THE SOCIETY FOR MUCOPOLYSACCHARIDE DISEASES



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CHARLES O'TOOLE (As above)

This newsletter is published quarterly and distributed free of charge to sufferers of MPS disorders in Great Britain and, where they are children, to their immediate families. It is also distributed around the world on subscription. Copies are sent to those members of the medical profession who are directly concerned with MPS, many of whom make a donation towards our costs, for which we are most grateful. If you wish to be added to the circulation list, please contact Christine Lavery at the above address. Annual subscription charges are £5 in Great Britain and £10 Sterling elsewhere.



The Society for Mucopolysaccharide Diseases

30 Westwood Drive, Little Chalfont, Buckinghamshire. Telephone: (02404) 2789

The MPS Society is a voluntary support group, founded in 1982, which represents over 300 families in the UK with children or adults suffering from the mucopolysaccaride and related diseases. It is a registered charity, which is entirely supported by voluntary contributions raised by its members and it is 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. The Society provides an information service for families and professionals. At the present time it funds a Consultant Paediatric Post at the Manchester Children's Hospital. It maintain links with sister societies in Europe, the United States, Canada, South Africa and Australia.

In most cases there is at present little treatment for MPS Diseases, but much can be done to improve the care of sufferers. The slogan of the Society is:

"Care Today, Hope Tomorrow".

In the last newsletter Christine welcomed me as the new Chairman of the Management Committee. For those who do not know us, we are what you might call original members of the Society and have been at all the conferences starting with Birmingham 1983. "Us" includes Judy my wife and our daughter Susan, and used to include our son Michael who had Hunter disease. He was at both the Birmingham and the Harrowgate conferences but died in 1985 shortly before the first conference at Heathrow.

At this moment I just want to emphasise one thing, and that is that this is Your Society. If you have any views or ideas on what directions the Society should take please let the Committee know. Among our members we have people with many different skills - again if you feel your particular expertise can be of use to the Society please do let us know. Drop me a line or give me a call!

Alf G. King

23rd April 1989	London Marathon. 4 MPS Runners. Who are you sponsoring?
1st June 1989	Deadline for Summer Newsletter.
3rd June 1989	Northern Ireland Family Day. (contact Margaret Kearney)
16th July 1989	Dr Garrow's Garden Party (see notice)
5th August 1989	MPS Group Holiday (see notice)
22nd September1989	Annual Conference: Stoke on Trent.
31st August 1990	Second International MPS Conference. Manchester

DIDMI

Raymond and Jacqueline Baird, of Halifax, Yorkshire, a son Stephen Charles born 31st January 1989, weighing 61bs 9oz. A brother for Jonathan.

NEW FAMILY

Danny and Jackie Reade from Cheshunt, Hertfordshire, whose son Christopher is suffering from Hunter disease.

DEATHS

Anthony Bayliss aged 3 from Fazakerley, Liverpool, who had Morquio disease, died unexpectedly on the 27th February 1989 after an operation. Our sympathies to Julie his mother, Diane and Bill his grandparents and to the rest of the Bayliss family.

Gareth Carden-Edwards from Colchester, Essex, who suffered from Hunter disease, died on the 10th ofMarch, aged 7 years. Our sympathies to his mother Hilary and to all his family.

Toni Bramford, who died on the 13th of December 1988 was eleven years and ten months and not as stated in the Winter Newsletter. She suffered from Sanfilippo disease. Our apologies to her family for the error.

FAMILY CONFERENCE AND INTERNATIONAL SYMPOSIUM ON MUCOPOLSACCHARIDOSES AND MUCOLIPIDOSES

31 AUGUST - 3 SEPTEMBER 1990

UNIVERSITY OF MANCHESTER, INSTITUTE OF SCIENCE AND TECHNOLOGY

As some of you will know the Society is in the early stages of making arrangements for our 1990 Conference at UMIST. we are hoping that this event will put the Society firmly on the map by bringing together MPS families and interested professionals from all over the world. Nevertheless in the tradition of past MPS Family Conferences we intend to make this event as family orientated as possible whilst at the same time hosting a major Symposium for interested professionals.

The Conference and catering facilities are booked and we will be offering two grades of accommodation ie. Family Units within the University or Family Rooms at the Chandos and Brittania Hotels located very nearby. Parents are invited to join the professionals on the Saturday at their meeting to hear presentations on the Clinical Management of these diseases including coping on a day to day level. We are hoping that new initiatives will come from these papers and I know it will be interesting to learn how attitudes to the management of our children varies from country to country.

On the Sunday we are organising a Parent Conference whilst the scientists are left to unscramble the genetical and biochemical aspects of MPS and ML.

The Conference will close at midday on Monday following discussions on the treatment of MPS children. We anticipate the papers will include Bone Marrow Transplant, Enzyme Replacement and Orthopaedic Procedures. Parents will be welcome to observe during this session.

Throughout special attention will be given to our children, MPS, ML and brothers and sisters, and a full programme of activities and creche will be organised under the careful eye of our trained volunteers.

The evening activities will provide families and professionals alike with ample opportunity to meet in an informal and relaxed atmosphere whilst safe in the knowledge that their children are being safely cared for.

Apart from wishing to keep everyone informed of our plans I would like to ask your help in raising the funds needed to subsidise the family accommodation and board, and the childrens' activities. As we have in the past, we intend to subsidise the costs so as to enable as many families as possible to afford to come. Because we are expecting many more families than usual to attend we need to double our conference budget for 1990 and hope that much of this money will come from Companies and industry. We have already made some approaches in this direction and £7,000 has already been pledged. Nevertheless to be able to proceed with comfort we need another £20,000. I would be very grateful if at least 20 mums or dads who work for companies who make charitable donations (most do) would let me know for whom they work so that supported by the employee, the Society can make the appropriate approach. If 20 companies each gave £1,000 we would be home and dry.

Just send your supporting letter stating the benefits and importance to you as a family that this event should take place and we will do the rest. If you would like to know more please contact me on 02404 2789 or leave a message with Linda or the answer machine and I will call you back.

Looking forward to hearing from you.

Christine



RESPITE CARE PROJECT

On behalf of Pauline Pruce, who is coordinating this research, I would like to thank all the families who so kindly took the time to sit down and complete what undoubtedly was a time consuming questionnaire. I am sure that you will be pleased to learn that 77 families have responded and we are hoping that those of you who haven, t as yet will as soon as possible. It maybe that you felt some of the sections of the questionnaire were not relevant. We appreciate this but collectively your answers, or non answers as the case may be will prove most useful.

Already the preliminary data is showing strong trends, not least the need for respite care comparible to the standards set by Helen House, Martin House and Acorns, at a more local level. Many families have stated that they do not use respite care locally because it does not suit the needs of their MPS child/children. Many of your answers could have been predicted but the value of this study is that it provides the Society with data and evidence that many local authorities may not be giving sufficient thought to the needs not just of MPS families but those of thousands of others families where a child is suffering from an often rare and degenerative condition.

It isn,t too late to return the questionnaire. If you need a replacement or failed to receive the original please do let me know. COPIES OF THE FINISHED REPORT WILL BE AVAILABLE TO PARTICIPATING FAMILIES and the findings published in a future newsletter.

Christine Lavery, 30 Westwood Drive, Little Chalfont, Bucks HP6 6RJ Tel: 02404 2789

FAMOUS MUG

We have two limited edition plates and one mug commemorating one million tons of coal being extracted from Bevercotes Colliery. If anyone knows someone who would like to make an offer for one or all of these items please call me.

Ron Snack

Last August, **Dr Ed Wraith** took up his post as a Consultant Paediatrician at the Willink Biochemical and Genetics Unit at the Royal Manchester Children's Hospital. This post is funded entirely by the Society for the first year and will be partially funded for the following two years until it is finally taken over by the District Health Authority. We have received generous donations from trusts totalling around £10,000 to date. Many families are working very hard to raise funds for the project but two in particular deserve a special mention.

Gemma Rollinson's parents are organising a sponsored cycle ride in June from St. Peter's Square in Rome to Scunthorpe. They are hoping eventually to present the Society with a cheque for £10,000. Wayne and Karen Hoather from Manchester, whose two small sons suffer from Hunter disease, have handed over substantial sums to the Society, including a number of Covenants. Keep up the good work, everyone; we may be not far off this year's total but we would all, including Dr. Wraith, feel more confident if we had next year's amount in hand!

Although this is called the Manchester Project, Dr. Wraith is very happy to see families from any part of the country whose GP or paediatrician wishes to refer them. Dr. Wraith tries to see MPS families in a special Friday clinic which gives them a chance to meet each other. Mary Gardiner aims to be there when she can to offer her support and to provide the sandwiches.

Note from Mary Gardiner

There have been a steady stream of new and familiar faces over the past few months at the Friday MPS clinic. For my part, it has been a pleasure to meet and chat with so many of you. For those who attended the clinic when I was not there, I'll catch you next time!

I know from these meetings how those of you who have seen Dr Wraith, especially those who came for the first time; benefitted from seeing him. Dr. Wraith is naturally charming and understanding, but what really matters is that he knows MPS and is committed to improving the care of our children.

Families have travelled from as far away as Norfolk, Somerset, Yorkshire and Ireland. I am beginning to understand the geography of Britain, something that has evaded me for the past thirty years! The things we learn through the MPS Society!

mary	Gardiner					
****	*****	******	******	*******	*******	******

AREA FAMILY PROFILE

Sean and Pauline Mahon.

(Yorkshire, Lincolnshire, Humberside and Nottinghamshire)

We are Sean and Pauline Mahon and we live in Sheffield which is fairly central to the area we cover. Sean is a chartered accountant for a national and worldwide firm of accountants. I keep myself busy with the house, children, some voluntary work at a special school, MPS and various other activities.

We have three children. Sean is seventeen years old and doing A levels, Victoria is sixteen and doing GCSE and Siobhan is thirteen and a half. Our fourth child Anne-Marie had Hurler disease and died aged two years two months following a bone marrow transplant.

We learnt a lot about the Society from around the time of Anne-Marie's diagnosis in 1984 but didn't meet anyone until the 1985 September conference just three months before she died. We all know what a tremendous shock it is to find that your much loved child has MPS but we found that being with people who understood and experienced the feeling we were going through was a great comfort and support, even though it didn't lessen the pain and the grief.

These children have made such an impact on all of our lives and when I'm flagging I only have to think of Anne-Marie and that helps me pick myself up and get on with doing something good and worthwhile to help all the other children who are still suffering with MPS.

So far we've only had one Area get together since we took over as Area Family in 1987 but I hope 1989 will be different. Quite a few people in the Area seem to be doing their own particular fund raising; it would be nice if a few families could get together and find a way of enjoying themselves and raising some money together.

If any family would like us to visit them or would like to chat on the phone, please contact me, I'd be delighted to hear from you.

Pauline and Sean Mahon

In November or December this year we are hoping to hold a giant Tombola in Milton Keynes city centre. If anyone has any items they think would be suitable please bring them to the conference or call in to see Linda and I at Milton Keynes. If you would like to come along and help please call me.

Ron Snack

Brave Chris strikes out for glory

BONE disease boy Chris Isaac has learnt to swim - an astonishing achievement for the Fiddington voungster who 18 months ago was languishing in a surgeon's steel cage.

Brave Chris, who suffers adding to the distance from a rare disease which and his achievement. has stopped him growing, swam a length of Burnham "It's the first-time the club special help from dedicated members of Bridgwater very grateful to them." Swimming Club.

The eight-year-old had tion." to beat a number of hurdles before he could

strokes to the edge of the efeet tall. pool, and about three! weeks ago he did a width." said Mrs Lock

Last week he was taken to the deep end, and, with experienced swimmer David Lock at our forgoty one

Sean and

Pauline

1988

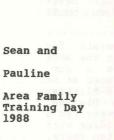
pool after receiving has taken on somebody so handicapped, and we are

Mrs Lock added: "Chris Club secretary Yvonne needs to be handled very Lock said: "We were abso- gently, but he's proved lutely amazed when he himself. His beaming did so well. It was incred- smile showed how pleased ible. A huge cheer went he was with himself. It up when he completed the shows what can be achieved with care and atten-

• The Mercury and Gazette featured Chris in, throw away his water his "cage of hope" in Aulem was the pressure on his lungs which made pressure was the device to his lungs which made pressure was the device to his lungs which made pressure and the device to his lungs which made pressure and the device to his lungs which made his lungs which was a like the device to his lungs which was a like the device to his lungs which was the device to his lungs which was the lungs which was the pressure of the lungs which was the lungs whi breathing very difficult. four months after sur-Chris was given individ- geons performed a deliual attention by the club care bone transplant oper for his first 14 weeks with ation. The surgery was them. "Eventually after successful, but Chris will many weeks he did a few never be more than three



Bone disease youngster, Chris Isaac is raised aloft by Bridgwater Club swimmers Louise Spooner and David Lock afte striking out for glory at Burnham Swimming Pool. Full story Page 20.





On reflection Peter's problems didn't become obvious until it was time for him to start at the local primary school. In some ways Peter had been a quick learner, knowing his colours and chatting twenty to the dozen. Even so it worried me that I couldn't teach Peter to write his name and that he failed to respond to simple commands. I used to take him to coffee mornings. The other children would stop touching the ornaments when told to. Not Peter, he would just carry on touching and so earned the name of "fingers Benbow".

The time it really dawned on me that all was not well with Peter was when we were trying to cross the road to the cinema. Peter was nearly five and we were going to see "Chitty Chitty Bang Bang" when he threw an uncontrollable tantrum. We had become accustomed to Peter's difficult behaviour but this was quite the worst we had encountered so far. From then on Peter's behaviour continued to deteriorate. By this time our younger son Martin was two and a half years old. Although Peter wanted to help him, if you gave him an instruction to fetch something he was unable to carry it out.

Peter started at ordinary school, but teachers found his unpredictable behaviour and lack of self discipline and concentration difficult to handle. Peter then changed to an assessment centre but escaped on a child's bike when the baker left the gate open. The next morning I was telephoned to come and collect him. Peter was expelled from school at the grand age of six!

Good news comes out of bad and Peter was offered a place at Neston Autistic Unit. There were only two children per teacher and only seven children at the centre and it was secure and safe for Peter.

Still no one had offered any solution to undertanding Peter's behaviour. He was repeating the same questions over and over again. He hated a change in routine as much as he hated hospitals. He insisted that our route to the shops never differed and watched Arrowe Park Hospital being built proclaiming "No hospital, no hospital".

By the time Peter was seven years old we were desperate for answers. Peter's balance was deteriorating, but the most worrying problem was his diarrhea. Often nine times a day. At about this time we took Peter to see Dr Vernon Jones at St Catherine's Hospital. Four days after Peter's tenth birthday we were told of Peter's diagnosis. Sanfillipo Disease. Initially there was a sense of relief that at last Peter had a label. Shock followed when we were told that children with this condition don't usually live beyond the age of ten years. (This information was innacurate: Editor). Following the diagnosis Peter went to Woodvale, a unit for autistic children and this meets his needs very well.

When it was time for Peter to go to Adult Training Centre he had calmed down considerably and the worry of him escaping and coming to harm subsided. Now, at twenty four, Peter still goes daily to the ATC where he is in special care. It was only fourteen months ago that we surrendered to incontinence pads and even so we still take him to the toilet regularly; sometimes we catch him especially if we give him a magesine with lots of smiling faces. Peter likes this and if he laughs we seem to have more success! We have recently had a disabled toilet and walk in shower installed which certainly makes life easier. Peter can only manage the stairs if two adults lift him and he can only get down using two cushions under his bottom while furiously hitting me! Despite Peter's limitations he gets a lot of pleasure from the television, especially "Pippin Fort", and "Soldier Boys". He can still manage to feed himself his cornflakes with a spoon.

When Peter was eighteen he suffered two strokes down the left side but has since completely recovered from these. One thing we put down to Peter's longer than usual life expectancy is that he has never had a convulsion (fit) and generally keeps well.

We have now come a long way from those early days. Peter's brother Martin is 21 now, a sales executive for Nissan and engaged to be married. From time to time we get to go out as a couple. Besides Martin and his fiance we are able to depend on Wirral Crossroads Scheme to help with caring for Peter in our own home. They are marvellous with him.

There have been very bad times, like the occasion when the Consultant wanted Peter admitted to Greaves Hall (a psychiatric hospital) but that was a red rag to a bull and made us more determined to care for Peter within the family.

Peter has brought us an awful lot of joy and we don't know what we would have done without him. Even our friends think the world of him. If there are regrets, perhaps it is the wish that we had more time to spend with Martin when he was younger when perhaps we concentrated too hard on getting through the day with Peter.

Sheila	and	Ken	Benbow,	as	told	to	Christine	Lavery.

ADVANCE NOTICE!!!
The Society has secured a slot for a radio appeal.
Sunday 17th September, Radio 4, 8.50am
"This week's good cause".

THE RED-HAIRED PRINCESS AND THE GENTLE GIANT

Have you noticed how fairy tales always end, "... and they lived happily ever after." Of course as a child I believed and expected such an ending, without question. But as a cynical 45 year old I think this kind of story ending is a cop-out. If there are no worries and concerns to mar domestic bliss, how can you measure the happy times and moments?

Whether in a fairy tale or in real life a red-head and a gentle giant sound a very powerful combination and you might expect that anywhere they went the effects would be immediately obvious, and noisy. Dan does of course have a booming laugh. (Yes, if you are a regular attender of MPS events you will know immediately that I am writing about Sue and Dan Butler). There the similarity ends. It has puzzled me for the last six years how the Butlers can do so much for the Society in such an unassuming way. I can't think of many important Society milestones where their contribution has not only been significant but often essential to the successful outcome to a major problem or project. Yet their touch has always been light and discreet. They surface when needed and then merge into the background.

Sue has been a member of the Management Committee from the earliest days. You will know her for a variety of roles, including Sales Coordinator and Overseas Liaison. Sue has also arranged at least one major fundraising event each year, including some very good fun themed-fancy dress parties/dances in their local village in Chinnor, Buckinghamshire.

Dan is the proprietor of BBW Transport, a small (at present) go anywhere, do anything freight company. We have used BBW often, to go anywhere and do anything, and although the Society offers Dan payment we are not very successful. It is not that we wish to exploit him for free (after all he has a business to run). Just that Dan and his staff really know about MPS. His compassion, and humour and his reliability are so reassuring. We want the best for our families. Dan and Sue deliver.

The Butlers and their specialist business experience have got us quickly out of difficulty, eg taking care of import documentation for special halos (Morquio treatment) from the United States, a large consignment of Jersy carnations for Lady Mountbatten's event in Tenterden. Our conferences over the last four years have been much improved by Dan and his brother Paul, who have engineered the audio-visual and discotheque.

Sue and Dan did once have a starring role when they represented the Society at the first Canadian Conference in Winnipeg in 1986. There are other stories, some of them sad, others happy and a few hair raising, that I am unable to write about because the details are confidential to one or two people on the Management Committee, to the Butlers and to the families concermed.

Above all the Butlers are important to The Society for themselves as an MPS family. Alexander, his Mum and Dad and Lucinda and Grand Mum are a particular inspiration to <u>Hunter</u> families and the rest of us as well. At 15 Alex is challenging the world and confounding the usual run of Hunter prognosis. Yet Alex and his family seem to find so much

time to shoulder and solve others' problems, and in a very cheerful way.

Grey clouds come and go so I do hope Sue and Dan are having their fair share of happiness-ever-after too. They deserve it, and more.

Robin Lavery

Incontinence supplies inadequate

Parents of children who are incontinent are being denied adequate provision, according to the Association of Continence Advisers.

"Parents are often left uninformed of what facilities they are entitled to, and the Government has been very woolly about the regulations," said Helen White, a continence adviser in north east England.

On average, 70 children per health area are incontinent, but standards of provision for them differ wildly between regions.

In Bloomsbury, North London, there are four continence advisers running clinics and assessments for children.

In neighbouring Hampstead, however, there is only one adviser who sees no patients and deals mainly with stores and supplies.

The Department of Health maintains that children who are registered disabled and are in full-time education are entitled to an adequate supply of equipment from their local health service.

But Helen White stresses that this supply is infrequent and inadequate.

"There has been confusion between the health authority's power to provide equipment, and parents' entitlement to it."

"It's often a matter of budget," said Dorothy Mandelstam, chairman of the Association, "and some health authorities give incontinence a different priority."



HOUSE OF COMMONS

Extending Mobility Allowance

An amendment submitted by the Mobility Allowance Campaign (a group of voluntary organisations) was moved by John Battle (Lab) at the committee stage of the Social Security Bill on 19 January.

It aimed to extend mobility allowance to people with a mental handicap whose behaviour is unpredictable and to people who are both deaf and blind.

Peter Thurnham (Con), supporting the amendment, said that in the light of cases produced by MENCAP and The Spastics Society, extra provision should be found to help people who have real problems in qualifying for the allowance.

Minister for Social Security Nicholas Scott was sympathetic to the cause, but said it was complex. It would be best to "await

the results of the OPCS survey and consider the matter comprehensively against the background of other changes to benefits that we might make at that stage".

The amendment was withdrawn. If any families are having difficulties obtaining adequate and suitable supplies of nappies please let us know.

If your child is mentally handicapped due to MPS disease and you are having difficulty in getting Mobility Allowance, please let us know.

Children with Special Educational Needs is the first detailed explanation of the new legislation for children with learning difficulties in Northern Ireland. Published by the Northern Ireland Council on Disability, the 72page book has a clear style and explains the legal changes, assessment procedures, statement contents and appeals advice, plus questions and answers for parents and sample letters, £1.50 + 52p p&p from NICD, 2 Annadale Avenue, Belfast BT7 3IR.



A quilt to grace any child's bedroom. From left Dominique Brooks-Daw, Mrs Pat Tozer, Mrs Irene Beech, Mrs Margaret Sunter, Mrs Judy Brooks-Daw, Mrs Jenny Wheaton, Mrs Michelle Brooks-Daw with baby Antoinette on her lap.

SOUTH Devon's Moor to Shore Patchwork and Quilting Group has made a fine applique quilt to raise funds for research into degenerative diseases.

The quilt is to be donated to the Society for Mucopolysaccharide Diseases who will put it in a national raffle. Last week it was pre-

sented to Mr and Mrs David Brooks-Daw, of Taunton, and will be taken to the MPS Parent Conference in London.

Mr and Mrs David Brooks-Daw's daughter, Alevique, died in October 1986, aged ten, of Hurlers Syndrome, and the quilt is in her memory.

It is intended it will travel round England, Scotland and Wales to area support families, to

ets. Moor to Shore hopes it will be possible for the quilt to return to Newton Abbot where it can be shown in an exhibition of quilts by members of the

About 300 hours went into the making of the quilt which began in May this year. It was made by Mesdames Judy Brooks-Saw, Mary Pearson, Mildred Marchington, Irene
Beech, Pat Tozer, make a donation to the Elizabeth Hawkins, Ceciley Coates, Margaret Sunter, Sue Edmunds, Jenny Wheaton and Barbara Paul.

Society for Mucopolysaccharide Diseases should send it to the society at 30 Westwood Drive, Little Chalfont Rucks

The patchwork quilt which was displayed at the annual conference in September is to be raffled in aid of MPS. The draw will take place in September 1989 at the next conference. The quilt has been presented to the Society by the "Moor to Shore Patchwork Quilting Group" of South Devon.

Anyone can help promote the draw by arranging with local stores to sell tickets on their premises. (Sainsbury's can be very profitable on a Saturday!) If you would like to display the quilt please let me know the dates you have in mind and I can have the quilt forwarded to you. Please write or phone me if you think you can sell some tickets.

David Brooks-Daw, St Mary's Lodge, Wellington Rd. Taunton, Somerset TAl 5AS Ph 0823 74661

MARK SIMPSON

Dear Friends,

Mark will be nine years old in July. He suffers from Morquio disease. He attends a special school, Northfield, which provides transport to and from school. Mark loves playing football but soon gets tired. He hates school holidays as he never sees any of his friends because they live all over York. He spends all day and most of the night playing with his Lego and he would love to go to Legoland. He also loves playing on the computer and also very much enjoys doing DIY with his Dad. Mark has an older sister Clare who is fifteen years old and is studying to be a nursury nurse for disabled As a back up she is doing a computer studies course. In her free time (when she gets it!) Clare sings in the local Church Choir and is a member of a Christian Youth Group.

I work part time in a local supermarket. I haven't much time for any hobbies but I enjoy gardening and helping Mark with his Lego.

With best wishes from Barry, Clare, Mark and Ruth Simpson. 14 Oak Tree Lane, Haxby, Yorks.





Hurler Disease takes its name from **Gertrud Hurler**, the doctor who described a boy and a girl with the condition in 1919. It has been estimated that 1 in 100,000 babies born in British Columbia would have Hurler Disease. The Society hopes to be able to publish British figures very soon, but based on the British Columbian figures one would expect seven Hurler babies to be born in Britain every year.

Children with Hurler disease lack an enzyme called alpha-L-iduronidase which is essential in cutting up the mucopolysaccarides called dermatan and heparan sulphate. The incompletely broken down mucopolysaccharides remain stored in cells in the body causing progressive damage. Hurler disease is caused by a recessive gene -- both parents are carriers and it is possible for prenatal diagnosis to be carried out.

Hurler babies may be quite large and may grow fast during the first few months, but growth will usually have stopped altogether by the age of three.

The brain of the Hurler sufferer becomes enlarged by storage of mucopolysaccarides. Sometimes the fluid which protects the brain from jolting is prevented from circulating and accumulates (hydrocephalus).

Most children will have nose, throat and chest problems. The chest is abnormally shaped and the tissue of the lungs becomes thickened so that it is hard for the lungs to work efficiently. Many children breathe very noisily at night.

Heart disease is common but may not develop or cause problems until later in life when drugs can be prescribed to relieve the condition. The liver and spleen are enlarged and hernias occur quite frequently.

The eyes are affected by corneal clouding and the ears are affected by both conductive and nerve deafness.

Sadly the intelligence is affected in Hurler disease and skills learned will eventually be lost. There is great variation in the severity of the condition and while some may learn to say only a few words, others learn to talk well and even to read a little.

Life expectancy is short and few Hurler sufferers live beyond twelve years. Hurler children are usually happy and friendly and much loved by all who know them. Jemma's diary of which another instalment is printed in this newsletter gives a delightful picture of a period in the life of a little Hurler girl.

This is an abridged version of leaflet which is being printed and which will shortly be sent to all Hurler families.

Mary O'Toole

(Extracts from a diary which travels between school and home with Jemma Corbett. Jemma is now six and has Hurler's disease.

October 7th 1986

Julia Bowtell, Gemma's teacher writes to Sarah who has just given birth to Elizabeth.

Thanks for the news. We are all delighted everything went so well. Jemma has told everyone she has a baby, even our school secretary when she took the register up. Our love to Sarah and a big cuddle for Elizabeth.

October 9th 1986

Absolutely no problems in school. Every so often reference is made to baby and we had a delightful example of Jemma knowing what's what. When Jackie asked a group where does milk come from Jemma piped up "Mummy", even though a cow and a bottle of milk were in the picture.

At long last we've had the children's hooks lowered to their height. Jemma has proved her self-help skills are all present. She came into school, greeted everyone and then went to hang her coat on the correct peg. She even managed to hook her suitcase up as well.

Today after a classroom chat using Makaton pictures about doing a poo, she set off and when asked where she was going she signed toilet, took herself out of the room, sat herself down and performed.

Later we had PE and she loved exploring the apparatus. Her favourite was walking up three steps along a platform and then launching herself onto a soft blue mattress. The 'launch' required a fingertip hold from us but that was all. I had thought she might be uncomfortable landing on her tummy but she came up giggling and laughing.

Have a lovely weekend with calm days and quiet nights!

October 16th 1986

Sarah to Julia.

We were very amused to read about where milk comes from, I've not started to moo yet.

7th November 1986

Annabel Edwards (Speech Therapist) who is expecting a baby in January came for the last time today and was delighted with Jemma's progress. I fall about every time Jemma answers "All right", when asked to do something, and when I listen to her giving Colin a verbal clip round the ear while playing in the Wendy house. "Naughty boy!" and "Stop it!", who does she take after? Its now the place names that need to be understood, "Next to", or "Beside", "Under", "On", "Behind", "In front". It is a bit hit and miss at the moment, even when stress is put on the vowels, but it is coming. She is also matching colours well but when asked to choose a named colour sometimes gets it right, sometimes wrong. I don't usually teach colours but she and Ashley seemed so close that for the next week we shall be working on RED. Any spare red items do send in. I'll send them back again later. Sorry about the dirty skirt but Jemma had a session of trying out various potties and little toilets and en route did a poo

and sat on her skirt. She is very good now and tells us each

12th November 1986

Sarah to Julia. | adam | martilles and bad an aware lead |

Jemma has been very good over the past few weeks using the toilet at home. In the mornings she sits on the loo seat before getting dressed. I usually ask her to poo and wee and on Moday she replied "I'll try", and did! I had to leave the bathroom it was so funny. I didn't want to upset her by laughing at her, she was so serious.

28th November 1986

The dentist looked at Jemmma's teeth and said they were fine. She has all her baby ones and may be ready to lose her front ones soon. Jemma has been marvellous this week, all her social skills are just fine. She's been given an impressive list of jobs because she is so capable. Her self help skills are coming along fine too, its just her little shape that gets in the way sometimes.

FUNDRAISING

Flag Days

We have held a flag day in Milton Keynes for the last three years and in 1988 we collected over £550, a good deal of money for very little work. All you need is permission from the local council by way of a license, a few helpful friends and some collecting tins. If anyone is interested in organising a flag day please get in touch and I will try to help in any way I can. Linda and I have found that MPS posters are a great help for flag days.

Ron Snack

Our first week back to routine and everyone breathes a sigh of relief. Could write a book about Jemma this week. She has been marking the register. She sits with a pencil in her hand and asks where is? Then she asks them if they want dinner. She gets the names in the same order as the register, hows that for memory? Then she writes a squiggle in the square I point out beside each child's name. As she is getting a bit precious about HER jobs I have asked each child to take a turn returning the register to Pat Wheeler. Jemma has learned to part with it but always give her own instructions. When it was her turn I hope you noticed Pat gave Jemma her own wage slip.

I've also tested Jemma's comprehension and listening skills with a lot of dollies, teddies, a cup, bed, spoon, dolls clothes etc. Without me signing Jemma picks up the right things and performed the correct tasks without me having to accentuate any part of my sentence.

Today we had PE and a real increase in confidence. She needed no coaxing, watched others and tried it out for herself. She asked me to hold her hand as she walked along an upside down form along a piece of wood no wider than one of her little feet. A good test of balance. She tried swinging from a ladder and lifted her feet an inch from the ground. She was highly amused with crawling through a tunnel.

February 10th 1987

Life is all one 'allright' or 'ok' from Jemma. Everyone wishes she was in their class and to be quite honest she is so good at keeping everyone on their toes in our class. I shall soon be out of a job.

Today for cooking she collected all the aprons together and brought the same one each child has for dinner. Then she helped me get the bowls, scales, patty tins and spoons with baking cases, all good fine finger work, and had time to tell Ashley he was naughty because he squashed his. Next she weighed, beat, stirred, and told Ashley to get his fingers out of the mixture. Then she gave instructions to Nazim and had time to tell Ian not to scatter the flour.

Apologies, we all ate rather a lot of raisins waiting for the cake to ${\sf cook}$ - while Jemma was telling Ian off for pinching her raisins.

Jemma also helped me to wipe up all the bowls and put everything back. She was so good it was tempting to let her do her little woman bit but work has to be shared out so I let her chat while the boys did a bit of tidying up. Women's Lib in the nursery.

Feb 10th (continued)

Unfortunately it was all marred a little because while Jemma was holding the door open for me the door closed on her fingers. She seemed fine after a good wail and soaking the fingers in witch hazel.

Also it was partially my fault that she had dirty knickers. The school Governers came round and while we were chatting Jemma took herself off to the toilet. It was when she came back with the knicks at half mast that I realized she was in a pickle. Never mind, I thought she coped beautifully.

February 11th 1987

Sarah to Julia And Andrew Sarah to Julia Sarah to Julia

Have you noticed a strong smell of Nivea Gift Set contents following Jemma this week? As if she needed anything else to add to her morning ablutions! She takes so long to wash, clean her teeth, sit on the loo, comb hair, twirl in front of the mirror, put on perfume, take vitamin drops, instruct me on her hair ties for the day, inspect the style, choose clothes etc. She was thrilled with her raffle prize and if we touch it we are told it's Jemma's.

Mick and I have a new title --Corrrbit. We have told Jemma she is a Corrrbit too, much to her amusement, and she thinks baby Corrrbit is hysterical. Have you been talking surnames at school?

Letters are this week's topic. Yesteday she had to buy a postcard, draw on it, put it in an envelope, stick a stamp on it and then walk to the postbox and send it to Grandma. This morning while waiting for the bus, the postman who thought he was being kind by giving a parcel for Lizzy was given a very strong talking to by Jemma about her drawing for Grandma and Postman Pat!

February 12th 1987

The 'Corrrbit' bit. Since September one of our rituals has been to ask each child "What is your name?" Those who can have been prompted to answer surname too. It was my insurance should any parent be so careless as to mislay an offspring.

Thanks for explaining about Postman Pat. She started talking about it to Jackie who thought I had resurrected an old story about him while she was having coffee. Ironically the story I have is rather similar. It was about giving Grandma a parcel - and Jemma understood it almost word for word.

Many of our families know that the Society sells 'little things' to help advertise and at the same time make some money for MPS. For the benefit of new families we have on offer the following items:

MPS coloured pens	25p each
MPS pencils	15p each
Key fobs	50p each
Bookmarks	60p each

If you would like any of the above items we would prefer money to be sent with the order, it makes the book keeping easier.

We also have sweatshirts and T-shirts. The money for these needs to be sent with with the order, otherwise no T-shirts! The range of colours is as follows, Burgundy, Red, Royal Blue, Yellow, Pink. (Black or White by special request.)

The sweat shirts are available in

Sizes 24" to 32" Childrens	£7.00
Small, Medium, Large, X Large	£9.50
XX Large	£10.50

Please expect to wait three to four weeks for delivery, as we do not keep the adult sizes in stock. We do have some children's sizes in stock at the present time.

We have a number of white T-shirts with the Society's logo on the front in blue.

Sizes 22" to 32" £3.00 each 30p p/p

Adults, Medium, Large, Xlarge £4.50

If you have friends who are running in Marathons this year, don't forget the MPS running vest which is supplied free.

Please phone your orders on 024 026 3185 or write to Sue Butler, Spriggs Holly House, Chinnor Hill, Oxon.

If you get an article in a newspaper could you please try to get the reporter to put in a plug for our famous "Fit and Famous " Cook book @ £1.95 each plus 35p p/p. available from Sue Butler. Sue has sold 40 copies recently through one article. You too can buy them at this special price!

We hope to have a regular column of hints and tips, but this will depend on you sending them in! Editorial health warning: People send in hints about what they found useful for their child in their special circumstances. The newsletter takes no responsibility for the suggestions offered.

FRIDGE LOCKS can be obtained from B&Q Stores from around £1.50. This type has been fully tested by Kristina Briggs and has defeated all her attempts! (It is however no longer being manufactured). Boots sell another type, but Kristina found this flimsy and poor fitting and easy to open.

A RAIN HOOD for the Mc Laren Major Buggy with 7" wheels can be obtained from John Paxton Products Ltd, Unit 14, Stockton Close, Minworth Industrial Estate, Sutton Coldfield, West Midlands, B76 8DH. Cost £20.78 + p/p and VAT and £27.01 for the twin.

David and Monica Briggs

Sue Bramford offers the following tips from caring for her daughter Toni, who suffered from Sanfillipo disease and whose death was announced in the last newsletter.

To stop a child chewing clothes, put a bib on; it will be cheaper to replace than jumpers shirts or dresses.

Can't stop child falling out of bed? Why not consider letting the child sleep on a mattress on the floor? A word of caution...This may offer better support for the child's back, but think of your own back as well.

Have a good supply of soft toys with safety eyes nose and mouths, also Fisher Price toys, teethers, even cloth books.

Buy a two way intercom so you can hear if your child is crying or sleeping soundly.

If a child is very active during the night you may need to consider putting a lock on the outside of their bedroom door to prevent them wandering and perhaps falling down stairs.

FUNDRAISING

Collecting boxes in local shops or post offices are always a good way of raising money. A friendly word with the shopkeeper will usually ensure a good position for our 'tin'.

HINTS AND TIPS (Sue Bramford; continued)

Don't force feed. Give little and often. Don't let them know you are frustrated because they won't eat if they sense this.

If the child won't drink with you facing them, stand behind them (if in a wheelchair), tip chair back so the handles are either side of your hips and give them a drink slowly. If you hold the jaw with one hand and close the mouth after the first sip the child will swallow. (You will of course need to seek professional advice about managing persistent feeding problems).

If the child won't eat solids give "Build Up" either in a feeder cup or by spoonfeeding. You can get different flavours.

Talk to the child, call them by pet names and praise them when they eat. Make a game out of it.

When the child goes to bed at night give them and old terry nappy to chew on. It saves on bed linen and night clothes.

I hope these tips are of some use, and if you find them helpful please write and let me know. May I finally say a big thank you to those of you who wrote to Pete and myself in our recent sad loss. Your letters were very much appreciated.

Remember every time the sun shines Toni is smiling down on us and radiating warmth on us all.

Love to all of you,

Sue, Pete and girls.

Ode to Spring

With Spring and hopefully Summer just around the corner now is the time to think of booking an MPS stall at any local fetes that are taking place. Sue Butler has plenty of MPS items that you can sell. Tombolas are always popular at fetes and it is not difficult to get small prizes from local shops and businesses. As well as raising money and promoting more public awareness of our society you can also enjoy a great day out with your family - enjoyable fundraising!

We still have one or two coffee snowball bits available, so if you are interested and have a few friends who enjoy coffee mornings, please get in touch.

Ron Snack

MPS GROUP HOLIDAY 5th August 1989

Once again it is time to think of the Summer holidays, and the TV is showing far away places in the sun.

Well, there are still five places left on the MPS Group Holiday at Filey, Near Scarborough, Yorkshire, in August. We cannot quarantee the weather, but if last year is anything to go by, we can guarantee plenty of sunshine of the "human kind".

As you will have seen from the form sent out with the last newsletter, we have chosen one of the Haven holiday parks again. It really offers a great choice in activities yet provides scope for us to organise get-togethers like barbeques and days out etc. The site is mature and the staff are helpful and kind.

The MPS holiday was first thought of in 1983/84. was to bring families together to share each others company for a short time. It provides an opportunity for brothers and sisters of MPS children to make friends with each other and to enjoy being part of a group who understand and care.

The holiday is for everyone regardless of income or whether your MPS child is with you or not. As a bereaved family we have enjoyed being with all the parents and the children on these holidays and in many ways it has given us a lot of help.

If you feel your family would like to join in such a holiday, please return the form to me as soon as possible to secure one of the remaining places. Please note that the holiday is self catering.

Mary Gardiner Holiday Organiser

DR GARROW'S GARDEN PARTY

16th July 1989

12.00 noon for Buffet lunch

Roughwood Farmhouse, Roughwood Lane,

Chalfont St Giles, Buckinghamshire

We do hope that we will see as many families and extended families as possible at our Grand Garden Fayre. hired a marquee for the day and there will be plenty for the children to do. The grounds are extensive and on a fine day the swimming pool is a big success, so dont forget your The Bucks Advertiser is loaning us a giant bouncy castle and there will also be a magic show, donkey rides and sidestalls.

If you plan to bring your own stall, want to help in any way, or just relax to the sound of the Misbourne Orchestra and enjoy a leisurely day out in the countryside, (only two miles from Junction 17 or 18 on the M25), then a warm welcome awaits

Do let us know if you plan to come, and a map and programme will be enclosed with the Summer Newsletter.

GRANDPARENT CONTACT *********

Diane Bayliss the Society's Northern grandparent contact, has recently suffered the demastating loss of her grandchild Anthony. Anthony who was three years old had Morquio disease and had just undergone surgery to correct a defect in his cervical spine. Although the surgery itself seemed to have gone well something went wrong afterwards and Anthony died three days later in the arms of Diane and his mother Julie. There is to be an inquest and it is hoped that any lessons learned from Anthony's tragic death will be used to make the operation safer for others.

I know that everyone who has got to know Diane and her family since they joined the Society will share their sorrow. I am sure that Diane would appreciate letters from the grandparent contacts and I know you will understand that she may not be able to respond just yet.

Mary O'Toole

Brave fight by parents By Heather Bennett HEARTBROKEN parents Alison and Nick Pullin are coming to terms with a terrible wasting disease that terrible wasting disease that

terrible wasting disease that is killing their young daughter by raising money for research.

Abigail, aged five, was was a normal baby, just a little light, weighing just six pounds,

She was a cute toddler with a shock of unusual coarse bright blonde hair, and chubby face but these proved to be early symptoms of the disease which will soon claim her life.

Incurable -

A pre-school check revealed a few learning problems, and earlier this year the Pullins, of Poplar Way, Colerne, were told Abigail had the incurable genetic disorder

Mucopolysaccharide.
In the last six months Abigail has lost the ability to talk and eat with knife and fork - she is now incontinent, hyperactive and cannot sleep.

As the disease takes over her brain she will forget how to walk and be unable to breathe or digest her food properly.

Doctors at Manchester Children's Hospital have reasured Mrs Pullin, aged 34, and her plasterer husband that their daughter is blissfully unaware of her tragedy.
"She is probably quite happy,

the sorrow and the guilt comes from us," said Mrs Pullin. She said: "She went to

Middlefield special school in Chippenham a little star, but now she is incapable of learn-

She could die in just three years time, or live a dependent existance until 30, although the average age of victims is 14.
Their young son Tim, two, has

a one in three chance of being a carrier of the disease, although tests show he should not develop it himself.

There are 350 children in Great Britain suffering from the disease and parents are raising cash for research.

It was a chance in a million that the Pullins both had the killer gene, and they had no idea until Abigail was diagnosed.

Mrs Pullin's eldest daughter Katie, 15, by a previous marriage, is perfectly healthy. Abigail has the Sanfilippo

Syndrome of the illness which affects the nervous system.

Mr Pullin, 32, and his fellow players in Colerne Rugby Club are raising money for the research in a 24 hour sponsored darts, skittles, cribbage and

pool marathon on August 28-27 at the village Liberal Club. "I wanted to do this fund-raising because it is a positive aspect, I know there is no hope for Abigail but we can still help those yet to be born," said Alison.

Money raised will go to The Society for Mucopolysaccharide Diseases which funds the work of the Manchester hospital.

MPS RUNNERS DOMINATE MARATHON !!!!

There will now be four London Marathon runners sponsored by MPS on the 24th April, Roger Bennett, Ken Goss, Glyn Bush, and Charles Pike. There is still plenty of time to get a sponsorship form from Ron Snack. Go to it!

******************** FUNDRALSING EVENTS

Our thanks to all those who sent donations. These have all been acknowledged separately. As usual there have been many and various fund raising events held. We need to hold more to keep the money rollling in. Perhaps the following list will give you some ideas; personally I think that sponsored pub crawls are the best thing since sliced bread!

Ron Snack

J Eyre (Avon)

Sponsored Bike ride J R Townsend Sale of dried flowers P&JM Dagnell A&C Martin Pub Collection SR&V Harrison Sponsored Run Sponsored Dinner P Mahon Car Boot Sale and Walk JA Sweeney Sale of Stamps FA Sandon B Carol Sponsored Bike Ride Hythe Methodist Sunday School Concert Brennan family Charity Evening W Blackburn and Sponsored Swim E Mc Garry Xmas Party P&M Archard Raffle and Sales D Brookes Daw Sponsored fast and clothes sale P&M Archard C&R Lavery Xmas Party Two collection boxes C&M O Toole Coffee Morning M&P Short Disco and Raffle S&E Wallace R&J Macintyre Sponsored Caterpillar Walk. Jewellery Party Mrs Masters Collection Box at Boots Basildon J. Towning Garden Fayre J&E Broughton Downend Baptist Church Event. Crowborough Townswomen Guild Copper Collection Collection Boxes W Robins New Year Party and Collection Box A Byrne Stonelaw High School Sponsored Swim (Glasgow) Penny Warner, Newbury Garden Party Sue Butler Luncheon Party Raffle Carol Eastwood British Airways (Heathrow) Collecting box. Collection box Darkes family Sarah Lowry (Herts) Carol Singing Coins in a bottle Barry Lowry 24 hour Marathon Events Colerne Rugby FC Wetherby half Marathon W Fawcett (Yorks) J Winsley Coins in a bottle A Clamp (Derbys) Bingo Evening Christmas Activities Wirksworth Rotary Club 10k Road Race Harrogate Lions CS Smith (Somerset) Perfume Sales Valentine Dance P Isaac Sponsored Cycle Ride

AREA SUPPORT FAMILIES

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Alan and Deirdre Beavan Tumbleweed', West Gate Lane, Lubenham, Market Harborough, Leics. Tel: 0858 62182

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