

PSPCAN

PROGRESSIVE SUPRANUCLEAR PALSY
SOCIETY OF CANADA

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ASK THE NEUROLOGIST- TRANSCRIPT
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Theme 1: PSP Causes & Risk Factors

MODERATOR

So, the question was, is PSP hereditary? And are there any studies about it running in families?

DOCTOR

- PSP belongs to a family of diseases, called frontotemporal lobar degeneration (FTLD) and that family of diseases has a strong genetic component
- PSP itself has a minor genetic component
- There are a few families that have PSP in them and there can be some genes that are associated
- One of the genes that we usually test for is called microtubule associated protein tau (MAPT)
- That is the gene that is most associated with PSP but usually PSP is not genetic and MAPT mutations are associated with other diseases of the FTLD family
- At a research level, we are continuously looking for genes associated with this illness.
- In terms of studies, yes, we do have studies. We are still searching for genetic predisposition to that disease.
- There may be genes that are associated, but we have not found them yet. In my lab, I collaborate with groups around the world and we try to look for genetic variations. So far, we have not found very much, but we have not given up looking.

MODERATOR

Next question, are there any new studies about pathogens and etiology? And on the same vein, are there risk factors or causes or underlying etiologies such as mitochondrial dysfunction?

DOCTOR

- We do not know what are the causes of PSP and we do not know why people get PSP. When a person dies, we see abnormalities in the brain that are consistent with PSP pathology
- Right now, although we can tell what it looks like in terms of the pathology with

abnormalities in the brain, we do not know why it happened

- Yes, there are many studies and researchers are looking in all directions
- People are looking at environmental exposures. People are looking at dysfunction of mitochondria
- There are many processes that get done by the brain, work to eliminate bad proteins, work to package things, and abnormalities in all these areas are being investigated as possible causes.
- When you do not know what the cause is, you must keep an open mind and look. You have to cast a very wide net. In the brain diseases, we have not actually made as much progress as we have in the heart and in cancer, but they have been at it for a much longer time.
- The brain is more complicated than the heart. And we have a lot of different types of cells in the brain. And there is a lot of machinery in all the cells that keep the cells alive. So, there is many areas to look at. Like there are many parts of this assembly line or processing plant to look at
- Although we have made some headway, there have been clinical trials that have tried to eliminate tau in the brain, which before that we did not do. We still do not know why it is that the tau starts to accumulate in the brain of people with PSP
- Everybody has tau, and that abnormal tau exists in other diseases, but it is malformed in different ways.
- It is interesting because mitochondria are the workhorses of our bodies, they are essential. There is more and more interest in the role of mitochondria in all diseases that are neurodegenerative.

MODERATOR

Is there any connection between PSP and recessive lysosomal?

DOCTOR

- Lysosomes are part of the machinery to eliminate things. So, it makes sense that you

would look at that since abnormal tau accumulates

- There are many studies being done and there are some studies that find things, some that do not.
- We do not have clear evidence to say, let us go after this, because that is clearly going to get us to the end goal.
- Low hemoglobin is very, very prevalent in the world, it is the cause of anemia. And when you think of the prevalence of PSP, it is actually very low compared to the prevalence of anemia in our society. I would not put that one too high on the list, but I would not take it off completely.

Theme 2: Movement & Physical Therapy

MODERATOR

We have been adding movement sessions while respecting fatigue. Are there specific movements and exercises that we should be implementing to slow the progression of the disease? This is specific to CBD.

DOCTOR

- There is a lot of evidence that aerobic exercise is good for the brain. Aerobic means things that make your heart rate go up.
- The reason that it is good for the brain is that there is a molecule that is increased when you do cardio activity. It is called brain derived neurotrophic factor. That molecule does not come in a pill form, it goes up when people exercise.
- In my practice, I prescribe exercise to everybody and you know, stationary bicycle when people are unstable is usually fairly well tolerated by people as long as you get some help sitting on a recumbent bicycle and you can still pedal. Some people can still get in the water and move around.
- The evidence is for aerobic exercise so anything that makes the heart rate go up, it can be fast walking, cycling, running, dancing, swimming . We tell people to do what you can.
- Sometimes CBD patients have a hard time because they do not even have control over the limb, even if they want to move it. So even passive movement is good because in that case, you keep the joints and the range of motion in the joints
- Movement, if it is safe is good.

MODERATOR

Are there any references for simple arm and neck exercises the caregivers can model and deliver to help support, mobility, and flexibility?

MODERATOR

- The PSP society has a physical therapy program. We have partnered with a physical therapist who is a specialist in neurological movement disorders and she is helping us with exercises that we are doing demonstrations of and posting on our

YouTube channel, which is accessible through our website.

MODERATOR

Are there any signs and symptoms that I should be watching for that indicate inappropriate time to use movement or stop movement?

DOCTOR

- If you are causing pain, we do not like that.
- If you are doing some exercises, there can be a little bit of pain as when you exercise and you haven't done that in a while but if there's constant pain, that should be checked out.
- Some people will develop what we call dystonia where you have a limb in a certain fixed position and that can be painful.
- In that case, you can talk to your doctor because sometimes we can do Botox to relieve some of the contraction of the muscle.
- Trying as much as possible to keep the limb from getting into a position where even any slight movement causes pain is something we should be aiming to do before a person gets to that point.

Theme 3: **Other PSP, CBD & MSA Symptoms**

MODERATOR

Is heavy breathing part of PSP when doing simple tasks like doing the dishes or cutting a piece of bread?

DOCTOR

- Heavy breathing is not usually part of these illnesses.
- Before I would accept that as part of PSP, I would want to make sure that there is no lung pathology first. You would want to make sure the lungs are okay. The heart is okay. But I would not start off with thinking this is related to PSP.

MODERATOR

What kind of swallowing problems are caused by PSP and how is it best dealt with?

DOCTOR

- PSP is often associated with swallowing problems, especially as the disease progresses
- Think about what affects the other muscles. That rigidity and stiffness that you see in the neck and in the trunk and in the limbs, it is also happening in the muscles of swallowing
- It is very common for people with PSP or CBD to have swallowing problems
- We always ask, if there is any coughing with eating or drinking, if there is any regurgitation and something is coming out of the nose. And then if the person says yes, and if it is happening consistently, we send people for a swallowing assessment.
- A speech language pathologist can come to your home and assess the swallowing. And if they feel like you need a more invasive assessment, they will refer them for a test where they have to drink something and they take X-rays at the same time
- Swallowing can be assessed, and it is often assessed in PSP.

MODERATOR

Is there a consistency or a type of food that is most difficult to manage at different stage of the illness?

DOCTOR

- Yes, it is very variable. Some people have trouble swallowing liquids/fluids and they have no problems with most foods. I would say most people have to cut up their food pretty small. Most people cannot eat big chunks of meat and things like that and be okay.
- As the disease progresses, some people have no trouble if the food is cut up, but if they drink something they are really struggling. In that case, we do an assessment, and we will say, thicken your fluids.
- So, you buy this stuff, and you make your water less liquid and it looks more like those Yoplait drinks.
- Other people really have trouble more with solids. Even if they cut things up small, and are diligent about chewing, they are still having a lot of trouble. Those people we will switch to a minced diet initially, and then if they continue to struggle, put them on a pureed diet.
- Sometimes it is just a matter of not mixing the solids and liquids at the same time
- Swallowing is one of the impairments that progresses and so often people do lose the ability to swallow.
- What do you do if that happens? If they cannot eat, they cannot drink, what are we going to do? One of the solutions people talk about, is a feeding tube. A tube that goes directly to the stomach
- But we always have this discussion on what stage this is happening. Because even though you can get rid of all the food going in, and all the liquid going in, you are not going to get rid of people's saliva.
- Eventually people are choking on their saliva, and what are we going to do about that? And so, you know, I think it is really important to have discussions about goals of care and what are we looking to do?
- Swallowing is a big issue. And sometimes people are at high risk of aspirating, which gives people pneumonia, which is, you know, the way people die from these diseases.
- Some people really like to have food in their mouths. And so, you think, well, that's for quality of life.

- It is important to recognize that the swallowing difficulties that happen with PSP are really common, and they can happen at different stages of disease.
- They are usually towards the later stages and I think it is important to have conversations about goals of care around that.
- Although it might stretch life out a little bit longer, it is not going to solve the problem because eventually you cannot swallow your saliva.

UNKNOWN SPEAKER

Does the tongue eventually get affected as well? And could that cause choking or gagging at night?

DOCTOR

- the tongue is a muscle and eventually it does get stiffer and moves much more slowly but it is not the cause of the choking
- the choking is really related to the liquids and solids not going down the pipe properly
- when saliva pools because you do not swallow as much, it becomes thicker, and that does not go down so easily
- Over time there is some stiffness that can, worsen and there will be more choking when that happens
- It's always important to rule out an infection when someone is coughing but if it happens only with food then it is likely a swallowing problem
- once you rule out infection, even people who have tubes and are sitting up right, eventually they have trouble swallowing their saliva

UNKNOWN SPEAKER

Can the introduction of more nutritious pureed foods into the feeding tube help to combat a drop in hemoglobin?

DOCTOR

- It is not food that is going to make your hemoglobin go from 100 to 70. There might be a bleed somewhere. It is unusual just with food.
- I do not think giving pureed food is going to make a difference. This is related to bleeding. This is not related to him not having enough iron or b12.

MODERATOR

How can you slow the progression?

DOCTOR

- So, unfortunately, you cannot slow the progression. That's the nature of this illness because swallowing is a coordinated muscle activity and these illnesses affect that.
- In some people it happens early in other people, it happens late. But it happens in most people with these illnesses.

UNKNOWN SPEAKER

Is there any exercise, to practice or to exercise the muscle to prevent the progression from speeding up? Like eating more chewy foods that exercise the jaw.

DOCTOR

- More for speech language pathologists because I personally cannot think of anything that you could do.
- I do not think exercising those muscles by eating more helps
- People who are eating more should have preserved swallowing, and we do not see that.
- Ask the speech language pathologists, maybe there is some tongue exercises to help.

UNKNOWN SPEAKER

Maybe chewing gum may help?

DOCTOR

- Chewing gum will just make more saliva so we do not actually want them to chew gum.

UNKNOWN SPEAKER

Maybe vocal exercises like singing?

DOCTOR

- That would help your vocal cords, but I do not know what it would do because they are not exactly the same muscles. And they work in different ways.

MODERATOR

We are also posting recipes by a dysphasia expert, we have a dietitian, who is a scientific advisor now, we are posting recipes that are easy to swallow on YouTube for people to see on this channel as well.

MODERATOR

Next question is if there are any tests, we can do to determine progression of disease or the rate at which it is progressing?

DOCTOR

- We follow people longitudinally (over time) and a lot of the testing that we do, whether it is a neurological test, or a questionnaire are really to look at the rate of progression. So, yes, we can follow people.
- We do MRIs also to look at the brain as it degenerates (shrinks) over time. So, there are their tests
- They are not testing like doing a blood test and checking how your thyroid function is doing. We do not have those kinds of markers yet

UNKNOWN SPEAKER

Is there stuff physically we can be doing to test for rigidity and muscle function to see how they are progressing?

DOCTOR

- The neurological test that we do is really about that. We check for rigidity, we check for slowness, we check coordination, imbalance, eye movements, muscle strength, so we check all those things. And yes, we grade them.
- If you do a test at home like a bicep curl and if you do it once a week for 50 weeks, you are going to see if that person change. So, in that way, yes, you can do it as part of a fun movement routine.

MODERATOR

Are extreme shivering and cold sweats combined with high blood pressure related to MSA? Are increased stiffness and slowing of movement following such episodes related?

DOCTOR

- In MSA, there is an extreme drop in blood pressure that can occur
- When we give some of the medications, we try to counteract that it can actually cause hypertension.
- Sweats and chattering can happen when your blood pressure drops

- What will happen is there will be a reflexive high blood pressure right after that and so, these episodes do go with MSA.

Theme 4: PSP Patient Tools & Treatment

MODERATOR

Are there any negative effects of people with PSP taking melatonin or CBD oil?

DOCTOR

- Melatonin is benign. Melatonin exists already in our brain and is important for setting your circadian rhythm.
- Often, we give melatonin to people, especially as a first line for people who have trouble getting to sleep.
- Everybody can have side effects to vitamins and minerals. So obviously everybody is different. We want to take note of that, but it is usually quite well tolerated.

DOCTOR

- The problem with CBD oil is that there are no good studies
- We do know that CBD with the THC can be associated with psychosis and depression and does affect cognitive function and is associated with cognitive impairment.
- They did a study on people with multiple sclerosis and showed that it was quite good for pain, except that it cost them some cognitive points. So, it is a cost benefit ratio
- Some people have terrible pain, MS has a lot of spasticity. CBD can help with that. So, I think that the issue right now is we need to have good studies to show where and when to use it.

MODERATOR

Are there any clinical trials Canadian patients can participate in? Are any trials showing promise?

DOCTOR

- There are two drug trials that were done in PSP in the last few years but both of them failed.
- There is one drug trial, which will hopefully start in the next year for PSP here. It will be aimed towards tau.
- The main focus is trying to get tau out of the brain.

Theme 5: Healthcare Resources

MODERATOR

Moving on to the healthcare resources theme.

MODERATOR

Can you speak to the fragmented nature of the healthcare system? There does not seem to be unified and coordinated programming for allied healthcare workers to help those affected by these rare illnesses. There also seems to be a surprising lack of knowledge about PSP and related illnesses.

DOCTOR

- I think what you are experiencing is exactly what a lot of people are experiencing
- Our healthcare system does not have very much room for Allied services (PT, occupational therapy, speech therapy)
- We would love to be able to offer these services, they do not really exist on an outpatient clinic basis at our hospital and as part of our center.
- The issue is that all these allied health services are integral to patient care. So we do try advocate for this
- For years, my memory clinic has advocated for OT and PT
- We are lucky we have a social worker and a nurse because a lot of times people cannot even get that
- It is unfortunate that they are not all OHIP covered services. So, you must pay for them out of pocket and not everybody can afford that, or has an insurance plan for that
- You are in good company with other people
- In terms of educating people, when my patients go to a long-term care facility, or they get some service that do not know about PSP or one of these diseases we do educate them
- On my website I have some information pamphlets that I give to the patients to bring to the physiotherapist

MODERATOR

What can patients and or caregivers do to ensure earlier diagnoses?

DOCTOR

- I think part of the problem is that the eye sees what the mind knows. So unfortunately, less people know about PSP. So, it does not actually even come up in the differential as a potential cause.
- Secondly, we do not have a good test. So, it is not commonly thought of
- Some rare illnesses have easy tests you can take to rule them out, we do not have that for PSP.
- We do recognize now that PSP does not only present with falls, and that PSP can present with other symptoms. It can present with cognitive changes, it can present with behavioral changes, with language changes.
- The new criteria will hopefully cause less delay in the diagnosis

MODERATOR

Finding ways to reduce wait times to see Neurologists.

DOCTOR

- There are very few cognitive neurologists. In movement disorders, there are more but not that many.
- I support primary memory care clinics across the province, there is about 140 of them and those people will send me consults and I coordinate with neurologists across the province.
- It still has to get from the family doctor to one of these primary memory care clinics and to us so people have to advocate, sometimes you're the one to have to bring it up to the family physician

Theme 6: Later Stages of PSP, CBD & MSA

MODERATOR

Keeping in mind every situation can be different. Could you talk a bit about what the final stage looks like?

DOCTOR

- These are very heterogeneous diseases and so not everybody progresses in the same way
- The things we look for in the later stages of the disease is the swallowing part which we have already discussed where a lot of people will develop swallowing difficulties.
- The other one is the immobility. So, in the later stages of the disease people become wheelchair-bound
- Sometimes it does happen where people are having falls and it is an earlier stage, and their cognition is perfectly preserved, and they have no swallowing problems.
- There is heterogeneity but as people have swallowing problems and are confined to a wheelchair, we say that is the final stage.
- If they start to have pneumonias, then that is what people die from because you cannot treat them over and over again and we think that is the final phase.
- When people have a lot of immobility where they develop posturing where their arms might be stuck in some position then that lends itself to sores and to infection. That too, is an advanced stage.

MODERATOR

When do you know it is time for MAID? (Medical Assistance In Dying)

DOCTOR

- That is a very personal choice and people choose that at different stages
- You cannot get MAID if you are demented so people are conscious of that and keep that in mind.
- I refer people to MAID when they ask me to, I do not question their motivation for it. All I must do is make sure that they are not depressed and that they are not demented.

UNKNOWN SPEAKER

Is MAID still an option if you are non-verbal.

DOCTOR

- No, not right now. If they are not able to communicate their wishes, then no.

MODERATOR

How much time is left when non-verbal and immobile, making reflexive movements unreliably and tiring quickly, cannot open eyes voluntarily and using a feeding tube due to serious choking.

DOCTOR

- We cannot know for sure. When people get to this advanced stage, we usually say that they die within a year.
- Of course some people are more resilient and can last longer. This can be a problem if referred for palliative care and we have said likely less than 1 year. We cannot always tell but in general, we usually say about a year.

MODERATOR

When do you know to admit to long term care?

DOCTOR

- If a patient is not safe at home, they should be admitted to long-term care.
- It used to be that if you were in crisis, you would get a bed within a week but there is such a shortage that that even crisis patients have to wait
- We work very hard to try to foresee that somebody is going into crisis
- I encourage people to get on several lists and pick places with reasonable waiting time. There is some that seem great but there is a seven year wait and that does not make sense for an illness like PSP.
- You want to be reasonable and pick a nice place, but also one that has a timeline that makes sense for this disease.
- This is not a disease like Parkinson's or Alzheimer's or diseases of 20 to 30 years. This is not the same type of illness.

MODERATOR

I would like to understand What to expect with respect to the progression of MSA under the following conditions?

- 82 years old
- Frequent falls and stiffness in
- Orthostatic hypotension for almost 15 years
- Lost the ability to walk or stand
- Can only sit for a very short time
- Having difficulty with speech and swallowing
- Blood pressure can increase as high as 200
- Blood pressure can drop to 80 or below

DOCTOR

- I would describe this as the final stage of this illness. In that case, we usually say maybe a year or two depending,
- The main issue is how bad is the swallowing? Are they choking every day and choking on their saliva?
- How frequent are falls knowing that frequent is different for people
- Some can fall 5 times per month and others 5 time in one day.
- It sounds like end stages. I would say less than a year

MODERATOR

Can you tell us what kind of cognitive declines are progressive with CBD?

DOCTOR

- CBD affects different networks. Some people with CBD will have a profound language problem and cannot speak. They know what they want to say but they have trouble putting all the letters and sounds together to get out the right word.
- Sometimes people have trouble understanding with CBD and they can speak very well, but they do not understand very much over time because of that language network being affected,
- They also lose their ability to spell and to write words. And then you think it is just a language network that is affected. But over time, the other networks also get affected
- Their memory might start to decline and their visual spatial processing. So, knowing how to

navigate to get somewhere or how to reach for something can be lost in CBD

- Some people will develop apraxia. Sometimes it is just speech where you cannot organize all the sounds to get the word out
- Sometimes it is movement apraxia so you want to brush your teeth and you must pick up a toothbrush, bring it to your mouth, and you have to make this kind of movement. It looks so easy for us but if you have CBD, that motor programming does not work.
- CBD has a lot of that inability to organize things in the brain properly and causes an inability to effectively communicate what you want the limb to do.
- In terms of the cognitive it can be quite heterogeneous. It could be language, it could be what we call executive like organizing, being able to pay attention to things. It could be memory, just regular verbal memory, visual memory, and it could be visual processing. It is a very heterogeneous disease and so people present with different patterns.
- It is one of the reasons it is hard to diagnose, and often gets diagnosed fairly late.

END