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Navigating the Crisis: How to Diagnose and Treat Malignant Hyperthermia

Dr. Turck:
Welcome to *Clinician's Roundtable* on ReachMD. I'm Dr. Charles Turck, and joining me to discuss malignant hyperthermia is Dr. Henry Rosenberg. He's a member of the Board of Directors of the Malignant Hyperthermia Association of the United States. Dr. Rosenberg, thanks for joining me today.

Dr. Rosenberg:
My pleasure. Thank you.

Dr. Turck:
So why don't we start with some background, Dr. Rosenberg. What is malignant hyperthermia? Is it inherited? And how do you recognize the signs and symptoms?\

Dr. Rosenberg:
Malignant hyperthermia is an inherited disorder. As there are few people who have any clinical signs of the disorder outside of anesthesia, most people who know about the disorder are anesthesiologists, nurse anesthetists, surgeons, and geneticists. Because it's rather uncommon, there are very few other specialists who have an in-depth knowledge of this syndrome.

The syndrome derives its name from when it was first described in the early 1960s where it was characterized in a patient who was receiving a general anesthesia—that's with a gas anesthetic—who develops a high body temperature, and if not recognized and treated, it was associated with a 70 percent mortality. It's, fortunately, not that way, but the syndrome was very dramatic, and the heart rate went up, the blood pressure, the temperature, the carbon dioxide, all the signs of hypermetabolism, which is very different than under the situation of a normal general anesthetic where the body temperature tends to decrease a degree or two.

The other prominent feature of the disorder was muscle rigidity, and that also is very atypical in anesthesia, where the muscles get rigid, the arms, the legs, and the jaw, and that's uncharacteristic. And that rigidity was not relieved by the usual muscle relaxant drugs.

Dr. Turck:
You started to touch on this a little bit earlier. You were saying that the traditional muscle relaxants, at least back in the '60s, weren't very effective at treating malignant hyperthermia. What are the current treatment options?

Dr. Rosenberg:
The current treatment is really a single drug: dantrolene. Dantrolene is specifically indicated in the treatment of this syndrome, and it's described as working like magic. It was discovered in a peculiar way, but nevertheless, it is very effective and used all over, and the recommendation is that any place that provides general anesthesia should have dantrolene immediately available because the treatment of the syndrome was critically dependent on the speed of recognition and the speed of treatment.

Dr. Turck:
For those just tuning in, you're listening to *Clinician's Roundtable* on ReachMD. I'm Dr. Charles Turck, and I'm speaking with Dr. Henry Rosenberg about malignant hyperthermia.

Now turning to critical care in the operating room settings, Dr. Rosenberg, how do you ensure patient safety when administering anesthesia to individuals with a known or suspected risk of malignant hyperthermia?

Dr. Rosenberg:

The first question that should be asked to anyone receiving a general anesthetic or anything other than just a straight local anesthetic is, “Have you or a member of your family had problems or even death related to anesthesia, per say? Is there a family history of unexplained death or elevated body temperature during an anesthetic or shortly after an anesthetic?” And if the answer is yes, the anesthesiologist, the nurse anesthetist, or anesthesia assistant should know immediately that this is a patient that should not receive one of the gas anesthetics, such as sevoflurane or desflurane, nor should they receive the paralyzing drug succinylcholine. Now succinylcholine is a drug that causes muscle relaxation and is used very, very commonly. However, it in addition to the gas anesthetics is a very potent trigger for this syndrome.

To be prepared for the disorder, patients should be monitored in the standard way. They should have their temperature monitored optimally through the esophageal probe or, if necessary, axillary probe or any other place that temperature can be monitored. And the other parameter that is measured during all general anesthetics is carbon dioxide exhalation.

Dr. Turck:
Now you’ve talked a little bit about the early detection of malignant hyperthermia during anesthesia. Is there a diagnostic test for a suspected case of malignant hyperthermia?

Dr. Rosenberg:
Before I mention that, in terms of the diagnosis, it’s important to note that the syndrome does not always present in a uniform way. It doesn’t always present at the beginning of the anesthetic or the middle of the anesthetic. There are cases where the syndrome begins very close to the end of the anesthetic, so it’s not a characteristic time of onset.

Now the diagnostic test has a very interesting story. The diagnostic test that was first developed in the 1970s and ‘80s involved taking a piece of muscle, usually from the thigh, and dissecting it and hanging the strips of muscle in a standard muscle bath, as you would do in a pharmacology lab. And then as the liquid is bubbled through with the gas of oxygen to keep the tissue alive, you add halothane, which is an anesthetic drug that was formerly used. What will happen is that the muscle will contract. It won’t relax, and it will develop an increase in temperature that is very dramatic. The test was used and is still used for many years in many places because it has a very good accuracy.

Since the disorder was known to be inherited, which could be documented in families, there was obviously a genetic defect, and there were many years of exploration of what that defect is, where it’s located, and where the problems are within the gene. And that was worked out progressively and more recently with the increase in the technological facility of doing genetic analysis. Particularly, DNA sequencing has become much more feasible to detect where the gene is and where the variants in the gene are located. And so that genetic testing, which is done quite commonly for many conditions in medicine, can be and is applied to the diagnosis of malignant hyperthermia.

Now the problem with the test is that even though a person may be at risk for malignant hyperthermia, maybe 20 percent of the people who have the genetic test will have a negative test, and that’s because all the variants within the genes are not explored as to their likelihood of causing or being related to malignant hyperthermia. So there are actually three genes that have been associated with malignant hyperthermia, and one of them is called the ryanodine receptor 1. That’s RYR1. The other one is a NA CACNA1S, which is a different gene, also related to muscle structure and function. And the third one is called STAC3. There are very, very few people with that one, so most of the work that’s been done has been on the first two genes.

Dr. Turck:
Now switching gears here a bit, should a malignant hyperthermia emergency occur, how do healthcare professionals, like surgeons, anesthesiologists, and nurses, collaborate to ensure a coordinated and effective response?

Dr. Rosenberg:
That’s a really very important question. It’s not a disorder that the anesthesiologist or anesthesia provider recognizes, injects the dose of dantrolene, and everything works out just fine. It really has to be a cooperative adventure because people have to, first of all, bring in the dantrolene, and the Malignant Hyperthermia Association recommends having a specific cart where the dantrolene is located because you’re not going to store dantrolene in every anesthesia machine because the disorder doesn’t happen that frequently. So you need someone to bring it in. You need someone to mix up the drug. If the patient is hyperthermic, you need to have someone bring in ice or a cooling blanket to put on the patient. The surgeon has to be aware of what’s happening and may have to modify or suspend the surgical procedure as the syndrome takes place because it’s so dramatic that the patient’s body temperature may rise three, four, five degrees in a matter of minutes.

So the other thing that needs to be happening is that the laboratory should be alerted because the important diagnostic or helpful test in management of the syndrome are blood gases, so there will be a series of blood gases sent during the management course. And then

the ICU needs to be alerted that the patient is not going to go back to a regular bed or discharged. They're going to need to be sent to an intensive care unit. And one of the reasons for doing that is because even though the syndrome was treated in the operating room, the person still needs to get dantrolene for another 24 or 36 hours because there is a recrudescence of the syndrome even with successful initial treatment.

Dr. Turck:
Now before we close, Dr. Rosenberg, are there any global take-home messages or additional thoughts you'd like to leave with our audience today?

Dr. Rosenberg:
Certainly. Anybody who is providing anesthesia that involves the gas anesthetics and/or succinylcholine should be aware of the syndrome. The facility should have a drill should a case occur. In other words, the facility needs to know what to do. If it's an ambulatory facility, there's no ICU, so there needs to be a transfer mechanism in place. And a lot of this information can be found on the Malignant Hyperthermia Association website, mhaus.org.

Dr. Turck:
Well, this is a condition that can have significant consequences on a patient's health, including their likelihood of surviving surgery. So I want to thank my guest, Dr. Henry Rosenberg, for sharing his valuable insights on malignant hyperthermia. Dr. Rosenberg, it was a pleasure speaking with you today.

Dr. Rosenberg:
Well, thank you very much. It's my pleasure to provide the information because, as you say, it can be a fatal syndrome in some cases.

Dr. Turck:
For ReachMD, I'm Dr. Charles Turck. To access this and other episodes in this series, visit *Clinician's Roundtable* on ReachMD.com, where you can Be Part of the Knowledge. Thanks for listening.