NEWSLETTER mps SPRING 1984

THE SOCIETY FOR MUCOPOLYSACCHARIDE DISEASES

National Registered
Charity No: 287034

CHAIRMAN OF TRUSTEES	Mr. G.C. Nichols C.B.E., D.F.C.
CHAIRMAN OF MANAGEMENT COMMITTEE	Mr. R.J.O. Lavery
HON. SECRETARY	Christine Lavery, 30 Westwood Drive, Little Chalfont, Buckinghamshire. Tel: Little Chalfont 2789
HON. TREASURER	Diana Fudge F.C.A., 22 Kingsley Park, Whitchurch, Hants. RG28 7HA.
HON. FUND RAISING OFFICER	Susan Heath, "Prospect House" 133 White Lion Road, Amersham Common, Buckinghamshire.
NEWSLETTER EDITOR	Marlene Sanderson, "Greenhill", Downend, Horsley, Glos.
AREA SUPPORT FAMILY (WALES) and REPRESENTATIVE	Mike Skidmore, 5 Chapel Close, Wyesham, Monmouth, Gwent.
MEDICAL ADVISORS	Dr. B.L. Neal M.B.B.S.,M.R.C.P.G.,D.Obst.,R.C.O.G. Dr. D. Garrow B.M.,F.R.C.P.
GENERAL MEMBERS	Mr. P. Archard Mrs. S. Butler Catherine Grant S.R.N.,R.S.C.N.,H.V.Cert.

AREA SUPPORT FAMILIES

Northern England
Colin and Mary Gardiner; 35 Church Road, Banks, Southport, Merseyside.

East Central England
Neil and Jane Reid; "Meadowlark", 9 Huddleston Way, Sawston, Cambs.

South East England
Robin and Christine Lavery; 30 Westwood Drive, Little Chalfont, Bucks.

South West England
Peter and Marlene Sanderson; "Greenhill", Downend, Horsley, Glos.

WHAT ARE THE AIMS OF THE SOCIETY?

1. To act as a parent support group.
2. To bring about more public awareness of MPS Disease.
3. To raise funds in order to further research into MPS.

SECRETARY'S REPORT

Since I last wrote for the newsletter, the Society has continued to evolve. The success of the First Parent Conference has resulted in the increase of workload and it has been necessary to divide up the secretary's job. We are most grateful to Pat Skidmore for agreeing to take over the secretarial duties for our oversees MPS families who continue to join the Society at a steady rate. It is thanks to all our families who have been beavering away raising money or letting the public know of our existance resulting in a large number of donations being made that has made it necessary to appoint a Donations Secretary. Sue Butler has kindly agreed to take on this task. In January the Trustees and Committee made some changes:- Geoffrey Nichols is now Chairman of Trustees and Robin Lavery takes over as Chairman of the Management Committee. We also offer a warm welcome to Mike Skidmore who has joined the committee as Area Support Family representative. One of our priorities has been to firm up the role of Area Support Families and to help them become more supportive in their areas. Steps were taken when in early March all the Area Support Families met in Amersham to discuss their roles and learn about future plans for the Society. By now many of you will have heard from your ASF although we still haven't appointed ones for Scotland and Northern Ireland. We are also still studying the area of Central England (West) as sadly we have to say goodbye and thank you to Anne and Robin Ridley as Area Support Family for that Area for personal reasons. Many ideas came out of this meeting including that of families in their areas holding Open House, or arranging outings for the group. In some areas such as Mary and Colin Gardiner's (Northern England) it may be impossible for reason of distance to get all families together but then on the other hand within Yorkshire their are enough MPS families to organise their own gathering and as you will see from Audrey Toker's letter further on in this newsletter, she is willing to organise just such an event. Essex and Hampshire also have enough families to go it alone. It may be that you don't know who the other MPS families are and for that reason we are asking you to complete the form accompanying this newsletter indicating whether you would like to be contacted by other MPS families in your area.

Since Professor Hobbs expressed the need for control studies of the Natural History of MPS Diseases paying particular attention to the regressive mental handicap that occurs in 4 types, our Medical Advisors have given a lot of thought to the subject. As a result we now have someone interested in taking on this study over a three to five year period. Dr. Garrow will cover this in greater depth later on in the newsletter. I only mention this simply because the project will require substantial funding and we would be pleased if as many families as possible might help in this task. We are a little behind schedule with getting our 'Coffee Snowball' packs which if carried to their conclusion could raise £49,000. 17 families have already received their bundle of tickets and we hope to get the packs out at a rate of 10 a week. The value of each families pack is just under £1,000 so do try to encourage your friends to continue the chain. Please do let me know if you are unable to participate.

After the success of the First Parent Conference in Birmingham we have now made a provisional booking for a 2nd Parent Conference 27/28/29th September 1984 at the Crown Hotel, Harrogate, North Yorkshire. This will be a much more leisurely occasion than last year allowing plenty of free time for families to get to know each other and relax, although we do plan a Saturday Programme paying particular attention to the Educational and Sociological aspects of caring for an MPS child. Unlike last year we don't think it likely we will be able to secure the monies required from companies and private businesses so along with the Committee the Area Support Families have each agreed to raise between £1,000 and £1,500 towards the weekend, enlisting everyone's support. If you feel able to help your Area Support Family in this task please do let them know.

We have had a steady trickle of new members and would like to give them a warm welcome. Over the past months I have had the opportunity to visit a number of families for the first time. Geographically it is easier for me to visit families in Southern England. If at any time you would particularly like a personal visit do let your Area Support Families Know.

Christine Lavery

PAMPER A SUNFLOWER

You'll remember perhaps that the Autumn 1983 Newsletter reported that the 1983 Sponsored Sunflower Competition raised £444. Well believe it or not the sum in the end was nearer £1,000. Yes, £1,000.

We hardly tried at all last year; it was a first experiment. So Christine is organising the event again and we hope the necessary information (and seeds) will accompany this Newsletter. I'm going to have a go: well at least Andrew and Ben will do the work and I'll manage. If Simon had been here he would have taken charge - he was always most interested in sunflowers. This is really a project for the children and I hope we can spend the odd 5 minutes from time to time supporting their efforts during the summer months.

The height to beat is 215 cms and for the most money raised by a single sunflower £143.62.

Robin Lavery

LEGAL AND GENERAL CHARITEX '84

Our last Newsletter gave advance notice of this event which had been scheduled for July 1984 at Alexandra Palace. Sadly this event has been cancelled. Firstly because it would have given many of us another opportunity to meet again and secondly because we could have put a good act together since there are members in the Society with professional experience of exhibition organisation and participation.

Let's hope Legal and General might make another attempt at what seemed a very good idea.

Robin Lavery

A PILOT STUDY OF THE NATURAL HISTORY OF MPS

Christine has asked me to write something for this Newsletter about the research project that I have proposed and which as a Society might be able to finance.

I have discussed some of the problems with Dr. Martin Bax whom I hope will supervise the project. He knows an educational psychologist who is interested and would be suitable. The tentative plan is for them to carry out a pilot study for say 6 months to be followed by a 3 year study of a selected group.

I would hope that they could visit a good number of families without excluding any one on grounds of age or severity of illness. The pilot study would clarify the number of possible participants and assemble information available to the local GP., Paediatrician and auxiliary workers whose agreement and co-operation would of course have to be sought. The study is not intended to surplant any assessments or investigations that are underway but to coordinate them. In some cases parents may feel that they have had more than enough of tests and questions and people wanting to help.

Tests of cognitire function in physically handicapped children all have limitations and this pilot study will it is hoped suggest how best this can be measured in the different age groups.

The definitive study will provide on-going background information in a sufficient number of children to enable a proper assessment of the value definative treatments directed to remedying the enzyme disorders of function. Is intellectual deterioration averted or prevented for instance?

The reason for the variation in the clinical picture produced by a single enzyme deficiency is not understood.

An XL iduronidase deficiency may result in Hurler syndrome with mental retardation or the Scheie syndrome without mental retardation. An intermediate Hurler/Scheie syndrome also occurs. An individual with this enzyme defect cannot in infancy be precisely diagnosed.

A Sulphoiduranate Sulphate deficiency may lead to a severe or mild form of Hunters syndrome.

What is the natural history of each and how early can they be clinically differentiated? If treatment is given to examples of either deficiency it is obviously essential to know to which sub group they belong, if its effectiveness is to be accurately assessed. Dr. Young's studies of the mild and severe forms of Hunters syndrome show that there is a clear tendency for older children to be more handicapped than younger but they do not show the progress of individual children followed for a number of years. Does every individual deteriorate at the same rate? Does spontaneous improvement occur?

It is hoped that these and many other questions will be answered. For those for whom treatment is not possible a greater understanding of the human needs of parents and siblings should follow from this longitudinal study.

Dr. Donald Garrow B.M., F.R.C.P.

THE ASSOCIATION FOR RESEARCH INTO RESTRICTED GROWTH

I had already written an article for your newsletter, but it got tossed straight into the wastepaper after I read the report on your first Parent Conference. What can I tell you about the benefits of support groups when you already know them very well. The tremendous enthusiasm and sense of unity generated at the Birmingham Conference very much echoed the atmosphere at our last and perhaps most successful Convention. New bonds and friendships are forged together with a new committment to the organisation we belong to. And it leaves the organisers wondering 'How on earth do we follow that!! It is understandable that no-one can contemplate a repeat Parent Conference in 1984, once every two years we find is quite enough to cope with.

Our Association (ARRG) has been around rather longer than the MPS Society but our philosphy's appear much the same - based firmly upon self-help. The number of affected adults is of course much smaller in the MPS Society, dictated by the particular nature of mucopolysaccharide diseases. Thus ARRG is governed largely by adults of short stature together with parents of affected children. ARRG aims to serve the needs of anyone with a disorder causing restricted growth, the needs of their families and their professional advisors. The majority of our short statured members have a wide assortment of untreatable bone growth disorders collectively called the 'chondrodystrophies', the remainder having a variety of syndromes and chromosome disorders. We disseminate information about growth generally, to lay people and medical professionals and are the representative voice for all short statured people on a national and international level.

Over the years few families with an MPS child have been referred to us, perhaps because the short stature aspect of the disease has been the least of the parent's problems. It was most necessary that your own Society be formed, and commendable that it has made so much progress so soon. Other groups with similar aims are being formed all the time and ARRG likes to maintain close links with them and lend support. However, some conditions are so rare that independent support groups are not feasible, so ARRG will always be their main support, as well as for those with chondrodystrophies.

Since ARRG's inception information resources have been set up and sub-committees formed to meet certain needs. The most notable being the clothing and aids resource and the Medical and Family Affairs sub-committees. The Family Affairs Committee, run by parents of average stature and restricted growth (all of whom have children of restricted growth), organise contacts for new parents and answer queries of a non-medical nature. Together with ARRG's Governing Committee, the Family Affairs Committee organise meetings and courses for parents, their children of restricted growth and siblings of average stature. We also feel that adults of restricted growth have a responsibility towards parents and that they can learn much from one another. These meetings also allow short statured children to meet and make friends with adults and other children similarly affected, helping them come to terms with their future as adults of restricted growth.

One of the rewards of being an adult member of ARRG is to see the transformation of parents as they lose their anxieties, fears and mis-conceptions about restricted growth and come to realise that their child can have a bright future with the prospect of a normal, happy and fulfilled life. The prospect for many MPS children is not so bright, yet I am sure you come to regard those short lives as special and very precious, and not the unremitting gloom you perhaps anticipated. I am sure this is due not only to the larger than life personalities of the children themselves, but in some measure to the work of the MPS Society.

May I wisn the Society every success, and trust that ARRG will always work in the best interests of all the members of both our organisations.

Pam Rutt

JEFFREY SMEIJERS - NEW ZEALAND

Hello, my name is Judith Smeijers. My husband Laurie and I have two children, Leah 13 years and Jeffrey 8¾ years who suffers from Hunter's disease. We live in Dunedin, New Zealand and Jeffrey, we believe, is one of only two Hunter's boys in New Zealand.

For 3½ years we thought we had a normal but active, healthy and intelligent boy. Sure, he had begun to get fairly frequent middle ear infections, but that's not uncommon for quite a few New Zealand children, and we were sure he had a hearing problem, but that we were dealing with,and nor was he toilet trained. Then the warning bells started. Our GP felt Jeffrey should go back to see the paediatrician at Dunedin Hospital for a check-up. His head size was out of proportion to his height and weight. X-rays and other tests followed. Arrested hydrocephalus they thought. But the paediatrician was not entirely happy with this diagnosis and asked us if we would see the geneticist. He was fairly certain of his diagnosis of Hunter's disease as soon as he saw and examined Jeffrey and tried to let us down gently. More tests were required to confirm the verdict. This time the samples and a set of colour photographs of Jeffrey were sent to Adelaide Children's Hospital in Australia - the nearest centre with a specialist interest in Hunter's disease and, therefore, able to carry out the sophisticated biochemical tests needed to confirm the diagnosis.

Since this diagnosis was made five years ago, we have been trying to find out more than the dry, factual, scientific information medical books and journals could tell us. Being such a rare disorder, the medical doctors here have no first-hand experience or knowledge of these children and were able to give us very little specific practical guidance. It was the blind leading the blind. So it was with great excitement and intrepidation that we read Christine's letter, published in the Mencap magazine. Here at last was a chance to actually contact other parents of Hunter's children. Since our initial contact, we have become members of the MPS Society and received all the newsletters. Reading them has been a real bitter-sweet experience, but hopefully we may be a little better prepared for what is yet to come.

Jeffrey no longer lives at home because we finally admitted that coping with him at home was slowly destroying our whole family. For the past 18 months he has lived in one of the New Zealand Intellectually Handicapped Society (IHC) family homes here in Dunedin. It is only one mile away, so we are able to maintain regular contact with him. I also still make, or supply all his clothes and take him to the doctor if and when he needs to go. The home looks just like an ordinary house. There are five other intellectually handicapped "children" living there - all older (15-22 years) and willing and able to help Cathy, the house mother, with the caring and mothering of Jeffrey. Each day during the week from 9 a.m. to 3 p.m. he goes to the IHC "Pre-School and Junior Centre", which he has attended now for 4½ years. The Centre, with its dedicated and caring teachers and volunteer workers, has been a constant source of support through all Jeffrey's ups and downs, Of course, there have been many other people who have given us support and help as well. We feel, however, that the experiences shared through the MPS Newsletter have been just as valuable, and we have asked the New Zealand National IHC magazine to publish a letter telling its readers about the MPS Society and we hope it will be of benefit to other New Zealanders.

Judith and Laurie Smeijers,
123 Hargest Crescent,
Dunedin, New Zealand.

A REPORT ON JEFFREY SMEIJERS FROM THE IHC PRE-SCHOOL/JUNIOR CENTRE IN DUNEDIN, NEW ZEALAND

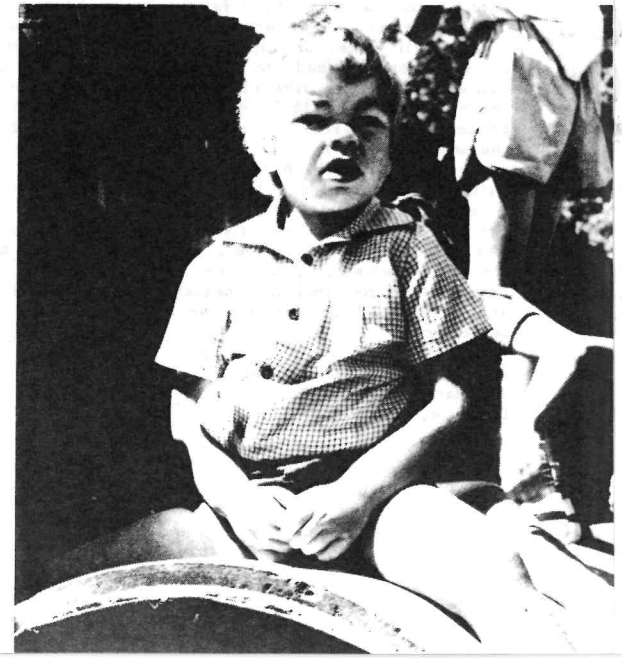
We first met Jeffrey when he was transferred from The Crippled Children's Kindergarten in August 1979. He was four years old. At this time he had more normal behaviour than abnormal. During 1980/81 he attended a normal playcentre, had a trial period of three months at a Physically Disabled School, and when that failed, attended an infant class in a normal school for 1 hour a day. Because of deteriorating behaviour this programme ceased in September 1981 and Jeffrey spent the next five months in a Psychopeadic Unit within a psychiatric hospital.

Jeffrey returned to the Intellectually handicapped Pre School/Junior Centre (P.S.J.C.) on 10th February 1982, where the staff noticed a marked deterioration in his behaviour. Jeffrey's assessments showed that functioning in all areas had regressed. During this period Jeffrey was maintained in the centre on a one-to-one basis. His behaviour included aggressive and destructive acts and he would not comply with instruction. It was very difficult for those who worked with Jeffrey to see the language return to echolic speech, the concentration span decrease to nil, the destruction and aggression that had been self-controlled, once more rise to the surface. This was Jeffrey for 1982/83.

Now in 1984, Jeffrey is a very different child, passive, with eating problems, chest infections, a marked change in his physical appearance, the use of a wheelchair for going small distances as walking has now become a painful experience. The frequent bowel motions have decreased in size and number, from up to six a day to less than 1 a day.

Throughout the five years we have known this lad he has added more to our thinking and has given more pleasure with his humour and cuddles than many of the children who pass through our centre. Since we ceased having specific programmes and goals for Jeffrey he has been a more contented boy, who enjoys especially the close contact of an adult reading stories to him. Our rewards are smiles and cuddles. He has found a special place in all our hearts, staff and volunteer workers alike.

Coral Davies
Supervisor, P.S.J.C., Dunedin.



Jeffrey Smeijers, aged 8 years old.

SOUTHERN AFRICAN FAMILIES

At last I've got round to sending you the article on our South African MPS kids together with a map to show how scattered we are. I have not met them all personally as the distances are too great to travel so some of this is information from Angela Kemp.

I have numbered them from 1 to 10 in descending order of age.

1. Steven Edkins - age 11 - Sanfilippo. SPRINGS
Steven is in a home for handicapped children as his mother found it very difficult to cope, but he goes home for weekends and holidays. He is quite severely affected. His parents lost a daughter aged 18 months before he was born in a car accident, so it is not known if she was affected or not and he has a younger sister born before he was diagnosed who is luckily fine.

2. Shaun Thomas - age 11 - Hurler. WESTVILLE, nr. Durban
Shaun is my younger son. Brett, my elder is fine and I've never attempted to have more children. Shaun is very healthy (I can't recall his last infection, but he no longer walks. He's never spoken but he's a cheerful content little boy and very easy to manage.

3. A boy aged about 8 or 9 - Hurlers. VEREENIGING
We have never met this child. We were told by the Genetic team in Johannesburg that he was diagnosed about 1 or 2 years after Shaun and has been deserted by both parents and is in the care of an aged and poverty stricken grandmother.

4. Muhammed Timol - age 8 - Hunters. DURBAN

Muhammed and his younger brother Ibrahim were diagnosed as Hunters. Ibrahim died after BMT a year ago. Muhammed is fairly well - very active still and attends special school. He's a friendly noisy little fellow and speaks well. He is deteriorating now but not severely.

5. Patrick Piron - age 6 - Hurler. PORT ELIZABETH
Patrick has always been a cheerful independent little character but during the last year has deteriorated quite a lot. He suffers terribly with abdominal pain. He has a younger brother and sister born after amniocentesis.

6. Simon Kemp - age 5 - Hurler. PORT ELIZABETH
Simon is a frail sickly little boy who is plagued with infections, but is far the "brightest" of our South African MPS kids. He speaks very well, does all sorts of puzzles and can identify a few written words. He has terrible problems sleeping. He is an only child.

7. Diane Bam - age 6 - Hurler. PORT ELIZABETH
Diane is a surviving twin. Her sister Jane died aged 22 months. Diane does not walk or talk and has not enjoyed very good health over the last year. Her hearing (and Shaun's) appear to have been impaired at a much earlier age than the other children.

8. Candice - age about 3 - Hurler. PORT ELIZABETH
WE have not met the parents and she was only diagnosed recently.

9. Johan Duvenhage - age 3 - Hurler. ARANOS
Johan lives on a farm in the arid area of South West Africa/Namibia. He appears to be the most healthy easy going child of all. The climate and the healthy farm life style probably account largely for Johan's good health and cheerful disposition. He walks, talks and plays happily outdoors. He has three elder siblings - all fine.

10. A little boy aged 2 - MPS type as yet unknown. AMALIA
This little boy does not appear to be severely affected and is almost certainly not a Hurler. He was only recently diagnosed and we have not yet heard the results from the parents. He has an older brother and younger sister.

We have heard vague rumours of a Hunter child in Pietermaritzburg and one in Port Shepstone and a Morquio in Durban but have been unable to trace them. I, personally, met a family from Durban whose Hurler son died about 8 years ago and a family from Johannesburg whose Hurler daughter died 6 years ago and I heard of a family in Cape Town who lost a Hurler daughter and after 2 unsuccessful attempts, finally had a normal daughter.

Our worst problem in South Africa is the vast distances by which we are separated. For example it is 960 kilometres (+- 600 miles) from Durban to Port Elizabeth which is a 12 hour drive - 3 hours more to Knysna - not something to undertake lightly with a Hurler child. It is 640 kilometres (400 miles) from Durban to Springs - more to Amalia - and somewhat over 2000 kilometres to Aranos. Airfares in South Africa are expensive and so are long distance phone calls. When one is feeling low and in need of support or advice one must think twice of the cost involved before just 'phoning for a chat.

Another of our problems is lack of interest and therefore aid for MPS. Because we are so few apathy seems to start at local doctor level and continue through to State Health Dept. We all have the same story to tell of battling to get a diagnosis following months of frustration and despair.

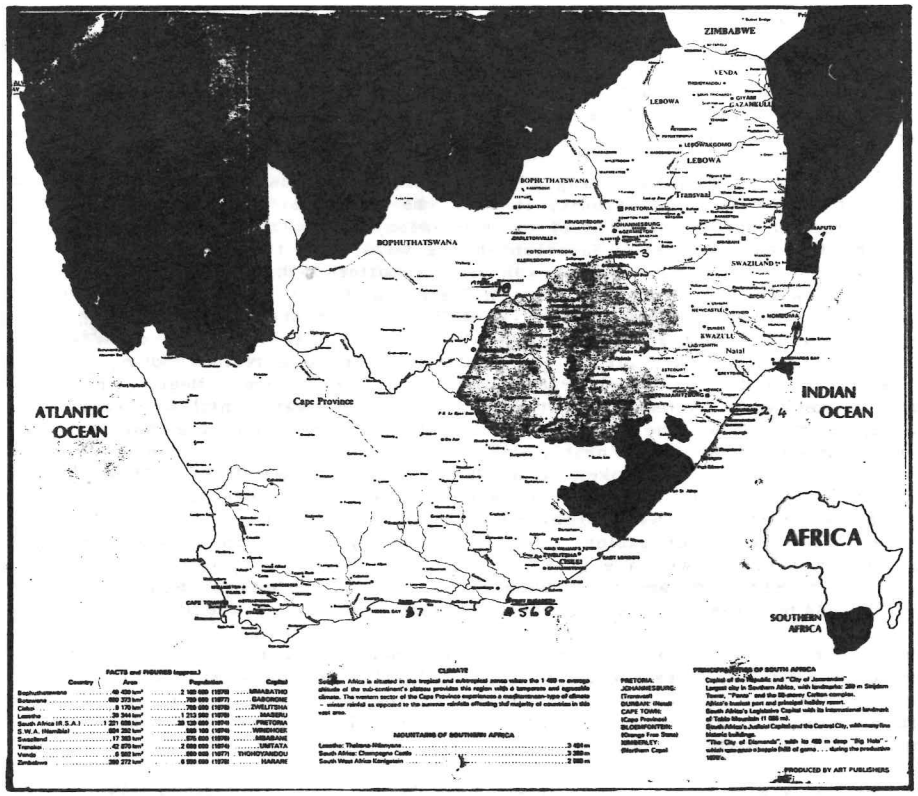
Medical care in South Africa is fairly expensive. Most of us belong to Medical Aid Societies. A compulsory deduction is made from one's salary and paid to these Societies who then pay a proportion of all medical expenses. They do however have a limit of what one can draw in any one year and with an MPS child it is very easy to exceed that limit.

State aid in the form of a grant of R132 (+-£61) is available only to indigent families and by that they really mean poverty stricken. Provincial hospitals provide free or reduced medical treatment but again only for indigent cases.

Homes and institutions are scarce in South Africa and of a questionable standard in most cases. There are a few excellent private homes but they charge exorbitant fees. Special education is only just beginning to receive attention and I can't think of one school in South Africa geared to our children's needs. Shaun attended a small private nursery school for a few years where he was very happy but their fees have also rocketed.

I am exceptionally lucky in having the help and devoted support of the first hospice team in South Africa, which I find of inestimable value but unfortunately the other mums are not so lucky.

However I must not make it all sound ghastly. On the credit side we have a wonderful climate, most conducive to rearing healthy MPS children with only the Cape Province experiencing wet winters. In Natal where I live it is often popssible to swim in winter and Shaun has spent many happy hours in swimming pools. We also have a lovely range of fresh vegetables and fruit available all year round and if one can persuade



these little monkeys to eat there is no reason why they can't enjoy a varied and balanced diet. Shaun certainly does, and food is cheap in comparison to Britain.

Holidays are a bit of a problem but we have solved that by staying at resorts where chalets are available which is very like home from home. Disposable nappies and bottled baby food are a help too.

I hope this will give you all some idea of MPS - South African style. If anyone is interested in contacting me, I'd be only too happy to write to them.

Love
Gail Thomas
12 Medway Rd., Westville, 3630 South Africa

AN UNUSUAL MEETING

Since joining the MPS Society, Christine Lavery has given me the address of a young lady named Pam who suffers from the same disease as myself and my sister Linda, which is Maroteaux-Lamy. Pam and I have corresponded regularly ever since.

When I heard that Pam was being admitted into Moorfields eye hospital I thought what a good opportunity it would be for me to go and meet her. Although not in the best of circumstances, it was much nearer to me than her home address. So arrangements were made for the day I would go and visit Pam. Soon it was only a matter of hours to go when my train would be pulling into Holborn Station and that meant a matter of minutes to when I would actually meet Pam. My heart was beating pretty rapid as I approached her room, but I was greeted by her mother as Pam had been whisked off by the doctor to have some tests. So my husband Paul and I waited for Pam to get back to her room. Meanwhile Pam's mother made us feel very at ease and chatted away, until we heard footsteps coming down the corridor and a head popped around the door. It was Pam and I think she nearly dropped as she thought it was herself sitting in her room. We looked so much alike it was incredible. She is a lovely person and we got on so well. To actually speak to somebody who really understands what problems I suffer was marvellous. Although for me things have been easier, because I have my sister Linda, who has the same disease, so we have grown up together sharing our problems. But for Pam, she has been very much alone with her problems. However poor Pam had lost her voice and found it very difficult to speak.

Anyway our afternoon was wonderful and had many photographs taken together. Even the nurses came in turn to Pam's room to have a look at us both together, we should have had one of the MPS collection boxes with us and made all the nurses who came pay. I think we would have raised quite a lot of money.

Our meeting was over and our journey home had to begin but its a day I will always remember.

Carol Hubbard.

STAMP NEWS

Hi there,

I thought you would all like to know that the stamps are coming on well, so far to date they have raised over £100, so do keep them flowing.

I would also like to add that I cannot take all the credit for all the money that has been raised from these stamps. I owe quite a lot to Christine Lavery, whom has also been Involved with the stamps.

Bye for now
Carol Hubbard.

A MOST WELCOME OFFER OF HELP FROM WEST YORKSHIRE

On February 14th my daughter Alisons first baby Jonathan (and our first grandchild) died in our local General Hospital, Special Care Baby Unit, aged 3 months, with a diagnosis of Mixed Sulphate deficiency.

Jonathan was born to Alison and Gerald on November 3rd, by Ceasarian Section. He had unusual signs and symptoms and had the Medical and Nursing staff perplexed and concerned over his condition. He was transferred to Leeds General Infirmary at six days old, and five weeks later, following the intensive tests and investigations, was found to have one of the mucopolysaccharide diseases. We experienced (as must all of you have done) the awful pain, anguish and despair associated with these tragic diseases, and although Alison and Gerald were told the diagnosis in a kind, gentle and sensitive manner, they were informed that the prognosis was very poor.

Jonathan was returned to our local District Hospital, Special Care Baby Unit one week later. He received so very much love and care in his little lifetime, and became the favourite of all the nurses in the Ward. He was such a sweet and lovely little boy.

Alison and Gerald never had Jonathan home, but the love we have all received through him is unbelievable.

We have become so aware of the need for support for parents, families and children with these diseases and will always be so grateful to the Staff at Leeds for sending us the leaflet on the MPS. From the moment we knew of its existence we felt some of our load to be lifted and the ignorance, bewilderment, anger and frustration to be understood and acceptable.

Because we only had Jonathan for such a short while, we would dearly love to meet other families in this area and if possible to get to know the children too.

I am taking the opportunity through the Newsletter, to ask if we may get in touch with families, especially from West Yorkshire and if at the same time we can offer any practical help then we would be so very pleased to do so.

We hope to have a list of families from West Yorkshire sent to us by Christine so if there is a letter in the post from a stranger, then this will be from "us".

We do hope that this will be possible and would love to hear from you all.

Yours sincerely
Audrey Toker.

STEVEN WHITTINGTON

Hello, we are Jean and Derek and we have three children, Terri aged 5½ years, Steven aged 3½ years and Garry 2 years.

Our problems began when Steven was born and we were told he was Achondroplastic. We found out that this meant he was a dwarf. The first two years of his life he was in and out of hospital with chest infections. The last time he was in hospital (just before his second birthday) the doctors seemed to think his symptoms suggested something else. Tests confirmed that Steven suffered from Hurler syndrome. What shattering news when we finally found out what it meant.

As we looked for signs of things starting to go wrong with Steven, we noticed that he seemed different to other children we had seen and read about in the MPS Newsletters. We questioned Steven's doctor about this but he was adamant that Steven had Hurler syndrome. He actually told us that we could take his tests to Timbucktoo but they would come back the same. That is that Steven had Hurler syndrome.

The family conference was soon to be with us and we looked forward to meeting and seeing other MPS families. Whilst we were there we compared Steven with other Hurler sufferers and we noticed that he was very different. Most people who saw Steven were of the same opinion including Dr. Wraith. They, like us, thought that Steven may not have Hurlers. We left the family weekend puzzled and decided to tackle Stevens doctor again. But what with eye tests and Christmas coming, we thought we'd leave it till the new year. He was also fitted with a hearing aid just before Christmas. However, in early January whilst talking with Christine, we discussed the possibility of having Steven re-diagnosed, because so many people at the conference had commented on how different he was. Christine thought that as I was so worried, I might go back to my own doctor and suggest that samples of Steven's urine should go for tests to The Institute of Child Health, Enzyme Lab. So we sent off the urine sample and waited with baited breath. Eight days later the results came through. Test results suggested that Steven did not have Hurler syndrome. An appointment was made to see Dr. Rosemary Stephens at the Hospital for Sick Children. On the 2nd February we went to see Dr. Stephens who confirmed that Steven did not have Hurler syndrome or any other type of MPS disorder. After many hours and many doctors, we were told that he has Kniest disease. After 3½ years of not really being sure of his sickness we are finally relieved, or are we.

We now find out that in the past 3½ years, we could easily have paralized or even killed Steven, just by giving his head a sudden jolt. This is because the top two vertebrae of his spine are not connected and could easily come apart. It sounds silly, but even pushing his head forward to trim his hair could have done untold damage.

The most apparent feature is his size, he's only 2ft long. What makes him look even smaller is the severe curvature of the spine, which the doctors are most worried about. It could curve even more and collapse in. The nerves could also become trapped.

Deafness, ear infections and short-sightedness are all connected to this condition, but it is the bone structure that is affected most by Kniest Disease. If Steven overcomes the spinal complications, he could live to be an adult.

Kniest unlike Hurlers is not heredetry, Steven being a fresh mutation. We are told that the genes mutated at the time of conception and that it is very unlikely it would happen to us again.

We now feel very bitter towards certain parts of the medical profession because over 12 months ago a doctor in Oswestry told us that she believed Steven had Kniest but it was never followed up. Other doctors who will remain nameless, told us that the tests done for Hurlers were positive.

Thank you Christine and the Society for helping us discover the truth.

Jean Whittington.

CHRISTMAS CARD COMPETITION

Ask your children to design a Christmas Card for MPS and send it to:

Mary Gardiner,
35 Church Road, Banks, Southport, Merseyside
by the 30th May

Mr. Bill Tidy the North Country illustrator has agreed to do the judging and there will be a prize for the winner.

BEREAVEMENTS

It is with great sadness that we learned of the death of the following children:

Paul Ryan, aged 3½ years old, who died on the 7th April following bone marrow transplant. Paul had been suffering from Hunter disease.

Jonathan Dore, aged 3 months old who died in The Special Care Baby Unit at Keighley General Hospital.

David McAleese aged 2 years old after bone marrow transplant at the Westminster Street Hospital.

Our deepest sympathies go out to the parents of all these children.

NEWS IN BRIEF

We would like to send our congratulations to Jean Silvey on her recent marriage to Mr. Richard Hopwood. Although Jean has one son Glen, she has acquired a new son and daughter. The family will live together on Canvey Island and we wish them the best of luck for their future together.

A donation of £1,000 was made by the Society to Prof. Adinolfi, M.D., Ph.D., Professor of Developmental Immunology, Paediatric Research Unit, Guy's Hospital.

Prof. Adinolfi and his team are researching into the causes of Hunter Disease.

Prof. Tadao Orii, of Gifu University School of Medicine is arriving in England on 3rd May for a stay of 1 month In order to study various aspects of MPS. He is particularly interested in the Society's support activities and have invited Prof. Orii to address some of our families situated nearer to London at a meeting the date yet to be agreed.

SAVE 'GOLDEN WONDER' CRISP PACKETS

'Golden Wonder' crisps have £1million giveaway on at the moment. This means that if you can save your packets, they will give 1p for each. Get your local school or pub and friends to help and see how much we can raise for MPS.

WANTED 'MONEY OFF' COUPONS

Please can everyone send 'Money Off' Coupons for groceries or cigarettes to Pat Kirkman, 73 Richmond Terrace, Darwen, Blackburn, Lancs. Pat will collect them all together and redeem them with the appropriate companies giving the money to MPS.

So come on turn out the kitchen drawers for those 5p off Persil or 3p Heinz Beans etc and help MPS children.

MPS CHRISTMAS CARDS DESIGNS

Despite the small number of children who sent Mary their ideas for the Society's 1984 Christmas Card we do hope that a choice can be made and that we will be able to have our own Christmas Cards on sale in late September.

PLEASE CAN ANYONE HELP?

Mrs. Wright from New Zealand would very much like to hear from a family who have or have had a child suffering from I-Cell Disease.

Mrs. Johnson from Chester would very much like to hear from anyone who has come across a child with Sanfilippo Disease complicated by Hereditary Diabetes.

Please refer any suggestions to these requests through Christine Lavery.

NEW FAMILIES

Mr. & Mrs. Kraft from Austria, whose 4yr old daughter Barbara is suffering from Hurler disease.

Mr. & Mrs. Hall whose son Duncan aged 9 yrs is suffering from Sanfilippo disease. They live in Leicestershire.

Mr. & Mrs. Sullivan from Co. Cork, Eire. Their son Kieron is believed to be suffering from MPS.

Gerald and Alison Dore after learning that their newborn baby Jonathon was suffering from Complete Sulfatase Deficiency disease. Sadly Jonathon died on St. Valentines Day aged just 3 months. They live in West Yorkshire.

Lorraine Wright wrote to us from Manurewa, New Zealand after learning that her 3 year old daughter Frances-Lee is suffering from I-Cell Disease.

Maitland Morgan whose only son, David aged 8 years is suffering from Hunter Disease. They live in Surrey.

Mr. & Mrs. Johnson from Chester. Their daughter Victoria is 6 yrs old and suffers from Sanfilippo disease. Victoria also has Diabetes and is a resident at Heswell, the country branch of the Royal Liverpool Children's Hospital.

Mr. & Mrs. Bielby from Yorkshire. Their eldest son James is believed to be suffering from MPS.

Mr. & Mrs. Devereaux whose son Bobby aged 14 years is suffering from Hunters disease. They live in Somerset.

Mr. & Mrs. Kearney from Co. Antrim, Northern Ireland. Their son Iadin aged 6 years is suffering from very mild Morquio disease.

Mr. & Mrs. Lowry from Hertfordshire. Their younger daughter Sarah aged 9½ years is suffering from Maroteaux-Lamy disease.

Mr. & Mrs. Hayward from Gwent. Sadly their daughter Angeline who suffered from Hurler disease died last year aged 10½ years. In memory of Angeline her parents have kindly offered to help raise funds for MPS.

Mr. & Mrs. Long from Coxwold, York, whose daughter Sarah suffers from Morquio and is aged 13 years old.

Mr. & Mrs. Hunt from Banbury, Oxford, whose son Nicholas suffers from Hunter disease and is aged 5 years old.

Mr. & Mrs. Shorthouse from Coventry, whose son Christopher, aged 2 years suffers from Hurler disease.

We now have parent contact for the following types of Mucopolysaccharide diseases

Hurler
Hurler/Scheie
Scheie
Hunter - Severe
Hunter - Mild
Sanfilippo 'A'
Sanfilippo 'B'
Sanfilippo 'C'
Sanfilippo 'D'
Morquio
Morquio - Very mild type
Maroteaux - Lamy Severe
Maroteaux - Lamy Mild
Gangiliosidosis
I-Cell disease
Complete Sulphate Deficiency disease

We wish to thank the following companies and people who have so kindly made donations to the Society.

The donations will go into the General Research and Holiday Homes Funds.

Hiro Tsungyoshi, British Embassy, Japan.
S.C. and A.J. Hodgkinson
Sandra Stone and Colleagues
D.M. Oldakes
The Perolin Company had a no smoking day.
Mandy O'Sullivan had a win in a pool competition.
C.A. and M.O.Toole
Mr. & Mrs. Warburton had a collection box in Cyncoed Post Office.
Medlock Manufacturing Sports Committee
Bequests and Donation from Richard Gardners Fund.
Alison James

The following strong willed people had a Sponsored Slim.
Ian and Catriona Ogilvie
R. Eden
Anne Ogilvie
A.D. McDougall all from Scotland.

Christine Lavery and Marlene Sanderson both held Coffee Mornings.
M.E. Davidson had a Book Sale.
Carol Hubbard, Linda and their mum have been collecting Tin Foil and Stamps.
Jean Bailey of Cheadle Heath
Jenny & Roger Broome and family who had a Spring Fayre.
Audrey Toker and Family who held a Bazaar.
Reynolds Boughton Ltd
Inverurie and District Ladies Circle held a Social Evening and Sheena McCarthy was invited along to accept the cheque.
Sue Bramford and Marlene Sanderson held a Charity Stall at Swindon Plaza.
Pat Hewitt and Friends who had a Clothes Sale.
Mrs. E. Sanderson
Tetbury Post Office - Donation Box.
F. & W. Data Systems
Cow and Gate
American Express
Uttlesford D.C. Staff and Social Fund
Mrs. Jean Cowles
M. F. Garde
Cadbury's Schweppes
National Westminster
Nationwide Building Society
Chorley Wood Women's Fellowship
Potteries Marathon
Louise, Nicola, Helen, Zoe, Laura and Tracey, all aged 13 years held a bazaar outside their home and raised £28.00 for MPS. They come from Pentwyn, Cardiff.

Donations in memory of Matthew Goldsworthy who tragically died in January . Matthew was a good friend of Matthew Hodges.

H.J. and J.F. Goldworthy
K.G. and J.M. Bowsor
Harby Village Institute
J.M. Downes
P.M. & M.A. Daynes
Freda Savige
Guildford Knapwood

Donations made in memory of Jamie Stenson.

R.J. and C.A. Lavery
Mrs. D. Kris
Welwyn Garden City TRades and Labour Club held a Disco.
Hallett Silberman Ltd.

The following donations were made in Liew of Flowers in memory of Jonathon Dore who sadly died in February aged 3 months.

A.M. Carlisle
D.A. Bartle
E. Hartley
G. Sooley and C.A. Sooley
N.J. and J. Hutchinson
Jennifer and Alan Hudson
Robert Preston
H. Shaw and Mrs. I. Shaw
G.S. and S.A. Newriss
E. Midgley
A. Toker
Mrs. I. Wilkinson
Alice Webster
J. Fort
Mr. & Mrs. Driver

Family Weekend Sponsors
Weetabix
Cardiff Lions
Cardiff Police Social Club

We still need many more sponsors and donations for the second family conference to be held at the Crown Hotel, in September, so put on your thinking caps for ideas for your Fund-Raising and tell your area family you could come up with something really good.

Donation Secretary
Sue Butler.