

# PROTOCOL FOR SCHOOL

(Date) \_\_\_\_\_

RE: (\_\_\_\_NAME\_\_\_\_)

D.O.B. \_\_\_\_\_

(\_\_\_\_(NAME)\_\_\_\_) is a \_\_\_\_ year old child with a diagnosis of mitochondrial disease, specifically a defect in \_\_\_\_\_. Mitochondrial disease is a defect in energy metabolism. (\_\_\_\_(NAME)\_\_\_\_)'s symptoms include:

\_\_\_\_\_.

## **BACKGROUND**

Mitochondrial disorders are diseases of the mitochondria, “the powerhouse of the cell,” which generates the energy (ATP) that powers cell and organ function. When mitochondria do not function properly, organs stop working effectively. It is a multi-system disease that is genetic in origin and can be inherited in a number of ways. Multiple family members (within the same generation or sometimes in multiple generations) are sometimes affected and may manifest with different symptoms or different degrees of symptom involvement.

Mitochondrial disorders may involve any combination of a variety of body systems including the brain and muscles (causing poor stamina, altered muscle tone, muscle weakness, seizures, stroke-like episodes); autonomic nervous system (temperature dysregulation, heart rate abnormalities, dizziness, poor heat and/or cold tolerance, too much/too little sweating, skin pallor and blotching that comes on spontaneously); eyes (vision loss); hearing deficit; endocrine disease (diabetes, thyroid or parathyroid abnormalities, adrenal gland failure); heart (cardiomyopathy, arrhythmia); liver (dysfunction, cirrhosis); kidneys (failure of the kidneys to reabsorb metabolites); and metabolic issues (high blood acid and/or ammonia levels). Symptoms become especially severe during ordinary infections, with exercise, significant psychological stress, and sometimes with excessive heat or humidity. The disease is progressive and organ dysfunction can become more apparent with time.

## **CHALLENGES**

Mitochondrial disease has been recognized more since the 1980s. It continues to be a challenging disease - difficult to diagnose, medically complex, variable in its symptom intensity, progressive in its course, and completely unpredictable. There is no good treatment and no cure. Because of the nature of mitochondrial

disease, it can significantly impact a child's life - physically, psychologically and cognitively.

Living with a rare, "invisible" disease can be incredibly challenging. Many of these individuals may look "normal" without any apparent problems. Because of a lack of awareness about the disease and the variable nature of its symptoms, patients are often viewed with suspicion and the diagnosis questioned. This poses great difficulties because affected individuals already have very limited stores of energy. Managing their disease can be overwhelming; when a mother and her child (children) are affected, the challenges are multiplied. Safety nets within the health care system are inadequate for children, and poor for adults.

The following recommendations should help support a mitochondrial patient's good health and enable (\_\_\_\_(NAME)\_\_\_\_) to be more productive in a school environment. Parents know their child best and are encouraged to teach other caregivers, teachers, and therapists about (\_\_\_\_(NAME)\_\_\_\_)'s symptoms to enable them to better interpret what they are seeing in the classroom.

On a day-to-day basis, the goal for these children should be to preserve their limited energy reserve. The energy demands required for physical, cognitive and emotional activities might be significantly draining for these children. Individual support from a 1:1 aide and/or nurse may provide the support that these children need to conserve their energy, tolerate the classroom setting for longer, and avoid the risks or decompensation associated with severe fatigue. A nurse that goes to school with a child allows the child to attend school with needed medical supports on "off days." This improves classroom attendance and supports the child's education. In some cases, such as (\_\_\_\_(NAME)\_\_\_\_)'s the school day can be too exhausting and home schooling may need to be considered as an option. It is important for children that are home-schooled that their therapies, including physical therapy, occupational therapy, and school therapy be continued either at home or in the school setting.

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### **GENERALIZED/MUSCLE FATIGUE**

The hallmark of mitochondrial disease is easy fatigability or decreased stamina, often with low muscle tone. This can be apparent in the way these children carry themselves, their posture when standing or sitting, and their gait. After resting or sleeping, a child can appear to have a lot of strength but the energy dissipates rapidly, before that of his/her peers, even with ordinary activity and play. Hand fatigue may occur with writing and the quality of penmanship can change with extended writing assignments. These children may have difficulty with oral motor dysfunction due to muscle fatigue. This results in slurred speech, fatigue with chewing, and choking or gagging when swallowing. The voice can become hoarse with use as a result of vocal cord dysfunction. Eyelids can become droopy. Their breathing muscles can become weak and they may become easily short of breath, complaining they cannot “catch their breath” even without prior exertion.

Like all people but more prominently in this population, energy levels fluctuate from day to day. Physical exertion, psychological stress (especially anxiety and depression), heat and humidity, and infectious illnesses are big “energy drains.” Mornings generally offer a better opportunity for learning although it may be hard for some patients to get up and get going in the early morning (since sleep is often disturbed due to sleep apnea).

Students should also be encouraged to pursue interests and activities outside of school. It is optimal for individuals with mitochondrial disease to stay conditioned; deconditioned muscles use up more energy to do the same work. School days may need to be tailored to accommodate these interests. This is important not only to enhance mitochondrial function but to improve self-esteem and socialization skills. Parents, children, and schools should work together to fashion a reasonable plan.

Guidance counselors, school psychologists, social workers and teachers should consider advising and supporting these students to pursue course schedules/careers that are “product-oriented” (focusing on the end result – projects, exams) rather than time- or process-oriented (e.g., regular attendance during set hours, class participation). This kind of flexible approach enhances performance and productivity. For the child in preschool and elementary school provisions should be made for child to meet academic expectations and complete projects at home or on a flexible schedule. Parents and/or tutors should be instructed how they can best support their child’s learning at home. The educational process for these children may need to be a year long, shorter day program rather than the traditional 180 school day calendar. Home schooling and/or summer learning programs may need to supplement the regular school year calendar.

***Recommendations:***

1. Parents, the student and the school need to work together to develop a day schedule that alternates “higher energy” and “lower energy” activities according to what the child can handle.
2. The day should include regular periods of rest or breaks. This might need to be in a location away from stimulating environment like the classroom.
3. Because a mitochondrial student's energy level can be very fluctuant in an unpredictable way, it is important that the school day schedule be flexible enough to accommodate this unpredictable variability. On "good days", a child may need only a few breaks or rest periods mixed in with more active subjects (physical education and recess are considered active times). On "bad days", most of the day may need to be a "low activity" day with expectations for learning that day dropped to a more realistic, manageable level; learning may have to be more passive. This kind of flexibility may, in the long run, allow a student to be more productive and able to demonstrate better what he knows and what he is capable of learning. Some children do even better if the school day is shortened, others if they attend school two, three, or four days a week with home tutoring during "home days". At times, it is important to introduce a child slowly to a particular routine to determine how he tolerates it. Whatever approach is begun, it is important that it be re-evaluated from time to time to make sure it remains the most appropriate plan.
4. Where children are more limited developmentally, those activities in which a student can show achievement, maintain motor skills and gain self-esteem should be identified. Energy should be conserved to allow the focus to be on the most important activities.
5. Mornings are usually the most productive time of day for children with energy disorders; as the day goes on they are often less able to attend or maintain the stamina to stay on task.
6. Activities involving reading and writing should include regular breaks, and could be augmented by approaches that reduce fatigue. For reading, someone could read a work assignment. Using a computer keyboard may reduce energy demands on the muscles of the hand while also contributing to a higher quality of finished product, something a student can feel proud of.
7. Physical support may be needed during circle time, or school assemblies when sitting on the floor. Assistance may be needed for standing. Strollers or wheelchairs may be required for field trips.
8. An adaptive physical education program might be more appropriate for some children.
9. Ongoing physical and occupational therapy may be needed to maintain strength, tone and flexibility. Speech therapy can help maintain and enhance oral motor skills and can teach the child about voice conservation.
10. Daily communication/check off form from home to school and school to home to report level of activity, exertion, stamina, eating, drinking and other comments.

11. Based on these individual's low stamina, unpredictability and their inconsistent attendance, alternative educational programs may need to be considered. There may need to be a combination of school and home based educational services to meet these children's educational needs.
12. School and educational programs may need to be provided on a full calendar year schedule rather than a school calendar schedule in order to accommodate their learning style and needs.

## **PAIN AND DISCOMFORT**

There are many reasons for children with mitochondrial disease to have pain, including:

- muscle pain (often following physical exertion)
- headaches and migraine (often related to poor eating or inadequate liquid intake)
- stomach aches from reflux, bloating, constipation
- nerve pain or burning or numbness/tingling in the limbs

Children in pain may become irritable or withdrawn, pale, and/or appear to be in pain. They may describe their symptoms.

### ***Recommendations:***

1. Assess for behavioral changes, triggers of discomfort.
2. Work with the parents re: measures for preventing triggers and treating pain.

## **AUTONOMIC DYSREGULATION**

The autonomic nervous system regulates certain body functions without our conscious control, including heart rate, blood pressure, temperature regulation, sweating, skin color (pallor, flushing), gut motility, and so on. Most mitochondrial patients have some degree of autonomic dysfunction.

Symptoms can include unusually low or very fluctuant body temperature, quick overheating or getting cold easily (heat and cold intolerance), dizziness and lightheadedness, fainting, rapid or slowing of heart rate, reduced sweating (as in heat) or inappropriate sweating, and skin color changes (like pallor or flushing or mottling of the skin). Seasonal changes (particularly spring to summer) are often associated with difficulties. When a patient is coming down with an infection, or when other medical issues become more active (e.g., constipation, bad reflux), the autonomic symptoms generally become more prominent as well.

### ***Recommendations:***

1. Note coloring and complexion (pallor, flushing, mottling), balance.

2. Ensure a comfortable ambient temperature. Learning programs in hot weather should occur in air-conditioned classrooms. Transport vehicles for field trips should be air-conditioned during warm days.
3. In warm weather, ensure adequate hydration. A water bottle (or equivalent) should be available at all times and they should be reminded to drink.

## **NUTRITION/LIQUIDS**

People who don't eat well can become weak. Those with energy disorders can become very fatigued if they don't eat or drink their daily requirements. Because of any number of problems (such as swallowing issues, or bad reflux, or a slowing of gut motility), some children eat slowly or can only eat and drink small volumes at a time. It is only through "grazing" that they can meet their nutritional goals over the day. Failure to eat or drink adequately can directly impact their energy levels and can lead to headaches and irritability or poor behavior.

### ***Recommendations:***

1. Know the child's cues of fatigue and hunger
2. Allow child a more flexible schedule for eating and drinking, as needed.
3. Children should be allowed water or recommended liquid at their table or desk and be reminded to drink.

## **BEHAVIOR**

Children with mitochondrial disease may have developmental and/or behavioral issues secondary to their disease. These may be related to fatigue, poor calorie intake, pain, or the way the disease impacts their brain. The children may have difficulty with focusing or being attentive to a task. They may seem irritable, or withdrawn, or uncooperative, and may have difficulty connecting with their peers.

Fatigue can have a direct bearing on behavior. If a child's behavior deteriorates significantly primarily during periods of fatigue, s/he might not have the energy to control his/her behavior at those times. A general approach that reduces fatigue can help reduce the frequency and severity of such "melt-downs". Measures to help the child control behavior are also useful in reducing the "energy drain" caused by those behaviors. Sometimes a child will "hold it together" while at school and decompensate at home; other times, the classroom is more aware of it.

Addressing the fatigue by pacing learning and activities, allowing breaks as needed, and optimizing calorie and liquid intake can also improve behavior.

An anxious child uses large amounts of energy to feed that anxiety; that energy is diverted from more productive pursuits. Anxiety needs to be identified and treated aggressively.

***Recommendations:***

1. Know those triggers that set off behavior.
2. Address the factors that cause fatigue.
3. Children who struggle with meeting their calorie and/or fluid need support to accomplish these goals. This might include regular snacks and/or liquids available at the desk.
4. Structure and predictability in the classroom are often critical to success. Small group sessions when possible are probably better tolerated.
5. Identify the child's strengths. "Catch them doing something good!"
6. Assist the child in making and maintaining friends. This takes a lot of energy. Provide opportunities to help him "fit in with peers."

**PREVENTION OF INFECTIONS**

Children with mitochondrial disease can become unusually weak during infectious illnesses. The fatigue can be prolonged and debilitating and may outlast the other symptoms of the infection, going on for days or weeks. Precautions should be taken to reduce the number of infections that these children contract.

***Recommendations:***

1. Reduce the number of exposure to other children with infections
2. Inform parents when there is an infectious illness in the classroom
3. Enforce regular hand-washing at school (and home)
4. Keep vaccinations should be up to date, including seasonal vaccinations, e.g. influenza
5. Implement any reasonable routine that helps promote personal hygiene in the classroom

**SUMMARY**

Children with mitochondrial disease can be a challenge medically. Their needs are complex. However, with adequate support, they can often shine academically. Without support, they are frequently unable to achieve their full educational potential. The goal for children is to meet the academic requirements to advance to the next grade level. Please call with any questions or concerns you may have about (\_\_\_\_(NAME)\_\_\_\_)'s or his mitochondrial disease and any assistance we can provide in developing a medical plan for (\_\_\_\_(NAME)\_\_\_\_) for educational program.