Doose Syndrome (MAE Myoclonic Astatic Epilepsy)

What is Doose Syndrome?

Doose Syndrome is an epilepsy syndrome of childhood, that is often resistant to medication and for this reason it is typically difficult to treat. It is usually characterised by generalised seizures, which may vary in type and frequency; many children can experience large numbers of seizures daily, part of what makes this condition so difficult to manage.

Onset generally occurs between ages one and five, usually in children with an uneventful history. In some cases, there is a positive family history of seizures, and family studies over the years have supported a genetic basis.

As with most medical disorders, the spectrum of severity seen in Doose Syndrome ranges from mild to those more severely affected. Children mildly affected by Doose Syndrome may have their seizures quickly and easily controlled with first-line medications, alone or in combinations.

Those children on the more severe end of the Doose Syndrome spectrum may have difficulty finding an effective medication or treatment.

How common is it?

This type of epilepsy is uncommon and happens in one to two out of 100 children with epilepsy. It is more common in boys than girls. In about a third of cases some other family member also has epilepsy though not always the same sort.

Epilepsy and Doose Syndrome

- The seizures, in Doose Syndrome, can be very different, consisting of jerks, sudden falls to the ground, or sometimes a jerk followed by a fall. Absences can happen when consciousness is lost briefly, and in some cases there may be major seizures with stiffness and jerking all over (generalised Tonic-Clonic seizures).
- Approximately one-third of people with Doose Syndrome will have a very long seizure at some time during their childhood or adolescence.
- The treatment of Doose Syndrome can be difficult. The medications which have been most successful
- are sodium valproate (Epilim), lamotrigine (Lamictal) and occasionally clonazepam, clobazam or nitrazepam.

- Certain medications should be avoided, including Carbamazepine and Vigabatrin.

- Doose syndrome is a condition which may respond to the ketogenic diet or the Modified Atkins Diets, special diets used in difficult to control seizures. For this reason, the diet is considered relatively early in the treatment course for children not responding to standard treatment measures.

- The outlook for people with Doose Syndrome is variable depending on the type and frequency of seizures as well as the individual’s response to medications.

- The spectrum ranges from complete remission and totally normal intellectual development to therapy-resistant epilepsy which can result in mild to severe developmental delay.

- Although there are advances in therapy and research in Doose Syndrome, prognosis and outcome tends to be unpredictable and varied.

- Prognosis is more likely to be unfavourable in children who have generalised tonic clonic seizures from the outset.

Information updated August 2013
To be reviewed in 2015

Although every effort has been made to ensure accurate and up to date information is provided, Epilepsy Queensland Inc and its advisors cannot accept any liability in relation to the information provided. It is strongly recommended that you discuss any information with your doctor as to whether it applies to you / your child.