

PERIODIC PARALYSIS INTERNATIONAL

Do I Have Periodic Paralysis?

Pursuing a Diagnosis

A Guide for Patients



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The information in this publication is based on current medical knowledge but should never — at any time — be substituted for the advice and care of a qualified medical consultant. For medical advice seek the services of a physician.

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Visit the Periodic Paralysis International website at:

<http://hkpp.org/>

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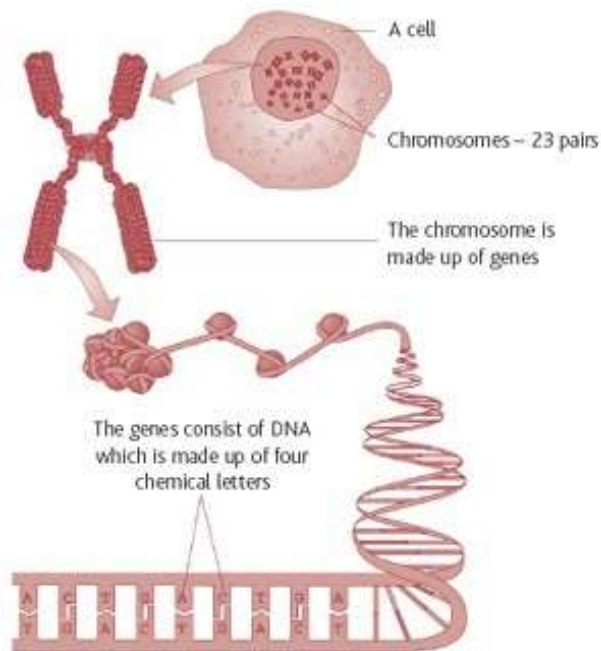
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Do I Have Periodic Paralysis?

We are asked that question frequently by people with a wide variety of health challenges. We cannot diagnose patients, but we can give you information about periodic paralysis which you can study, compare with your own symptoms, and take to your physician.

What Is Periodic Paralysis?



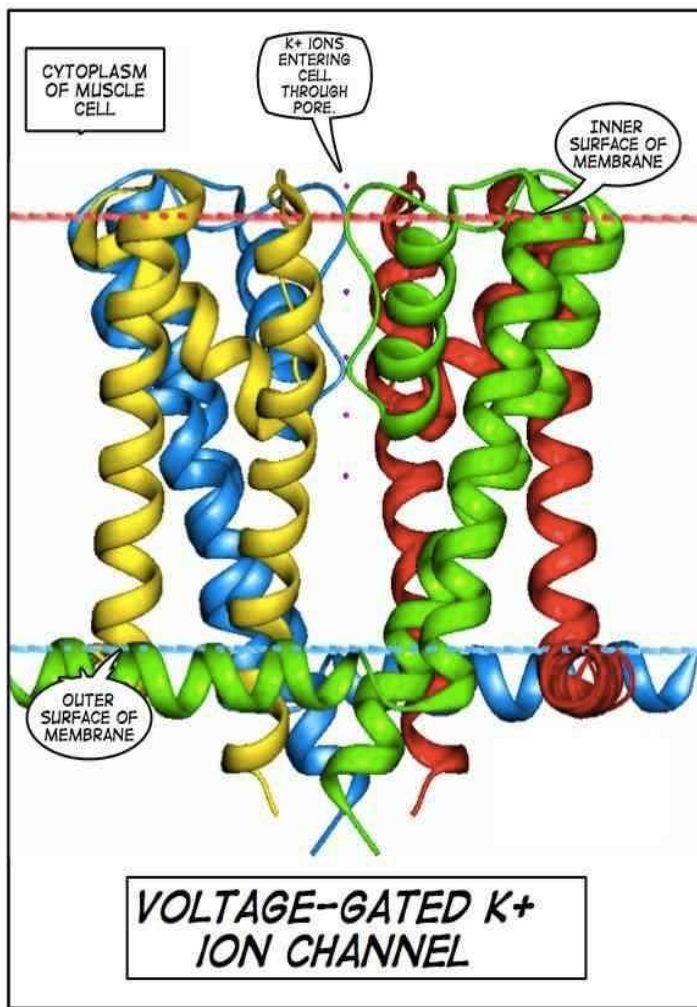
The Periodic Paralysis are a group of muscle disorders which are caused by genetic mutations which affect some of the ion channels located in the skeletal muscles. Skeletal muscles are sometimes called voluntary muscles because they are muscles we have control over. They are the muscles which enable us to move from place to place, throw a ball, and pick up a glass of water.

Genes are made of DNA. They contain information which is passed from parent to child. Genes contain instructions which determine a child's eye and hair color, as well as the structure and function of all the body's organs and tissues.

A genetic mutation is a mistake in these instructions. In periodic paralysis the mutation (or genetic mistake) causes a change in the structure or function of certain ion channels in skeletal muscle. Periodic Paralysis is found in all races and in both sexes. There are several different types of periodic paralysis, but all of the types have some features in common.¹

- 1) They cause attacks of episodic muscle weakness and/or paralysis. The patient's muscles become flaccid and floppy, like a rag-doll. The patient's strength initially returns to its normal level between attacks, but as the patient ages lingering weakness may develop after and between attacks.
- 2) Almost without exception attacks begin before the age of 25 but *attacks may go unrecognized until a later age, explained away as laziness or attention-seeking behaviour, a medication reaction, or the result of another illness*. If attacks of weakness haven't begun by age 25 they are almost never due to periodic paralysis. Two exceptions to this are patients who develop exercise intolerance in the legs, which progresses to a fixed and unvarying weakness of the legs first noticeable in the 40s but affecting function by the age of 60s or 70. The second exception is the patient with Andersen-Tawil Syndrome, which may surface at any age.
- 3) The level of potassium in the patient's blood fluctuates shortly before an attack. It may be lower or higher during attacks, but potassium level is in the normal range between attacks.
- 4) The attack may affect only one muscle or group of muscles, or it may affect the entire body.
- 5) When weakness is on-going or affects only one group of muscles the change in the potassium level may be too small or temporary to measure.
- 6) Speech, swallowing and breathing are not usually affected, but may be in severe attacks.
- 7) Rest after exercise may provoke an attack, but mild attacks may sometimes be "walked off", that is, if the patient can keep moving a mild attack may simply go away.
- 8) Becoming chilled can provoke attacks.
- 9) Deep tendon reflexes diminish and may disappear altogether during attacks.

What Causes the Symptoms of Periodic Paralysis?



Over 50 different genetic mutations have been identified in association with periodic paralysis. Each of these mutations affect the structure or function of either a sodium, calcium or potassium ion channel. These mutated ion channels are located in the skeletal muscle membrane. The muscle membrane is an active, living tissue which wraps around the entire surface of each muscle cell. This membrane has an outer surface and an inner surface.

Ion channels are formed of groups of flexible coiled tubes. These create and surround an opening (or pore) which extends from the outside surface of the membrane to its inside surface. The coils open and close the pore by changing shape. When the sensor on the ion channel receives the proper signal the coiled tubes react. They change the shape of the opening, allowing ions of potassium, calcium or sodium to pass through. The channel remains open only for a fraction of a second, ions flow through, the tube closes and ion flow stops. The ion channel then goes into a "resting" phase, relaxed but ready to respond to a new signal.

The symptoms of PP are caused by fluctuations of serum K^+ which primarily affect muscle tone, but a potassium imbalance in either direction also affects other functions in the body, as potassium and sodium are the two ions that "fire up" the electrical grid that our cells operate on.

The muscle cell works best when the membrane is maintained at a certain level of

electrical resistance. This resistance is "set" or maintained by the ratio of potassium and salt on either side of the muscle membrane, a ratio controlled by the ion channels. When an ion channel is defective, as in PP, it affects the resistance of the muscle fibre. The resistance of the membrane goes up, making it more difficult to overcome. As membrane resistance rises the muscle becomes less responsive to stimulation, thus weaker. When resistance gets so high that the muscle quits responding at all the result is paralysis.



You might visualize this increased membrane resistance as a dimmer switch controlling a light with an infinite number of settings. It will handle a 100 watt bulb, but



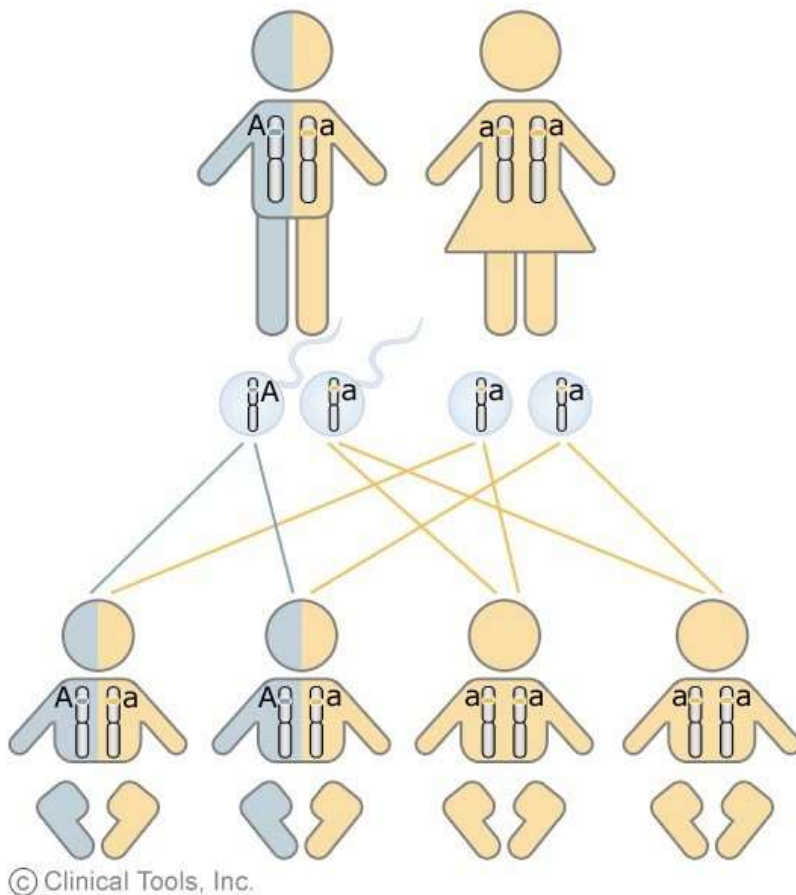
let's say we only had a 50 watt bulb in the drawer, so we stuck that in the socket. When the mark is straight up potassium is at a normal level. When you turn the dial down (dim the lights) you are actually increasing the resistance to the flow of electrons in the wire. That's just what happens when the level of potassium drops in the blood serum. Cell membranes **all over** the body become more resistant to stimulation. The muscles get weaker, because nerve stimulation can't overcome the increased resistance.

If you turn the dial UP the light gets brighter because electrons flow more easily, and resistance to stimulation is reduced. It's the same in the body. Too **much** potassium in the

blood causes the ion gates to become too sensitive to stimulation. They respond too vigorously, or too often, and they may fire repeatedly until they become stuck in a half-closed position, allowing sodium ions to pour through the membrane into the muscle tissue. This would be like turning the dimmer dial too high, so high that the light bulb pops. In hyperkalemia this is the point where weakness becomes paralysis. The muscle packs up and says, "No more movement until you get this wiring problem worked out!"

Fluctuations in potassium don't just affect the skeletal muscle, they affect every cell and organ, because we literally run off a power grid created by the exchange of sodium and potassium ions in our cells. In PP it's not the absolute value of potassium in the blood that determines a person's weakness, it's the ratio of potassium and sodium inside and outside the muscle cell. The weakness hasn't anything to do with nerves or nerve signals. The nerve signals arrive and are received, it's the response (or the lack thereof) which causes the weakness.

How Do You Get Periodic Paralysis?



About two-thirds of cases of periodic paralysis are inherited, though a family history may not always be obvious. All the periodic paralyses are inherited in an autosomal dominant manner. That means that only one parent need carry the affected gene, and that parent may not have symptoms of periodic paralysis. About half of the females and a few males who carry a periodic paralysis mutation have no symptoms, or have such mild symptoms that they are never recognized as periodic paralysis. These people can still pass the gene on to their children.

Periodic paralysis doesn't "dilute" over the generations. People who inherit the mutation may have symptoms as severe as an affected ancestor several generations earlier. A parent with few symptoms may have a child who has a severe case, or a person with a severe case may have children with very mild symptoms, or no symptoms at all.

Children in the same family may have different levels of symptoms. Some may

not be affected, some may be severely affected, some may be mildly affected. Each child of an affected parent has a 50% chance of inheriting the gene, but this doesn't mean that half of an affected person's children will inherit PP. Each time pregnancy occurs the child has the same 50-50 chance of inheriting as every other child of an affected parent.

In about one third of all cases of PP the mutation occurs at conception and is not inherited. These are called *sporadic** cases. The person who has a sporadic case has the same chance of passing the mutation on to their children as does the person who inherited the condition.

* *sporadic*: occurring at irregular intervals

What Determines Which Kind of Periodic Paralysis a Person Has?

The mutation determines the type of periodic paralysis. Over 50 different mutations have been identified in sodium (Na⁺) and calcium (Ca⁺) channels. DNA testing is now available for many of the mutations which cause periodic paralysis, but cannot be relied on to diagnose every case. There has been a slow but steady trickle of newly identified mutations since the first one was identified in 1994. Yet many clinically diagnosed patients with clear family histories of periodic paralysis still test negative on DNA tests. It's clear there are more mutations waiting to be identified.

What Are the Different Types of Periodic Paralysis?

The periodic paralyses are classified into types by how the patient reacts to potassium. The normal level for serum potassium is 3.5 – 5.0. Even a healthy person will get weak when their potassium goes too far outside of the normal range, so it is necessary to do more than measure potassium levels to establish a diagnosis of periodic paralysis.

It takes a very little shift of potassium in the body to move the serum potassium up or down. If a patient weighs 50 kg (110 pounds), there are 20 litres (50 pints) of fluid inside the cells in their body. The volume of fluid outside the cells, in the blood and other fluids, is 10 litres (25 pints).

There's an enormous amount of potassium inside a cell. The potassium level inside the cell is 150 mmol/l - compared to a serum level of 3.5 – 5.0 mmol/l. Only 20 mmol of potassium needs to shift from the blood into muscle cells to cause the serum potassium level to fall 2 mmol/l. A fall of 2 mmol/l in the serum potassium can paralyze even a normal person. The opposite is also true. Only a relatively small amount of potassium needs to leak from the cells into the serum to raise the serum potassium level to a point which causes symptoms.²

Hypokalemic Periodic Paralysis

In the most common form of periodic paralysis patients have attacks of flaccid (rag-doll) paralysis or weakness which go away when treated with potassium. This form is called Hypokalemic Periodic Paralysis (HypoKPP). HypoKPP is associated with mutations on the Calcium (Ca⁺) and Sodium (Na⁺) channels. During severe attacks the patient may become totally unable to move and may appear unconscious. Despite being unable to communicate, the patient is awake and completely aware of their surroundings during attacks.

Patients with HypoKPP experience a drop in potassium levels during attacks. The potassium may fall a lot or only a little. It may even stay within normal limits, but it falls from the patient's normal level. This makes it essential to know a person's baseline potassium level. This is obtained by testing the potassium level several times when the patient is feeling well and is not experiencing any weakness. This level is then called their baseline. In some patients with HypoKPP it takes only a fall of 0.5 mmol/l to cause severe weakness and paralysis.

Weakness in HypoKPP most commonly affects the muscles of the arms and legs but may affect the trunk. In a few patients the muscles involved in breathing and swallowing can be affected during severe episodes. An irregular or weak heartbeat can occur during episodes. Most patients have normal muscle strength between attacks, but muscle tissue is damaged by attacks. This damage may eventually cause permanent weakness in some patients by their 50s or 60s.

Patients with HypoKPP typically begin showing symptoms in childhood or adolescence, often as they enter puberty. About 65% develop symptoms before the age of 16, but symptoms may first be recognized as HypoKPP at any age. Attacks may be infrequent at first, but eventually may occur daily. Some patients have attacks only during sleep and may be unaware of being paralyzed.

2 Alazami M, Lin SH, Cheng CJ, Davids MR, Halperin ML. Unusual causes of hypokalaemia and paralysis. QJM. 2006 Mar;99(3):181-92. PMID: 16469765

Attacks may last anywhere from an hour or two to two or three days. They may vary in intensity from episode to episode, with mild localized weakness in a hand, foot, jaw or limbs one time and generalized paralysis the next. Episodes may be accompanied by extreme muscle tenderness and aching or be entirely painless. Attacks are

triggered by high carbohydrate foods, large meals, a high intake of salt, sleep, stress and medications which lower the serum potassium.

Licorice root (*Glycyrrhiza glabra*) is known to cause hypokalemia. Some HypoKPP patients experience weakness after consuming St. John's Wort, American Ginseng, and Ginkgo Biloba, all of which increase insulin sensitivity.

Some patients with HypoKPP also have what are called abortive attacks – so called because they abort (stop) before progressing into paralysis. Abortive attacks are periods of fluctuating weakness which lasts for extended periods. They have even been mistaken for permanent weakness, but unlike permanent weakness abortive attacks respond to therapy. Abortive attacks become more common in patients as they enter their 40s and in some patients abortive episodes replace paralytic attacks as they grow older. Patients often find abortive episodes are more disabling than the brief attacks of paralysis were.

Some patients have only exercise intolerance and weakness of leg muscles which increases with age. They may have trouble walking up hills or climbing stairs and may become quite fatigued by the normal activities of daily living and as a result may appear to be “lazy” to others. Their symptoms are rarely diagnosed as HypoKPP except in families already known to be affected.

Treatment for HypoKPP focuses on keeping the potassium level from falling and reducing the shift of potassium from the blood to the muscle. The most effective treatment is often reached through trial and error and a bit of experimentation with different medications and dosages.

Hypokalemic Thyrotoxic Periodic Paralysis

Another form of HypoKPP is associated with an overactive thyroid gland. This is called Thyrotoxic HypoKPP (TPP). As opposed to inherited HypoKPP, TPP often first occurs between 20-40 years of age, when thyroid disease becomes prevalent. TPP has three characteristic symptoms which all occur at the same time:

- Elevated thyroid hormone (which may be normal in the early stages of the illness)
- Low levels of potassium in the blood (hypokalemia)
- Muscle weakness or paralysis

Potassium levels during attacks of TPP are usually quite low, often as low as 2.0. TPP is far more common in Asian males but can be found in people of all ethnicities and genders. When physicians say periodic paralysis is only found in Asian males TPP is the form of periodic paralysis they are thinking of. Except for the fact that an overactive thyroid triggers TPP, its symptoms are identical to HypoKPP, although patients with TPP rarely have a family history of PP. There are clues when HypoKPP is caused by thyrotoxicosis. Unintended weight loss, heavy sweating and heat intolerance are all associated with an overactive thyroid.

Grave's Disease is the most common cause of hyperthyroidism in TPP patients, but any cause of thyrotoxicosis (including taking too much thyroid hormone) can trigger attacks in people who are susceptible. Recently mutations for TPP have been found in KCNJ18, in a gene coding for Kir2.63. TPP is the only type of periodic paralysis for which there is a cure. Once the overactive thyroid has been successfully treated the symptoms of HypoKPP go away.

3 Ryan DP, Dias da Silva MR., Soong TW. et al. Mutations in potassium channel Kir2.6 cause susceptibility to thyrotoxic hypokalemic periodic paralysis. Cell 140: 88-98, 2010. PubMed: 20074522

Andersen Syndrome

Andersen Syndrome (Also known in the US as Andersen-Tawil Syndrome) is yet another type of periodic paralysis. In this type, the potassium shifts during attacks are inconsistent. Some patients with AS have low potassium during attacks, some have high potassium during attacks. Some have no change in their potassium levels during attacks and some have low potassium during some attacks and high potassium during others.

Patients may also have generalized weakness between attacks.

In addition AS patients experience irregular heart rhythms including a prolonged QT interval, especially during activity. Heart rhythm irregularities are often a distressing feature in AS. Some AS patients faint during exercise or during periods of intense emotion. Almost all patients need heart medication and some require the surgical implantation of a pacemaker or defibrillator.



Many, but not all, AS patients are short and slender, and have unusual facial and hand characteristics, such as curved little fingers, webbed or fused 2nd and 3rd toes, widely spaced eyes, low-set ears, a broad forehead and small jaw. These physical signs may be absent or very subtle, or they may exist in other family members who do not have paralytic attacks. Gene mutations which encode for AS are on the potassium channel.

AS is treated from a variety of approaches. The goal is to stabilize any cardiac arrhythmias and treat the fluctuation in potassium. AS can be difficult to treat, as what helps the weakness may worsen the cardiac problems and vice versa.

Hyperkalemic Periodic Paralysis

Patients with Hyperkalemic Periodic Paralysis (HyperKPP) react with flaccid weakness and/or paralysis when they eat a diet high in potassium or after they exercise. The potassium level of these patients is often just a little on the higher side of normal between episodes, but it may not go above normal at any time. Serum sodium levels may fall as potassium rises. This results from sodium entry into the muscle. Water also moves into the cell causing further hyperkalemia.

Attacks of HyperKPP begin during the first ten years of a person's life. Attacks last from 10 minutes to one hour, rarely up to two days. Some patients experience only a few attacks in their lifetime, others have one or more attacks every day at the same time.

HyperKPP symptoms are caused by malfunctions of the sodium ion channels. These malfunctions cause the ion channel to stay slightly open when it should be closed, allowing ions of sodium to leak through the channel like water through a drippy tap. The sodium eventually causes an imbalance in the electrical potential of the cell membrane. Weakness and paralysis can follow. Attacks may come on very rapidly. Falling may occur in sudden attacks. Hunger or rest after activity are common triggers. Changes in the daily level of activity and periods of inactivity (sleep, sitting through a movie, car or plane trip, especially in cool temperatures) triggers weakness in many patients. Many patients learn to fidget or move around to abort episodes. Some patients chew gum constantly to stave off episodes. Patients may describe muscle stiffness (myotonia). The stiffness in HyperKPP can often be “worked” or “walked” off and is dismissed many times as rheumatism or fibromyalgia.

Sometimes patients with HyperKPP are misdiagnosed as having HypoKPP because they go to the Emergency Room during an attack, and when blood is drawn a slightly lower than normal potassium level is discovered. This often happens as the attack is ending, as the potassium level normalizes. The kidneys work very hard to remove the excess potassium, and may overcompensate for a short time, leading to a temporary mild hypokalemia.

A high carbohydrate food (candy bar) or beverage (sugar cola, sweet tea, GatorAde) taken at the first sign of weakness may abort or relieve episodes of HyperKPP. HyperKPP is treated with medications which cause the patient to excrete potassium, and with medications which alter the function of the mutated ion channels. (See also Myotonia and Paramyotonia)

Normokalemic Periodic Paralysis

The term “Normokalemic Periodic Paralysis” is rarely used any longer. Those patients who were originally diagnosed as “Normokalemic” have been genetically identified as carrying the same kind of sodium channel mutations which cause Hyperkalemic PP.

Myotonia

Myotonia is not a type of periodic paralysis, but a symptom which often accompanies it. Myotonia is the inability to relax voluntary muscle after effort. The muscle fibers contract and, instead of relaxing immediately, they relax very slowly. This is experienced as muscle stiffness or rigidity. Despite its definition, myotonia often develops while the affected person is inactive. It is more likely to occur in cool temperatures. Sitting in an air-conditioned room for an hour or two can lead to a feeling of stiffness when the person tries to move. This stiffness can be “worked off” the muscles loosen up when the person moves around for a few minutes.

Patients with Hyperkalemic PP usually have myotonia. Patients with Hypokalemic PP may have myotonia in their eyelids, so that they have difficulty opening their eyes or keeping them open. Myotonia in the eyelids makes the lids feel tight and uncomfortable. It usually occurs only on awakening, or when using the eyes (reading) for extended periods.

Paramyotonia and Paramyotonia Syndromes

The word “paramyotonia” is made up of two words: paradoxical, which means seemingly absurd or selfcontradictory, and myotonia, the inability to relax voluntary muscle. Paramyotonia is defined as myotonia that appears during exercise and becomes much more severe upon exposure to cold. Paramyotonia may be severe enough to interfere with breathing and make movement difficult. Rest relieves paramyotonia, but attacks triggered by vigorous activity may take weeks to resolve. Paramyotonia can interfere with facial expression, especially in cool or cold weather. There are a number of different Paramyotonia Syndromes:

Pure Paramyotonia: Paramyotonia ± Episodic Weakness With Exercise:

Muscle stiffness which worsens with exercise. More severe attacks produce weakness. Weakness or stiffness may occur alone. Symptoms are worst in face, neck & upper extremities. Onset during infancy is common, onset is always by adolescence. May be associated with HyperKPP. Some patients are worse after potassium intake.

Paramyotonia Congenita Without Paralysis:

Spasms of muscle stiffness: Mostly in hands and face. Symptoms worsen with exertion, worst trigger is cold but there is no weakness or paralysis.

Paramyotonia Congenita (PMC) (aka PMC Von Eulenburg):

Paramyotonia is increased by exposure to cold; potassium level may be increased, decreased or normal during attacks. Weakness in PMC is triggered by a drop in potassium level but attacks of flaccid weakness/paralysis occur which are not caused by a change in potassium level. PMC is not progressive. There is no muscle wasting or overly large muscles though patients who have both PMC and HyperKPP may have muscle wasting or large muscles associated with HyperKPP.

PMC is treated with medications (diuretics) which make the patient excrete potassium through the urine, and with medications which alter sodium and potassium ion channel function. Paradoxically, some patients with PMC respond very well when a potassium supplement is added to their medication regime.

The Potassium Aggravated Myotonias (PAMs):

In the PAMs attacks of myotonia are triggered by potassium intake. None of the PAMs cause weakness. The PAMs are treated with medications which make the patient excrete potassium, and with medications which alter sodium channel function.

Myotonia fluctuans; Mild myotonia that varies in severity from day to day. There is no weakness or cold sensitivity. Stiffness develops during rest in the half-hour period after exercise; lasts for about an hour, worsens with potassium intake, and may be severe enough to interfere with breathing.

Myotonia permanens; Severe continuous myotonia which may interfere with movement and breathing. The person appears very muscular as the constant activity causes the muscles to enlarge but does not increase their strength.

Acetazolamide responsive myotonia; The person appears very muscular. There can be significant muscle pain. Symptoms improve with acetazolamide treatment.

Summary

In summary, all patients with periodic paralysis, with the exception of those with Myotonia Fluctuans, experience weakness and/or paralysis. In those with a Paramyotonia Syndrome the most prominent feature of the disorder is muscle stiffness, but in all other forms muscle weakness and/or paralysis are the dominant symptom. As a rule, patients with periodic paralysis tend to have attacks triggered by:

1. Unusual exercise. For example: a weekend tennis match, a long walk, or **any** activity that is 10-15% more than that person is accustomed to on a daily basis.
2. Or conversely, inactivity. For example: sitting through a long movie, a long car ride, sleeping longer than usual.
3. Intake of food which is high in carbohydrates for those forms associated with hypokalemia; or food which is high in potassium for forms associated with hyperkalemia.
4. Becoming chilled.
5. Stress, fear or fright.
6. Medications which cause a fluctuation in potassium levels, block ion channels (many blood pressure meds do this), relax muscles, the epinephrine added to local anesthetics (i.e. Dental freezing).
7. Alcohol intake.
8. General anesthetics in the halogenated ether group (isoflurane, ethrane, sevoflurane, and desflurane), as well as halothane and succinylcholine. In the PPs these can cause uncontrolled release of calcium in the muscle cell, leading to intense muscle contraction. This is called Malignant Hyperthermia (nothing to do with cancer) and can be life-threatening.

Do My Symptoms Point to Periodic Paralysis?

It is very important that you have carefully read the material to this point before attempting to answer these questions!

Symptom	Yes	No
I experience episodes of weakness/paralysis which have a recognizable beginning and end?		
I experience episodes of weakness/paralysis which develop while I rest or sleep, especially after activity or exercise?		
I experience muscle stiffness or rigidity which develops while I rest or sleep, especially after activity?		
I am fully conscious during paralytic attacks?		
I experience muscle stiffness or rigidity which develops while I am active, which is relieved by rest?		
I experience a fast or irregular heart beat during weakness, paralysis or activity?		
I have fainted during exercise or stress?		
Becoming chilled makes my symptoms worse?		
Becoming overheated makes my symptoms worse?		
Becoming too hungry triggers weakness?		
Eating sweets, starchy foods or a large meal makes my symptoms worse?		
Eating sweets makes my symptoms better?		
Becoming fatigued triggers weakness?		
During attacks my serum (blood level) of potassium is low?		
During attacks my serum (blood level) of potassium is high?		
Between attacks my serum potassium is in the normal range?		
My symptoms began before I was 25 years old?		
I have relatives (or ancestors) who are known to have (had) episodic paralysis?		

Notes:

No Patient Will Have Every Symptom

Some of the questions in the foregoing table apply to only one or two types of PP. Some people with PP have no family history, and some types of PP do not cause muscle stiffness. But almost all patients develop symptoms by age 25, and in all forms the patient is alert and aware during paralysis. A patient may lose consciousness due to irregular heart beat, or due to the inability to breathe adequately, but this happens very rarely.

If you are over 25 years old and never had any episodes of weakness or paralysis when you were younger, even ones explained away by low blood sugar, epilepsy, migraine equivalent, chronic fatigue or similar illnesses, then you probably don't have periodic paralysis caused by an ion channelopathy. You may well have an illness which causes potassium fluctuations, weakness and episodes of paralysis, but you probably don't have ion channelopathy PP. However, having said that, there are patients who have awakened weak or paralyzed so frequently since infancy that they believe it to be normal.

Documenting Your Triggers

How do I find out if I have ion channelopathy periodic paralysis? One of the first and most important things you need to do is to identify the factors that trigger your episodes of weakness, paralysis or stiffness. You start by writing down all the food you eat, everything you drink, all your activities and any emotional events of note, plus your symptoms, on an hourly basis.

You will not remember accurately enough what you ate, or how you felt, if you wait until the end of the day, or the end of the week, to log it. So, on your phone or in a small notebook that you can keep with you all day, record your data as you go. Do not use abbreviations. If you need to look back at your log from a year from now, you may not remember that you used to write RLP for “right leg paralyzed.” Also, your doctor may ask to see your log and he will understand it faster if you spell out the details.

How to Keep a Triggers Diary

Get an app which allows you to record data in a chart or set up a simple chart in your notebook. For each hour of the day reserve several lines on which to record everything you've had to eat or drink in that hour - including juice, pop and alcohol. Record the amount, and the time you ate/drank it. Also leave space to record your activities during that hour. (Some examples might be: I walked three blocks. I mowed the lawn. I laid on the sofa and watched TV.) For the same hour, on a scale of 1-10 record your general strength level. One means you couldn't get out bed if it were on fire, 10 means your strength is as good as it gets for you.

Be objective. Think *carefully* about the intensity of your symptoms. This is one situation which calls for absolute clinical objectivity, observing your body and your symptoms as if they were a brick you found lying on the sidewalk. Don't allow your emotions to describe your symptoms, make your logical brain do it. If you are experiencing localized weakness record where (arms, legs, trunk, hands etc.) and grade it separately 1-10, with one being a floppy paralyzed limb and 10 being normal strength. If you had generalized (overall) weakness or fatigue within the last hour record grade it on a scale of 1 – 10. If you have pain say where and grade it on a scale of 1-10. One is a mosquito bite and 10 is akin to having your leg amputated with a rusty crosscut saw. Record any other significant symptoms like nausea, chest pain or heart palpitations, a feeling of pins and needles, shortness of breath, etc. These symptoms may of course last longer than an hour, if so, indicate when they began and when they stopped. Also record at what times you void urine (as “U”) and have bowel movements (as “BM”).

In some forms of periodic paralysis, muscle stiffness or rigidity is a particularly significant symptom. This stiffness may cause a slight feeling of hesitation when moving, or a problem “letting go” of grasped objects. Stiffness may come and go or be a more or less constant companion, but watch for increased stiffness following exercise or the ingestion of certain foods. If you have muscle stiffness, rate it and notice if it improves or grows worse as you “warm up” or use your muscles. This may change from episode to episode with the HyperKPP/PMC combination (explained below), so close observation is necessary.

Example of what a diary entry might look like

Monday 6th Jan 4:00 PM

Generalized strength level: (7) able to walk around house with no problem

Localized weakness: (5) hands (unable to use manual can opener, had to get son to open cat's food.)

Other sensations: generalized muscle aching (3) pins and needles in fingers (3)

You may have to keep this record every day for two-four weeks. It takes time and it's *really* annoying, but it is vital for several reasons. One, it can establish whether you actually have a pattern of episodic weakness and two, if you do, it will reveal your triggers and thus allow you and your doctor to establish if your weakness responds to potassium or is caused by potassium. This information will be very valuable when you start to design an effective management program.

You'd be surprised at how many patients have come to us over the last 20 years, having been diagnosed with periodic paralysis, not knowing what form of periodic paralysis they have, or diagnosed with the wrong form. Many patients are diagnosed in the Emergency Room, based on their paralysis and a blood draw, and some physicians don't realize there is more than one form. However, the misdiagnosis happens most frequently because many blood draws are done toward the end of an attack and may not be reliable. Not that the patient doesn't have periodic paralysis, but the physician misreads the test result.

In HypoKPP as the triggers push the body toward an attack, potassium in large quantities moves from the blood serum into muscle cells. Potassium in the blood falls lower and lower and potassium in the muscle cells gets higher. All during the attack potassium keeps being moved from the blood into the muscle. All this potassium in the muscles upsets the sodium/potassium balance that they need to respond to the stimulation from nerves, causing weakness and even paralysis.

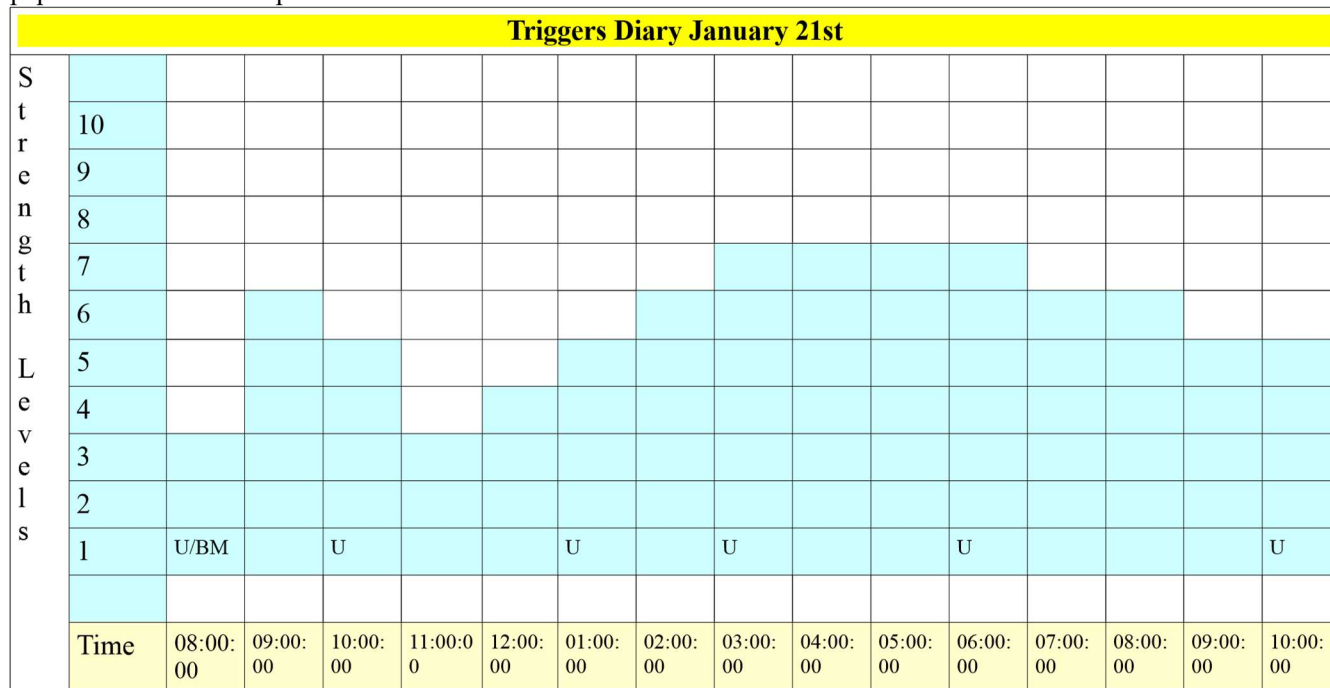
As the attack begins to resolve all the potassium pours out of the muscle in a flood, and for a brief time, the blood potassium level may go high enough to register as *hyperkalemic*! The kidneys get busy and within a half-hour or so, in most patients the potassium level will go down to normal levels. But if the blood draw happens in that brief time when the muscles are flushing potassium back into the blood, and the patient is diagnosed based on that one blood draw, they may be misdiagnosed as Hyperkalemic PP.

In Hyperkalemic patients the opposite happens. During attacks muscles push potassium into the blood, thus they have high potassium readings. However, as the attack resolves their potassium level falls as potassium moves back into the muscle a little too enthusiastically. If they have a blood draw at this time they can look dangerously *hypokalemic*. They also may be misdiagnosed as HypoKPP.

So, it is very important not only to know what form of periodic paralysis you have, but to use the information in your diary to learn and understand your triggers so you can design a management plan which will preserve your muscles and your health in the best of health.

What Do I Do With the Diary Information?

At the end of each week sit down with a piece of graph paper and make a chart for each day. Begin at the bottom line and number each line on the graph from 1 to 10 as it goes up the side of the paper. Across the bottom mark down the hours of the day, beginning each morning when you get up and ending when you took the last reading of the day. Transfer the information about your hourly strength levels from your notebook to the graph paper. Then look at the pattern.



		Woke up weak	2 eggs toast milk took bus to work	At work 2 donuts at 10:30	Weak leg pain level 4	Tuna salad milk pear	Walk of 10 blocks leg pain level 3	work	work	work	Bus home Groc shop	Beef patty bake p'tato green beans cake	TV	TV	TV Cake	Bath bed
--	--	--------------	---------------------------------------	------------------------------	--------------------------	-------------------------	---------------------------------------	------	------	------	-----------------------	--	----	----	------------	----------

Look at your charts very carefully for the entire time period. Does your strength fluctuate from hour to hour? Does your strength dip from say - seven - down to three and then in a few hours it's back up to six or seven again? Or maybe it falls gradually over a period of days? Monday you had a strength level of seven – nine all day and on Tuesday you woke up at a level three and stayed there until noon? Look at the dips in strength and look at what you ate and your activities in the 24 hour period before the dip. Some people may have to go back as far as 72 hours to find the trigger. It may take looking at a week or more's worth of activities and food to find a pattern. Women, especially those with HypoKPP, may find they are more likely to have attacks at particular times during their monthly cycle, at ovulation and in the days before menstruation.

Look at your episodes. Notice what you did or ate during the 24 hours prior to your episode of weakness. Did you eat a plate of mashed potatoes, a Big Mac and fries, four pieces of apple pie? A cantaloupe or a bag of dried apricots? Maybe you played 18 holes of golf or went dancing. Make a note of your diet and activities in relationship to your episodes of weakness. Is there always a dip in strength an hour after eating sweets or 30 minutes after your jog around the park? Is there anything you eat that consistently makes you stronger? Maybe drinking a soft drink or eating candy makes you stronger, perhaps a salty, high carbohydrate meal makes you feel better?

If you are experiencing continual weakness the variations may be very subtle. Some patients, especially older patients or those who have ATS, may have what is called an *abortive attack*. This kind of episode can literally last for weeks, even months, at a time, leaving the patient feeling weak/greatly fatigued almost continually, yet the attack never develops into paralysis. In these cases it is important to watch for fluctuations in strength that may follow a pattern. There may be a consistent pattern of morning weakness with greater strength in the evening, or a slightly greater period of strength at one particular time of the day.

Food Triggers

Food triggers for episodes vary, depending on the type of periodic paralysis. Those who have HyperKPP may find that high-potassium foods like cantaloupe, bananas, dried fruit, avocado, nuts and beans may trigger weakness. Those with HypoKPP are more apt to be bothered by sweets, starchy foods like bread, pastry, cakes and cookies, pasta, rice and by salty foods. Patients with either HypoKPP or HyperKPP may find that potato, orange juice and other fruit juices are triggers. This is because these foods have a high amount of potassium, which bothers the person with HyperKPP. At the same time these foods have a high amount of starch or sugar, which can trigger HypoKPP. Patients with HyperKPP accompanied by Paramyotonia Congenita, and some Andersen Syndrome patients can have episodes which can be triggered by potassium fluctuations in either direction.

Preparation of foods can affect their ability to trigger an episode. Many patients report that while a baked or boiled potato will trigger weakness, French fries don't seem to be a trigger food, unless they are heavily salted. This is because the fat content slows digestion and both the carbohydrate and the potassium enter the blood at a slower rate. High fat foods are less likely to trigger an episode, so that pasta with a very rich sauce may not trigger episodes but pasta with a low-fat sauce will. The same is true of desserts. A person with HypoKPP may tolerate a small dish of full-fat ice cream, where the same size serving of low-fat sherbet would trigger an episode.

Sodium is a trigger for HypoKPP. Patients are advised to eat a diet low in salt, under two grams daily. Many patients report feeling better when they eat less than one gram of salt a day. This is a challenge. It requires that practically everything one eats be prepared from "scratch", with no added salt, and that no salt be added at the table.

Patients with any type of periodic paralysis may find sleep, rest after exercise, eating a big meal, or getting too hungry triggers an attack. Becoming chilled or overheated will provoke an episode in many people.

Once you've correlated a trigger with an episode look at ALL your episodes and see if they have a common trigger. They may not have. It may take a combination of several triggers to initiate an episode, and they may not all be the same triggers every time.

With a graph like this, you can begin to recognize a pattern to your weakness, and see what triggers episodes. If the foods which trigger episodes are high in carbohydrates then you may suspect that your episodes are associated with low potassium. On the other hand, if high potassium foods make you more likely to have an episode and eating sweets make you feel stronger when you are weak, you are likely to have one of the hyperkalemic forms. You might see that it takes a combination of two or more triggers to trigger weakness. You might be able to enjoy a high carbohydrate food if you were not under stress, but a combination of the two might make you susceptible to an attack.

Documenting Potassium Fluctuations

If you have paralytic attacks it is a good idea to arrange with your family or friends to take you to the Emergency Room of the nearest hospital as early as possible during an attack – if possible as soon as it is apparent that you are going to have a paralytic attack. You need to go early because the potassium fluctuations found in periodic paralysis return to normal well before paralysis begins to resolve. If you wait until well into an attack it may be too late to catch the abnormal potassium level.

You should speak to your doctor and ask for an order for a stat potassium draw. This means that when you go to the hospital during an attack and your family member or friend gives the doctor's order to the admitting desk they should arrange for a potassium draw to be done as quickly as possible. Ask that the doctor write on the order that it is okay to give a photocopy of the results of the test to you. This will go into your documentation file. It's absolutely vital that you get copies of all test results as these will be very important in determining whether you have periodic paralysis or not.

Do I Have a Family History of Periodic Paralysis?

If your mother or father, brothers or sisters, cousins, a grandparent, or any other blood relative has symptoms which are similar to yours you should document that. If you think you have inherited periodic paralysis it's a good idea to put together a family medical history which shows who else in the family is affected.

Talk to your relatives in person or on the phone, or send them a survey. Gather information on as many generations of relatives as you can, including your parents, grandparents, sisters, brothers, any half brothers and sisters, aunts, uncles, nieces, nephews, children and grandchildren.

Gather as much accurate information as you can. If no one can remember information about a family member, don't guess. Incorrect information will give you incorrect results. Do your best to collect solid information about your closest relatives — parents, children, sisters and brothers. Include each person's name, who their parents were, their birth date, age at death and cause of death. Then include any major health conditions this person may have had, like cancer, heart attack, stroke, rheumatoid arthritis, kidney disease or other disorders.

You may find that relatives do not want to talk about a disorder which may be inherited. Some people still see genetic illnesses as “taints”. This calls for tact and compassion. Consider these ways to get family members to open up and share personal information:

Explain your purpose. Emphasize that your purpose is to create a record that will help you determine whether you and your relatives have a family history of periodic paralysis. Offer to make the medical history available to other family members so that they can share the information with their doctors.

Provide several ways to answer questions. Some family members may be willing to share health information face-to-face. Others may prefer answering questions by mail or e-mail. **Word questions carefully.** Don't start with personal questions. Begin by asking questions about the whole family and let your relative volunteer their own health information. **Be a good listener.** As your relatives talk about their health problems, let them speak without interruption. Listen without judgment or comment.

Respect privacy. As you collect information about your relatives, respect their right to confidentiality. Some people may not want to share any health information with you. Or they may not want this information revealed to anyone other than you and your doctor.

To find out about periodic paralysis you might ask questions like these:

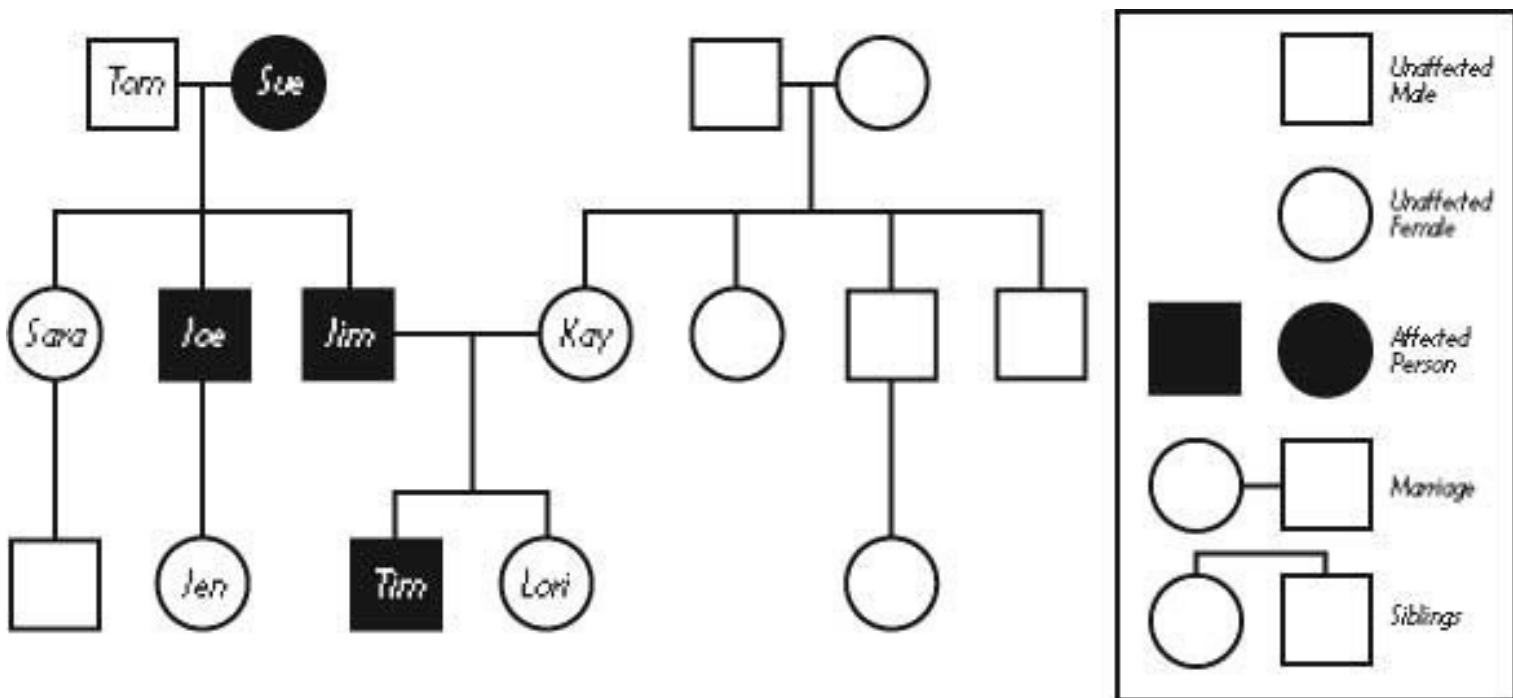
1. Do you remember any older relatives having trouble climbing stairs?
2. Did any complain of morning weakness
3. Did any have weak spells, where for hours or days they couldn't do daily chores?
4. Did any family members complain of muscle stiffness in the cold?
5. Did any family members lose the ability to walk in their 60s and 70s?
6. Did any family members ever have a trouble with surgery, for example running a very high temperature, developing rigid muscles or having muscle paralysis?

Putting Together a Family Tree

Once you have assembled your family medical information you will be able to build a chart which shows at a glance who may have been affected, who was definitely affected and who was not. The following illustration shows how a simple chart is drawn up and labeled. You can see that unaffected persons are indicated by white and affected ones are indicated by black. Males are represented on the chart by a square. Females are indicated by circles. A straight line joins a male and female who are married, or who have children; their children are indicated by drop brackets. Although not shown on this chart a person who has died is indicated by placing an x across their symbol.

The accompanying chart illustrates an autosomal dominant pattern of inheritance, as is found in the PPs. An affected parent, or a parent who is a carrier (one who has the gene mutation but has no symptoms) passes it to their children. Two or three generations of carriers may pass before an affected child shows up.

In this example you can see that the mother, Sue, is affected, while her husband Tom is not. Their children Joe and Jim are affected. Jim marries unaffected Kay and they have two children, Tim and Lori. Tim is affected but Lori isn't. The person preparing this chart might have been Tim, Jim, Joe or Sue, but is probably Jim, because his wife Kay's unaffected family is shown.



You should also number each person on your chart, and make a reference sheet which gives their full name, birth and death (if applicable) dates, cause of death, and their relationship to you. This should make it instantly obvious if you have a family pattern of inheritance. Take your completed family medical history with you to your doctor's appointment, as part of your documentation.

What If I Don't Find a Family Connection?

About 30% of all cases of periodic paralysis are sporadic, that is to say they are spontaneous mutations which occurred at the moment of conception. So the lack of a family history of PP doesn't mean you don't have PP. It may also mean that the affected relative was two or three generations previous and no one remembers that they had weakness.

Testing for Periodic Paralysis

The periodic paralyses are rare disorders. Most physicians will have never heard of any of them. Neurologists are the specialty physicians who care for patients with periodic paralysis, but even most neurologists

have no experience with PP. It is not at all unusual for patients to go for years without obtaining a diagnosis, or to never obtain a diagnosis. Some patient's symptoms are only recognized as periodic paralysis years after their death, when one of their children, grandchildren or other close relative is diagnosed.

It is our goal that, if you have periodic paralysis, you will be able to obtain a diagnosis. For this reason we are going to focus on testing which can be done by a local physician who is not a specialist or expert in periodic paralysis. There are highly specialized tests which we will discuss later, but these are available only in a very limited number of centers and from a limited number of neurologists. Experience has taught us that the best care is frequently found at a local level, from a physician one has developed a long-term and trusting relationship with.

What to Do If Your Potassium is Low During Attacks

Let's assume that you have kept a triggers journal and have determined that your episodes are triggered by high carbohydrate meals, by rest after unusual activity and by stress. You have had several blood tests which show that between episodes your potassium level is well within the normal range, and you may have had a low serum potassium reading during one or more episodes.

At this point it is important that your Family Physician or Primary Caregiver determine the origin of your hypokalemia. There is a simple way to determine the origin of hypokalemic paralysis. It requires that blood be drawn and a urine sample be obtained during an episode of hypokalemic paralysis. The information about this test is medically complex, and you may wish to print off the webpage found at:

<https://hkpp.org/articles/determining-the-origin-of-hypokalemic-paralysis-2/>

and share it with your physician. In plain English it says that when the level of potassium in the blood drops the kidneys stop excreting potassium in the urine, unless the kidneys are the problem.

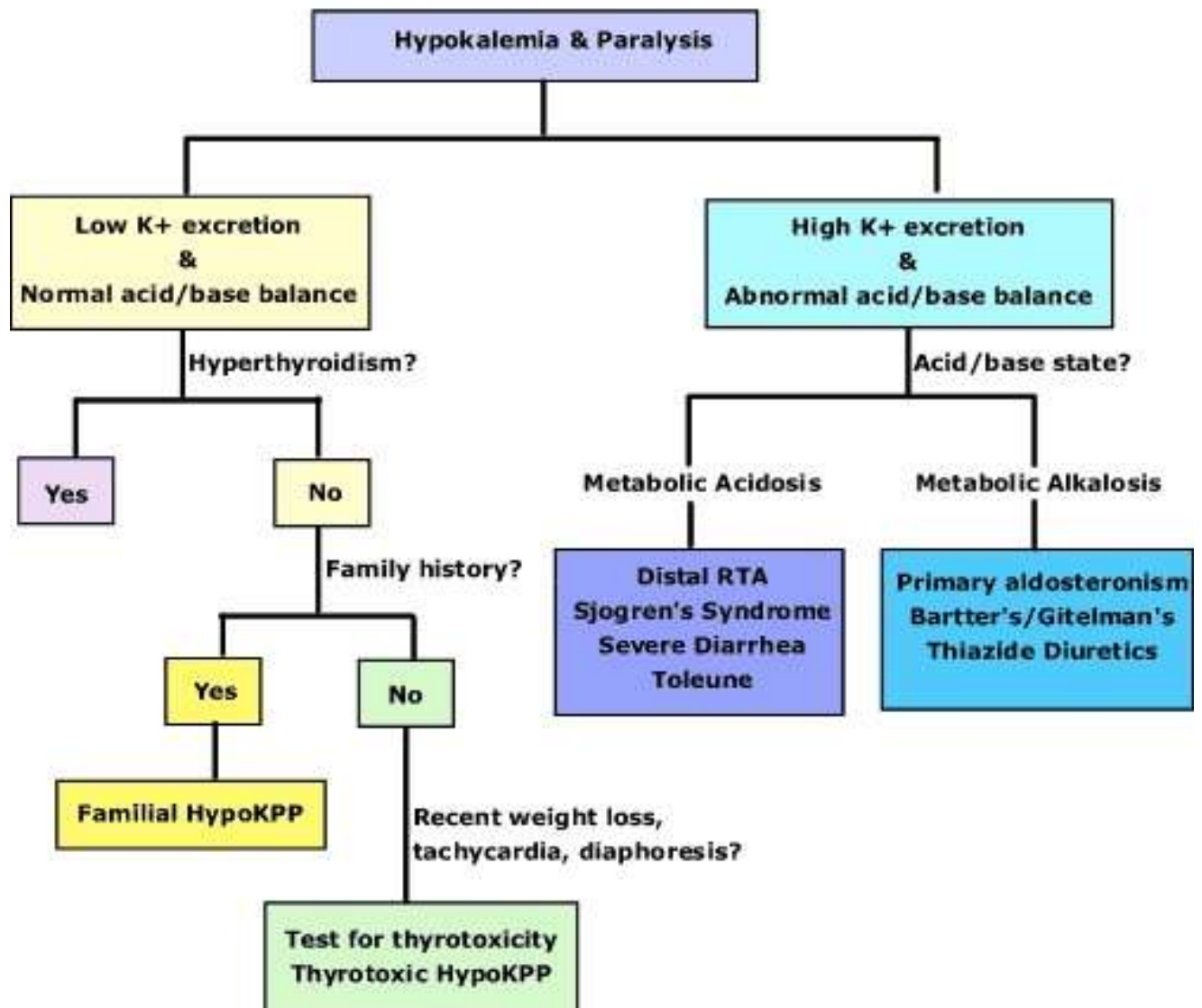
For example: A 13 year old boy with inherited Hypokalemic Periodic Paralysis woke up at 5:00 am completely paralyzed. Because he vomited and could not take oral potassium he was admitted to the hospital at 11:00 am for treatment with intravenous K⁺. Laboratory tests were done beginning at 11:00 am and for the next 22 hours, until 9:00 am the following day, when he was fully recovered. Urine was collected at 14:00 after 15 hours without urination.

Time in hours	11:00:00	14:00:00	17:00:00	22:00:00	09:00:00
Serum potassium	2.3 mmol/L	3.3 mmol/L	4.6 mmol/L	4.6 mmol/L	4.7 mmol/L
Total Urine Potassium excreted	-	5 mmol	8 mmol	10 mmol	25 mmol

During an attack of Ion Channelopathy Hypokalemic Periodic Paralysis the kidneys excrete practically NO potassium. The potassium moves into the muscle cell temporarily. By comparing the amount of potassium in the blood to the amount in the urine, and factoring in other properties of the urine, it's possible to tell whether the hypokalemia is due to the shift of potassium into the muscle cell or if it is being excreted by the kidneys.

Not all hypokalemic paralysis is a result of one of the channelopathy periodic paralyses. The diagnostic flowchart below shows some of the more common disorders associated with hypokalemic paralysis, and how basic testing can clarify where symptoms are coming from. After the origin of the hypokalemia is determined the patient and FP can then seek a referral to the appropriate specialist.

Diagnostic Flowchart



Testing for Thyrotoxic Hypokalemic Paralysis

Thyrotoxic Hypokalemic Periodic Paralysis (THPP) develops as a result of an overactive thyroid gland. The thyroid gland is located in the neck and produces hormones that regulate growth, digestion, and metabolism. A complex set of mechanisms control the rate of thyroid gland activity. Hyperthyroidism or thyrotoxicosis results from too much thyroid hormone within the body.

Hyperthyroidism (an overactive thyroid gland) is not a specific disease, but a symptom of some underlying condition or disease. The causes of hyperthyroidism include Graves' disease, tumors of the thyroid or other endocrine glands, inflammation or infection of the thyroid, taking too much thyroid hormone and taking too much iodine.

Graves' disease accounts for 85% of all cases of hyperthyroidism.

THPP occurs most often in men of Asian descent, including Japanese, Chinese, Vietnamese, Koreans and Filipinos. It also occurs more frequently in those of Native American and Latin American descent. Almost all cases of THPP occur in men, but the occasional woman develops THPP.

The hypokalemic paralysis found in THPP is almost indistinguishable from inherited HypoKPP, with a few extra features thrown in. THPP patients are apt to report recent weight loss, an intolerance to heat and heavy sweating, in addition to muscle weakness.

To check for THPP physicians test the levels of thyroid-stimulating hormone (TSH; reference range: 0.45-4.5 μ U/mL) as well as free thyroxine (FT4; reference range: 8-20 pg/mL) and free tri-iodothyronine (FT3; reference range: 1.4-4 pg/mL). A low level of TSH together with high levels of FT3 and FT4 indicates that hyperthyroidism is present and is probably the cause of paralytic attacks. THPP is curable. When treatment corrects the thyroid problem the THPP goes away.

Testing For Hyperkalemic Periodic Paralysis

In Hyperkalemic Periodic Paralysis the attacks of flaccid muscle weakness usually begin in the first decade of life. Potassium-rich food or rest after exercise may trigger an attack, as may exposure to cold or stress. Patients with HyperKPP usually have a high normal potassium level during attacks, but the potassium level returns to baseline after the attack is finished.

The physician will want to rule out Addison's disease and other adrenal disorders, as well as inherited kidney disorders which cause hyperkalemia. After these disorders have been ruled out testing for HyperKPP can begin.

The patient's diary showing that relatively short attacks are triggered by exercise and foods high in potassium can help suggest a diagnosis. A test which may be performed by the local physician includes one in which the person rides a stationary bike (or walks on a treadmill) for 30 minutes to increase the heart rate to 120-160 beats/min, followed by absolute rest in bed. An affected person's serum potassium level rises during exercise, declines after exercise, and rises a second time 20 minutes after exercise ends.

Testing for Paramyotonia Syndromes

Information from the patient's diary and the physician's observations that attacks of muscle stiffness are associated with potassium fluctuation, are triggered by repetitive movements, exertion and exposure to cold should lead the physician to refer to a neurologist familiar with the Myotonias. Paramyotonia can be difficult to diagnose without specialized testing. Most tests call for cooling the muscle, as symptoms often do not show up at room temperature. There are no easy or simple tests a Family Physician can do, though in some PMC attacks the serum potassium rises to above 4.5 mmol/L and there is a high urinary excretion of potassium.

The neurologist may cool a hand or arm muscle and do Electromyographic (EMG) studies in order to reveal the paramyotonia pattern. Sometimes it takes several attempts before this is successful.

Testing for Andersen Syndrome

Patients with AS have the same kind of paralysis attacks as patients with other forms of periodic paralysis, but they also have problems with irregular heart rhythms. The heart problems found in AS are in the ion channels which regulate the heart rhythm, the force with which the heart beats and the time it takes for the heart muscle to recharge and prepare to beat again. If left untreated, the irregular heartbeats can lead to discomfort, fainting, or cardiac arrest.

Physical abnormalities associated with Andersen Syndrome typically affect the head, face, and limbs. These features may include a small lower jaw, dental abnormalities, low-set ears, widely spaced eyes, an unusual curving of the fingers or toes and webbed toes. Aside from establishing that the person has potassium fluctuations during episodes, and noting any physical abnormalities, the family physician can probably do little to diagnose AS. It is the combination of paralysis, the physical characteristics and the typical heart problems which distinguish AS. The heart arrhythmias found in AS are much like those found in the Long QT Syndromes. Assessing the heart problems which come with AS requires consultation with an Electrophysiologist, a Cardiologist who specializes in heart rhythms and a Neurologist who is familiar with periodic paralysis.

The CMAP Test

There are some highly specialized tests for periodic paralysis. Any neurologist who is well trained in electromyography can perform the McManis Protocol, also called the Compound Muscle Amplitude Potential test (CMAP). Many do not simply because they have never been called upon to do so. In the body, groups of skeletal muscle fibers are “wired” to a single nerve ending. When that nerve ending is stimulated all of those fibers contract at the same time, to the same tension. The Compound Muscle Action Potential (CMAP) is the medical name for that “leap to action” of a group of muscle fibers.

The CMAP test is based on the observation that patients with PP have a greater than normal increase in the strength of the compound muscle action potential immediately after two to five minutes of exercise. In the CMAP test the “exercise” is usually provided by contracting the thumb and forefinger against resistance, like a rubber band, or tape. The patient exercises in an off-and-on pattern for two to five minutes. The patient then rests the muscles for next 30 to 40 minutes while measurement of the CMAP amplitude continues.

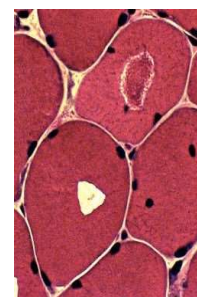
In people without muscle disease the CMAP amplitude declines an average of 15% regardless of age or sex. In those with Periodic Paralysis there is a steady decline in CMAP amplitude to less than 40% of the patient's rested baseline level. In the hands of the experienced examiner the CMAP test is reliable 70%-80% of the time.

In studies, patients with hyperkalemic PP, on average, have much greater amplitude increases and decreases than patients with hypokalemic PP, but there is overlap between individual patients.

It is important to note that factors such as the time of day, temperature, serum potassium levels, and recent meals may affect the susceptibility of muscle-induced exercise in PP. Day-to-day variability may also be present in a given individual and affect the outcome of the test. Katz et al. suggest that the CMAP test can be used to determine the effectiveness of therapy by comparing CMAP decrements pre and post treatments, so it is obvious *the patient must be unmedicated for results to be accurate diagnostically. A medication-free period of 72 hours is required before the patient undergoes a CMAP test.*

Muscle Biopsy

A muscle biopsy, a procedure in which a small piece of muscle tissue is removed with a special needle during or following an episode, may be done in some cases. The biopsy sample is sent to the lab for analysis. Muscle tissue is often slightly abnormal in periodic paralysis, reflecting changes which occur in muscle tissue over time. It is often possible to look at a small section of affected muscle and confirm the diagnosis.



Genetic Testing

Periodic paralysis is caused by a mutation in genes that control the development of certain ion channels in the muscle membrane. Ion channels are openings that pierce the muscle membrane and act as gates for the movement of ions in and out of the cell.

The ion channel which is affected determines the type of periodic paralysis. So far about 50 different mutations have been identified in the sodium, calcium or potassium channels. DNA testing is available for a limited number of mutations, but cannot be relied on for diagnosis. Many patients who have clear family histories of periodic paralysis and have been diagnosed by timehonored and medically accepted standards are still testing negative on commercially-available DNA tests.

Commercial labs test for only the most common gene mutations found in HypoKPP and HyperkPP although more than 50 mutations have been identified. There are no commercial lab tests for Paramyotonia Syndromes or Andersen Syndrome. There are some genetic research projects which patients may be able to join once they are diagnosed. These research programs are trying to identify the remaining “mystery” mutations. This should make it obvious that someone with a negative DNA test may still have periodic paralysis.

While Pursuing a Diagnosis

- 1) Keep a careful Triggers Journal.
- 2) Organize a family medical history.
- 3) Ask for and keep a copy of all lab reports, Electrocardiograms, doctor's reports etc.
- 4) Organize your information into categories for easy reference.
- 5) Report only the three most distressing symptoms to the doctor, let him/her ask the questions from that point.
- 6) Stay objective when reporting symptoms. Don't dramatize.
- 7) Stay informed. Keep reading and learning. Be prepared to ask questions.
- 8) Realize that diagnosing a rare disorder often takes quite a long time.

Glossary

Terms commonly encountered when reading about periodic paralysis:

Abortive Attack: Long-lasting period of weakness which stops short of paralysis, often characterized by daily fluctuations. Abortive attacks can be so persistent that they can be confused with Permanent Muscle Weakness (PMW). [More information](#)

Anaphylaxis / Anaphylactic shock: A severe allergic reaction.

Arrhythmia: Disturbance of heart rate and rhythm.

Bradycardia: An abnormally slow heart rate and pulse, less than 60 beats per minute

COPD: Chronic obstructive pulmonary disease. Group of diseases and conditions in which the lungs suffer a decline in their ability to exchange gases.

CVA: Cerebral vascular accident or stroke. Damage or blockage to the blood vessels of the brain resulting in a lack of blood supply to the brain.

EKG: Electrocardiogram (also called ECG).

GERD: Gastroesophageal reflux disease. [More information](#)

GI: Gastrointestinal.

GU: Genitourinary

HDL: High density Lipoproteins (“Good Cholesterol”) [More information](#)

Hyperglycemia: Excess of sugar in the blood. [More information](#)

Hyperlipidemia: High Cholesterol. [More information](#)

Hypertension: High blood pressure. [More information](#)

Hypoglycemia: Too little sugar in the blood [More information](#)

Hypotension: low blood pressure [More information](#)

IDDM or NIDDM: Insulin dependant Diabetes Mellitus (Type I Diabetes) or Non-insulin dependant Diabetes Mellitus (Type II Diabetes). [More information on Diabetes Mellitus](#)

LDL: Low density lipoproteins (“Bad cholesterol”) [More information](#)

Malignant Hyperthermia: A syndrome characterized by increased body metabolism and muscle rigidity that can result in death. It is triggered in susceptible individuals by commonly used general anesthetics. [More information](#)

Milliequivalent (mEq): A system used to quantify electrolytes. Specifically, an equivalent is the amount of substance needed to combine with one mole (6.02 \times 10²³) of H⁺ ions or OH⁻ ions. A milliequivalent is 1/1000 of an equivalent. **mmol/L:** the abbreviated form of millimoles per litre, a term used to describe how much potassium or other substance (like glucose) is present in a specific amount of blood.

NSAIDs: Non-steroidal anti-inflammatory drugs

OTC: Over the counter - Medication that does not require a prescription.

Palpitations: Heartbeats that are unusually strong, rapid or irregular enough to make a person aware of them. They are felt in the chest, throat, and neck and may be regular or irregular in nature.

Paralysis: Complete or partial loss of the ability to move a body part. Sensation in the area may also be lost.

PMW: Permanent Muscle Weakness. Weakness caused by permanently flaccid muscle cells, most likely progressive in nature and occurring over decades.

Syncope: Loss of consciousness due to many causes, usually a dramatic fall in blood pressure, which leads to fainting.

Tachycardia: Rapid heart rate greater than 100 beats per minute

HKPP Website

Our website at: <http://hkpp.org/> has many useful and informative articles on the PPs. Many of the concepts discussed in this booklet are covered in greater detail on the website. We hope this booklet is of help to you in your quest for a diagnosis.

The author gratefully acknowledges the assistance of Don, Tina, Gill, Linda, Leilani and others who proofread this manuscript and offered valuable advice.

The information in this publication is based on current medical knowledge but should never — at any time — be substituted for the advice and care of a qualified medical consultant. For medical advice seek the services of a physician.

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Notes