ABDULRAZZAQ HINDU MA’AJI

17/MHS01/004

MBBS 300LEVEL

NEUROANATOMY ASSIGNMENT (ANA 303)

**DEVELOPMENT OF THE CEREBELLUM**

The cerebellum ‘little brain’ lies at the posterior cranial fossa and contains over half of the mature neurons in the adult brain. In adult, the weight of the cerebellum is about 150g, this is about 10% of the weight of the cerebral hemispheres. Like the cerebrum the cerebellum has a superficial layer of grey matter, the cerebellar cortex and an inner white matter. Because of the presence of numerous fissures, the cerebellar cortex is much more extensive than the size of this part of the brain would suggest. It has been estimated that the surface area of the cerebellar cortex if 50% of the area of the cerebral cortex. The cerebellum is separated from the cerebrum by a fold of dura mater called the tentorium cerebelli. Anteriorly, the fourth ventricle intervenes between the cerebellum (behind) and the pons and medulla (in front). Part of the cavity of the ventricle extends into the cerebellum as a transverse cleft. This cleft is bounded cranially by the superior medullary velum, a lamina of white matter. Histologically the cerebellar cortex is composed of a very basic structure comprising a monolayer of inhibitory purkinje cells sandwiched between a dense layer of excitatory granule cells and a sub-pial molecular layer of granule cell axons and purkinje cell dendritic trees

DEVELOPMENT

During the early stages of embryonic development, the brain starts to form in three distinct segments: the procencephalon, mesencephalon and rhombencephalon. The rhombencephalon is the most caudal segment of the embryonic brain; it is from this segment that the cerebellum develops. Along the embryonic rhombencephalic segment develop eight swelling called rhombomeres. The cerebellum arises from two rhombomeres (rhombomere 1 caudally and isthmus rostrally) located in the alar plate of the neural tube. Two primary regions are thought to give rise to the neurons that make up the cerebellum. The first zone is the ventricular zone in the roof of the fourth ventricle. This area produces purkinje cells and deep cerebellar nuclear neurons. These cells are the primary output neurons of the cerebellar cortex and cerebellum. The second germinal zone (cellular birthplace) is the rhombic lip, neurons then move by human embryonic week 27 to the granular layer. These layer of cells- found in the exterior of the cerebellum- produces granule neurons. The granule neurons migrate from this exterior layer to form an inner layer known as the internal granule layer. The external granular layer ceases to exist in the mature cerebellum, leaving only granule cells in internal granule layer. The cerebellar white matter may be a third germinal zone in the cerebellum; however, its function as a germinal zone is controversial.

**CEREBELLAR DISORDER**

JOUBERT SYNDROME this is a disorder of brain development that may affect many parts of the body. It is characterized by the absence or underdevelopment of the cerebellar vermis ad malformed brain stem. Signs and symptoms include weak muscle tone (hypotonia), abnormal breathing patterns, and abnormal eye movement.

RHOMBENCEPHALOSYNAPSIS is a midline brain malformation characterized by missing cerebellar vermis with apparent fusion of the cerebellar hemispheres it can in isolation or together with other nervous system and extra-central nervous system malformations. Severity of rhombencephalosynapsis is also associated fusion of the tonsils, as well as midbrain abnormalities including aqueductal stenosis and midline fusion of the tectum. It is associated with abnormalities like absent olfactory bulbs, dysgenesis of the corpus callosum, absent septum pellucidum and atypical forms of holoprosencephaly.

DANDY-WALKER MALFORMATION is an abnormally resulting in absence of the central part of the cerebellum or abnormal positioning. The left and right sides of the cerebellum may be small as well. The fourth ventricle and the posterior fossa are abnormally large. These abnormalities often result in problems with movement, coordination, intellect, mood, and other neurological functions. People with dandy-walker malformations experience muscle stiffness and partial paralysis of the lower limbs seizures may occur. Rare hearing and vision problems can be features of this condition.

References:

1. Muller F, O’Rahilly R (1990). “The human brain at stages 21-23, with particular reference to the cerebral cortical plate and to the development of the cerebellum”
2. NINDS joubert syndrome information page. NINDS. January 21, 2016
3. Joubert syndrome and related disorders foundation. 2016
4. Joubert syndrome. Genetics home reference. January 2011