Name: Awe Ayomide .B.

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Question assignment: Cerebellum and it's connections.

During embryonic development, the anterior portion of the neural tube forms three parts that give rise to the brain and associated structures:

* + Forebrain (prosencephalon)
  + Midbrain (mesencephalon)
  + Hindbrain (rhombencephalon)

The hindbrain subsequently divides into the **metencephalon** (superior) and the **myelencephalon** (inferior). The cerebellum develops from the metencephalon division.

Function: The cerebellum receives information from the sensory systems, the spinal cord, and other parts of the brain and then regulates **motor** movements. The cerebellum coordinates voluntary movements such as posture, balance, coordination, and speech, resulting in smooth and balanced muscular activity.

* + The internal structure of the cerebellum reflects an intriguing paradox; the arrangement of its cells is relatively simple and repeated throughout, yet the connections between its neurons are wired into a complex array of gene expression domains and functional circuits. The developmental mechanisms that coordinate the establishment of cerebellar structure and circuitry provide a powerful model for understanding how functional brain networks are formed. Two primary germinal zones generate the cells that make up the cerebellum. Each zone expresses a specific set of genes that establish the cell lineages within the cerebellar anlage. Then, cohorts of differentiated projection neurons and interneuron progenitors migrate into the developing cerebellum. Thereafter, a number of remarkable patterning events occur including transformation of the smooth cerebellar surface into an intricately patterned series of folds, formation of three distinct cellular layers, and the demarcation of parasagittal gene expression domains. Together, these structural and molecular organizations are thought to support the proper connectivity between incoming afferent projections and their target cells. After birth, genetic programs and neural activity repattern synaptic connections into topographic neural networks called modules, which are organized around a longitudinal zone plan and are defined by their molecular, anatomic, and functional properties.

* + HUNTINGON DISEASE (CEREBELLAR DISEASE)

It's a progressive brain disorder that causes uncontrolled movements, emotional problems and loss of thinking ability(cognition).

Genetic bases:

This condition is inherited in a autosomal dominant pattern which means one copy of the altered gene in each cell is sufficient to cause the disorder. An affected person usually inherits the altered gene from oeo effected parent. In rare cases, an individual with H77 ( Hungtington) disease doesn't have a parent with the disorder. As the altered H77 gene is passed from one generation to the next, the size of CAG trinucleotide repeat often increase

* + MYOCLONUS DYSTONIA

It is a movement disorder that typically affects the neck, torso and arms. Individuals with this condition experience quick, involuntary muscle jerks or twitches (myoclonus). The movement problems usually first appear in childhood with the development of myoclonus, and it remain stable throughout life.

GENETIC BASIS

This condition is inherited in an Autosomal Dominant pattern.In cases in which the cause of the condition is unknown, the inheritance is unclear. When caused by SGCE(Sarcoglycan epilon) genetic mutations,myoclonus-dystonia occurs only when the mutation is inherited from a person's father.People normally inherit one copy of each gene from their mother and one copy from their father. For most genes both copies are active.

* + PROGRESSIVE SUPRANUCLEAR PALSY(PSP)

This is a brain disorder that affects movement, vision, speech and thinking ability. The signs and symptoms of this disorder usually become apparent in mid to late adulthood, most often in a person's 60s,loss of balance and frequent falls are the most common early signs of Progressive Supranuclear Palsy.

GENETIC BASIS

Most cases of the PSP are sporadic which means they occur in people with no history of the disorder in their family. However, some people with this disorder have had family members with related conditions, such as parkinsonism and dementia.

JOUBERT SYNDROME

a rare neurological disorder featuring absence of the cerebellar vermis (i.e. midline: a part of the brain that controls balance and coordination) and a malformed brain stem (connection between the brain and spinal cord).

The most common features are lack of **muscle** control (**ataxia**), abnormal breathing patterns (hyperapnea), sleep apnea, abnormal **eye** and tongue movements and low**muscle tone**.

GENETIC BASIS

Joubert syndrome is inherited as an autosomal recessive genetic disorder.

In recent years, mapping of three genetic loci and the identification of mutations in two genes, AHI1 and NPHP1 have tremendously helped with identifying this disorder has it has many overlapping symptoms with other disorders. These genes encode proteins with some shared functional domains, but their role in brain development is unclear. Clues may come from studies of related syndromes, including Bardet-Biedl syndrome and nephronophthisis, for which all of the encoded proteins localize to primary cilia. The data suggest a tantalizing connection between intraflagellar transport in cilia and brain development.

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