

# What is Opsoclonus Myoclonus Syndrome (OMS) /Opsoclonus Myoclonus Ataxia (OMA)?

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**Opsoclonus Myoclonus Syndrome (OMS)** also known as **Opsoclonus Myoclonus Ataxia (OMA)** is an extremely rare neurological disease which appears to be the breakdown of an autoimmune process involving the nervous system . While OMS can occur at any age, most cases involve children under the age of three years old. Most OMS children have a normal birth and development until age 12-36 months (average). At that point, something triggers the immune system into overdrive and it can't shut itself down. About half of all OMS cases occur in association with neuroblastoma (a cancer of the sympathetic nervous system usually occurring in infants and children). When the immune system cannot find a neuroblastoma, it begins attacking the base of the brain.

- \* Gross and fine motor functions affected
- \* Speech impairment occurs
- \* Vision (“dancing eyes”) problems are exhibited
- \* Severe ataxia (balance) problems arise

"Because this rare movement disorder is so unique, some neurologists may never see a case in their entire career," Dr. Pranzatelli – [www.omsusa.org](http://www.omsusa.org) . This disease affects roughly one child in 10 million annually, or about 60 people per year worldwide.

## **Signs and symptoms**

OMS is commonly mistaken for other illnesses such as Cerebellar Ataxia or inner ear infections. Prompt diagnosis often determines the severity of the long term impact on the child. Symptoms of OMS include:

- \*[Opsoclonus](rapid, involuntary, multivectorial (horizontal and vertical), unpredictable, conjugate fast eye movements without intersaccadic quick rotation of the eyes intervals)
- \*[Myoclonus] (brief, involuntary twitching of a muscle or a group of muscles) \*Ataxia both truncal and appendicular \*Dysphasia (a language disorder in which there is an impairment of speech and of comprehension of speech, caused by brain damage)
- \*[Mutism] (a language disorder in which a person does not speak despite evidence of speech ability in the past, often part of a larger neurological or psychiatric disorder) \*Lethargy \*Irritation or malaise \*Drooling \*Strabismus (a condition in which the eyes are not properly aligned with each other) \*Vomiting
- \*Sleep disturbances.

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\*Loss of balance, inability to walk

\*Loss of fine motor skills

A child afflicted with OMS can quickly regress from healthy and active to a child who constantly cries, throws tantrums, and loses all sense of balance and fine motor skill. This regression happens seemingly in a matter of a few days or weeks.

## **Cause**

About half of all cases are associated with neuroblastoma cancer, and most of the others are suspected to be associated with a low-grade neuroblastoma that spontaneously regressed before detection. It is hypothesized that a virus infection causes the remaining cases. OMS is not generally considered an infectious disease. OMS is not passed on genetically.

## **Treatment of OMS**

There is no known definitive cure for OMS. However, several drugs have proven to be effective in its treatment. Some of the medications used to treat the symptoms are: \*Corticosteroids Used at high dosages, this treatment can accelerate regression of symptoms. Subsequent very gradual tapering generally follows. Most patients require high doses daily or every other day for months to years before tapering. These medications go by the name ACTH and Prednisone, to name a few. \*Intravenous Immunoglobulin (IVIG) These infusions are given on average every 4-8 weeks for years. Most OMS children are required to have an outpatient hospital visit for these treatments.

\*Chemotherapy treatments include Cytosin and biotherapy treatments include Rituxin at various levels and for various lengths of time.

\*Other medications are used to treat symptoms without influencing the nature of the disease (symptomatic treatment. These include:

\*[Trazodone] can be useful against irritability and sleep problems \*Plasmapheresis ("washing the blood", showing similarities to dialysis for severe, steroid-unresponsive relapse .

Treatment of OMS should not be delayed. As soon as OMS is diagnosed, immunotherapy should be given, even before tumor removal. Surgical resection of the tumor may not help the symptoms of OMS; other treatment is usually necessary. Because chemotherapy is also immunotherapy, it

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should work to treat the tumor and the OMS. While “the cure” remains elusive, a handful of doctors are researching OMS.

Often, our OMS children are part of their research and their experiments in order to progress a cure. This often requires traveling great distances to meet with one of these specialists. They will often require spinal taps to determine the B cell and T cell levels of the child. Also, CT scans, MIBG scans, and other testing is normally a semi-annual or annual event for these kids. Once the specialist sets the treatment protocol for the child, they work with the local neurologist and/or pediatrician. At times, an OMS child may be working with a team of doctors including pediatricians, neurologists, OMS specialists, oncologists, endocrinologists, and others. When there is not agreement on treatment, parents need to resolve the conflict and the child normally suffers during the wait.

## **What is the Prognosis?**

Remission may be more common in viral or idiopathic cases than in children with neuroblastoma. Relapses frequently occur with minor illnesses; viral infections may play a role in the reactivation of disease in patients that are exposed. Children with severe opsoclonus-myoclonus at the start have the highest risk of permanent neurologic problems. Other problems that may arise are learning and behavior problems, such as attention-deficit hyperactivity disorder, conduct disorder, and obsessive-compulsive disorder, which often require treatment with medications specific for those problems. These problems emphasize the need for early diagnosis and treatment.

## **Relapses**

Neurological symptoms may return during illness, fever, stress, sedatives or anesthesia, tapering or discontinuation of immunotherapy, and after immunizations. Because of the

susceptibility to relapses, it is imperative to keep OMS children away from sick individuals. What is a common cold or a flu bug can cause a relapse in an OMS child and send them back to the hospital. Such relapses are common and usually occur more than once. Rarely, many years may elapse between episodes. Relapses should be treated. Usually children who responded initially to immunotherapy will do so again, even to a single agent.

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## **Treatment Complications**

As much as we all would like to eradicate opsoclonus-myoclonus, overly aggressive chemotherapy or immunotherapy can devastate the immune system, causing life-long after-effects. Infections in children receiving immunosuppressive drugs must be taken seriously and receive medical attention early.

## **Common myths about OMS children**

OMS children have special needs that will ultimately help them be successful citizens in the future. These children do not necessarily need dependent care for the rest of their lives, but they do need some help to achieve these goals. Some common assumptions about OMS kids that friends, family, and school administrators make include:

- MYTH:** These kids will always be behind and require dependent care.  
**FACT:** With proper treatment and focused care OMS children can lead a life independent of facilitated care. OMS children have grown up and gone to college, gotten married, and raised their own children. They need a hand, not to be looked down upon.
- MYTH:** Special Education for all activities is required.  
**FACT:** While OMS children may have certain needs, careful attention needs to be made to ensure a child is not labeled as “Special Education” for all classes. Often, the kids are just needing special accommodations for certain classes; such as one on one facilitation for certain subjects.
- MYTH:** Speech development is another indicator that OMS children will be behind.  
**FACT:** While OMS children may have certain needs, many OMS children can achieve scholastic milestones when given the opportunity. Delays in speech development should be seen as a teaching opportunity to assist the child in achieving milestones, not as a predetermined sentence on their future.
- MYTH:** Attendance issues are because parents are overprotective regarding their OMS child’s illnesses.  
**FACT:** The immune system of OMS children is greatly compromised. Parents need to be mindful that when illnesses are rampant in the classroom, it can cause great harm to their children. A minor illness passed to an OMS child can begin a relapse with potential to hospitalize the child.

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**MYTH:** Parents allow misbehavior in their children. Their poor behaviors, mood swings, non-responsiveness, and OCD tendencies are because parents enable these behaviors.

**FACT :** OMS children routinely exhibit behavior issues and have bad moods. This disease attacks the brain and creates issues which do not allow children to function normally at times. Their behavior issues and bad moods are not “learned” issues, but rather outward evidence of the damage that has been done to that part of the brain. These bad behaviors may come on suddenly or the child may be aware it is coming on. OMS children at times may also go into a non-responsive state and give the appearances of being rude and disrespectful. In reality, the child is trying to process the instructions or the present situation. Processing certain instructions such as directions somewhere, may seem simple to many, but create tremendous confusion for an OMS child.

**MYTH:** Parents are mistreating their children by not immunizing their OMS child.

**FACT:** While not proven, there are strong links between OMS children and immunizations given to young children. It is a strong enough case that the pharmaceutical industry has now created a fund which provides compensation for certain OMS cases filed within 2 years of diagnosis.

**MYTH:** OMS children don't fit in well at school.

**FACT:** OMS children have not always had an easy childhood so far. They have spent a lot of time in hospitals and doctors' offices and may be apprehensive about those visits. They often are secluded from the public for months and years and may be behind in social skills. They don't always get an opportunity to be in large groups and that may scare them.

## **How teachers and administrators can help**

The teachers, counselors, and school administrators for these OMS children play a vital role in their development. There are some straightforward steps that can be taken to ensure a successful school year:

- \* Educate yourself on OMS.
- \* Encourage the use of hand gel in the classroom, hand washing in the rest rooms, and proper coughing and sneezing etiquette to avoid the passing of germs.
- \* Just as schools promote the diversity of children from different cultures and beliefs, introduce and promote the diversity of children with special needs and disabilities.

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- \* OMS kids and their parents just want a normal life for the child. Classmates need to be encouraged to openly interact with OMS kids and engage them in activities.
- \* OMS kids may have speech delays, but that does not mean they cannot master activities in school. Assist the child in their speech development and encourage school officials to find additional resources to enable the child's speech to develop.
- \* Develop an action plan with the parents. Unlike mainstream children that may get a development plan when problems arise, it is necessary to develop expectations up front. Also, develop a regular review of that plan.
- \* Identify the child's strengths and weaknesses; both intellectually, emotionally, and physically. This will allow everyone to agree on a course of action. For example, teachers may find that OMS children are at level or above level in reading, but behind in speech. They may also find they have difficulty in physical education activities.
- \* Identify areas where group participation is beneficial and where one on one attention is required. OMS kids do not necessarily need individual attention in everything. Likewise, putting them in group activities where they are clearly struggling serves no benefit for anyone.
- \* Be aware of illness in the school and classroom. A minor bug can be devastating to these kids.
- \* Push your school to be ingenious in their approach for educating this child with some special needs. Some schools utilize Skype during times when OMS children must be away from school. Also, there are many applications on iPad, iPhone, and so on that can help OMS children develop.
- \* Despite so many setbacks, OMS kids are some of the happiest children. By providing some individual attention, these kids will respond positively and can go far!

## **Reference materials for more information**

- \* Wikipedia – Opsoclonus Myoclonus Syndrome
- \* Dr Pranzatelli – [www.omsusa.org](http://www.omsusa.org)
- \* OMS Parent Web Site – [www.omslife.org](http://www.omslife.org)
- \* Facebook groups
- \* OMSLIFE - Opsoclonus Myoclonus Syndrome Support Group,  
Opsoclonus Myoclonus Syndrome Support,  
OMS Families – Opsoclonus Myoclonus,  
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**Some specific details about *fill in child's name here* that will help *fill in child's name here* and the school succeed:**

**What *fill in child's name here* is most interested in (what gets her juices going):**

- Zzzzzz
- Zzzzz
- Zzzzz

**What *fill in child's name here* NEEDS EXTREME PATIENCE with:**

- Zzzzzz
- Zzzzz
- zzzzz

**What LEARNING STYLE works best for *fill in child's name here*:**

- Zzzzzz
- Zzzzz
- zzzzz