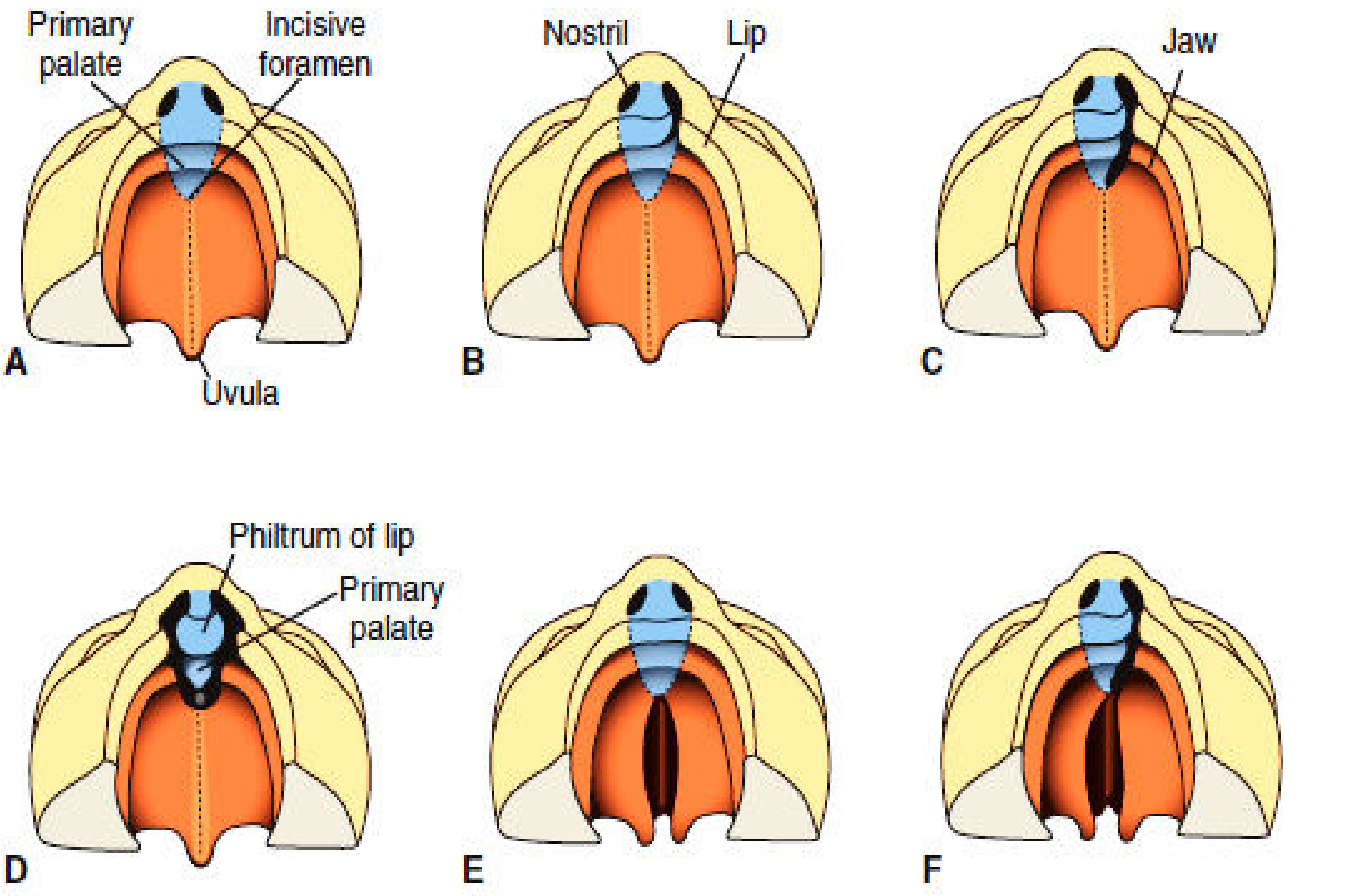


Figure 17.28 Ventral view of the palate, gum, lip, and nose. **A.** Normal. **B.** Unilateral cleft lip extending into the nose. **C.** Unilateral cleft involving the lip and jaw and extending to the incisive foramen. **D.** Bilateral cleft involving the lip and jaw. **E.** Isolated cleft palate. **F.** Cleft palate combined with unilateral anterior cleft lip.

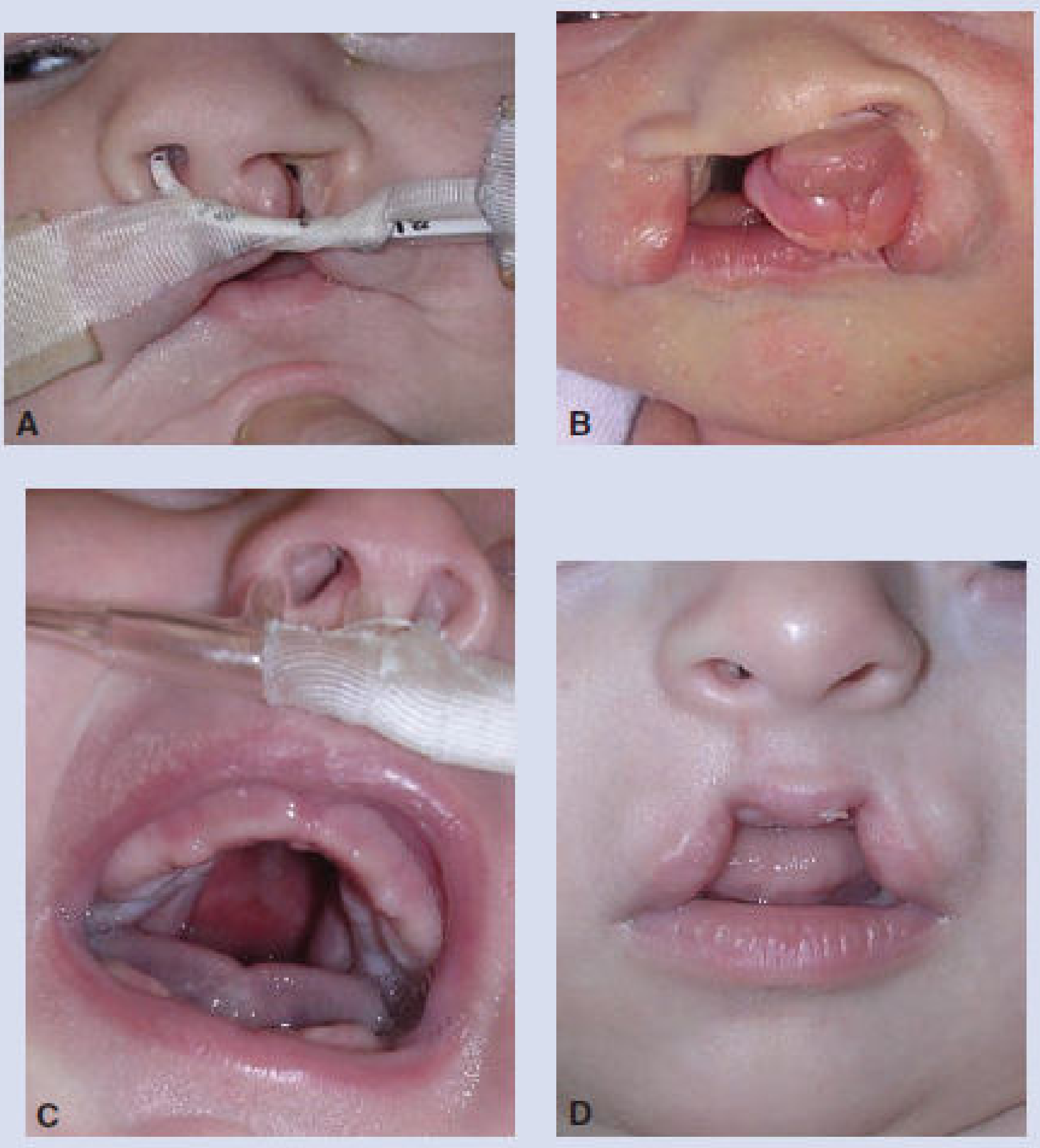


Figure 17.29 **A.** Lateral Cleft lip (compare to Fig. 17.28C). **B.** Bilateral cleft lip (compare to Fig. 17.28D). **C.** Cleft palate (compare to Fig. 17.28E). **D.** Midline cleft lip

MACROSTOMIA

As a resident in paediatrics, you are called to see a newborn who has a unilateral cleft lip and a unilateral cleft of the primary plate. This condition is most likely the result of:
a) failure of fusion of the mandibular prominence
b) failure of fusion of the medial nasal processes
c) failure of fusion of the maxillary prominence with the medial nasal prominence
d) failure of fusion of the lateral palatine processes with the nasal septum

Cleft Lip & Palate

The term cleft lip & palate is commonly used to represent two types of malformation

- cleft lip with or without cleft palate (CL/P)(fig.1)
- cleft palate (CP) (fig.2)

Bilateral cleft of lip & palate

(fig.1)

cleft of the hard & soft palates

(fig.2)

Unilateral cleft of lip

(fig.3)

cleft of lip & palate

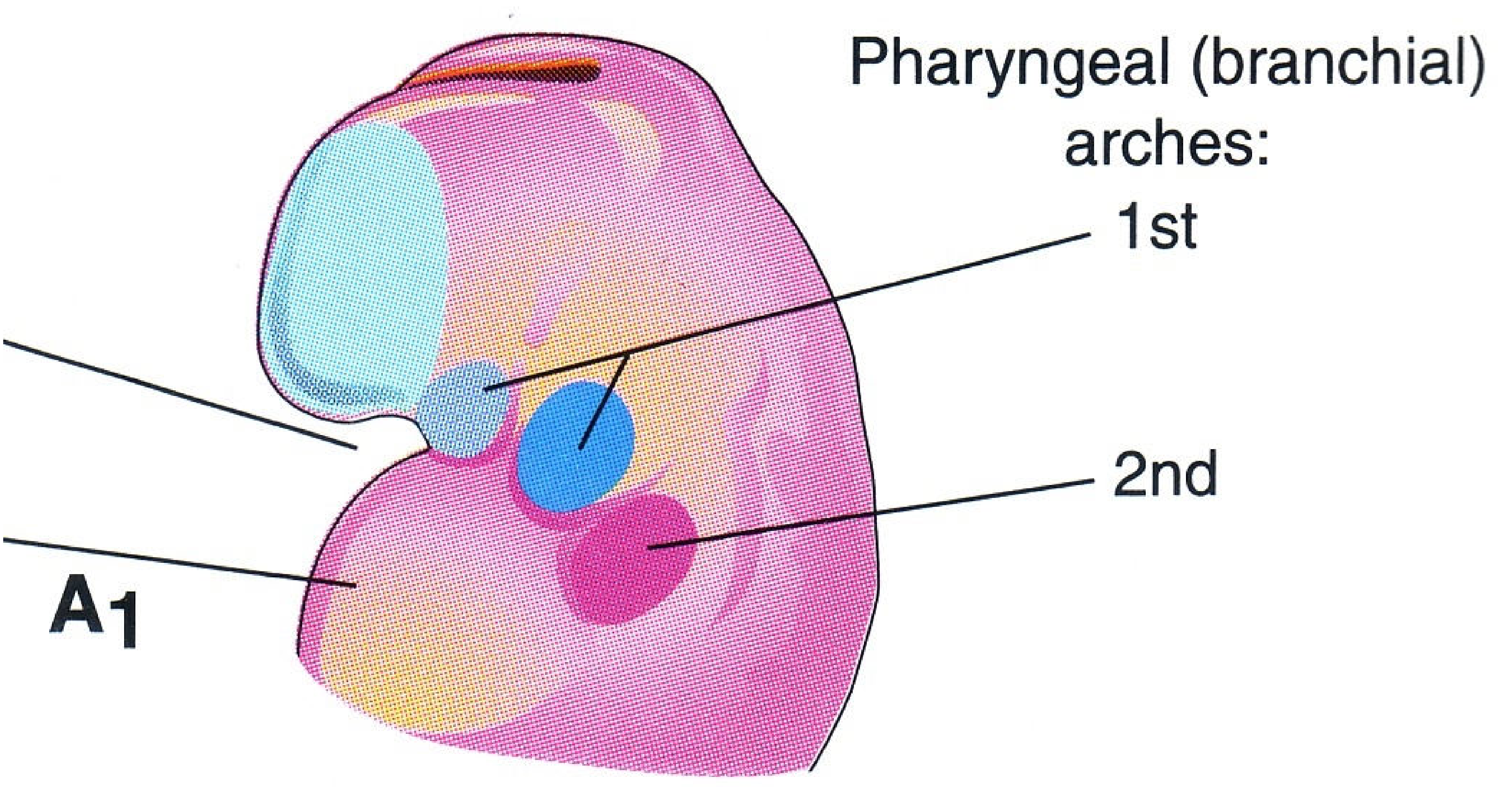
(fig.4)

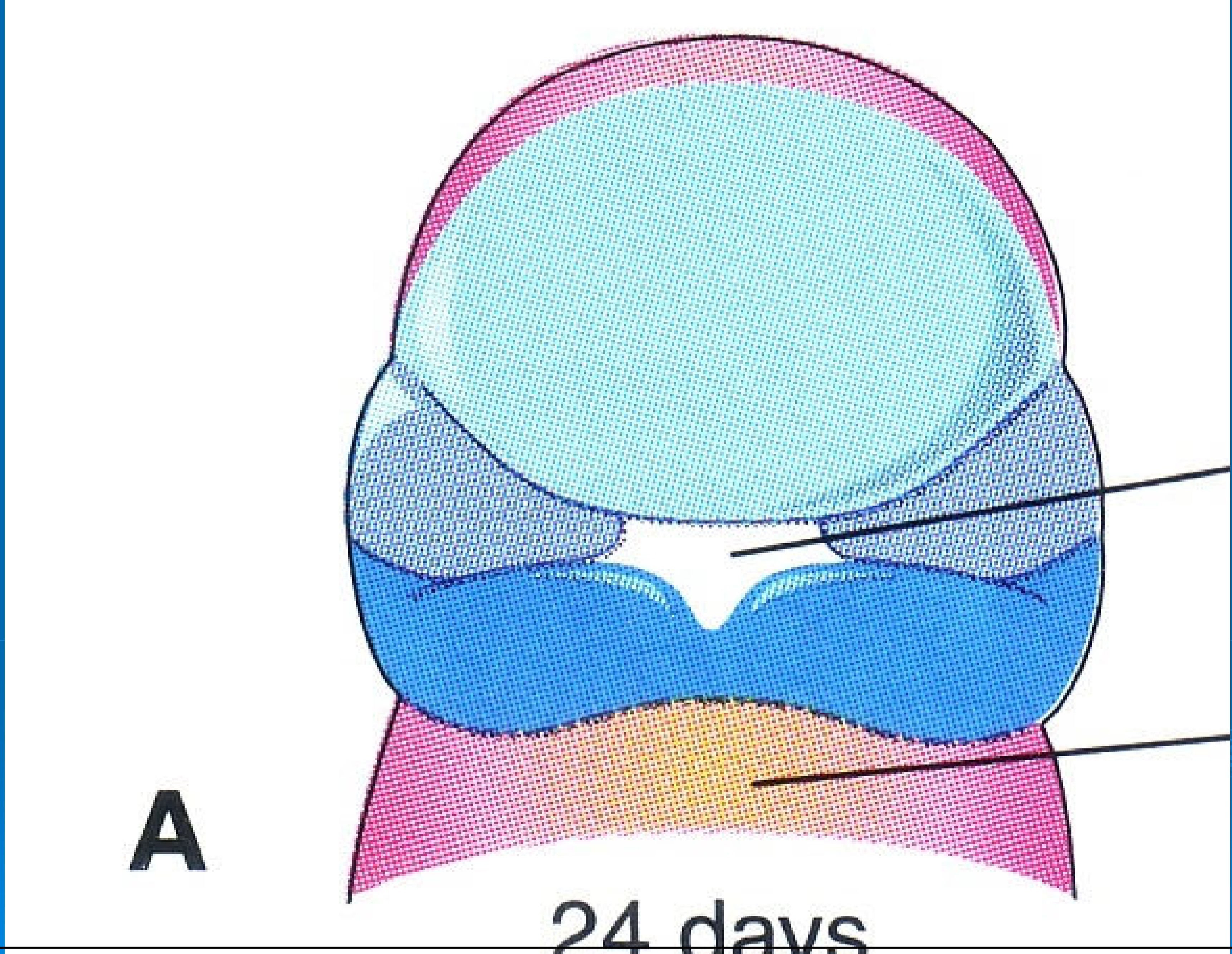
Bifid uvula

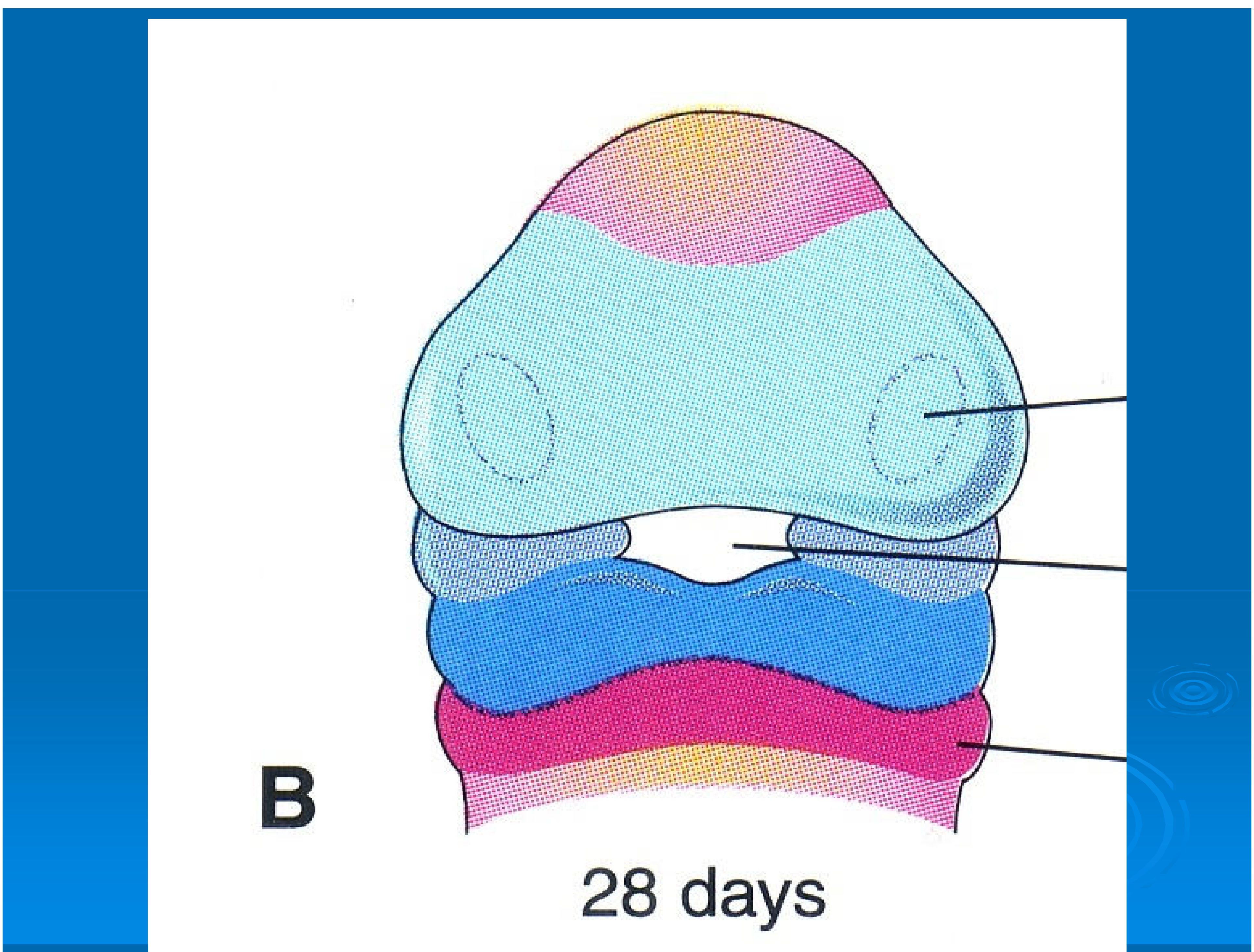
(fig.5)

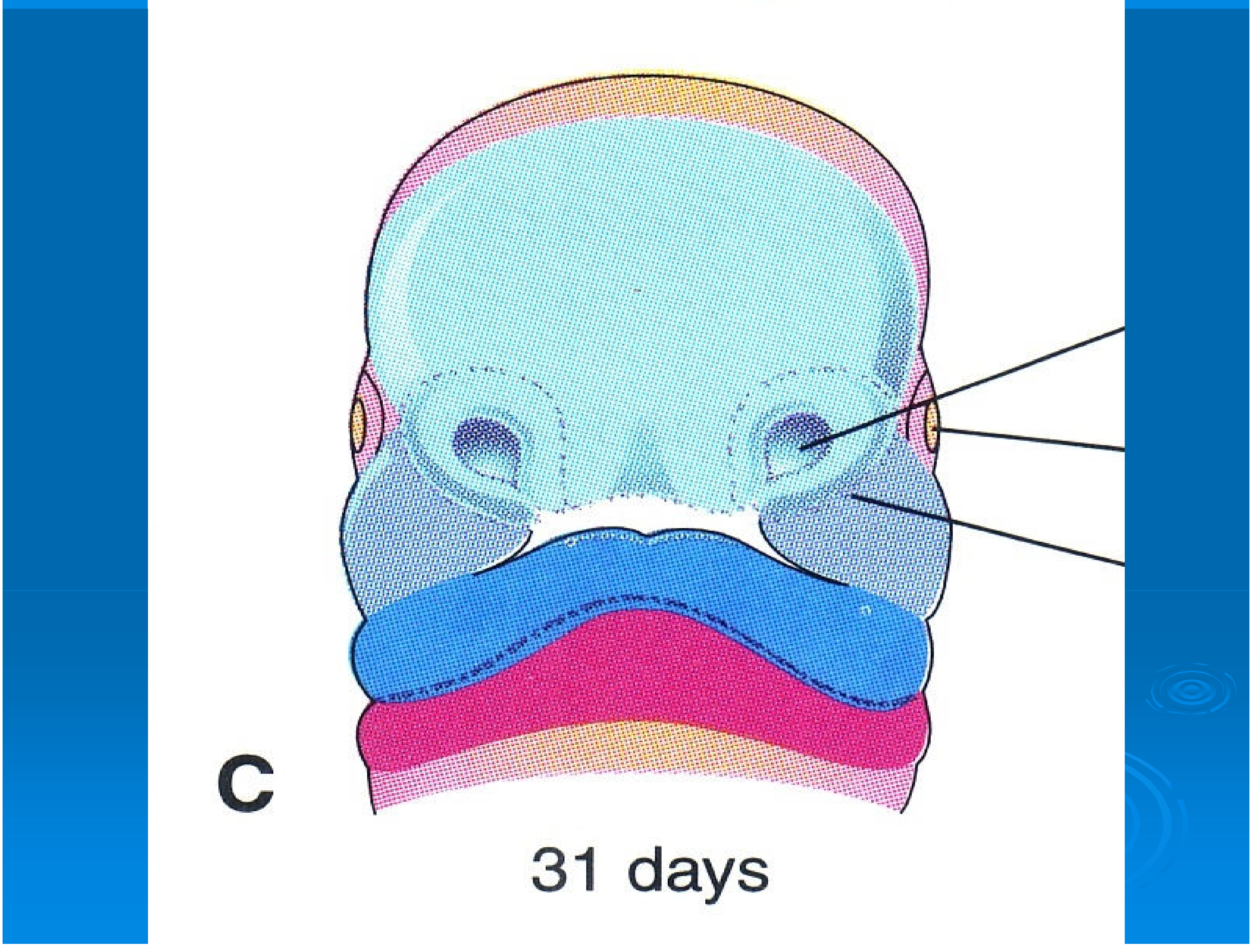
Congenital Anomalies of Face & Palate

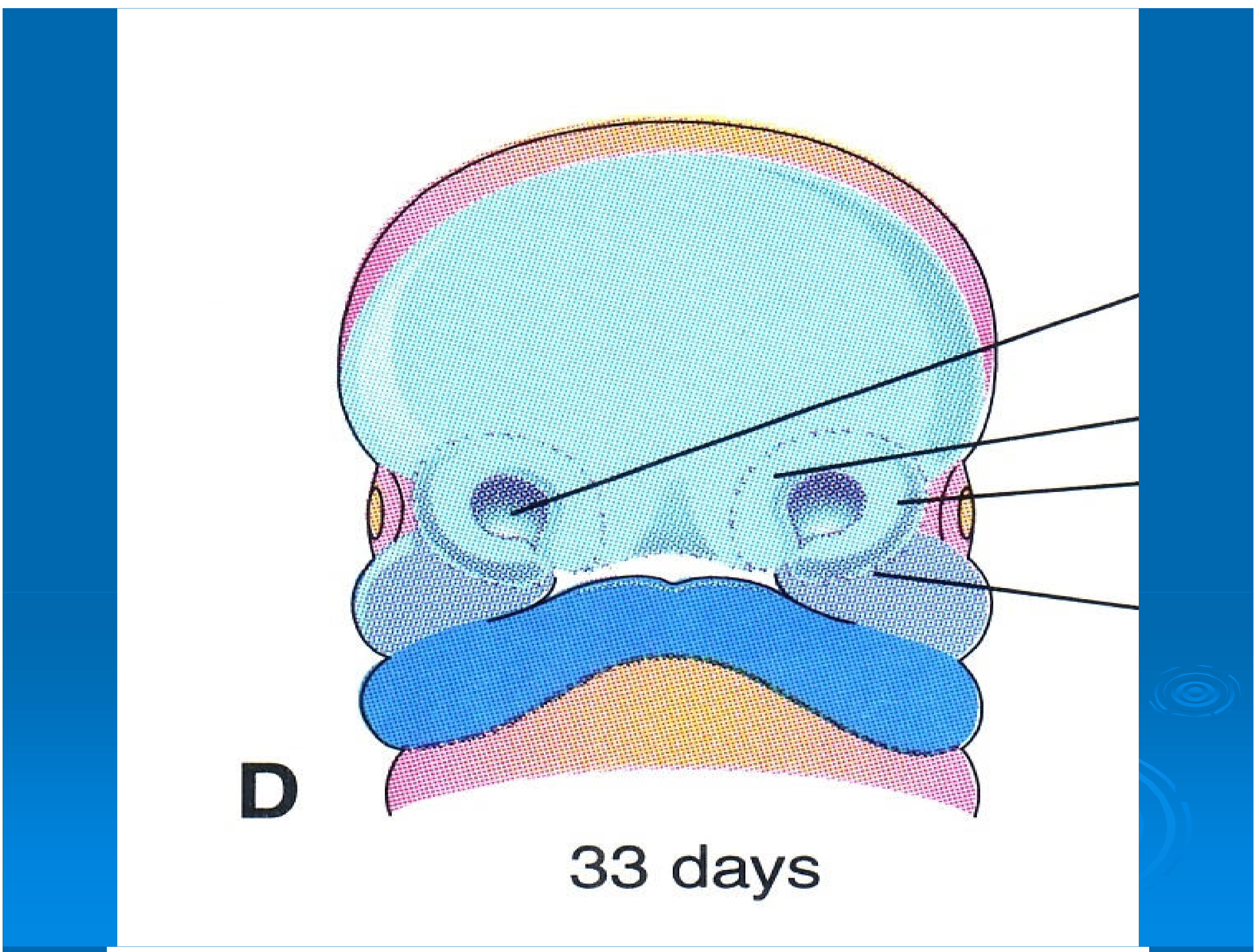
- 1- Inclusion dermoid: cystic swelling at line of fusions between processes of the face.
- 2- Oblique facial cleft: failure of fusion between maxillary and fronto-nasal processes.
- 2- Macrostomia or Microstomia : defective or marked fusion between maxillary and mandibular processes.
- 3- Cleft (hare) lip: cleft lip due to failure of fusion between maxillary process and intermaxillary segment.
- 4- Cleft palate: failure of fusion between different parts that form palate. It could be:
  - A- Unilateral complete cleft palate.
  - B- Bilateral complete cleft palate.
  - C- Partial cleft palate.
  - D- Cleft uvula.

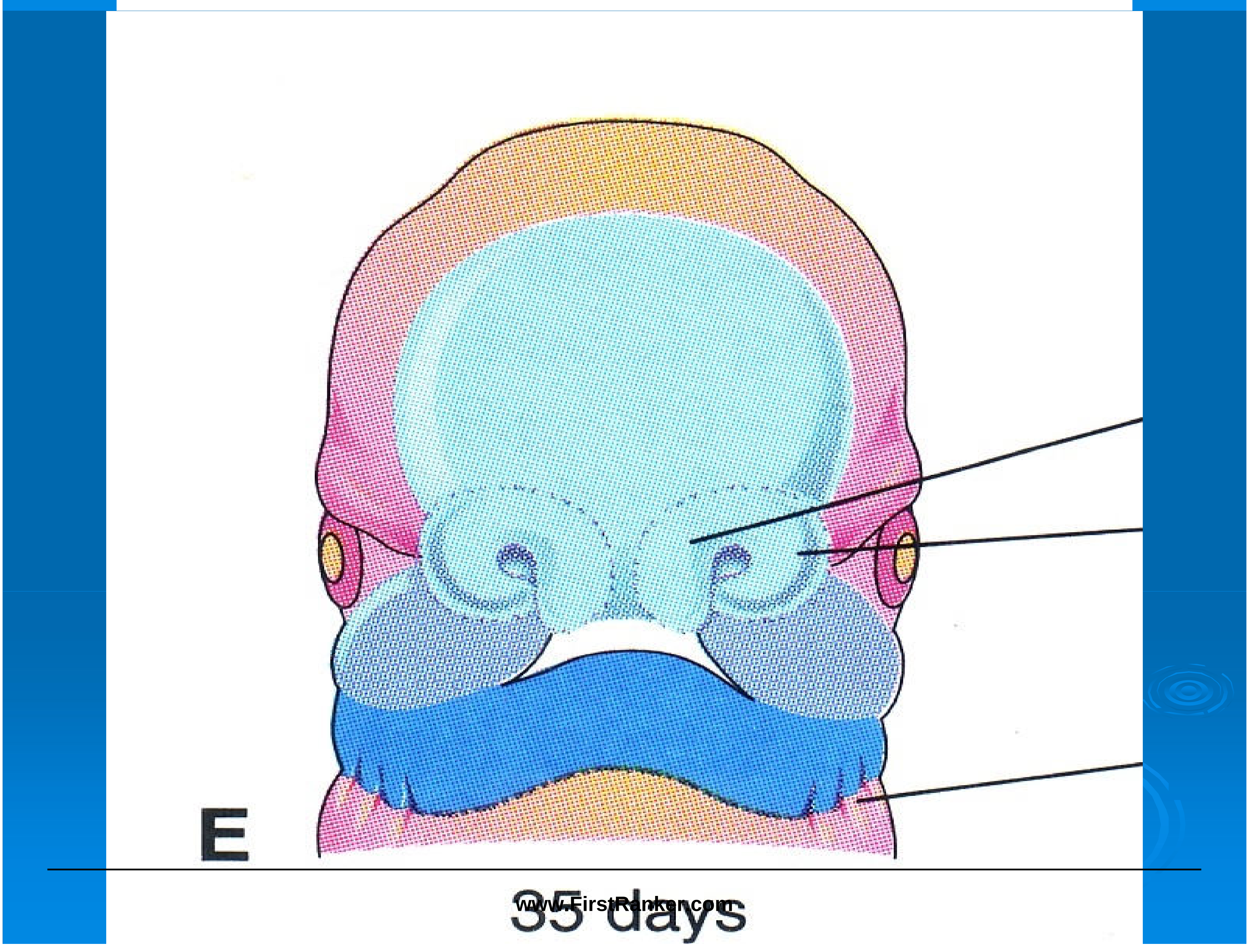


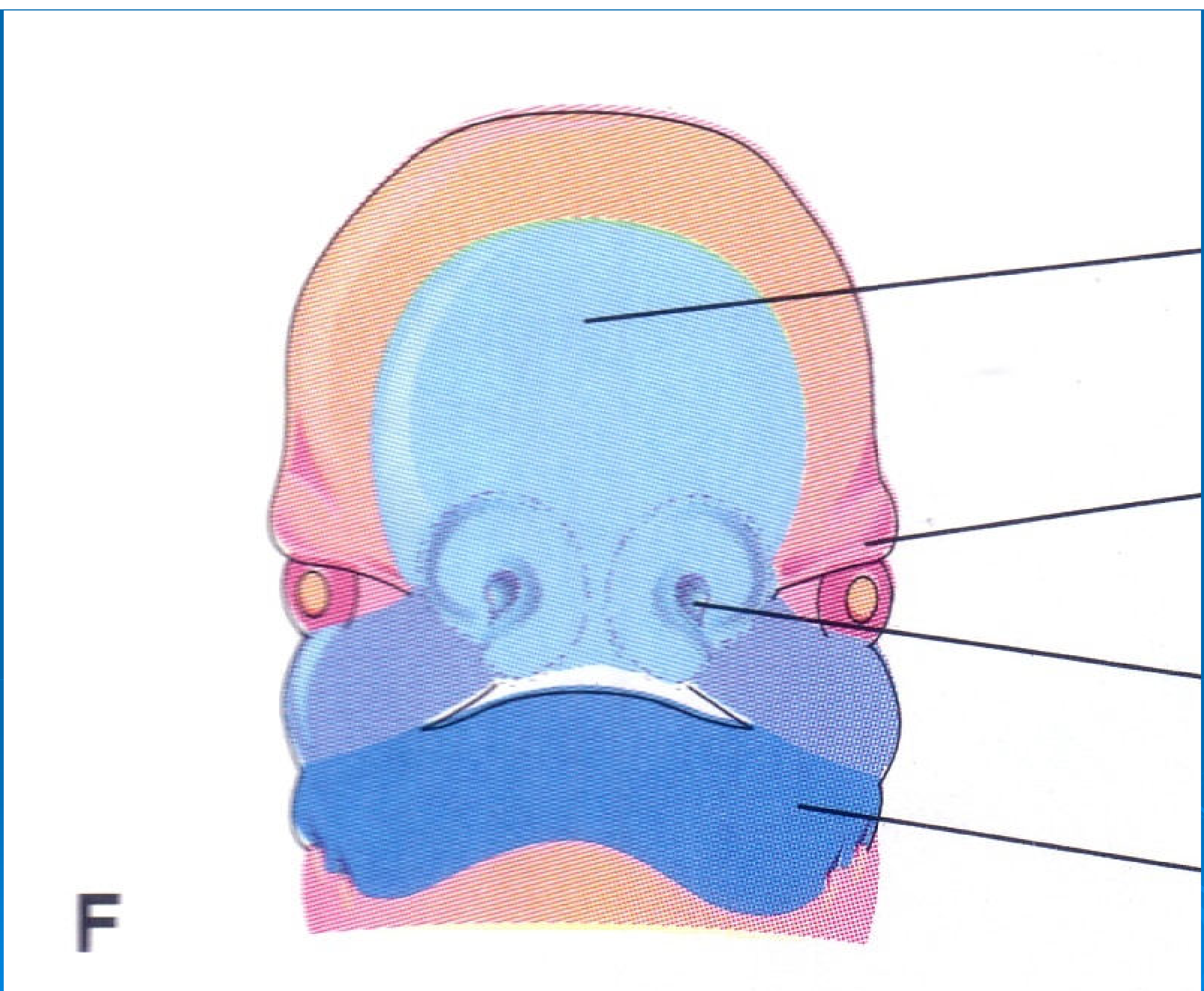



B
28 days


C
31 days

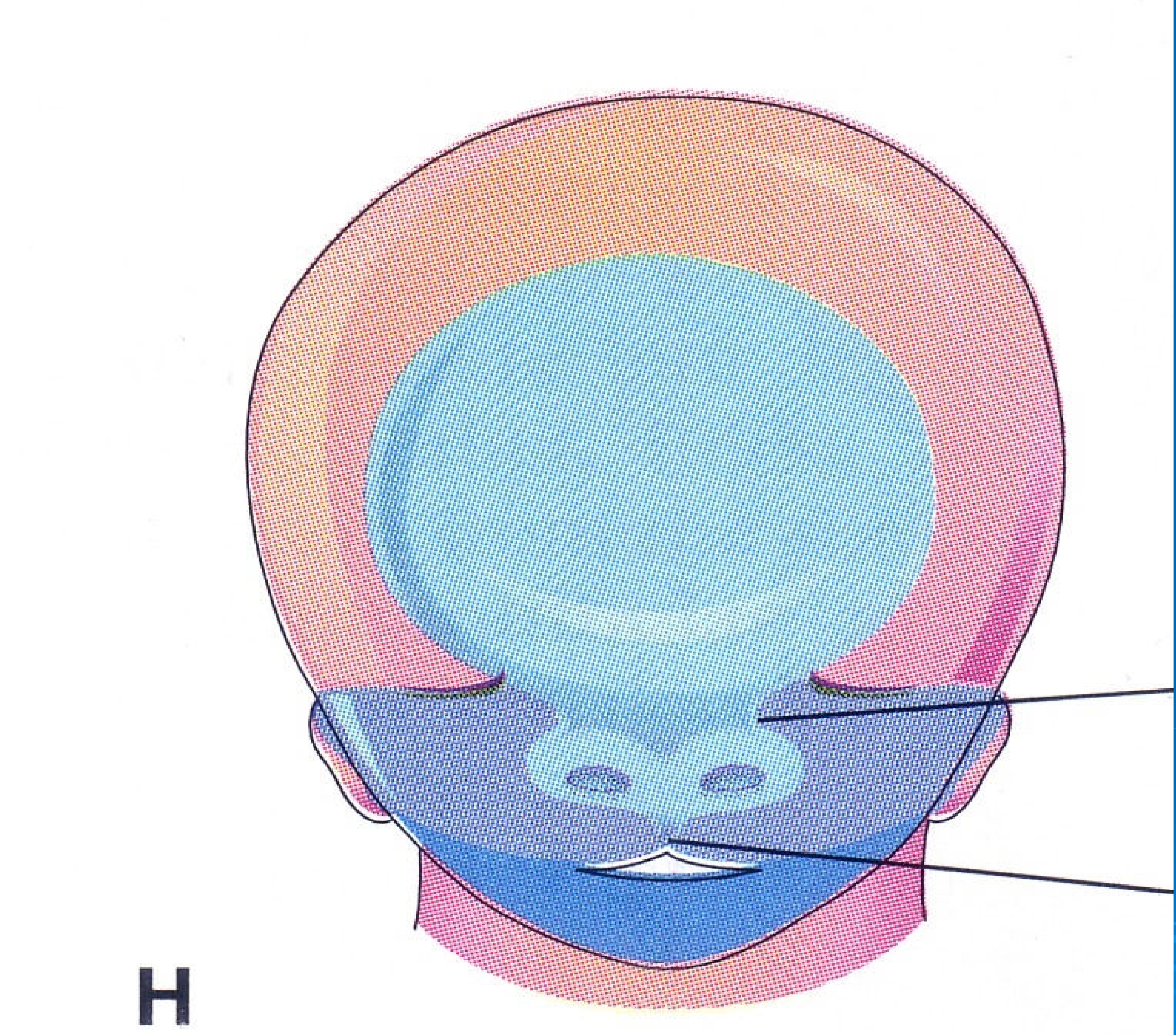

D
33 days


E
35 days



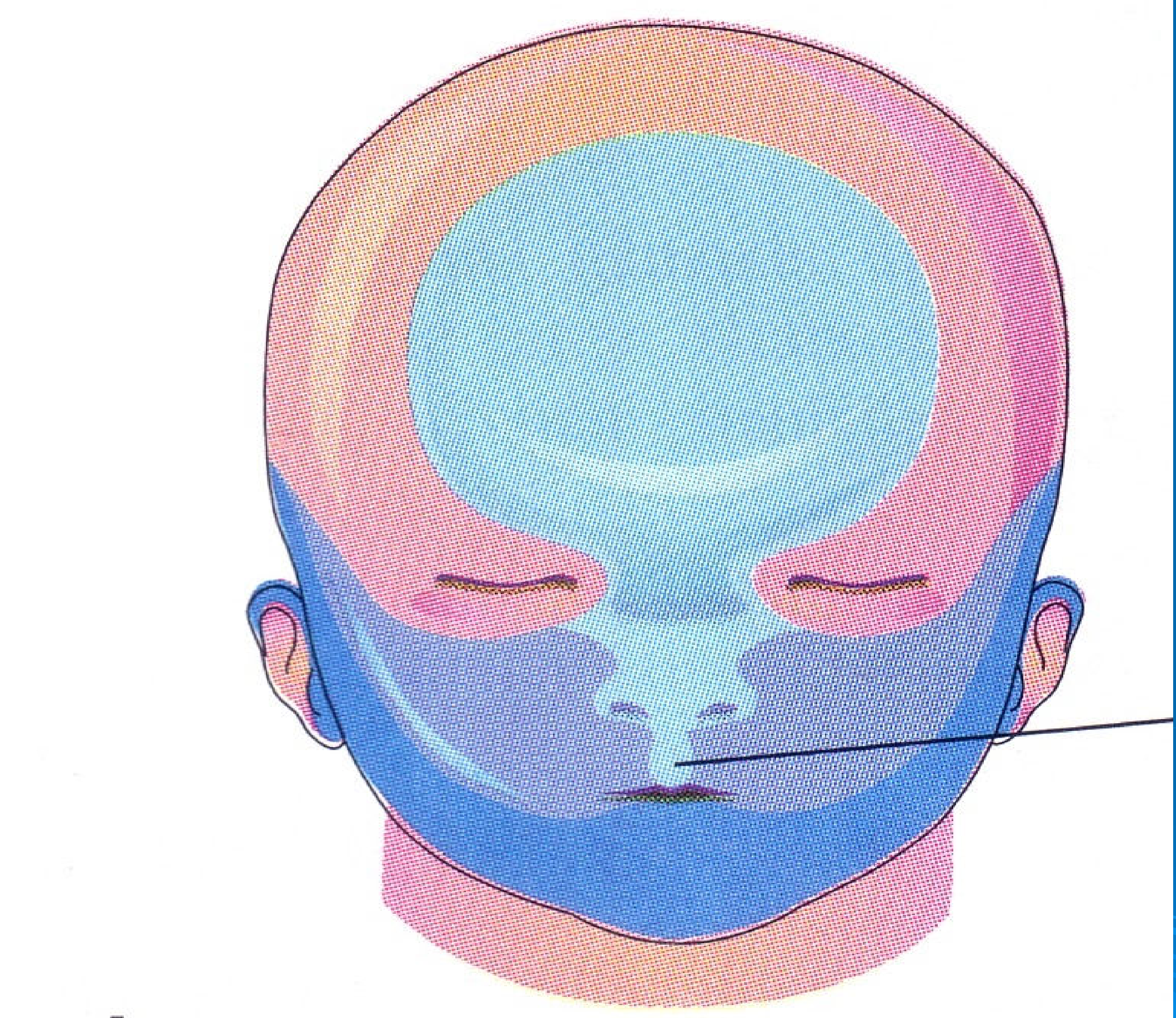
F

40 days



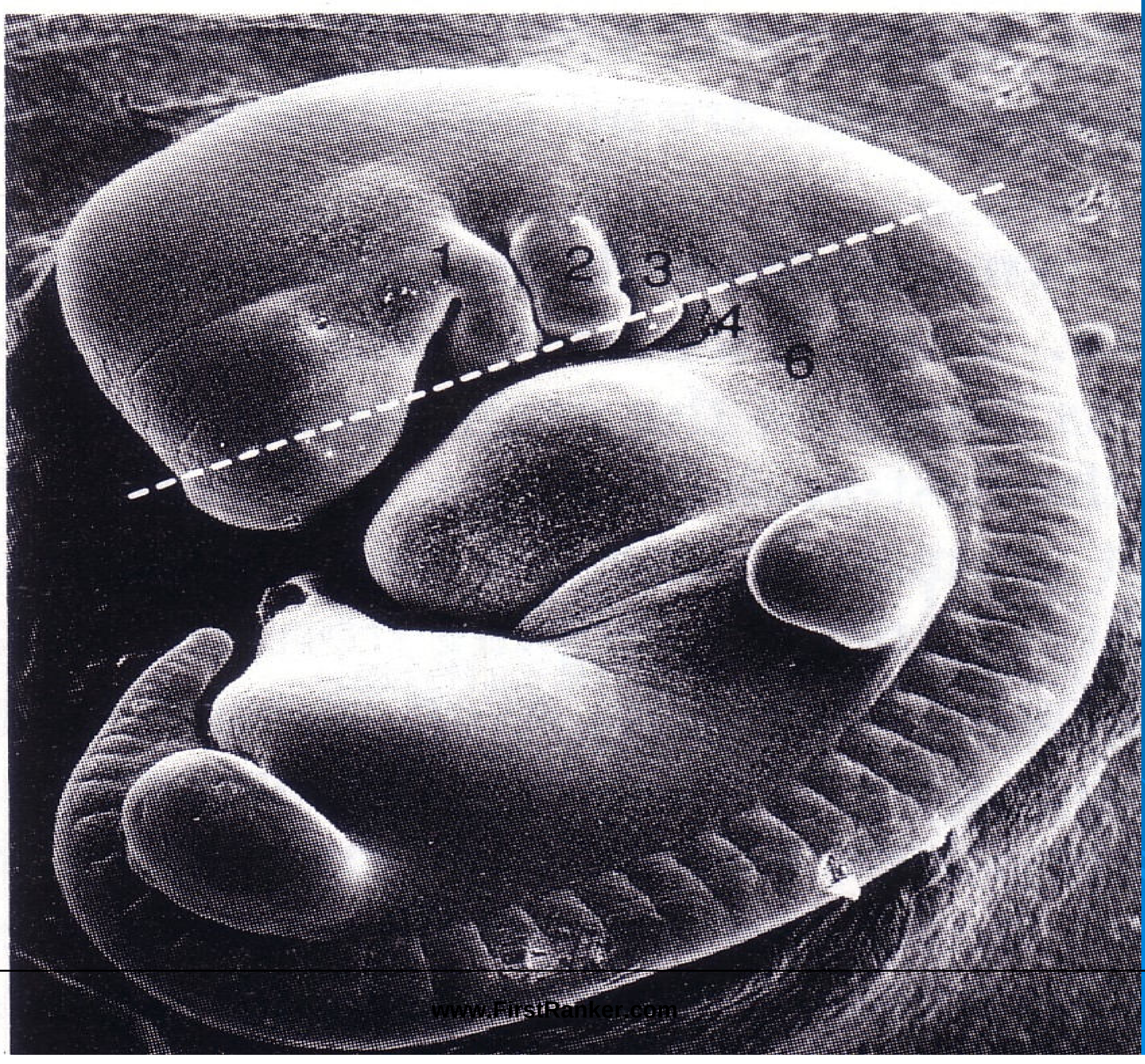
H

10 weeks

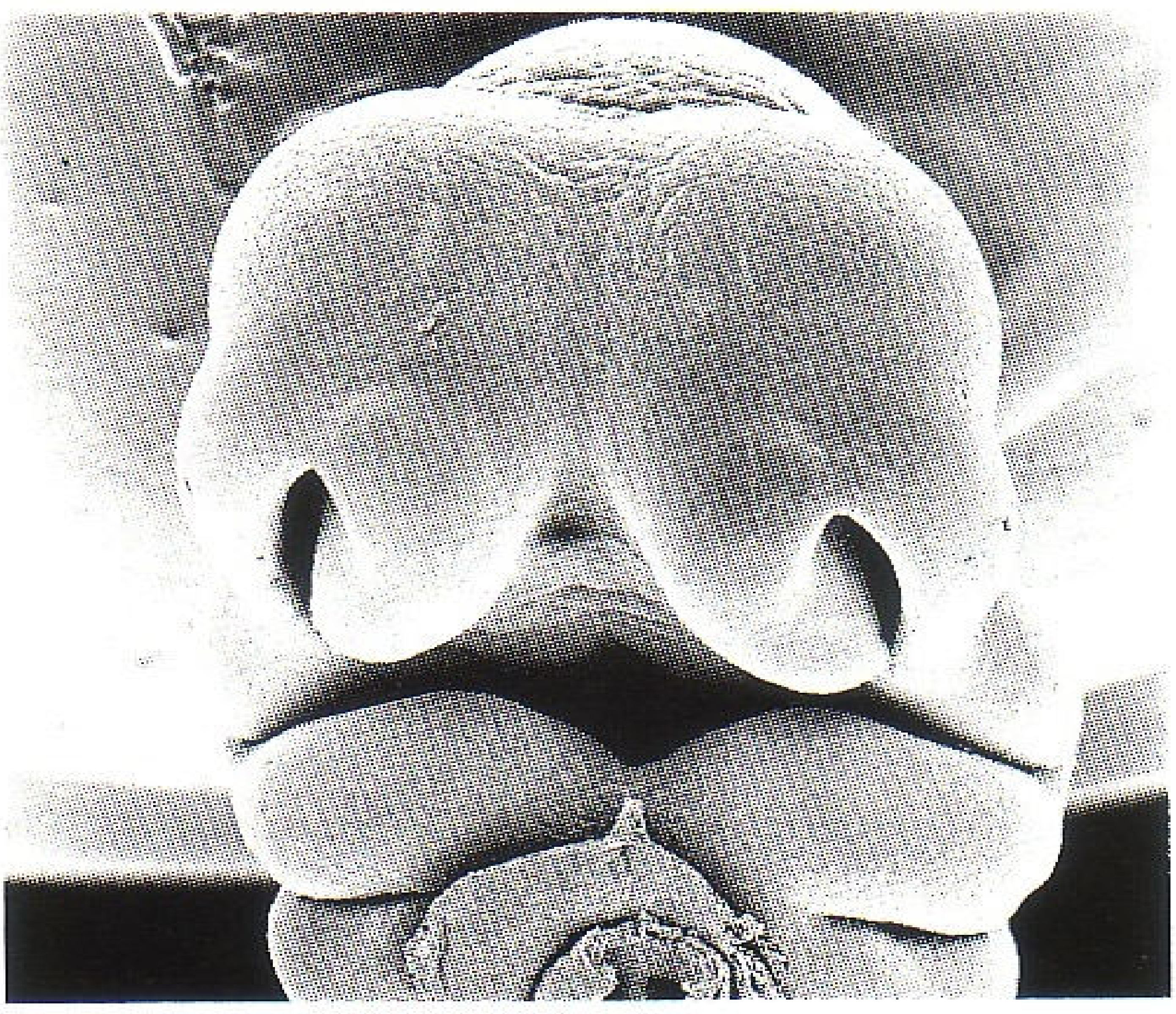


I

14 weeks



A 4th week



A Early 6th week

OTHER CLEFTS

- Lack of fusion of maxillary & mandibular processes ➔ Lateral facial cleft
- May be associated with 'Mandibulo facial dysostosis'
- Extends from commissures towards the ear
- May be unilateral or bilateral
- Failure of fusion of lateral nasal process with maxillary process ➔ oblique facial cleft

Dr. Ali Tahir

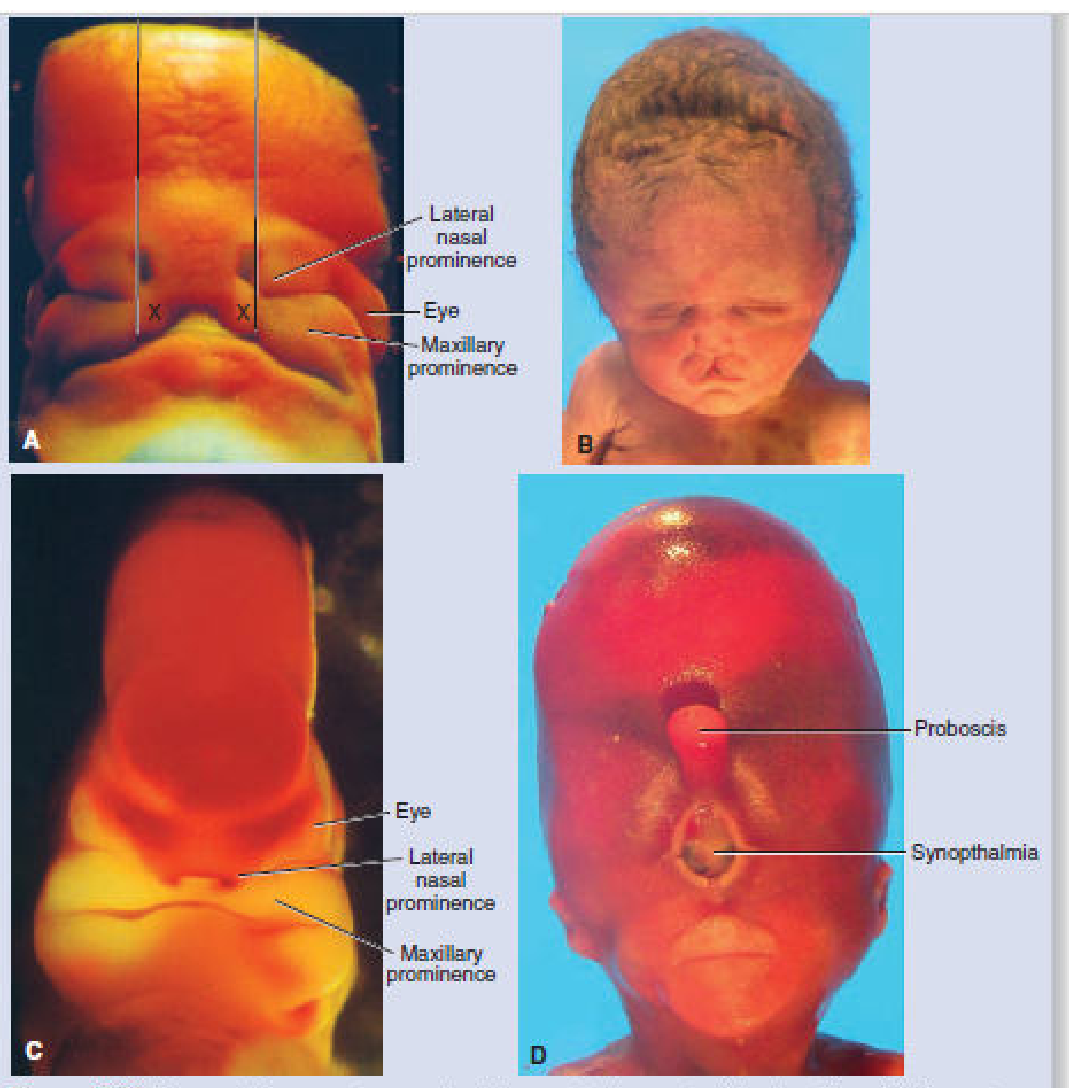


Figure 17.30 Photographs demonstrating normal and abnormal development involving the midline of the face and brain. **A.** Normal face of an embryo at the beginning of the sixth week. Note the distance between the nasal openings (*vertical lines*) and between the medial nasal prominences (*Xs*). **B.** Face of a newborn with a midline cleft lip. In this case, there is insufficient tissue in the midline to fill in the space between the medial nasal prominences. **C.** Face of a 6-week embryo showing an extensive deficiency of midline tissue. As a consequence, the medial nasal prominences have not formed, and there is a single nasal opening created by fusion of the two lateral nasal prominences. The deficiency of midline tissue is reflected in the brain as well, and as a result, the head is narrow, and the eyes are positioned more anteriorly and slightly caudal to the nasal opening. **D.** Face of a newborn with holoprosencephaly characterized by such an extensive deficiency of midline tissue that the eyes have fused (synophthalmia), and a proboscis with a single nasal opening has formed from fusion of the lateral nasal processes. An upper lip is formed by fusion of the maxillary prominences. The head is narrow, and the brain wo[illegible] (alobar holoprosencephaly) due to such a loss of midline tissue that the two lateral ventricles fused. The condition may be caused by mutations in *sonic hedgehog (SHH)*, the gene that estab-

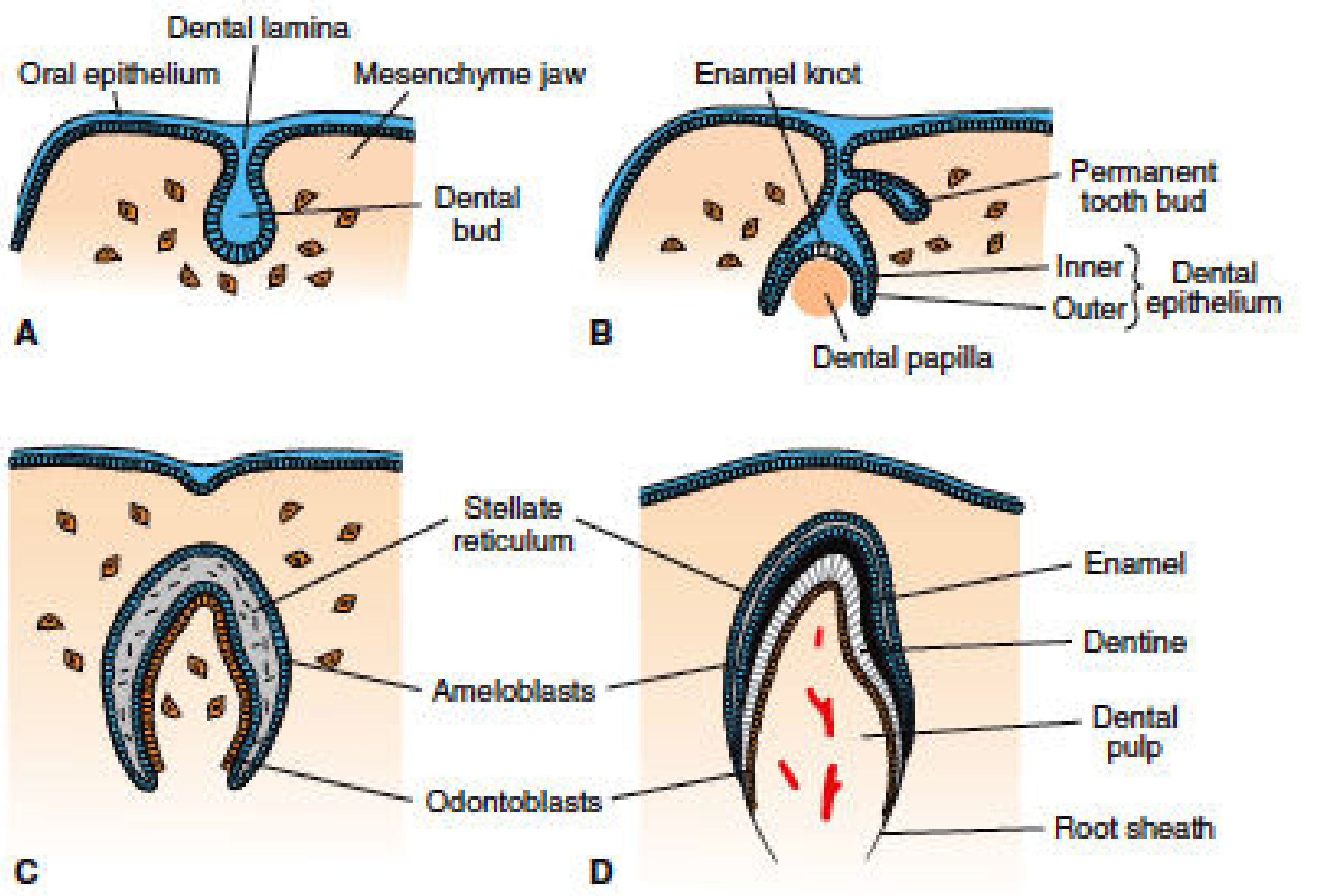


Figure 17.32 Formation of the tooth at successive stages of development. **A.** Bud stage; 8 weeks. **B.** Cap stage; 10 weeks. **C.** Bell stage; 3 months. **D.** 6 months.

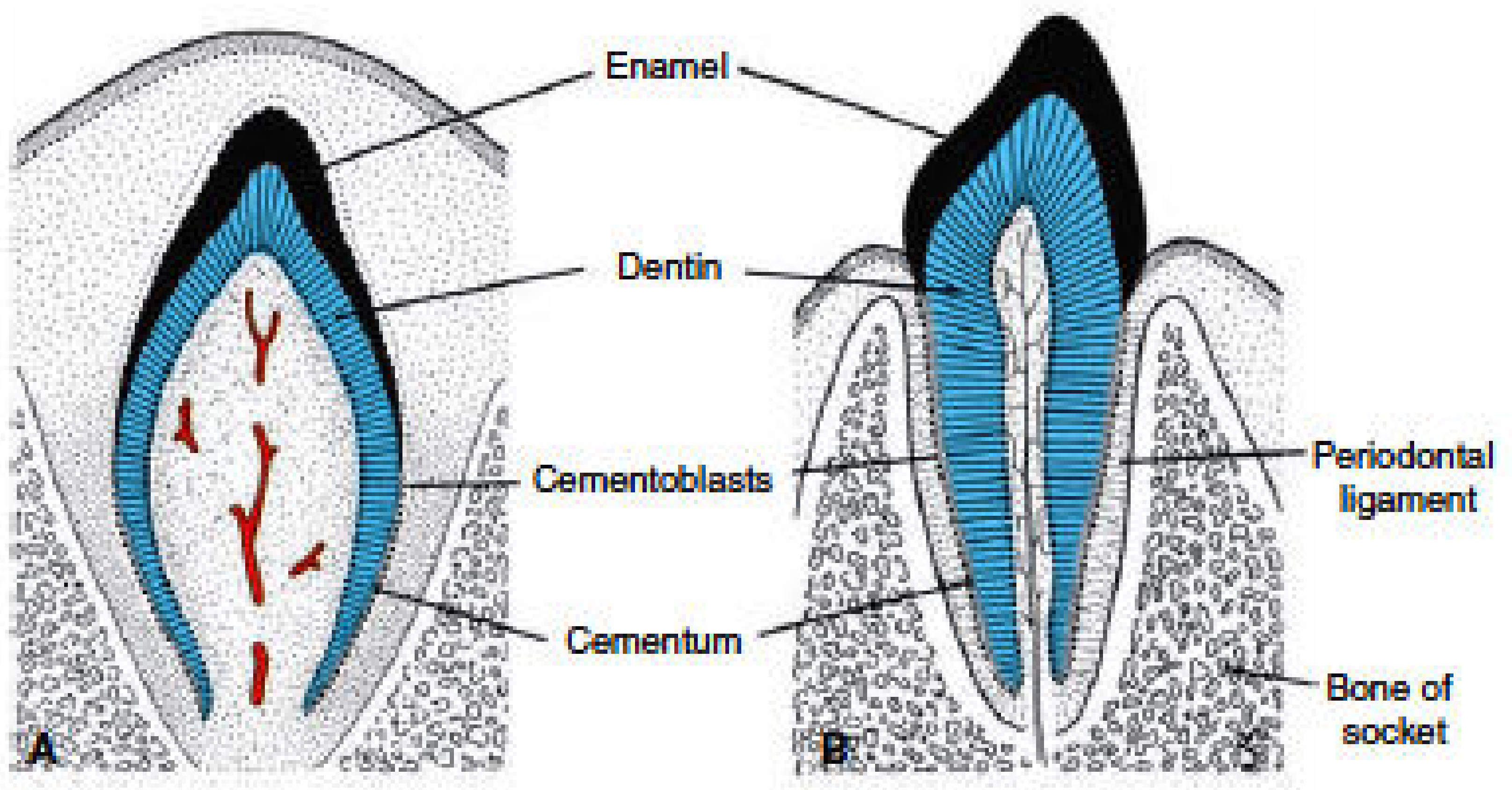
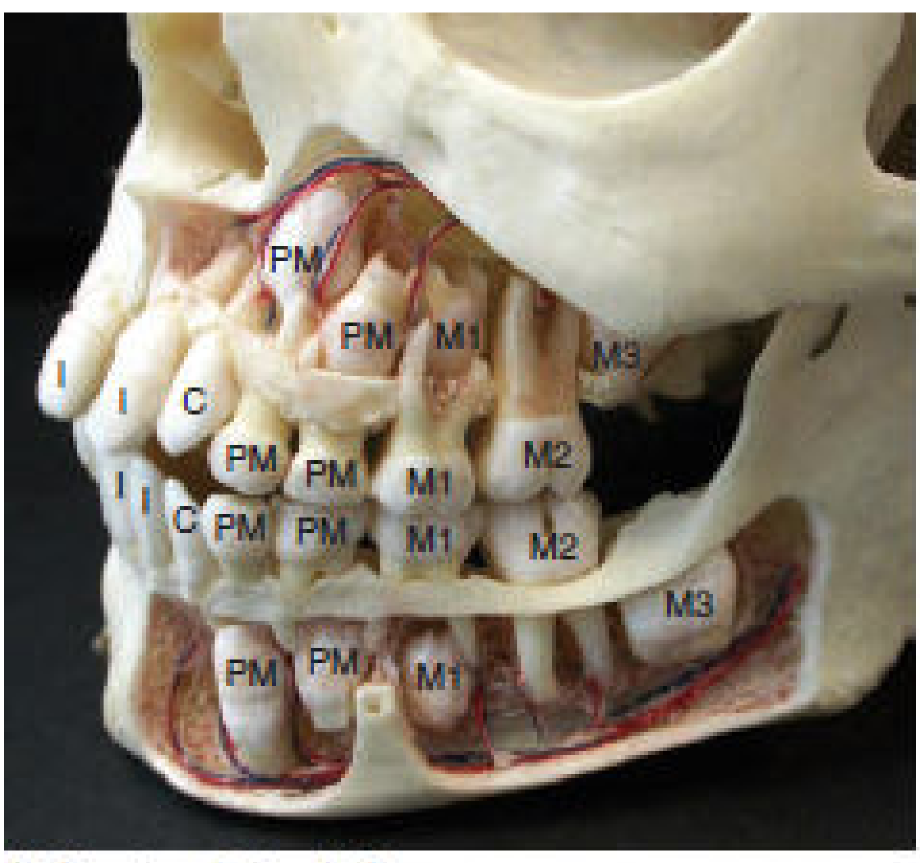


Figure 17.33 The tooth just before birth **A** and after eruption **B**.



Left anterolateral view

Figure 17.34 Replacement of deciduous teeth with permanent teeth in a child. I, incisor; C, canine; PM, premolar; M1, M2, M3; 1st, 2nd, and 3rd molars. (From Moore, KL and Dalley, AF. *Clinically Oriented Anatomy*, 5th ed. Figure 7.47B, p. 993. Lippincott Williams & Wilkins, Baltimore: 2006.)

Which pharyngeal arch is associated with Treacher Collins syndrome?

a) Pharyngeal arch 1
b) Pharyngeal arch 2
c) Pharyngeal arch 3
d) Pharyngeal arch 4

The stapedius muscle that moves the stapes ossicle is innervated by
a) CN V3
b) CN XII
c) CN III
d) CN VII

The Palatine tonsil and Tonsilar fossa are derivatives from which pharyngeal pouch?
a. Pharyngeal pouch 1
b. Pharyngeal pouch 2
c .Pharyngeal pouch 3
d. Pharyngeal pouch 4