The National Hospital for Neurology and Neurosurgery

National Commissioning Group: McArdle Disease Service

Exercise and McArdle Disease
Contents

1. Introduction ........................................... 3
2. What is McArdle disease? ......................... 3
3. How does McArdle disease affect exercise? 3
4. Current exercise recommendations ............ 5
5. What exercise is safe for me to do? .......... 5
6. Using the CR-10 Scale ............................... 5
7. What can physiotherapy help with? .......... 7
8. Where can I get more information? .......... 7
9. References ............................................ 7
10. Contact details ..................................... 8
1 Introduction

This leaflet provides information on how to exercise safely for people with McArdle disease.

2 What is McArdle disease?

McArdle disease, also known as glycogen storage disorder type V, is a rare metabolic muscle disorder characterised by a deficiency of the enzyme muscle phosphorylase.

This enzyme is essential for the process of converting glycogen to glucose. Therefore people with McArdle disease are unable to use the glycogen stored in the muscle as an energy source in the first few minutes of exercise as they are unable to break this down to glucose.

3 How does McArdle disease affect exercise?

Energy (ATP) stored in the muscles supplies the first 10 seconds worth of energy and is used for short bursts of exercise such as picking up shopping bags. After this has been used up, the body will move onto anaerobic and aerobic metabolism to fuel exercise.

Anaerobic metabolism provides energy by the breakdown of glucose without the need for oxygen. Anaerobic metabolism produces energy for short, high-intensity bursts of activity, such as walking up stairs, running and squatting. In McArdle disease, the enzyme used to convert glycogen to glucose is absent and therefore people with McArdle disease are unable to use the anaerobic pathway as an energy source.

Aerobic metabolism fuels most of the energy needed for less intense, longer duration activity, such as walking or cycling on the flat. It uses oxygen to convert nutrients (carbohydrates, fats, and protein) to ATP. This system is a bit slower than the anaerobic system because it relies on the circulatory system to transport oxygen to the working muscles before it creates ATP. When this energy source is used by people with McArdle disease, exercise becomes easier and you may feel yourself getting into a ‘second wind’.
Anaerobic exercise
Use of glycogen stored in the muscle at the start of activity and during strenuous activity such as running, carrying shopping or squatting.

People with McArdle Disease are unable to convert this glycogen into glucose, which results in pain and muscle cramps within a few minutes of starting exercise.

Slowing down or stopping to rest for 30 seconds helps to manage the cramps during this painful period.

Continuing to exercise through intense pain will result in muscle damage. This should be avoided.

Aerobic exercise
Use of alternative energy sources from fats and sugars supplied from the liver.

This energy source becomes available 7-8 minutes after the start of exercise, as the aerobic pathway requires oxygen and is dependent on the circulatory system to transport the fats and sugars to the muscles.

The switch to this energy source is known as the second wind in people with McArdle Disease. Symptoms of fatigue and cramping ease and exercise becomes easier.
4 Current exercise recommendations

The current guidelines recommend that adults should aim to do moderate intensity aerobic exercise for 150 minutes each week, in bursts of at least 10 minutes (Department of Health, 2011; American College of Sports Medicine, 2013). This includes activities such as walking or cycling.

Research has shown that aerobic training in people with McArdle disease increases exercise capacity and cardiovascular fitness without muscle damage (Haller et al, 2006, Quinlivan et al, 2011). Therefore, we recommend that people with McArdle disease participate in gentle aerobic exercise, such as walking or cycling, on 3-5 days per week, aiming to exercise for at least 30 minutes on each occasion.

There is emerging evidence that strengthening exercises may be beneficial for people with McArdle disease. However, these should only be done under the guidance of a physiotherapist experienced in treating people with McArdle disease. Incorrect advice and exercises may cause you to have muscle pain and cramps.

5 What exercise is safe for me to do?

Aerobic exercise, such as walking, is the safest type of exercise. It is important to start off slowly. If you experience leg pain that is moderate to strong (4 on the CR-10 scale, see below), slow down or stop and rest for 30 seconds until the pain starts to subside, before starting to walk slowly again.

When in your second wind (exercise becoming easier with less muscle pain), try to exercise at a moderate intensity. You should still be able to talk while exercising.

Aim to exercise for 30 – 40 minutes, three to five times per week, but remember to gradually build the time up slowly over several days or weeks.

6 Using the CR-10 Scale

In the McArdle Clinic at The National Hospital for Neurology and Neurosurgery we use the CR-10 scale, as shown below (Borg, 1970). You may find it useful to use this scale when you are exercising at home to monitor your muscle cramps and identify when you reach your second wind. Your pain should not exceed Level 4 on this scale (moderate to strong pain). If it does, slow down or stop and rest for 30
seconds before continuing with exercise. Pushing through intense pain will cause muscle damage and therefore, this should be avoided.

**CR-10 Scale**

<table>
<thead>
<tr>
<th></th>
<th>Description</th>
<th>Rating</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Nothing at all</td>
<td>No pain</td>
</tr>
<tr>
<td>0.3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>0.5</td>
<td>Extremely weak</td>
<td>Just noticeable</td>
</tr>
<tr>
<td>1</td>
<td>Very weak</td>
<td></td>
</tr>
<tr>
<td>1.5</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Weak</td>
<td>Light</td>
</tr>
<tr>
<td>2.5</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Moderate</td>
<td></td>
</tr>
<tr>
<td>3.5</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Strong</td>
<td>Heavy</td>
</tr>
<tr>
<td>6</td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>Very strong</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9</td>
<td></td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>Extremely strong</td>
<td>Maximum pain</td>
</tr>
<tr>
<td>11</td>
<td>Absolute maximum</td>
<td>Highest possible</td>
</tr>
</tbody>
</table>

You may also find it helpful to measure your heart rate during exercise. You will notice that your heart rate initially increases as a physiological response to the muscle pain. When the muscle pain starts to subside and you enter your second wind, your heart rate will reduce. As you continue to exercise, any further increase in heart rate is related to cardiovascular fitness. Your heart rate should be maintained between 60 – 70% of your maximum heart rate during exercise once you are in your second wind. Your maximum heart rate is worked out as 220 minus your age.
7 What can physiotherapy help with?

The McArdle disease specialist physiotherapist at The National Hospital for Neurology and Neurosurgery can help with:

- Exercise training and practice identifying your second wind
- Guidance on appropriate exercise to strengthen weaker muscles, such as your core stability muscles
- Treatment of musculoskeletal problems that may be exacerbated by your McArdle disease, such as back pain or shoulder pain
- Liaison with your local physiotherapist, including providing information on McArdle disease

8 Where can I get more information?

The Centre for Neuromuscular Diseases
http://www.cnmd.ac.uk/

The Association for Glycogen Storage Disease UK
http://www.agsd.org.uk/

Muscular Dystrophy Campaign
http://www.muscular-dystrophy.org/

9 References

- Department of Health (2011) *Start Active, Stay Active: a report on physical activity from the four home countries*. London: Department of Health

**10 Contact details**

If you would like any further information, or you would like to book an appointment with the specialist McArdle disease physiotherapist, please contact:

Specialist Neuromuscular Physiotherapist – McArdle disease  
Direct line: 020 3448 8034  
Extension: 88034  
Switchboard: 0845 155 5000 (there is no additional service charge for using a 0845 number. The cost is determined by your phone company’s access charge) or 020 3456 7890  
Fax: 020 3448 3633  
Email: enquiry.mcardle@uclh.nhs.uk  
Website: www.uclh.nhs.uk

**Space for notes and questions**