



Hyperkalaemic periodic paralysis: information for families

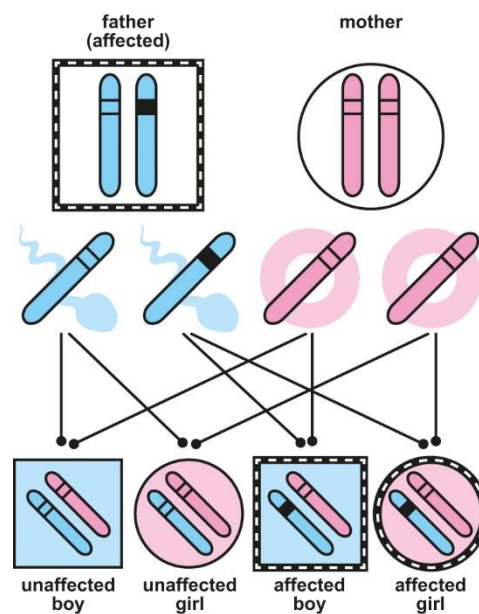
Hyperkalaemic periodic paralysis is a condition that causes attacks of muscle weakness that come and go (episodic) in response to high levels of potassium in the blood. Attacks may be focal – affecting one limb only - or can affect the entire body. Potassium is a mineral electrolyte that is important in lots of body functions, such as heart rate, muscle function and nerve impulses. This information sheet from Great Ormond Street Hospital (GOSH) explains about hyperkalaemic periodic paralysis, what causes it and how it can be treated.

What causes hyperkalaemic periodic paralysis?

Every cell in the body has a thin membrane wall that lets a mixture of water, sodium and chloride in and out through ‘channels’ that act a bit like gates. How these channels work is controlled by our genes.

Hyperkalaemic periodic paralysis is caused by a fault in a muscle channel gene – two different genes can both cause this condition. These gene faults affect the amount of potassium flowing in and out of the muscle cells. As potassium is needed to make the muscles contract, if the amount of potassium in the muscle is not right, the muscles become weak.

The faulty gene can be passed on from parent to child. If one parent has the faulty gene, there is a 50 per cent (1 in 2) chance that their child will also have it. This is called autosomal dominant inheritance. This chance is the same for every pregnancy.



In many cases, however, the faulty gene can occur sporadically (out of the blue) in a child so is not inherited from either parent.

Who is affected by hyperkalaemic periodic paralysis?

Attacks can start at any age, but often start during primary school – that is, between 5 and 10 years of age.

What are the symptoms of hyperkalaemic periodic paralysis?

The main symptom is weakness affecting one limb (arm or leg) or the entire body. This weakness can be triggered in a number of ways including:

- Cold
- Fasting (not eating regularly)
- Resting after exercise
- Eating foods that are naturally high in potassium

Attacks tend to last minutes although sometimes can be hours and usually happen during the day.

Some people with hyperkalaemic periodic paralysis also have myotonia – stiff muscles that cannot relax after activity. The effect of myotonia varies – some children can have more frequent weakness attacks with little or no stiffness but others may have more stiffness but fewer or less severe episodes of weakness.

How is hyperkalaemic periodic paralysis diagnosed?

The doctor will take a careful description of what symptoms occur and when, along with a physical examination. A gene test from a blood sample is needed to make a definite diagnosis.

How is hyperkalaemic periodic paralysis treated?

The aim of treatment is reduce the frequency and severity of attacks of weakness. This is done with a combination of medication alongside dietary and activity changes.

Medications

The main medications prescribed are called diuretics - these get rid of excess potassium in the body by making your child pass more urine. The most common tablet used is called acetazolamide.

At GOSH, this is usually supplied under the brand name Diamox® in the form of capsules.

The biggest impact of taking acetazolamide is the increase in trips to the toilet. If this is problematic, please discuss this with the team. The Clinical Nurse Specialist will follow up with you how your child is responding to treatment with acetazolamide.

All medicines have the potential to cause side effects. There is a chance that your child could develop mild tingling (pins and needles) in their fingers and toes, but this usually improves after a few days. If it continues, the GOSH team may suggest reducing the dose that your child is taking.

Rarely, acetazolamide can cause kidney stones to form. We recommend that your child drinks plenty of fluids each day and also has an ultrasound scan of their kidneys ever year to check for stones. If your child develops kidney stones, we have a specialist team at GOSH who can treat them.

Diet changes

We usually suggest starting a food diary to keep track of what your child eats and when. It is then easier to identify simple swaps that reduce the amount of potassium they are eating. The Clinical Nurse Specialist will give you more detailed advice about dietary changes and may refer you to our Dietitians if needed.

What is the outlook for children and young people with hyperkalaemic periodic paralysis?

Episodes of weakness can usually be reduced in severity and frequency by making simple lifestyle changes and taking medication. The vast majority of children and young people grow up living a near-normal lifestyle.

Further information and support

Please contact the Clinical Nurse Specialist in the Dubowitz Neuromuscular Centre at GOSH. Call 020 7405 9200 ext 1195 or email nmchan@gosh.nhs.uk.

Muscular Dystrophy UK is the main organisation offering support and advice to anyone affected by a neuromuscular disorder. Call their helpline on 0800 652 6352 or visit their website at www.musculardystrophyuk.org