

Myotonia Congenita

Pregnancy and Birth Considerations

Myotonia congenita, or MC, is a condition caused by chloride ion channel mutations affecting skeletal muscle cells. The negatively charged chloride ions are needed to allow a muscle group to relax after contracting. In most people this takes place without any delay, but with MC it can be significantly extended. It will be worsened by cold exposure, sudden changes in extracellular potassium, most pesticides and herbicides, epinephrine, certain medications (especially inhalers, statin drugs and quinolone antibiotics), and hormones like progesterone and testosterone.

A person with MC may get up out of a chair and start walking, and rather than contracting and releasing as you take the next steps, the muscles don't relax quickly enough to have a normal walking pattern. It feels like the muscles grab or lock up, and if you try to force them to move, this exerts a significant isometric force on the muscles and can even cause cell damage. This results in hypertrophied muscle groups (calves in particular) as well as pain and cramping in the muscles. This is usually only noticeable when first initiating movement after being still for several minutes. Once the muscles are flexed a few times, chloride ions are pumped into the cells more efficiently and the stiffness appears to let up.

MC is not a risk for pregnancy complications. It only affects skeletal muscles, so the uterus is not impacted. Because progesterone does tend to worsen myotonia, pregnant women with MC need to be extra careful about fall risks and make sure their muscles are kept warm and flexed when rising, walking or climbing stairs. Some women with milder MC only have symptoms during pregnancy.

The main concerns have to do with obstetric anesthesia. While MC does not cause true malignant hyperthermia, it can cause similar symptoms related to sustained depolarization and hyperkalemia. Because of this it is essential for the patient to have a consultation with the anesthesia department before delivery so that they will have notes on file and use appropriate anesthetics during labor and delivery. Epidurals and spinals are safe as long as any locals used to initiate insertion are free of vasoconstrictors. Locals used for suturing also need to be vasoconstrictor-free (bupivacaine, mepivacaine or lidocaine w/o epinephrine are fine). Emergency cesareans are the major concern. It has not been proven that anesthesia gases will trigger the MH type symptoms with myotonia congenita, but most anesthesiologists prefer to avoid them when possible. The primary triggering agent is succinylcholine and similar depolarizing drugs.

Because most pregnancy caregivers are not familiar with MC, they usually do an internet search on myotonia and find a great deal of information about complications for both the mother and the baby. That is because myotonic muscular dystrophy comes up in the searches. While the two conditions do share muscle myotonia, that is the only similarity. Myotonic dystrophy is associated with very high risk and mothers need to be under close supervision throughout

pregnancy, labor and birth. A neonatologist is always involved because of the risk of the congenital form of myotonic dystrophy which can be life-threatening for the baby. None of these complications are a factor in myotonia congenita. Women with MC do not need special monitoring unless they have an additional condition associated with high-risk pregnancy.

In summary, here are the main things to consider when you have a patient with MC:

- Stiffness will be worse during pregnancy because of the high levels of progesterone. This can affect any skeletal muscle including arms and legs, ocular muscles, tongue, upper part of esophagus and diaphragm. This is completely normal and assuring them their condition is not progressing is helpful. It will get better after birth (as long as progesterone-related birth control methods are avoided)
- Medications used to treat myotonia (most women get off meds for pregnancy and nursing based on their doctor's recommendations):

Mexiletine and flecainide (anti-arrhythmics) - Category C
Carbamazepine, oxcarbazepine, phenytoin - Category D
Lamotrigine - Category C
Acetazolamide - Category C
Quinine Sulfate - Category C

- During labor, avoid hypoglycemia from fasting or from giving dextrose IVs too quickly. Rising insulin will trigger myotonia. Avoid IVs containing potassium unless blood tests show low levels, and even then, administer very slowly.
- Have a plan in place for emergency cesarean surgery
- Keep the mother warm if she feels chilled and warm IV fluids
- Avoid use of local anesthetics containing epinephrine

Women with myotonia congenita are just as likely to experience the typical complications of labor and childbirth, but no more likely than the general population. They don't need additional monitoring and should be given the opportunity to experience a comfortable birth setting (especially laboring in warm tubs which will reduce myotonia), family support and the least intervention necessary.