



Intermediate (Type II) Spinal Muscular Atrophy

What are the Effects of Intermediate (Type II) SMA?

People with the intermediate form of SMA show generalised weakness and wasting of the muscles. Despite a set pattern of weakness, each person is different in the extent to which they are affected. These children are able to sit unsupported and some are able to crawl or stand, although these abilities may reduce with increasing body weight. There is often susceptibility to respiratory infections, but the danger from these depends on the extent to which the respiratory muscles are affected.

Although the condition does not usually get significantly worse after the original onset, bouts of infection may be associated with further apparent weakening. Scoliosis (curvature of the spine) and deformities of the hands, feet and chest may develop in childhood and joints may be affected by tendon contractures which reduce movement of the limbs. However, medical advice should always be sought since, with careful management, these effects can be minimised or prevented.

People with SMA are often of above average intelligence and can develop considerable talents and skills. With appropriate aids to mobility and help with other aspects of physical independence, even the youngest children can take an active part in family and social activities. Most children with Intermediate SMA should be capable of being educated in mainstream schools, if that is what their parents want, and will have much to contribute to others. Special equipment can be provided to enable children to develop a positive attitude to their abilities and make the most of them.

Is There Treatment For SMA?

Research into SMA is progressing, but at present there is no known cure, and no treatment which will repair the damage to the anterior horn cells, or reverse the weakening of the muscles. However, who knows what the future will bring?

Physiotherapy (stretching exercises) and orthotic appliances may improve the posture of those children with the intermediate type of SMA. Active exercises to maintain muscle strength are to be encouraged, such as supported standing and assisted walking if possible.

Respiratory exercises and other breathing techniques are important to reduce the effects of chest infections. Advice on techniques can be obtained from a physiotherapist.

What Does the Future Hold?

The Jennifer Trust for SMA and other organisations throughout the world are funding research into SMA in the UK and abroad.

From past research it is now possible for parents of affected children to have available pre-natal or post-natal testing on future children. The tests will indicate whether the child is affected, unaffected or is a carrier.

Researchers are studying affected children to pin-point the reason for degeneration of the anterior horn cells, and how this occurs. They are also investigating the anterior horn cells of unaffected people to see how damaged nerves might be repaired. It is possible that this might lead to the development of drugs that will be able to improve the regeneration of nerve cells.

Research is advancing in many areas. The organisations listed at the end of these pages can keep you up-to-date with developments.

What is the Jennifer Trust for Spinal Muscular Atrophy (JTSMA)?

The JTSMA is a support group run by parents of children with SMA and adults who have SMA. The main aim of the group is to provide support, information, understanding and friendship to those whose lives are affected by SMA. A quarterly newsletter, "Holding Hands", is produced, and an annual weekend conference is held offering the opportunity to meet each other and share experiences in a relaxed atmosphere.

The Jennifer Trust is open to anyone who comes into contact with SMA. It now has contact with several hundred families, who, between them, have a wealth of experience to share. The Jennifer Trust was founded in 1985 by Anita Macaulay, whose daughter, Jennifer, had severe SMA.

JTSMA has an Area Contact Network of people who provide local support and advice and are experienced in SMA and aware of the wider issues. The Trust will be able to give you the name address and phone number of your local contact family in the UK, who should be able to answer many of your questions, offer advice and just be there whenever you need them. Many areas also produce regional newsletters on a regular basis informing you of all the events taking place in your area. As well as the annual conference, there is a national Christmas Party, Regional Fun Days and Regional Lunches which all provide you with an excuse to get together. These are often invaluable meetings with other families who have similar problems. It can be very positive to meet other families who have had the same experience as yourselves and have come through it.

What Other Support Services can Help?

The Muscular Dystrophy Group provides some information and assistance for those affected with SMA and their families. The MD Group has a network of Family Care Officers attached to some neuromuscular clinics who give help and advice.

General Practitioners (GPs) and hospitals should also be able to offer you help, however SMA is not so common that all GPs or hospitals know much about it. They may need to refer to specialists in other hospitals. If you are not getting as much information or help as you want, keep asking.

Where Can I Get More Information?

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