NEWSLETTER mps SUMMER 1983

THE SOCIETY FOR MUCOPOLYSACCHARIDE DISEASES

N E W S I N B R I E F

CONGRATULATIONS

John and Sandra Hartley from Warrington on the birth of their second daughter, Rebecca, on May 12th. Their first born daughter, Emma suffers from Hurler disease and recently left hospital after a Bone Marrow Transplant.

Susan and Robert Heath from Amersham Common on the birth of their third child, a son, Christopher on May 26th. Susan has managed to combine pregnancy with the hectic job of co-ordinating fund-raising events. Her husband Robert is a Trustee.

Mr. & Mrs. Taylor, on the birth of their second son Nicholas. Christopher, their first son suffers from Hunter disease.

RICHARD TURNER

Richard and his parents returned home recently after a successful Bone Marrow Transplant at the Westminster Children's Hospital. Richard's ordeal lasted just 6 weeks as he was lucky in having a perfect match donor in his 10 year old sister Claire.

CHILDREN IN NEED FUND - BBC

Back in April, The Society received a donation of £700 from the fund to help towards our Family Weekend/Conference.

CALLING ALL MUM'S IN ESSEX

Jean Silvey who lost her own daughter Treena from Sanfilippo disease last December has very kindly offered to hold a coffee evening for other MPS mums in the Essex area. Everyone is welcome and we thought it would be a good opportunity for myself (Christine) and some of the other Committee members to meet and get to know one another. We will bring our MPS stationery to sell on the evening and anyone wishing to take a selection away to sell to friends, will have the opportunity to do so. If you would be interested in coming to this coffee evening, please would you either write or telephone Jean Silvey, 5 The Larches, New Thundersley, Essex. Tel: South Benfleet 56623.

HTV WEST - PUBLIC SERVICE ANNOUNCEMENT

Sue Bramford from Swindon, travelled to Bristol on the 11th July, for the filming of a Public Service Announcement for The Society. This should be shown sometime in August. Let's hope we get good response. Sue's daughter Toni suffers from Sanfilippo disease.

MARATHON AND SPONSORED EVENTS

Congratulations to our runners in the London Marathon, the Guinness Grand Union Canal Medical Meander, Tetbury Sponsored Walk, Pottery's Marathon, Piccadilly Charity Marathon and the Vicker's Worthing Marathon, for their marvellous runs. All completed the distance. Thanks for being sponsored for MPS.

We should also congratulate our sponsored parachutist - David Bower, who made his very first jump in aid of MPS.

HEALTH VISITORS JOURNAL

Catherine Grant our Liaison Officer, wrote an excellent article - entitled (The Society for MPS) for (The Health Visitor), which was published in the July 1983 issue. Perhaps you might like to tell your own Health Visitor about the article. Maybe she will let you see a copy.

MPS COFFEE SNOWBALL

The Coffee snowball is gathering momentum and we have just topped the £4000 mark, so we have a long way to go. Due to the enthusiasm shown, we have extended the completion date to the end of September. Many thanks for all the effort which so many of you have put in, into turning the coffee mornings into major fund-raising events. Please do continue the good work and encourage your friends to keep the Snowball going.

'WOMAN' MAGAZINE

Aaron Fitts, aged 20 months, who suffers from Hurler disease and the Society, will be featured in an article in 'Woman' magazine in July/August.

DON'T FORGET

We now have a large stock of Charity Boxes, labels and MPS Posters. Just drop Christine a line if you would like some posted off. We have found a surprising number of shops, building Societies and Post Offices willing to display these and the posters are ideal for spreading the word about forthcoming MPS events, Coffee mornings etc.

IMPORTANT NOTE: The articles and views expressed in this publication are not necessarily the views of the Society.

FAMILY WEEKEND/CONFERENCE 28th - 30th OCTOBER 1983

What a Weekend it should be. Over 40 families will be at the Conference to be held at 'The Post House Hotel, Chapel Lane, Great Barr, Birmingham. (Junction 7 of the M5/M6).

We have been fortunate in securing experts in the various fields of MPS to come and present talks on many aspects including Pre-natal diagnosis, clinical and biological diagnosis, bone marrow transplants and amnion implants. There should be ample opportunity for those attending to ask questions.

We have arranged an outing to Dudley Zoo for all the able bodied MPS children and their brothers and sisters. For those unable to go on the outing there will be a creche with expert supervision.

Sunday morning will provide you with an opportunity to learn more about and enter into discussions on Sociological aspects of caring for your MPS child. Details will be available on Mobility and Attendance Allowance.

Those who have indicated they wish to attend the Weekend will be receiving final details soon.

We all look forward to meeting you in October.

If any members of the medical profession known to you are interested in attending the Conference on the Saturday, we have approx 50 spare seats which will be allocated strictly on a first come first serve basis. The cost of the 1 day conference, including coffee, lunch and afternoon tea is £15.00 per head payable in advance.

Christine Lavery
for the Core Committee.

SPONSORED SUNFLOWER COMPETITION

Many of you have been in touch to say how much you are enjoying our simple, although I admit not original, idea of The Sponsored Sunflower Competition. As we had hoped it is proving to be a fun event for the family and friends.

We look forward in September to hearing who has the greenest fingers and don't forget to try and get a photograph record of your success as we shall feature the winner of the tallest sunflower in our October Newsletter. In the meantime we would like to express our thanks to Hyrons Nurseries, Amersham for supplying the seeds and to Fothergill's seeds for donating £10.00 towards the prize for the tallest sunflower.

Christine

AMNIOTIC CELL TRANSPLANTATION

One of the most important scientific mysteries that faces modern medicine and particularly modern immunology, is that of the survival of the human baby during its development in the mother's womb. Put simply, the baby shares half of its genetic complement with its mother, which should, therefore, not evoke any hostile reaction. However, it also has a half complement of genes from its father, who is obviously genetically different and this represents a potential antigenic challenge to its mother. In immunological terms it is as if a piece of foreign tissue has been presented to the mother to which she would normally mount a reaction. This, of course, does not happen and on the contrary the mother accepts and nurtures the foetus and allows it to reach maturity. Occasionally things go wrong and the mother does indeed mount a reaction against the foetus and the best known of these is that of the Rhesus incompatibility, when a mother who is Rhesus negative has been exposed to blood from a Rhesus positive first baby and develops an immunological memory against the Rhesus factor which then leads to the destruction of a subsequent Rhesus positive bay's red cells.

Much research has been invested in trying to find out how the baby survives in this immunologically foreign environment because the benefits of this knowledge could be extended to many aspects of tissue transplantation and cancer work. Needless to say we do not know the answer as yet but we do know that a layer of cells separates the baby completely from the mother at every point throughout gestation and this is called the trophoblast. The biggest concentration of this tissue occurs at the placenta or afterbirth and is very important in separating the baby's blood circulation from the mother i.e. the two circulations normally never mix. The trophoblast is a fascinating tissue and behaves very much like a tumour cell in that it is able to survive In a foreign environment and can actively invade the mother's tissue to establish a communication with the mother. It is thought that one of the ways in which the trophoblast cell acts is that it does not express on its surface its antigenic identity and so is not recognised as a foreign cell by the mother's immune defences. The situation is not dissimilar to two football teams in which each player knows his opponent by the colours of his jersey, but the situation would be very difficult if all the players had no distinguishing features. In this way, therefore, it is possible to imagine that the layer of trophoblast "Hides" the baby within its protective environment and so the mother is not able to pin point or identify antigens from the baby and cannot, therefore, mount a reaction against it. I must at once say that this is a very simplistic description and serves only to illustrate a principle.

My interest in this work originated from an attempt to recreate the special circumstances of the baby for transplants of pancreatic islets. These islets are specialised groups of cells which normally are found in the pancreas and are responsible for secreting insulin and other hormones necessary to control sugar metabolism. The failure of the correct function of these cells leads to insulin dependent diabetes and its complications. These islets can be isolated but transplantation from one individual to another has been a great problem because of the immunological factors involved.

It seemed reasonable, therefore, to try and wrap these islets up in some sort of membrane, which would prevent a hostile reaction from the host and yet enable the secretions of the tissue to escape and help to treat the patient.

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Attempts at doing this with artificial membranes have been partially successful in experimental models, but we have yet to find a suitable membrane which will not itself produce some sort of reaction and end up getting walled off by the host. The search for a natural membrane led to consideration of the trophoblast, but this was discarded because trophoblast, as I said before, has a malignant potential and indeed a significant number of pregnancies where a small amount of trophoblast is retained can lead to the condition of hydatidiform mole and the highly malignant chorion carcinoma. The dangers of implanting live trophoblast, therefore, are obvious and this tissue was, therefore, not suitable.

In man, however, there is a second membrane within the trophoblastic layer called the amnion, and this too separates the baby from the mother at almost every point of contact. This membrane is much stronger, highly differentiated, has a suitable pore size and has never been known to turn malignant.

The membrane was, therefore, investigated in more detail and like the trophoblast it was found that it too does not express on its surface its antigenic identity. This discovery confirmed the observations of surgeons and doctors, who in the past used the amnion as a dressing in burns and ulcers and indeed have even implanted it into patients who had poor circulation to the limbs in an attempt to try and improve the blood flow. In all these patients very little in the way of reaction was noted with these implants and none were found to have come to any harm from them. This circumstantial evidence allied with our laboratory work encouraged us to try an experiment in which I and a number of colleagues implanted each other with a small piece of tissue to see what happened. I am pleased to report that, in fact, very little happened and that the tissue was shown to survive up to two months without any obvious immunological reaction when we tested our blood and our cells.

It was at this point that we decided it would be worth trying to use the cells as a living chemical factory to replace enzyme deficiencies in suitable patients. Thus, although we had started with the idea of wrapping up islets of Langerhans in a bag of amnion, when we looked at the biochemistry of the cells we found that they were a 'factory' of numerous lysosomal enzymes and it seemed reasonable, therefore, to try the first experiment of implanting the cells directly into the patients who had appropriate deficiencies. This led us to consider patients with Hunter's and Hurler's disease because we know from laboratory work that if one replaces the enzyme the deficient cells can pick it up and utilise it and that in vitro at least a biochemical cure can take place. This idea was confirmed by an experiment where the secretions of amniotic cells were fed to fibroblasts of patients with both Hunter's and Hurler's disease and these fibroblasts were shown to effect a biochemical cure.

A number of children have been transplanted with amniotic tissue and in a number of cases the results have been extremely encouraging with a significant increase in the enzyme levels being achieved. It has been very difficult to be objective about the physical changes in the children so transplanted as I am sure any parent would confirm. When you are looking for an improvement in your child you are grasping at every straw in the hope that you really are seeing some improvement.

Suffice it to say that there is some evidence that some physical improvement did take place in some of the children. It is a shame that a simple idea cannot have a 100% success rate, but nothing is simple in nature.

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Nevertheless, although the early work was not a resounding success, it certainly warrants further study and further transplantations in the hope that we can modify the technique to make it simple and more successful. I do not think there is any doubt that the technique has the same potential for enzymatic improvement as does bone marrow transplantation. Indeed, it has none of the side effects or risks and it has the enormous advantage that no specific donor is required and that the cells last for only a matter of 4-6 months before dying off. If, therefore, a biochemical improvement does not lead to a physical and mental improvement, then it would be a simple matter to stop the transplants, which can be performed by a simple injection as an out-patient procedure every six months, and allow nature to take its course. On the other hand, if a successful bone marrow graft takes, it is there for life and there is a risk that we could be left with a patient who has a long life but no mental improvement worth the trouble.

This seems to be a hard and thoughtless way of looking at the situation, but I think that it is the feeling of most parents who have children with Hurler's and Hunter's diseases and certainly it is a criticism that has been levelled by the medical community. Much work therefore, remains to be undertaken in the field of amniotic tissue transplantation but given that it is theoretically sound, cheap, easily available and readily controlled, surely it justifies at least as much interest and investment in terms of research as does anything else that is going on in this particular field.

Mr. Charles A. Akle, BSc., MB., BS., FRCS. Consultant Surgeon.

BEREAVEMENTS

ELIZABETH PEACH

We learned with great sadness of the unexpected death of Elizabeth Peach, at the Westminster Childrens Hospital on July 9th 1983, following a bone marrow transplant. Our thoughts are with the Peach family at this time.

GARY LOVE

We wish to extend our deepest sympathy to Becky, John and Kerry Ann on the sudden death of Gary on July 2nd 1983. Gary was 8 years old and suffered from Mucolipidosis. We would also like to acknowledge all the donations made to the Society in memory of Gary.

JOHN McCARTHY

It was with great sadness that I learnt from Sheena McCarthy that her husband, John had been killed in a tragic road accident, and I know you would wish to join the Committee in offering our deepest sympathy to Sheena and her two young children, Vicky aged 3 years and 10 year old Scott who suffers from Herler/Scheie disease. They live in Aberdeenshire.

RICHARD'S PROGRESS

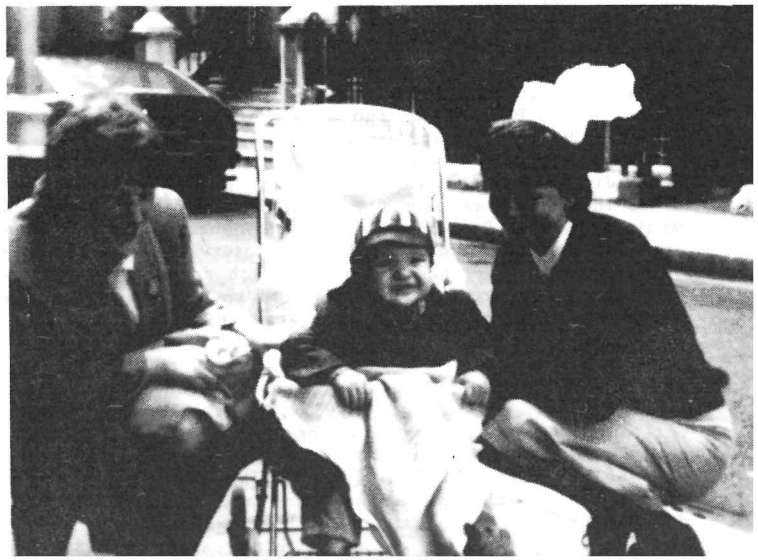
As you may remember, our story appeared in the last Newsletter; regarding our son Richard. We now write again to update you all on his progress. Richard had his Bone Marrow Transplant on the 7th May 1983, using our 9 year old daughter Claire as his donor. (We must emphasise, she was a perfect match sibling donor). We were in Westminster Childrens Hospital altogether for 8 weeks. Richard's treatment began almost immediately on admission (which was 17th April). The care and attention which was given to Richard by both the doctor's and nurses was marvellous and very reassuring during such an anxious time. Both my husband and myself feel very honoured and privileged that our little boy received his treatment at the Westminster Childrens Hospital.

How is Richard now? At present he is very well, indeed he did not suffer any ill-effects from the treatment and has settled down and re-adjusted to home-life very quickly. He has not suffered any more of the nasty viruses, which he seemed to get very frequently.

We now look forward to a much happier future for Richard and hopefully with lots of good stimulus and loving care from us, he will blossom in all aspects. We still live very much from day to day, as one has to remain in a realistic frame of mind, for after all only time will tell.

As a footnote, we would like to thank Christine and the MPS Society, all the families who contacted us, particularly Mary & Colin Gardiner for the valued support and help they gave us in the days of not knowing what was going to happen to Richard. Their kindness and concern shown will always be remembered!

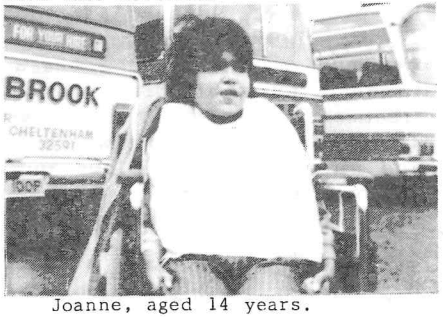
Val Turner



Richard Turner

JOANNE'S SECOND HOME

Our names are Peter and Marlene Sanderson and we come from Gloucestershire. Our first daughter Joanne died last November from Sanfilippo at the age of 16½ years old.



Joanne, aged 14 years.

However, I do not intend to go over all the problems which we had with Joanne, because I know that most of you reading this, have had and are still having, exactly the same traumas that go with an MPS child. What I would like to tell you about is the place where Joanne spent the last four years of her life.

Quite obviously one of the main problems of having an MPS child is finding somewhere to place them to give a break to the parents. We were no exception. Our family consisted of Joanne aged 12 years, Suzanne aged 8½ years and a baby girl Elizabeth aged 4 months. We were a long way from our relations in Lancashire and there was at that time no help available in the area either on National Health or privately. We had ourselves already looked into lots of possibilities. Joanne had already spent a year in a Rudolph Steiner School for the handicapped which was marvellous, but try as they would, she became impossible to cope with and came back home. We spoke to our Social Worker who found that there was a place available in a hospital in Wiltshire but it was the usual sort of institution, housing both very badly disturbed adults and children. Joanne was still at the very hyperactive stage, wrecking everything in her path. We were desperate for a break, took the place and very reluctantly allowed Joanne to stay there in the week, coming home Friday till Monday. However we began to notice that a rapid deterioration in her physical health could take place during the few days that she was there. Joanne was always given wholefood at home but was fed on pureed food in the hospital. From being a very strong, robust little character, Joanne looked like something out of a concentration camp. Again we felt desperate and knew we could not allow her to stay there. However she suffered what seemed like a massive stroke and was rushed into the Princess Margaret Hospital in Swindon. She couldn't walk at all and seemed to lose her sight temporarily. We didn't expect her to survive. However due to marvellous nursing Joanne pulled through. Joanne came home and we begged our G.P. to help find somewhere better and nearer to home for Joanne. He was pessimistic about the chances of finding somewhere but said he would try his best. A few weeks later we received a letter from the Senior Medical Officer in Gloucestershire, who suggested to us that as an experiment, we could try Joanne on the Children's Medical Ward at Standish Hospital, Stonehouse, Glos. We were to go along to see the Sister there for an interview. At our meeting Sister explained that they had never had such a severely handicapped child on the ward before for long stay and she was very apprehensive. However she agreed and we were very relieved.

The hospital itself is situated amongst rolling hills and farmland above Stonehouse and is very beautiful. The children's ward was in another part of the grounds away from the main hospital. In fact a little unit on its own. There is a playground with swings, roundabout, see-saw and the most beautiful real Jet Aeroplane which the children love to play in. There is also a duck-pond and a family of Peacocks, which delight the children constantly. Joanne had a small room to herself and the staff soon took to her. I think partly because of her lovely smile and she became known affectionately as Jo. One of the nurses Daisy loved her dearly and insisted that only she feed Joanne. She treated our daughter just as my own mother used to and who had died a little while before. I think Joanne felt a certain security with Daisy as she had with her Nana. In the end it was only Daisy who could raise that lovely smile, though I must emphasise that the rest of the staff were also very dear and kind to Joanne.

At first Joanne used to go out from Hospital to the The Shrubberies Special School in Stonehouse and everything was tried to stimulate her. Soon the journey to and from school became too much for her to bear and it was decided to keep her in the ward. Actually she was happy just lying in bed or in her wheelchair. It was soon realized by the nurses that the best place for a good old chat was around Joannes bed. The drone of their voices seemed to relax her. The atmosphere was always homely with the other children running around, who incidentally accepted Joanne completely, helping wherever possible. Another thing which was nice was that the nurses were not in uniform, which put the rest of the little patients at their ease.

Although in the four years that Joanne was in Standish we saw lots of new faces; patients and staff alike, some faces never changed. For instance Wally and Samantha, two more long-stay children, with completely different diseases but who are two very important familiar faces. Not least of all the staff - Sister Goodrich, Heather the Nursery Nurse who got Joanne to walk again, if only for a short time. Val, Shirley, Carol, Judy, Mr. Wills and Barbara. Night staff - Nell, Diane, Roz and Carol in particular. They were marvellous. Joanne was never in distress for long, they saw to that. Talking from a laymen's point of view, I am sure her drugs were changed at exactly the right time to suit her constantly changing degrees of illness. We trusted everyone implicitly. On many occasions Joanne could have easily choked to death but thanks to the staff and all the necessary facilities around her she always pulled through. However the main problem which was feeding, became impossible around October of last year and Joanne had to be put on drip-feed. This was the big important step back that we had all dreaded taking, but for Joanne there were no more traumas of choking every meal-time. something which we all had to be grateful for. She lived five weeks after this, most of the time asleep and it was in her sleep that she passed away, very, very peacefully. For that we will be eternally grateful to the most marvellous people in Ward A at Standish Hospital, Stonehouse, Glos.

To conclude, I know that Joanne is still greatly missed on the Ward and I also know that if there are any parents of an MPS child, living as near to Stonehouse as possible and who at this moment might be fighting with their emotions about allowing their child to be looked after in hospital most of the time, they need have no worries about letting them go into Standish. This staff are now well experienced in the nursing of an MPS child through day and night. Indeed - night being the most vulnerable time for any sick person, was the time when we were most grateful to the staff, because on many occasions - at our own request - we were called out in the middle of the night to be with Joanne. I often shudder to think that if she had been at home, perhaps we would have been too tired to deal with her and anything could have happened.

I know that to the majority, it seems a terrible thing to let a child be looked after in hospital, when they already have a loving, caring home but for ourselves, I know that what we did was for the good of the whole family and we felt that nearer the end of Joanne's life, that Ward A and it's staff became just an extension of our own home.

Marlene Sanderson

CHILDREN'S UNIT, Standish Hospital, Stonehouse, Glos.

In the Summer of 1978, a Senior Medical Officer who had close ties with my Ward, asked whether I would consider taking a mentally and physically handicapped child. Joanne was a patient in a large mentally handicapped hospital, outside our Area Health Authority. She had only a limited life expectancy and the parents would be able to visit more often and have her out for days and possibly week-ends if she came to us.

Our ward was a reasonably long stay one but catered for chest and other medical conditions with 4 psychiatric beds. None of us had ever looked after this type of case and we were most reluctant to take Joanne, expecting that this would create a precedent and other's would follow. However, the paediatrician added his persuasive powers and a date was fixed for her admission.

We were then given a diagnosis - Mucopolysaccharide, Sanfilippo type. Fortunately a new text book was delivered to the Medical Library as none of the others made more than a passing reference to the disease. We could obtain no nursing information, nor could anyone predict how the disease would progress and what other handicaps would develop. A "wildly optimistic" guess gave her a life expectancy of 15 years - "with a few miracles".

So on 30th August 1978 Jo, as we all called her, came to Standish Hospital. Her physical state was poor, lots of healed and partially healed bed sores, under-nourished, hair brittle and dull with a bald patch where she rubbed her head. She was doubly incontinent, could not stand, could do nothing for herself. She could not talk but was extremely vocal and one could almost make out swear words when she was cross!

Our first task was to heal her bed sores and improve her physical state. We gave her a normal diet with chopped food (not minced) and she chewed well. We added fresh fruit to her diet with plenty of vegetables and added bran to her cereals. She had an iron deficiency which we corrected and we nursed her on a sheepskin with sheepskin elbow and heel pads. Slowly her hair improved and her skin healed. Her constipation was a thing of the past, although later on she was put on suppositaries twice a week and in the end every other day. She was able to sit on the side of her bed and take a few steps, holding on to a nurses hands. Hydrotherapy began once a week and the Artificial Limb and Appliance centre assessed her needs and provided an excellent 'Avon' chair with a head support either side and slowly her whole conditioin improved.

School was the next target and before long Jo was going from the ward daily by taxi to The Shrubberies, an excellent school for the handicapped. As we grew to know one another, we realized what a definite personality Jo had and we grew to love her. The (so called!) normal children were very protective towards her and they accepted her as part of their every day life.

We had lots of problems but tackled each one as they arose. Her teacher at The Shrubberies, a dedicated young man called Steve was especially helpful as feeding problems started. As nurses we had all been taught to sit in front of our patient when feeding, it being very important to present an unhurried, relaxed atmosphere. Jo merely spat it all back, especially fluids which she disliked but obviously needed. Steve taught us to feed from behind and to get the spoon

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over her tongue, so that she had to swallow. In fact towards the end we would tip the whole chair back on to our lap, so that she was lying flat. As professional nurses we found this a difficult concept at first.

Jo began to develop severe chest infections and in less than 4 hours from the first symptoms, she would be fighting for her life. She was no longer able to stand but the fighting spirit never left her. She had her favourites on the staff, at whom she would beam and dimple but she would play the new students up. Luckily we have a permanent staff who remained fairly constant.

Jo was able at first to come on ward trips - once a month. The sewing room made a large canvas bag to hold all her changes of clothes and cloakroom ladies in many towns were very good to us, as were our coach drivers. The back seat often became Jo's changing room.

Nursing care was constantly reviewed. Respectfully school and trips had to be stopped. A.L.A.C. provided a full length bed/chair which was a great boon. Jo was turned at 2 hourly intervals day & night. She was never left wet - Jo saw to that!. From her admission she never developed another bed-sore. She was tip and topped twice a day by the Ward staff and once by the Physiotherapist. Her swallowing reflex slowly went until at the end she was tube fed.

We celebrated all birthdays but Jo's was always special and her 16th one especially so. She had a super new outfit from Mum and Dad and lots of 'Smellies'.

On the 26th November 1982 on my day off, I had a very sad telephone call, to tell me that Jo had died in her sleep the night before, aged $16\frac{1}{2}$ years old. Her death left a gap in all our lives that will never be filled. Her short life enriched all who knew her. However because of her, two other handicapped children are living a much fuller life here with us, than they could in some large institution. Seven severely handicapped children, all but one mentally as well as physically handicapped, spend holidays and week-ends with us, so helping their families to cope. We watched Jo's small sister grow and her older sister become an attractive young lady and the ward has two wonderful friends in Jo's Mum and Dad but perhaps even more important; in the four years that Jo was with us, countless student nurses and even more children have learnt to accept handicapped children and to love them.

Sister Goodrich
Standish Hospital

AUSTRALIA

LET'S MEET THE AUSTRALIA BRANCH SECRETARY.

Firstly to introduce myself. I am Helen Wheatley 36 and my husband is Phillip 38. We in complete but blissful ignorance, had four beautiful healthy kids, when our fifth and final child Colin was born.

Our story was similar to many I have read in this newsletter. At nine months, Colin was diagnosed as having Hurlers disease. For four years and nine months, we shared Colin's little life, years we would never have been without. Last October Colin's Life ended after a short illness and even now in our sadness, we still thank God for his little life and enormous love.

When we first found out Colin had MPS, we desperately sought information and contact. The information was hard to find and to understand and was often traumatic in its effect. Contact proved impossible until I saw Donna's letter In the Woman's Weekly. We quickly formed our friendship and the idea for a Society was born.

In commencing this society branch, we hope to alleviate some of the loneliness which we felt and to help provide information in a more gentle and understanding way.

In Australia, we inherit the problem of distance, being seperated by many miles but we are fast building bonds with each other and know the help it is to have someone who understands.

We are very thankful for the help we have had from the Doctor's and staff of the Department of Chemical Pathology in Adelaide Children's Hospital. Without their help our society would be much harder to set up.

Helen Wheatley

Dear Readers,

My name is Donna Bryan 23 and my Husband is Barry 29, our only child Rebecca Tarnia is 4 and was diagnosed at 16 months as having Hurlers disease. Becky was born 6 weeks premature, a foot breech and with a cord prolapse, it was picked up at 6 weeks that she had a congenital hip that eventually corrected itself. It took hours to get a small amount of milk into her and she had never ending trouble with colic, as well as suffering with continual E.N.T. infections. We took Becky to many Doctors before she was diagnosed, hoping each time that the new Doctor would finally clear up the continual infections. With every new Doctor there was always the same barrage of questions about her facial features, head and those little fat fingers and each time I would say she looked like me. Finally we took her to a hospital in Queensland with yet another Infection and once again came those barrage of questions but this Doctor came out and said he thought she had Downs Syndrome and after a visit to the paediatrician it all started. Becky went into hospital in Brisbane for 'all those tests' about September 1980 and diagnosis was confirmed in February 1981 at the Royal Childrens Hospital in Melbourne when we went for Genetic Councelling.

Becky Bryan, aged 4 years.

Within 6 months of moving to Tasmania, Becky had an umbilical hernia repaired and got two hearing aids and started going to a special school. Since getting her aids her speech has gone from around 12 words at the most to over 1000 and she is still going full steam ahead. We have only had one major sign of progression of the disease and that was when her liver and spleen had a sudden visible amount of growth in a matter of about 6 weeks. Naturally she is getting stiffer and the features are becoming more 'Hurlers' but mentally she is great.

Near the end of 1981 we decided to have another baby with the intention of terminating if it was affected also with Hurlers. Sadly the amniocentesis results were positive and we had the pregnancy terminated. Six weeks later I fell pregnant again but this pregnancy also resulted in a termination after another positive amniocentesis test. I have no doubt that we will try again but not for quite a while, as neither of us feel strong enough to tackle another pregnancy yet.

At the beginning of 1982, we felt we desperately wanted and needed contact with other families and the only way I could think of doing that was by writing to the Australian Womans Weekly and wrote a letter requesting contact with other MPS families. The response we got to this letter was well beyond anything we imagined. Letters came from as far as Pakistan, The Republic of Nauru, New Zealand and all over Australia. By the way the one from Pakistan wasn't from an MPS family. Many of the letters were from well-wishers and some from families whose children had died as far back as 10 years. In turn I have been able to put families in contact with one another and we have quite a good contact group going already. I will leave it up to Helen Wheatley to explain our intentions about forming an Australian Branch of the British MPS Society.

Donna Bryan

Postscript

Since Donna wrote this article, we have learnt with sadness that Becky had complications with the anaesthetic during the removal of her adenoids and is now on a respirator. Our thoughts are with Barry and Donna at this time.

KNIT ONE, POT ONE, EAT ONE

Our names are Lorraine and Paul Stenson and we have four boys. Jamie 7 years, who has Hurler's disease. Philip 6 years, Anthony 3 years and Russell 9 months. When Jamie was first diagnosed as having Hurler's, I was unaware of how serious the disease was. I lived in a world of my own with Jamie getting worse and changing into a different child for nearly 7 years. Wanting help and not knowing where to turn, until my occupational therapist gave me a slip of paper with the name and address of the MPS Society on it. I 'phoned Christine the same afternoon and my world started to change. Here was someone who had been through the worries and heartache of having a child like Jamie. Christine suggested we should think about having an article on Jamie published in the local paper. At first I thought I didn't want other peoples pity, until one day an old lady said "If he was mine, I wouldn't take him out". That did it, I decided that maybe a few people needed educating on the disease. I had the article done and the response was fantastic. People now come up to talk to Jamie with understanding, even the taxi drivers who have been taking him to school for the last 5 years have changed towards him. The article has brought about a lot of interest in fund-raising as well. Recently a 24 hour pool match was held. Jamie went to see the end and he was given the time of his life. We have been asked to attend a Shredded Wheat eating competition at our Trades and Labour Club and after the competition is over they are holding a disco and band. More than 100 people will be there. People are going all out to help raise money for the Society.

Do you have problems with clothes? That seems to be one of my biggest headaches with Jamie. I have a small sewing machine, so I started making clothes for him. Then came the problem with knit-ware. I wrote to the Joseph Rowntree Fund, asking for help with a knitting machine. They sent a cheque for £100 towards the cost. I have had my machine for about 6 weeks now. I am still trying to alter patterns for him and it is slow going, but we now have a few jumpers that fit everywhere. So if any of you have any ideas, please let me know. When I get styles and fitting correct, I would be more than willing to knit clothes for any MPS children.

Thank you Christine for being my life-line, although things are getting worse, I do not feel alone anymore and thanks for the MPS Society.

Lorraine Stenson

Jamie Stenson, aged 7 years
Picture by courtesy of The Welwyn and Hatfield Times.

NEW FAMILIES

We would like to welcome these families to our Society.

Mr. & Mrs. Astbury whose son James 5 years, suffers from Sanfilippo disease. They live in Staffordshire.

Mr. & Mrs. Bagshaw and their two children, Pamela 2½ years and Paul 5 years. Both the children suffer from Sanfilippo disease and live in Derbyshire.

Mr. & Mrs. Bellman from London. Their 1 year old son, Edward has recently been diagnosed as suffering from Hurler disease.

Mr. & Mrs. Blackburn and their only son Matthew. Matthew suffers from Hunter disease. They live in Cheshire.

Mrs. Elmer and her son Martin. Martin is 20 years old and suffers from Mild Hunter disease. They live in Hertfordshire.

Mr. & Mrs. Fitts and their son Aaron. Aaron is 18 months old and suffers from Hurler disease. Recently Aaron and his mother Maxine made their television debut on ITV's breakfast programme - TV AM. They live in Manchester.

Mr. & Mrs. Green from Lincolnshire. Their 3 year old son, Charles who had Sanfilippo disease, died last September after receiving a Bone Marrow Transplant.

Mr. & Mrs. Greenwood and their daughter Joanne. Joanne suffers from Morquio disease and is 10 years old. They live in Cheshire.

Mr. & Mrs. Ireland from Scotland. Their 10 year old son Iain suffers from Hunter disease.

Mr. & Mrs. Taylor whose son Christopher has Hunter disease. Christopher is 4 years old and they live in Staffordshire.

Mr. & Mrs. Westland and their son Daryl from Berkshire. Daryl is 14 years old and suffers from Sanfilippo disease.

Mr. & Mrs. Wheatley from New South Wales. Their youngest child, Colin who died 6 months ago suffered from Hurler disease. Helen Wheatley has just been elected secretary of the Australian Branch of The Society for Mucopolysaccharide Diseases.

Gill and Eddie Farwell from North Devon. Very recently their 3½ year old daughter Katie was diagnosed as suffering from MPS. Their 4 month old baby son Thomas is awaiting clearance.

Mrs. Darke and her son Ian from Tyne-Wear. Ian is 8 years old and has Hunter disease.

Mr. & Mrs. Smeizers from New Zealand. Their 8 year old son Jeffrey also has Hunter disease.

MORE FUND-RAISING IDEAS

I thought you might be interested to hear what fun it has been fundraising for MPS. My escapades started in late April when I was on our Local Radio Station, publicising the sponsored parachute jump by David Bower, a ladies hairdresser from Amersham in Buckinghamshire. Unfortunately I didn't get much response, although I did raise £30 in sponsorship overall.

I wrote to the radio station a week before and they telephoned me asking me to do a radio phone in interview. I was asked all about the disease and our Society and where the money was going. It was quite an experience. At first I was on tender hooks waiting for the phone to ring but funnily enough, once we got talking I was quite relaxed.

On May 17th I held a clothes party and sold over £114 in clothes and have donated the 10% commission to the Society. Soon after my husband, Pete came home with the news that two friends from work, John Flynn and Clive Taylor were running in the Swindon Marathon. Both men completed the 26 miles and as a result they raised £75 for MPS and £50 for the Institute of Child Health at Great Ormond Street.

Recently we have decided to close our daughter, Toni's fund and split it down the middle, giving half to The Institute of Child Health and the rest to MPS. This will enable us to concentrate fully on helping MPS.

If any families have local radio stations why not get in touch with them and tell them all about MPS. I wish you all luck and hope we can raise money to further research and purchase a holiday caravan.

Sue and Pete Bramford
196 Ferndale Road, Swindon Wilts., SN2 1MP.

Dear Readers,

My name is Anne Palmer and my husband's name is Mick. We have two children, Jane who is a normal 11 year old and Neil aged 13 years who suffers from Sanfilippo disease.

When Neil was diagnosed at 3 years old it was very hard for us to accept because apart from his inability to string words together , he appeared to be quite normal, in fact quite an active and robust toddler. Gradually over the years he began to deteriorate until now he is unable to walk unaided , cannot talk and has to have everything done for him. He also has to have his food liquidised. Nevertheless throughout all of this Neil has remained a very plensant child. He suffers from fits which we have learned to cope with and each year we manage to take Neil away for a short holiday. Recently Neil has been going to Ryegate, whilst we have a weekend break and so enable us to devote more time to Jane.

We consider ourselves very lucky to have such a lovely daughter. who is so understanding to Neil. She never moans about him and encourages all her friends to accept him and become his friend too.

We are very pleased about the Society and hope it continues to expand.

Anne Palmer

DOUBLE TROUBLE

Our names are Helen and Peter Rock and we have two children, Lorraine 8 years and Christopher 6 years who suffer from Morquio-Brailsford disease.

We first noticed with Lorraine at the approximate age of twelve months that she had a curvature of the spine and sternum. We took her to see a paediatrician who, at the time, did not know what she was suffering from. He had made a cloth corset with metal stays, to support her spine. Weekly visits to the physiotherapist involved a lot of spine stretching. At a much later date physiotherapy was discontinued as no real benefit would come of this. Lorraine also developed a hernia which was surgically repaired, at the age of three. Little did we know that this was just the beginning of our problems.

Just before Christopher was born in May 1977, we were told that Lorraine was suffering from Morquio-Brailsford disease. At six weeks old Christopher was examined and thought to be normal; however, at the age of twelve months he showed signs of a curved spine and sternum and was also diagnosed as suffering from Morquio-Brailsford disease.

It was late 1979 when we began to realize that it was not just the children's spines and sternums which were affected but also their legs, feet, hands, ribcage and most of the bone joints. Enlarged abdomens, were also noticeable at this stage. Normal growth is retarded so they will remain small but in proportion. They have had several operations on their legs to aid their walking. Calipers have been of help; Christopher still has to wear his. A noticeable hearing loss was detected with both children. The E.N.T. specialist has helped the children by having their tonsils and adenoids removed and the ears drained and grommets inserted. They have had two sets of these inserted and are due for more later this year. This has made things better for them but they will always have a hearing loss.

The children attend normal school where they are very popular despite being unable to join in most activities for any length of time. However, they are very cheerful children and keep me on my toes. Lorraine is very interested in books and is always reading. Christopher's main interest is in cars and lorries - the more the merrier. We have learned in the last nine months that a bone marrow transplant is a possibility for M.P.S. children and we hope that one day all children with M.P.S. will have the opportunity to have a normal life although sadly only a few will benefit at present.

Mrs. H. Rock

Lorraine, aged 8 years and Christopher, aged 6 years.

Dear Readers,

I would like to tell you about my son who suffers from Hunter Syndrome. He is now 20 years old. Unfortunately the condition is inherited from my side of the family, but even with this in mind Martin was not diagnosed until he was eight years old because he was so different from previous sufferers in the family.

He had a double hernia repaired at six months old, tonsils and adenoids removed at four years old because of persistent ear abcesses and grommets inserted for the first time at five years old which brought his hearing level up considerably, but even then his condition was not diagnosed. It really was not noticeable as a baby or indeed prior to the condition being confirmed. Facially it just did not show and photos we have can prove it. When people who did not know him as a baby see these photos now they cannot believe it. We are only grateful for the first few happy years.

He was checked at fifteen months for deafness as I was not happy about him, but we were laughed at. It was not until we saw a paediatrician just before his third birthday because he was not talking that he referred us to another Hearing Specialist who pronounced him severely deaf. He got his hearing aid just after his third birthday and as soon as he had this he started talking and went on to school in the normal way - a private school because it was suggested that he should attend a school with smaller classes than average and we were very lucky in that the County Council paid for him. He used to play football and is still an avid Chelsea supporter. His hobby is football and he knows everything there is to know about players and teams etc., including players names, what teams they have played for and how long they have been with their respective Clubs.

At the age of eleven when he was due to go to secondary school it was then realised that he would not cope in a Comprehensive school so he attended a local school which caters for children with varying handicaps and difficulties.

When he came to leave school it really was a problem of what he would do and where he would go, but fortunately Papworth was suggested and even more fortunate he was accepted, as we had been warned that he could be turned down. He has been there for almost four years now and loves every minute of it. He earns a small wage, has his own room which is like a second home as he is very very tidy, and he comes home every three to four weeks.

He is definitely deteriorating now. His hands are very stiff and his handwriting is not as good as it was. His breathing is much more noticeable. He doesn't like to walk far, so much so that at Papworth they have provided him with a battery operated chair which he rides about in, but all in all he is still very cheerful and very lovable and affectionate.

Of course, at the time Martin was born there was no treatment available, or indeed, no one to talk to prior to going in for a family. We feel now that there is little chance of help for him mainly because of his age and also because of the limited treatment available for the number of children who are young and would benefit from the treatment.

Beryl Elmar
Hitchin, Herts.

AREA SUPPORT FAMILIES:

Colin and Mary Gardiner; 35 Church Road, Banks, Southport, Merseyside.
Derek and Pat Kirkman; 63 Higher Shady Lane, Bolton, Lancs.
Robin and Ann Ridley; 2 George Road, Lutterworth, Leics.
Neil and Jane Reid; "Meadowlark", 9 Huddleston Way, Sawston, Cambs.
Robin and Christine Lavery; 30 Westwood Drive, Little Chalfont, Bucks.
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 Diseases.
3. To raise funds in order to further research into MPS.

SECRETARY'S REPORT

Here in Little Chalfont The Society's work does not let up and we are still continuing to have a steady trickle of new members. Doing this job one is rarely surprised by who is at the other end of the telephone, but as you can imagine, it was hard to disguise my amazement when about 7 weeks ago, just as I was resigning myself to the inevitable chores that go with Monday mornings, the 'phone rang and it was a call from The Princess of Wales' private office. As a result of a letter written by one of our families, The Princess of Wales was sufficiently interested in the work of the Society to request further information. A package was promptly sent to Buckingham Palace. The Society has now officially invited Her Royal Highness to become Patron and our request will be considered when Her Royal Highness next considers requests for patronage, most likely at the end of 1983. As you can imagine Her Royal Highness receives numerous similar requests compared with the small number of patronages she can accept. In the meantime we are very grateful to The Princess of Wales for her kind donation from the 'Princess of Wales' Wedding Dress Charities Trust, which will go towards the cost of the Family Weekend/Conference.

By the time you read this, those families who expressed a desire to attend the Family weekend will have heard from Catherine Grant our Liaison Officer, concerning the special needs of each family. We will also have visited Birmingham and made a final decision on the venue. There is an awful lot of work involved in arranging such an event and not least is the problem of finance. For some time various members of the committee and families have been churning letters out in earnest to attract sponsorship and gradually we can see the budget will be met by donations and sponsorship specifically for the weekend. If anyone feels that they can help either because they work for a company that might consider sponsoring this type of venture or feels they want to hold a fund-raising event in aid of the weekend, please do let me know.

Recently our Charity Registration was granted and you will note our registration number is displayed on our new Information Sheet and Newsletter.

Several families let us know that they were unhappy at the use of the word 'Gargoylism', when describing features of MPS diseases, in particular Hurler disease. The Committee agreed that all mention of 'Gargoylism' should be deleted from official MPS literature and we are very grateful to members of the Medical profession who have helped to improve the accuracy of the new information sheet.

The Committee of the Society have had to say goodbye to Mary Hourigan for personal reasons. We would very much like to thank Mary for her help and support in the initial stages of setting up MPS. We are glad to say she will continue to be available to comment and advise on Special School Education.

We are very pleased to welcome to the Committee Peter Archard and Sue Butler. Peter and his wife Maggie have two sons, the elder of whom suffers from Hunter disease and live in Letchworth. Sue and her husband also have two children, a daughter aged 7 years and Alexander 10 years who has mild Hunter disease. They live in Chinnor. Behind the scenes both these families have already made a considerable contribution to the workings of the Society. Lastly, but by no means least, we welcome an additional area family. Marlene and Peter Sanderson whose eldest daughter Joanne died last November from Sanfilippo disease and who will be available to talk to and meet new families as well as co-ordinate fund-raising in the Gloucester/Avon, South Wales area.

Christine Lavery.

WE WISH TO THANK THE FOLLOWING PEOPLE FOR THEIR GENEROUS DONATIONS

Kingsthorpe Upper School Playgroup.
Mrs. Sweasey
Lady Lidderdale
Mrs. T. Vipond
Mrs. E. Sanderson
Miss A. Marshall

Mrs. Pat Miller
Local Bakery, Blackpool
Mr. Seary, Hampstead
Mrs. Barrand, Blackpool
Asquith Designs

The Mayor of Worthing
Mrs. Moon
Sawston Free Church Playgroup
Neighbourhood Committee, Marlow Methodist Church
Chemical Pathology Dept., Adelaide Children's Hospital
Mrs. Walters
Flaky Pastry Pop Group
Silver Wings Catering
Mr. & Mrs. Palmer
Mrs. Twigger

Our thanks also go to:- Mr. B. Coole, Clifton LC. and Baines Endowed C of E School who held fund-raising events after reading about Samantha Walker from Blackpool, who has Scheie Disease.

Kirkham Carr Hill County High School who held sponsored events.

Mr. Michael Burchell who made a donation in Memory of John McCarthy.

Mrs. Appleton and Little Kingshill Morning WI who held Coffee Mornings.

Mrs. Jane Reid, who held a garage sale and made flowers to sell.

Amersham College of Further Education, who held a Charity Day in aid of MPS.

Mrs. Yvonne Fuller from Inverurie Aberdeeenshire, who did a sponsored walk and made a donation to MPS.

Mrs. Susan Lee's Dance School, Blackpool, who held a Charity Concert.

Mrs. Walker who held a Tupperware Party.

Friends of Grahame and Samantha Walker, who collected ½p's.

Mrs. Ross from Welwyn Garden City, who held a Jumble Sale after reading an article about Jamie Stenson in their local paper.

We would also like to thank the following who have specifically donated money to the Family Weekend:-

Saab (Great Britain) Ltd.
Mr. & Mrs. Hodges from Harby, Leics.
National Panasonic (U.K.) Ltd.
BBC 'Children in need Fund'.
The Princess of Wales Charities Trust.
Roussel Laboratories.
Boehninger Ingleheim Ltd.
Tricentrol (McMinn).

CBS Records and Alberto Culver have kindly given some of their products to help raise money for the Weekend.