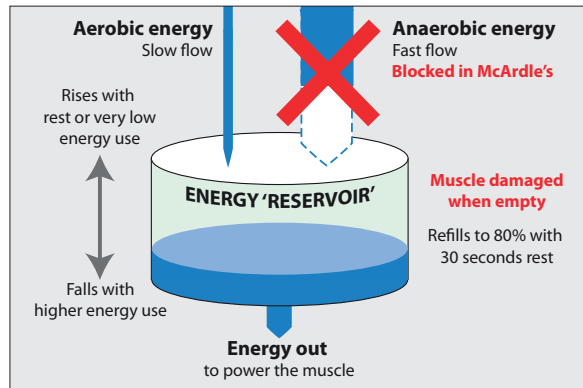


## This is the problem

- The condition arises as a result of an inherited gene mutation.
- The 'fast' anaerobic energy pathway is blocked.
- This results in a severe shortage of energy at times.
- Cramping/pain occurs if the energy 'reservoir' is emptied.



## Degrees children are affected

The degree to which children are affected varies greatly, from those in a wheelchair to those playing sports. Due to delay in diagnosis, children may be very incapacitated at diagnosis, but with advice and support should improve over time.

## The risks they can face

Pain, cramping, exhaustion, embarrassment are usual. Here are just two examples of larger issues.

### An everyday problem to avoid

- Walking on the level at their friend's pace, in a minute their legs feel heavy. Embarrassed, they struggle on. Cramping starts. They slow down but it *really* hurts. Legs like cement, they can hardly move. Heart pounding, feeling sick and light headed, they grind to a halt. Their friend says "You are *really* unfit."

### A risk of a rare complication of a contracture

- They carry a grocery bag from the car, it's heavy but they manage. Their hand is making a fist, their fingers won't open. More than a cramp, a fixed contracture - muscles swollen, locked solid. The pain is extreme. Next morning their hand is tingling, numb and an odd color. Off to hospital. After tests, the doctor says "You have compartment syndrome, we have to operate immediately".

## Further information

The child's parents can probably share these publications with you, or you can request copies via [IamGSD](http://IamGSD.org).

### Information/Emergency Card

Folding card. Suitable to be carried at all times. Brief explanation of the condition, plus reminders of when to seek medical assistance.

### 101 Tips for a good life with McArdle Disease

164 page paperback. Easy to read guide with practical tips.

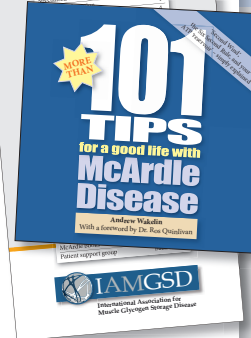
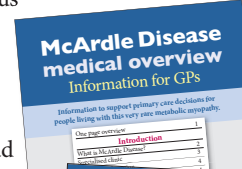
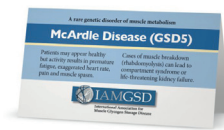
### McArdle Disease: medical overview

20 page booklet. A quick guide, primarily for the family GP and other medical professionals.

## More questions?

If you have any further queries, the [IamGSD](http://IamGSD.org) will be happy to assist.

[www.iamgsd.org](http://www.iamgsd.org)



# At school with McArdle's

## Information for schools

A plain language guide to McArdle Disease and how to safely accommodate and support a pupil with this muscle disorder.

- McArdle's is an ultra rare inherited condition which restricts fuelling of skeletal muscles.
- Children with McArdle's cannot utilize the energy stored in the muscle.
- They have a serious shortage of energy early in any activity, and throughout all *intense* activity.
- They can develop cramping and pain. If activity continues, muscles become stiff, swollen, very painful and hospitalization may be required.
- They have a 'second wind' somewhat reducing their symptoms after about 10 minutes of careful activity.
- They need to keep active to keep their muscles functioning and healthy.
- They achieve a lot by utilizing practical and management techniques to protect themselves.
- Some simple adjustments and allowances by the school are an essential contribution to the child's safety and on-going improvement.



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International Association for  
Muscle Glycogen Storage Disease

## Strategies used by children

You will notice that children use these key strategies to help them handle their physical activity and avoid pain. *They are not being lazy or intending to be disruptive.*

### Slowing or pausing to balance energy

During even simple activity like brushing teeth, drying after a shower or walking on the flat or up a flight of stairs, children will:

- Slow down to ease the energy demand.
- Pause for 30 seconds rest, then continue, perhaps more slowly.
- Do these frequently in the first minutes of activity.
- Find symptoms easing after about 7 to 10 minutes, when a 'second wind' occurs as other energy pathways start up.
- With each muscle group used, this process has to be repeated.
- In 'second wind', a child may behave normally but is still at risk.
- 'Second wind' will be lost due to inactivity during a lesson.

### Breaking intense tasks into sections

During maximum intensity tasks (such as carrying a heavy object or standing on tip toe) children with McArdle's will:

- Interrupt the task after 6 seconds.
- Rest for 30 seconds so the energy 'reservoir' refills, then try again.
- This avoids a painful muscle contracture which would otherwise occur and last for days, possibly needing medical attention.



### Resting before a risk

Before a risk, such as crossing a road or lifting something heavy, children will often first rest for at least 30 seconds, to ensure enough energy is available to get them safely through.

## In the event of overdoing it

Occasionally, a child may accidentally do something which causes them longer term pain.

- Allow them to sit down, rest and have water to drink.
- If they are not recovered within 10 minutes, call a parent.
- If severe, consider whether they should be taken to hospital†.
- The emergency card (see back page) reminds of the decision points to consider.

† Once diagnosed, advised, taught techniques and returned to aerobic fitness, most children are very seldomly hospitalized.

## Ideas on allowances

Schools and must liaise with the parents and discuss any concerns. The following notes will help to explain the areas of need.

### ■ Make sure that they stay active, at their own pace

It is vital that children are as active as possible, but always utilizing their strategies, such as outlined opposite. If restricted from activity children will quickly suffer loss of aerobic fitness and muscle mass, both of which will make symptoms worse and increase the risk of muscle damage.

### ■ Brief the sports and physical education staff

Children must be allowed time to get into 'second wind', see opposite. They must have the right to not engage in activities or stand out mid-activity, at their own decision and without question. Isometric and other anaerobic activity at maximal effort must be limited to 6 seconds.

### ■ Don't keep them in a wheelchair

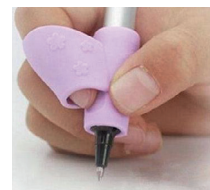
Diagnosis is often delayed, with children wrongly treated. If put in a wheelchair they become very deconditioned. Support them to walk as much as they can, with the wheelchair only for when they want it, not for the convenience of others; e.g. do not enforce use of a wheelchair on school trips, but instead provide support.



### ■ Provide an extra set of books

If printed textbooks are used, provide two sets so that they do not need to carry them between home and school.

### ■ Provide help for writing



McArdle's affects all activity, especially repetitive movements such as writing and isometric activity such as gripping. Encourage the child not to grip or press hard. Offer rubbery grips for pens and pencils to make gripping easier. Consider the need for an iPad/tablet.

### ■ Allow extra time for written exams

For many children there is a requirement for extra time in exams due to a slow speed of writing and a need for rests. There is no known link to intellectual impairment.

### ■ Authorise to use the elevator/lift

At an appropriate age, and with training, allow the child to use the elevator/lift to move up floors when changing classrooms.

### ■ Allow time to avoid rushing

Rushing between classrooms can be very harmful, especially if caught up in the throng. Consider allowing children to leave the classroom a few minutes early.

### ■ Be understanding of lateness

Despite best efforts, children can be late for school. They should not be admonished in front of other children.

### ■ Provide an accessible locker

Children may struggle to reach their locker and return to the classroom, especially if carrying extra items such as PT kit. Provide an easily accessible locker or an alternative safe place for them to leave possessions.

### ■ Allow water in the classroom

Muscles are 80% water, so good hydration is important. Allow the child to have access to water to drink in the classroom.

### ■ Watch out for peer pressure and bullying

Staff need to be aware of the danger of isolation and of bullying. But also children should be supported to resist peer pressure to perform activities that they know will hurt them.

### ■ Support during days off sick

Illness can be very debilitating with McArdle's, often requiring time off that other students would not need. Even a simple cough can cause cramps due to over stress of chest and stomach muscles. Children should not be penalized, but supported with work to do at home.



## For older students

### ■ Consider anxiety and depression

As a chronic condition, with self-image issues and a risk of episodes needing hospitalization, depression and anxiety may be an issue. Do not single out or draw attention to the student, even with good intention.

### ■ Respect their own decision making

With training and experience, students will become adept at making their own decisions about their need to return home or to seek medical attention.

### ■ Understand always at risk

Older children may have learned to manage their condition well. At times (especially in 'second wind') they may appear normal in their activities. However, this is misleading. They always remain at risk of severe episodes.