

# Time to Say Good-Bye to "OPCA"?

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## Summary

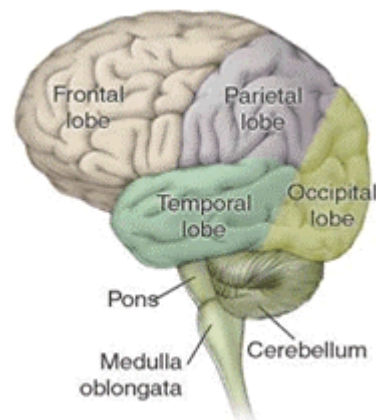
**This short article has been written to try and clarify the numerous acronyms surrounding the various ataxias. There is much confusion among those afflicted and their care-givers. The article suggests that the term "OPCA" be replaced by MSA and SCA. The article concludes by suggesting we all share our lived experiences with one another and not get wrapped up in diagnostic titles.**

When OPCA first came into my life in 1998 I was told by my husband, Greg, that his disease was either called OPCA (olivopontocerebellar atrophy) or it could be called OPCD (olivopontocerebellar degeneration). At the time, there was practically no information available online for the disease. Over the past seven years it seems that there is more online buzz about the disease each year. Many online journals such as eMedicine, MedHelp, HealthGuide, MedFriendly, and Answers.com have cropped up providing users with a plethora of definitions for the disease. The internet is wonderful, providing information to users in seconds about symptoms, treatments, history, and diagnosis. However, it has also created much confusion about what the disease should be called among patients, doctors and care-givers. It is my hope that this article will help organize the information for you.

Not only is OPCA a group of diseases all which differ in slight ways, they are hard to diagnose. Often people who experience symptoms are given numerous diagnoses before a neurologist rests on one diagnosis. If "patients" (I use the term as "those afflicted with ataxia symptoms") are lucky enough to receive a specific diagnosis: "You have SCA-1" then the "patient" is often left to figure out what the really means on their own. It is more likely they are given a general diagnosis: "You have cerebellar ataxia." "Patients" and care-givers are left to discover what this means on their own in many cases. We have visitors to our discussion board every day that find themselves confused by the terminology and titles used.

This article is published with the support and contributions of the "OPCA Awareness Support Forum." We invite you to join us online in creating awareness about these groups of diseases. Our website is:

<http://www.alyshia.com/opca>





### Why is it so confusing?

It's probably so confusing for many reasons. These are rare diseases. (Friedreich's ataxia affects 2 out every 100,000 people.) With fewer cases there is less research done on these diseases. Research could be very scattered and with different research happening in different spots all over the globe there could be little consistency between what terms are used. Also, with the SCA#'s, it seems common practice for Dr.'s to just create a new number when they're doing research on a string of families that don't fit within the other already defined SCA's. This creates more confusion among what these diseases are called and how they are categorized. I hope this article will dispel some of the confusion for you.

### Acronyms

**OPCA:** olivopontocerebellar atrophy

**OPCD:** olivopontocerebellar degeneration

**SCA:** spinocerebellar ataxia

**MSA:** multiple systems atrophy

**FA:** Friedreich's ataxia

### Are you drunk?

Often confused with being drunk, people who are afflicted with cerebellar ataxias have a wide gait and walk as though they appear drunk. This leaves them having to "explain" why they appear drunk. Explaining this disease is a very challenging thing to do.



## Ataxia

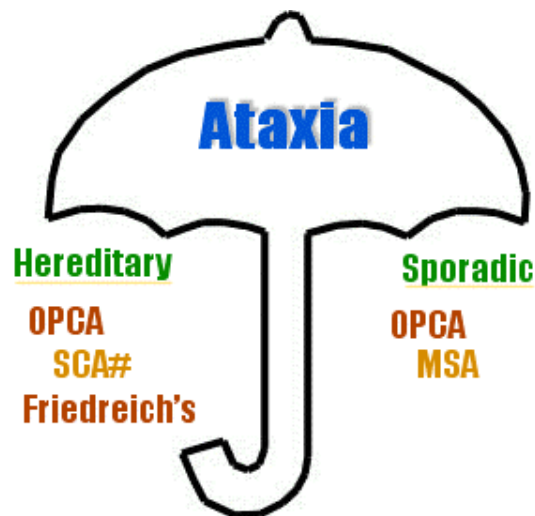
Let's start with Ataxia. It is the over-arching diagnosis if you will. Think of Ataxia as the umbrella.

Ataxia is an inability to coordinate muscle activity during voluntary movement, so that smooth movements occur. Most often due to disorders of the cerebellum or the posterior columns of the spinal cord; may involve the limbs, head, or body.

"The ataxias" are rare neurological disorders that can affect anyone of any age. They can destroy balance and co-ordination, often limiting speech, hearing and even sight.

Therefore, falling within the ataxia spectrum are the following diseases: OPCA, SCA, MSA, Friedreich's, etc.. If you've been told you have one of these then you all have an ataxia. Ataxia is the main symptom of these diseases that are sometimes referred to as "cerebellar ataxias". Ataxia can also be a symptom of multiple sclerosis or cerebral palsy.

From here, it is best to determine whether you have hereditary or sporadic ataxia.



### Hereditary Ataxias

Think of hereditary ataxias as one side under the umbrella. The hereditary ataxias are genetic. Meaning that something inside some gene "mal-functioned." The hereditary ataxias can be divided into those that are dominantly inherited and those that are recessively inherited. I won't get into this because the ins-and-outs of genes is not my purpose here. In short, all Spinocerebellar ataxias (SCA#) are dominant genes while Friedreich's ataxia (FA) is from a recessive gene. Friedreich's ataxia is the most inherited ataxia.

The SCA#'s are hereditary forms of OPCA. **This means that OPCA is the "parent" disease while the SCA# defines the type of OPCA a "patient" has.**

This means that if you have been diagnosed with a specific SCA# then your specific strand of OPCA has been determined. Lucky you! Some of us are not even sure which SCA# we have and therefore, we're stuck with using the parent diagnosis: OPCA, until more SCA#'s are defined (that is to say, until the specific genes for the other SCA#'s are discovered). The prevalence of SCA's is estimated to be about 1 to 4 people per 100,000.

### Sporadic Ataxias

Think of the sporadic ataxias as falling on the other side of the ataxia umbrella.

This is where the confusion can easily begin. On the sporadic ataxia side of the umbrella we find OPCA again. Well how can this be on both sides: both hereditary and sporadic at the same time?

Ataxias are sporadic when there is no known family history of this disease. This type of ataxia can be very difficult to diagnose. A number of acquired and hereditary causes of ataxia must first be ruled out before a diagnosis of sporadic ataxia can be made.

**MSA**

I think it is important to know that MSA (Multiple Systems Atrophy) is a disease which often follows a diagnosis of sporadic OPCA. MSA can develop from three diseases. The term MSA encompasses striatonigral degeneration (SND), the Shy-Drager syndrome (SDS), and many cases of sporadic olivopontocerebellar atrophy (sOPCA). Between 25,000 and 100,000 Americans have multiple system atrophy. However, many will not receive the correct diagnosis during their lifetime.

It is not possible for someone with hereditary SCA (or "OPCA" if you like) to develop MSA.

### Erasing the Confusion

Since OPCA may exist as a sporadic **or** hereditary disease, categorizing sporadic OPCA as MSA and inherited OPCA as SCA may be appropriate. Differences between sporadic and inherited OPCA in microscopic pathology support this division.

**This means the term "OPCA" *should* be changed to avoid the confusion.**

We should be calling "OPCA which is sporadic" MSA  
 We should be calling "OPCA which is hereditary" SCA



But what challenges is there for re-naming an entire disease? Is this feasible? Does it make sense? Is it necessary?

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| <p><b>Sporadic Ataxias</b></p> <ul style="list-style-type: none"> <li>• Marie's ataxia</li> <li>• Holmes ataxia</li> <li>• Menzel's ataxia</li> <li>• ataxia with parkinsonism</li> <li>• autonomic neuropathy</li> <li>• corticospinal features</li> <li>• dementia spastic ataxia</li> <li>• episodic ataxia syndrome, called EA1 and EA2</li> <li>• Multisystem atrophy</li> </ul> <p><b>Hereditary Ataxias</b></p> <ul style="list-style-type: none"> <li>• SCA #</li> <li>• "slow-eye movement" ataxia</li> <li>• Machado-Joseph disease</li> <li>• "pure" cerebellar ataxia</li> <li>• OPCA #</li> <li>• ataxia with ophthalmoplegia</li> <li>• ataxia with retinopathy</li> <li>• Marie's ataxia</li> <li>• Holmes ataxia</li> <li>• Menzel's ataxia</li> <li>• spinopontine atrophy</li> </ul> |
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## Problems

I see that 'doing away' with the term "OPCA" leaves three problems:

1. "Patients" can have sporadic OPCA that has not yet manifested into MSA. So it's pre-mature to call their disease MSA if it doesn't have the "autonomic failure" which is prevalent in MSA cases.
2. "Patients" that do not have their specific type of SCA defined will be left with using just "SCA."
3. What about all those internet websites that are still calling this disease OPCA? Even if doctors, "patients," and care-givers discard the term OPCA for MSA and SCA, there are still a multitude of internet resources, research papers, and medical journals using the term OPCA.

## Let's Be Realistic

We need to be realistic, we are not going to be changing the name of this disease. We are not going to be saying "Good-Bye" to OPCA and "Hello" to MSA and SCA. It's not the name or title that matters. Whether you call your disease OPCA, MSA, SCA or whatever, the symptoms are similar (but far from the same), our challenges and plights are comparable. As an "ataxia community" we need to come together and rather than be hung-up on the titles of our diseases we need to share our lived experiences with each other and provide support for everyone living an ataxia life.

I hope this article has dispelled some of the confusion for you about the titles and names given to the face of your disease. Remember, it's not what you've been diagnosed with that matters, it's what you do with and how you react to that diagnosis.

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## Sources

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<http://www.ataxia.org/generations/2002/spring02/koeppen.html>

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- From our Own Website  
<http://www.alyshia.com/opca/hereditary.html>  
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<http://www.dizziness-and-balance.com/disorders/central/movement/msa.html>



Disclaimer: The information in this article is purely based on my own understanding from the research I have done on these diseases. The information should be understood to be my personal opinion and not fact.