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**MATRIC NO: 17/MHS01/086**

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 also 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.

The internal structure of the cerebellum reflects an intriguing paradox; its cytoarchitecture 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

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