NEWSLETTER SUMMER 1990

THE SOCIETY FOR MUCOPOLYSACCHARIDE DISEASES

National Registered Charity No. 287034

The Society for Mucopolysaccharide Diseases

30 Westwood Drive, Little Chalfont, Buckinghamshire.
Telephone: (024 04) 2789

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

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"

****

The address of the Society has changed from Westwood Drive to:

Christine Lavery
7 Chessfield Park,
Little Chalfont,
Amersham,
Bucks HP6 6RU

The telephone number remains the same. We will continue to use the old stationery with the Westwood Drive letterhead while we have it in stock. Letters sent to Westwood Drive will be forwarded by the post office.

Pauline Mahon has very bravely taken over as Treasurer from **Stella Hale** who has resigned for family reasons. Our very warm thanks to Stella for her very efficient service to the Society over a long period. She has done an extremely professional job at a time when the finances of the society expanded enormously. Best wishes to Pauline in this key role. (see note about paying in books).

****

BIRTHS

****

Congratulations to **Caroline and Robert Fisher** of Saffron Walden on the safe arrival of **Francesca Louise**, a sister for **James.**

Congratulations to **Karen and Wayne Hoather** on the birth of **Katie Suzanne** on the 11th of June, a sister for **Simon** and **Michael.**

****

NEW FAMILIES

Mr and Mrs Ward, from London, whose daughter **Linda** aged three and son **Michael** aged one have both been diagnosed as suffering with Hurler disease.

Nora and Tom Corcoran whose son **William** aged one and daughter **Marian** aged nine months have both been diagnosed as suffering from Hurler disease.

John and Delores Byrom from Sale in Cheshire whose daughter **Rebecca** born in December 1986 has been diagnosed as suffering from Sanfilippo disease.

****

DEATHS

Our thoughts are with **Mr and Mrs Eileen Taylor** who on the 5th of June 1990 sadly lost their much loved son **Liam** who suffered from Hurler disease.

Paul Evans from Ashford in Kent who died on the 8th of June, aged almost fourteen. Paul suffered from Maroteaux-Lamy disease. Our thoughts are with his parents **George and Jill** who not long ago suffered the loss of Paul's sister **Cheryl.**

Jemma Corbett who died on the 22nd of June 1990 aged seven. Many of you will know of Jemma through her diary in the newsletter. Our thoughts are with **Sarah, Mike and Elizabeth,** her grandparents and all her family.

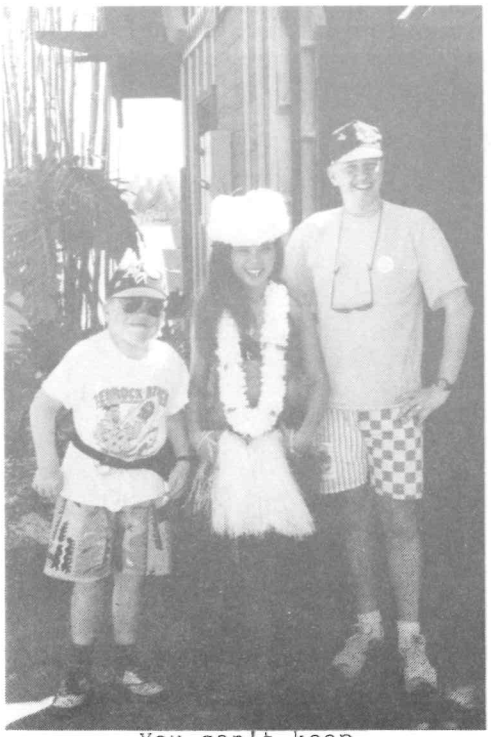
James Piromalli who died on the 18th of July aged eight. James suffered from Hunter disease. Our thoughts are with his sorrowing parents and family.

****

DATES FOR YOUR DIARY

31st August	Conference UMIST Manchester.
7th October	Deadline for Autumn Newsletter
1st December	Christmas Party - South Wales

Outing to Alton Towers - Annual Conference 1989



You can't keep him out! Alex and friends in Florida.

A pensive moment at the conference tea party 1989.

****

POLISH MPS SOCIETY

****

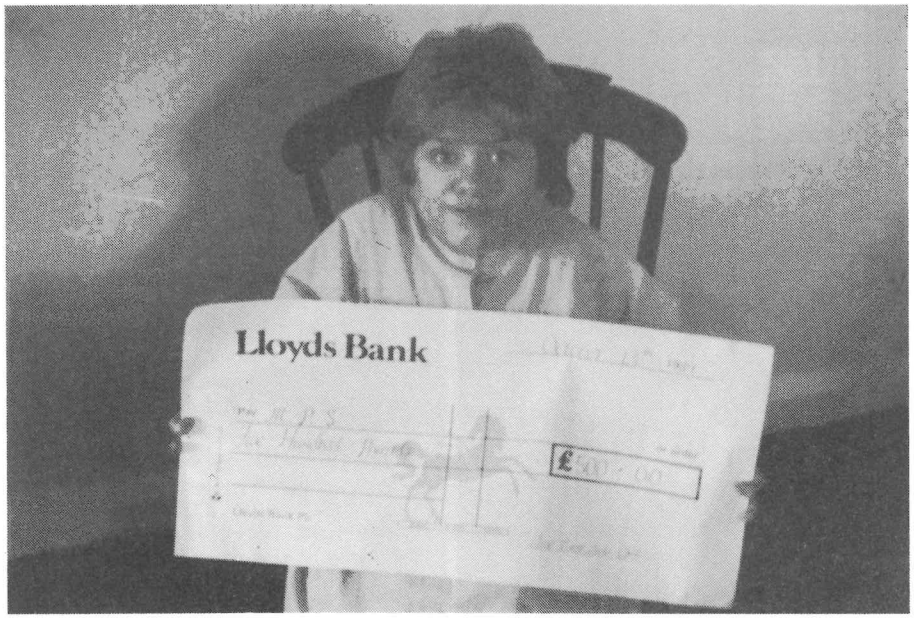
Readers will be delighted to hear that an MPS Society has been established in Poland by Joanna and Marek Popek. Mr and Mrs Popek will be attending the UMIST conference with their daughters Kamila,aged eight, who suffers from Sanfilippo Disease, and Magdalena who is six. Christine Lavery has overcome enormous obstacles to obtain funding for them and to make travel arrangements. Travel to the west is now possible, but not necessarily easy!

If you have Polish friends or acquaintances please let them know of the needs of the Polish MPS Society.
(Further information about their needs from Christine Lavery)

The address of the Society is:

c/o **Joanna and Marek Popek**
Os Kochanowskiego 41 IV 13
43-190 MIKOLOW
Poland

****



Sarah Kilvert age seventeen, from Newtown, Powys, receiving a cheque for £500 on behalf of the Society from the Motor Cycle Club.

****

SOUTH WALES FAMILY DAY

****

Duffryn House and Gardens, said to be the finest in Wales, was the venue for our first meeting of the year on 6th of May. Almost every Welsh family was there and we were joined by some of our 'cousins' from across the water. All in all there were about forty five of us - our family in this part of the world is certainly growing!

The weather was glorious - Remember the summer we had in May? After the buffet lunch laid on for us by the staff of Duffryn house we were able to stroll around the magnificient grounds. It was their May family day and there was a pet show, Punch and Judy, a Hawking display, a butterfly house and a palm house which the children enjoyed, as well as a display of motorbikes through the ages which I think the dads enjoyed more than the children.

We all met back at the house to finish off the food and catch up with all the news. We had a room set aside for us in case it rained so we were able to sit and linger while the kids ran about and generally created mayhem - not possible in the public tea rooms. Our thanks to the staff of Duffryn House for making us feel so welcome - whatever we asked of them they were able to provide. Nothing was too much trouble for them and as I said we felt "special" rather than "different". Here's looking forward to the next get together! Welsh families get your thinking caps on for some suggestions as Tony and I are running out of ideas. Mind you the weather will have to improve a great deal otherwise we will be stuck in the pub all day, which would be absolutely unbearable -- or would it? Now there is a thought for next time.!

****

CHRISTMAS PARTY Ist Dec 1990

****

The powers that be (ie, the Committee) in their wisdom have decided to hold the MPS Christmas party in South Wales this year. The date is December the first and the venue is the Perkin Elmer Staff Social Club. The club is located next to the Royal Mint at Llantrisant (otherwise known as the hole with the Mint in it). Sorry, we have already asked for some samples but nothing doing! It is not far off junction 34 on the M4 so is easily reached by even the worst navigator. If you prefer someone else to navigate it is planned to send a coach from the North West and another from Milton Keynes and the London area. Full details in the next newsletter.

So note the date in your diaries and 'we'll keep a welcome in the hillsides' for you until then.

Mary and Tony Lockyer
Welsh Area Family

****

MOVING DAY

****

On Saturday 16th of June a group of us assembled at 30 Westwood Drive. It was the day of the moving of the office. Some grumbled. "Why was it necessary to have ten people come from all over the country to move some papers?" But we quickly shut up and got down to work under the firm direction of Christine and Mary Gardiner. First of all we moved half a ton of papers out of the shed. Then we sorted it into four lots, one for the temporary office at Bryn Neal's house, one to go to Manchester for UMIST, one lot for storage and one lot, for some reason that I couldn't grasp, to go back into the shed. Dan Butler turned up with a van almost as big as the house (his little van which is only half as big as a house had been a bit temperamental) and deftly reversed to the kitchen doorway with millimetres to spare. So we put things on the van and we took things off the van and put things in and out of the shed and eventually it was done.

Robin went away with a hundredweight of plastic spaghetti and came back to announce that the computer was safely up and running with hard discs and floppies doing whatever discs do. By this time we were feeling pretty floppy ourselves so we adjourned for a photo opportunity and coffee. We left the Laverys with an enormous amount of work to do. Some of us returned to our grotts and caves while the hungry ones followed the smoke signals across the valleys to where the ubiquitous Dan Butler was already starting his barbeque.

****

MOVING FORCE!

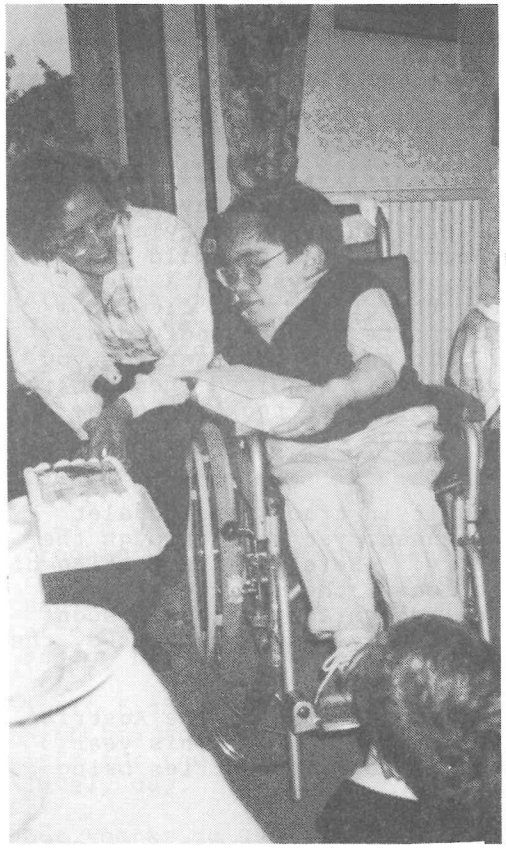
Charles O'Toole, Dan Butler, Christine and Robin Lavery, Peter Robins, Jenny Broome, Ann Neal, Mary Gardiner, Wilma Robins, Bryn Neal, with Dan's little van.

****

BUTLER BARBEQUE 16th June

Laverys, Snacks, Culleys, Neals, Gardiners, Rocks, Isaacs and O'Tooles were treated to a most wonderful afternoon at the Butler household. The food was mouthwatering and lavish, the ambiance and the company was agreeable. The entertainment was provided by Dr Bryn Neal trying to outdo his sons at football in the little paddock. After the barbeque we had strawberries and cream and after the cream we had cake and after that.....It was Lorraine Rock's birthday so we had more cake and sang birthday songs.

Our thanks to Sue and Dan for a wonderful afternoon.



Alex Butler presenting Malcolm Haddow (Organiser) with a thank you gift on on behalf of holiday makers and helpers after the Florida trip.

Lorraine Rock from Hinkley, Leics, with her mother Helen and her brother Christopher on her sixteenth birthday.

Robert Culley enjoying the air at the Austrian Conference at Pertisau.

****

EURO-MPS

****

It all started with a phone call from Christine Lavery. "Would you like to represent the British MPS Society at this year's Austrian conference?" Well, our ten year old son Robert, a Hunter disease sufferer, has always been a great traveller so we decided to accept the offer. We flew from Heathrow to Munich and at both airports we were individually escorted to and from the plane. To ensure this service you must state that wheelchair assistance is required when booking your ticket. From Munich a two hour drive took us to the conference venue, the beautiful village of Pertisau in the Austrian Tyrol.

The hotel, facing the lake and built in traditional chalet style was so unlike the British venues, yet once through the doors we were in familiar territory. Here was the meeting of old friends and the making of new ones, the efficient organisation, and the surprise of recognising children soon followed by the realization that you have been mistaken by the 'family likeness' of MPS children.

That Friday evening was a real United Nations. The Austrian Society plays host to Europe every other year. This year fifty five families attended, most European countries being represented.

Saturday morning, and the view from our bedroom was a bright sun shining on snow capped mountains. The Post House Hotel was never like this! After breakfast the children were handed over to the volunteer carers, mainly young students, and taken to the village sports hall. A day of fun and games for them and lectures for the adults. I must confess that I skipped the talks, my tourist German not being up to that and spent some of the day walking round the village.

We all came together for the children's tea, with much good humour and table cloth pulling. The evening meal and dance was a chance for the parents to relax, exchange ideas and discuss experiences. We were surprised at the different levels of care available across Europe. Certain countries that we perceive as advanced were inferior to Britain in this respect.

Sunday morning was tinged with sadness as our new found friends began to leave for their homes. We were staying on another day, a time to reflect on how our lives are affected by MPS, yet somehow enriched by the comradeship of the common bond. We left the conference, as indeed all MPS gatherings a little stronger and better able to cope. Many thanks to all the societies and helpers that make these events possible. We owe them all a great debt.

Andrew and Vivienne Culley, with little Robert.

****

NORTH WEST NEWS

****

The North West Area held its annual barbeque on Sunday 17th of June at Gilfords Dairy, Willaston.

After two weeks of unsettled weather Sunday turned out to be a glorious day. Twelve families arrived and over eighty people were catered for. With four barbies on the go and a handful of helpers there was plenty of good food for everyone. The children enjoyed rides on Marigold the donkey, who must have thought it was her birthday with the number of carrot consumed. The kids had a bouncing time in the "Bouncy Castle" - even some of the mums had a bounce!

A warm welcome was extended to a new family, Mike and Pamela Croghan with their son Daniel, who came along with their relatives. They even brought their own entertainment! Daniel's uncle did a juggling act towards the end of the afternoon and was really very good - I think we will have to engage him for future MPS events!

Apart from Dominic Hall trying to paddle in a bucket of water with his shoes and socks on, we had no mishaps. Everybody left for their long journeys home after having enjoyed a lovely day.

Our thanks to June and David Gilford (Matthew's aunt and uncle) for allowing us to hold this annual event in their garden.

Thanks to David's dad for scraping the new potatoes - all twenty pounds of them! Next year we will leave the skins on.

Bill and Sylvia Blackburn

****

DAVID CRIDDLE

Living with a shunt and a tracheostomy.

****

My son David is aged twelve and suffers from Hurler Disease. About September to October last year he started having headaches. He complained of "baddy head" and asked me to "pick it off", pointing to his head. Later he started banging his head, also he became bad tempered, which is really out of character for him.

My usual GP was away at the time so I saw another doctor who didn't know much about him. He told me to find a paracetamol that suited him and that he could take ten 5mls spoonfuls per day. As you can imagine I wasn't very happy about this so I rang Dr Ed Wraith.

One week later David and I went to Manchester by train. We stayed overnight and after several tests it was discovered that he had water on the brain and that there was no alternative but to place a shunt in his head. Within the space of a few days we were installed in Heywood ward and David had his shunt. Unfortunately they had trouble with the anaesthetic. I then discovered that he had to have a tracheostomy. David spent the next five days in the intensive care unit. The staff were marvellous. From the time i entered the unit they explained everything and involved me with the care of David. By the third day they started to bring David round and that was when the trouble started. The little so and so wouldn't keep his tubes in and by the fifth day the staff gave up and sent us packing back to Heywood ward.

Over the next fortnight I started to learn how to suck him out. Then the doctors started to put a smaller trachy tube in until we reached size 0. Unfortunately, about four o'clock one morning David got into difficulties and during the course of the day returned to a sized 2 tube.

After a month we brought him home and for the first time in his little life he could eat and breathe at the same time. It is lovely to watch him - we've always had trouble with his food, but now his favourite, spagetti and sausage and chips covered in tomato sauce, dissappears in a flash.

Manchester sent me home with a small portable suction machine, about the size of an ordinary camera. (By the way, Cardiff didn't have one of these). This enables me to take David out shopping and to the park, etc.

Well, there has been a great deal of ups and downs over the last few months. David couln't go back to school for more than one and a half hours per day because they didn't have enough nursing staff. One day at Debenham's stores, ladies toilet, David started to go a nice shade of red/purple because his tube was blocking --- but I won't go into that episode.

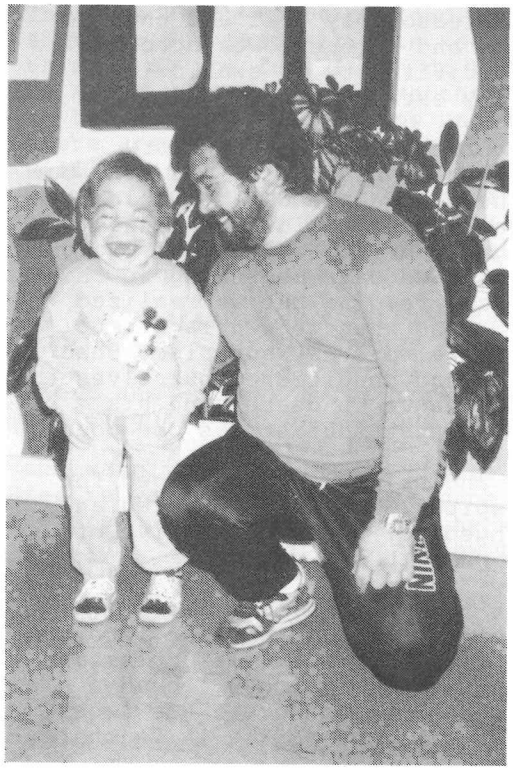
Once again we returned to Manchester for three days. They increased his tube to size three and David is breathing even better. I have now got him into school for two and a half hours in the mornings, so I am back to my 'soap box' again to get him in full time.
I know David is well enough for it, because anyone who can throw a 7lb 2oz frozen turkey at his sister in the kitchen and hit his target, is, in my way of thinking, fit for school!

It has been quite an experience and very hard work, but to see the courage of this little bionic boy has shown and the way he has fought to live and cope with his tube is worth all the effort. I know I am biased but I think he deserves a medal.

I would like to thank the doctorss and staff at Pendlebury for making our stay bearable. They were considerate and kind, especially Dr Ed Wraith. I also send my thanks to all the MPS families who came to see me, also to Mary Gardiner and her daughter who made several long trips to see David and myself.

Pat Criddle
88 Bryn Pinwydden
Pentwyn, Cardiff, CF2 7DF
Tel: 0222 732 788

****



ANGEL DAVID

You're not a burden David
You're an angel from above
Sent from God in heaven
For your mam and dad to love
With a smile that warms the winter
And brings back the summer sun.

A cuddle from your mum and dad
You're loved by everyone
I have a sister David
She reminds me so much of you
She too was sent from heaven
For us to love her too.

So when we feel depressed
And life looks all in vain
We'll look at your angelic smiles
That'll chase away the rains.

So David you're a classic
To your friends you're a chum
But truly you are loved the most
By Sister, Dad and Mum.

**** * ****

(Left) David and Kim,
his swimming instructor,
who wrote this poem for him.

****

TUBE FEEDING

****

Many of you will know us, but for those who do not, we are Bill and Sylvia Blackburn and our son Matthew aged thirteen years and nine months has Hunter Disease.

Those who do know Matthew will know him as a chubby placid boy with a lovely smile. However, things changed twelve months ago. Matthew became ill during the early part of last summer with a virus which after a number of tests turned out to be German measles. Until he was ill with the virus Matthew was very well and still mobile, albeit very slowly. However with the illness his mobility went completely and he became very lethargic and began to lose weight because he was not eating properly. He seemed to lose his co-ordination to chew and swallow.

During the summer and autumn Matthew became more and more difficult to feed and although by this time we were liquidizing his food he was not able to swallow much at all and one mealtime was leading into the next and of course his weight was continually dropping.

Bill and I were becoming more and more anxious about our little boy - he was losing interest in many things and his smiles were very few and far between. He could only go for a few weeks eating liquidized food reasonably well and then he would become ill again with colds and sore throats accompanied by excessive saliva which he had difficulty in swallowing. During these times he would hardly eat anything.

Just before Christmas our paediatrician suggested we try Matthew on "Build Up" foods which come as liquid in a tin, but to have the amount of calories he needed per day he would have to drink 1000 mls. There was no way he could drink that amount of liquid. He did manage to drink some and his weight stabilized a little. By this time we were beginning to think about and dread the thought of tube feeding but we realized that things were coming to this. So when we went again to see the paediatrician in February we were not surprised that tube feeding was suggested and we had conditioned ourselves to this. We could not let Matthew starve to death, and his weight had by this time dropped to just four stones one pound.

Our paediatrician was very kind and helpful, he arranged for us to see a dietician at the hospital. He also ordered an Enteral Nutrional Pump and arranged for a representative of the firm who were to supply the pump to see us.

Matthew had a nasal gastric tube inserted (a fine tube inserted up the nose, down the back of the throat and into the tummy). Although it does not hurt it is a little unpleasant but only takes a few seconds to insert. Matthew looked a little strange at first with the tube taped across his face but we soon got used to it and do not even notice it is there now.

There are two golden rules about managing the tube.

(1) Always remember to aspirate before feeding. You have to draw some of the acids from the stomach with a syringe and test on blue litmus paper - if it turns pink you know that the tube is safely in the tummy.

(2) Tape the tube securely across the face. If not those little hooked fingers or thumbs will soon find a gap and out comes the tube!

Matthew had a reaction to the tube for a few weeks. He was ill with a very sore throat but after this cleared up he was fine and now it does not trouble him at all - I do not think he is aware of it, except when it is due to be changed.

The pump is worked by electricity (although I do believe there is a battery operated one) and Matthew has most of his calorie intake over ten hours through the night in the form of a liquid food which contains all the nutrients he needs. The liquid food is put in a container which has a tube attachment and this in turn attaches to the end of Matthew's tube. The food is then pumped into Matthew's tummy very slowly while he is asleep and the pump works very silently so as not to disturb. He has just a little liquid food and water during the day via his tube, which we put in manually with a big syringe. The school nurse also does this for him during the day while he is at school.

The tube needs to be changed about every six or seven weeks. Although our paediatrician keeps asking me to learn how to do this, I do not feel I can do it; I am not a nurse and as it is a bit unpleasant for Matthew he does not want his mum whom he trusts completely doing unpleasant things to him. However there are no problems, either the hospital or district nurse will change the tubes and Matthew's school nurse has changed it when it has become blocked at school. The liquid food is obtained on prescription and all the attachments are supplied by the Health Centre. The dietician guides us as to how many calories per day are needed.

It is remarkable how Matthew has gained weight in the last few months (back to his chubby self). He is now five stone five lbs. He now looks so well again, he is brighter and more alert and we have a lot more eye contact. We are getting smiles again and on the odd occasion he has a fit of the chuckles and for the first time in years his hands are nice and warm. It is lovely to see him. Before Matthew was tube fed he was having terrible problems with his bowels with severe constipation because he was not getting enough food or liquid. This problem has now resolved itself with the tube feeding.

Whatever other problems Matthew may have we do know one thing - he is no longer hungry. Bill and I feel a lot happier and relaxed knowing this. Tube feeding came at the right time for Matthew. Anyone who has to face the task of tube feeding is likely to be a little nervous to start with as we were, but do not worry, before long you will be quite professional at it.

Sylvia Blackburn 11 Beatty Rd Nantwich, Cheshire. Tel:0270 626809

****

JEMMA'S STORY
How a family prepared for a child's death.

****

Many of you will have read parts of Jemma's diary in previous newsletters. Jemma died on the 22nd of June after a long fight against Hurler disease. We feel it was her determination that gave her such a full life. We always had to adapt to Jemma's needs and demands and not the other way round.

She was diagnosed in July 1984 at Milton Keynes Hospital after being admitted for a chest infection and dehydration. The consultant recognised the visual signs and gave us the diagnosis, which was confirmed after tests were taken. The doctors said to us, "You have had the best already, - it is downhill from now on."

It was an awful time. As you all know, your whole world just falls in and yet you have to carry on. We had thought that there was something wrong with Jemma but we couldn't begin to imagine something as awful as MPS.

Jemma started school part-time in February 1985. She always enjoyed school and was soon leading a very busy life as her school diaries have shown. She was keen to learn and school encouraged her and many milestones were crossed.

In June 1989 Jemma stopped walking. One day she could, the next she couldn't. This was a shock for us as we had expected a gradual loss of movement as learning to walk is a gradual learning. By now Jemma was deteriorating in many other ways. In February 1990 she lost the ability to sit up unaided. She did enjoy many things still and was at school four afternoons a week. We would read her favourite stories and sing songs that she knew well and she would respond.

Jemma's breathing at night when she was asleep was her biggest problem. She would not breathe for very long periods and this was traumatic for her and for us. When she went to relief care for a night about once a month she was on a monitor.

As the last few months got worse we were told her chest was very weak and that the next infection might not respond to antibiotics. We discussed the use of sedative drugs instead. This was the help we felt best for Jemma in June. It was given to her at bedtime and when she went to sleep it was a relaxed and peaceful sleep with none of the trauma she usually suffered at night. Mick and I were staying at the hospital with her and Mick describes it as almost beautiful. Jemma slipped away in the early hours of the morning.

Both Mick and I felt relieved and confused but amazed at Jemma's strength to fight and to give so much to us and to others. We are lost and miss her but life was so special to her and she had the ability to show that to others. This is why our children are called "Special Needs". It never made any sense to me before as we always led a normal family life, or so I thought.

The diagnosis is the hardest part of being an MPS parent, we think. The acceptance that the doctors can do nothing for your child. Coping with other people's shock as you tell them what is wrong with your child and their inevitable response that there must be something that can be done. If people asked me what the future held for Jemma I would always give the truth - it was their problem if they could not cope with it, not mine.

Last year Mick and I arranged to see our funeral director. We had found out about Hurler's syndrome and we wanted to find out about funerals. Some people who were trying to be kind had told a friend who had lost a baby with a cot death some dreadful stories so we went to find out for ourselves.

It was a very difficult time for us both but we felt so reassured that we knew what choices we could make and what happens about funeral arrangements. We wrote our instructions down and left them with him. Then when the time came we knew that we had arranged everything at a time when we were both thinking straight.

Planning a funeral in advance may seem an unusual thing to do, but we are not the only parents to consider this and arrange it. Nobody wants to talk about funerals, that's all. I have seen articles since in magazines and our crematorium recently had an open day. I found out that you can arrange to visit your local crematorium by ringing the director during office hours or by contacting a funeral director. We found it did help to make our choices in advance. It helps us to know that we did the things that were right for us and for our child.

This may be an odd read for the newletter and I know some people have been concerned about the effect on new families of talking about the death of a child. I hope it will help some families and not confuse or shock anyone. If you want to know more please contact us.

Sarah and Mick Corbett
25 Alledale Place
Hodge Lea
Milton Keynes
Bucks MK12 6JN
Tel: 0908 312440

****

"A life is not measured by the number of days, months, or years that it is lived,
but by the effect that life has on other people who touch it,
It is not measured by the length of it's days or the height of it's achievements,
but by the breadth of it's influence."

Alex and Rhonda's Trip to America

Two children in the Society Alex Butler and Rhonda Brierly were lucky enough to go to Florida with the National Holiday Fund. The President of which is Richard Branson.

Alex and Rhonda along with ten more girls and boys aged 16-18 travelled with helpers, organizers, a doctor and their very own video camera man, a party of 28 in all.

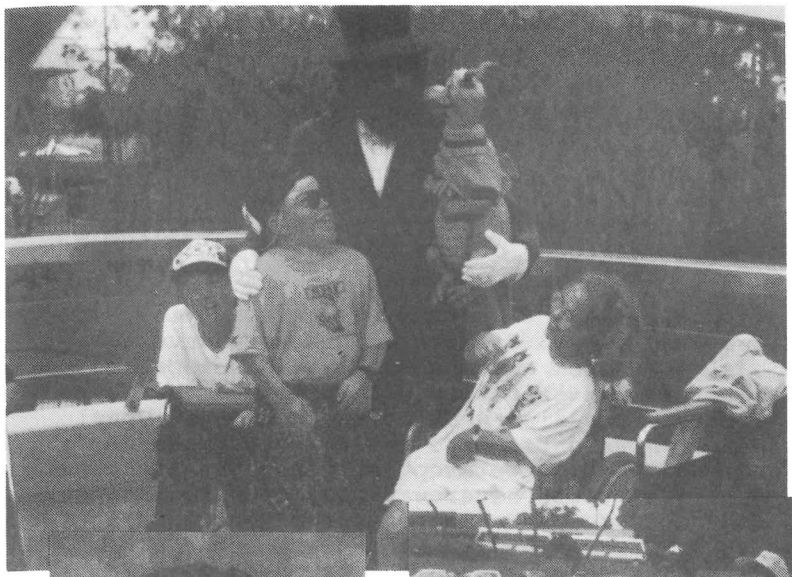
They travelled Virgin Airlines and stayed at the Hilton International Hotel in Granada, and spent two wonderful weeks in ideal temperatures around 80 F. They travelled around in three mini buses named "Beanz " Meanz" "Heinz". After a good flight lasting eight hours they were ready for a fun filled holiday. Visiting the Epcot Centre, the Kennedy Space Centre and Cyprus Gardens. Two magical long days in Disneyland etc. Being kissed by Snow White and Cinderella, cuddled by Mini Mouse, they were all made to feel like the stars.

Nicola, one of the children had a birthday party in Disneyland it continued all day, "the most fantastic birthday I have ever had" she said. Alex's helper Andy a policeman and Alison his wife a nurse who was Rhonda's helper were fantastic, they had as much fun as the children. I would like to thank Carol Hubbard, Christine Lavery and Mary Gardiner for suggesting Alex and Rhonda for the trip. Alex is one for smiling, but his face when he was told he was going was incredibly happy, it brought a lump to my throat.

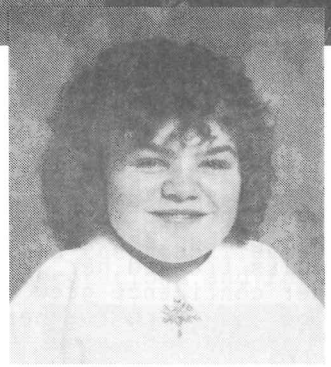
I hope that in the future more of our children can benefit from the generosity of the National Holiday Fund, and they too can have the most fantastic holiday of a life time.

Sue Butler - a very grateful mum.

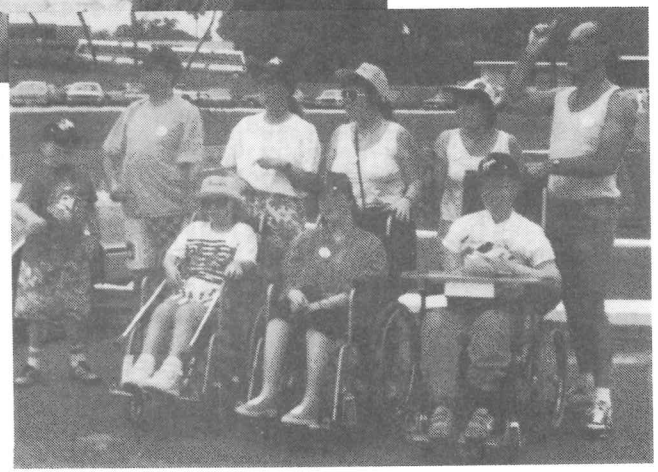
Rhonda Brierley at the pool with helper Alison.



Alexander and friends at the Epcot Centre.



Rhonda at school May 1990, age 16.



All together at the Cyprus Gardens Orlando, Florida.

They're coming to take us away!

****

WHICH WHEELS?

****

It is very important for children who cannot walk, or who can walk only a short way, to have the freedom and independence which can come from driving themselves in an electric wheelchair or battery operated scooter. Over the years, by trial and error and by hearing of other people's experience, we have built up some knowledge of what to look for in choosing an electric chair and thought it might help others if we shared it.

Our daughter Helen has Morquio disease and is very severely affected. She has never been able to walk any distance and only once managed with much effort to walk home from her school. Helen's wrists are also weak so she could not wheel herself, even in a lightweight chair. We delayed getting anything electric, not wanting to stop her using her legs, but it became clear that she could never walk any useful distance and would always have to be pushed to and from school in a buggy.

When Helen was five we went to the NAIDEX aids for the disabled exhibition and tried out a number of scooters. We chose the Pony made by Ortho-Kinetics as it was the smallest and most attractive looking for a child. Helen learned extremely quickly to drive the Pony with great skill, but I make sure I keep up the insurance just in case! It was wonderful to see her driving home in a gang with her friends instead of being stuck with the mothers as I pushed her. We noticed at once that other children and adults treated her differently when she was mobile and that her confidence grew accordingly. At that age we felt it would be very hard for her to keep her friends if she could not "run" around with them. Once the school was used to the idea, they allowed her to use it in the playground so she could join in games instead of sitting on a bench.

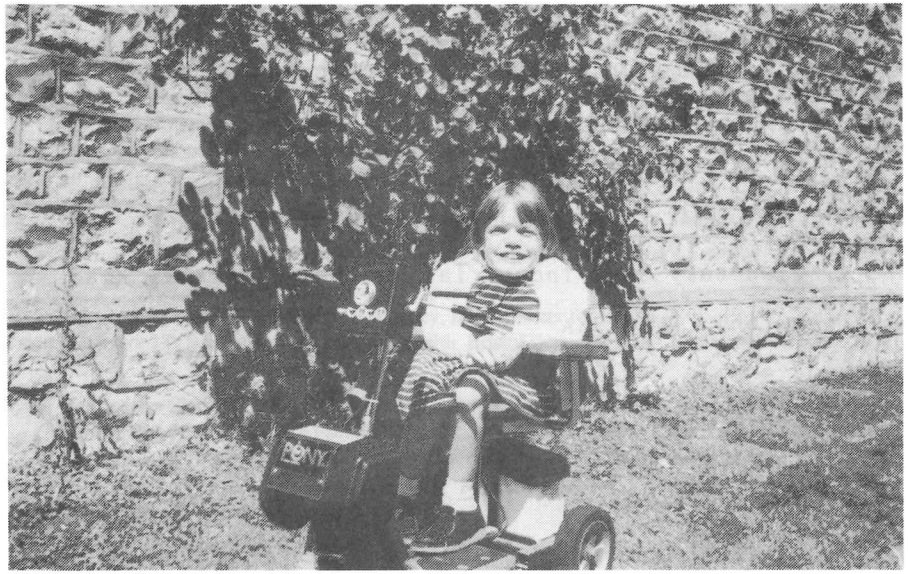
We still feel the Pony has been the best choice for Helen's needs, although I must say I have not always been impressed with the after sales service.
Helen cannot walk at all now and uses the Pony to get around indoors as well as to go on errands to the shops. Being small, the Pony can be driven round all but the smallest shops. It can be taken apart and put in the back of our estate car. I can manage to lift it, but was finding it a bit of a strain on my back so we have recently bought a folding ramp from a firm called Rolac which was adapted to fit against the rim of our car boot. Helen drives to the car and climbs into her seat while I drive the Pony up the ramp into the boot. The ramp folds to fit in the back and is available to unload the other end. We also find it useful as a ready made adaptation when visiting other people's houses.

Helen started her new secondary school last September. It is a mainstream comprehensive adapted for disabled pupils. She is taken by taxi so it was not practical for the Pony to go to and from school with her. We were asked to consider whether a Turbo chair which has a seat that can be moved to any height would be suitable but decided against it.

We felt it was important for Helen to maintain what little strength she has in her legs and were afraid that if she sat all day she would become even weaker. The Education Authority therefore provided a number of Tripp Trapp chairs which can be adjusted to any height to be placed in all the classrooms Helen uses.

We applied, with a letter from our Consultant Paediatrician, to the Department of Health for an electric wheelchair for use at school. The DoH will not provide powered chairs for use out of doors (I believe this is something to do with their responsibility should a user become stranded somewhere unsafe by a battery running out). They will provide them on free loan however for use indoors and in the playground.

After the usual long wait, I was told that there was no saying when a chair could be provided as BEC, the manufacturers, no longer made them and existing chairs were being reconditioned. I telephoned BEC and learned that they had indeed discontinued the old model which they felt did not go fast enough to enable a child to keep up with friends, but were still making a better, more expensive model which of course the DoH would not buy. Through BEC we were put in touch with Finchley Golf Club which was looking for a child to whom they could donate a BEC wheelchair and Helen became the proud owner of a very smart and exceedingly fast model which she uses at school. In some ways she prefers it to the Pony; the seat is more comfortable and it goes faster, but I find it heavier to load in the car and the stabilisers at the back play havoc with the decorations and furniture.



Hell on Wheels or Heaven on Wheels -
The choice of machine can make all the difference -
says **Helen O'Toole**

Helen would love to have yet another type of scooter - a larger adult one which would climb kerbs and enable her to go out on longer journeys more safely - I worry that cars will not notice her on the Pony because she is so low down. She has tried a larger model at NAIDEX and rode it round a type of assault course to the visible alarm of the salesman who could not believe such a small person could possibly manage safely when adults were having such difficulty. We do not have anywhere at present to store a large scooter, it would not fit in the back of the car and Helen would not be able to get into all the shops she can manage on the smaller Pony, so for the moment the idea is shelved.

There are also choices to be made between lead-acid batteries and the safer gel batteries which are more expensive. If the scooter is to be used a lot or if you live in a hilly area you will need a more powerful battery but the extra cost may be compensated by a longer battery life.

I hope this gives some idea of the considerations to be taken into account when choosing the right model. No one model is perfect in all situations and it is worth spending time to work out what will suit your child and family situation. I list a few manufacturers below. Their sales representatives will bring a model to your home for you to try out without obligation to buy.

Electric wheelchairs and scooters are not cheap. The indoor version can be acquired free through the DoH but others must be bought privately. There are a number of Charities which fund the purchase of powered chairs and the Society can help you to apply for help. It must be remembered however that the cost of repair, battery replacement and insurance will have to be met by the family. Mobility allowance will of course help towards these costs.

Happy wheeling! **Mary O'Toole**

Manufacturers

Ortho-Kinetics (UK) Ltd.
Unit 4, Planetary Road Industrial Estate, Planetary Road
Wednesfield
Wolverhampton. WV13 3XA

Tel: 0902 866166 **PONY**

BEC Mobility Ltd
Fens Pool Avenue
Brierley Hill
West Midlands DY5 1QA

Tel: 0834 263191 **SMALL ELECTRIC CHAIRS**

Everaids
38 Clifton Road
Cambridge CB1 4ZT **TURBO**

Rolac Ltd
Unit 16, Enterprise Centre Two
Chester Street
Stockport SK3 OBR

Tel: 061 429 8477 **PANDA FOLDING RAMP**

Stokke Fabriker AB **TRIPP TRAPP CHAIR**
Box 122 57301 Tranas
Sweden Tel: 0140 19010

NEWS FROM GROUPS FOR SPECIFIC CONDITIONS

ACTION FOR THE CARE OF FAMILIES WHOSE CHILDREN HAVE LIFE-THREATENING AND TERMINAL CONDITIONS

The Institute of Child Health
Royal Hospital for Sick Children
Bristol BS2
Tel: (0272) 221556

ACT has carried out a national survey of existing support services (medical, social, educational and befriending) and continues to monitor resources available to families as a whole, whose children have life threatening conditions: the emphasis being on those services which make it as easy as possible for the family to care for their child/children themselves, in the main at home. The survey included educational and personal support services for professional careers.

ACT aims to:

* collate information on all support services for families (statutory, voluntary and parent support groups)
* improve communication and liaison between all those involved in this field of family centred care
* establish a national information and resource centre

"LISTEN, MY CHILD HAS A LOT OF LIVING TO DO"

1st National Conference on The Care of Children with Life-threatening Conditions and their Families.

Monday 1st October 1990, Regent's College, Regent's Park, London

Opening by
Virginia Bottomley, MP, Minister for Health
Chairman: Professor J D Baum (Institute of Child Health, Bristol Department of Child Health, University of Bristol)

Guest Speakers include:
Bernardette Cleary (Rainbow Trust), Peter Jeffery (Brookfield House School, Waltham Forest), Christine Lavery (Contact a Family, MPS Society), Jacqueline Mok (Edinburgh City Hospital), Bob Woodward (CLIC-Cancer and Leukaemia in Childhood Trust)

Details from ACT at above address.

There are a limited number of bursaries available to parents of special needs children wishing to participate in this meeting. Enquiries to ACT at the above address.

****

A TEACHER'S POINT OF VIEW

****

Three years ago, beginning at a new school, I was first introduced to MPS. Two of my four pupils were diagnosed with Sanfilippo disease. One of the students is still in my classroom. The other has moved on to an adult day programme in the area.

In the past three years I have painfully watched as Danny's skills have diminished. Two years ago this past June, I remember being eight months pregnant and chasing after Danny down the long hallway in our school because he decided it was time to go for a quick run to use up some of his extra energy.

Now Danny only walks with the assistance of one or sometimes two staff members and is in a wheelchair most of the time. His wheelchair even reclines for those lethargic periods when he is already sleeping when he arrives at school.

The goals I have set for Danny are "fun" goals. They teach him socialization, and leisure time activities and they help him to maintain his upper body strength (which is unbelievably good). But these goals have been modified as the years go on. During ball play I used to have to dodge the ball as Danny threw it at me and laughed. Now I have to sit in front of him and encourage him to hold the ball and pass it to me and he still laughs sometimes.

Danny has taught me much more in the past three years than I could ever have taught him. He has shown me that although he can't run through the halls, or try to hit me with the ball, or clear the top of my desk with one swoop of his arms, his need for love has not diminished by one slight bit. Nor his ability to love. Those mid morning hugs and wonderfully sloppy kisses mean more to Dan and to me than any ball I ask him to pass or any scooter board I ask him to push.

Some people who work with people with severe disabilities tend to forget the emotional side of life, and continue to concentrate on work, work, work! I know the work is important but so are other things. We hug and listen to music. Danny will lie on the floor mat and I will just sit and rub his back. I want him to know that I love him. I want him to know that I am there when he needs me and when he gets his bursts of independence I am there just to let him do it by himself and be proud of him.

One problem we have recently encountered is that it has been requested of the staff I work with to put Danny on a behaviour programme to address his "hand in mouth" behaviour. Danny has been putting his hand in his mouth for at least three years. As his ability to do other things diminishes, his time with hand in mouth increases. We prompt his hand down as often as possible, but still it continues. Once in a while he will get an open sore on one of his hands or fingers.

At these times Dan is placed in splints which prevent him from bending his elbows and putting his hands in his mouth. He has only to wear these for a few days at a time during waking hours until the medication on his hands takes effect and the sore begins to heal.

I know that this behaviour is part of the disease. He doesn't always have control over the things he does. I don't feel a behaviour programme will do anything but frustrate Dan and make him angry. Fortunately the professionals and residential staff who work with Dan feel the way I do. Unfortunately the inspectors who come to review our facility feel differently.

I have spoken to Marie Capobianco concerning this and I am in the process of getting in touch with Dr Steven Amato, in Baltimore, to hopefully get something, anything in writing to prevent the behaviour programme from going into effect.

I understand that the inspectors are doing their jobs, they just don't seem to fully understand Sanfilippo disease and its ramifications. There are too many people out there who don't understand it. I used to be one of them until three years ago, but I learned and am still learning with frustration and many many tears.

Danny's mom visits him frequently and comes to see him at school when she can. He loves her so much. There are times he tries to hold her hand. She once asked me if I thought he still recognises her. He doesn't always respond to her with smiles and handholding, but I know he remembers her. It's just something in his eyes and attitude, nothing specific or tangible, but it is there. She cries, I tell her it is okay to cry, I give her my box of tissues and I listen. Then after she leaves I cry. I don't know why I wait until she is gone. Maybe I'm trying to be strong for her and hold her up as her son changes right before her eyes. She knows Dan is special to me and she probably even knows that I cry.

It is frustrating to be doing the best you can for someone and have somebody else tell you it isn't enough. Thank goodness I have found someone to turn to who can lead me in the right direction to teach others that strict programming isn't always the best thing. The teaching that I do with Danny is good and I feel he benefits from it. But flexibility is important and if he just wants to hug or have his back rubbed, well that is just as good!

Maureen Whelan
Special Education Teacher
Norwalk, Connecticut, USA.

(Reprinted from the newsletter of the American MPS Society).

****

All is now set to go for the International Conference at UMIST, Manchester 1990. For those who can come we look forward to seeing you there and hearing all your news. If you cannot come, look out for the Autumn newsletter and the conference report.

****

****

THE EDITOR SQUEAKS

****

Several excellent magazine articles were sent to me for this newsletter. Unfortunately they could not be used because they were photocopies. The edges were faded and the photos were completely dark.

Please send the originals from newspapers or journals! Our printers can usually get very good results from these and other families will be delighted to see you in print!

Trust me! Your copy will be sent back to you in good condition without fail.

Many thanks for the photos and articles sent in. We have some wonderful items in this edition. Remember if your family does not feature, it's because you haven't sent anything! Members really do want to hear from you. Several of the articles this time show how members can pass on the fruits of their hard earned experiences with their accompanying joys and sorrows.

If you send photos please write the names on the back and stiffen the envelope with card. One last thing, I am not an ironmonger! Keep your paper clips and staples for the man who comes round with the cart. They damage the paper and make the printer very unhappy. Copy for Autumn by 7th of October please.

Charles O'Toole

****

VIDEO CAMERAS

The Society owns several video cameras which it loans out to members.

WILL MEMBERS WHO ARE CURRENTLY USING A CAMERA, PLEASE LET LINDA GOLDING AT THE OFFICE KNOW.

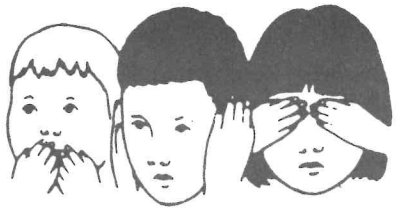
The cameras are needed at the UMIST Conference at the end of August. If you are not coming to Manchester, we need to arrange to collect the camera in good time.

PAYING IN BOOKS

The Society has a new Treasurer, Pauline Mahon, and will be changing its banking arrangements. If you have a paying-in book, please return it to Pauline who will send you a new one.

Pauline's address is:-

41 Stumperlowe Crescent Road,
Sheffield 10.
South Yorks.
Tel:- 0742 - 304069



AFASIC

Association For All Speech Impaired Children

Children with Speech and Language Disorders

Educational Facilities Available for Communication Impaired Children

AFASIC, 347 Central Markets, Smithfield, London EC1A 9NH

CHRISTMAS FUNDRAISING

With the swift onset of the Festive Season, I thought I would remind you of a fun way of raising a few pounds for MPS - hold a Sherry and Mince Pie Morning.

With a little bit of persuasion you should be able to get the Sherry on "sale or return" from your local off-licence, they may be able to lend you the glasses as well. So much posher if you have all the glasses the same! Of course, if you try really hard you may get the Sherry donated.

Either make your own mince pies or buy them from the supermarket or, better still, have friends and neighbours make them for you.

Make up your own tickets, just small ones, and have them photocopied, showing the date, place time and cost. I would suggest that 1 for the first glass of Sherry and a Mince Pie is not too extortionate. You then decide on a charge for any further glasses of Sherry, coffee, tea or Mince Pies.

You could also have an MPS Stall selling our pens, keyrings, pencils, tea towels, ties etc, and perhaps your friends - after making all your mince pies for you - would knock up a few cakes for sale!

Dig out a few Christmas records to put on the stereo or get a local entertainer to come along for an hour, and everyone will soon be enjoying themselves and getting in the mood to spend some money.

I know it may seem a little early to be thinking of Christmas, but it pays to start arranging things like this in plenty of time.

Have a good Sherry Morning.

Ron Snack

****

VINTAGE CONFERENCE REPORTS

****

On sorting out the MPS shed we had unearthed lots of back numbers of conference reports. 1986 was a particularly fruitful year. It has discussions on the different diseases and two important articles on anaesthesia and a wealth of other information. We have three boxes in stock. If new families have not had a copy of this report please contact Linda Golding at the new MPS address or ring her at the office number.

Families can have an extra copy if they send a 34p stamp to cover the cost of the postage. If there are other years you wish to have Linda may be able to help you. We have a good stock of the 1989 conference report available and extra copies are available by consulting Linda.

****

PETROL TOKENS

If anyone still has any Shell, BP, or Esso petrol tokens that they do not need, could you please pass them on to myself or David Briggs who will make good use of them.

Ron Snack

IN MEMORY

Our sincere thanks to everyone who has made donations to the Society in memory of Mrs Gregory, Mrs Blumenfield, Iain Ireland, Monica Briggs's mother, and Paul and Cheryl Evans.

MENTAL HEALTH FLAG DAY

Although only five families decided to take part in the collection on Mental Health Flag Day in May, between us we collected £942.27p. That means we will recieve over £800.00 from the Mental Health Foundation, after they have deducted their percentage for doing all the paperwork involved in arranging the whole day.

This seems a very worthwhile amount to have raised in one day, especially as everyone involved seems to have enjoyed the day. Perhaps next year a few more of you will feel like joining in with the collection.

Ron Snack

LONDON MARATHON 1991

If you know of anyone who is planning to run in the London Marathon next year, and is looking for a good cause to run for could you please ask them to consider running for us. We can supply a running vest with our own logo and as many sponsorship forms as necessary.

Ron Snack

SALE OF GOODS PARTIES

Don't forget that you can have a good time with not very much organising, and raise some money for the Society, by holding a Tupperware party, or Make-up, Sale of Linen etc. The Tupperware dealer will often donate a few items that can be raffled on the night.

Just one thing to remember, if you are holding an Ann Summers party please let me know in good time so that I can keep the date free.

Ron Snack

****

BACK TO BABYHOOD - Adult Sufferers from Sanfilippo

****

Buddy Armstrong looks lovingly at a picture of two healthy toddlers, and then turns to smile at his children, now nearly thirty years old, but still babies.

Terry aged 29 and his sister Susan 27, were no different from other children. They were inseparable best friends, loved to climb trees, ride bikes and play in the sand - until they reached the age of seven.

When Terry started school it was thought he had behavioural problems. He was then diagnosed as having the rare syndrome Sanfilippo disease. By this time it was also obvious that Susan's slow speech development was something more serious.

The children's normal growth and development had stopped at early school age. Since then they have regressed to babyhood. It was a gradual process - just like growing up. They stopped talking and nine years ago they stopped walking. Since then Buddy and his second wife Fran have been caring for two adult babies in their home on Queensland's sunshine coast.

Buddy who is now fifty six, was told that his two beautiful toddlers would die before they reached fourteen but he refused to place them in an institution, believing a loving home environment was best. They have been able to continue to care for them at home.

According to US experts Terry and Susan are now the oldest Sanfilippo survivors. The odds against having the genetic disorder are enormous (an estimate from a study in the Netherlands suggests an incidence of 1 in 24,000 - Ed.). At the time of Terry and Susan becoming victims there were only four known cases in Buddy's home state of New South Wales. "And I had two of them," Buddy says.

"We were told to put them in an institution and go home and have another baby. But that would have been like playing Russian roulette," says Buddy.
Terry and Susan suffer from the type A form of the disease. Their stepsister, Colleen, is a normal healthy teenager.

Adapted from the New Zealand Newsletter.

Editor's note:
The oldest living Sanfilippo sufferers in the UK who are known to the Society are **Peter Benbow** who will be twenty six in December and **Sally Ann Brookfield** who was twenty five this July.

****

New Family

Pamela and Mike Croghan from Bramhall, Cheshire, whose three year old son **Daniel** has been diagnosed as suffering from Hunter Disease.

FUND RAISING EVENTS

Once again, many thanks to everyone who has raised money for us, in whatever way, in the past few months. This time the fund raising ranges from collecting boxes in corner shops to a Dance Night held in Glasgow and a Safari Supper in the wilds of Lancashire, via Bric-a-Brac sales, marathons and raffles.

T Hurdle, London	Swanley half marathon
P Chandegra, Birmingham	Works Raffle
Horwich Inter Church Fellowship	Bric-a-Brac Sale
Mrs Clearie, Glasgow	Bingo Night
Forresters/Young Forresters, Glasgow	Various Fundraising events
Mrs Todd, Glasgow	Two Collecting Tins
Clydebridge BSC, Glasgow	Bottle Fund
Alan and Amy Bottrell	Saving 20ps
Alan and Fiona Byrne, Glasgow	Dance night
Burnside Hotel, Glasgow	Collecting Tin
Workers at Hinckley Point	Donated Vouchers
G and L Finch, Bridgwater	Sponsored Slim
The Harveys, Oxford	Copper coin collection
Bridgwater Swimming Club	Raffle
Margaret Horsley, Cumbria	Collecting Tin
Mrs Hardy, Peterborough	Copper coin collection
G Rowe, Erith, Kent	Collecting Tin
W Robins, London	Levy on drinking during Lent
Mr and Mrs Rock, Leicester	Collecting Tin
Dr and Mrs Dagnall, Bolton	Plant Sale & Coffee Morning
Rampton Hospital, Notts	Collecting Tin
A Vardy, Notts	Sponsored Slim
D Briggs, Notts	Sponsored Slim
Willesborough C P School, Kent	Various sponsored events in memory of Cheryl Evans
Sandra Black, Cleveland	Sale of Satin and Lace
Bet Williams, Liverpool	Sale of Home Made Marmalade
Westhoughton Inner Wheel, Lancs	Safari Supper
S Cully, Kent	Fun Run
Mrs S Beamond	New Years Eve Dance
W Robins, London	Collecting Tins
Mrs Freeborn	Fundraising by Class 2T
Sue and Dan Butler, Oxon	Cheese and Wine Party
Ian Marshall, Oxon	Money saved by not smoking
Sue and Dan Butler, Oxon	Luncheon Party
Ashford Junior School	Sponsored Swim
Ballyholme School	Various sponsored Events
Geoff and Selma Oulton, Lancs	Xmas Party
G and A Barnett	Raffle
K and B Houston	Night at the Races
R and C Lavery	Collecting Tin
J and H Dean	Coffee and Mince Pie Morning
Mrs Le Page, Channel Islands	Xmas Carols
M and P Skidmore	Carol Concert
Kingsey Carol Singers	Raised Money
St Mary's School, Haddenham	Raised Money
G and P Criddle	Collecting Tin
C and P Hubbard	Sale of Stamps
F A Sandow	Sale of Stamps

RESTRICTED GROWTH ASSOCIATION

* PRESS RELEASE * PRESS RELEASE * PRESS RELEASE * PRESS RELEASE *

The Medical Committee of the RESTRICTED GROWTH ASSOCIATION have produced a 3rd edition of their helpful booklet "The Layman's Guide to Restricted Growth". The Association have also reprinted their booklet "Coping with Restricted Growth"; a booklet giving everyday advice and help to families.

There are a number of rare conditions causing severe short stature and in around 90% of cases, affected children are born to parents with no history of restricted growth in the family. The information provided by these booklets is helpful to families understanding their child's condition; to affected individuals and to their professional advisors.

The Layman's Guide
To Restricted
Growth

- describes types of short stature and particular disorders.
- associated medical complications common patterns of inheritance, pregnancy and parenthood
- how a diagnosis is made.

WITH NEW CHAPTERS ON:

- feelings of families at the time of diagnosis.
- new developments in limb lengthening surgery as a posiible treatment for short stature.

Please return the slip below to:

RGA Administrator
103 St Thomas Avenue
Hayling Island
Hants PO11 OEU

PLEASE SEND ME (remittance payable to RGA)

---------- copies of "The Layman's Guide to Restricted Growth" @ £1 each

--------- copies of "Coping with Restricted Growth" @ £1 each

Please send me details of bulk orders (tick)

Name & Address _____