What is Periodic Paralysis ?
"PERIODIC PARALYSIS IS NOT YOUR FRIEND"

By Susan Q. Knittle-Hunter

Periodic Paralysis (PP) is a rare, hereditary disease characterized by episodes of muscular weakness or flaccid paralysis without the loss of sensation or consciousness. It is a channelopathy; a disease involving dysfunction of an ion channel for potassium, sodium, chloride and calcium. Periodic Paralysis is listed by the Muscular Dystrophy Association (MDA) as a myopathy**;** a disease of the muscle.

There are three types:

Hypokalemic Periodic Paralysis (HypoPP): Paralysis results from potassium moving from the blood into muscle cells in an abnormal way. It is associated with low levels of potassium during paralytic episodes.

Hyperkalemic Periodic Paralysis (HyperPP): Paralysis results from problems with the way the body controls sodium and potassium levels in cells. It is associated with high levels of potassium during paralytic episodes. In

Andersen-Tawil Syndrome (ATS): Paralysis results when the channel does not open properly; potassium cannot leave the cell. This disrupts the flow of potassium ions in skeletal and cardiac muscle. During paralytic episodes, ATS can be associated with low potassium, high potassium or shifts in the normal ranges of potassium.

I have Periodic Paralysis (PP). I am 62 years old and was newly diagnosed on February 7, 2011. I was misdiagnosed for many years. The type I have is a form of Andersen-Tawil Syndrome. I have had episodes of partial and total paralysis for many years. During the episodes, my potassium shifts are low (hypokalemia), high (hyperkalemia) and within the normal ranges (normokalemia). Due to several misdiagnoses and a lack of proper diagnosis and treatment for over 50 years, I have become totally and permanently disabled with weak muscles throughout my body including those involved with my vision, digestion, breathing and my heart. I must be on oxygen constantly and cannot exert myself in any way. I have had a heart loop monitor inserted in my chest to monitor the tachycardia and arrythmias, that include long QT interval beats. I sit in a recliner for most of my day and can walk (with a walker) only from one room to another or stay on my feet only short periods of time to do simple tasks like brushing my teeth. I must use a motorized wheelchair if I leave home or need to go any distance. If I did not have the help of my husband, I would have to live in an assisted living program.

I have discovered there are many triggers that set the partial and total paralysis into motion. It has been important for me to discover these triggers because I need to stop the episodes, if possible, in order to regain some quality of my live and to prevent further damage to my organs as the potassium shifts and depletes in my body. This damage has lead to the permanent weakness and disability. Because I have Andersen-Tawil

Syndrome, the paralysis leads to tachycardia and serious arrythmias, including long QT intervals, which can lead to cardiac arrest. Avoiding paralysis is absolutely necessary for me, due to these life-threatening conditions. From my research, I find the latest literature indicates certain foods, medications, conditions and activities can trigger most the paralytic events. Every moment of my present life I must control my symptoms. I must walk a constant "tightrope" of sorts.

The following list/plan was put together after trial and error in my own quest for treatment and management. I had no doctor assisting me and gleaned as much as possible on the internet and in discussion with other people who live with Periodic Paralysis and Andersen-Tawil Syndrome.

My triggers:
simple carbohydrates,
complex carbohydrates,
most meat,
salt,
sugar,
caffeine,
medications including over-the-counter medications,
exercise,
rest after exercise,
sleep, all aspects: falling asleep, during sleep, waking up and napping.
stress (good or bad),
dehydration
msg
food in general
large meals
fatigue
fasting
and ? I still have not discovered all of my triggers. No matter how careful I am, I can still go into paralysis without knowing why.

How I relieve or ward off my symptoms:
avoid triggers
following a proper ph balanced diet, eating from the farm; not the factory
take no medications including over-the-counter medications
avoid stress
no exercise
no exertion
get plenty of rest
stay well hydrated
constsnly monitor vitals
take potassium when needed (if low potassium) (If under 3.0 go the the hospital)
take sugar or glucose tablets as needed (if high potassium) (If over 6.5 go to hospital)
24/7 oxygen

How I diligently monitor my vitals:
I use several pieces of medical equipment for measuring my vitals. These items are necessary for my caregiver to monitor me while in paralysis or for me to know which direction my potassium shifts for proper treatment.
cardy meter,
finger pulse oximeter,
blood sugar monitor,
stethoscope,
wrist blood pressure monitor,
a thermometer and a digital
ph balance reader
litmus paper

I had to gather a team of medical professionals:
(I have an MD as my primary care provider, a neurologist and a cardiologist. I no longer have a renal specialist. He felt he could do no more for me. I have a counselor who helps me deal with my failing health and the changes in my life related to that. I have a geneticist who is studying my blood in Germany. Except for the geneticist, none know about my disease. I must provide them with information on how to treat me.)

I educate myself: I must educate myself, my family, my friends, my neighbors, my community, my doctors, my hospitals, my dentist, my optometrist and my local EMT's about every aspect of my condition. Knowing and understanding the syndrome eases my fears and the fears of those around me and assists me with proper management and treatment. Knowing others will be able to aid me during paralysis episodes is essential.

I have joined and created Periodic Paralysis Groups: Being part of a Periodic Paralysis community is vital. I have discovered I am not alone. I receive encouragement, support, sympathy and empathy. I will gain information and knowledge from others who live with the same enemy daily. I ask questions and share ideas.

The above plan is followed constantly and diligently. It is a constant "tightrope" I must balance, day-by-day, minute-by-minute; second by second. I cannot let up for even a minute or I can go into paralysis and the cycle begins anew.

When I slip into an episode, I now become totally paralyzed after years of partial paralysis. I wrote the following in May 2010, describing what happens when I am paralyzed.

> *Usually, I wake up in the morning and I am paralyzed. I find I can't move. I can't open my eyes. My mouth is open. I can't breathe through my nose. I have urges to swallow but can't so there is a choking sound in my throat every few minutes. Sometimes my heart will race or beat irregularly, though usually, there is no problem with my heart. I don't*

have any or much feeling in my body. My mouth is very dry. I cannot speak.

As I begin to come out of it, my mouth will start to get saliva, my eyes will open but I can only see what is in front of me, since I can't move my head. Sometimes my eyes will jerk around when I first open them, usually jerking up. My body will sometimes jerk a little. Sometimes there is a big breath my body will take

Sometimes, I will go back into it. My eyes close, I feel very hot and all the symptoms return. Sometimes there will be a few jerks as I go back into it.

During all of this I am awake and am aware of everything going on around me. Sometime I begin to cry, due to the frustration, and fear. I can feel the tears running down my cheeks.

If I have these at times other than upon waking, the symptoms are the same. I get a strange sensation of heat body wide, usually beginning in my back. My eyes will close and then my body goes limp. I may have a few jerks as I am going limp. My mouth will open and I am in it…unable to move, speak or open my eyes.

Sometimes, I don't go too deep. It is all the same but I am able to open my eyes and can speak a little with a tight tongue and tight lips. My mouth is still open, however. I can't move my body.

Once one of these begins, it may last up to 45 minutes to an hour, or can be as short as about 10 minutes, if it is a second or third one in a row.

It takes about 15 to 30 minutes to come out of it all the way. I am always left with lingering weakness for many hours that can linger into days. Speaking is difficult. Walking is difficult. My arms and hands come back sooner than my legs. I begin to get feeling back in my body. I can move my lips. I begin to breath thru my nose again. It is difficult to speak or move but it gradually comes back. Speech is very difficult, my lips don't want to move. My tongue is difficult to move. I will suddenly have an urgency to urinate. If, at this point, I get help to the bathroom, I am like a rag doll, especially my legs. My arms flail, like a child just learning to stand and walk; balancing herself.

For many hours, I remain too weak to do much of anything but sit up in bed or sit in a recliner. I must use my walker or a wheelchair..

It is difficult to know what brings these episodes on. I know that sleep has something to do with some of them, but not all of them. I know that

> *sometimes, when I wake up during the night with an urgency to urinate, I am coming out of one, because I have all of the symptoms previously discussed. My arms and hands and legs are numb and feeling is just coming back. My mouth is tight and dry. Walking is difficult."*
> (May 28, 2010)

Through the past years of my physical decline, I have had to give up my career as a special education teacher, my hobbies to include hiking, walking, swimming, exercising, fishing, camping, traveling, shopping, cooking and baking. I can no longer drive. I have lost many friends, because I could not keep up with them or entertain any longer. I have lost contact with family members who did not understand or did not want to watch my decline or who thought I was a hypochondriac. I have lost the connection I once had with my grandchildren because I can no longer keep up with them or continue a meaningful relationship with them. The relationship with my husband has changed from husband and wife to caregiver and patient. Most of the over 30 doctors I have seen in the past 6 years have treated me poorly and like I was mentally ill.

I have heard it referenced that Periodic Paralysis is our "friend". A friend is a person whom one knows, likes, and trusts. A friend is a supporter, an ally and a well-wisher. A friend is a person you know well and regard with affection and trust. That being said, I must disagree with this ideation. Periodic Paralysis is not our "friend". After all, a friend does not leave you totally paralyzed. A friend does not leave you unable to walk. A friend does not leave you unable to talk. A friend does not leave you in fear. A friend does not take away your quality of life. A friend does not leave you disabled. A friend does not keep away your friends and family. A friend does not keep you from doing your favorite things. A friend does not change the relationship between you and your spouse. A friend does not make you appear to be mentally ill. A friend does not treat you poorly by the very people you most need to help you. A friend does not take away your hope, dreams and desires. So, I must disagree with this ideation that Periodic Paralysis is our "friend". Periodic Paralysis is not my "friend". Periodic Paralysis is not our "friend".

We cannot, we must not treat it as our friend. We cannot embrace it. We must treat Periodic Paralysis as an enemy that we must battle diligently; daily; minute-by-minute; second by second. Only by doing so, is it possible to keep our "foe" regulated and managed in order to decrease the number and severity of paralytic episodes and the amount of possible permanent damage. We must walk a constant "tightrope" of sorts.

In conclusion, Periodic Paralysis is a very rare disease that in some cases steals one's life away. It is not our "friend", but rather an enemy that must be battled every second of our waking hours and sometimes while we sleep. Many people who suffer from it live in fear and despair as they seek help from doctor after doctor. Some will die before they get the help they desperately need.

This is not the normal, technical description of Periodic Paralysis that you will find in the textbooks, but rather the honest description as seen through the eyes of one who lives with a form of Andersen-Tawil Syndrome, the most rare form of Periodic Paralysis.