Society of Rural Physicians of Canada
26TH ANNUAL RURAL AND REMOTE MEDICINE COURSE
ST. JOHN'S NEWFOUNDLAND AND LABRADOR
APRIL 12 - 14, 2018

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Dr. Gavin Parker • PINCHER CREEK • AB

MALIGNANT HYPERTHERMIA

Malignant hyperthermia is a rare and serious complication of providing a general anesthetic or a potential reaction to medicines given in the rural ED. Using a case from a rural OR we will discuss management of this case and how to avoid a similar outcome in your facility.

1. Compare MH Physiology and epidemiology 2.Discuss Clinical manifestations 4.Differential diagnosis and treatment 5.Case review 6.MH resources

Malignant Hyperthermia

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The Pincher Creek Echo

Community

Pincher Creek doctor saves child

By Brian Vossen
Pincher Creek Echo Editor

One Lethbridge mother is thanking her lucky stars at the staff at the Pincher Creek Health Centre for saving her son's life.

Last month Chelsea Bretzke brought her six-year-old son Aaron to Pincher Creek to have some general dental work done. Aaron is a non-verbal autistic and extremely sensitive to sensory information, so it was difficult, if not impossible, for him to have any kind of dental work done. A friend of Bretzke's recommended that she come to see Pincher Creek dentist Dr. Mark Leishman, who she said performs regular dental surgery on patients under general anesthetic at the hospital.

However, this time, something went terribly wrong when Aaron had an extremely adverse reaction to the anesthetic. Bretzke said that Aaron's temperature rose and he started to exhibit sign of an incredibly rare genetic disorder called malignant hyperthermia, one that anesthesiologist Dr. Gavin Parker managed to recognize despite never seeing the disorder himself.

Bretzke explained what she could about the disorder, saying that it caused Aaron's skeletal muscles to metabolize at huge rate on a cellular level. Essentially, it was as if Aaron was having a very intense physical work-out without actually moving. She added that she was beyond concerned when the doctors came out halfway through the surgery to inform her that there had been complications.

"It was terrifying," Bretzke said, "So much of our life has been invested into Aaron."

Bretzke added that she was not completely aware of how much danger Aaron was in at the time, but after seeing her son in stable but still critical condition, hooked up to various medical implements while he awaited the arrival of STARS, she knew his condition was serious.

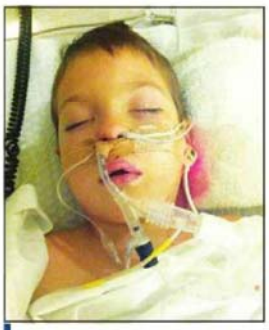
"I remember Gavin saying to me, 'He's not out of the woods,'" Bretzke said.

According to Bretzke, it was the quick thinking of Dr. Parker and other staff at the hospital that saved her sons life. Aaron was hooked up to a breathing tube and had cooling blankets placed all over him to keep his temperature down. STARS air ambulance was quickly dispatched and air lifted Aaron to the Calgary Children's Hospital, Bretzke said. She added that that Dr. Parker immediately called a specialist to further advise him on how to proceed during the crisis.

Later, it was confirmed (within a 1 per cent error margin) that Aaron did have malignant hyperthermia. When Bretzke arrived at the Calgary Children's Hospital, where Aaron spent the next several days recovering the hospitals pediatric ICU, a specialist told her that Aaron would not survived without the quick recognition and response to the rare condition.

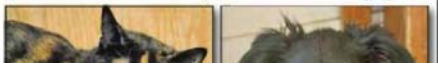
"The specialist in Calgary said, 'Yeah, he saved your son's life for sure," Bretzke said.

However, recovery in Calgary was no picnic either, Bretzke said. She said that Aaron had another attack in Calgary later that same week and seeing her son in that condition was



Submitted photo
Aaron Bretzke recovers at the Calgary Children's Hospital after an attack of malignant hyperthermia, a rare genetic condition the Calgary Children's Hospital had not seen in a decade.

Pincher Creek Humane Society
Phone: 403-627-5191 www.pinchercreekhumanesociety.org



Mac's Clubhouse Diner

Overview

- Physiology and epidemiology
- Clinical manifestations
- Differential diagnosis and treatment
- Case review
- MH resources

Physiology

- Inherited disorder of skeletal muscle
- Problem w/ reuptake of intracellular Ca^{2+}
- Exact cause uncertain
  - Ryanodine receptor
- Disease inheritance is autosomal dominant

Epidemiology

- Definite association: central core disease
- Possible association: Duchenne, Becker, King-Denborough, other myopathies
- Coincidental association: neuroleptic malignant syndrome, SIDS, Lymphomas, and Heat Stroke

Epidemiology (cont)

- Frequency 1:3,000 - 1:60,000 anesthetic cases
- Approximately 600 cases per year in U.S[1]
- Increased incidence in young adult males
- 1:100,000 hospital discharges complicated by MH

Diagnostic Testing

Muscle Contracture Test

- Caffeine Halothane Contracture Test(CHCT)
- Gold Standard
- MH Muscle Biopsy Center

Genetic Testing

- Ryanodine receptor (RYR1 Gene)
- Primary genetic focus
- DNA blood test or biopsy

Trigger Agents for MH

MH Triggers

- Potent volatile anesthetics
  - Sevoflurane
  - Desflurane
  - Isoflurane
- Depolarizing muscle relaxants
  - Succinylcholine

NOT MH Triggers

- Nitrous oxide
- IV induction agents
- Non-depolarizing muscle relaxants
- Opioids

Clinical Signs of MH

Specific	Non-Specific
▪ Muscle rigidity*	▪ Tachycardia
▪ Increased CO_2 production	▪ Tachypnea
▪ Marked temperature elevation	▪ Acidosis (respiratory; metabolic)
▪ Rhabdomyolysis	▪ Hyperkalemia

Differential Diagnosis

- Insufficient anesthesia or analgesia
- Insufficient ventilation or FGF
- Anaphylactic reaction
- Pheochromocytoma
- Thyroid crisis
- Cerebral ischemia
- Neuromuscular disorders
- Procedural causes
- Malignant neuroleptic syndrome

Treatment of MH

- Stop triggering inhalation agents/succinylcholine
- Hyperventilate high flow 100% O_2
- Dantrolene 2.5mg/kg push, repeat prn
- Continue monitoring & interventions
- Treat hyperthermia, acidosis, and arrhythmias

Dantrolene (20mg/vial)

- Muscle relaxant
- Indications
  - The ***only*** specific and effective treatment for MH
  - Neuroleptic malignant syndrome, muscle spasticity, serotonin syndrome, and 2,4-dinitrophenol poisoning
- Drug Interactions
  - CCBs, NDNMB, CNS depressants & benzodiazepines

Preparing for at risk patients

- Shut down/disable vaporizers
- Flow O_2 > 10L/min for 20 minutes through machine and ventilator
- Change CO_2 absorbent
- Use *non-trigger* agents and methods
- Monitor for early signs of MH

Case Study

- 6 year old for dental surgery, no family history of anesthetic issues
- Gas induction with sevoflurane, tube changed halfway through case with succinylcholine assistance
- EtCO2 increased, never did get hyperthemic
- Eventually had muscle bx, confirmed spont. mut.

MH Resources

- Site specific policy
- Malignant Hyperthermia Cart
- MHAUS
  - Malignant Hyperthermia Association of the United States @ 1-800-MH-HYPER
  - www.mhaus.org

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Summary

- Disorder with intracellular Ca^{2+} effecting skeletal muscle
- Triggered by inhaled anesthetics & succinylcholine
- Specific and non-specific clinical signs
- Definitive treatment with Dantrolene

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Summary

- Call for help (let surgeon know)
- Turn off potential triggering agents
- Administer dantrolene 2.5 mg/kg every five minutes
- Cool patient to 38C
- Monitor and correct blood gases, electrolytes and glucose

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