



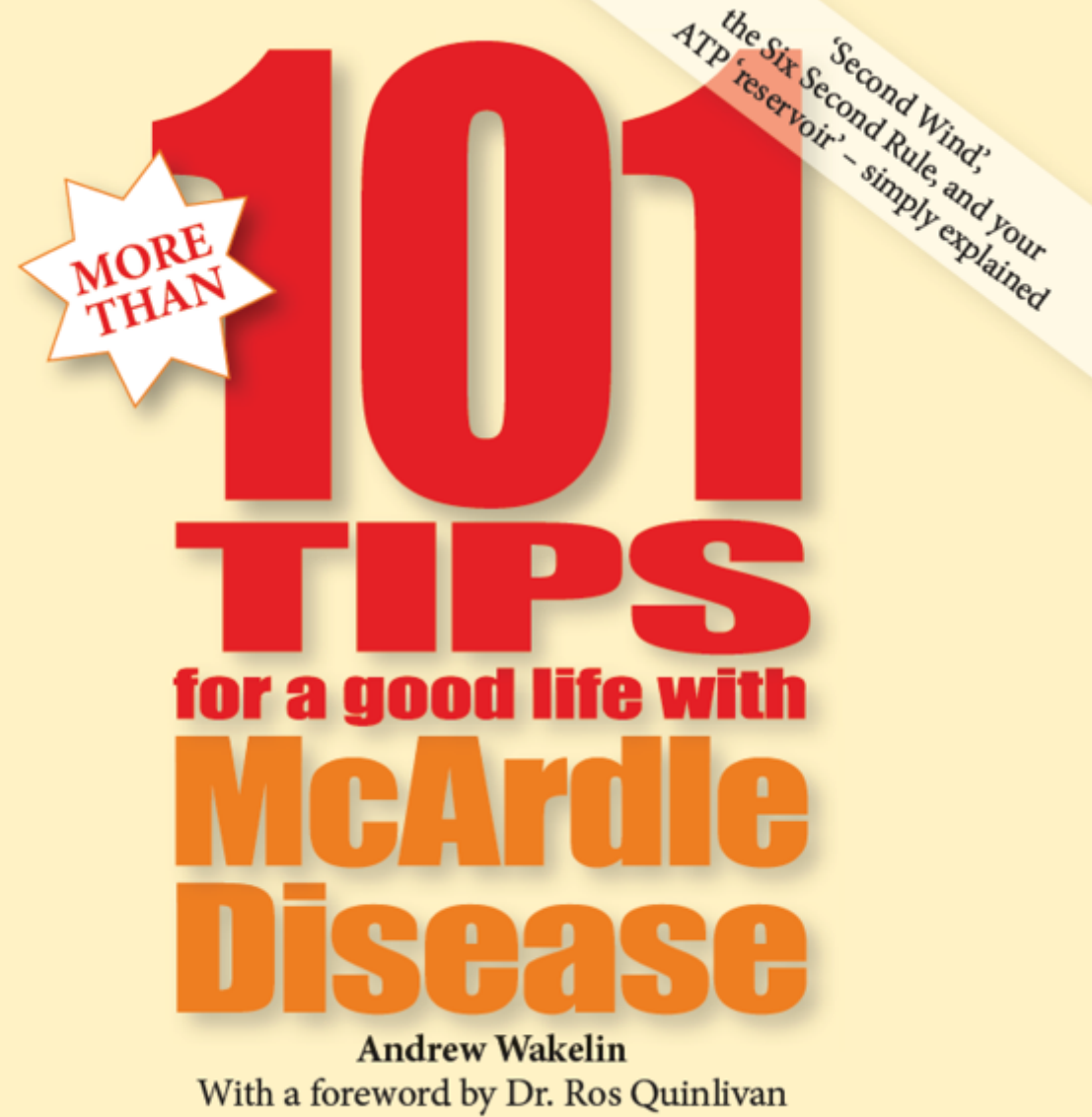
Top Tips

for managing McArdle activity
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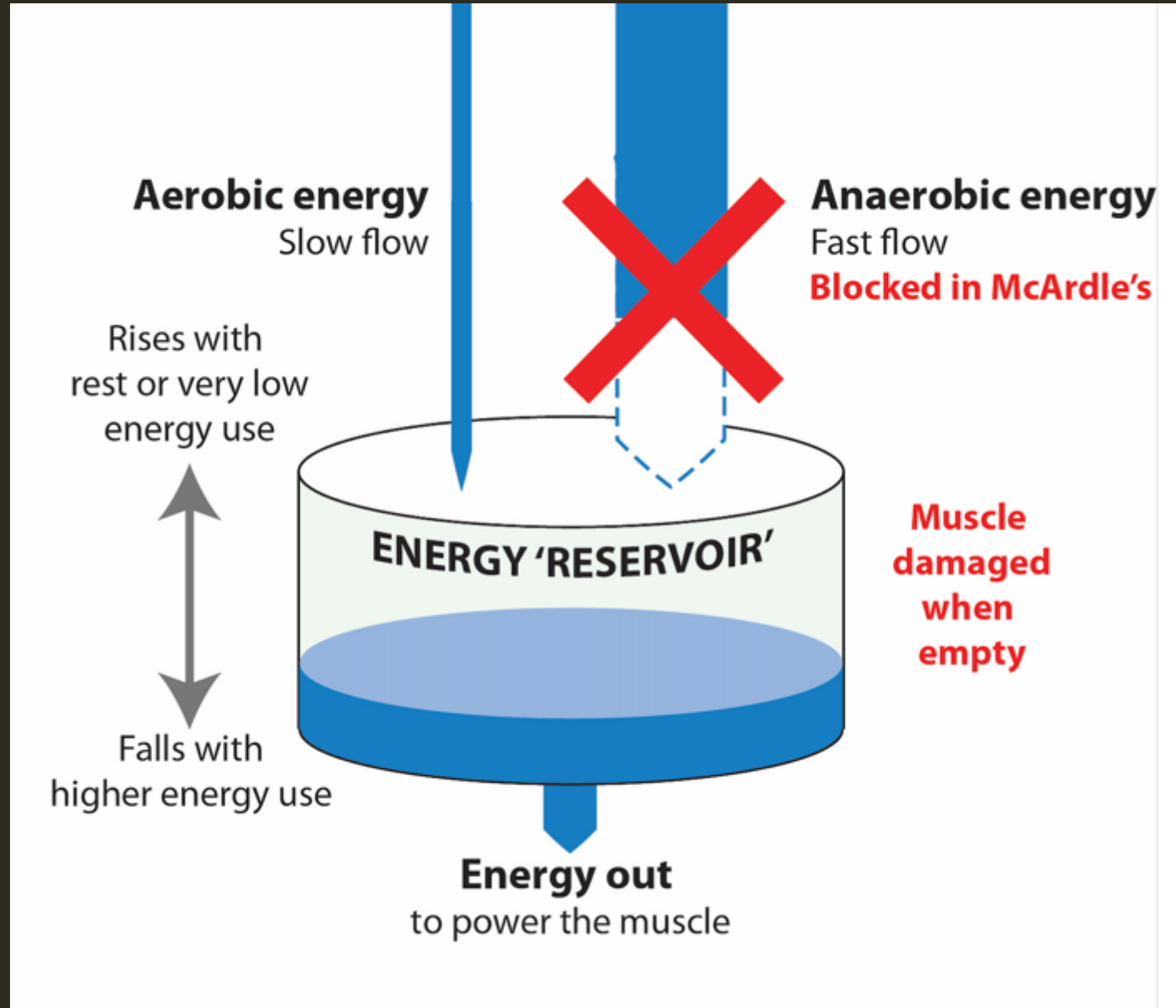
Key aspects underlying all tips

- The immediate energy 'reservoir'.
- The four Rs.
- Second wind.
- The 6 second rule.
- 30 for 80.
- Rest before risk.



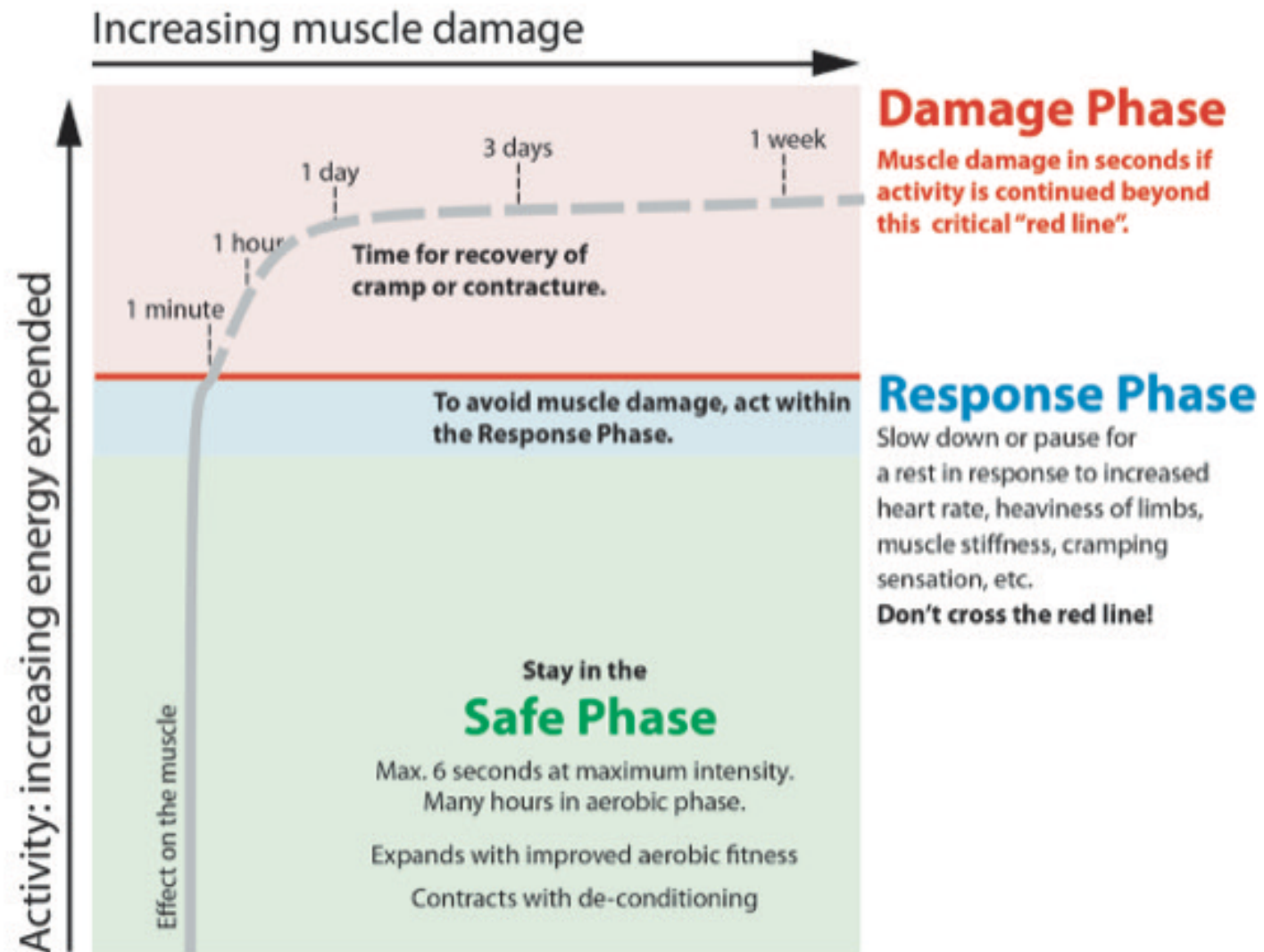
The immediate energy reservoir

- Energy store in the muscle cell.
- ATP - last metabolic step ready to activate muscle.
- Visualise as a 'reservoir'.
- Don't empty the 'reservoir'.
- That is when muscle damage occurs.



The four Rs: Recognise Respond Recover Resume

- Once McArdle pain starts, if activity continues muscle damage accelerates rapidly.
- Lear to recognise the early signals.
- Slow down or pause for a rest.



Second wind

- This is when light to moderate activity/exercise becomes noticeably easier.
- A vital aspect of McArdle's.
- Universal to all McArdle's people.
- Some people need help to recognise it.

'Second wind'

- When starting activity if you feel your muscles run out of energy, or a cramping sensation, slow down. If the pain doesn't fade, stop for a rest. Once it has faded you can resume activity.
- After 6 to 10 minutes more blood and oxygen is flowing to the muscles, which means they can rely on fat stored in the muscle, and on fat and glucose circulating in the blood.
- Our liver releases glucose from its store of glycogen.
- 'Second wind' partly relieves inaccessibility of muscle glycogen.
- You will learn to recognise it. Heart rate is a good indicator, it will drop as you enter 'second wind'.
- It is not fully systemic. Get each muscle into 'second wind'.
- Even when in 'second wind' if you raise the exercise intensity you can get muscle pain and contracture, so pace yourself.
- Exercising in 'second wind' will train the muscles, improving their aerobic capacity – this makes 'second wind' easier to achieve and enables you to work harder.
- Ideally we should get into 'second wind' and then continue exercising for another 30 to 45 minutes, and do this most days.

There is more about 'second wind' on the AGSD-UK web site.

Second wind demonstrated

- Heart rate increases gradually to minute 7.
- Pain increases to minute 6.
- Both reduce in minute 8 onwards.
- At minute 12 both similar to start, even though pace now 66m compared to 55m.

Time	Walking speed	Distance covered (m)	Heart rate	Borg Pain Rating
1	-	55	90	0
2	-	120	95	1
3	Slowed down	180	100	2
4	-	240	109	3
5	Slowed down	290	108	3
6	-	344	113	2.5
7	-	404	116	2
8	-	460	111	1.5
9	-	530	111	1.5
10	-	590	106	1
11	-	664	107	0.5
12	-	730	104	0

Resting heart rate: 67; Baseline symptoms: none; Distance completed: 730 m.

The 6 second rule

- When making maximal effort (e.g. opening a jar, lifting something very heavy or sprinting).
- The energy reservoir will be emptied in <10 seconds.
- Count to 6 seconds and stop.
- Rest, or use other muscles.
- Then try again.

Six second rule

- When doing something at maximum intensity the instantly available energy lasts for only 5 to 10 seconds².
- Examples: opening a firmly stuck jam jar, standing on tip toe, lifting or pushing a heavy object, rushing up a flight of stairs.
- This relies on the muscle's own fuel store – ATP (adenosine triphosphate) – and creatine phosphate.
- When these are used up people unaffected by McArdle's then convert glycogen stored in the muscle into glucose for energy.
- McArdle's people can't do this due to lacking an enzyme called myophosphorylase.
- A painful fixed contracture of the muscle can then develop and last for hours or days.
- It is best to avoid these activities.
- If you *have* to do them, count up to six seconds.
- If the task is not complete by then, stop or put it down.
- Take a break, let the muscles recover.
- ATP will substantially recover in a minute and fully recover in about three minutes. Then try again.



There is more about the six second rule on the AGSD-UK web site.

30 for 80

- Pain lags behind the damage we do.
- Learn to recognise the early signs.
- Slow or rest before you do damage.
- 30 seconds rest replenishes 80% of reservoir.

Physical activity.

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for 80

% of your ATP

Once you need to pause for a rest,
count to 30 seconds.

Pain in McArdle's lags behind the damage you do. Learn to recognise the early signs and slow or rest before you do damage. 30 seconds rest will replenish 80% of the energy reservoir in your muscles. Then you have some leeway.

Rest before risk

- When about to do something risky.
- Rest for 30 seconds or a minute.
- Ensures you have energy available to get through.

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Physical activity.



Rest before risk.



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When you are about to do something which might involve risk, like crossing the road or jumping in a swimming pool, rest for a minute before proceeding. This is to ensure that you have energy available to get you through.

The McArdle Mantra

- **Memorable phrases.**
- **Remind us of important things to help look after ourselves.**
- **Detail on each in “Living with McArdle disease”.**

The McArdle Mantra

This set of memorable phrases makes it easier for us to remember the things that are important to looking after ourselves. All the tips in this book are encompassed by one or more of these phrases.

- **Stroll before Second**
- **Pause before Pain**
- **Embrace our Embarrassment**
- **Rest before Risk**
- **Stop after Six**
- **Shorten our Static**
- **Avoid the Awkward**
- **Reduce our Repetitions**
- **Think before Tasks**
- **Temper our Tension**
- **Halve our Hurry**
- **Eat before Effort**
- **Aim for Activity**
- **Extend our Exercise**
- **Watch our Weight**
- **Mind our Medications**
- **Carry our Card**

For anyone who would like to know more, there are some explanatory notes for each phrase on the AGSD-UK web site.

