Nervous System

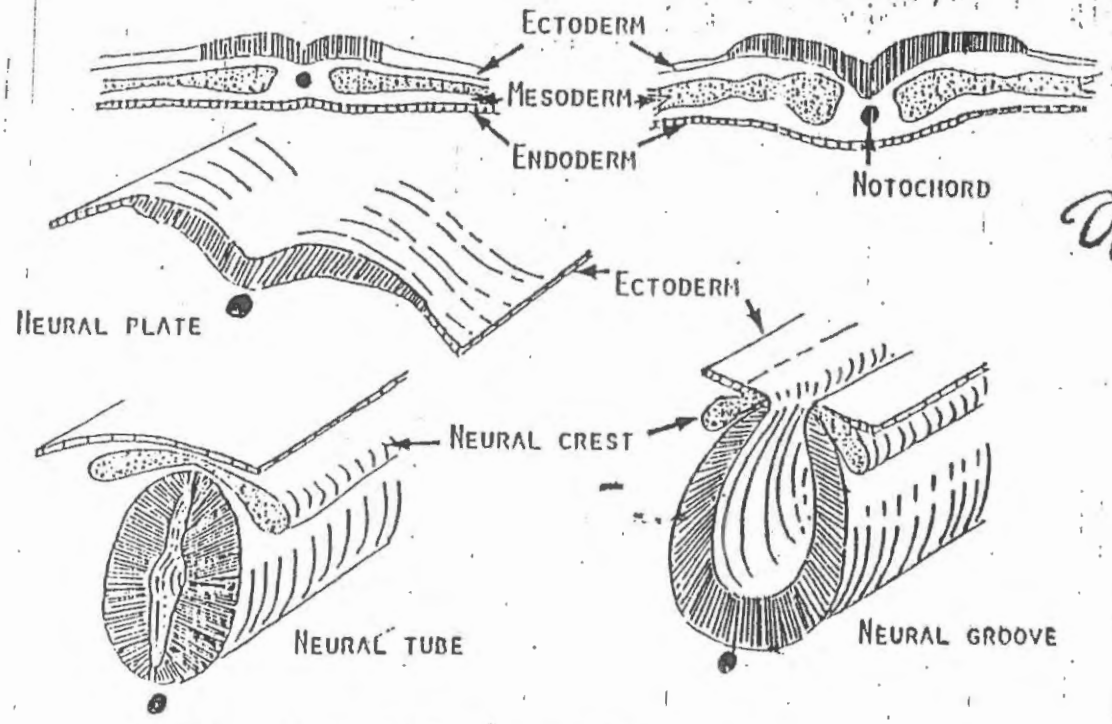

Fig. 21-1. Formation of neural tube.

NEURAL TUBE FORMATION

I. At the beginning of the third week, under the inductive influence of the notochord, the dorsal ectoderm thickens in the midline to form the neural plate (Fig. 21-1).

II. Due to the changes in the shape and size of the neural epithelial cells and the changes in their connections with surrounding cells, the lateral margins of the plate become elevated to form the neural folds.

III. The depression between these folds is known as neural groove.

IV. At about the 25th day the neural folds fuse to form the neural tube. The fusion begins at the fourth somite and progresses rostrally and caudally.

V. For a short time the neural tube remains open at both ends as the rostral and caudal neuropores (Fig. 21-2).

VI. The rostral neuropore closes at about the 25th day, and two days later the caudal neuropore closes.

VII Some cells at the margin of neural fold do not incorporate into the neural tube, and thus form the neural crest.

VIII The neural tube detaches itself from the ectoderm and sinks into the underlying mesoderm.

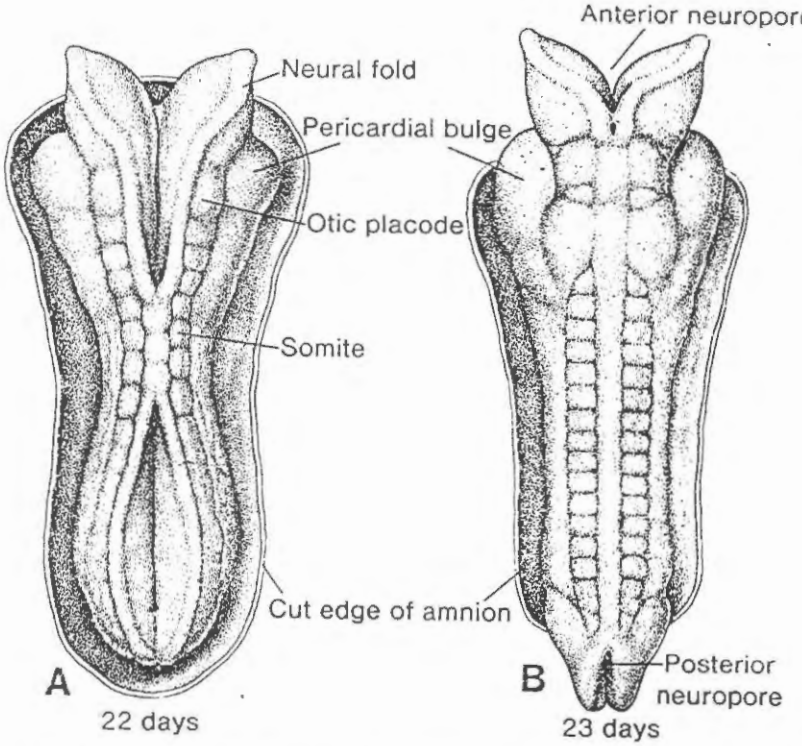

Figure 5-4. A, Dorsal view of a human embryo at approximately day 22. (Modified after Payne.) Seven distinct somites are visible on each side of the neural tube. B, Dorsal view of a human embryo at approximately day 23. (Modified after Corner.) Note the pericardial bulge on each side of the midline in the cephalic part of the embryo.

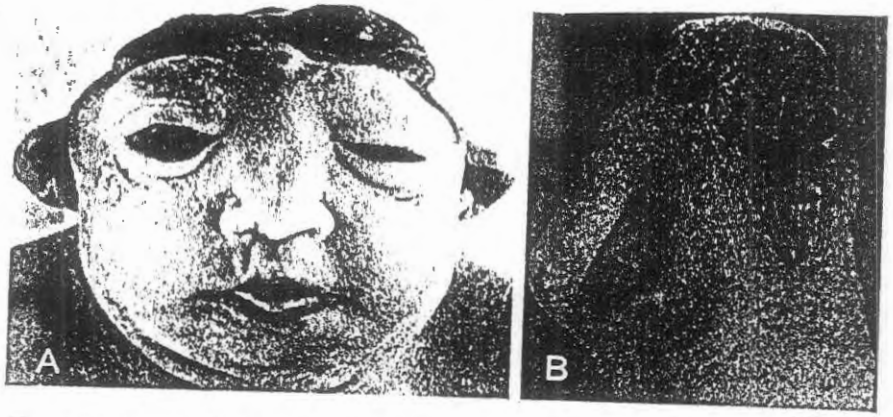

Figure 20-31. A, Photograph of anencephalic child. Ventral view. This abnormality is frequently seen (1:1000 births). Usually the child dies a few days after birth. (Courtesy Dr. J. Warkany. From Warkany J: *Congenital Malformations*. Chicago, Year Book Medical Publishers, 1971. Used by permission.) B, Dorsal view of an anencephalic child with spina bifida in cervical and thoracic segments.

Anencephalus → failure of the cephalic part of the neural tube (anterior neuropore) to close

At birth →
- the vault of the skull is absent
- the brain is represented by a mass of degenerated tissue exposed to the surface
- often → rachischisis (open spinal cord) in the cervical region + the neck is ~~absent~~ present

* The foetus lacks the central mechanism for swallowing
→ the last 2 months of pregnancy are characterized by ⊕ polydramnios (2000 ml) → high level of α-feto protein (AFP)
* more common in ♀ than ♂ (4 : 1)
* common abnormality (1 : 1000)

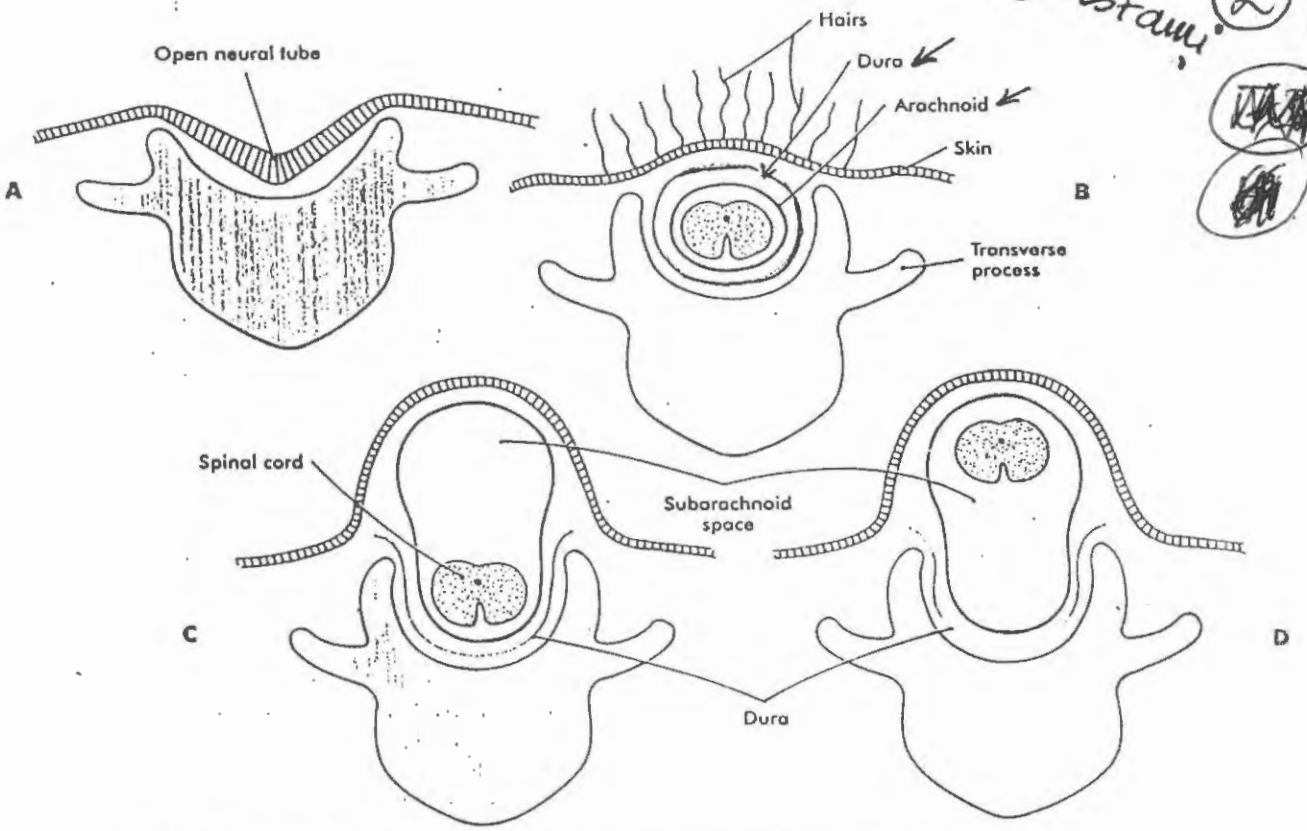

FIG. 12-38 Varities of closure defects of the spinal cord and vertebral column. **A,** Rachischisis. **B,** Spina bifida occulta, with hair growth over the defect. **C,** Meningocele. **D,** Myelomeningocele.

Other Closure Defects

A defect in the formation of the bony covering overlying either the spinal cord or brain can result in a graded series of structural anomalies. In the spinal cord, the simplest defect is called **spina bifida occulta** (Fig. 12-38, *B*). The spinal cord and meninges remain in place, but the bony covering (neural arch) of one or more vertebrae is incomplete. Sometimes the defect goes unnoticed for many years. The site of the defect is often marked by a tuft of hair. The next most severe category of defect is a **meningocele,** in which the dura mater may be missing in the area of the defect and the arachnoid layer bulges prominently beneath the skin (Fig. 12-38, *C*). The spinal cord, however, remains in place, and neurological symptoms are often minor. The most severe condition is a **myelomeningocele,** in which the spinal cord bulges or is entirely displaced into the protruding subarachnoid space (Figs. 12-38, *D* and 12-39). Because of problems associated with displaced spinal roots, neurological problems are commonly associated with this condition.

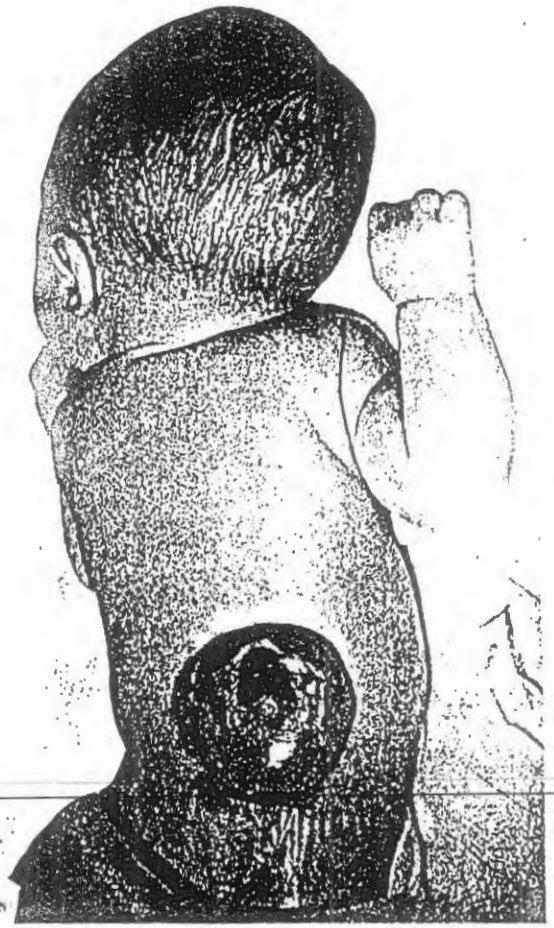

FIG. 12-39 Infant with a myelomeningocele and

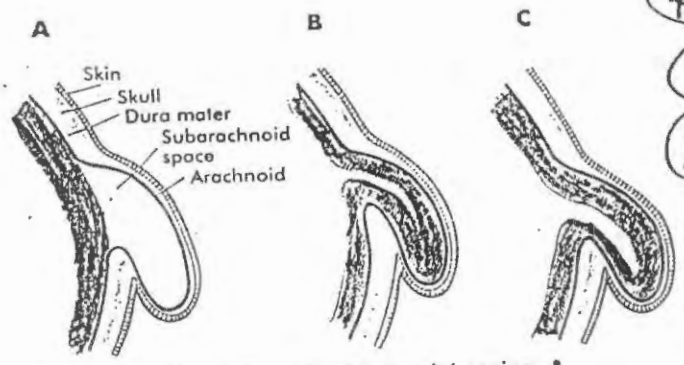

FIG. 12-40 Herniations in the cranial region. A, Meningocele. B, Meningoencephalocele. C, Meningohydroencephalocele.

A similar spectrum of anomalies is associated with cranial defects (Figs. 12-40 and 12-41). A **meningocele** is typically associated with a small defect in the skull, whereas brain tissue alone (**meningoencephalocele**) or brain tissue containing part of the ventricular system (**meningohydroencephalocoele**) may protrude through a larger opening in the skull. Depending on the nature of the protruding tissue, these malformations may be associated with neurological deficits. The mechanical circumstances may also lead to secondary hydrocephalus in some cases.

Microcephaly is a relatively uncommon condition characterized by underdevelopment of both the brain and the cranium (see Fig. 10-9). Although it can result from premature closure of the cranial sutures, in most cases its etiology is uncertain.

Many of the functional defects of the nervous system are poorly characterized, and their etiology is not understood. Studies on mice with genetically based defects of movement or behavior due to abnormalities of cell migration or histogenesis in certain regions of the brain suggest there is likely a parallel spectrum of human defects. **Mental retardation** is common and can be attributed to many causes, both genetic and environmental. The timing of the insult to the brain may be late in the fetal period.

FIG. 12-41 Fetuses with (A) an occipital meningocele and (B) a frontal encephalocele.
(Courtesy Mason Barr, Ann Arbor, Mich.)

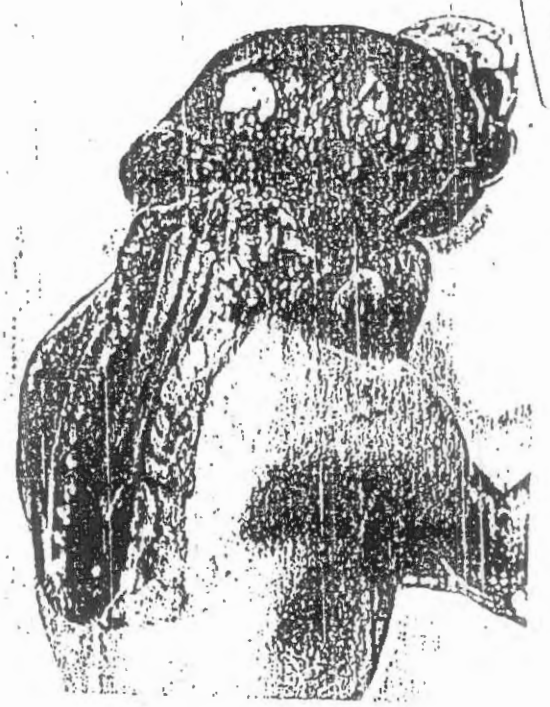

FIG. 12-37 Fetus with a severe case of rachischisis. The brain is not covered by cranial bones, and the light-colored spinal cord is totally exposed.
(Courtesy Mason Barr, Ann Arbor, Mich.)

A number of the closure defects can be diagnosed by the detection of elevated levels of alpha-fetoprotein in the amniotic fluid or by ultrasound scanning.

Defects in Closure of the Neural Tube

Failure of closure of the neural tube occurs most commonly in the regions of the anterior and posterior neuropore, but other locations are also possible. In this condition the spinal cord or brain in the affected area is splayed open, with the wall of the central canal or ventricular system constituting the outer surface. A closure defect of the spinal cord is called rachischisis and, in the brain, cranioschisis. Cranioschisis is incompatible with life. Rachischisis (Fig. 12-37) is associated with a wide variety of severe problems, including chronic infection, motor and sensory deficits, and disturbances in bladder function. These defects commonly accompany anencephaly (see Fig. 8-4), in which there is a massive deficiency of cranial structures.

Myelination in the Spinal Cord

In the spinal cord the nerve fibers are heavily myelinated or slightly myelinated. The myelin sheath is formed and maintained by the *oligodendrocytes* of the neuroglia. The cervical portion of the cord is the first part to develop myelin, and from here the process extends caudally. The fibers of the anterior nerve roots are myelinated before those of the posterior nerve roots. The process of myelination begins within the cord at about the fourth month, and the sensory fibers are affected first. The descending motor fibers are the last to myelinate, which process does not begin until term; it continues during the first 2 years of postnatal life.

Myelination in the Brain and the Onset of Function

Myelination in the brain begins at about the sixth month of fetal life but is restricted to the fibers of the basal ganglia. Later the sensory fibers passing up from the spinal cord myelinate, but the progress is slow so that at birth the brain is still largely unmyelinated. In the newborn there is very little cerebral function; motor reactions such as respiration, sucking, and swallowing are essentially reflex. After birth the corticobulbar, corticospinal fibers, and the tectospinal and corticopontocerebellar fibers begin to myelinate. This process of myelination is not haphazard but systematic, occurring in different nerve fibers at specific times. The corticospinal fibers, for example, start to myelinate at about 6 months after birth, and the process is largely complete by the end of the second year. It is believed that some nerve fibers in the brain and spinal cord do not complete myelination until puberty.

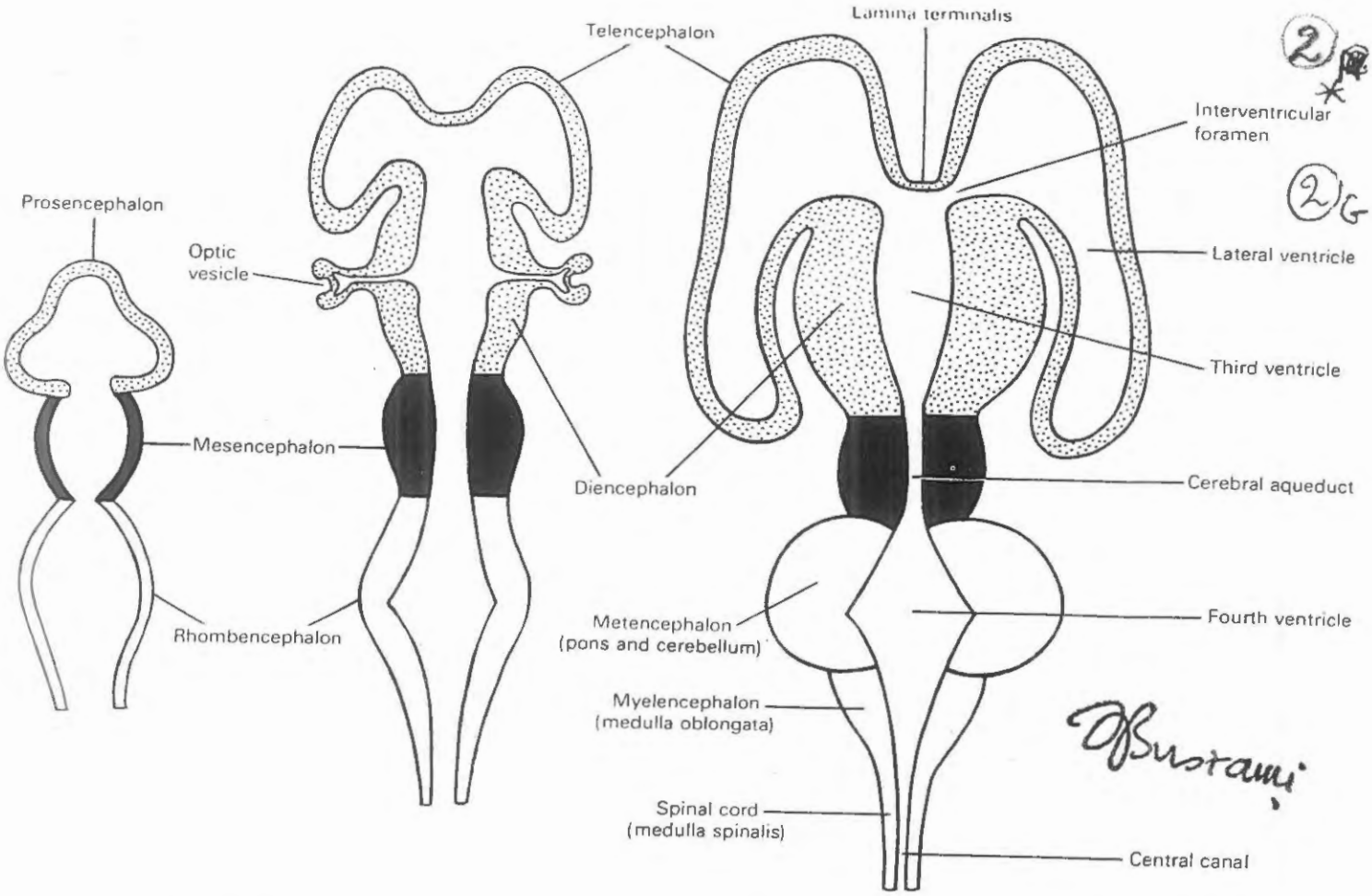

Fig. 1.3 Diagrams of stages in the differentiation of cerebral vesicles and the ventricular system.

Developmentally → 3 brain vesicles ← forebrain, midbrain, hindbrain develop from the rostral (superior) part of the Neural tube → the cavities of these vesicles become the ventricular system of the adult brain as follows :-

1. The cavity of the telencephalon (each cerebral hemisphere) will form the LATERAL VENTRICLE
2. The cavity of the diencephalon (thalamus and hypothalamus) is the Third Ventricle
3. The cavity of the mesencephalon (midbrain) remains a narrow canal called the CEREBRAL AQUEDUCT
4. the cavity of the rhombencephalon (hindbrain) will form the Fourth Ventricle (a cavity bounded by the cerebellum, pons and medulla oblongata)