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A Review: Malignant Hyperthermia-A Genetic Disease Of Sodium Channel Function

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Abstract

Malignant Hyperthermia is inherited as a dominant gene in pigs and in humans. It is characterized by a rapidly increasing body temperature up to 118oF with a metabolic rate over 10x normal, an intense peripheral vasoconstriction with blood pressures over 400 mmHg, and a fatal outcome in most cases.

The MH syndrome is triggered by exercise, hauling to market, breeding activity, hot weather, and other stress factors. In humans, the cases are triggered by depolarizing muscle relaxants, Halothane anesthesia, and other halogenated anesthesia compounds. Non-depolarizing muscle relaxants such as Pancuronium and Vecuronium are safer to use. Organon 9426 (Rocuronium) is safe to use and provides carry over protection against the development of MH.The widespread use of Sevoflurane in humans has reduced the incidence of MH to 1:550,000. Only three cases of MH have been reported in the USA during the past 30 years. They developed in WI, FL and SC.The MH susceptible pig is an outstanding animal model for research purposes and has enabled the development of new muscle relaxants and anesthesia agents that are safer for human use. The sodium channels in MH susceptible animals are leaky and allow the influx of sodium into muscle cells which must be pumped out by sodium-potassium ATPase, thereby using ATP and generating heat.

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