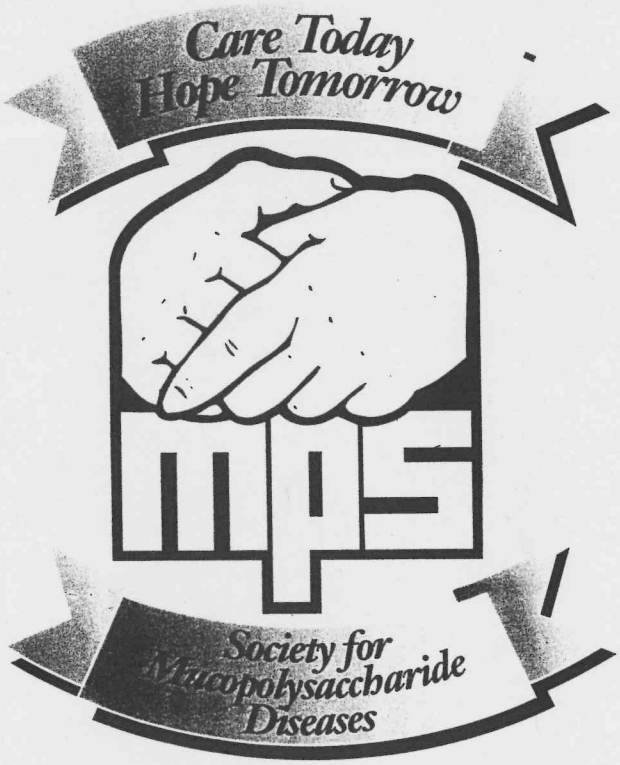
NEWSLETTER AUTUMN 1993

The Society for Mucopolysaccharide Diseases



National Registered Charity No. 287034

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The Society for Mucopolysaccharide Diseases

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The MPS Society is a voluntary support group, founded in 1982, which represents over **650 families in the UK with children or adults suffering from Mucopolysaccharide and related diseases.** It is a registered charity, entirely supported by contributions raised by members, and run by the members themselves. Its aims are:

- **To act as a parent support group**
- **To bring about more public awareness of MPS**
- **To promote and support research into MPS**

The Society operates a network of Area Families throughout the UK and Northern Ireland, who offer support and links to families in their areas. It provides an information service for families and professionals. At the present time it supports a Consultant Paediatric Post and two biochemists, one at Manchester Children's Hospital and one at The Christie Hospital. It encourages and assists contact and co-operation between parents and professionals and maintains links with sister societies in Europe and throughout the world.

There is at present no cure for MPS diseases, but much can be done to improve the treatment and care of sufferers. The slogan of the Society is:

``Care Today, Hope Tomorrow"

Please note changes to the committee and list of Area Families and corrections to office Fax number and to phone number of Kieran Houston.

Contents

Deadline for Winter Newsletter
Friday 24th December 1993.

The Annual General Meeting at Stoke

Some seventy families were represented at the Annual General Meeting of the Society, which took place on the 19th of September at the annual conference at Stoke on Trent. A brief resume was given of the Society's activities over the previous twelve months. Fuller details will be forthcoming in the Annual Report to be published early in 1994. Sean Mahon spoke on behalf of his wife Pauline, the Treasurer, giving a graphic explanation of the finances and the outlook for the future.

*Two new committee members were elected, **Lynn Grandidge** who comes from Chester, and **Tony Eyre** who lives near Bristol and who, with his wife Shirley forms an area family partnership.*

New blood is most welcome to the committee and although September 1994 may now seem a long time ahead, there will be further vacancies which will then need filling, so I make no apologies for reiterating what I have said many times before. Do please seriously consider putting yourself forward for nomination if you think you can make a useful contribution to the running of the Society.

*Two members left the committee, **Sue Butler and Robin Lavery.** Thanks were expressed to both of them at the meeting and I am sure you will wish to join me in thanking them for all their support, loyalty and input over many years. Sue, you will be pleased to know, will be continuing as Sales Co-ordinator and we are all grateful to her for all that she does in this respect.*

Robin was of course at the heart of the Society from its inception and he has contributed more than most over all these years. I am sure he will continue to take a more than usual interest in the Society and that he will continue to express his views, which are always most welcome.

Thank you, Sue and Robin.

Perhaps this is the time to remind all members that if you have any views on how the Society should be run, or any particular topics in which we should we should be interested, please do let me or any other member of the committee know.

A.G. King

Chairman

Tea time at the conference.

Tree Planting at the Childhood Wood

The Childhood Wood is looking particularly beautiful at present, with the heather blooming between the rows. Christine has visited the wood recently and reports that the trees are flourishing and have almost doubled in height. The forest rangers have recently had a weeding party and cleared the undergrowth.

The Society is now organising a second planting. This will take place on Friday, **February 25th 1994.** We will be writing to those who have lost children during the year and inviting them to join us there or to commission a tree if they wish.

A working party of forest rangers, MPS families and friends will take place at the end of June 1994 to assist with weeding and to clear deadwood from the site. We thought this might be an excellent time for bereaved families to return to the wood. If sufficient people are interested we will consider hiring a marquee and organising a picnic lunch. We could also negotiate a special rate for hotel accommodation if enough families wished to stay overnight. There will be more news about this proposal in the next newsletter. If you are interested in joining the weeding party please let the office know.

* The MPS Marathon Team 1994?

Mike Kilvert has made an application for an MPS team to run in the London Marathon next April. He will not know until November whether this is successful. If it is successful he will need runners! It is much more encouraging to run in a team so if you know anyone who would like to run please persuade them to join up with Mike and get a place on the team. Full support available on the day.

Mike Kilvert,

Windy Waye, Nantoer, Newtown, Powys, SY16 1HH, **Phone 0686 624387**

* Marathon man takes to his wheels!

Mike Willoughby, who has run the marathon several times for MPS, decided to give it a miss this year. Is he getting old and tired? Don't bet on it. With three colleagues from work, he plans to cycle non stop from Land's End to John o'Groats. They intend to do it in two hour relays.

Campervan Needed.

I have offered to go along as a driver. I would like to ask if anyone has a camper van, to use for the cyclists to sleep in when they are not riding, or a support van to carry the bikes that are not being used. Also, does anyone know of any decent cheap bed and breakfasts, one near John o'Groats and one near Land's End? The ride will, hopefully, take place in the last quarter of next year. If anyone can help it would be much appreciated.

More news about this next year and also sponsorship forms.

Ron Snack.

(Address inside back cover)

Fundraising in Bedfordshire

Albert and Trudi Deacon, of Cranfield, lost their son to MPS in 1958, but only found out about the Society recently via their daughter. Although they are both pensioners they decided they would raise money for us. They held a very enjoyable sale and raffle in the garden of their home in August. A total of over £450 was raised. I can do no better than let you read what they have to say:

Trudi Deacon with her two daughters and friends at their sale

"We were glad we have at last found a charity which is of great interest to us. We have attended all the various charities in Cranfield, but never an MPS one. We were pleased our sale went off well. I dreamed we would be looking for people to attend, but I need not have worried - we could not believe where they all came from.

We were pleased to let people in Cranfield know about MPS so they now know why Michael was as he was, because so many went to school with him.

Thanks to you all.

Trudi and Albert Deacon

Congratulations!

To Alex Butler on passing his driving test on the 12th of October

1993 Marathon Sponsorship.

The final amount for Mike Willoughby's sponsorship for this year's marathon was £3077.17. I would like to thank all those who collected sponsors and to Abbey National who matched £1000 of the money.

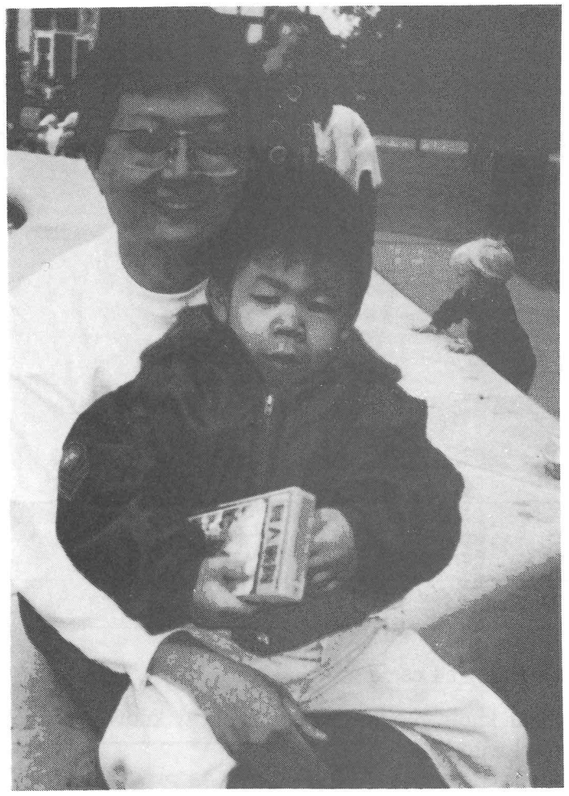
Ron Snack.

MPS Radio Appeal

We are delighted to have been awarded a Broadcast Appeal by the BBC on Radio 4 at 8.50 am on Sunday 14th of November. This will be a marvellous opportunity to publicise our work and to raise much needed funds.

Elinor Bennett, International Welsh Harpist and mother of two sons who had Sanfilippo disease will make the appeal on behalf of the Society.

Area Family Day at London Zoo



David Chou with his father Eddie at London Zoo. The Chou family live in Ashford, Middlesex. David suffers from Hunter disease.

It was a real treat to join the MPS family day at London Zoo in the beginning of June. I had not been there for ten years and in that time it has had it's ups and downs. We had extremely pleasant surroundings with good company and an atmosphere of enjoyment.

Driving into London early on a Sunday morning was really easy. We had no trouble parking opposite the main entrance - which meant we did not have to carry our lunch about with us.

Despite all the doom and gloom about its financial standing, the Zoo is still a very large place, full of attractions. It was difficult to know where to start. The noise coming from the chimps drew us that way. It was quite frightening to see how boundary disputes were settled between them.

Going towards Three Island Pond and the pelicans took us past the pandas, where Ching Ching and Chi Chi had resided for so long. We saw a photo of the world's most destructive animal - Man!

We were given some helpful advice from a member of the Zoo staff on how to tell the different species of big cats. He said the easiest way was to read the labels!

After lunch we had a photo call by the fountains. Then we were off to the Children's Farm and the opportunity not only to see the animals but to touch some of them as well.

The tunnel to the north side of the Zoo is decorated with prehistoric cave paintings, which Alex, our daughter, has been trying to copy ever since.

By the end of our visit our feet were dropping off. A quick cup of tea and a bun, more photos and farewells as we started our journeys home.

Thanks to Ron and Linda Snack for organising another excellent day.

Alan and Tina Marchbank

Annual Conference

The annual conference took place from the 17th to the 20th of September at the Stakis Grand Hotel in Stoke-on-Trent, a venue that we now regard with the affection of long acquaintance. Over seventy families attended for a relaxed and informative weekend. A report from a talk by Ed Wraith is included in this edition and other talks will be reported in future numbers. **Paul Butler**, uncle to Alex, surpassed himself with the disco on Saturday night. We had to stop the dancing at 1.00am because the walls began to sway to the music. Particular thanks to **Ann Neal** and **Jenny Broome** who organised the conference. A wonderful vote of affection and thanks to all the volunteers who looked after the children on the outings and who did the babysitting. (See photos, pages 14 to 17). A special mention too for **Ann Ridley** and friends who cared for eight severely disabled children in the creche during the weekend. My apologies that I have been unable to give the names of the children in the photographs.

Dear Sam,

I didn't want you to go and I knew you did have to go, but it just wasn't fair. It was just coming up to the Christmas Party for MPS and Christmas with your family. Your mum has taken it very well. On Sunday it was Carly's dancing concert. She was a flower and a mouse. On Thursday I'm glad you laughed and smiled as I can remember you as a cheeky little boy. I first remember you from the time Laurie and I knocked at the door selling Yatton Junior School Fete programmes and you were sitting at the table smiling and laughing as always. After about a year you recognised my voice. If your mum said something you would smile and when I spoke you laughed and your mum would call me all the names under the sun. After a while you felt like a little brother to me I loved and cared for you and still do as I know you are still with me and watching over me. Sam I will never forget you through all my life and I will still go and visit your mum and dad and your sister Carly. And also I will never forget the trouble we had with your hearing aids. Sam I love you.

I will always want to know why now, why not in the next century, I don't care when it is as long as it isn't now. I kept saying,"Not my Sam dead, it can't be, they have made a mistake.

Celebrating the memory of Sam Eyre

Everyone who knew you said you were a lovely little boy. We had conversations about different and weird things. We used to play on your keyboard and your bike. You used to sing along to "Neighbours" every evening.and then you knew it was time for your dad to be home soon. You loved your chocolate buttons and mini cheddars and you always liked your red cup with the two handles.

What will I do without you Sam? When it was my birthday you and Carly and mum came up to my house and you rode your bike around the house and you liked the birds out in the garden.

I love you Sam, take care, see you soon,

Your helper and friend, whatever you want to call me,

Yvonne Julie Miles

This letter was written by Yvonne two days after Sam Eyre died on the 29th of November 1992. Yvonne continues to be a close friend and neighbour of the family. She was fifteen when she wrote the letter. **Tony and Shirley Eyre** have asked for it to be included in the Newsletter. They send their best wishes to everyone.

Milestones

Births

Congratulations to **Ann and John Brown** of Tunbridge Wells on the birth of their daughter, **Lucy,** born 16th August 1993.

Congratulations to **Elizabeth, Richard, Hendrick, Emily and Tilman Volk** of Sinzig, Germany on the birth of **Sophie Leonora**, born 25th September.

New Families

Peter and Julia Andrews of Thurmaston, Leics. whose sons, **David** born 21st of September 1989 and **Jamie,** born 19th of August 1991 have been diagnosed with Hunter disease.

Wendy and Karl Hitchin of Beddington, near Croydon, whose son **Sam,** born 6th of September 1988 has been diagnosed with Sanfilippo disease.

John and Emily Slater of Droitwich, whose daughter **Emma,** born 25th December '92 has been diagnosed with Hurler disease.

Sandra Mc Intyre of Greenock, Strathclyde, whose daughter **Jade** Docherty, born 12th of December 1990, has been diagnosed with Maroteaux Lamy disease

Deaths

Ibrar Ali, of Small Heath, Birmingham, died on the 21st of August 1993 aged five. Ibrar suffered from Morquio disease.

Leanne Woods from Bracknell, Berks. died on 28th of August 1993 aged eight. She suffered from Hurler disease.

John Kennedy , father of **Andrew Kennedy** of Stockport, Cheshire, died tragically on 27th of August 1993, age 39.

Matthew Blackburn of Nantwich died on the 30th of September 1993 aged seventeen. Matthew suffered from Hunter disease.

Bill and Sylvia Blackburn wish to thank all the many friends for their sincere message of sympathy which have been a great comfort at this sad time. Also thank you for the flowers and donations received in memory of Matthew. Everyone has been so kind and this will always be remembered.

Sylvia and Bill

Marian Corcoran of West Ealing, London, who died on the 10th of September age four years. Marian suffered from Hurler disease.

Our thoughts are with the grieving parents relatives and friends of all these children. We pray they will find hope, courage and strength in this sad time.

Congratulations!

Eleanor (Ellie) Dronfield and I were married on the 31st of July this year in Leeds. We are living in Nottingham where I have begun a job as a support assistant in a local college. I work with people with physical disabilities. Ellie has recently graduated and qualified as a social worker and is seeking employment.

We were lucky to have two honeymoons; one in Ireland and the second the MPS young peoples holiday on Exmoor. Not only did we we survive the activities but also the teasing about being a newly married couple as well!

We would both like to thank all those involved in organising the Exmoor holiday. We would also like to thank everyone in the Society for their best wishes, card and voucher - the latter received at the conference dinner and dance - thank you all.

Love,

Paul and Ellie Gunary

Elena Mc Gauran in her own write

My name is Elena McGauran and I have Morquio syndrome. I was diagnosed at four years of age. My first contact with MPS was in 1986 at the age of fourteen. That same year my parents and I went to the International Conference in London. To have the opportunity to meet with Morquio children was a dream come true. Up until then I thought there was no other child like me.

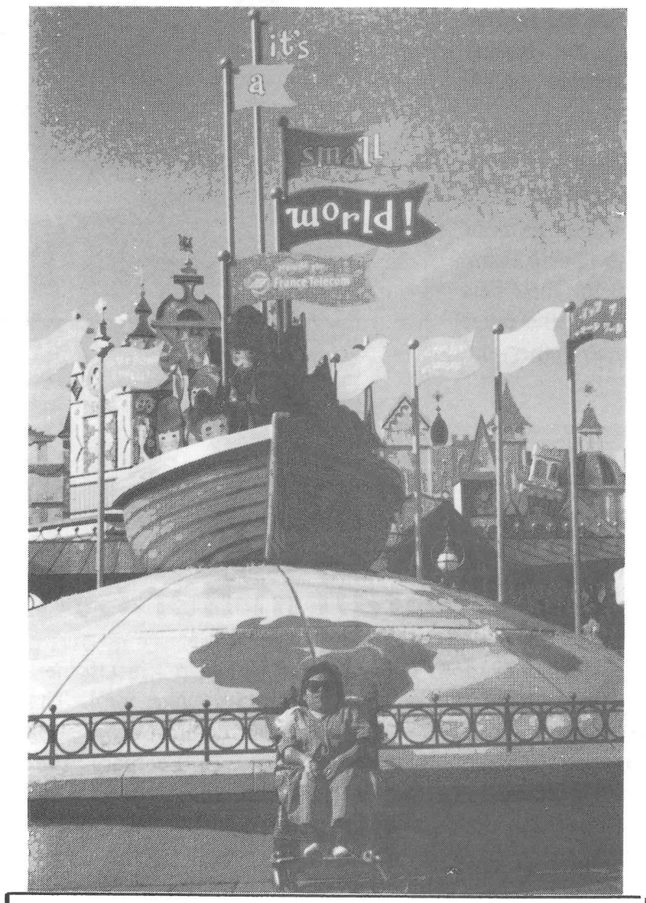
"My dad told me that everybody had a disability - it was just that some were more apparent than others"

My parents have given me great confidence for which I am most grateful. From age seven onwards I knew I was different but my Mum and Dad assured me everyone has a disability of handicap - just some are more obvious than others - ''like me''. This thinking has been a great comfort and strength to me over the years. In February 1988 I had my cervical fusion and I was in a plaster cast and halo for six and a half months. As it was fairly lightweight I was able to use my battery operated chair -"The Pony" - so I was not totally restricted. Living in a provincial town people soon got relatively used to me buzzing around with my antennae!

For years I was a keen cyclist (on a two wheeler)! until I was eleven years old. I also

Elena's story (continued)

enjoyed Brownies and Girl Guide activities, I went to Speech and Drama classes and have a few awards to my credit. I took piano lessons up until two years ago. I also enjoyed swimming but last year I was getting a lot of ear and chest infections. So that was the end of the water scene. Secondary school/Graduation Day, Graduation Dance are all happy memories and now I am doing a Secretarial Course in my local town. Our college has relocated to a bright new wheelchair friendly building - thus I am now able to study all skills associated with the course. I have received Word Processing and Typing Certificates, a City and Guilds Certificate and I will sit City and Guilds Exams in book-keeping and computers in the Autumn. I get work experience in my Dad's office but 1994 will see me job hunting.



Elena goes to Paris to find a world that's the right size.

Generally my home town of Sligo is wheelchair friendly - I have total access to churches, secretarial academy and bank (The one with wheelchair access gets my business!) and most shopping areas. Crossing the streets of Sligo can be difficult for people because it is a very busy town. But being three foot nothing in a wheelchair creates a problem too, but my Dad - a real live ''Dad'll fix it'' - fitted a blue flasher - in competition with the police! So I flash my way across to the shops, bank, post office whatever. This has greatly increased my independence.

On May 9th I celebrated my 21st Birthday - I had a party and the biggest surprise of the evening was when my family presented me with airline tickets to Paris and four nights accommodation in EuroDisney. Two weeks later saw my Mum and I flying out to Paris. I was so excited.

Three cheers for EuroDisney! It is so wheelchair friendly. All that talk about long queues, well forget it, one advantage of being wheelchair bound is you skip the queues. It certainly was dream holiday for me. I even experienced the Metro in Paris at rush hour - quite overpowering, to say the least.

I love being in touch, always checking the mail box - one of my favourites through the letter box is the MPS Magazine. I read it from cover to cover - always looking for news of Morquio people. So Dear Editor and team a very special "Thank You" for your dedication and delivery.
Good luck to you all in the future.

Elena

Ann Canton tells her story

Believe it or not I have been asked many times to write something for the magazine, at last I'm putting pen to paper.

I was thirty one years old when diagnosed as having Scheie Syndrome, one of the MPS diseases. Until that time it was thought that I suffered from Stills Disease (juvenile arthritis) as a child.

I was quite relieved to have a proper diagnosis and name to what I had. When Dr. G. Ruki, who was the rheumatologist I saw at the Heath Hospital, Cardiff wrote the name of "Mucopolysaccharide" at first I found it quite strange. Imagine how I felt when I saw this word in print for the first time. It was in the "Woman's Own", that I read an article written by Christine Lavery. I wrote to the "Woman's Own" for the address and telephone number and eventually I received them. So this was how I found out initially about the society. What lovely memories we have of all the people we have met and are still meeting in this way.

The first conference Mervyn and I attended at the Post House Hotel, Heathrow we were a little apprehensive but what a joy it was to be made to feel welcome when we arrived at the hotel foyer and at the MPS registration desk. Since that time the society has grown a great deal, but a lot of the old faces, like ours , are still around.

I have been fortunate even though as a child I spent time in different hospitals and was unable to do all the things I would have liked to do. I did obtain a few 'O' levels and got a diploma in Clinical and Commercial subjects at Portland Training College, Mansfield. I was in full employment with accountants for many years. Twelve years ago Mervyn and I were married. Until a year ago I worked part time as a book keeper for a firm of solicitors. I was told at this time by my doctor to give up work. I was disappointed having to do this and for a while I was quite depressed, now though I realise it was for the best.

I spend some time now at the local junior school in a voluntary capacity listening to youngsters reading. I enjoy this very much as I am told some amusing stories and find it hard to keep a straight face. I get frustrated at times as there are certain things I cannot do but I do realise that I am very lucky to be as well as I am. I am fortunate for I have a very caring husband who has plenty of patience.

I am sure that parents and sufferers of Mucopolysacharide Diseases find the Society very helpful whenever there are any problems. Mervyn and I would like to express our grateful thanks for the special friendships we have made through the Society. We are very sorry because of the distance of travelling we do not do as much as we would like for the Society. May I take this opportunity to thank all members of the committee and officers for all the hard work they do for the Society.

Ann Canton.


Ann and Mervyn Canton at the Conference

The Calvert Trust Experience

My name is Andrew Butler and I am twenty seven years old. I come from near Chesterfield in Derbyshire. Until this year I had little contact with the MPS Society. Then suddenly out of the blue I received a call from Christine Lavery about a possible place on an adventure holiday at a centre down in Devon. My first thoughts were to refuse the opportunity but after sitting back a while I decided to grab the bull by the horns and give it a go. So on the 29th of August, with my parents help I set off on the long journey to the Calvert Trust farmhouse.

The house was surrounded by fields and trees. On my arrival I was greeted by Christine and shown to my room. This was plain and simple, but adequate for the amount of time I was going to spend there. After unpacking I got rid of my parents and began to meet the other people. This included Christine's family, the army gang of helpers, the ever bubbly Mary Gardiner plus the other MPS members.

We then set off on a little expedition exploring the surrounding land. This took us along a zig-zag path down a steep slope, which led us to a small picturesque reservoir. There we had our first look at what has become the famous Anneka Rice boathouse.

The following day the holiday truly began. Kitted out with waterproofs, rucksack and lifejacket we went down to the sun trap of a landing bay on the reservoir. This was to be our first spell of many of sailing and canoeing. We took our boats on a circuit of the reservoir and were very lucky not to get wet either by accident or by design.

Our second activity of the holiday was orienteering. We divided into small groups and were Then, by using a map and compass we had to get round a small course in the neighbouring fields as quickly as possible.

Another activity which I think I enjoyed most was archery. We were given bows and arrows and tips on how to use them correctly. This was our chance to imagine being Robin Hood in Sherwood Forest or a Red Indian from the Wild West.

We also spent time at places of interest and natural beauty such as the small horses centre and the coastal towns of Lynmouth and Lynton. Lynmouth is at the bottom of the cliff and Lynton at the top. To get to Lynton we had to ride on a water powered cliff lift. We also each got a chance to drive a horse and cart.

The evenings were usually spent at the farmhouse having lovely barbecues cooked by the army people and chatting about the day's events.

On the final evening we went to a little public house for a goodbye dinner. After the meal we were all presented with a small gift linked to something we have become associated to on the holiday. I was given a bar of soap in the shape of a rhinoceros for my snoring and the effect it had on the group.

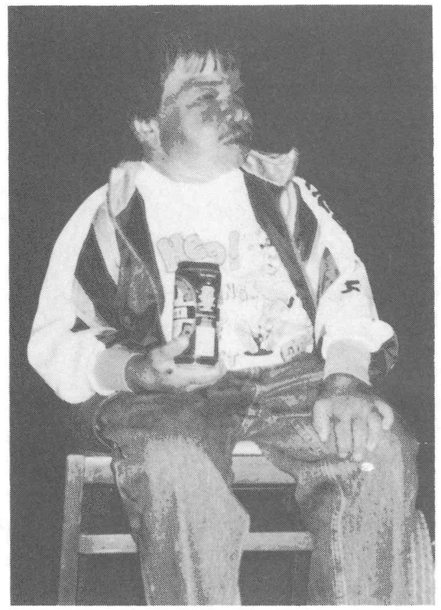
The holiday was full of adventure and new activities for me. It allowed me to make new friends. It was a complete success and I hope there will be more to come.

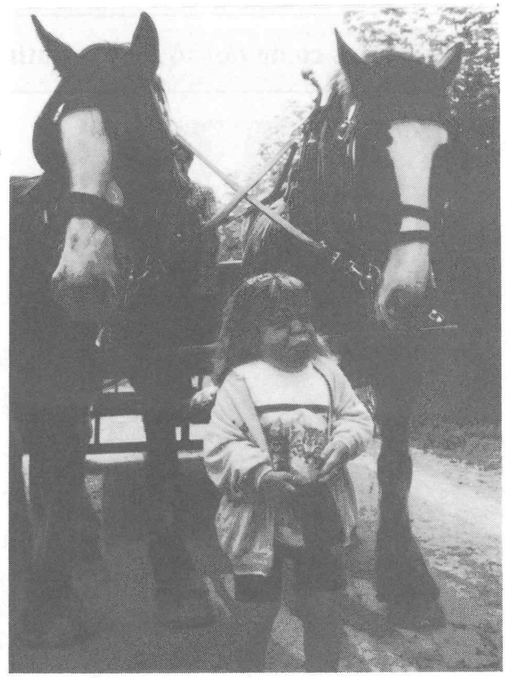
I would like to thank everyone involved for all their time and hard work to get this project off the ground.

Andrew Butler

"Rutland", 48 Chesterfield Rd.,

North Wingfield, Derbyshire.





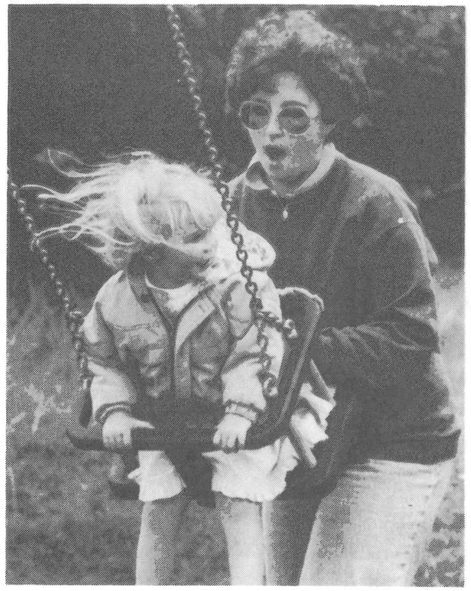
Top left: Andrew Butler enjoying a pint.

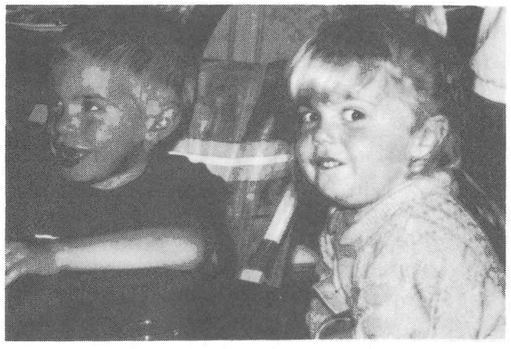
Above: Sarah Lowry, animal lover!

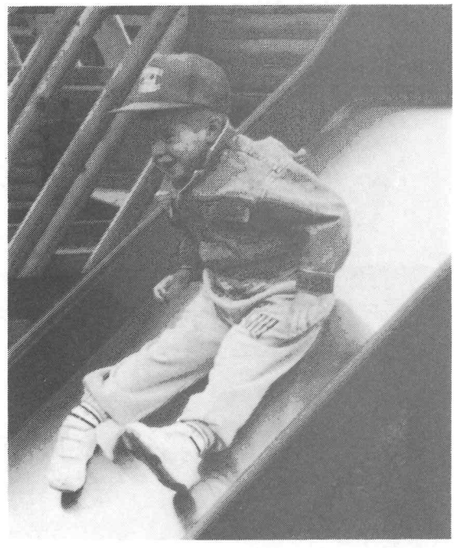
Left: Sarah Kilvert takes to the water, with just a little help from her friends.

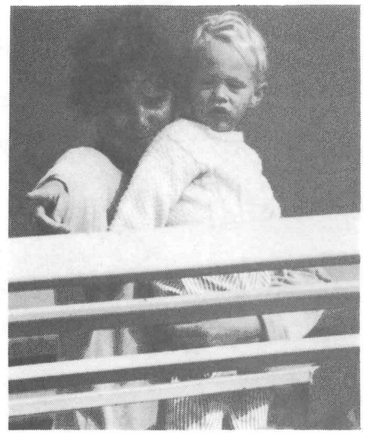
Ellie and Paul Gunary, Derek Denhan and volunteer John take to the woods, while in the background the "ever bubbly" Mary Gardiner takes pictures.

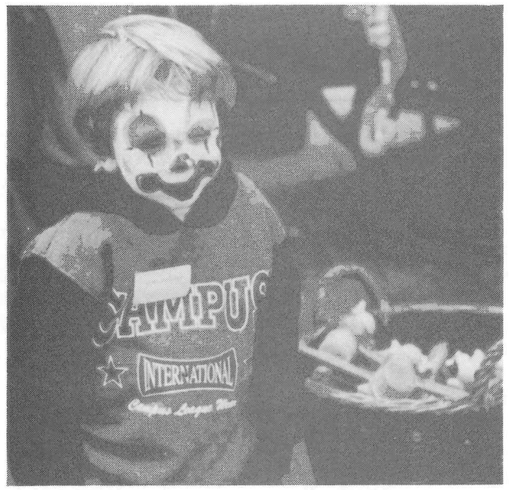
Girls and boys come out to play - outings at the annual family conference in September







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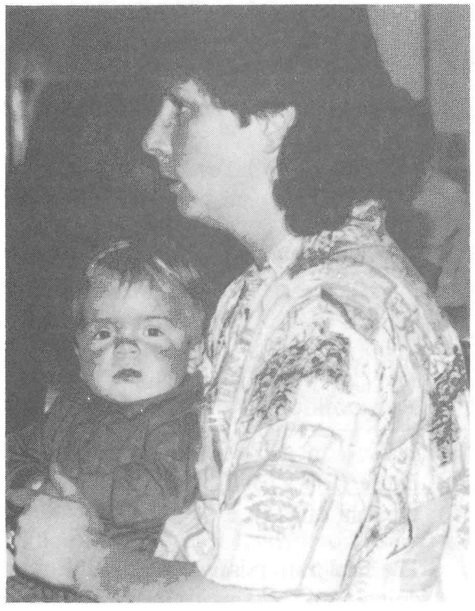
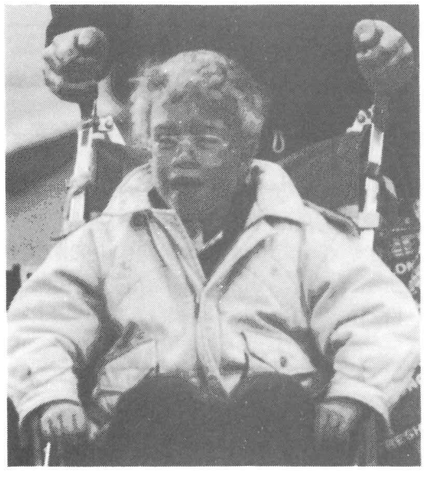
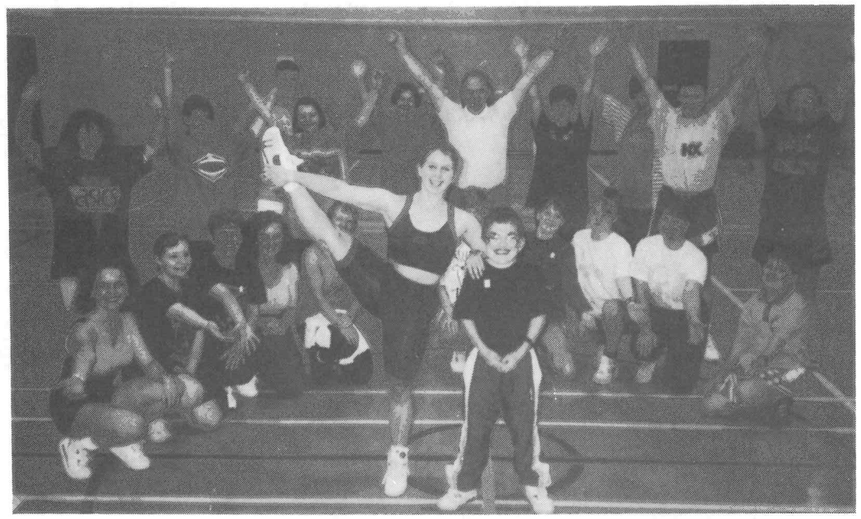




Fundraising for MPS - Derek Denham



Earlier this year I organised two events to raise funds for MPS. The following is a brief account of each event.

Twelve Hour Game-athon

Three of my friends (Sandy Baird, Darren Vance and Richard Mc Call) and I played a variety of games for 12 hours non-stop. The games were:

Pool, Snooker, Dominoes, Cards, Chess, Sega Games, Car Racing.

Even though it was very tiring, we enjoyed ourselves and raised a lot of money. The final sum was £250.

Aerobics Session/Fitness Challenge.

This event was held in two stages. First of all the Aerobics Session was held. This was a simple work-out for anyone of any age. I participated for some of the event. Some of the participants were sponsored, so they had to complete the routine. The session was guided by aerobic instructor Jackie Harkness. The second half of the event was a fitness challenge. This was a contest for the fitter people who came along. The participants had to do as many exercises as possible within a forty five minute period. We had prizes for both men and women and they were won deservedly. The sum raised in this event was £170.

Pub collection

I also had a collection tin in my local public house (Black Bull), and they managed to raise the sum of £150.

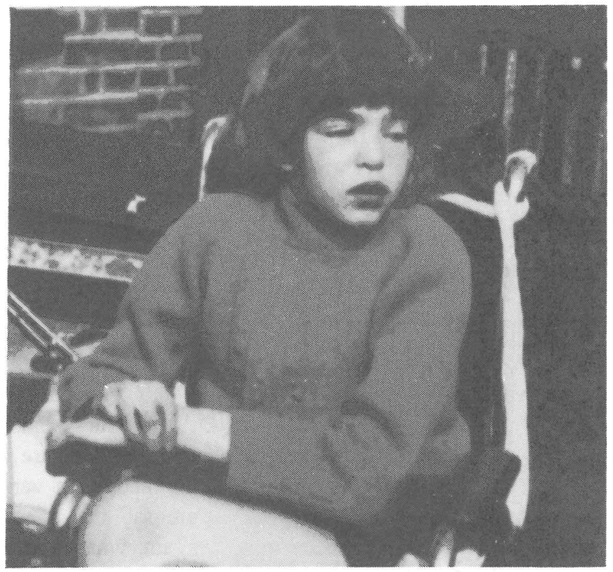
The final sum raised was £570. It was good fun doing the events as well as organising them. I hope to do more fund-raising in the future and to beat £570. Thanks to all involved in the events!

Derek Denham.

Tracey Brigden

My daughter Tracy was born in January 1962. She was a premature baby so I attended a clinic with her once a month. I became concerned when she did not seem to be doing as well as she should. No one listened to me. I was told that all children develop at different rates and that I worried about her too much because she was my only child.

They agreed that she was hyperactive and a little slow in learning. I was told not to worry because she would improve as she got older. By the time she was ten years old she had stopped doing all the things she had learned and she was limping badly.



Tracy Brigden age 12 in 1974

Finally the doctor said she had Perthes disease (a hip condition). She was admitted to hospital for a hip replacement operation. After the operation her condition deteriorated and she never walked or talked or smiled again.

"She never walked or talked or smiled again".

She was sent to Great Ormond Street Hospital for some tests and she was diagnosed as having Sanfilippo disease. At that time very little was known about the condition. All I was told was that it was progressive and that it was incurable. She was returned to my local hospital where she stayed until she died in May 1980.

In all the years since then I have never met anyone who had ever heard of the disease, until quite by chance I saw a letter in my local paper from a charity organiser who had collected money for a child with Sanfilippo disease, to help to send her on a trip to Disneyworld in Florida.

I contacted him and I met the child and her family. They put me in touch with the MPS Society. I had no idea that the Society existed until then They have been very kind to me and brought me up to date with what is being done for these tragic children.

They have also told me about "The Childhood Wood" and I am to have a tree planted for Tracy. What a wonderful memorial for her. At last I know that I am not alone and that there are people who care and know how we feel about our children. I can't tell you how good that makes me feel. I only wish I had their support years ago, when I needed it so much.

Sheila Brigden

Flat 12, Nita Court,

152 Burnt Ash Hill,

London SE12 OLJ

Ilia Kandel

Our little girl Ilia was diagnosed in May '92 as being a sufferer of "Sanfilippo disease". She was just turned seven years old. What a shock! Neither I nor the local doctors, schools social services etc., had ever heard of MPS. Well they sure have now. Everyone knows when I have had to be in contact with them. "Once met never forgotten", that's me! I have found out how to make a damn nuisance of myself, it helps you get further. Everything I have ever asked for I have received, so I must praise my local Council at Barnsley.

Later the soft room will convert to Ilia's bedroom, when I can't get her upstairs. The local services have even installed a hoist for her, to get from bed to bath with a turntable to take her to the changing bench.

I count my blessing for having a lovely daughter like Ilia, I feel that she was given to me for a reason, a special reason? Yes, it has certainly changed my life, opened my eyes, to handicapped children & life.

"Everyone knows when I have had to be in contact with them" says Gail

I would like to tell you how I first met Mary Gardiner, it was on our first visit to see Ed Wraith at Royal Manchester Children's Hospital in '92. She was really nice to us, "Anything I can help you with just ask," so I did! I organized a meeting in my home between our local services. - Aids & Adaptations - Social Worker - Head Paediatrician - Chief Physiotherapist and of course Mary Gardiner & Pauline Mahon for MPS were all there, tea cups a flying. They sat for over two hours, photos and facts of all Ilia's needs going across the table.

Now I am sitting in my dining-room looking at a wonderful extension thanks to all at that meeting. Ilia has a lovely soft room all padded out. The padding company "Sutcliffe Leisure" even donated a ballroom ball - magic lights and soft shapes which I thank them for, so now Ilia has a lovely soft room where she can't hurt herself, she is "Free" again and its great, no more sitting in her chair while Mam does housework or cooking. There is also a lovely bathroom next to it with an Island bath and a padded changing bench.

It has also made me realize my family, all of them, Ilia's Daddy, James, her sister Sally, my mam & dad, are all very special people in my life and I am very happy with that, I know I am sad that Ilia won't always be with us but again, what time we have with her is special.

Thank again to the M.P.S Society for all your help.

Yours most sincerely

Gail Kandel.

50 Yvonne Grove

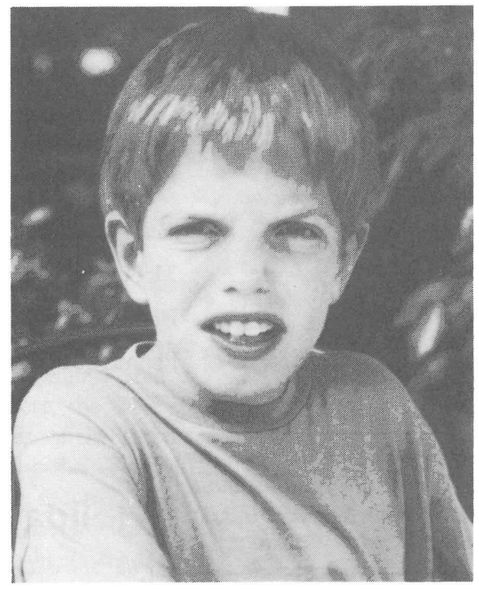
Wombwell, Barnsley,

South Yorkshire, S73 8NA

Andrew Hawkins

There is so much that we would like to relate through this article, but space does not permit. First let us introduce ourselves, **Peter and Jean Hawkins** and our four children, **Tracy 23, Debbie 20, Lindsay 16, and Andrew 10.**

When we finally had a son our excitement was immediately muted by his having apparent breathing difficulties at birth. On examination Andrew was pronounced fit, but during the night he had to be rushed to another hospital with a partially collapsed lung. After seven days intensive care he was allowed home, jaundiced, but apparently normal. A diagnosis that we came to question as he barely slept at night for the first eight months of his life.



Andrew Hawkins, Sanfilippo A, aged ten.

Behaviour problems

He then settled until he was approximately two and a half years old, when slow development in his speech was identified. By four years of age Andrew showed signs of behaviour problems. He instigated situations which worsened over time and this resulted in very serious rifts with both family and friends which have not been reconciled since then.

Better at school - a nightmare at home.

At five Andrew went to mainstream school where he caused havoc. After fifteen months he was given a ''Statement of Special Educational Needs'' and was transferred to a school for children with 'learning difficulties'. He behaved better at school but became a nightmare at home!.

Domestic life worsened. Eventually the medical profession identified Andrew's problems as being due to a chromosome abnormality. This 'red herring' clouded the issue for two years. The saga of what was wrong with Andrew became very prolonged. We began to have questions about the response and support of the medical profession. The saga ended when Andrew was diagnosed Sanfilippo A in May of this year at the age of ten.

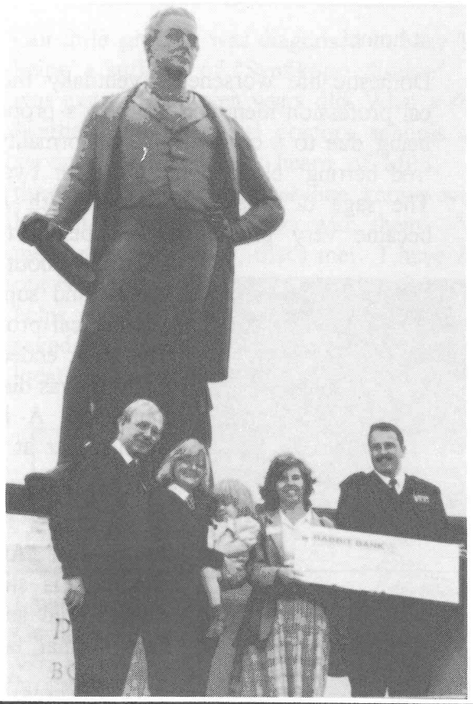
At present Andrew's condition is still considered mild as he has near normal faculties. We love Andre dearly so we have been able to derive much enjoyment from him, especially on his good days. However we know that his condition will deteriorate. This thought is forever in our minds and particularly when Andrew demonstrates the horrendous side of his behaviour. In fact it is this dilemma of bad behaviour from an incurable disease that clearly identifies Sanfilippo for being the wicked disease that it is.

Today at least we know the truth and hopefully we can cope with what lies ahead.

Peter and Jean Hawkins,

17 "Wimbushes", Manor Park Drive, Finchampstead, Wokingham, Berks., RG11 4XG

Jessica Rabbit Raises £2000



Sue and Peter Stuart are both police officers in the Metropolitan Police. Their two and a half year old daughter has Hurler disease. When Sue's work colleagues at Harrow heard of Jessica's plight they wanted to do something to help. Sgt Barry Simons designed a small enamel badge to sell throughout the police service. The design was a rabbit (one of Jessica's favourite toys) in a police helmet.

Peter says, "The badge has proved to be a firm favourite, both with our police colleagues and with the general public. We shall continue to sell them to raise money specifically for Gene Therapy.

If you are interested in helping to market the Jessica Rabbit Badge please contact:

Peter and Sue Stuart

31 Stevenage Crescent,

Borehamwood, Herts WD6 4NS

Phone: 081 953 2095

Sgt Barry Simons presenting a cheque for £2000 to Christine Lavery by the famous statue of Robert Peel at Hendon with Sue and Jessica looking proudly on.

Dear Friends,

It is with with deep sorrow that I am writing to let your know that my grandson, Tyler Smart age twelve passed away peacefully on the first of June. Tyler suffered from Hurler disease. He lived in Winnipeg Canada with his mum Hazel and his brother Neil.

Tyler left us wonderful memories. His beautiful smile will be fondly remembered by the many friends whose life he touched.

May God bless all MPS sufferers and their families.

Joyce Brown

24 Headlam Green

Newcastle upon Tyne NE6 2PG

Holiday Home Exchange

We are a German MPS-family with a nine year old Sanfilippo boy, called Julian and an eleven year old boy, called Marius. Our names are Kornelia and Werner Weber. We live in Olpe which is a little town (20,000 inhabitants) 75 kilometres east of Cologne. It's very easy to visit a lot of towns in the middle of Germany by motorway from Olpe. Our town is situated in a nice landscape. There is a famous lake (Biggesee) nearby with hills and forests, where you can walk, swim and go by ship. We live in a little house with a garden and three bedrooms, a livingroom, a diningroom, a kitchen and two bathrooms.

Next year we would like to go to England, Wales or Scotland for three weeks during our holidays which are from 26th of June to 3rd of August 1994. We are looking for a British MPS-family, who are interested in spending their holiday in Germany and who would like to swop homes for three weeks during this time.

Please contact us if you are interested in spending your holiday in Olpe. Our address is::

Kornelia and Werner WEBER

Kupferweg 6, 57462 OLPE, Germany.

First Russian Conference

The first Russian conference of families with children with lysosomal storage diseases (LSD) was held in Moscow on October 10-12 1993. The motto of this conference was "Love and hope". This conference was a result of a special programme of LSD prevention which started in the National Research Centre of Medical Genetics in 1982 (the head of the programme - Prof. X.D. Krasnopolskaya). Nearly 400 patients had been diagnosed as having 21 various LSD including 64 families (74 pregnancies) at risk who had prenatal diagnosis over a period of 10 years.

Families have to overcome the difficulties of the present situation in Russia.

Sixty five families from all over the country gathered to solve their problems. Like all such families they need contacts with one another, communication to overcome loneliness and a phobia for doctors. What is more, the families have to overcome the difficulties of the present situation in Russia and fight for their right to diagnosis, treatment and social security. The country being in an economic crisis there are constant shortages of drugs, reagents for diagnosis, physiotherapeutic equipment and wheelchairs. (Hardly 1% of the patients are provided with a wheelchair).

Every family got a special "information envelope" which was which was prepared in the laboratory of inborn errors of metabolism (National Research Centre of Medical Genetics). Their travel expenses were covered by the Russian Society of Invalids. Their accommodation was provided and subsidised by the Department of Social Security. The conference took place in the Institute of Youth (former - Central Komsomol School), where a special faculty of social help was established two years ago. All the students of this faculty were participants of this conference. and helped to look after the families.

They acted as personal tutors to the sick children, they arranged cultural programme and family's meals. The guests of conference - Mrs. Marion Kraft chief of Austrian "Gesellschaft fur Mocopolysaccharidosen" and Dr. Anna Tylki-Szymanska (from Warsaw) - took part in this conference and told its participants about their experience in the organization of special societies for affected families in their countries.

During the conference the patients were given a medical counselling service and their needs were attended to. They also had a meeting with Father Amvrosij, who talked to the families and gave them encouragement and hope. The participants enjoyed a performance by a children's ensemble from a local factory.

Aim to establish a centre for families with children with inherited diseases.

The main aim and outcome of the conference was the establishment of a Centre for families with children having inherited diseases at the National Society of Invalids. According to the charter of this Centre, parents of affected children have a right to engage in tax-exempt business activity to support their children.

Please contact us!

The Russian Centre of affected families would like to get in touch with similar organizations in other countries and would appreciate any information about their work, especially informative booklets to be distributed among affected families.

Prof. X.D. Krasnopolskaya.

National Research Centre for Medical Genetics

Moscvorechie str.,1

115478 Moscow, Russia.

Fax: 007 - (095) - 324 0702

Gastrointestinal Disorders in children with mucopolysaccharidoses

Dr J.E Wraith
Consultant Paediatrician
Royal Manchester Children's Hospital

The following is a digest of a talk given to parents at the MPS conference in September 1993.

Introduction

The digestive system can be regarded as a long muscular tube beginning at the mouth and ending at the anus. Its primary function is the digestion of food and it contains many special glands and organs to help it to do this. It is a complex structure and its normal function can be severely impaired in children with MPS.

Mastication

This is the process by which food is fragmented and prepared for swallowing. It is mainly a voluntary process but the control of highly coordinated movements are reflexively initiated.

The jaws and teeth are responsible for mastication and can produce a force equivalent to a 90 kg. weight on the first molar of an adult. In MPS, gum disease, dental caries and malocclusion of the teeth can reduce the efficiency of mastication, which is further handicapped by the loss of coordination later in the disease.

Swallowing.

Swallowing is the process by which food is conveyed from the mouth to the stomach. Adults swallow approximately 600 times in 24 hours and are largely unaware of this important activity. Swallowing frequency is greatly reduced during sleep as salivary secretion almost ceases during the night. Normal saliva production is around 600/800mls. in 24 hours. This becomes a significant problem if the swallowing mechanism is no longer intact.

Swallowing can be divided into three phrases.

- **Oral**
- **Pharyngeal**
- **Oesophageal**

The oral phase depends on the tongue pushing the "bolus" of food to the back of the mouth. The food enters the pharynx and then moves into the upper oesophagus. Once the food is in the pharynx it reaches a "point of no return". The oral and pharyngeal phases are highly coordinated and involve some twenty five muscle groups and ten nerves working in harmony to produce normal swallowing. It is easy to see how this can become disrupted in children with degenerative disease of the nervous system like MPS.

Safety mechanisms

In addition to the movements of food to the oesophagus, a number of safety mechanisms are brought into play to prevent food entering the nose or windpipe (trachea) rather than the oesophagus. If these mechanisms fail, a common end result is nasal regurgitation and "choking" on feeding. These problems are both commonly seen later on in various MPS disorders.

The oesophagus

The oesophagus is the only part of the gastrointestinal tract that has neither absorptive or digestive functions. Its sole role is to convey food from the pharynx to the stomach. It has control valves (sphincters) at each end and propels food by using a "snake-like" movement (peristalsis) from top to bottom.

The Stomach

The stomach acts as a reservoir for ingested food. Its functions include:

- **Storage, mixing and controlled emptying of food.**
- **Acid secretion.**
- **Vitamin co-factor synthesis**
- **Initiation of protein and fat digestion.**
- **Synthesis of hormones.**
- **Absorption of iron.**

The stomach contains a "pacemaker" which tells it when to contract. The rate at which it empties depends on the nature and volume of the food.

The small bowel.

The small bowel is made up of different areas. The first part (the duodenum) neutralizes stomach acid and presents the food to the next part (the jejunum), where it is finally digested. The surface area of the small bowel is greatly increased by the "finger- like" projections on its surface (villi).

The large bowel.

The large bowel absorbs water from the intestinal contents prior to their excretion. Bowel motility is largely under involuntary control via the "autonomic" nervous system, except at the point of exit, the anal sphincter.

Specific MPS problems:

- **Mastication.**

Many sufferers from MPS are prone to gum disease, gum hyperplasia, large tongue (hyperglossia). They also tend to breathe through their mouths. All these problems impair mastication. Dental caries can be an additional problem.

- **Swallowing.**

Lack of coordination in swallowing is a common feature of many of the MPS disorders during the later stages of the illness.

- **Large and small bowel motility.**

Decreased transit time of the contents of the stomach through the bowel and the resulting diarrhoea are very common problems, particularly in MPS II and III.

As the disorders progress, immobility, inadequate fluid intake and poor diet make constipation very common.

Treatment:

- **Mastication and swallowing.**

Food texture and consistency has to be altered as the disease progresses. In many children nasogastric tubes or gastrostomies become inevitable.

Saliva production is a positive handicap in many children at this stage. **"Scopaderm"** patches have been used to inhibit saliva production and can often lead to a marked reduction of "choking" episodes during feeding. Unfortunately scopaderm is no longer readily available and has to be prescribed on a "named patient basis".

HOME TRUTHS AND REFLECTIONS

from The Isle of Wight

by **Mary Tubb.**

Congratulations to Mary Moulding on her slim book of poems which is selling like hot cakes on the Island. Mary's daughter Pam who suffered from Maroteaux Lamy disease sadly died six years ago. Her first husband Neville Tubb died ten years ago and she married a childhood friend, Tony Moulding in May this year. Copies of her book can be obtained at £1.20 plus 20p postage from

Mary Moulding

23A Princes Esplanade

Gurnard, Cowes, Isle of Wight.

Ph: 0983 200354

Thank you Mary for your generous donation!

Diarrhoea

Diarrhoea can be managed by a combination of diet and drugs. A number of families recognise "trigger" foods which precipitate bouts. By avoiding these they can succeed in reducing the diarrhoea.

Where this approach is unsuccessful drugs can be used, once constipation with overflow has been excluded. **Immodium** (loperamide hydrochloride) is the preparation that I have found most useful. It binds to the receptors in the bowel wall responsible for the bowel's movement. Overdosing can lead to constipation and a severe overdose can depress respiration (causes difficulty in breathing). Severe overdoses, which are usually accidental, can be treated with a specific antidote.

Constipation.

Constipation can sometimes be treated simply by increasing fluid intake or by altering the diet. Often drugs are required and there are many available:

- **Osmotic purgatives - e.g. lactulose, is the safest initial preparation. It's major side effect is to produce flatulence and diarrhoea will result if the dosage is grossly overestimated.**
- **Bulking agents - e.g. normacol.**
- **Lubricants - e.g. liquid paraffin.**
- **Large bowel irritants - e.g. senna.**

If oral therapy is impractical or unsuccessful, resort to suppository (e.g. glycerin) or enemas will be necessary.

Conclusion

As may be expected when dealing with a complicated structure such as the gastrointestinal tract, many problems are encountered in MPS children. Fortunately most have an effective therapy which, if promptly and appropriately supplied, can lead to an improvement in quality of life for the child.

MPS Research Database

In July the Society was awarded a grant to set up and develop an international database of MPS sufferers. We already keep details of all MPS sufferers and families in Britain who have contacted the Society. This new database will include more detailed clinical information.

To get this information we need to complete a new questionnaire for each sufferer. Information will only be collected with the agreement of families and of patients, where they are able to agree. It is planned that Christine Lavery and Mary Gardiner contact most families personally either by visiting them or at meetings or by telephone.

Some of the medical information needed may not be known by you. We will be asking your permission to approach your doctor for this.

The information gathered will only be used for statistical purposes and we stress again that no information held by the Society about individuals will ever be passed on without the express agreement of the persons concerned.

The overall aim of the study is to gain as precise a picture as possible of the incidence and spread of MPS diseases and their clinical variations throughout the world. We consider this will be key information in assessing the possibility of development of treatment in the future.

If you have any questions or concerns about this, please contact Christine Lavery at the MPS office.

Our grateful thanks and acknowledgements to all those who fundraised on behalf of the Society. Without you the work of the Society would not be able to continue.

Mrs Lakeman, Plymouth	London Marathon
Alan & Fiona Byrne, Glasgow	Dance & Raffle
Alan & Amy Bottrell, Glasgow	Saving 20ps
St. Andrew's Fellowship, Glasgow	Donation
British Steel, Glasgow,	Donation
Calderwood School, Glasgow	Bring and Buy Sale
Stonelaw High School (PTE), Glasgow	Raffle
Haylie Hotel, Largs	Hogmonay Raffle
Telethon, Scottish Application	Donation
Mr Koeglmeier, London	London Marathon
Mr & Mrs Gooch, Kent	London Marathon
Sid Shiff, Family & Friends, L'pool	Collection and sale
Mrs Graham, Dumfries	London Marathon
Pam Croghan, Stockport	" "
Mr Morris, Aberystwyth	" "
Mr & Mrs Bullimore, Malaga, Spain	" "
Mrs Allen, Birmingham	" "
Messrs Lindleys, London	City of London Road Race
Mrs Seymour, London	" " " "
Adam Ridley, London	" " " "
Hill Taylor Dickinson, London	" " " "
H T D, London	" " " "
Mr Webb, Mr Bleasby & Mr Reynolds	" " " "
Gary Martin & Michelle Lucking	" " " "
Amanda Colgate, London	" " " "
C Pitman-Jones, London	" " " "
Julie Meade & Carl Annison	" " " "
Cora Joscelyn & Paul Annison	" " " "
Cheryl Hughes	" " " "
Nicki Smithers, Dorking	Sponsored Step Class
Ruth Brozaitis, Bristol	Bath Marathon
Elaine Prangnell-White, Selby	London Marathon
Mrs Hosking, Norfolk	" "
G Waldron, Solihull	" "
Dr. Milton, Swansea	" "
Mr & Mrs Murray, Scotland	" "
Linda & Ron Snack, Milton Keynes	" "
Carol Westland, Reading	" "
Mrs Turner, Bristol	" "
Mr & Mrs Puddy, Shepton Mallett	" "
R Strawbridge, Swansea	" "
Helen O'Toole, London	" "
Gay Hosking, Dyfed	" "
Mr & Mrs Mort, Swansea	" "
D Middleton, Milton Keynes	" "
Abbey National, Matched Funding	" "
Mrs Jowett, Chepstow	" "
Mr & Mrs Patrick, London	" "
N Parker, Jersey	" "
T James, Bristol	" "
St Andrews Club, Hull	Sponsored Run
Avon County Transport Fund	Donation
Arthurlie Church, Glasgow	In lieu of design of Group Emblem
North West Area Family Day	Raffle
Sid Shiff, Family and Friends	Collection Boxes and sales

Our grateful thanks and acknowledgements to all those who fundraised on behalf of the Society. Without you the work of the Society would not be able to continue.

Lee Jennings and Martin Keogh	270 mile sponsored canoe trip
Edward Nowell	Edward Nowell Appeal
Mr and Mrs C Grandidge	Saved 20p Coins
John Bolton and Peter Snelgrove	60 mile sponsored cycle race
Mr and Mrs K Ballard	Sale of goods at Conference
Mrs R Nurse	Fundraised
Spiffing Stationary	Fundraised
The University of Buckingham	Rag Week
Mrs M A Moulding	Book of Poems
Mr and Mrs Gooch	Fundraised
Alison Coles	Fundraised
Derek Denham	Fundraised
M & E Bin Hire	Fundraised
Peter & Sue Stuart	Sale of Jessica Rabbit badges
Sarah Sampson	24 hour sponsored fast
Mrs K Jordan	Fundraised
Ruth Payne	Fundraised
Tanya Denyer	Sponsored Slim
Pauline Mahon and Jo Woods	Baldwins Charity Dinner, Sheffield
Andy & Jenny Hardy	Coffee and Doughnuts Morning
Kieran Houston	Northern Ireland Golf Tournament
Amersham College	Focus-Pre Voc Group
Mr Cedric Gooch	Mental Health Flag Day
Mr and Mrs S Fraser	Gave Talks
Mrs K Murphy	Gave talk on MPS
Alan and Fiona Byrne	Dance and Raffle
Alan and Amy Bottrell	Saving 20ps
Calderwood School	Bring and Buy Sale
Stonelaw High School	Raffle
Haylie Hotel	Hogmany Raffle
Sid Shiff,family and friends	Collection and sale
Nicki Smithers	Sponsored Step Class
Ruth Brozaitis	Bath Marathon
St. Andrews Club	Sponsored Run
Arthurlie Church	In lieu of design of Group Emblem
North West Area Family Day	Raffle
Billy Ingham	Collection Box
Paula and James Connelly	Dance
John and Carol Westland	Folk Music afternoon
Mrs Todd	Collection Box
MPS Family Holiday	Raffles
20 Over Cricket Team	Raised
ARC Ltd	Collected
Billingham Rugby Club	Triathlon
Mr & Mrs Deacon	Garden party
Sheila Benbow & friends	Tea Bar at Royden Park
Miss N Ward	5km City Centre Run
Sara Rickard	Sponsorship of Canoe Ride
Scott and Stuart McNee	Glasgow Run
Perth Prison Officers	Walking the West Highland Way
R Williams	Rhondda Fun Run
Silk Cut	Cash back

Area Support Families

Martine and John Brennan.....**Tel: 0524 382164**
105 Barley Cop Lane, Lancaster, Lancs.

Robert and Caroline Fisher .....**Tel: 0799 86631**
The Horrells, Great Samford, Saffron Walden, Essex, CB10 2RL

Suzanne and Jeffrey Hodgetts.....**Tel: 0827 56363**
6, Godolphin, Tamworth, Staffs. B79 7UF

John and Barbara Arrowsmith .....**Tel: 091 2662999**
11 Penfold Close, Fairways Est. Benton, Newcastle on Tyne.

Sean and Pauline Mahon......**Tel: 0742 304069**
41 Stumperlowe Crescent Rd, Sheffield 10, South Yorkshire S10 3PR.

Mary and Robin Gooch......**Tel: 0435 883329**
Highbank House, Swifehill, Broadoak, Nr Heathfield, East Sussex, TW21 8XG

Bill and Sylvia Blackburn.....**Tel: 0270 626809**
11 Beatty Rd, Nantwich, Cheshire, CW5 5JP.

Ron and Linda Snack.....**Tel: 0908 666819**
16 Wandsworth Place, Bradwell Common, Milton Keynes, Bucks. MK13 8BT

Tony and Shirley Eyre .....**Tel: 0934 834537**
6 Westway Park, Yatton, Near Bristol, Avon. BS19 4JU.

Ann, Michael and Sarah Kilvert,.....**Tel: 0686 624387**
Windy Waye, Nantoer, Newtown, Powys , SY16 1HH

Contact for Scottish Families:-

Alan and Fiona Byrne......**Tel: 041 6430034**
3 Jedburgh Ave, Rutherglen, Glasgow, G7 3EN.

Northern Ireland Co-ordinating Committee:-

Kieran Houston (Chairman) .....**Tel: 0504 884168**
15 Barrack Street, Strabane, Co. Tyrone, BT82 8HB.

Margaret Kearney (Secretary) .....**Tel: 026 57 62073**
12 Coleraine Rd, Ballycastle, Co. Antrim, BT54 6DU.