

Malignant Hyperthermia



PLAY PICMONIC

Inheritance

Autosomal-Dominant

Domino

Malignant hyperthermia is inherited in an autosomal dominant manner; this means that one abnormal allele is enough for the individual to manifest the disease. However, it is important to note that the penetrance of this disorder is variable, hence not all individuals who inherit abnormal alleles will manifest the disease.

Pathophysiology and causes

Ryanodine Receptor 1 Mutation

Rihanna Receptor (1) Wand Mutant

Malignant hyperthermia patients have a genetic predisposition due to mutations in skeletal muscle ion channels. Ryanodine receptor 1, coded by the RYR1 gene, is a commonly affected calcium channel that controls calcium release from the sarcoplasmic reticulum. Mutations in this channel result in excessive calcium accumulation within muscle cells after exposure to anesthetics or succinylcholine.

Inhaled Anesthetics

A-Nest of Inhaled Anesthesia

A common trigger for malignant hyperthermia is inhaled anesthetics given during surgery, such as halothane and isoflurane. Therefore, suspect malignant hyperthermia if a post-surgical patient develops the acute onset of autonomic symptoms and muscle rigidity.

Succinylcholine

Sucker-in-cola

Succinylcholine is a depolarizing neuromuscular blocking agent often used during procedures to paralyze muscles and facilitate intubation. In myocytes with mutated calcium channels, however, succinylcholine will trigger excess calcium release and lead to malignant hyperthermia.

Symptoms

Autonomic Instability

Unstable Atomic-automobile

Malignant hyperthermia can manifest with autonomic instability, which can include signs like fever, tachycardia, hypertension, and diaphoresis.

Muscle Rigidity

Muscle-man of Stone

Due to the overwhelming release of calcium from the sarcoplasmic reticulum, these patients can develop muscle rigidity.

Treatment

Dantrolene

Dantrolene-lion

Dantrolene is the treatment for malignant hyperthermia. It directly inhibits the ryanodine receptor in myocytes, thereby preventing calcium release from the sarcoplasmic reticulum of muscle cells.