BIMDG
British Inherited Metabolic Disease Group

RHABDOMYOLYSIS IN YOUNG CHILDREN

- **Please read carefully.**
- **Meticulous treatment is important as there is a high risk of serious complications.**
- **If the instructions appear not to make sense or the problem is not addressed you must discuss your concerns with the consultant on call.**

1. Background

Rhabdomyolysis refers to the acute necrosis of skeletal muscle fibres. It can be caused by trauma or recreational drugs (particularly in adults), but it can also occur in several inborn errors, especially LPIN1 defects, long chain fatty acid oxidation disorders and McArdle disease. Most patients are healthy most of the time but episodes of rhabdomyolysis can be brought on by infections or exercise. Because episodes are often triggered by infections, early symptoms may include anorexia or vomiting; the other major early symptom is muscle aching or stiffness. Myoglobin, released from muscle, can enter the urine and cause dark brown or black discolouration. Apart from pain and weakness, **the main complications are hyperkalaemia (which can cause arrythmias) and acute renal failure** (due to heavy myoglobinuria). Treatment aims to prevent these by maintaining a good flow of urine and correcting any electrolyte disturbances that arise.

These guidelines apply to young children (less than about 10 years), particularly those with LPIN1 defects, who are at highest risk. The management in adolescents resembles that in adults, in whom rhabdomyolysis is commoner. There are published guidelines for McArdle disease, which seldom presents in young children (http://www.agsd.org.uk/tabid/2158/default.aspx).

2. Telephone call.

If you get a telephone call from a family with a child who is unwell, they should come to hospital without delay.

3. Initial Plan

Most patients who present to hospital will require admission. Only allow a young child home if it becomes clear that there is no rhabdomyolysis and there are no other problems. Some older patients have frequent episodes of exercise induced rhabdomyolysis and want to go home if the current episode is mild – but severity can be hard to judge in the early stages.

If there is any doubt at all, the child must be admitted.

4. In hospital

Put on cardiac monitor.

Check weight & vital signs, including blood pressure. Compare with a recent clinic weight if available.
Review the history & examine the child.

⇨ If the child is anuric, has heavy myoglobinuria, hyperkalaemia or hypocalcaemia, arrange for admission to ITU/High dependency. Consider this in all LPIN1 patients.

The following urgent tests should be done:

BLOOD	Urea & electrolytes Bone chemistry, Mg Liver function tests CK pH and blood gases Glucose, Lactate Full blood count
URINE	Dipstick (looking for a false positive for haemoglobin) Myoglobin (may not be available urgently)

ECG if there is hyperkalaemia or reason to suspect it (e.g. heavy myoglobinuria). Features of hyperkalaemia include peaked T waves, prolonged PR interval, widened QRS, loss of P wave and ultimately a sine wave appearance.

5. Management
The most serious complications are hyperkalaemia/hypocalcaemia (which can cause arrhythmias) and acute renal failure. Treatment aims to prevent these by maintaining a good flow of urine and correcting any electrolyte disturbances that arise.

a) Fluids
Obtain intravenous access and start fluids immediately if there is rhabdomyolysis, even if there is not currently heavy myoglobinuria.

If hypoglycaemic, give glucose 200 mg/kg at once (2 ml/kg of 10% glucose or 1ml/kg of 20% glucose) over a few minutes.

If shocked or obviously dehydrated (>7% weight loss), give 20 ml/kg 0.9% saline, with a further 10ml/kg subsequently if necessary.

Initial infusion rate:

- Calculate the deficit and maintenance & divide this by 24 hrs.
- Fluid depletion is common as it is sequestered in damaged muscle.
- 24 hr maintenance fluid = 100ml/kg for 1st 10kg, 50 ml/kg for next 10kg, then 20ml/kg

- Give as 10% glucose with 0.9% saline.
- If this is not immediately available, use 10% glucose with 0.45% saline, 5% glucose with 0.9% saline or 5% glucose with 0.45% saline.
- Potassium should not be added.

Subsequent adjustments:
If urine output is >0.5ml/kg/hr after 2-4 hrs, increase fluids to 3L/m^2 of 10% glucose with 0.9% saline.
See section (d) Oliguria if urine output is less than 0.5 ml/kg/hr (<1 ml/kg/hr in infants).

Sodium bicarbonate administration is not recommended routinely unless there is hyperkalaemia (see below) or acidosis. If the base excess is more negative than -10 mmol/L, consider giving 8.4% Sodium bicarbonate 1-2 ml/kg IV (diluted 1:10 with 10% glucose unless there is central access) over 2-3 hours. Note that this lowers the ionised calcium fraction and may cause tetany, convulsions, hypotension or arrhythmias if there is hypocalcaemia (which is common in rhabdomyolysis).

b) Monitoring:

- Patients should be on a cardiac monitor.
- Consider catheterisation. Monitor urine output, fluid balance, twice daily weight.
- Reassess after 1-2 hours if the initial potassium was >5mmol/L, if there is oliguria or heavy myoglobinuria or if patient has LPIN1 defect, otherwise after 4 hours (unless there is any clinical deterioration).
- Clinical assessment should include Glasgow coma scale and blood pressure.
- Blood tests: U&E, Calcium, pH and blood gases, CK
- Continue to monitor 2-8 hourly depending on the results; frequent monitoring is essential if there is hyperkalaemia.

c) Hyperkalaemia

- This is the most life-threatening complication and is caused primarily by release of potassium from damaged myocytes. If the patient is on PICU, follow the local guidance. If the child is not on PICU, discuss transfer.

- Ensure the patient is not receiving potassium (oral or IV) or potassium-sparing drugs. Check plasma calcium as there is often hypocalcaemia, which increases the risk of arrhythmias.

- Repeat ECG & start continuous cardiac monitoring.

- If hyperkalaemia is associated with heart block or ventricular arrhythmias, give 0.5ml/kg of 10% calcium gluconate (maximum 20ml) as a slow IV bolus (e.g. over 5 minutes) via a large peripheral vein or central venous line. This stabilises the myocardium but does not lower the potassium. The line should be flushed afterwards (e.g. with saline), particularly before sodium bicarbonate is given as otherwise calcium carbonate will precipitate!

- If the potassium is over 6 mmol/L, some or all of the following should be undertaken and they should be considered if potassium is over 5.5 mmol/L:

  i) Salbutamol 2.5mg nebulised (5mg if over 5 yrs) or
  Salbutamol 4 micrograms/kg IV diluted to a concentration of 50 micrograms/ml in 5% glucose or 0.9% saline and given as a slow bolus over 5 minutes.

  ii) 8.4% Sodium bicarbonate 1 ml/kg IV (diluted 1:5 or 1:10 with 10% glucose) over 30-60 minutes if the plasma bicarbonate concentration is less than 21 mmol/L. This lowers the ionised calcium fraction and may cause tetany, convulsions, hypotension or arrhythmias, particularly at higher pH, if there is hypocalcaemia (which is common in rhabdomyolysis).

  iii) Increase fluids to 5ml/kg/hr 10% glucose 0.9% saline = 0.5g/kg/hr glucose.
  If BM >10 mmol/L, add insulin, initially at 0.05 unit/kg/hr and adjusted according to blood glucose (use local sliding scale for diabetes).

  iv) Furosemide 1-2mg/kg IV if patient is not anuric.

  v) Calcium resonium 1g/kg oral/NG or PR.

  vi) Dialysis should be considered if there is refractory hyperkalaemia, acidosis or oliguria and fluid overload.

d) Oliguria

Ideally, one wants the urine output to be approximately 3 ml/kg/hr. Seek advice from paediatric nephrologists if the urine output is less than 0.5 ml/kg/hr (<1 ml/kg/hr in infants). Transfer to PICU may be needed for haemofiltration or dialysis but first they may suggest
i) a fluid challenge with 20ml/kg of 0.9% saline as a bolus.
ii) diuretics (1-2mg/kg furosemide or mannitol), particularly there is fluid overload.

e) Other treatment

- Treat any infection.
- Analgesia (paracetamol or opiates) may be needed for muscle pain.
- Seek orthopaedic advice if there is concern about compartment syndrome.
- Though hypocalcaemia is common, intravenous calcium is only given if there are symptoms (e.g. tetany, seizures) or severe hyperkalaemia (e.g. with arrhythmias).

6. Progress:

If myoglobinuria is improving, allow patient to drink & consider substituting this for some of the intravenous fluids. Ensure the carbohydrate intake remains adequate, particularly in fatty acid oxidation disorders.

7. Going Home: Only allow the child home if you and the family are entirely happy and you have discussed the problems with the consultant on call. The family must have a clear management plan and be prepared to return if the child deteriorates.

8. HELP: If there are any questions or concerns, contact the regional metabolic centre and/or paediatric nephrologist.

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