NEWSLETTER SUMMER 1989

THE SOCIETY FOR MUCOPOLYSACCHARIDE DISEASES

National Registered Charity No. 287034

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This newsletter is published quarterly and distributed free of charge to sufferers from MPS disorders in Great Britain and to their immediate families. It is also distributed around the world on subscription. Copies are sent to 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 nearly 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, and provides support to several research projects. It maintain links with sister Societies in Europe, the United States, 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".

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My article for the previous newsletter was curtailed by lack of space - the Editor rarely has this problem! This Newsletter is welcomed by you all so if you have not done so already, why not drop a line to **Charles O´Toole,** about who you are, what you do, and anything that can be shared by the members. I assure you it will make most interesting reading and anything on how you cope with ordinary living can give ideas and inspiration to other people. Some people find it difficult to express things in writing -- Charles will be only too pleased to help in editing your material!

On the back cover are the names and addresses and telephone numbers of the Area Support Families. Do not forget that if you have any problems or just want a sympathetic ear, to give your local family a call, or if they are not available, particularly during holiday time, try one of the other numbers. The family may not be able to answer your problems, but if not they will soon put you in touch with someone who can help. These comments are addressed mainly to new members but they do of course apply to everyone.

Alf G King Chairman of the Management Committee.

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DATES FOR YOUR DIARY

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9th July 1989	Family Day Out, Cotswold Wild Life Park. (Ron Snack)
17th July 1989	Dr Garrow´s Garden Party (see notice)
5th August 1989	MPS Group Holiday (fully booked).
1st Sept 1989	**Deadline for Autumn Newsletter.**
16th Sept. 1989	´Kids Count´ Fun Day at Battersea Park, London.
22nd of Sept. 1989	Annual Conference, Stoke on Trent.
31st August 1990	Second International MPS Conference UMIST, Manchester.

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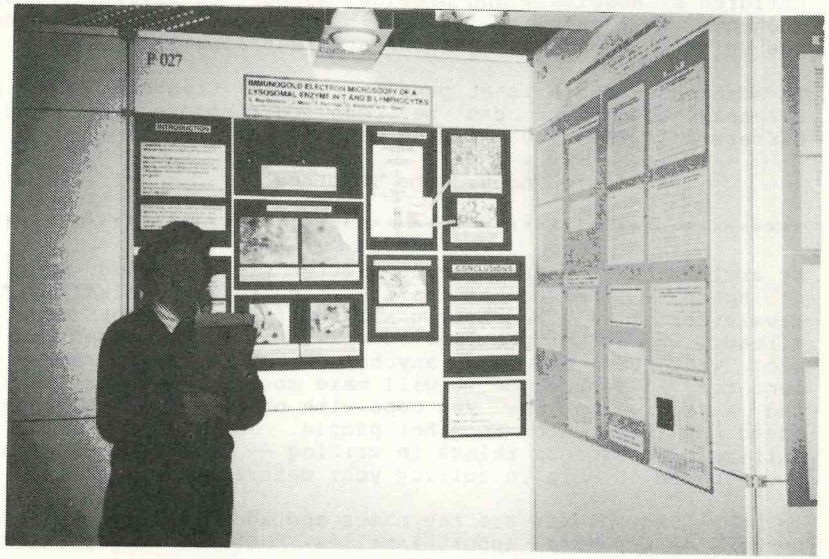
The absolute deadline for the Autumn Newsletter is the first of September so that it can come out in time for the conference.



Christine Lavery at the International Conference for Lysosomal Storage Diseases at Münster, Germany, in March.

garden fayre

"fun for all the family"

SUNDAY 17TH JULY

2p.m.~5p.m

Roughwood Farmhouse, Roughwood Lane
Chalfont St Giles

ADMISSION BY LUCKY PROGRAMME: £1
Children under 10 years - FREE

STOP PRESS!

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FAMILIES ARE INVITED TO COME AT 12.00 noon
To have lunch together
Before the Fair (Fayre?) begins

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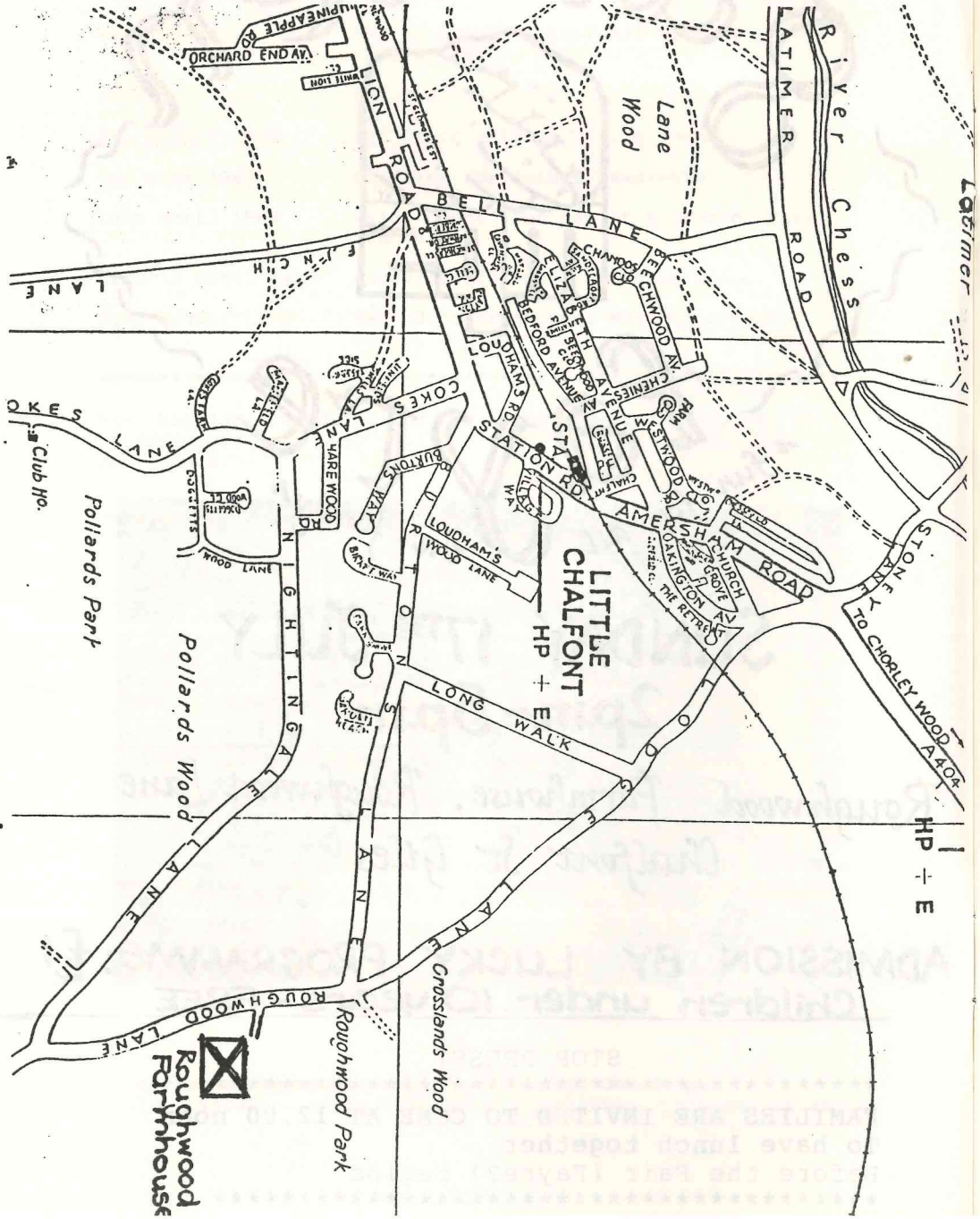

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NEW FAMILIES

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Mr and Mrs Rasen from Newark, Notts whose daughter **Michaela** was born in 1982 and suffers from **Sanfilippo Disease.**

Kevin and Vicky Berry from Eskington, near Sheffield whose baby **Michael** died from **Hurler Disease** in March 1989, only 3-4 weeks after diagnosis.

Mr and Mrs Hall from Ashton Under Lyme, Lancashire, whose nine month old baby is suffering from **Sanfilippo Disease.**

Jackie and Peter Poole from Frodsham, Cheshire, whose son **Alan** aged fifteen months is suffering from **Hurler Disease.**

Thelma and Geoffrey Oulton from Runcorn Cheshire, whose son **David** aged eighteen months has been diagnosed as suffering from **Hunter Disease.**

John and Carol Hicks, from Heamoor, Penzance who have a three year old son **Jamie** suffering from **Hunter Disease.**

Robert and Caroline Fisher, from Great Samford, Saffron Walden, whose son **James** aged three has been diagnosed as suffering from **Hunter Disease.**

Mrs Woods from Speke, Liverpool, who has a daughter **Leanne** suffering from **Hurler Disease.**

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DEATHS

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Christopher Pheeney, from Craigneux, Airdrie, who suffered from **Sanfilippo Disease** died on 26th April aged eleven years. Our thoughts are with his mother Frances, grandparents Betty and Tom and with Gabriel, Gerald and Noreen.

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To Mary O´Toole

Many thanks for the latest newsletter and for the booklets on Morquio, Hunter and Hurler/Scheie which I think are **EXCELLENT.** They are beautifully presented, clear and concise, and I think that the colour coded format is a brilliant idea and looks very smart. They are really marvellous and will certainly fill a great gap in the currently available literature.

Dr Diana Chase
National Institute for Medical Research

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BANGERS BEANS AND BONNETS !

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The Bryans, Butlers, Corbetts, Hardys and Snacks all got together on Sunday 12th March at Bradwell Common Community Centre. For the uninitiated Bradwell Common is that little part of Bucks that is forever Milton Keynes --V5 to V9 and up past the concrete cows.

Colin couldn´t believe his luck when dinner was served - bangers and beans with baked potatoes, hand picked by his dad at the MK Sainsbury´s Food Store.

Gateaux and ice cream followed. Poor old Dan wasn´t allowed a sweet - yet another diet (his third this year) so Jeanine had his portion, and his seconds and his.....
We even had a cabaret! Tom brought his guitar and Claire her flute and voice. Matthew was itching to have a go but was suitably restrained for most of the time.
Then everyone had an opportunity to make an Easter Bonnet. Gemma´s mum did her Blue Peter bit with card, feathers, ribbon, glitter and sticky backed tape. Hats off to all those who entered into the spirit of the occasion.
Nathan won a prize for his bonnet - and he hadn´t even finished it! But he did look lovely in the completed article - eat your heart out Gertrude Shilling.
With the wind blowing a gale and the rain pouring down outside, it was good to be indoors, and with such good company too.
Thanks Ron and Linda (and Linda´s mum and dad) for organising it. Roll on the warmer weather when we can meet again, this time in the open air.

Andy Hardy.

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Bryans

Butlers

Corbetts

Hardys
and
Snacks

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FAMILY DAY OUT

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On Sunday 9th of July there will be a Family Day Out at the Cotswold Wild Life Park at Burford in Oxfordshire. The plan is to meet in the car park at 11.30 am and to bring a picnic. Everybody is welcome to what promises to be a really good day out. Contact **Ron and Linda Snack** for further information.

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NORTH WEST AREA FAMILY DAY

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Fourteen families and friends in the North West had an enjoyable day in Nantwich on Sunday 9th of April at the Crowsfoot Centre.

After a buffet lunch the children were entertained by Paul Presto, a comedy magician, whilst the adults were able to circulate and have a good chat. It was lovely to see old friends and new. A warm welcome was extended to **Mike and Ann Kilvert** from Newtown who came with their daughter **Sarah** to their first MPS get-together.

Heather Pope, a dentist with Crewe Health Authority gave a very good talk on dentistry and dental hygiene for handicapped children, and she has promised to do an article for the magesine.

Here´s to the next event which will be a Barbeque on Sunday 11th June.

Sylvia Blackburn

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CANADIAN FAMILY

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Kharis and Malcolm Collins from Canada wrote to Christine Lavery in the Spring and sent her a photo of their son **Colin,** born 9th March 1975, who suffers from **Sanfilippo Disease.** They said that Colin likes to be cuddled and he does not suffer too much discomfort.

Unfortunately the Canadian MPS Society, founded in 1984 with encouragement from Christine, has had problems and has decided to close down. Kharis and Malcolm would be pleased to hear from other families in Britain with children suffering from Sanfilippo Disease. Their address is:

Kharis and Malcolm Collins
14706-34 Street
Edmonton
Alberta
Canada TSY 2KG

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BROOMES SWEEP NORTH - NIGIL CUTS SOUTH!

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On ´Red Nose´ day Roger and I were browsing about Barnsley Market, having a tremendous time. We were spending a few days in York and the trip to Barnsley was the highlight of the short break. Thanks to all who live in the North - we thoroughly enjoyed your hospitality.

Meanwhile, far away in the south **Nigil Wiggins** was having his head shaved in aid of charity. Our thoughts were with him. The MPS Society was to benefit from any sponsorship received at the College of Estate Management where I work with Nigil´s mother Margaret. He raised £125.50, with more to come. Our thanks to Nigil for his brave action. He came into work afterwards, just to prove he had the courage, and actually he was still as loveable as before.

Nigil has been a student holiday worker at the college for three years and so is well known to us. My thanks must also go to all at CEM who supported him, and to Margaret for eventually supporting him in his crazy action!

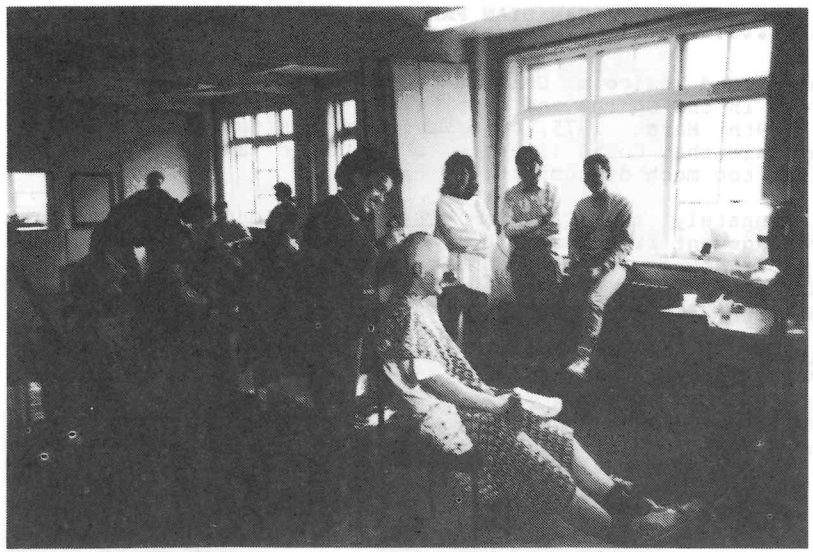
We wish Nigil every success in his studies at Ravensbourne College in Kent, and we will miss the pleasure of his company at work this summer as he will be on **genuine** work experience.

We hope all the MPS families we know are keeping well and we look forward to meeting up again soon. In the meantime,

Chins up,

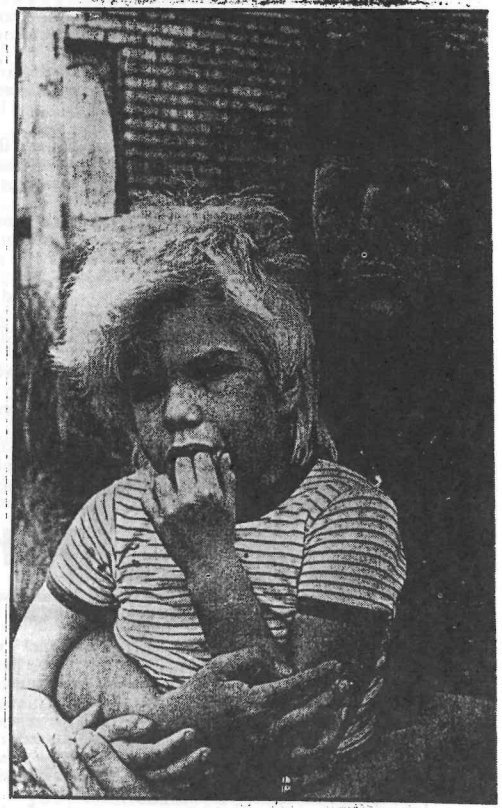
Jenny Broome

Nigil Wiggins bares all for MPS!



Focus on the incurable disease affecting one in 25,000 youngsters

Tragedy of the children with so little time



WENDY BEST and RODERIC ELDRIDGE talk to two families whose children suffer from the incurable Mucopolysaccharide disease.

IMAGINE that your bouncing baby is slowly dying from a rare incurable disease which will leave him severely mentally and physically handicapped.

Imagine your active toddler suddenly loses his speech, his joints stiffen, his face and body become grossly distorted, he loses the use of his legs and becomes incontinent.

Your child would be suffering from Mucopolysaccharide Disease, which affects only one in 25,000.

Aims

At the moment 450 children are suffering from the incurable illness which causes progressive handicap.

Children born with it often die before they are ten.

Christine Lavery formed the MPS society in 1982 after the death of her son from MPS II, Hunter's Syndrome. It is a voluntary charity receiving no help from statutory bodies. Parents work together to improve the quality of life for their child and those not yet born.

The aims of the society are to act as a parent support group, bring about more public awareness and raise funds in order to further the research into MPS.

Mrs Lavery, of Little Chalfont, Buckinghamshire, said, "Parents can take as much or as little from the society as they want. At this year's national conference 90 parents turned up, representing 120 children. We are in touch with about 90 per cent of all MPS children. We have financed a national post in Manchester for a consultant."

Children born with the disease cannot produce certain enzymes necessary for the correct chemical changes to take place in their bodies.

The MPS diseases are inherited from healthy parents who have no idea they carry the genetic defect.

If both parents have the defective gene there is a one in four chance of every child being affected.

There are several main types of the disease, Hurler, Scheie, Hunter, Sanfilippo, Morquio, Maroteauxlamy, and Sly, and associated diseases Mucolipidosis, Fucosidosis and Sialic Acid Disease.

Youngsters with Sanfilippo Syndrome are often not diagnosed until they are four years old.

Research

There is no cure and the society is always raising money to pay for research in to the disease.

Regulars at Filos pub in London Road, Bath, have organised a charity barrel push between Union Street and Southgate on Saturday, and a disco will be held at the pub in the evening from 8pm.

They plan to push the barrel through the town centre to raise money for the MPS society.

Anyone wishing to contact the society should call Christine Lavery on 02404 2789

Help for the future

FIVE-year-old Abigail Pullin from Colerne is suffering from the incurable wasting disease.

The youngster from Poplar Way, who is pictured above with her mother Alison, is hyperactive, has trouble sleeping, has lost her speech and is now incontinent.

She has the Sanfilippo Syndrome of the disease which affects the nervous system.

In August the village rugby club and Abigail's parents started organising a series of fund-raising events to pay for research in to the killer disease.

Mrs Alison Pullin said then, "I wanted to do this fund-raising because it is a p aspect. I know there is no hope for A or others like her who already have the d but we can still help those yet to be

She said that at first Abigail's disti features and bright blonde hair were s cute, but they soon realised somethin wrong with her.

"We were told of the diagnosis whe went for pre-school check-up," she sa

Abigail's features — a squat face and c hair — are typical of sufferers.

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Please note **Stella Hale´s** change of address inside the front cover.

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'He has more minders than Arthur Daley'

Smiling in the face of adversity: Jenny and Andrew Hardy with daughter Rebecca and son, Matthew, who suffers from Hunter's Syndrome.

LITTLE Matthew Hardy is blissfully unaware of his tragic future.

Because he suffers from an extremely rare disease he will never grow up. His life is in reverse.

His parents, Jenny and Andrew, suffered a strange kind of bereavement when they discovered their beloved son would not live beyond his teens.

Now they devote their time to making life as normal as possible for Matthew, who suffers from a degenerative condition called Hunter's Syndrome. It has affected him mentally and physically.

The couple have learnt to take life one day at a time - the future is something they prefer not to think about.

Matthew is so endearing it is difficult to accept he has such a hopeless future.

When I visited the family at their Haddenham home, seven-year-old Matthew was totally unreserved about his displays of affection for me, a person he had never met before.

He put his face close to mine and held my hand. A new visitor was fascinating to this happy little boy, whose brain has been affected to the extent that he knows little about his illness.

Hunter's Syndrome is one of several Mucopolysaccharide Diseases (MPS for short). They are rare, life threatening and cause severe progressive handicap, often both physical and mental.

Children born with these diseases cannot produce certain enzymes necessary for the correct chemical changes to take place in their bodies. Complex sugars become stored in all types of connective tissue causing progessive damage.

They are inherited from healthy parents who have no idea they carry the genetic defect. If both parents have the defective gene there is a one in four chance of every child being affected.

But with Hunter's Syndrome the mother is the unknowing carrier.

Like other MPS children, Matthew was born perfectly normal and healthy.

"As he was getting towards the age of three he was late with his toilet training, which concerned us," recalls his mother. "He was also late walking as a toddler and late talking."

When the couple took him for a check-up they were told not to worry because boys are often late developers.

But the doctor decided to send Matthew for tests and he was diagnosed as having Hunter's Syndrome.

"We never dreamed it would be anything as serious - we just expected to be told he was a late developer," says Jenny.

"After that he did progress, albeit very slowly. But he had no co-ordination,"

Gradually the disease set in causing stiffness of the joints, mental regression and respiratory and hearing problems.

"Over the past 18 months he has deteriorated more," Jenny told me, "Before that he was using seven or eight word phrases, now it's just the occasional word. It's been a noticeable deterioration."

It is touching but bizarre that traits of his old self remain. He's still able to repeat his favourite songs and nursery rhymes from memory. Matthew's favourite pastime is to stand at the window with his nursery rhyme book and sing every verse word perfectly.

Before his illness he loved to sit and do jigsaw puzzles. Now he can only complete them with a lot of help from his parents due to lack of co-ordination and the deterioration of his mind.

Music is another subject close to Matthew's heart.

His favourite television programme is Top of the Pops, but he'll listen to any kind of music from Songs of Praise to orchestras.

After attending the village school the decision was made to send him to Stoke Leys Special School in Aylesbury. A classroom assistant is always there with him to look after his needs.

Matthew is one of the most popular boys in the school and in the village for that matter.

"He has more minders than Arthur Daley," jokes his father, "people in the village are also very fond of him and always wave when they walk past the window."

The couple had already decided to have their second child when Matthew's tests came through.

"The doctor told us that the best thing Matthew could have would be a little brother or sister," says Jenny.

When she fell pregnant, tests on the unborn baby were found to be negative.

Sadly, some parents have two or more children with the disease. One particular MPS syndrome does not manifest itself until the child is nine or ten - by then the couple may have had more children.

Three-year-old Rebecca knows her brother is different and helps him as much as she can.

"She already understands there are certain things he is not able to do and she will help in her own little way. One day she said 'Matthew can't talk very well can he'. If he starts running away she chases after him," says Jenny.

But the couple are determined not to put any pressure on Rebecca to look after her brother.

"We don't want her to feel she's got to grow up being made to do it. She's got to want to help."

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David Criddle: By his mother and sister.

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Recently I received the Spring MPS newsletter together with a booklet on Hurler/Scheie Disease. I read through it with interest as my son David has Hurler Syndrome. At the end of the booklet I found a piece I had written when he was five years old. I suddenly realised how things had been changed in the last five years. David is now nearly eleven and I know that time doesn´t stand still, so I´ve decided to write an update as to what´s happened in the last five years.

When David was five he attended a local assessment school called "Presswood House." It was a very close knit, friendly school, where David was on the same mental level as the other children. The headmistress Miss D´eath was very helpful and friendly towards David and the family. With a great deal of effort from her and the other teachers David was taught the basic skills like reading simple words, matching pictures, drawing and some speech. All three surprised the doctors and consultants who were convinced that these things were impossible for David to learn.

I decided to try and keep things as normal as possible for as long as we could. We continued taking David on holiday every year to Spain. He has travelled great distances, especially to America with me, for a holiday for terminally ill children, sponsored by Les Evans. (David still talks about Mickey Mouse three years later.)

Unfortunately we can no longer take David away any more. He is too restless and wants to stay at home in familiar surroundings with his TV, toys and videos. David has to be occupied and requires a lot of attention.

The problems became worse when David had to leave "Presswood" to go to Pentwyn Special School for handicapped children. This school didn´t meet his special needs and the headmistress wasn´t very helpful. She insisted on treating David like a normal ten year old, when really his abilities were those of a three year old. It was during this time that we noticed a lot of changes in David. First, all of a sudden his sight became worse, and he didn´t have the ability to adjust his eyes when it became dark, so he kept asking for the light on all the time. He was offered a cornea transplant, but there was no way we could accept it. He would never have survived the anaesthetic and the transplant would probably have been no benefit to him. Sometimes David would end up talking to an empty chair because he couldn´t always tell if anyone was there or not, but we could all see the funny side of this, even David.

The best part is when he mistakes me for Jane, which makes my day! Apart from losing his sight he is also losing most of his reading skills and his short term memory, although he is good at remembering things from years back. He has lost some control of his bowels and sometimes if he laughs too much he has "a little accident", so to be on the safe side we put a nappy on him when we go out, much to his disgust! David has slowed down considerably and is especially slow in the mornings. He is beginning to lose the use of his legs and he walks with a gait, but he seems to have adapted to this disability very well indeed and still manages to run around.

David was not happy at school, so I looked around and found another school, "Ty-Gwyn". The headmaster Mr Dwyer was very understanding. David is in the toddlers special play unit with other handicapped children of the same mental age. Here he is able to play in the sand and draw and do all the things he loves all day long.

Recently David and I paid a visit to the genetics unit in Manchester, where we saw Dr Wraith. He explained fully and in great detail everything about David and his condition. I learned a lot as he was very honest with me, drawing pictures to show David´s situation. It is obvious that David is not the same as he was. He has an enlarged heart and a heart murmur and sometimes when he runs around or gets too excited he goes a funny purple colour! What we do now know is that David is in excellent condition for a child with his disease.

However this nearly all came to an end when David caught measles very badly. David´s doctors, Glenys Lindsey and Peter Groom, were very supportive and did everything they could to help. They made sure we knew what was happening and only took David into hospital when it was really necessary (because they thought he was dehydrating). David didn´t eat for about a week and was drinking less than half a cup of coke a day. Everyone thought it was a miracle that he could survive on so little, after what could be a fatal infection for children with his disease. Somehow David pulled through, but he has been left slightly weaker and for a while not as bright as he was. His sight has worsened and I hope he will not lose it totally.

A lot of things mentioned in the Hurler/Scheie booklet apply to David and it is helpful to know that these things are possibly going to happen. As he is getting older lots more things are going wrong. Frequently we have slight upsets at dinner time when David chokes on his food. (He insists on eating only bacon and drinks only coke!) We try to keep David away from hospitals and doctors as much as possible in order to keep things normal.

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27 Edna Ave
Fazakerley
Liverpool 10

Dear Charles,

It has taken me a long time to put pen to paper. I also find it hard to put what I feel into words. There are so many people I would like to thank for their kindness, support, reassurance and love shown to my daughter Julie my family and myself during the recent sad loss of my darling grandson Anthony. Without the support of the MPS society I really don´t know what we would have done.

Firstly I would like to thank Christine and Mary for being there with Julie and myself. The easiest thing for them would have been to run, but no, they stayed and supported us both through a very sad ordeal. No words can say how grateful we feel. I would also like to thank you and Mary for opening up your home for us all and making us feel a part of your family. My thanks to Mary Gardiner for being there when we arrived back in Liverpool, her love and support has been never ending. My thanks to Dr. Wraith for his reassurance and support to us both. I could go on and on, so I will end by thanking all the MPS families for their many cards letters and phone calls received.

On behalf of my daughter Julie and my family thank you all once again and God bless you.

Regards,

Diane Bayliss (Granparent contact)

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David Criddle (continued)

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He goes to see Dr Groom every so often for a "service", just to check that everything is in order. The doctors are always honest and constructive. The health visitor Mrs Mitchell calls to check that everthing is OK and just to see if I need anything.

One of the surprising things about David is that he has not lost his speech, in fact he seems to learn more and more words. He surprises us with words and sayings he has picked up from the television and from us. He is also very choosy about the clothes he wears, so in some aspects he is a normal ten year old.

Despite all these problems that he has and the way he is, it doesn´t change the effect that he has on the people he meets. The one thing that is perfect and which leaves a mark on everyone is his impeccable manners and his beautiful smile.

Jane and Pat Criddle

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TUESDAY´S CHILD IS FULL OF GRACE

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This is about **Tim**, a child who holds the hearts of many, who has a profound hold on the hearts of those who meet him, a child whose eyes really do mirror his soul.

His eyes express great understanding, an understanding far beyond his years. I feel priveleged to know Tim. He is three and a half years old. He has the body of a much younger child.

He is classified as handicapped. When I am with him I feel he is not handicapped. I feel that Tim is gifted. I feel it is I who am handicapped.

Tim is beautiful. His eyes laugh and smile at times of pleasure. He communicates just enough to keep attention. His eyes hold your heart. His gaze shows hidden depths of secret knowledge which I long to share.

I believe that God gave us all a job to do in life. Tim´s job must have been to bring great tenderness and beauty to this world. Before I met Tim I saw handicapped people as people to feel responsible for --- as people to be cared for, also as people to be a little wary of --- a fear of the unknown I suppose.

Through Tim I have come to see the remarkable beauty of these special people. It seems these people are chosen especially by God to show how dependence can nurture understanding and tenderness within the coldest and hardest heart.

Tim fulfills this purpose well. It would truly be a perfect world if we were all as diligent as Tim at doing God´s work. I love Tim for being just Tim.

Margaret

(Timothy Bryans is aged three and a half and suffers from ML II. He lives with his family at Borehamwood in Hertfordshire. His photo appears in the Winter 1988 newsletter.)

LONDON MARATHON

Could anyone who is collecting money from sponsors for the London Marathon please try to get it all in as soon as possible so that we can get a grand total together. I believe that we had five people running for us, although I didn't manage to spot any of them on the tele.

Ron Snack

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CHASING THE ENZYME (or The Hunting of the Snark)

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Dr Diana Chase spent five years trying to purify **the Hunter** enzyme. The last eighteen months of this work was funded by the Society. Sadly, Diana has had to abandon the attempt. She has written the following account of her work for the newsletter.

The active Hunter enzyme is one of many thousands in the body. At the 1988 parents conference I compared the task of separating that one protein from the mixture of proteins in the body to the task of separating the pinch of salt from the dry ingredients of a fruit cake mixture. I mentioned that two litres of amniotic fluid contains about twenty grammes of protein, of which I thought then that perhaps 0.0001 g. was the enzyme itself. I am now sure that this is a ten to a thousand fold over-estimation of the quantity of the enzyme: there is probably only 0.00001 g. or even 0.0000001 g. of the enzyme in two litres of amniotic fluid, i.e. 0.1 - 10 mg.

During any protein purification some of the protein is lost, just as when sieving flour some grains stick to the sieve. After all the steps involved in the purification procedure, I estimated that I had about 10% of the amount of enzyme that I´d started with (this may sound dreadful, but in protein purification terms, it isn´t too bad at all!) Thus, even if I began with the upper estimate of 10mg of the enzyme, I ended up with 1mg. This is a very small amount indeed and in fact too small for a sample to be clearly "seen" using the current means of detecting proteins. However it does explain a curious observation: frequently, after completing a purification, and removing all the unwanted proteins, which should have left just the Hunter enzyme, there was apparently nothing left! As you can imagine this was extremely frustrating. The answer is that the enzyme was really there, but in such a small amount that it could not be seen.

Machines that specifically analyse proteins can now use very small quantities of protein -- but not that small! Generally, if we can´t see the protein, then the machine can´t either. A single analysis on one type of machine could require about 10mg. of this protein. But realistically, more than one analysis would be needed, preferrably on two or three types of machine, to get valuable data, so you would need ten times this amount, ie 100mg. As I´ve mentioned two litres of amniotic fluid can, at best, give only 1mg. of the enzyme and even this might be a gross overestimate. So to obtain the required amount of enzyme I would to do at least 100 purification "runs" and use a minimum of 2,000 litres of amniotic fluid!!!

It takes two weeks to do a single purification run, so this work alone would take over four years to complete. Clearly this is impractical.

The quantity of Hunter enzyme in amniotic fluid is far too small for purification purposes. (However there is obviously sufficient enzyme there to determine whether or not the foetus is affected with Hunter´s Syndrome, which shows how sensitive and valuable that biochemical test is.)

For practical purposes, in order to purify the Hunter enzyme the starting material should contain at least 100 times the quantity found in amniotic fluid. With this in mind, I measured the amount of enzyme produced by a special type of human cell that can be grown in the laboratory. Unfortunately there was only about the same quantity of enzyme as in amniotic fluid, so those cells would not be any better as a starting material than amniotic fluid.

We know that the normal Hunter enzyme is present throughout the body, but perhaps it is there in far smaller quantities than we have so far thought. Maybe this enzyme is extremely efficient and only minute quantities of it are needed. Many research groups have tried to purify this enzyme, using a variety of different starting materials (blood, urine, placentas etc.) but, as yet no one has succeeded. It might be that there just isn´t enough of the enzyme anywhere in the body for it to be purified using current technology.

Identification of the normal gene is still a valuable goal, but perhaps the best route now is via analysis of the chromosome (DNA), rather than in analysis of the enzyme. There are many laboratories investigating numerous genes on the X-chromosome. eventually all the genes will be located and analysed in detail, including the normal Hunter gene, although this may take several years. The route to a gene via enzyme/protein analysis is more direct and thus usually quicker, but in this case the enzyme route is currently proving to be impractical.

In conclusion I would like to say how very grateful I am to the members of the MPS Society for all their support, not only financial, but also for their great interest and encouragement given to me throughout the five years that I have been grappling with this enzyme. I´m very sorry not to have achieved the goal I set for myself.

Dr Diana Chase

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JEMMA´S DIARY (Part 9) continued from previous newsletter.

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February 19th 1987

Does Jemma watch you cook? She had the whole sequence off pat. Finding saucepan and spoon, turning cooker knobs on, waiting for it to cook, stirring, turning knobs off and then serving up.

March 20th 1987

Thanks for the chat last night. Another anecdote I forgot to tell you. Jemma is chief nose wiper - other peoples noses. Sometimes she goes all mumsy and does a quick wipe job on those who need it; other times she tells me imperiously "Colin tissue", or "Ian dirty". Sometimes she checks up on the finished job.

We had a visit from the school dentist who brought an enormous pair of false teeth as big as a bath brush. Jemma thought it a huge joke.

23rd March 1987

Sarah to Julia

This morning after I had tied Jem´s hair she inspected her appearance and said "Oh nice!" I said yes, you can show Julia. She replied, "No, Ashley", with such feeling in her voice. Isn´t young love wonderful!

30th March

We had a lovely time today telling the children about pussy willow and catkins. Ashley insisted they were rabbits! He tried to help Jemma put it in a vase but Jemma insisted on doing it herself. We took delivery of a large dolls cot. Guess who decided to test it for size? Ashley helped. He even took her tea in bed. When it was Ashley´s turn Jemma decided to play hospitals. Tummies were displayed, which Colin thought a hoot, but refrained from joining in. Jemma even listened for Ashley´s baby. I hope our role descriptions of Mummies and Daddies fitted. I probably only got it right that Mummies have babies and Daddies don´t, because Jemma told me that Daddy does the washing up.

Jemma´s really our "blue eyed girl". It is such a joy for Jackie and me to tell her to go and do something and know that the instruction will be carried out. We have rather a lot of walk abouts in class lately. Jemma´s regular one is a detour coming back from lunch. She usually goes for a quick brush up on office skills with Pat Wheeler, but generally gets back to the right place at the right time.

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JEMMA´S DIARY (Part 10)

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7th May 1987

Thanks for the lovely flowers. After smelling them and touching them some even tried to grab them, but Jemma was there with her "stop it´s" and handbag bashing technique. She remind me of one of those grannies in the Beano.

This week I´ve moved Jemma next to me at table. I thought it was about time we worked on getting her to use her knife and fork. She resented it at first and threw both knife and fork across the table. I took her plate and moved it to the centre of the table and continued eating my lunch. I didn´t say a word. She tried to cry, looked at me, waited a little, and then realized she was hungry, stopped crying and tried again - fine. She then tolerated any help I proffered. She actually refused to eat her lettuce until I put it on the fork.

22nd May 1987

This week she dropped some stacking beakers accidentally and then proceeded to stack them back properly. No problem. Doing the same with a set of barrels was a little bit more difficult because she forgot to put the small barrel in first, but we shall work on it. Everytime she plays the little mother the sequence gets extended. Now the baby gets winded and the cups get washed up.

11th September 1987

At last a routine is gradually emerging and Jemma is the one who is helping most to establish it. This term she has preferred to have a quick play in the house corner and then sit down to table tasks. Her concentration has improved over the holiday and as one thing has finished she has demanded more. Her least favourite is doing jig-saws.

PE is no trouble. She tried all her old tricks and copied some new ones like sitting astride a very narrow form and pulling herself along, also crawling to the top of a sloping form attached to the wall bars and shouting to me to look.

14th September 1987

Sarah to Julia

Have you taught her to tell the time? She looks at her watch - thinks - and tells me what time it is! I hope the novelty wears off soon. Jemma is flower arranging for a show on Saturday.

To be continued.....

Car Boot Sales

An increasingly popular way of uncluttering your garage, shed, or spare room is to take everything along to a "car boot sale".

There is often no need to book a pitch - just keep an eye open for adverts in your local newspaper and turn up on the day. In our area there is a sale every Sunday at the local hospital car park, so the hospital benefits as well as those doing the selling.

Simply ask all your friends and neighbours for their unwanted antiques or objets d'art, and get as much money as you can for everything on the day. One or two MPS posters displayed prominently beside your pasting table will always help get people's attention and interest.

Ron Snack

Sponsored Events

As you will have seen in the list of fund-raising events sponsorship is a popular way of raising money.

As well as the more obvious sponsored events like the London Marathon,why not have some fun by arranging an unusual sponsoring. I am sure you can all think of some amazing things to do, but how about involving the kids. Eating Baked Beans, one at a time, using your fingers is a good starter - just think how much you can make, even at 1p for five beans.

How about getting all the children in school looking for four leaf clovers on the playing field - could keep them quiet for hours (well, minutes), or see how many items they can collect in a matchbox. I'm sure you'll think of loads more.

The main bugbear with sponsorship is that you always seem to be approaching the same people for cash. The secret is to get other people to actually do the events then they can go round their own group of friends.

Ron Snack

SAINSBURY'S

Please don't forget that we are an "authorised" charity with Sainsbury's. This means that they will look favourably on any request from members to collect outside their stores, or even inside in some cases.

Many Sainsbury's stores now have their own car park and area around the store, so all you need to do is approach the local Manager with a date when you would like to collect, then contact me and I will set it up with Sainsbury's Head Office. I have lapel stickers and collecting tins available.

You never know, some of the stores may even take up our cause themselves.

Ron Snack

Fund Raising Events.

Once again, a varied list of activities which have raised money for us recently. I feel sure that you can add to the growing number of ways that we use to obtain our funds.

M Horsley	Collecting Box, Cumbria
Mr & Mrs Love	Harmony Wirksworth Singers
Mr & Mrs Sutcliffe	Middleton WI, Derbys, Coffee Evening
Mrs Todd	Collecting Box, Glasgow
A Byrne	Westburn Nursery, Glasgow, Collecting Box
Effie Whitham	Collecting Box
J & J Norton	Marathon
Baines C of E School	Sponsored Spell
W & M Griggs	Collecting Box
P & G Freer	Book Party
C & M O'Toole	Collecting Box
S Blackburn	Sponsored Swim
Collern Parish Council	Sponsored Knit
S Lowry	Carol Singing
Collern Rugby Club	Marathon
Fawcett	Wetherby Half Marathon
D Brooks Daw	Patchwork Quilt Draw
Miss E Gould	Sponsored Swim
N Wiggins, Reading	Red Nose Day
Justin Eyre, Bristol	Sponsored Cycle Ride
Westfield U R Church	Collection
P Isaac	Valentine Dance & Jacket Potatoes
G Townsend	Sponsored Slim
Wembdon School, Bridgwater	Carol Singing
Downend Ladies Enterprise	Coffee Evening
C Gooch	Collecting Box
C S Smith, Taunton	Perfume Sales

Good Home Wanted

Is there anyone out there who will offer a good home to an old, well used, portable typewriter. It has given good service, but I have moved on to bigger and better things. If anyone could make use of this please give me a ring.

Ron Snack.

Flag Days

After quite a lot of research I have come to the conclusion that it is virtually impossible to organise a national flag day. Instead it has been decided that we will make use of the Mental Health Day umbrella and let someone else do all the work applying for licences, go along and collect and then give them five per cent. This does not mean that we are not interested in arranging flag days for ourselves where anyone wants to, just contact me for details.

Morden.

LANCASHIRE 7 PM 10 AUG 1989

Mrs C. Lavery.
one M.P.S. society,
30 Westwood DR.
little Crayont.
BUCKS.

Dear Charles,

This envelope was received in the post unsealed and empty. Could we please put a copy in the next Newsletter asking if anyone recognises it to get in touch.

Christine

Payment of Fares to Hospital

Mrs Janis McCulloch is a CF Adult and represents Scotland on the Association of CF Adults Committee. She works in a Citizens Advice Bureau. She has written the following very helpful resume on the vexed question of fares to hospital. I sincerely hope families on a low income will find it helpful.

People in receipt of either Income Support or Family Credit automatically have their travel costs, to and from hospital, for N.H.S. treatment paid. Payment is made at the hospital on production of bus/train ticket and order book or the letter that comes with the girocheque for those paid by giro. Unemployed people should take form B.3. Those who are sick should take form C.3.

Help may also be available for those on a low income who have savings of less than £6,000. Further information is available from form H.11 — N.H.S. hospital travel costs. Claims can be made on form A.G.1. All these forms are available from Social Security Offices, Family doctors, N.H.S. hospitals and The Agency Benefits Unit, D.S.S. Longbenton, Newcastle-upon-Tyne, NE98 1YX. If eligible, claimants will be sent back a certificate showing entitlement to full or partial help (either form A.G.2. or A.G.3.).

N.B. Those aged 16 or over may claim on the grounds of their own low income even if they still live with their parents.

Health authorities (in Scotland, the ambulance services) have a duty to provide, or arrange provision of suitable transport, free of charge to the nearest suitable hospital for patients who are considered to be medically unfit to travel by other means.

Requests for non-emergency transport will be met where a doctor, dentist or mid-wife certifies that ambulance transport is necessary. Children being conveyed by the ambulance service should be accompanied by an adult whenever possible.

The Agency Benefits Unit accept that hospital visiting is not only desirable but generally regarded as a social duty. Consequently when assessing a claim for fares to visit a patient in hospital, provisions for meeting the expense is normally on the basis of the actual number of visits made. Where the patient is a child, allowance is usually made for daily visiting by one parent.

If there is not enough money to pay for fares to hospital in the first place CRISIS LOANS are available for those without sufficient resources, whether on Income Support or not, to meet their own and their family's immediate short-term needs, at the date when the application is made. In some circumstances Crisis Loans can be given for living expenses for up to 14 days. Applications must be made in writing by the applicant either on an approved form from the D.S.S. or by letter. A decision should be taken as quickly as possible.

A COMMUNITY CARE GRANT may be available to help with the travel expenses to visit an ill person in hospital, for those on Income Support. These grants are non repayable. Applications should be made in writing by the applicant on an approved form available at any D.S.S. office. The decision should be made within 28 days.

With acknowledgements to the Cystic Fibrosis Newsletter: Editor.

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Mrs C Lavery
MPS Society
30 Westwood Drive
Little Chalfont
Bucks

20 March 1989

Dear Christine,

I would be most grateful if you could advertise in the literature produced by your organisation details regarding our Parents Conference to be held on 23rd/24th September 1989 at Crewe and Alsager College, Crewe. Cheshire. Please feel free to edit the enclosed programme.

Thank you for your assistance in this matter.

Yours sincerely,

Lesley Greene (Mrs)
Hon Sec - Founder - Parent Counsellor

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STAMP NEWS

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I would like to thank everybody who has been saving their used postage stamps. We are still working hard at them so do keep sending them. They have been raising quite a bit of money for MPS.

So get saving and bring them along to the conference if you are not able to send them on.

I would also like to take this opportunity to thank Paul's parents for helping us with the stamps. We have had so many we really have been snowed under. So they have really been a great help.

Carol and Paul Hubbard
71 Preston Rd
Harold Hill
Romford RM3 7YU

****

Kids Count

Bringing happiness to kids who have so little - yet give so much.
Charitable status applied for.

Registered Charity Number 327868

Britannia Fun Day

Mrs Christine Lavery
The Society for Mucopolysaccharide Diseases
30, Westwood Drive
Little Chalfont
Buckinghamshire

Dear Mrs Lavery,

In London's Battersea Park, on Saturday, 16th September 1989, 'Kids Count' will once again be hosting a fun-packed celebrity party for terminally and seriously ill, physically and mentally handicapped children. Last year's event was a great sucess despite the bad weather!

We would like to invite you and your children to attend. The party will be the same as before, however, the entire event will be hosted under a Big Top in order to prevent a repeat of last year. The party will start at 2.30 p.m. and last until 4.30 p.m. During this time the children will meet many Pop and sport celebrities; be entertained by top class circus acts and performers; join in fun games and eat as much as they want.

There will also be a 'Fun Run' that all children (whether wheelchair bound or disabled) are invited to participate in. The aim of the run is to give to each participant a sponsorship form. The child will then be sponsored to complete the course (no pressure is to be put upon the participant to win, merely to compete) and the money they raise will be donated to the charity of their choice, (i.e a child with Leukaemia to be donated to Leukaemia Research). If you let us know along with your numbers of attendees, who will be joining in the 'Fun Run', we will send you the appropriate number of sponsorship forms. We are eager for as many children as possible to enter the 'Run' and so if there are any problems, please do not hesitate to contact me. All correspondence or queries should be addressed to me at 192b, Stamford Hill, London, N16 6RA, tel. 01 800 5495.

We would be grateful if you could let us know as soon as possible how many children/helpers will be attending so that we can organise things accordingly.

Thank you for your support last year. We look forward to hearing from you and seeing you again this year.

Yours sincerely,

Mary Robey (Miss)
Childrens Co-Ordinator

55 Glencoe Avenue, Newbury Park, Ilford, Essex 1G2 7AL

TAKING A BREAK

A guide for people caring at home

Do you help to care for a relative or friend who is elderly, ill or disabled?

Do you sometimes feel the need for a break, but don't know how to go about it? If so, this booklet can help you.

Looking after someone often involves hard physical effort and long term commitment. It is an experience which can arouse strong feelings in both you and the person you care for. It's only natural that you should want and need a rest from such demanding work. But it's not always easy to admit this or to find an acceptable alternative to your care.

This 36 page booklet explores these issues. Based on carers' experiences, it offers information and practical advice which will help you and the person you look after to arrange a break. It includes:

- descriptions of over 20 different types of break
- information and advice on how to make practical arrangements
- hints on coping with emotional worries and resolving practical problems
- ideas for improving services for carers and the people they look after
- an extensive list of useful contacts and national organisations

You can obtain your copy from **Taking a Break, Newcastle-upon-Tyne X, NE85 2AQ.** The booklet is free of charge to people who are themselves caring for relatives or friends who are elderly, ill or disabled. Others (such as professional workers) should send £0.60 (sixty pence) to cover costs (cheques/POs payable to King Edward's Hospital Fund for London).

ORDER FORM

Please tick one box

☐ I care for a friend or relative who is elderly, ill or disabled. Please send me a free copy

☐ I would like to receive a copy of the booklet but am not myself a carer. I enclose a cheque/PO for £0.60 (sixty pence) to cover costs

Name _____

Address _____

_____

Postcode _____

Send to: Taking a Break, Newcastle-upon-Tyne X, NE85 2AQ.

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Tel: 0223 834570

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

Mary Gardiner
15 Sidney Avenue, Hesketh Bank,
Nr. Preston, Lancs. Tel: 0772 815516

John and Barbara Arrowsmith
140 Newtown Rd, High Heaton, Newcastle on Tyne. NE7 7NH
Tel: 091 2812062

Sean and Pauline Mahon
41 Stumperlowe Crescent Road,
Sheffield 10, South Yorkshire
Tel: 0742 304069

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Tel: 0908 666819

David and Michelle Brooks-Daw
St.Mary's Lodge, Wellington Rd, Taunton,
Somerset. TA1 5AS Tel: 0823 271661

Tony and Mary Lockyer
29 Llanberis Close, Tonteg, Pontypridd, Mid Glamorgan.
Tel: 0443 2032845

Area Family for Northern Ireland:-

Margaret Kearney
12 Coleraine Road, Ballycastle, Co. Antrim
Tel: 026 57 62073

Contact for Scottish Families:-

Alan and Fiona Byrne
3 Jedburgh Ave, Rutherglen, Glasgow G73 3EN
Tel: 041 643 0034