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Malignant hyperthermia: The genetic disease is characterized by a runaway futile cycle operating at the acetylcholine receptor (sodium channel) level

The Malignant hyperthermia gene is inherited as a dominant gene. There are variable degrees of penetrance so a colony of MHS pigs must be established to provide susceptible animals for experimental studies. We have used our MHS pig colony for over 20 years. The MHS pig colony was started in January 1969 at Wisconsin, it was moved to Missouri in 1973 and on to El Paso, TX in 1982. We completed an experimental study of sevoflurane in 1984 with MHS pigs. We found sevoflurane to be an exceptionally smooth anesthetic with no side effects. Sevoflurane usage in clinical practice has reduced the incidence of MH by eleven fold since it was approved for human use. It is an outstandingly good anesthetic with minimal side effects. Non-depolarizing muscle relaxants such as Pancuronium, Vecuronium, metubine iodide, and Orgnon 9416 (Rocuronium) do not trigger an MH episode in experimental MHS pigs. Similar results are observed in human patients. Succinlycholine is a potent trigger of MH in pigs and human patients. Organon 9426 offers protective effects after 100% muscle twitch response has been recovered in MHS pigs. MHS pigs have a 30-35 % decrease in acetylcholine receptors at the myoneural junction. These observations suggest either a decrease in the synthesis of acetylcholine receptor proteins or an increased degradation of dysfunctional acetylcholine receptors. The Ryanodine receptor appears to be involved in MH as a secondary or tertiary participant in triggering the MH syndrome.

Biography

Charles H Williams completed his PhD in 1968 and then a Post-Doctoral at the Institute for Enzyme Research with David E Green on mitochondrial studies. He also published with Henry A Lardy at the Enzyme Institute. He moved to Missouri as Associate Professor of Biochemistry and Assistant Professor of Medicine. He relocated to TTUHSC-El Paso in 1982, where MH research was his primary area of interest. He has published over 50 papers in refereed journals and has presented posters and lectures at many international events. He was a Full bright Fellow in Peru in 1970.

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