

Myotonia Congenita Information Sheet for School Staff

Introduction

This paper is to provide information for school staff regarding a condition known as myotonia congenita which has been diagnosed in your student,

_____ (student name).

The following is for informational purposes only and is not to be used for diagnosis or treatment of myotonia congenita.

Myotonia Congenita, or MC, is a condition caused by a mutation in the chloride ion channel of the skeletal muscle. While the degree of severity can vary widely from person to person, the most common and obvious symptom is the inability to relax muscles after they are contracted, especially after being in a static position such as sitting, lying or standing still. This looks like stiffness to an observer and is the same condition that you may have seen in “fainting goats.” Other factors which may influence the stiffness include sudden loud noises, intense exercise, stress, cold weather, exposure to pesticides and herbicides, and certain foods or medications.

The muscles affected by myotonia congenita include hands, arms, legs, abs, back, diaphragm, neck, face, throat (upper part of esophagus), eyes and even tongue. When the tongue is stiff it can impair speech and the student may sound drunk.

This stiffness may last for many seconds, for instance when rising up out of a chair or desk or grasping a doorknob. Usually the muscles begin to relax and work more freely after they are flexed and warmed up. However a sustained pause in activity or movement tends to “reset” the whole cycle and the warming up must take place again.

Myotonia Congenita is an inherited disorder. It is not contagious and classmates need to understand that they cannot “catch” MC. Unlike muscular dystrophy it does not become progressively worse over time and a person with MC will have a normal lifespan, but the amount of stiffness and resulting pain and cramping can vary from one day to the next and even one hour to the next. A child may come to school and seem perfectly normal, but after being exposed to cold at recess the stiffness may become so severe he or she could have trouble swallowing food at lunch.

While the physical manifestations of this condition are not usually that severe, the emotional impact can be devastating if children are subjected to constant teasing, bullying and condescending comments from staff. MC does not affect mental function and it can further add to humiliation and embarrassment if children are put in to special education classes. A child with MC is just as likely to have a learning disability from other causes as the rest of the population so screenings are appropriate, but any deficits are not caused by MC and should be investigated further with the parents and family physician.

The greatest risk of injury for a student with MC is falling. Most children learn to adapt and use caution on stairs, but just catching the toe of a shoe on a sidewalk can cause all the muscles to seize up suddenly and often it is impossible to break the fall. A sudden loud noise or a shove from a classmate can have the same effect. It is advisable to send any students to the nurse to be evaluated after a hard fall since the head often hits the ground. Sometimes lightly touching the arm of another student can help provide a little extra stability when crossing a street or open space. Most students will be unwilling to draw attention to themselves in this way and that should be respected. Their greatest desire will be to appear normal.

The rest of this information sheet is useful for all staff, but it will be divided into specific categories based on the relationship of the student to the staff person. It also is from a more subjective standpoint based on collective personal experiences with schools.

Classroom Teachers

You will probably be the most influential person in the student's life at school, especially younger students who are forming their self-image and awareness of the world around them. It is important to be sensitive to the impact of this condition without communicating to the child and their classmates that they are disabled. Many of us were accused of being lazy, uncooperative or attention-seeking because we were physically unable to perform at the same level as other students and did not know how to relay our feelings to the teacher.

There can be a strong sense of shame with MC because our bodies so often betray us and we find ourselves in embarrassing situations like sprawling on the floor when getting up to hand in a paper. Some children have a strong personality that can take the laughs and teasing and they even join in. Others withdraw and can even become depressed and dread going to school.

Here are some examples of situations that are especially stressful:

Reading or answering questions out loud: Anxiety or nervousness causes a release of adrenaline which directly affects the function of our muscles. This can cause the diaphragm to become stiff which impairs proper breathing, or the tongue can become stiff which affects how we talk. Some students will be just fine with public reading, but it helps to be aware that others may have actual physical reactions to the stress.

Writing: Our hand muscles can be quite affected by myotonia and may cramp easily. This can make our handwriting messy and illegible. It is useful for students to learn to type as early as possible and to be allowed to do homework on a computer if they prefer. This is much easier on the muscles.

Handing in papers: Getting up out of a chair or desk after a period of sitting almost always causes the muscles in the legs to be stiff for several steps. It is not unusual to stumble and fall if the student does not pause for several seconds and allow the legs to adjust before moving. Also be aware that *handing out papers* (which is usually considered an honor) is a prime time for other students to try to trip their classmate. It may be a normal prank, but for someone with MC it can cause a serious fall because they can't recover quickly from a loss of balance.

Giving reports: This is similar to reading out loud although getting up in front of a group of people is usually even more stressful since the student also has to walk up under the scrutiny of many eyes. It is helpful to have a podium of some sort to help with balance.

Changing Locations: Moving from one class to the next or going out for recess can be challenging. The sensory overload of being jostled and then having loud bells ringing is enough to make the stiffness of the muscles worse. Most children will adapt over time, but it helps if the teachers are aware of this potential reaction. Stairs are particularly troublesome...the first two or three steps may be normal, but then the muscles will seize and it can take several seconds between the next steps as the student waits for the contracted muscles to relax. Often children behind them will get impatient and shove which just makes the situation worse and can result in a fall. Even the anticipation of going up stairs creates anxiety and worsens the symptoms.

Recess: Most of us agree that we should be allowed to participate as normally as possible in physical activities and have the freedom to make adjustments to our activity as needed. Younger children don't have as much awareness of their bodies and may fall quite often, but if they jump right up and keep going they are usually fine. The student should be allowed to sit out any games or activities if he or she is feeling particularly stiff. While we are able to push through the stiffness to some extent, this can cause damage to the muscles and results in pain and cramping.

Performances: Any type of excitement, good or bad, can increase the rigidity of our muscles. A child may be looking forward to performing for their parents and yet suddenly have trouble walking. If they are in a play, it is helpful to think through the movements required. Stairs will require some extra lead time. If there are bleachers for a concert, the student may need to be on the floor level instead. Some children with MC have almost no symptoms until puberty, so the accommodations should be evaluated on an individual basis from year to year.

Boarding buses: Since many teachers assist young students in boarding, it is important to be aware of the difficulty climbing the steps onto the bus. The student may have to pause several seconds between each step to allow the stiffness to subside.

Cold exposure: Cold can greatly aggravate our symptoms. While it is the parents' responsibility to be sure the child has appropriate clothing for the weather, teachers should be aware of the potential for increased stiffness after being outside in the cold. Often this affects the facial muscles and can make speech difficult.

Physical Education Teachers and Coaches

Everyone with myotonia congenita has horror stories about physical education class in school. Most of us were forced to participate in activities that were not appropriate for our condition and it often resulted in injuries. Students with MC do not need to be in adaptive PE classes...most love physical activity and sports, and the exercise is good for us. However it needs to be tempered with common sense and an awareness of severity of symptoms on any given day.

Some activities that may be particularly risky would be gymnastics including vaulting, balance beam, uneven bars, etc. A climbing wall, trampolines and weight training are all fine with good spotters. Swimming may cause quite severe stiffness if the pool is cool. Chlorine can also

aggravate myotonia (delayed relaxation of muscles). Any child taking swimming lessons should be instructed in how to deal with muscle cramps since that is a very real possibility for us.

Many people with MC participate in competitive sports and students should be encouraged to play on teams or compete individually if they are comfortable with it. The best choices would be sports that allow continuous movement such as soccer, hockey, or tennis. While other sports are certainly acceptable if the student desires to participate, please be aware that any period of rest on the bench or standing still for long will necessitate warming and flexing the muscles again.

Softball is especially difficult for most of us because of inactivity of our legs while at bat. We may be able to hit the ball, but when we turn to run to first base we often fall flat on our faces. Flexing and stomping the legs can help but may subject the student to teasing. Track and field events are also very challenging, especially sprints, hurdles, vaulting or long jump. Many with MC do great with middle or long distance running since the muscles have time to loosen up.

As mentioned earlier, stairs can be particularly difficult because of the delay in relaxation of the muscles. An activity like running bleachers creates a higher than normal risk of falling for someone with myotonia. Push-ups and chin-ups can actually cause pain that persists for hours.

MC especially affects white fiber muscles like the calves, thighs and abs. We are unable to make functional Type IIB fibers to adapt to stress on the muscles. Repetitive flexing of those muscles like sit-ups can be quite excruciating. Living with MC is basically doing isometrics all day long because our own muscles are resisting us. This often causes micro-tears and releases creatine kinase into the bloodstream. While MC in itself is not painful, this constant tearing of the muscles and glycogen depletion can create a great deal of pain and cramping. A student with MC will often have hypertrophied muscles with minimal effort, but that does not necessarily correlate with strength. In fact there can be sudden weakness during exercise because of increased insulin output and a shift in potassium outside the cells. This is why responsible spotting with weight lifting is critical.

If you have a student who is having a rough time with sports, please try to involve them in other ways. They can be statisticians, water carriers or even a team mascot, but it's such a valuable experience to be a part of a team that it's worth the effort to help them find a place to participate.

School Nurse

Hopefully you will not have to deal with too many emergencies related to myotonia congenita. Falls will be the most common complaint, but students may also have quite painful muscles at times. Children are usually not given medication for myotonia because of the potential side effects, but some doctors will prescribe drugs, especially for teens. The most common are Mexitil, Tegretol, Dilantin or Diamox. Parents need to provide complete product inserts for your files so that you can be alert to reactions, especially heart arrhythmias from drugs like Mexitil.

The most dangerous scenario will be if a student has an injury serious enough to require intubation for transport to the hospital. Succinicholine (Anectine) can trigger a potentially life-threatening reaction in someone with myotonia congenita. Student should always wear Medic Alert type jewelry stating "No Anectine" and "Malignant Hyperthermia" to avoid a crisis. While we don't have the true malignant hyperthermia gene, all the same precautions should be taken for

anesthesia. Emergency personnel should also be very cautious about administering anything containing potassium. Unfortunately many doctors aren't even aware of these possible reactions.

If the student has asthma and is allowed to use inhalers, you need to know that any medication containing epinephrine or which increases adrenaline can aggravate myotonia. While the asthma may improve, the diaphragm may become so rigid it appears the student still can't breathe well. An Epi-Pen can have the same consequences. You can estimate the severity of the myotonia by having the student squeeze your hand hard and try to let go suddenly. The worse the myotonia, the slower the release. (Note – the myotonic response will diminish as this is repeated.)

For muscle pain and cramping, warmth is often the best treatment. Cold weather will make myotonia considerably worse. A heating pad may help, and you may want to have parents provide a few Therma-Care type wraps. Capsaicin can also relieve stiffness and pain. Chewing a few flakes of hot red pepper can have a dramatic effect, especially when the diaphragm or ocular muscles are affected. Parents may want to provide a small sealed packet for you to have on hand from a pizza restaurant.

Rising insulin levels will increase myotonia. Educating parents about limiting sweets can help minimize this effect. A low glycemic/diabetic diet is quite effective in controlling MC. Testing blood sugar isn't always helpful since the insulin sensitivity may not correlate with low blood sugar readings. A small amount of protein may help, but once the stiffness increases it will probably persist for hours or days. Potassium is another factor in myotonic response. Eating foods high in potassium can exacerbate symptoms, but the most extreme reactions will be seen with potassium preservatives (such as those added to lunchmeats and many prepared foods) and medications which contain or increase potassium. Many parents are totally unaware of this correlation and pay no attention to their child's diet.

Bus Drivers

Climbing steps is very difficult for anyone with myotonia congenita. Please be sure the student is allowed adequate time to maneuver the steps and protect them from shoving by other students to avoid falls. Since sudden loud noises can also cause a fall if the muscles seize up, be especially cautious about honking when the student is crossing a road near the bus. If there is an accident that requires emergency transport, it is crucial to notify emergency personnel if possible about the student's condition. They may not be wearing medical alert jewelry and certain treatments or medications can trigger a life-threatening reaction called malignant hyperthermia.

Maintenance

Students with myotonia congenita are very susceptible to the effects of certain pesticides and herbicides. The most dangerous are weed killers containing 2,4-D (Weed 'n Feed type products). The other dangerous class would be organophosphates like Sevin, Dursban, Orthene, Diazinon, etc. Hopefully these are only applied on weekends when students are not in the buildings or on the grounds, but it would be helpful if parents could be notified any time these products are used so they can choose whether or not to let the child attend school.

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