

# Guide to Understanding Mucopolysaccharidosis III (MPS III) Sanfilippo Disease



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Oliver (MPS IIIC)

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## What is Sanfilippo Disease?

Sanfilippo Disease is a Mucopolysaccharide storage disorder, also known as Mucopolysaccharidosis Type III. The disease takes its name from Dr. Sanfilippo who was one of the first doctors from the United States to describe the condition in 1963.

Whilst there is no cure for individuals affected by MPS III, this factsheet explores the presentation and clinical management of the disease.

This factsheet is produced by the Society for Mucopolysaccharide Diseases (MPS Society) drawing on the experiences of parents and doctors with reference to medical literature.

## What Causes Sanfilippo Disease?

Mucopolysaccharides are long chains of sugar molecules used in the building of bones, cartilage, tendons and many other tissues in the body. “Muco” refers to the thick jelly-like consistency of the molecules, “poly” means many and “Saccharide” is a general term for the sugar part of the molecule.

An alternative word for Mucopolysaccharides is glycosaminoglycans (or GAGS) but the term Mucopolysaccharides will be used for continuity throughout this Factsheet.

In the course of normal life there is a continuous recycling process of building new Mucopolysaccharides and breaking down old ones. This process requires a series of biochemical tools called enzymes.

Individuals with MPS III are missing an enzyme which is essential in cutting up the used Mucopolysaccharides. Incompletely broken down Mucopolysaccharides remain stored inside parts of the cells called lysosomes. The lysosomes become swollen and disrupt cell functioning causing progressive damage.

Babies may show little sign of the disease but symptoms start to appear as more and more cells become damaged by the accumulation of Mucopolysaccharides.

## Are There Different Forms of Sanfilippo Disease?

To date, four different enzyme deficiencies have been found to cause MPS III and thus the condition is described as MPS III Type A, B, C or D. Type A is the most common form found in most populations.

- MPS III A is missing the enzyme heparan N sulphatase
- MPS III B is missing the enzyme alpha-N-acetylglucosaminidase
- MPS III C is missing acetyl-CoA:alpha-glucosaminide acetyltransferase
- MPS III D is missing N-acetylglucosamine-6-sulphatase

It is important to note that there are no significant, clinical, physical differences between the different subtypes of MPS III disease, although there have been cases of late onset MPS III Type B where the individuals have remained relatively unaffected into adult life. The latest understanding is that some people seem to produce some enzyme activity which helps to slow down the progression of the disease whilst those with more severe symptoms appear to have no enzyme activity at all.

## How Common is Sanfilippo Disease?

The MPS Society, which co-ordinates the Registry for Mucopolysaccharide and Related Diseases, has shown that MPS III is a rare condition affecting one in 85,000 live births. Over a ten year period between 1988 and 1998, 97 babies were born with MPS III in the United Kingdom.

## How is Sanfilippo Disease Inherited?

MPS III is an autosomal recessive disease whereby both parents must carry the same defective gene and each pass this same defective gene to their child. Where both parents are carriers of the MPS III gene there is a 25% (1:4) chance of having an affected child in each pregnancy. There is a 50% (1:2) chance of a child receiving only one copy of the defective gene and therefore being a carrier. A carrier will not be affected but can pass the defective gene to his/her offspring. The remaining 25% (1:4) will be neither affected nor a carrier. Using information from an affected individual's DNA, it may be possible to determine whether brothers and sisters are carriers of, or if they are affected by MPS III.

## Can you Test for Sanfilippo Disease in Pregnancy?

For each pregnancy the chances of a baby inheriting MPS III are totally independent of whether a previous child was affected by MPS III. Pre-natal tests can be arranged early on during a pregnancy for those families who already have a child with MPS III. Both amniocentesis and chorionic villus sampling can be used to diagnose MPS III in utero.

## Genetic Counselling

All parents of children with a lysosomal storage disease should consider asking for genetic counselling before having other children. The counsellor should be able to advise on the risk to close relatives and to suggest whether the wider family should be informed.

## Clinical Presentation of Sanfilippo Disease

### Physical Appearance

Children with MPS III grow to a fairly normal height and changes in appearance may be less than in other MPS diseases. The hair is thick and coarser than usual and their bodies may be hairier than normal. The eyebrows are often dark and bushy and may meet in the middle. Noses tend to be upturned and flat on the bridge.

### Physical Problems

Of all the MPS diseases, MPS III produces the mildest physical abnormalities. It is important, however, that simple and treatable conditions, such as ear infections, are not overlooked because behavioural problems make examination difficult. Parents may need to search until they find a doctor with the patience and interest in treating a child with a long-term condition. Do not hesitate to consult your doctor if you think your child may be in pain.

### Intellectual Ability

Whilst the majority of children affected by MPS III will lose their intellectual ability progressively through childhood, a small number may retain intellectual skills into adulthood.

## Neurological Problems

During the later stages of MPS III some children may experience seizures. Seizures are as a result in disruption in the electrical activity in the brain, which may also be referred to as epilepsy or fits.

## Eyes

Corneal clouding does not occur in the eyes as it does in other MPS diseases.

### Night blindness

Many families have reported that children with Sanfilippo Disease do not want to walk in the dark or are afraid when waking up at night. Determining the reason for this problem is difficult.

Putting a night light in a hall or bedroom may prove beneficial.

### Seizures/Epilepsy

At a later stage of the disease a number of children with MPS III will start to have frequent, minor seizures when they momentarily alter their level of consciousness (absences). This could be a stare for a few seconds, lack of response or a slight twitch.

On days when this occurs the child may seem more out of touch or harder to feed. Some may have more generalised seizures (grand mal) involving either loss of consciousness or physical jerking.

During the seizure you should place your child on his or her side to prevent the inhalation of vomit. The child should be left in that position until the seizure is over. You should check that the airway is clear but do not put anything in the child's mouth.

Both forms of seizure can be treated with medication if indicated. Sometimes this may involve a number of drugs and the child may be drowsy until they become used to the medication.

## Ears

Some degree of deafness is common in individuals suffering from all types of MPS III. It may be Conductive Deafness, Nerve Deafness or both (Mixed Deafness) and can be made worse by frequent ear infections.

It is important that individuals with MPS III have their hearing checked regularly and for problems to be treated early to improve or maintain the ability to communicate and learn.

Conductive Deafness is concerned with impaired transmission of sound waves through the ear canal, the ear drum and the middle ear. Correct functioning of the middle ear depends on the pressure behind the ear drum being the same as that in the outer ear canal and the atmosphere.

This pressure is equalised by the eustachian tube which runs from the middle ear to the back of the nose. If the tube is blocked the pressure behind the eardrum will drop, the drum will be drawn in and the transmission of sound waves will be impaired.



Amy (MPS IIIB)

If this negative pressure persists, fluid from the lining of the middle ear will build up and in time become thick like glue. Hence the condition being referred to as “glue ear”.

### Conductive Deafness (Glue ear)

Under general anaesthetic a small incision behind the eardrum can be made (myringotomy) and the fluid sucked out. A small ventilation tube called a “grommet” may then be inserted to keep the hole open and allow air to enter from the outer ear canal until the eustachian tube starts to work properly again. Grommets will eventually fall out. If the conductive deafness recurs T-tubes may be used. T-tubes are a type of grommet which stay in place longer. Due to the anaesthetic risks for individuals with MPS III, the surgeon may decide to use T-tubes on the first occasion.

### Sensorineural Deafness (Nerve Deafness)

In most cases the cause of Nerve Deafness is damage to the tiny hair cells in the inner ear. It may accompany Conductive Deafness, in which case it is referred to as “Mixed Deafness”.

Nerve Deafness is managed by the fitting of hearing aids. Children with MPS III may keep pulling out their hearing aids at first but it is important to persevere at wearing them so that communication can be maintained.

### Respiratory Infections

Medication may affect individuals with MPS differently so it is essential to consult your doctor rather than using over the counter medication. Decongestants usually contain stimulants that can raise blood pressure and narrow blood vessels, both of which are undesirable for individuals with MPS III. Cough medicines that have a sedating effect may cause more problems with sleep apnoea by depressing muscle tone and respiration. Individuals with MPS III may end up with secondary bacterial infections which should be treated with antibiotics.

### Heart

It is generally accepted that the heart is not usually affected by the disease.

### Nose & Throat

The problems described in this section are common to children with MPS diseases but occur less often in individuals with MPS III. The severity of the problem depends greatly on the individual child.

Frequent coughs, colds and throat infections are common problems. The tonsils and adenoids often become enlarged and can partly block the airway. For this reason they may be removed (See breathing difficulties).

The neck is usually short which contributes to the problems in breathing. The windpipe (trachea) becomes narrowed by storage material and is often more floppy, or softer than usual, due to abnormal cartilage rings in the trachea. Nodules or excess induration of tissue can further block the airway.

Typically, the bridge of the nose is flattened and the passage behind the nose is smaller than usual due to poor growth of the bones in the mid-face and thickening of the mucosal lining.

The combination of abnormal bones and storage of Mucopolysaccharide in the soft tissue can cause the nose to become easily blocked. Some children with MPS III can have chronic discharge of clear mucus from the nose (rhinorrhea) and sinus infections.

### Mouth & Teeth

The lips are thick, the gum ridges are broad and the tongue becomes enlarged. Teeth are widely spaced and poorly formed with fragile enamel. It is important that the teeth are well cared for as tooth decay could be a cause of pain.

### Dental Hygiene

Teeth must be well cared for to avoid the need for extractions. If the water in your area has not been treated with fluoride then your child should have fluoride tablets or drops daily. Cleaning around the mouth with a small sponge or a stick coated in mouthwash will help keep the mouth fresh and avoid bad breath.

It may be safer for any treatment to be carried out in hospital. If teeth need to be removed under anaesthetic this should be carried out in hospital under the care of an experienced anaesthetist and never in the dental surgery.

Dribbling is a common problem and a plastic backed bib under the clothes may prevent soreness.

### Breathing Difficulties

Children with MPS III may breathe very noisily, even when there is no infection. At night they may be restless and snore. Sometimes the child may stop breathing for short periods while asleep (sleep apnoea).

This noisy breathing, which stops and starts, can be very frightening for parents to hear and they may fear that their child is dying. In fact many children may breathe like this for years. Many children with MPS III have frequent colds, blocked noses and chest infections. This may be managed using C-PAP.

## Night Time C-PAP

### (Continuous Positive Airway Pressure)

A Night Time C-PAP may be recommended where a sleep study has shown that an individual is experiencing sleep apnoeas. This is where the individual suffers from low oxygen levels at night and stops breathing for short periods, leading to day time drowsiness and headaches.

C-PAP involves placing a mask or canula on the face each night and having air pumped into the airway to prevent it from collapsing. This may seem to be an extreme measure but it can greatly improve the quality of sleep and help prevent or reduce the risk of heart failure caused by low oxygen levels at night.

In severe cases of sleep apnoea with heart failure, a tracheostomy (a hole in the airway made in front of the neck) may be needed. Those who have received an early tracheostomy claim to feel much better after improving their night time breathing.

## Liver, Spleen & Abdomen

The liver and spleen may be slightly enlarged due to the storage of Mucopolysaccharides (hepatosplenomegaly). The enlarged liver does not actually cause problems or lead to liver failure but its volume can interfere with eating and breathing.

## Skin

Individuals with MPS III tend to have thickened and tough skin which lacks elasticity. Excess hair on the face and back occurs in some individuals.

## Bowel Problems

Many individuals with MPS III suffer from loose stools and diarrhoea. Occasionally it is caused by severe constipation and leakage of loose stools from behind the solid mass of faeces. However, parents often describe it as “coming straight through”. It is thought there may be a defect in the autonomic nervous system which controls those bodily functions usually beyond voluntary control. Examination by a paediatrician or physician supplemented by an X-Ray if needed will establish the cause.

This problem may disappear as the child gets older but it can be worsened by antibiotics prescribed for other problems. If there is diarrhoea (and it is not secondary to constipation) simple medication, such as loperamide (Immodium) can be very useful. A diet low in roughage may also be helpful.

Constipation may become a problem as a child gets older and becomes less active, resulting in the muscles weakening. If an increase in roughage in the diet does not help or is not possible, the doctor may prescribe laxatives or a disposable enema.

## Bones & Joints

### Bones

Individuals with MPS III tend to have minimal problems with bone formation and growth.

## Joints

Joint stiffness is common in all MPS diseases. All the joints become stiff and their movement may become limited. Later in the child's life this can cause pain which may be relieved by warmth and ordinary pain killers. The limited movement in the shoulders and arms may make dressing difficult.

## Legs and Feet

Many individuals with MPS III stand and walk with their knees and hips flexed. Combined with the tight Achilles Tendon this may cause them to walk with their toes curved under, rather like the hands.

## Hands

The fingers of children with MPS III occasionally become bent over (clawed) and the arms may not be able to extend fully. Later on there may be some limitation of movement in the large joints such as shoulders, elbows, hips or knees.

## Hips

Individuals with MPS III may suffer from dislocated hips but treatment may not be advisable or necessary.

## Hip Disease

Many children with MPS III have periods of limping or apparent pain in their hips. If an X-Ray is taken it is often thought that they have a condition called Perthes' Disease. The changes seen are in fact very common with MPS III and are probably part of the bone disease that occurs in all children with the disease.



Harrison (MPS IIIA)

## Progression of Sanfilippo Disease

The disease will affect children differently and its progress will be much faster in some cases more than others. Change will usually be very gradual and therefore easier to adjust to. There could also be periods where there is a significant amount of change all at once, followed by a period of time where the rate of these changes slows down considerably. This is referred to as a plateau stage. The disease tends to progress through the following stages:

### The First Stage

The first stage takes place during a child's pre-school years. This may be a very frustrating time for the parents. They begin to worry as their child starts to lag behind their friends' children in development and they may feel that they are being blamed for the child's overactive and difficult behaviour. The diagnosis is often made very late due to the fact that some children do not look different in any way and their symptoms (such as diarrhoea, frequent respiratory and ear infections as well as boisterous behaviour) are among the most common seen in all children. The doctor has to be perceptive enough to recognise that something serious is wrong and to ask for urine and blood tests to help reach a diagnosis. It is not unusual for families to have had more than one affected child before the diagnosis is established.

### The Second Stage

The second phase of the disease is characterised by extremely active, restless and often very difficult behaviour. Some children sleep very little at night. Many will be into everything. Many like to chew hands, clothes or anything they can get hold of. Sadly, language and understanding will gradually be lost and parents may find it hard not being able to have a conversation with their child. Many will find other ways of communicating, perhaps by using sign language such as the Makaton System. Some children never become toilet trained and those who do will eventually lose this ability.

### The Third Stage

In the third phase of the disease children with MPS III begin to slow down. They become unsteady on their feet and tend to fall frequently as they walk or run. Eventually they will lose the ability to walk. Life may be more peaceful in some ways but parents will need help with the tiring physical aspects of caring for an immobile child or teenager. Parents will also need to be advised on how best to cope with these changes so they are equipped and able to provide their child with the best care possible.

## General Management of Sanfilippo Disease

### Anaesthetic

Giving an anaesthetic to an individual with MPS III requires skill and should always be undertaken by an experienced anaesthetist. Where a child is concerned this should be a paediatric anaesthetist.

The airway can be very small and may require a very small endotracheal tube. This is a tube that is inserted into the trachea through the mouth or nose in order to maintain an open air passage to deliver oxygen

or to permit the suctioning of mucus. Placing this tube may prove difficult and require the use of a flexible bronchoscope. In addition, the neck may be somewhat lax. Repositioning the neck during anaesthesia or intubation could cause injury to the spinal cord. For some individuals, it is difficult to remove the breathing tube after surgery is completed.

There is a more detailed explanation of this complex subject in the specialist anaesthetic booklet published by the MPS Society.

### Physiotherapy & Hydrotherapy

Both physiotherapy and hydrotherapy can be useful to help individuals with MPS III achieve specific and realistic goals in daily life. It can also help to drain mucus from the chest.

Individuals should be as active as possible in order to improve their general health. A physiotherapist may be able to suggest ways of achieving this. The best forms of physiotherapy exercises are those that are introduced through play in the younger individuals.

In adults it is important to remember that passive stretching maybe painful and should only be used with caution.

### Medication

Children with MPS may be affected differently by drugs so it is essential to consult your doctor rather than purchase over the counter medication.

Drugs may be tried for controlling mucus production but some may make the mucus thicker and harder to dislodge. They may also make the child more irritable. The use of sedatives can increase the problem of sleep apnoea by depressing respiration.

It is now recognised that frequent use of antibiotics may make them less effective when really needed. Repeated use can also cause thrush, a fungal infection which commonly affects the mouth or vagina and produces a white curd-like deposit. It causes irritation and discomfort and will need to be treated. Your doctor may, therefore, wish to limit the number of times antibiotics are prescribed for coughs and colds.

### Diet

Most children with MPS III have hearty appetites but if your child is faddy it can be difficult to achieve a balanced diet. Ask your doctor or dietician if supplementary vitamins should be given.

There is no scientific evidence that a particular diet has any beneficial effects. Symptoms such as diarrhoea tend to come and go naturally. Some parents, however, find that a change in their child's diet can ease problems such as excessive mucus, diarrhoea and hyperactivity. Cutting down on milk, dairy products and sugar as well as avoiding foods with too many additives and colourings have all been said to help individual children.



Oliver (MPS III) with his brother Samuel

It would be advisable to consult your doctor or a dietician if you plan any major changes to ensure that the proposed diet does not leave out any essential nutrients.

If your child's problems are eased by a new diet you could try reintroducing foods one at a time to establish if there are any foods in particular that increase your child's symptoms.

### Feeding

Children's early feeding very rarely causes problems, however, some do not progress to eating food that requires chewing. Others learn to chew but increasingly find solid food difficult to manage. Children with MPS III can find lumps in food particularly difficult, especially if mixed with food of a smooth texture. The child may also become quite faddy and reject a number of foods for no particular reason.

Children may become very difficult to feed. If possible parents should encourage their child to take an active part in feeding themselves. This, as well as seeing and smelling their food, encourages them to prepare for the process of swallowing. Some parents find a musical box or the television calms their child during a meal. As children lose the rhythm of swallowing they may start to splutter and cough while eating. In addition, dribbling or drooling may be more obvious. It is better to serve food of a mashed consistency. Meat will be coped with more easily if it is made soft through slow cooking rather than just chopped into small pieces.

Thickeners added to liquids can often make swallowing drinks easier. Moving the hand gently backwards under the chin and slowly down the throat can help the tongue to move and encourage swallowing.

Choking is frightening so you can provide reassurance by rubbing your child's back and holding hands.

### Cold Hands & Feet

It cannot be said whether icy cold hands and feet are a feature of the disease or purely due to lack of activity. If this becomes a problem it may be beneficial for the child to wear warm socks and gloves.

In the later stages of the disease the body's natural temperature control mechanism may become damaged and the child may sweat at night and have cold hands and feet by day. Some children have episodes when their body temperature drops (hypothermia). If this happens you should warm your child up and ask your doctor for advice on the best ways of managing the problem.

### Gastrostomy

For some individuals swallowing will become increasingly difficult and unsafe.

In these circumstances a gastrostomy is recommended. This is a tube placed in the stomach during a short operation. Most parents prefer a gastrostomy as this does not interfere with the airway or irritate the nose.

Although a gastrostomy requires a short general anaesthetic, there are usually no complications as long as the operation is done before the child becomes too frail.

Gastrostomies can leak and occasionally the skin around the tube insertion becomes inflamed. You will be given advice from your surgeon on how to deal with this should the need arise.

### Chewing

As they become more out of touch with their environment, many children with MPS III will amuse themselves by rocking or by chewing their fingers, clothes or whatever they can lay their hands on. As this behaviour cannot be stopped, it is best dealt with by providing an acceptable and interesting range of things to chew such as rubber toys or teething rings. If the problem becomes so severe that the child is causing harm to their fingers and hands, it is possible to splint the elbows for periods of the day to prevent the hands from reaching the mouth.

### Hyperactive Behaviour

Most Sanfilippo children go through a hyperactive stage when they are difficult to control and unaware of danger. It is not so easy to modify this by drugs and it is better, if possible, to adapt the home (as described earlier). A garden where the child can run about safely is a great asset. It might be possible to get a grant for fencing if this is necessary to make it secure. It is most helpful if the child can get out to a playgroup or to school where a variety of activities can be provided. Children with MPS III do not concentrate for long periods of time and cannot be expected to sit still. Ideally there should be space for the child to run about and use up energy as well as keeping fit for as long as possible. Many children are calmed by the movement of a car and will travel well.

## Sleeping Difficulties

Many Sanfilippo children are very restless at night, not sleeping for more than a couple of hours at a time. The reason for this is not known.

It is sometimes possible to improve the situation with medication, but it may take a period of trial and error to establish which drug will work best for your child. Drugs often lose their effect after a while so some parents choose to either ration their use to a few nights a week or accept that after a few weeks the drug will have to be discontinued for a period. Some parents find they can achieve a longer period of unbroken sleep by putting the child to bed later and with a regular routine. It is vitally important that parents get enough sleep if they are to cope in the day, so do not hesitate to ask your doctor for help.

## Living with a Child with Sanfilippo Disease

### Home Adaptations

During the hyperactive stages of MPS III parents find it very helpful if they can set aside a room or part of a room within hearing distance. The room, which can be fitted with a half door latched on the outside, can be made safe for the child to play without constant supervision so that the parent can get on with day to day tasks in relative peace and quiet. Furniture which is fragile or that has sharp edges should be removed and replaced by large cushions or a mattress on the floor. Windows may need to be fitted with strengthened glass and the floor should be easy to clean. The child's favourite playthings and durable toys should be provided and a television can be placed on a high shelf or suspended from the ceiling. These could be operated by the parents using a remote control.

Children in the later stages of the disease will become progressively less mobile and increasingly dependent on their parents and carers to meet their everyday needs in areas of incontinence, personal hygiene and nutrition.

It is important to give thought early on to the ways in which the families and carers will manage when weight bearing and walking or climbing the stairs is no longer possible. An ensuite bathroom for the child's bedroom is ideal with plenty of space for a buggy and a carer to manoeuvre around in. When weight bearing is no longer possible a hoist is beneficial with tracking from bed to bath directly in line for ease of use. Adaptations can often take a long time so it is prudent to plan ahead as far as possible. The MPS Society has considerable experience of the options available to families caring for a child with MPS III.

### Education

Whilst some children with MPS III may benefit from having a mainstream education in their primary school years and enjoy the social interaction with peers, a majority will equally benefit from a Special Educational Needs Placement with small class sizes and a range of communication systems in place. Children with MPS III may have a Statement of Special Educational Needs or need an Individual Education Plan (IEP) with regular reviews.

Many will need the help of a classroom assistant during the school day as well as during breaks and lunchtimes to ensure they are able to access all the educational opportunities open to them. This is also to maintain their health and safety throughout the day.

This provision will enable the class teacher to set specific tasks for the child. The classroom assistant will be on hand to help keep the child on task and maintain concentration. Many individuals with MPS III have a very short concentration span requiring them to be constantly kept on short tasks.

### Having a Break

Caring for a child affected by MPS III is hard work and parents or carers need a break to rest and enjoy activities which may not be possible when in their caring role. Many families use children's hospices; social services respite care or have someone close, such as a friend or family member, on hand to help out at busy times. Further details of hospices throughout the UK offering respite care to families are available from the MPS Society.

### Palliative Care

Palliative care is provided to both the family and the child with a life limiting disease in situations where curative treatment is not an option. This support encompasses aspects such as respite care, symptom management and bereavement support and may extend over a considerable period of time. In addition, considerable personal care may be required including feeding and personal hygiene. This can take up a large amount of time. The stress involved can put a family under immense strain. An assessment of medical needs and a care plan should lead to an approved package of support being provided to both the child and the family. Once in place this should enable both to experience a better quality of life.



Sophie (MPS IIIA)



Lewis (MPS IIIA)

### Puberty

Children with MPS III will go through the normal changes associated with puberty, sometimes at an early stage. There are a number of ways of managing periods in older girls and you should ask your doctor for advice.

### Life Expectancy

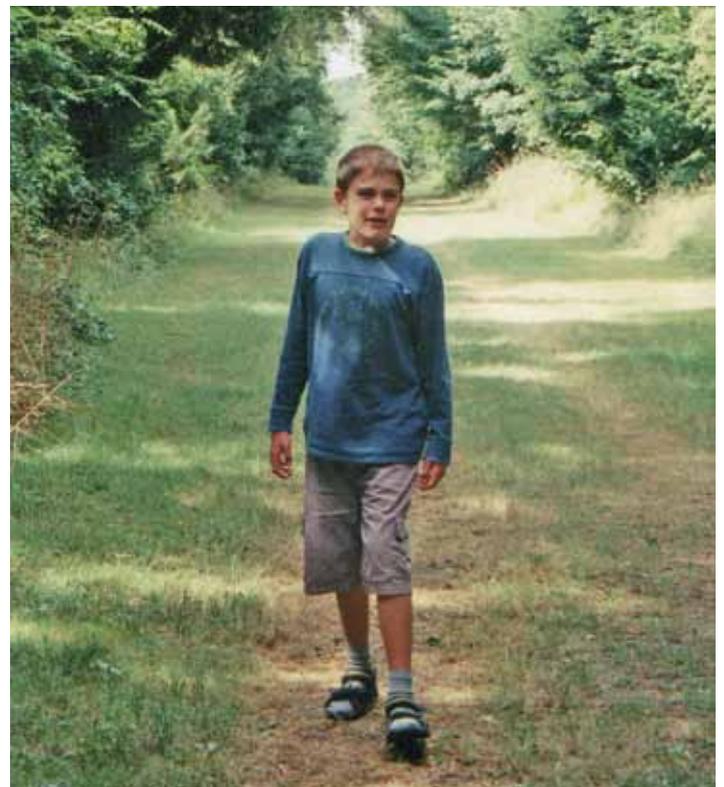
Whilst many children will lose their lives to MPS III in their teenage years, a significant number will survive into adulthood. Survival into the 30s, 40s and even 50s is known.

### Treatment of MPS III

At present there is no cure for MPS III. Various experimental methods have been used to try to replace the missing enzyme but none have proven to have any significant long term benefits. Bone Marrow Transplant has been tried on patients with MPS III but with disappointing results and is now never recommended for this condition.

### About the MPS Society

The Society for Mucopolysaccharide Diseases (MPS Society) was founded in 1982 representing over 1200 children and adults suffering from MPS and related diseases including Fabry, their families, carers and professionals throughout the UK. It is a registered charity entirely supported by voluntary donations and fundraising and is managed by the members themselves.



Nathan (MPS IIIA)

### Society for Mucopolysaccharide Diseases

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