

Why they need to exercise

Given the issues, it might seem best for people with McArdle's to avoid all physical activity. However...

- Increased aerobic fitness can make it easier and quicker to enter "second-wind" (see inside).
- Both increased aerobic fitness and increased muscle mass and strength make activities of daily living easier and avoids serious disability.
- Exercise helps with weight loss (there is a tendency to be overweight and at increased risk of insulin resistance).
- Regular exercise helps with mood (a chronic condition is challenging).

How you can help

- Guide/supervise use of gym equipment and facilities.
- Allocate equipment (e.g. desk cycle, treadmill, static cycle, cross-trainer) in a quiet area/time, whereby the McArdle person will not feel embarrassed by slow pace and rests in front of other gym users.
- Discuss/log results and help with motivation.
- If required, help them to recognize "second-wind" using the 12-Minute Walk Test by tracking their walking speed, heart rate and pain level [1].
- Guide strength training with low-number repetitions to be carried out after achieving "second-wind".
- Check on their symptoms during and at the end of the session.

Partnership

- Trainer and trainee need to form a partnership to bring together their respective training expertise and understanding of McArdle disease.
- Both parties listening to, respecting and trusting each other.
- The McArdle person needs to agree to the points in the box inside.

In event of overdoing it

Very occasionally, a person with McArdle's may get something wrong and hurt themselves.

- Allow them to rest and have water to drink.
- Ask if you should call a relative or friend or, in rare severe cases, take them to hospital.
- They carry a card to remind of decision points.

More: "Emergency" button on www.iamgsd.org.

Training Support

The following are available from www.iamgsd.org on the menu Medical / Training Support.

- [1] **12-Minute Walk Test:** guidelines for this test to show "second-wind" and monitor progress.
- [2] **"Second-wind":** a full explanation in the "Living with McArdle Disease" booklet.
- [3] **Rating scales:** to assess pain and exertion.
- [4] **Further guidelines:** video presentations and more detail on aerobic and strength training.

Further information

A range of publications available on our web site. Browse on-line or download free PDFs:

Medical overview: A quick guide for medical professionals. 20 pages.

101 Tips for a good life: Simple, practical tips for people living with McArdle's. 164 pages.

Questions? Contact us on info@iamgsd.org.

DISCLAIMER: IamGSD uses its best endeavors to provide accurate, factual and up-to-date information on this ultra rare condition. However, each person must take into account their own circumstances, diagnosis, and any additional genetic and medical factors and is advised to consult with their doctor before making use of our generic information and guidance. Please refer to the disclaimer on the Medical menu of our website.



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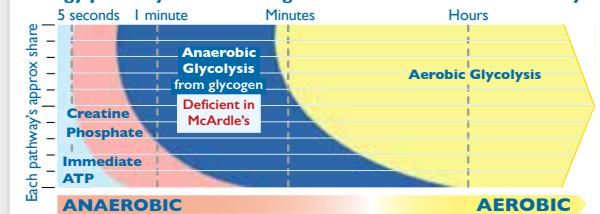
At the gym with McArdle's

Briefing for physical therapists and personal trainers

Explaining the problems for people with McArdle disease, why they need to be physically active and how you can help.

- McArdle's is an ultra rare genetic condition.
- Myophosphorylase is deficient, so glucose cannot be released from muscle glycogen.
- A severe energy crisis arises early in all physical activity and exercise.

Energy pathways at increasing duration of submaximal activity



- In activities such as walking at a normal pace cramping may start within a minute or two.
- In maximal-effort isometric and anaerobic activity, duration should be limited to about six seconds, otherwise cramping will start.
- Cramping may develop into a contracture with rhabdomyolysis, myoglobinuria and a risk of acute kidney injury or compartment syndrome.



“Second-wind”

- Early in aerobic activity, as the start of cramping is felt, people with McArdle’s slow their pace or pause for a rest.
- They rest for at least 30 seconds to recover approx. 80% of immediate ATP and the creatine phosphate system.
- By managing pace and pauses, after about 6 to 10 minutes a “second-wind” is achieved, largely specific to the muscle groups being used
- Activity then becomes somewhat easier.
- Glucose is released from the liver glycogen store, and fat metabolism and aerobic glycolysis start.
- “Second-wind” is lost within about 30 minutes of inactivity of the affected muscles.

More: In the “Living with McArdle Disease” booklet, from IamGSD [2].

Assess the individual

McArdle’s can manifest differently between individuals due to:

- Additional conditions or genetic factors.
- Loss of aerobic fitness and/or muscle mass, often due to under-use.
- Damage caused by incorrect management, often before diagnosis.

Assessment: Assess the individual by using the 12-Minute Walk Test [1] and discussing their current fitness, so as to agree their starting level. Plan to start gently, in case badly affected.

No pain = Gain

- “No pain, no gain” does not apply in McArdle’s as damaging cramps and pain develop much sooner.
- In the event of pain exceeding 3 on a scale of 0 to 10, the exercise should be slowed or paused.
- If pain rises rapidly, immediately stop the exercise.

The contents of this leaflet and the Training Support web page are based on the Clinical Practice Guidelines published in “Neuromuscular Disorders”, the journal of the World Muscle Society, December 2021.

Aerobic training

- Light warm-up during which the rating of perceived pain should not be allowed to exceed 3 (moderate) to 4, on the scale of 0 to 10, where 0 is no pain and 10 is maximum pain) [3].
- Once in “second-wind”, regular moderate-intensity aerobic exercise should improve muscle metabolism.
- In training, heart rate should be 50% to 75% of max. HR calculated as: $208 - (0.7 \times \text{age})$.
- Training 2 to 4 x week, 20 to 90 mins per session.
- After session, light dynamic stretching and good hydration are recommended.

More: Further guidelines on aerobic training [4].



The McArdle person must:

- Seek personal medical guidance before commencing training.
- Establish their baseline CK through several blood tests (it is typically 2 to 25 x top of the normal range, or approx. 400 to 5,000 IU/L).
- Accept responsibility for compliance with the “No pain = Gain” statement.

Creatine Kinase

- Plasma CK (also known as CPK) is a marker of muscle damage.
- If concern arises regarding pain or cramping, a CK test will help assess whether training has been too intense (higher than baseline CK).

Strength training

Warm up

Walk or pedal to achieve “second-wind” (as for aerobic training), continue for 12 minutes. Then do 12 minutes upper body exercise to achieve “second-wind” in upper body. You only get “second-wind” in the muscles you are using.



Strength training

Use equipment to exercise large muscle groups. Sets of 6 reps. Gentle dynamic stretching for 10–30 seconds after each set, to reduce stiffness. Circuit training structure, rotating on multiple equipment, recovery of 3 minutes between sets.

Example: 3 to 4 turns to circuit: Leg Press → Chest Press → Seated Rowing/Lat Pull Down → Abdominal Machine.

Load: should be set to elicit a rating of perceived exertion (RPE) of 6-7 and a rating of perceived pain (RPP) of 0–1 (both on scales of 0–10) [3]. Load can be increased when RPE is <6 for two consecutive sessions.

Warning: If perceived pain rises rapidly, immediately stop the exercise or risk contracture. While resting, undertake dynamic movements without load, and gentle stretching, until this perception disappears, then continue circuit.

Cool-down

Low intensity dynamic exercise. End with global passive stretching, hydrate with plain water.

Repeat

Up to 3 non-consecutive days per week.

More: Further guidelines on strength training [4].