Newsletter

The Society for Mucopolysaccharide Diseases

National Registered Charity No.287034

Spring 98

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The Society for Mucopolysaccharide Diseases

46 Woodside Road, Amersham, Buckinghamshire HP6 6AJ
Telephone: 01494 434156 Fax: 01494 434252

The MPS Society is a voluntary support group, founded in 1982, which represents over 900 families in the UK with children or adults suffering from Mucopolysaccharide and related diseases. It is a registered charity, entirely supported by voluntary donations and fund-raising, and run by the members themselves. Its aims are as follows:-

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 Great Britain 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 two specialist MPS clinics at the Royal Manchester Children's Hospital and at the Hospital for Sick Children, Great Ormond Street, London. The Society also funds research projects at the Christie Hospital, Manchester, Royal Manchester Children's Hospital, Bristol Children's Hospital and the Institute of Child Health, London. 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"

Front Cover:
Bilal Wali aged 8 who suffers from Hurler Disease with his sister and Cousin at the Manchester BMT Clinic.

CONTENTS

Deadline for the 1998 Summer Newsletter
26th June 1998

Please send us lots of photos

CHAIRMAN'S REPORT

Many of you will know that at the Management Committee meeting in January I became the new Chairman of the MPS Society. I was very honoured to have been asked to take this role and to have the unanimous support of my fellow Trustees. I am deeply committed to the work of the Society and will do my very best to support you all.

Many of you know my husband, Peter. Our son, Gethin, was diagnosed with MPS I (Hurler Syndrome) at Great Ormond Street in May 1980 when he was 10 months old. The MPS Society had not been founded then and we felt totally traumatised and isolated. Dr Rosemary Stephens was our consultant and she became our support, strength and friend. Our families were very caring but it was difficult to communicate to them and for them to understand such a complex subject as MPS! You will all know what I mean! I cannot describe the joy and relief of meeting, quite by chance, (the children were so alike) another MPS family in Guy's Hospital, and being told about Christine and her work in setting up the Society which was just starting then.

Sadly, Gethin died just after Christmas 1984 when he was 5½ years. He was our only child and we shared so many very happy times. We still miss him very much and now that he would have been 18, we wonder whether he would have been at University, another milestone in his life. Life is all about milestones and we shall always think what Gethin might have been.

I became involved with helping the Society first with some administration work and latterly as a member of the Management Committee and as a Trustee.

The most important news currently is that all the stops have been pulled out hasten the refurbishment of our new office premises in Amersham. We are indebted to the support we have received from a number of MPS families and we hope to move into the new office in late March - a very welcome relief for the staff in the present overcrowded office.

Joan Evans, Development Officer who has been a great strength to the Society leaves at the end of March to move to Germany with her husband, Geoff. We thank Joan very much for her loyalty and commitment and also Geoff who has provided invaluable support in setting up our computer systems. We wish them both a very happy time in Germany.

We welcome to the Southern office Anne Brockfield and Angela Ratcliffe and to the Northern officer Samantha Rymer.

The Training Weekend for Area Support Families took place 6th-8th March. A packed timetable included discussion groups on developing good support techniques and communication skills. It was a very successful event.

Negotiations with the Manchester Children's Hospital NHS Trust over the appointment of a third metabolic consultant to work with Doctors Ed Wraith and John Walter have been successfully concluded. The Trust has accepted the Society's offer of 50% of the funding for the post, over a period of three years. Thereafter the post becomes fully part of the Trust. The Society's funding contribution comes from the Jeans for Genes Appeal.

On that note, finally, the Jeans for Genes 1997 Appeal will total around £1.4 million. At the time of writing the MPS Society had received £200,000

Wilma Robins
18th March 1998

MILESTONES

New Families

I would like to apologise as I seem to have failed to include the Bailey family in the Winter newsletter. Nineteen month old, Olivia was diagnosed last year with Hurler Disease. Olivia lives in Wiltshire with her mum and dad, Claire and John.

Gail and Nicholas Barnett's daughter, Faye was recently diagnosed with Sanfilippo. Faye who lives in Birmingham is 4 years old.

Mrs Phillips from Surrey whose husband, Stuart recently died at the age of 75 years. Stuart suffered from Morquio Disease.

Mr and Mrs Home from Dorset were recently given a Hunter diagnosis for their son, Matthew. Matthew is aged 3 years.

Mr and Mrs Begum's daughter, Sannah was recently diagnosed with Sanfilippo Disease. Sannah who lives in Burnley is 3 years old.

Tina and Stuart Damen's daughter, Lorren was recently diagnosed with Sanfilippo Disease. Lorren who is 4 years old lives in Portsmouth.

Mrs Sandra Manion from West Midlands has recently contacted the Society. Sandra is an adult sufferer of Morquio Disease.

Karen and Robert McLean's son, Craig has been recently diagnosed with Sanfilippo Disease. Craig who is 4 years old lives in Liverpool.

Sylvia and Gilbert Watterson's daughter, Robyn has recently been diagnosed with Hurler Disease. Robyn who is 18 months old lives in Glasgow.

Lilly and Phil Whitely's daughter, Emma has recently been diagnosed with Sanfilippo Disease. Emma who lives in Manchester is 4 years old.

Barbara and Barry Wilson's daughter, Joanna has recently been diagnosed with Scheie Disease. Joanna who is 11 years old lives in Bromsgrove.

Ayshe and Enver Kilinc's son, Can has recently been diagnosed with Mannosidosis. Can who lives in Brixton is 3 years old.

Mr and Mrs Ali's son, Mohammed has recently been diagnosed as suffering from Morquio Disease. Six year old Mohammed lives in Glenrothies.

Elizabeth and Daniel Green's daughter, Daina has recently been diagnosed with Hurler Disease. Daina who lives in Winchester is one year and 10 months old.

MILESTONES

Deaths

We were sorry to learn of the death of James Astbury, aged 19 years on the 12th of January 1998. James who had a Bone Marrow Transplant in 1983 suffered from Sanfilippo Disease.

Sadly Christopher Rock, aged 20 years passed away on the 12th March 1998. Christopher suffered from Morquio Disease.

Sadly Waseem Ahmed from Glasgow died on the 16th March 1998. Waseem who was 15 years suffered from Maroteaux-Lamy.

Births

Congratulations to Clare and James Garthwaite on the birth of their daughter, Phoebe. Phoebe was born on the 9th of January 1998 and we are sure is being entertained by her brothers, Thomas and Louis.

Congratulations to Sandra and Ray Martin on the birth of their daughter, Amie Anna on the 21st February 1998 - a sister for Katie and Liza.

Congratulations to Mary and Andy Wragg on the birth of their son, George in March 1998. I expect Jacob is looking forward to playing with his new brother.

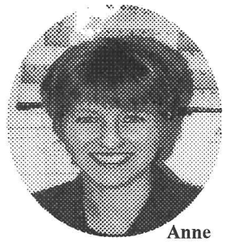
WE HAVE INCLUDED THIS PHOTOGRAPH FROM THE MPS ANNUAL CONFERENCE TO REMIND YOU ALL THAT BOOKINGS ARE COMING IN FOR THE 1998 CONFERENCE. SO IF YOU WOULD LIKE TO COME TO THIS YEAR'S CONFERENCE PLEASE BOOK SOON.

OFFICE NEWS

Changes to the MPS Amersham Office.

First of all the Amersham office will be moving to new premises from the first we of April 1998.

MPS Society
46 Woodside Road
Amersham
Bucks HP6 6AJ


Anne

There are also two new members of staff. Anne Brockfield who will be taking over the financial aspect of the day to day running of the office and Angela Ratcliffe.


Angela

Angela will be taking over much of the administration side of the work and liasing with the Area Support Families.

The work is going ahead to bring the new office up to a reasonable standard. Unfortunately the new office has come to late for me.

I am leaving the MPS Society at the end of March as my husband Geoff is taking up a new position in Germany.

I am very sorry to leave the Society and I will miss the daily contact with Area Support Families, families and children who have become very special to me over 4½ years.

Geoff and I would like to thank you all for the beautiful Waterford Crystal Glasses we were given as a leaving present. Every time we use these glasses we will think of the MPS Society. ***(Not that we drink, of course)***

I would like to thank lots of you personally but this newsletter does not have enough pages. So I would like to specifically thank Christine who over the years has supported and encouraged me and has taught me so much about the voluntary sector I hope to be able to use the skills I have developed in the future.

As this will be my last newsletter I would like to thank you all for the articles and photos that you have kindly sent (***some make us laugh and some make us cry).*** Sheila will be taking over and I hope you will all support her so that the newsletter continues to be a source of tears and laughter for a long time to come.

Good Luck to you all

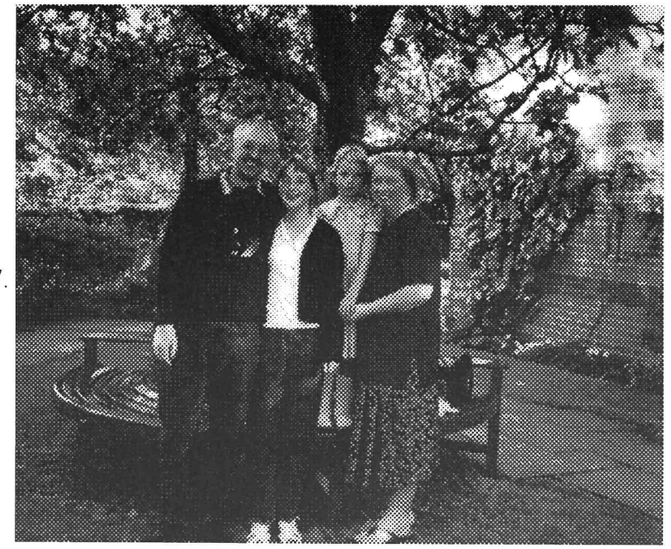
Love Joan


The MPS Removal Team, Anne, Sheila, Angela, Hannah and Joan busy packing

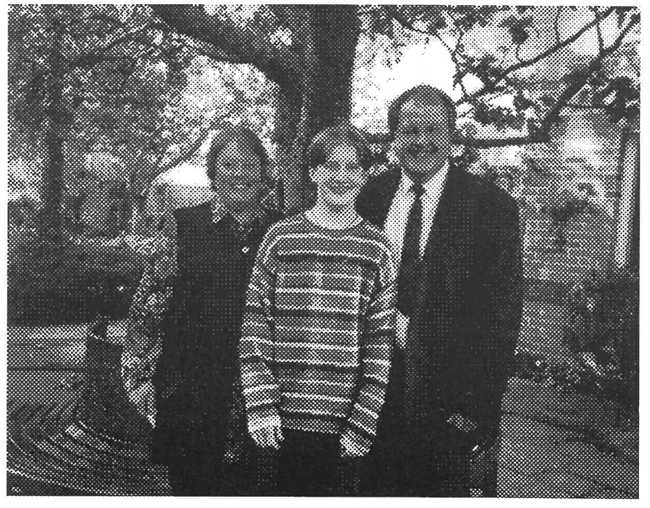
AREA FAMILY SUPPORT

Getting to Know Your Area Family

Pictured opposite are the Russell Family who are one of the Area Support families from the Midlands. Doreen and Monty are pictured with their daughters Gemma aged 15 and Hannah aged 6 years old.



Doreen and Monty's son, Matthew died in April 1997. Mathew who was 12 years old suffered from Hurler Disease.



Pictured above are the Hodgetts family who are another Area Support Family for the Midlands.

Sue and Jeff are pictured with their 13 year old son, Carl.

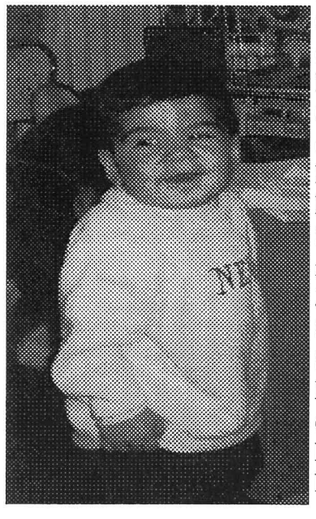
Sadly their son, John died in January 1995. John who was 14 years old suffered from Sanfilippo Disease.

FAMILY SUPPORT

Royal Manchester Children's Hospital
Bone Marrow Transplant Review Clinic
27th February 1998

Every six months, at the Willink Unit, R.M.C.H. they hold a review clinic for children who have undergone bone marrow transplantation for MPS conditions. These review clinics enable the multi-disciplinary team to focus on both the child as an individual and to identify common problems associated with MPS conditions post transplant.

The team consists mainly of Dr. Wraith, Consultant Paediatrician metabolics; Dr Will, Consultant Paediatrician BMT;



Dr. Clayton, Consultant endocrinologist; Dr. Williamson, Consultant orthopaedics; Lorraine Burnett, Specialist Nurse metabolics; Jackie Imrie, Staff Nurse; Sue Crook, BMT family coordinator and Mary Pagett, MPS Family Support North.

Another aspect of these clinics is to give parents the opportunity to meet each other and to share similar experiences.

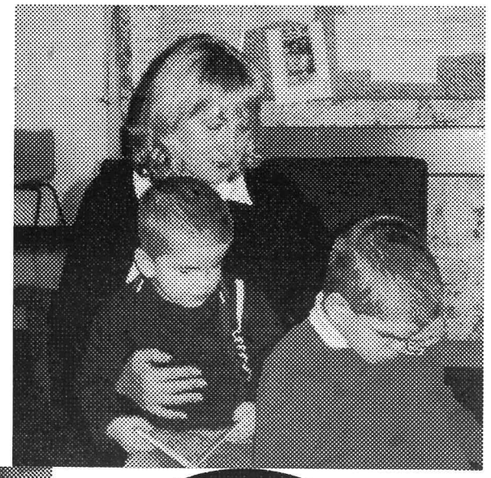
This is especially beneficial to parents of very young children who have recently undergone treatment. The majority of children suffering from Hurler have undergone two bone marrow transplants and several have gone on to have spinal surgery, making the need to be well supported by the medical profession and others vital.

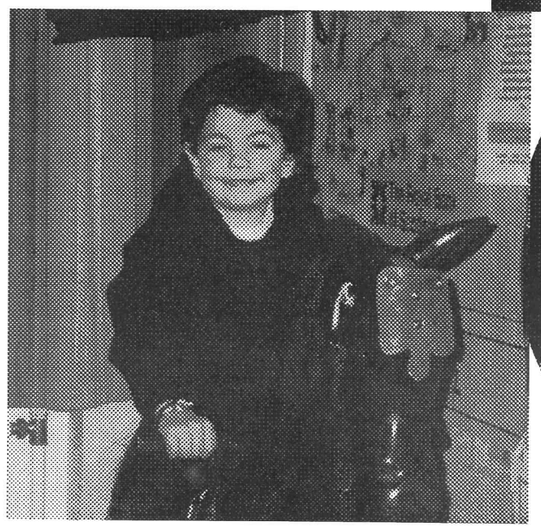
FAMILY SUPPORT

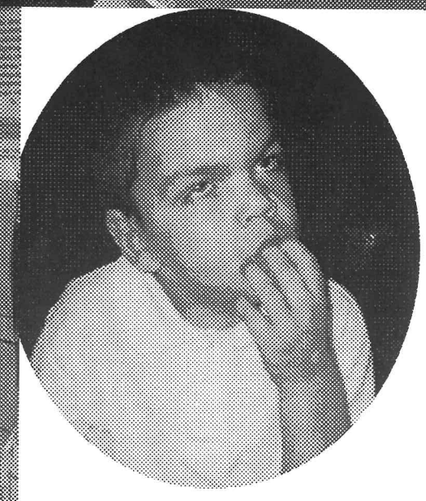
Pam Thomas, Development Officer from the Northern Office, was able to join me at this clinic, which made a difference, as an extra pair of hands is always needed.

As can be seen by the photographs the children are a happy little bunch.

Mary Pagett
Director of Family Support Services North.







FAMILY SUPPORT

CHRISTMAS AT HULLAVINGTON

Better late than never, as the saying goes. I was kindly invited to the "Christmas Do," at 90" Squadron, Hullavington. Mainly because they were holding a raffle for us, their adopted Charity and secondly, it was an ideal opportunity to recruit Army volunteers for the '98 Activity Week.

Photographed above are Major Wilding, Mary and Cpl. Mick Gillespie

Having been introduced to army life in short bursts over the past five years through our activity holiday, I left the MPS office at 3pm having decided that if I didn't go, then I would never make it to Wiltshire for the deadline of 6pm, ***orders from the army!*** At 6pm I was given a quick one - two around the guardroom and then told to be ready for the transport by 7pm. CPL Mick Gillespie, eased the blow by giving me a refreshing glass of wine to the tune of *"Hen be ready to go in five minutes"* (Mick's a Scotsman). The very same man who crossed the dance floor at the Conference last year, in a kilt, bright red wig and TamO' Shanter on his head, and told us all what a wonderful, amazing group of youngsters we have. By Midnight, it was my turn to let the folks know why they had been parting with their money all night and who these wonderful youngsters were who had captured Mick's heart, along with many other strong, manly army guys. Plus the feminine army girls.

By 1pm, Mick and companions, were back to getting more money, by auctioning the raffle prizes people had won. No-one had the nerve to keep hold of their prize, emotional blackmail had set in- well that's not entirely true, Mick did say they could have their raffle prize back providing they won the bid!

Somewhere in my mind, is the memory of the military transport arriving to take us to our beds- time is irrelevant when you're among friends, but I did debate the value of the energy I would use in getting into bed only to get out just as quickly. As ***more orders arrived*** - a meeting with Major Wilding at 9am to discuss volunteers for the activity week.

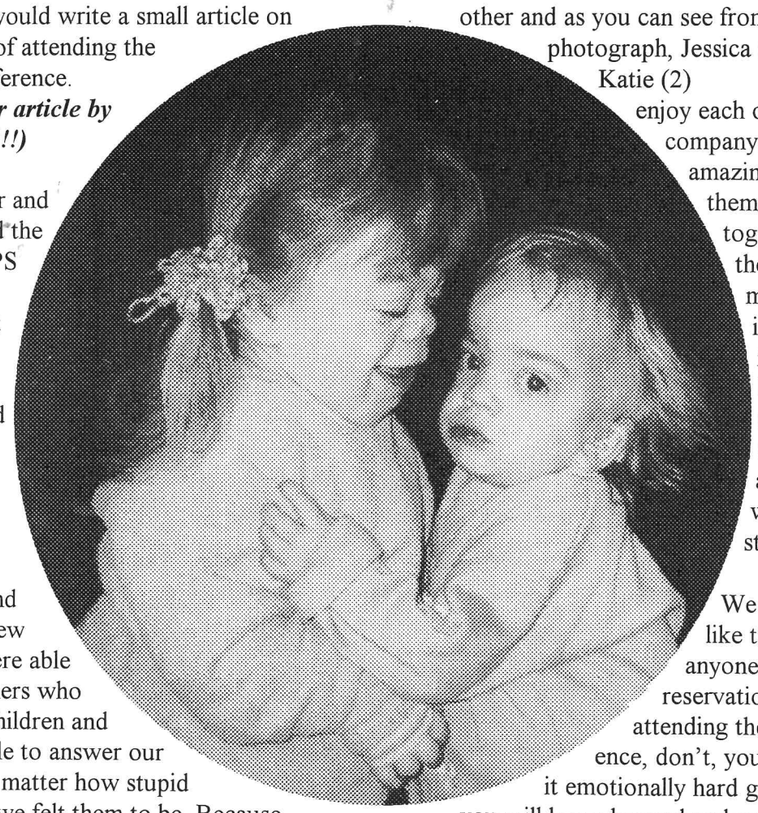
In May, our young folk, once again, will be steeling themselves for great fun with plenty of daring activities, like Zip Wiring, Rock Climbing and Caving to mention but a few. The great thing about the Activity Week, is not the experience of doing things able-bodied people do or don't do, or the achievements and boosts to our egos. The week is about being with friends, of renewing old friendships and discovering new ones.

Friendships come in various ways, and I have personally learned through our young people how precious friendships are, and that if they come, we should grasp them, and most of all, take the time to enjoy them. The 9" Supply Hullavington, will be answering the call for volunteers and I would like to send a warm Thank you for a wonderful "Christmas Do", and a special thank you for opening your hearts to our young people, who truly value you kindness and commitment to MPS.

Mary Pagett.

FAMILY NEWS

I thought I would write a small article on the benefits of attending the Annual Conference. ***(Not another article by Sue Stuart!!!!)***



Myself, Peter and Jessica found the first ever MPS Conference that we went to very confusing, upsetting and very emotional, but we were very pleased that we took the plunge and went. As a new family we were able to talk to others who had Hurler children and they were able to answer our questions no matter how stupid or awkward we felt them to be. Because of the support and advice we had from those families, we decided to extend our family and are now very pleased that we did and do not regret it as Jessica gets so much from her sisters.

We are now an "old family" but still attend the conferences (with our other children Hollie and Annie) so that we can answer questions and be of some comfort to "new families" We have met some wonderful people and still keep in touch even though we have moved house and so have they.

Last year we met the Devine family: Rob. Lindsay, Andrew, Laura, Gary and Katie. Katie has Hurler and reminded so much of Jessica when she was two years old. We get on very well and try to visit one another as often as possible. The children adore one another and as you can see from the photograph, Jessica (7) and Katie (2) enjoy each other's company. It is amazing to see them together as their mannerisms are identical, like the thumb sucking and the way they stand.

We would like to say to anyone who has reservations about attending the conference, don't, you will find it emotionally hard going but you will be welcomed and you will find so much to talk about. When you leave you will have made new friends and have lots of answers to your questions!!!!

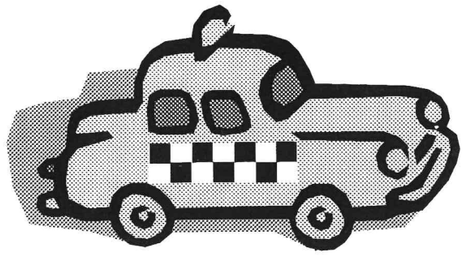
And much to your surprise you will almost certainly have helped someone else.

IT'S GOOD TO TALK!!!!!!!!

Sue Stuart

There is still time to book for the MPS Annual Conference 18th-20th September 1998.

FAMILY NEWS



Dear Readers

I am seeking advice regarding regarding motoring with your disabled child. I will try to explain a little about my dilemma and ask if anyone has experienced or is experiencing the same sort of problem and what is to be done.

I have a son called Thomas who is 9½ years old and has Sanfilippo Syndrome. He is a very large boy (6 stone plus) and is now at the stage where he can no longer walk without my assistance as he falls straight down. He sometimes finds it too difficult to walk and also he cannot get up of his own accord. He is also near to reaching my very slight height of 5ft. 2inches. So all in all I am finding it a real struggle manoeuvring Thomas. I also have a 7 year old daughter, Jade.

Eighteen months ago I joined the Motability Scheme and got a Volkswagon Sharran (MPV) which is fantastic from the point of view of space. The Obi buggy that Thomas has fits into the back space where I've taken out 2 seats and there is a good amount of space in between from and back seats. The major drawback is lifting Thomas into and out of the vehicle as it is higher than an average car. Not only is there the lifting of Thomas but also the buggy.

Recently I visited the Motability Centre to ask for advice and yes they came up with the perfect answer but unless by some miracle I win the lottery it is totally way beyond my financial means. Idealistically a vehicle with the hydraulic lift which would allow Thomas to travel in his buggy in the back would solve all the problems but however do you finance something like that which has a price tag in the region of £30,000?

So in eighteen months time when the 3 year contract finishes what do I do? Continue with the scheme with the same vehicle, change to a car and face the possibility of never taking Thomas out which is completely out of the question as I have to travel to use any sort of facilities.

To be quite honest dread sweeps over me now at the thought of just popping out the shops as it is such hard work.

Please if anyone could offer advice or any practical solutions to this matter I would be most grateful.

Julie Thacker

If anyone has any ideas to help with Julie's problem please let us know at the MPS Amersham office and we will pass them on to Julie.

FAMILY NEWS

BEREAVEMENT

MEN DO CRY

I heard quite often "men don't cry" Though no one ever told me why.
So when I fell and skinned a knee No one came by to comfort me.

And when some bully boy at school Would pull a prank so mean and cruel
I'd quickly learn to turn and quip "It doesn't hurt" and bite my lip.

So as I grew to reasoned years I learned to stifle any tears.
Though "Be a big boy" it began Quite soon I learned to "Be a man."

And I could play that stoic role While storm and tempest wracked my soul.
No pain nor setback could there be Could wrest one single tear from me.

Then one long night I stood nearby And helplessly watched my son die.
And quickly found to my surprise That all that tearless talk was lies.

And still I cry and have no shame I cannot play that "big boy" game.
And openly without remorse I let my sorrow take its course.

So those of you who can't abide A man you've seen who's often cried.
Reach out to him with all your heart As one whose life's been torn apart.

For men do cry when they can see Their loss of immortality.
And tears will come in endless streams When mindless fate destroys their dreams

Taken from: The Canadian MPS Newsletter.

INFORMATION

H.E.L.P.
Holiday Endeavour *for* Lone Parents

H.E.L.P. is a non-profit making charity run mainly by people who are or have been lone parents. They are dedicated to providing, high quality, low cost, half board, full board & self catering accommodation with entertainment packages which will allow any lone parent family REGARDLESS of income, to both afford and enjoy a family holiday.

H.E.L.P. operates an equal opportunities policy for all.

For a brochure and further details write to the Booking Officer enclosing a 9" x 6" self addressed envelope with 2 first class stamps to:
57 Owston Road
Carcroft
Doncaster
South Yorkshire DN6 8DA
or **Tel: 01302 728791**

DLA

The mother of a 29 year old man with cerebral palsy who was awarded Disability Living Allowance (DLA) for life is angry that her son is being reassessed.

Under the Benefits Integrity Project, 442,000 disabled people will receive questionnaires or visits from the Department of Social Security (DSS) during the next few months in a bid to cut fraud.

Violet Tallet from Kent, received a 34 page DLA 250 questionnaire for her son,Simon, even though he was told five years ago he would not have his care needs reassessed.

Mrs Tallett said, "It has come as a real shock to us. This will take hours to complete and puts an emotional strain on the person with the disability as well their carer."

A DSS spokesman said that the questionnaire was a way of keeping in contact with those who had been given life awards.

The Royal Association of Disability and Rehabilitation (RADAR) social security officer, Margaret Lavery, said "It is bad news that they are including people on life awards".

The Carers National Association are advising people with professional guidance. If they have any problems or questions.
Contact :The Disability Alliance on
Tel: 0171 247 8776

Social Security Secretary, Harriet Harman, announced last month that Incapacity Benefit was also to be reviewed.

Child Death Helpline

The child death Helpline is operated from Great Ormond Street and Alder Hey Children's Hospitals. It is a confidential helpline for anyone affected by the death of a child.

A bereaved parent is always available to answer your call. We provide an opportunity for you to talk to someone sympathetic to your situation.

The telephone helpline is available Monday, Wednesday and Friday evenings from 7pm to 10pm.
The freephone number is **0800 282986**

Suction Machine

If anyone requires a suction machine please let the Amersham office know as we have one available to MPS families.

INFORMATION

UPDATE OF MPS RESEARCH IN ITALY

During the past years, the Associazione Itliana Mucopolisaccaridosi have supported some MPS research projects carried out in laboratories around Italy.

Briefly researchers have focused their attention on investigations that will lead to a better understanding of the molecular and Biochemical character of the MPS disorders and attention has been focused upon the development of protocols for gene replacement therapy.

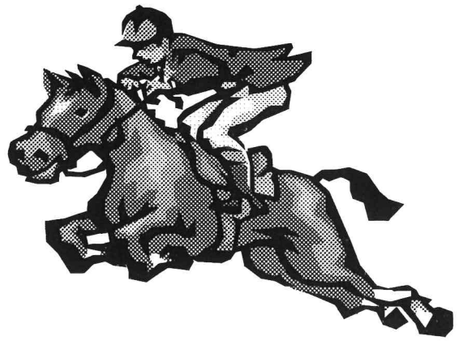
Specifically, mutational analysis has been carried out in a number of different MPS patients and carriers. Dr. N. Balzano University of NapIes, has conducted mutational analysis on 7 Hunter, 30 Sanfilippo A, 20 Sanfilippo B and 7 Maroreaux-Lamy patients. His efforts have lead to the detection of several common mutations. Similarly, Dr. G.R. Villani of Naples, has identified 17 mutations in MPS II patients and Dr. G. Bonurrcelli, Gaslini Institute, Genoa, has screened by SSCP (Single Stranded Conformation Polymorphism) and DNA sequencing 20 women, discovering 13 MPS II carriers. Dr. G. Bonuccelli has also improved a non-radioactive method for carrier diagnosis, when no mutational data is available. Results generated from this research will enable a significant advance to be made in understanding the molecular causes of these MPS Diseases with the possibility of developing an easier and more accurate means of detecting MPS in carriers and in prenatal diagnosis. Mutational analysis has also provided the opportunity to investigate biochemical parameters of stability and activity of in vitro expressed mutated proteins Research directed towards characterising the expressed mutated proteins is currently being conducted in the laboratories of Dr. Balzano and Dr. Villani. Particularly, Dr. Villani has expressed five mutated enzymes of MPS II patients. Results from this study have suggested a key role for Arg 88 in the active site of the iduronate-2-sulphatase. This work will lead to a deeper understanding of the relationship between structure and function of the enzymes.

The importance of therapy for MPS patients has also been addressed in several Italian laboratories with research currently directed towards gene therapy for MPS II and IVA. The results of the research for gene therapy for MPS II carried out by Dr. M. Scarpa's team, Dept. Paediatrics, CRIBI, University of Padoa, was recently published in Gene Therapy (Di Francesco et.al.,1997, Gene Therapy, Vol 4, 442-448 "In vitro correction of iduronate-2-suplhatase deficiency by adenovirus-mediated gene transfer"). The paper describes the invitrocorrection of primary cells obtained from MPS II patients, mediated by an adenoviral vector carrying iduronate-2-suplhatase cDNA. Moreover, Dr. Scarpa has developed a model system based on the generation of derma-equivalent, which is a suitable tool for long-term expression of adenoviral transduced cells. Yet another approach under investigation at the CRIBI: is the possibility of ex vivo gene therapy for patients, where an iduronate-2-suplhatase protein delivery system is being assessed. This system uses an encapsulated murine myoblasts C2C12 cell line expressing high levels of IDS. Gene therapy for MPS IVA patients is currently being evaluated by Dr. G. Toietta at the Telethon Institute of Gene Therapy for Genetic Diseases, HSR, Milan. The first in vitro pre-clinical data on gene therapy of MPS IVA has been collected. This data will enable the evaluation of a possible drug delivery approach mediated by retroviral infected cells of different origins (fibroblasts, muscle cells, keratinocytes, peripheral blood lymphocytes). Additionally, a local approach to gene replacement therapy will be assessed by the injection of retrovirally infected synoviocytes into joints.

These studies, that are still carrying on, represent the Italian contribution to the world-wide effort in MPS research, which involves many public and private centres. The lack of an effective cure underscores the importance of a basic and applied research in this field. We believe that understanding the molecular origin of MPS is a mandatory task for the development of effective preventive and therapeutic strategies.

Vito Brusco (Italian MPS Society)

INFORMATION

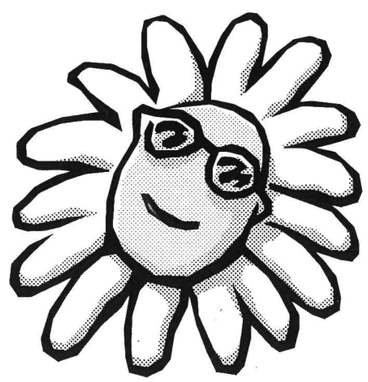


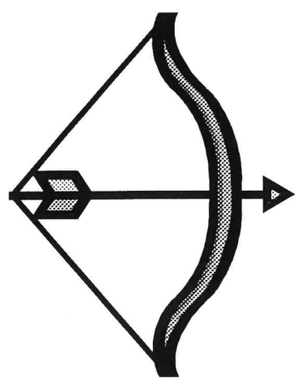
MPS Family Holiday
Saturday 25th July to 1st August 1998
Haven Holiday Centre, Primrose Valley
Near Filey, Yorkshire

There are still 7 places available for the MPS family holiday so if you are interested please contact Pam Thomas in the Preston office.

MPS Activity Holiday
Sunday 24th May to Saturday 30th May 1998
Bendrigg Lodge, Cumbria

There are still 5 places left for the MPS Activity Holiday so if you are interested please contact Pam Thomas in the Preston office





INFORMATION

LAUNCH OF THE RARE DISORDERS ALLIANCE-UK

After a lengthy research process producing very encouraging results, this new Alliance was launched on 20th November 1997 in London. There was an overwhelming response in favour of the proposed Alliance, but it was not felt necessary to set up another completely separate organisation. As a result the RDA-UK will be run from within Contact A Family (CAF).

With over 5,000 differing rare disorders the Alliance recognises that many of the problems experienced by families and individuals are due to the rarity of the conditions rather than the specific diagnosis. It is hoped that CAF, together with an advisory group made up from members, will be successful in trying to redress some of the inequity felt by these families in every aspect of their care. The Alliance will represent all those affected by rare conditions of any age, including those with late onset conditions.The launch programme included a talk by Dr Michael Paton, Clinical Geneticist, who has been a very good friend to CAF for may years and acted as an advisor for The CAF directory. The other speaker was Stephane, Korsia, Executive Director of a comparatively new European organisation for rare disorders called EURODIS. Together with EURODIS this new Alliance hope to improve the quality of life for all those affected by rare disorders.

Christine Lavery represented MPS at the launch in London and the MPS Society will be joining the Alliance, taking a close interest in it's planning and development.

WANTED

Anne MarieWatson, mother of Zara aged 8 years old (Sanfilippo) has written (by hand!) to hundreds of celebrities over the past year. We now have a collection of famous peoples belongings to be auctioned this summer.

If anyone has a computer going spare, perhaps your office is upgrading, Anne Marie could put it to good use for the MPS Society.
Please contact the Amersham office if you can help.

INFORMATION

ACT Charter

1. Every child shall be treated with dignity and respect and shall be afforded privacy whatever the child's physical or intellectual ability.

2. Parents shall be acknowledged as the primary carers and shall be centrally involved as partners in all care and decisions involving their child.

3. Every child shall be given the opportunity to participate in decisions affecting his or her care, according to age and understanding.

4. Every family shall be given the opportunity of a consultation with a paediatric specialist who has particular knowledge of the child's condition.

5. Information shall be provided for the parents, and for the child and the siblings according to age and understanding. The needs of other relatives shall also be addressed.

6. An honest and open approach shall be the basis of communication which shall be sensitive and appropriate to age and understanding.

7. The family home shall remain the centre of caring whenever possible. AII other care shall be provided by paediatric trained staff in a child centred environment.

8. Every child shall have access to education. Efforts shall be made to enable the child to engage in other childhood activities.

9. Every family shall be entitled to a named key worker who will enable the family to build up and maintain an appropriate support system.

10. Every family shall have access to flexible respite care in their own home and in a home-from-home setting for the whole family, with appropriate paediatric nursing and medical support.

11. Every family shall have access to paediatric nursing support in the home when required.

12. Every family shall have access to expert, sensitive advice in procuring practical aids and financial support.

13. Every family shall have access to domestic help at times of stress at home.

14. Bereavement support shall be offered to the whole family and be available for as long as required.

We copied this Charter from the Association for Children with life threatening or Terminal conditions and their families 65 St Michael's Hill, Bristol, BS2 8DZ. Tel: 0117 922 1556. Registered Charity No. 1029658

INFORMATION

THE SANFILIPPO CODE OF CONDUCT

by Penny Lister

Thou must never, under any circumstances, allow parent or carer to entice thou into a daytime nap. Should the disgraceful situation ever occur the offending parent/carer must be punished by at least 24 hour of sleep deprivation.

Thou must be constantly on the lookout for open doors and gates; these are invitations to forbidden pleasures and adventures.

Thou must always leap into active mode when thou hears the first tweet of a bird at sunrise. The only exception to this rule is on the first day of each school term, when a sleep-in until at least 9.00 am is recommended.

When travelling in a car thou must always buck violently when stopped at traffic lights- this prevents the driver from becoming bored and dozing off to sleep.

There is only one correct way to empty they bath of its water and it is not be pulling out the plug.

To avoid the indignity of being washed with soap, choose one of two alternatives:
a) eat it
b) hide it in the toilet bowl.

Thou must taste-test the environment at every given opportunity. Even the stringiest leaf, grittiest mouthful of dirt, crunchiest rock and mushiest pet food will taste better than the content of thy lunchbox.

Thou art quite entitled to swipe food and drinks from other if they are within thy arm's reach.

As a matter of self-preservation thou must save thou most charming and disarming smiles for the hours between midnight and 4.00 am.

Thou must always remember that buttons knobs, switches and taps are there for a reason - do not allow them to stay idle.

Singing at the top of thy voice, decorating the floor with magazines, and pruning the plants are highly recommended activities for Doctor's waiting rooms - thou will usually not have to wait long for attention.

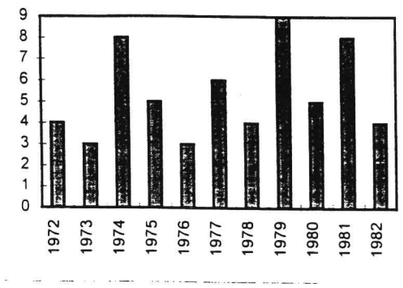
Within five minutes of arrival at parks thou must fill thy pants. Ditto, regarding arrival at swimming pools.

This article was found in the Australian MPS Newsletter in September 1997. We thought that parents of Sanfilippo children would ***relate*** *to this code ...*

INFORMATION

The following are graphs showing the incidence of MPS Disease in the United Kingdom over two ten year periods.

1972	4
1973	3
1974	8
1975	5
1976	3
1977	6
1978	4
1979	9
1980	5
1981	8
1982	4



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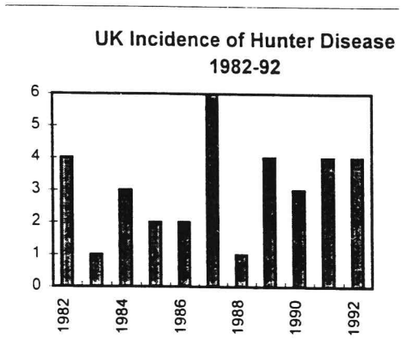
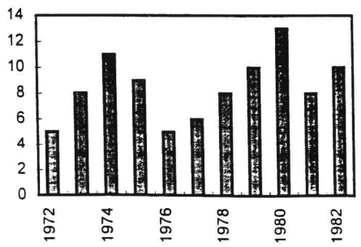
1984	3
1985	2
1986	2
1987	6
1988	1
1989	4
1990	3
1991	4
1992	4



1972	5
1973	8
1974	11
1975	9
1976	5
1977	6
1978	8
1979	10
1980	13
1981	8
1982	10



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1984	2
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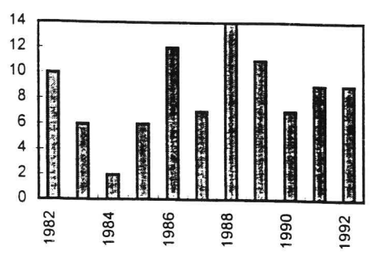
1988	14
1989	11
1990	7
1991	9
1992	9



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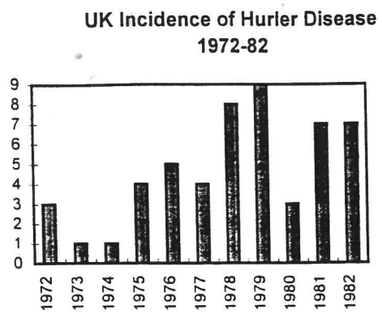
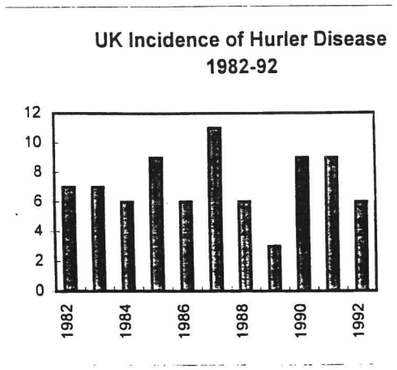


1982	7
1983	7
1984	6
1985	9
1986	6
1987	11
1988	6
1989	3
1990	9
1991	9
1992	6



1972	1
1973	4
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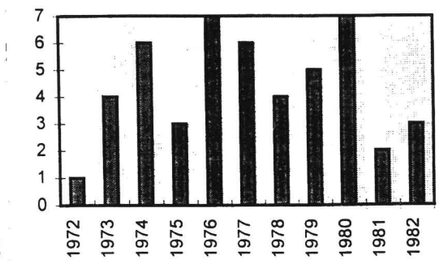
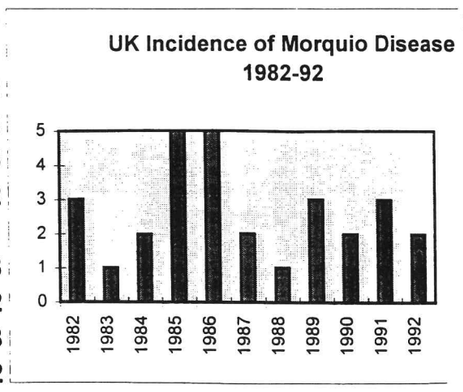
1977	6
1978	4
1979	5
1980	7
1981	2
1982	3



1982	3
1983	1
1984	2
1985	5
1986	5
1987	2
1988	1
1989	3
1990	2
1991	3
1992	2



INFORMATION

SEIZURES IN CHILDREN WITH MPS DISORDERS

by Dr. Joe T.R. Clarke. Division of Clinical Genetics, Hospital for Sick Children,

Introduction

In general, seizures, or convulsions. are not a common or difficult problem in children with MPS disorders. However, in some conditions they are more common than in others and occasionally controlling them is a major challenge. This article is intended to answer. bnefly and in understandable terms. some of the more common questions parents of children with MPS disorders ask about the significance and the management of seizures.

What are seizures?

Seizures. or convulsions. are defined formally as transient. paroxysmal behaviors often associated with alterations of consciousness accompanied by involuntary movements and accompanied by EEG abnormalities. However some seizures only occur once, some involve very little or no obvious change in the level of consciousness, some are not associated with involuntary muscle movements, and in some the EEG may be normal. particularly between attacks. Epilepsy refers to recurrent. spontaneous seizures.

There are several types of seizures. The type most familiar to people is the generalised tonic-clonic seizure typical of classical epilepsy. These occur in many children with Sanfilippo syndrome (MPS III) generally late in the course of their disease. However. there are other, more subtle, types of seizure which are more difficult to identify. These include classical generalised absence attacks. characterised by extremely brief impairment of consciousness, with no aura or postictal state and a classical EEG abnormality of a 3 per second spike and wave pattern. Complex partial seizures. which are characterised by stereotypic motor behavior. associated with disturbances of consciousness, are also common in some patients with Sanfilippo syndrome. Myoclonic seizures consist of brief muscle jerk and may be a major problem in some other lysosomal diseases. like Tay-Sachs Disease and Sialidosis. They are not typical of MPS Disorders, though they may occur.

In other MPS disorders, such as in children with Morquio syndrome (MPS IV), Maroteaux-Lamy syndrome (MPS VI) or Scheie syndrome (MPS IS), seizures are very rare. In Hurler syndrome (MPS IH) and Hunter syndrome (MPS II), they are quite variable. Seizures may occur as a complication of the hydrocephalus in some infants with Hurler syndrome or Hunter syndrome. Some boys with Hunter syndrome develop very severe seizure disorders, though most appear not to be at particularly high risk.

It is important to remember that MPS children. like any child. may develop seizures for reasons unrelated to their MPS disorder. If a child with an MPS disorder has a seizure, particularly if it is the first one they have ever had, the possibility that it might have been caused by something else should be considered Some of the conditions, such as meningitis, are treatable.

How do we know a child is having seizures?

The most important information needed to determine if a child is having seizures is a reliable account of the event itself. When did it occur? Was there anything

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that seemed to bring it on? What did the child do? What movements did he or she make? Could you interrupt the episode by holding the child or stimulating him or her in any way? Did he or she seem to be "out of it" Or did he or she respond to you in any way during the event? How long did it last? Did he or she go into a deep sleep after the event? Was there any kind of warning? What. if anything, seemed to precipitate the event? Was it associated with urinary incontinence?
The EEG is helpful. but it may not be abnormal. A CT scan or an MRI scan are not helpful in determining if a child is having seizures. though they may help to determine why a child with seizures is having them. Seizures tend to be unpredictable, may be uncontrollable, and are often associated with unresponsiveness. Generalised tonic-clonic seizures are often followed by a period of deep sleep. Susceptibility to seizures is greatest when a child is tired, particularly shortly after waking in the morning. or at bedtime.

Many conditions may mimic seizures, such as fainting or breath-holding spells. certain types of behaviour disturbances (such as temper tantrums). nightmares, tics,
apneic spells. excessive startle. "sleep jumps'. (sleep myoclonus or sudden massive jerks of the body occurring, just as one is falling asleep), migraine. narcolepsy. even masturbation. The determination of whether a child is having seizures, or some other seizure-likeparoxysmal problem, is
sometimes very difficult.

What causes them?

Seizures are a symptom. not a disease. They are caused by some insult to the brain and treatment sometimes simply involves removing whatever is causing them. Insults to the brain most commonly associated with seizures can be classified as:

- disturbances of brain energy metabolism, such as low blood sugar (hypoglycemia). lack of oxygen (hypoxia), low blood calcium (tetany), to mention a few.
- accumulations of toxins, such as certain drugs, carbon monoxide,strychinne.
- inflammations of the brain tissue, such as meningitis.
- disruptions of the circulation or bleeding into or around the brain
- excessive fever
- distortion of brain tissue, such as occurs with injuries and in some congenital malformations of the brain.

In children with MPS disorders, the seizures are probably caused by distortion of the brain cells caused by accumulation of MPS or other complex metabolites in brain cells (neurons) or disruption of the supporting tissue of the brain, either as a result of pressure (hydrocephalus) or the death of brain cells. Some things tend to lower the threshold for seizures, such as fever, exhaustion, and some common drugs. like antihistamines.

Are they dangerous?

Seizures can be dangerous. The sudden and unpredictable loss of motor control occurring in generalised tonic-clonic seizures may cause falls resulting in injuries which can be serious. particularly if they involve the head.

Other types of seizures are less likely to cause falls resulting in serious injury. Head protection is sometimes prescribed for these children to try to avoid head injury. Unfortunately. MPS children with seizures often resist wearing helmets. On the other hand, many of the MPS children with seizures are older and will be confined most of the time to a wheelchair or to bed, and injury from falls is not such a major problem. Because the reflexes that normally protect the airway are often depressed, children are at increased risk of aspirating stomach contents if they vomit during a seizure. Sometimes this causes sudden death. At other times, it may

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cause pneumonia.

When a child is having a seizure, you should lay them on the floor or on their bed, if they are already there turn them on their side, face turned downward, to ensure that if they vomit, the stomach contents run freely out of the mouth. Hold the child's head. but **DO NOT** try to hold their mouth open. Most seizures stop by themselves within a few minutes. If a seizure persists for more than 5 minutes you should call for help and take the child to the nearest hospital. You may have been given drugs to give your child if they are particularly susceptible to prolonged or very frequent seizures. One of the most commonly used is lorazepam (Ativan) which can be given safely by the rectum.

Whether repeated seizures themselves cause brain damage, in the absence of injuries caused by falls and airway problems. is debatable. It is sometimes impossible to separate direct effects of seizures on the brain from the effects of lack of oxygen, injury, and the underlying disease. Severe. frequent or prolonged (more than 30 minutes), tonic-clonic seizures probably do cause some damage to the brain. However. the seizures in children with MPS disorders are rarely that severe or difficult to control.

Do they cause further deterioration?
Seizures do not appear to accelerate the course of any of the MPS disorders. However there is no doubt that the transient disturbances of brain function occurring during seizures interferes with memory, thinking and other higher cerebral activities. These disturbances are generally considered to be rapidly reversable when the seizure stops. However, if seizures occur so frequently that the brain does not have time to recover from one before it is hit by another, the effect can be quite profound. This is often only fully appreciated when the seizures are brought under control. and a child who was thought to be deteriorating suddenly shows an improvement in cerebral functioning. Unfortunately. the drugs used to control seizures may also interfere with brain functioning. causing apparent deterioration in the patient. Finding· the right balance between seizure control and anticonvulsant drug-induced drowiness is sometimes difficult and may take some time and several measurements of plasma anticonvulsant drug levels.

What types of medications are used to treat seizures in children with MPS disorders?
The selection of the drugs to use to control seizures depends on the type of seizures. their frequency, the age of the patient and the stage of their disease. Because the seizures are generally not difficult to control, treatment with one of the more familiar and safe drugs like phenytoin (epanutin) is often all that is needed. Some of the more commonly used drugs are shown in the table on the following page.
Consistency and appropriate dosing are as important in seizure control as the choice of drug.

Routine management with anticonvulsants includes periodic measurements of drug levels in plasma. blood counts, and liver functions tests, to ensure that drug dosages are appropriate and that the child is not developing avoidable adverse side-effects. Occasionally, satisfactory seizure control will not be achieved with the use of one drug alone. If this is the case, getting the help of a neurologist is important. Parents should be aware that some anticonvulsant drugs interact with other commonly used medications, with undesirable effects. For example, the antihistamines in common, over-the-counter cold medicines may decrease seizure threshold, making seizure control more difficult. The antibiotic, erythromycin increases the toxicity of carbamazepine;

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appearing on the market and some of these will probably be effective in the treatment of seizures in children with MPS disorders. As a general rule,however as long as the treatment is effective, it is generally better to use a medication that is familiar rather than a new drug with uncertain side-effects.

Concluding remarks
Seizures are a dramatically frightening event, when they occur. Fortunately, although they often occur in children with Sanfilippo syndrome and in some boys with Hunter syndrome, they are uncommon in the other MPS disorders. When they occur, they are generally relatively easy to control, usually with the use of one of the more conventional anticonvulsants. If the seizures do not appear to be adequately controlled
Consider:

- Is the child actually having seizures? or is he or she having some other seizure like episodes?
- Is the drug being used to treat the seizures appropriate?
- Is the dosage appropriate?
- Are there other treatable factors such as hydrocephalus a contributing factor?

Table: Drugs used in the treatment of seizures in children with MPS disorders.

Drug	Indication	Possible adverse reactions
Phenytoin Epanutin	Generalised seizures, partial seizures	Drowsiness, unsteadiness, thickening of gums, skin rash, enlargement of lymph nodes, learning problems, allergic reactions, rickets (rare)
Phenobarbital	Generalised seizures, partial seizures	Drowsiness, hyperactivity, skin rash, learning problems.
Carbamazepine	Partial seizures, generalised seizures	Drowsiness, unsteadiness, skin rash, low white blood count.
Tegretol® Valproic acid, Epilim	All kinds of seizures	Drowsiness, nausea and vomiting, abnormal liver function, low platelas, carnitine depletion, pancreatisis
Clobazam Frisium® Nitrazepam	All types of seizures, except absence attacks (Development of tolerance is common) Myoclonic seizures	Drowsiness, fatiguability, dizziness, irritability
Mogadon® Vigabatrin	Partial seizures, generalised seizures	Drowsiness, emotional depression
Sabril®* Gabapentin	Partial seizures	Drowsiness
Neurontin®* Lamotrigine Lamictal®*	Partial and generalised seizures	Rash, drowsiness

* Some of the Newer Types of anticonvulsants.

The above article was copied from the Canadian MPS Newsletter

INFORMATION

CHORIONIC VILLUS SAMPLING
by Dr. P Ferreira, MBBS, FRCPC, FCCMG

Chorionic Villus Sampling (CVS) is an alternative to amniocentesis for prenatal diagnosis of many disorders. Its main advantage is that it can be performed earlier in pregnancy: 9 to 12 weeks vs 15 to 17 weeks for amniocentesis. If termination of-pregnancy is being considered, the earlier it is done the easier, both physically and emotionally.

With CVS, a small sample of the developing placenta is taken either via the cervix with a catheter, or via the abdominal wall with a needle. In amniocentesis, amniotic fluid, which contains some fetal cells, is withdrawn with a needle. Ultrasound is used for guidance in both procedures.

The fetal cells obtained in either procedure may be grown in culture to provide enough for analysis, however CVS provides a larger number of cells and many tests can be done directly on the sample, saving a lot of time, since cell cultures may take several weeks. The types of tests that can be done are
a) examining the chromosomes to check for conditions such as Down syndrome,
b) biochemical analysis, where there is a known risk of a specific disease. usually enzyme deficiencies as in the MPS disorders and/or
c) DNA analysis which can be done in specific instances where the gene itself, or another gene close by on the same chromosome has been isolated. An additional test that is routinely done on amniotic fluid from amniocentesis is to screen for neural tube defects (spina bifida and anencephaly); this cannot be done with CVS.

The risk of miscarriage due to amniocentesis has been estimated to be 0.5% (1:200), at a time of pregnancy when the spontaneous miscarriage rate is 2.5-3.0%.
The spontaneous miscarriage rate is higher earlier in pregnancv when CVS is usually performed about 3 -5%. The additional risk of the procedure is estimated to be between 0.5-1.0%. Prenatal diagnosis has been successfully carried out in most MPS disorders by biochemical (enzyme) analysis using both amniocentesis and CVS. In the future, greater use may be made of DNA techniques as more of the genes are isolated. It is difficult to go into much further detail since the field is rapidly changing, and there are many specifics to be considered with each individual family and in each disease. I would strongly urge anyone who thinks they may wish to request prenatal diagnosis for any of the MPS disorders to contact their local Genetics Centre for advice preferably well in advance of becoming pregnant.
(see diagram opposite)

GLAUCOMA IN MPS CHILDREN

by Dr. P Ferreira MBBS, FRCPC, FCCMG

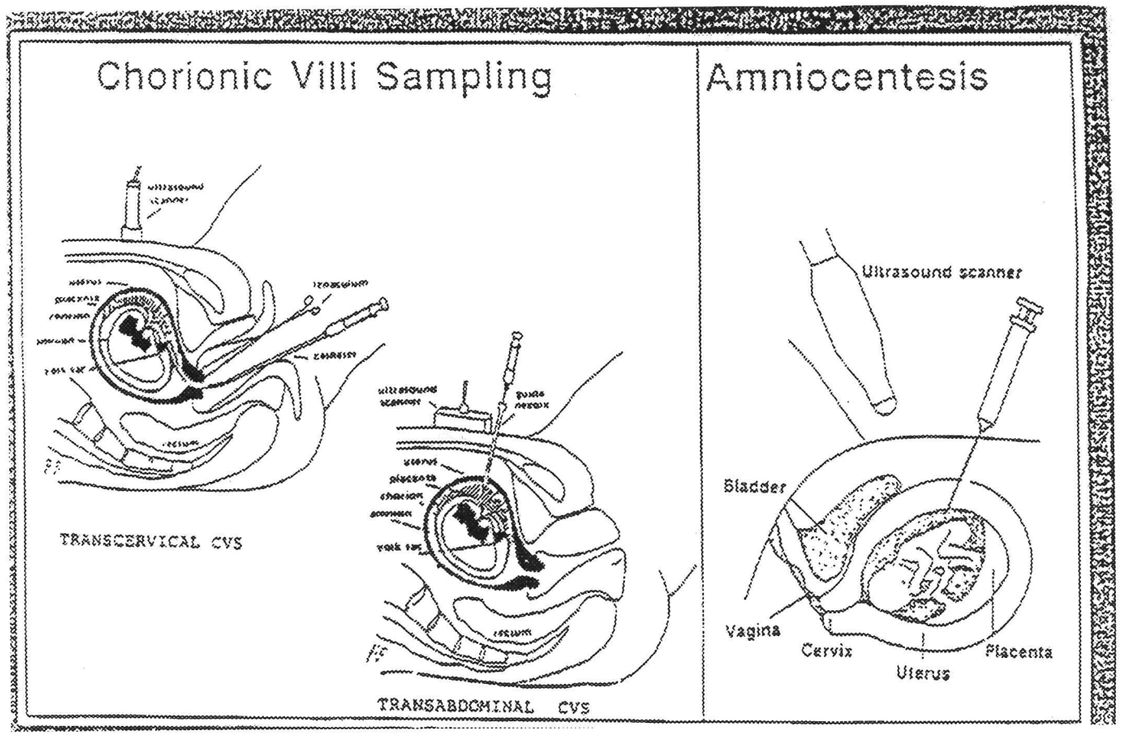
In all MPS diseases, the basic problem is accumulation of mucopolysaccharides in many organs and tissues of the body, and the eye is no exception. This causes clouding of the cornea (the transparent part of the front of the eye) which is usual in many but not all MPS disorders; vision may be also affected in several other ways. One of these is glaucoma, which in MPS diseases is caused by increased pressure resulting from blockage of the filter that normally removes fluid from the eye. because of accumulation

FAMILY NEWS

of mucopolysaccharides. Glaucoma has been reported in Hurler, Sheie, Hunter, Morquio and Maroteaux-Lamy syndrome but it is difficult to know how frequent this is in any particular disease because most of the studies report one or two people with this complication. and to my knowledge no one has published a long-term systematic ophthalmic study of many MPS patients.

Unfortunately diagnosis may be difficult. In some other situations where glaucoma occurs it can be quite sudden, causing pain, decreased vision. aversion to bright lights, and haziness of the comea. In very young children (under 2 years) there may even be noticeable enlargement of the eye. Although acute glaucoma like this can occur in MPS diseases, the development of increased pressure is more likely to be gradual, and could be easily missed by both the patient and family. Also, it may be difficult for physicians to diagnose it because of the lack of symptoms. and since it can be difficult to examine the eye properly through a cloudy cornea.
Glaucoma can cause permanent visual problems. but can be treated medically (with drugs) and/or surgically. Therefore it is important to be aware of this possibility and try to diagnose it as early as possible. For these and other reasons, I think it is a good idea for all MPS patients to be examined by a specialist ophthalmologist with repeat visits as necessary. This would be especially important if there are any symptoms suggestive of an eye problem.

The above articles were copied from the Canadian MPS newsletter.

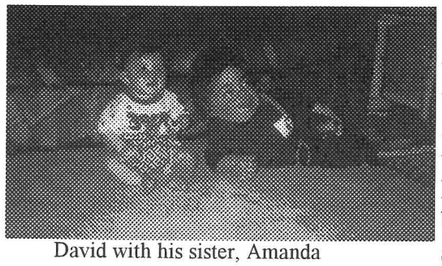


OVERSEAS NEWS

News from New Zealand

Please let me introduce myself, my name is Trudy and my husband is Mike. We have 2 beautiful precious children, David is 5½ years and Amanda is 14 months old. Sadly my sweet David has Hurler Disease.

He was diagnosed at the age of 16 months and I have been receiving your newsletter ever since. I would like to thank you all very much for the many stories and the information I have received through it. We lost our wee David to Hurler Syndrome on the 12th June 1997. It was just so sudden we didn't even have time to say "goodbye". We had no idea that this was the time. Six weeks before I started to get concerned with the amount of sleeping David was doing. He would go to bed at 7 o'clock and sleep through the night and I would have to wake him up around 11 o'clock in the morning. This was for the first two weeks. His eating seemed to improve and he was still feeding himself, mainly bread and chocolate.

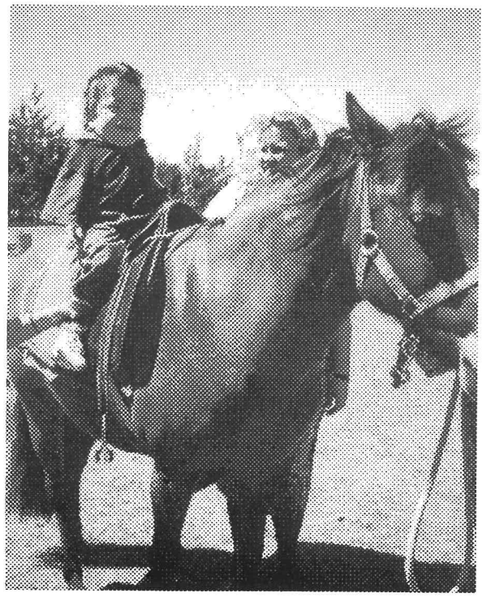

David with his sister, Amanda

We took David into hospital for two days (not overnight). The Doctor was concerned with his blood. They tried him on oxygen overnight which made the blood worse. Anyway went we home and his sleeping continued to get worse as he was sleeping 18 hours straight through. On the Sunday before he was so blue in colour it really frightened me and then on the Monday his hands, feet and legs swelled up so much that he was admitted to the hospital on the Tuesday. The Doctor examined David and he had an enlarged liver and kidneys. We took him home overnight and back the next morning. David had an ECG and a scan the next day. The results of which were good. We took him home on Wednesday afternoon. David was wonderful, he never complained about anything and he always gave us his beautiful big smile. He only said about 5 words all his life and never 2 words together. The only thing David lost in the last 6 weeks was his ability to walk. Wednesday night he looked very tired. We cradled and kissed him good-night. David was his usual happy smiling self. We never dreamt for one moment that this was his last smile we would see. Thursday morning 8.20 Mike went in to wake David up. He was breathing very shallowly and Mike picked him up just as David breathed his last in Mike's arms.

David is my pride and joy I was so privileged to be his mother. David was such an easy child to love. My very special Hurler child sent from God. I was so proud of him.

I once again thank you for your newsletters with love,

Trudy, Mike and Amanda Fermor.
God bless you all.



OVERSEAS NEWS

Hi! Greetings from Bancroft, Ontario

My name is Rhoda Dickinson and I'm five years old and I have Fucosidosis. Pictured with me is my little sister, Tanya whose diagnosis is PESKY SISTER!

I am a very busy girl. I attend kinder-garten now and I ride on a big school bus. I seem to be always catching colds and getting sore throats. In the past six months I have had my adenoids and tonsils out and tubes in my ears now five times.

I will soon be wearing leg braces to prevent me from falling down so much.

Not a lot has changed with my condition, since being diagnosed two years ago.

My speech is staying consistent with around sixty words. I'm not potty trained yet, Mum is still on my case about that, but I'm not giving in to her yet!!!!!

My favourite T.V. programme is still "Barney" and I'm teaching my little sister to love him as much as I do. I really enjoy music and I love to dance fall down and dance some more.

I was finding it difficult to ride my tricycle without toppling over so my parents bought me a battery powered "Jeep". I can drive all over. It's a great toy for a kid to have. It doesn't go very fast and I can't get hurt, and most importantly I can operate it all by myself. My only problem is now my sister thinks I'm her own personal chauffeur.

Well this concludes the highlights of my life and I will try to writie more often. However it depends on how much homework my teacher has me doing, that is if I ever start to get any.

Rhoda Dickinson.

MY DAUGHTER

My daughter ,my love, my first born child
Her temperment so calm and mild
Her eyes so brown, hair so blond
A mother and daughter so close in bond

At the age of three
It was plain to see problems lay ahead
It was news I would dread

A rare disease the Doctor said
Research I did, information I read
His news to me seemed so unfair
Her disease for sure was extremely rare

something we live with everyday
Nothing anyone can do or say
Hope for the best and pray and pray
And enjoy my little girl each and every day
Because she is different, not the same

It's not her fault she is not to blame
Her hair so blond, with the odd little curl
Take the time to know her, a sweet little girl

Extra help she will need
So to eveyone I plead,please don't shy away
Give her a smile and help make her day

Yes she may be slow to learn
Of mine this is a big concern
Please give this little girl a chance
Her life I know I can enhance

Her life is slowly passing away
Don't let her waste another day
give her a chance to learn and play
And smile to help make her day.

Rhonda's Mum

FUNDRAISING

Sponsored cycle ride from Great Ormond Street Hospital - London to Paris -France.

Day 1
Tania and Mark Brown and colleagues left GOSH at 8.00 am and cycled to Bognor Regis (70 miles) and the weather was raining all the way.
Lunch in the coldest pub *great when you are soaking wet* and arrived at Bognor at 6.00 pm. Had a refreshing shower and changed in the local leisure centre then into the minibus and drove to Portsmouth to catch the overnight ferry to Le Harve. *rest at last!*

Day 2
Woke up with a sore head in the morning and had a quick drive to Fecamp (directly opposite to Bognor Regis). From here we re-started our cycling,70 miles through lovely pretty villages and the sun shone on our backs, so what a contrast from yesterday. Cycling most of the way along the River Seine until we reached Louviers.

Day 3
In the morning we started cycling again through pretty scenery but it then became suburban. Passed through the Bois de Boulogne, then St. Germain. Then alas we got lost but eventually ended up in Paris at the Eiffel Tower at 7.00 pm on the Sunday night for a much welcomed champagne reception. 68 miles today making a grand total of 208 miles.

Mark and Tania along with their colleagues would like to thank everyone that sponsored them and helped raise a great total of £5,413.for the MPS Society.

Congratulations to you all and thanks to Tania for the diary of events.

Picture from left to right shows: Tom Parker, Mark Brown,Dan Griddin,Barry Burgess, Mark Ireland and Jamie Parker at the Eiffel Tower - Paris.

FUNDRAISING

Please find enclosed forms and cheques for the raffle we did for MPS at the dance shows of the Arabesque Dance Academy back in December.

Hannah, our six year old, attends the Academy and so were able to do a raffle for the MPS Society for the second time, as the shows are produced every other year.

Before the event we were able to sell tickets to friends, families of the girls who were also in the shows. This resulted in a grand total of £560 being raised for the MPS Society.

Enclosed is a photograph of Hannah with her friend Katherine in their '*German*' costumes at the shows.

Best Wishes
Doreen and Monty Russell

Jeans for Genes Day 1997

When my husband Adrian asked his boss if Bertram Books could take part in Jeans for Genes Day, he met with an enthusiastic response.The big day dawned and Dominic and I went along to take some photographs. As I walked into the building, I couldn't believe my eyes. Everyone was in jeans, including management! A big difference when everyone normally had to wear suits!

There was even a box on reception so that all the reps that came in could be encouraged to contribute! Although I heard a few 'moans' about not being able to sit down and having to hold stomachs in, they all seemed to enjoy the day! I left feeling proud and touched that so many people wanted to get involved. A total of £500 was raised and a few more people were made aware of MPS diseases, in particular, Sanfilippo disease which Dominic has.

I would like to say a special 'thank-you' to Amanda for organisingeverything.

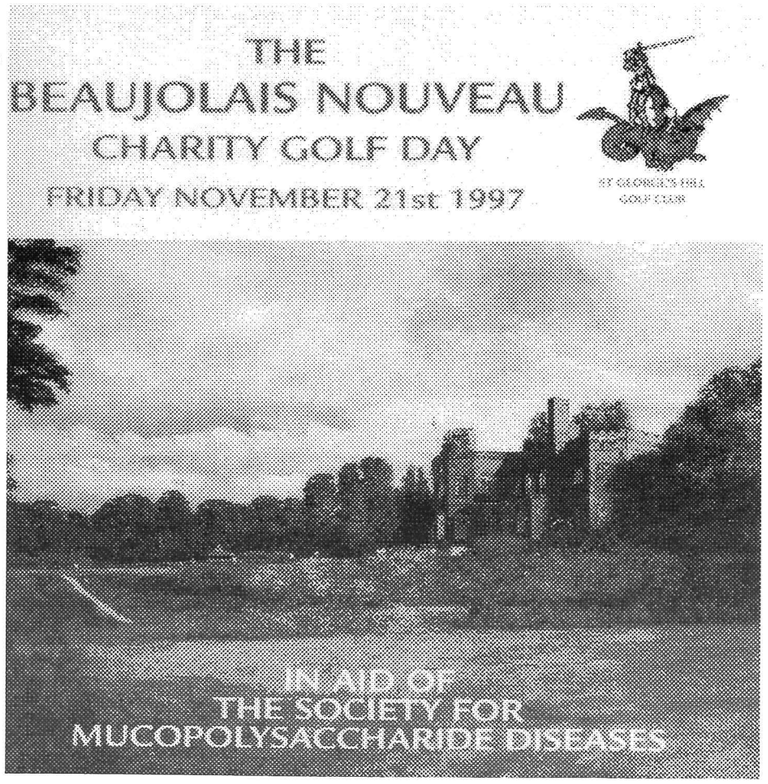
Here's to Jeans for Genes Day 1998.

Maianne Stimpson

FUNDRAISING

Charity Golf Day at St. George's Hill Golf Club

A Charity golf day was held at St. George's Hill Golf Club, Weybridge, Surrey on Friday, November 21st, in aid of the Society for Mucopolysaccharide Diseases. This proved to be a very appropriate venue, as the present Lady Captain of the Club, Heather Gordon, has two grand-children who suffer from Hunter Syndrome.



The Competition was timed to coincide with the arrival of the 1997 Beaujolais Nouveau, which had been collected from France the previous day. Twenty teams of four players each (male or female) competed for valuable prize of wine. A shot-gun start enabled everyone to sit down together to enjoy a three course lunch, whilst sampling the Beaujolais Nouveau.

After lunch Heather gave a most interesting and informative talk about Mucopolysaccharide Diseases and the work of the Society, and this was followed by a very successful auction and raffle.

An enjoyable day had been had by all and in the process raised a net sum of £4,745 for the Society.

Anne Cotton

Mrs Cotton has since held another Charity Golf Day on the 26th February 1998 when she raised £1,569 for the MPS Society.

We would like to thank Mrs Cotton very much for the superb effort she has made in raising the grand total of £6,314 for the Society

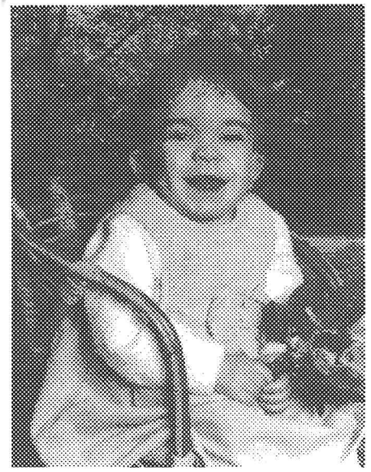
FUNDRAISING

Thank you to the Scunthorpe Streaker

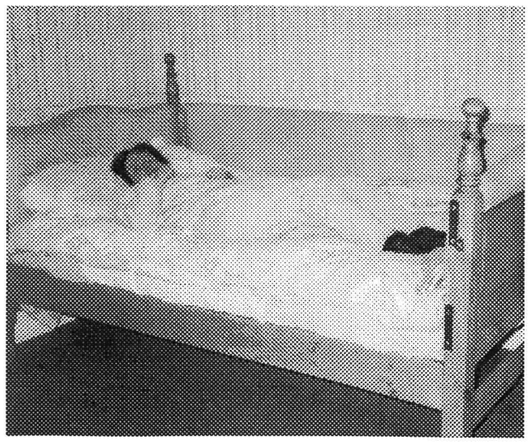
I have enclosed a cheque for £60.00 raised at the Crosby Hotel (where the Bike events have been held). It was raised unexpectedly on New Year's Eve when a collection was made prior to a man removing everything bar his tattoos and running naked through and around the two bars in the pub, much to the delight of the ladies, especially one who decided to make sure it was for real and not just her imagination! Could you please say a big thank you in the newsletter to the Streaker, customers and regulars of the Crosby Hotel.

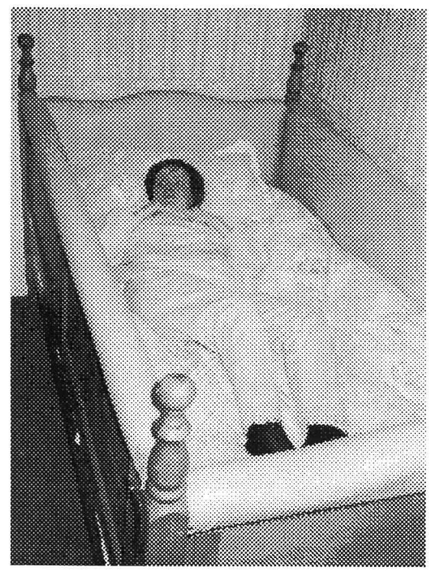
Barbara Rollison

P.S. Funnilly enough there are no photos of this event enclosed for the newlsetter!



Pictured above is Barbara's daughter, Gemma Rollison who died in July 1994 aged 7 years old. (Hurler)





Rosemary Nurse, mother of Lisa who is 16 years old and suffers from Sanfilippo Disease, kindly sent us some photographs of Lisa's new bed.
The bed was made at Pine Wood in Crystal Palace and cost £508 plus an additional £200 for the padding all round the bed.
The bed measures 6ft long and 3ft 6in wide.

FUNDRAISING

DONATIONS

The Society is grateful to the following who have made donations.

Vauxhall, South Wirral
C Gardner
K Hauley
M Hermans
John Ellerman Foundation
B Susijin
Nycomed, Amersham
Boots Charitable Trust
Cornwell Parker
Joseph Rochford & Sons Ltd.
Shell UK
J Avison
Pyke Charity Trust
Jenny and Andy Hardy
The Fitton Trust
F Wilson
K Maslen
A Paige
John Crawley
H Denman
Michael Skidmore
Blair Foundation
Alison Pickard
S Stephens
D Ellmore
I Vicks
Dawn Nelson
Atlantic Foundation
Clydebank Health Centre
Starlight Santuary
Alison Gunary
M Meaker
Holly Nowell
Fred Lacey G M Buses
N Wilson
S Mossman
Dr Bale
The Dee Group
Abbey National Charity Trust
Ron and Linda Snack
Tony Lockyer
Aimley Jail
Boys Brigade NI
St Mary& St Paul's Primary School
Dr and Mrs Kingstone
Mike & Anne Kilvert
Holly Bank School
R F Walker
Debbie Onion
Christine Butt
The Stocks Inn
Catholic Women's League
Anglea Marriott
Somerset Trading Standards
John Laing Group
Peter and Jean Maver
W D Close & Sons Ltd

STAMPS

DVLA, Wimbledon Ann Thompson, Darlington.
Dominic Stimpson's Family, Norfolk
Hannah Chisling's Grandmother, Hilperton Marsh
Sue Vivier, Tunbridge Wells

FUNDRAISING

FUNDRAISING EVENTS

The Society is grateful to the following who held fundraising events.

Anne Cotton, Surrey - 2 Charity Golf Days at St George's Hill Golf Club.
A Hardy, Ayesbury - Chrisitmas Play and Guides and Scouts
Joseph Kearney, Belfast- Oddballs Golfing Society
Maureen Lord, Saffron Walden - Charity Lunch
H Gordon, Weybridge - Golf Club Charity Event
Ecclesfield Priority Players - Christmas Coffee Morning
A Reed, Bournemouth - Freemasons Raffles etc.
Haddenham Morris Men - Show BOC - Parachute Jump
Louise Crogan, Stockport - Sale ofToys Ann Kirkpatrick. Antrim - Praise Service
St Georges Gold Club - Charity Event E Matthews, Herriott - Charity Event
Gavin Brown,Hemel Hempstead - Sponsored Parachute Jump
Paul & Barbara Harriss, Rochester - Sale of Stevens Dr Who items
Mark and Tanya Brown, Letchworth - Sponsored Cycle ride London-Paris
John Crawly, Hemel Hempstead - Sale of obsolete furniture
Rachael Wheeler, Reading - Raised by members of Reading Modern Sequence Dance
Gordon/Russell families, Birmingham - Arabesque Dance Academy, Raffllle
North East Families - Christmas Raffle Jenny Hardy, Haddenham - Soup Lunch
Hannah Chisling's Grandmother - Sale of homemade fudge and marmalade
Stokenchurch Middle School, Stokenchurch - Mufty Day
Marina Foster and Brian Baker, Bristol - Car Boot Sale
Crosby Hotel, Scunthorpe - Collection at New Year
Jean Cockman, Verwood - Collection of Alluminuim cans
Comber Liverpool Supporters Club - Sponsored Walk

CHARITY BOXES

Pat Lomas, Pinxton G Plummer, Cardiff Pam Croghan, Stockport
Ann Kirkpatrick, Antrim Val Brockie, Alcester Figure Shapers, Southampton
Harry Nurse, London

IN MEMORY

Mr M Andrews, Knareborough Marjorie Wookey, Hounslow
Christopher Read's Grandfather Helen Skidmore's Grandmother James Astbury
Jordon Mills, Windsor Gethin Robins M Kennedy's Mother Margaret Wright

SPECIAL OCCASIONS

Mr Varsani's Birthday Celebration

Area Support Families

East Anglia

Robert and Caroline Fisher
The Horrells, Great Sampford, Saffron Walden, Essex CB10 2 RL
Tel: 01799 586631

Julie Thacker
20 Herolf Way, Harleston, Norfolk IP20 9QA
Tel: 01379 854204

Zelda and Paul Hilton
17 Stanley Drive, Sutton Bridge, Nr Spalding, Lincolnshire PE12 9XQ
Tel: 01406 351524

South East

Robin and Mary Gooch
High Bank House, Swifehill, Broadoak, Nr Heathfield, East Sussex TW21 8XG
Tel: 01435 883329

Potteries

Bill and Sylvia Blackburn
11 Beatty Road, Nantwich, Cheshire CW5 5JP
Tel: 01270 626809

Lynn and Chris Grandidge .
41 The Boulevard, Broughton, Chester CH4 0SN
Tel: 01244 531163

South West

Bill and Fer Pidden
5 Westbury Leigh, Westbury, Wiltshire BA13 3SE
Tel: 01373 865117

Gordon and Anne Hill
Bowhayes Farm, Venn Ottery, Ottery, St Mary, Devon EX11 1RX
Tel: 01404 813823

Home Counties

Mark and Rachel Wheeler
26 Tamarisk Avenue, Reading, Berkshire RG2 8JB
Tel: 0118 9541293

Gavin and Denise Brown
395907
32 Ellingham Road, Adeyfield, Hemel Hempstead, Herts HP2 5LE
Tel: 01442

Wales

Ann and Michael Kilvert
Windy Waye, Nantoer, Newtown, Powys SY16 1HH
Tel: 01686 624387

Area Support Families

Midlands

Sue and Jeffrey Hodgetts
56363
6 Godolphin, Tamworth, Staffordshire B79 7UF
Tel: 01827

Zerina and Sajjad Shah
37 Lowe Street, Wolverhampton, West Midlands
Tel: 01902 656147

Monty and Doreen Russell
71 Templemore Drive, Great Barr, Birmingham, west Midlands B43 5HF
Tel: 0121 6864779

Yorkshire and East Coast

David and Monica Briggs
7 Humber Street Retford, Nottinghamshire DN22 6LZ
Tel: 01777 700046

Barbara and Trevor Rollinson
43 Crosby Avenue, Scunthorpe, Humberside DN15 8PA
Tel: 01724 864115

North West of England

Joanne and Gary Adshead
10 Church Lane, West Houghton, Nr. Bolton, Gt Manchester BL5 3PP
Tel: 01942 810109

Geoffrey and Selma Oulton
37 Saville Road, Liverpool 13, Merseyside
Tel: 01514 752941

North East of England

John and Barbara Arrowsmith
11 Penfold Close, Fairways Estate, Benton, Newcastle on Tyne NE7 7UQ
Tel: 0191 2921234

Ann Thompson
7 Sunningdale Green, Darlington, County Durham DL1 3SB
Tel: 01325 254985*

Elizabeth and William Armstrong
7 The Crescent, Hartlepool, Cleveland TS26 8LY
Tel: 01429 273703

Scotland

Cath and Jim McLean
"Woodlee" 47 Oakdene Court, Culloden, Inverness, Highland IV1 2XL
Tel: 01463 791816

Clint and Karen Stevenson
6 Viewfield Street, Harthill, Shotts, Lanarkshire ML7 5SN
Tel: 01501 752712

Alan and Fiona Byrne (*Telephone contact only*)
3 Jedburgh Avenue, Rutherglen, Glasgow G7 3EN
Tel:0141 5695376

Northern Ireland

Kieran Houston (Chairman)
21 Cavanalee Road, Strabane, County Tyrone BT82 8HB
Tel: 01504 884168

* Ann Thompson has a new telephone number 01325 254985

The Society for
Mucopolysaccharide Diseases

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