Newsletter

The Society for Mucopolysaccharide Diseases

National Registered Charity No.287034

Winter 96

Our Home Page on the World Wide Web : http://www.vois.org.uk/mps

* Address may have changed by the time you receive this newsletter.

The Society for Mucopolysaccharide Diseases

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The MPS Society is a voluntary support group, founded in 1982, which represents over 800 families in the UK with children or adults suffering from Mucopolysaccharide and related diseases. It is a registered charity, entirely supported by voluntary donations and fund-raising, and run by the members themselves. Its aims are as follows:-

To act as a parent support group
To bring about more public awareness of MPS
To promote and support research into MPS

The Society operates a network of Area Families throughout Great Britain and Northern Ireland, who offer support and links to families in their areas. It provides an information service for families and professionals. At the present time it supports two specialist MPS clinics at the Royal Manchester Children's Hospital and at the Hospital for Sick Children, Great Ormond Street, London. The Society also funds research projects at the Christie Hospital, Manchester, Royal Manchester Children's Hospital, Bristol Children's Hospital and the Institute of Child Health, London. It encourages and assists contact and co-operation between parents and professionals and maintains links with sister societies in Europe and throughout the world.

There is at present no cure for MPS diseases, but much can be done to improve the treatment and care of sufferers. The slogan of the Society is:-

"CARE TODAY, HOPE TOMORROW"

Front Cover:
Roseena Ismail aged 8 years (Maroteaux/Lamy)
Miriam Saud aged 10 years (Morquio) with her sister, Attia.

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Deadline for the 1997 Spring Newsletter
20th March 1997

Please send us lots of photos

DIRECTOR'S REPORT

On behalf of Mary and Pam in the Northern MPS office and Joan, Sheila, Sue and I at the Amersham MPS office we would like to express our heartfelt appreciation for the hundreds of Christmas and New Year greetings received.

We know and understand that for each and everyone of you this time in particular brings to the surface a mixture of emotions. We were only 30 hours into the New Year when 3 year old James Edwards (Hunter) chose his time to leave this earth. Our thoughts are with his Mum, Jackie , family and friends at this sad time.

Many of you who telephone the Amersham office or have attended the conference will have spoken to Sue Balmforth. We all said our farewells to Sue at Christmas after 3 years of working for the Society. Sue with her son George has now joined her husband, Mike to live in Munich. Sue loved her work with the Society and we will miss her very much. Sheila who job shared with Sue is for the next few months working full-time for the Society.

As you will read in the Chairman's Report the Society has said goodbye to several long serving Trustees. I would also like to express my gratitude to Alf King and Pauline and Sean Mahon for their support and contribution to the progress of the Society over the years.

I would also like to welcome the newly elected Trustees, Alison Pullin, Vic Lowry and Sarah Long. Sarah is the first person to be appointed who suffers from MPS (Morquio). I also look forward to working with our new Chairman, Paul Leonard and Lynne Grandidge who has taken over as Treasurer.

In the Autumn I applied for four grants under the National Lottery Charities Board. Just before Christmas we learnt that our applications to develop our support networks in Wales, Scotland and Northern Ireland have been successful. Regrettably the application for England for two advocacy posts (one for ethnic minority families) was not successful. What are the implications for the Society? It means that we, staff, Trustees, families and supporters are going to have to work harder at raising funds if we are to improve our advocacy services to MPS families. For Mary, myself, Joan, Sheila and Pam, we will continue to try and respond to families as we always have.

There are few tasks more pleasurable for a charity Director than informing succesful grant applicants. Following Peer Review the Trustees agreed to fund the following projects.

1. **Dr E Wraith - A Proposal to Support the Clinical Development of Gene Therapy** ***Year 1 - £41,100***

2. **Dr B Winchester - Mutation Analysis in Patients with Mucopolysaccharidoses Types I, IIIA and IIIB as a Prerequisite for Gene Therapy or Enzyme Replacement Therapy**. ***Year 1 - £36,797***

3. **Dr L Lashford - Optimising Retroviral Vectors for Gene Delivery to the Haemopoeitic System .** ***Year 1 - £27,188***

DIRECTOR'S REPORT

4. **Dr B Winchester - Experimental Gene Therapy for Fucosidosis.**
   Year 1 - £5,000

5. **Dr C Pennock - To Identify Mutations at the Iduronidase Gene.**
   Year 1 - £2,500

All these projects are being funded with monies raised for research and the Jeans for Genes Appeal.

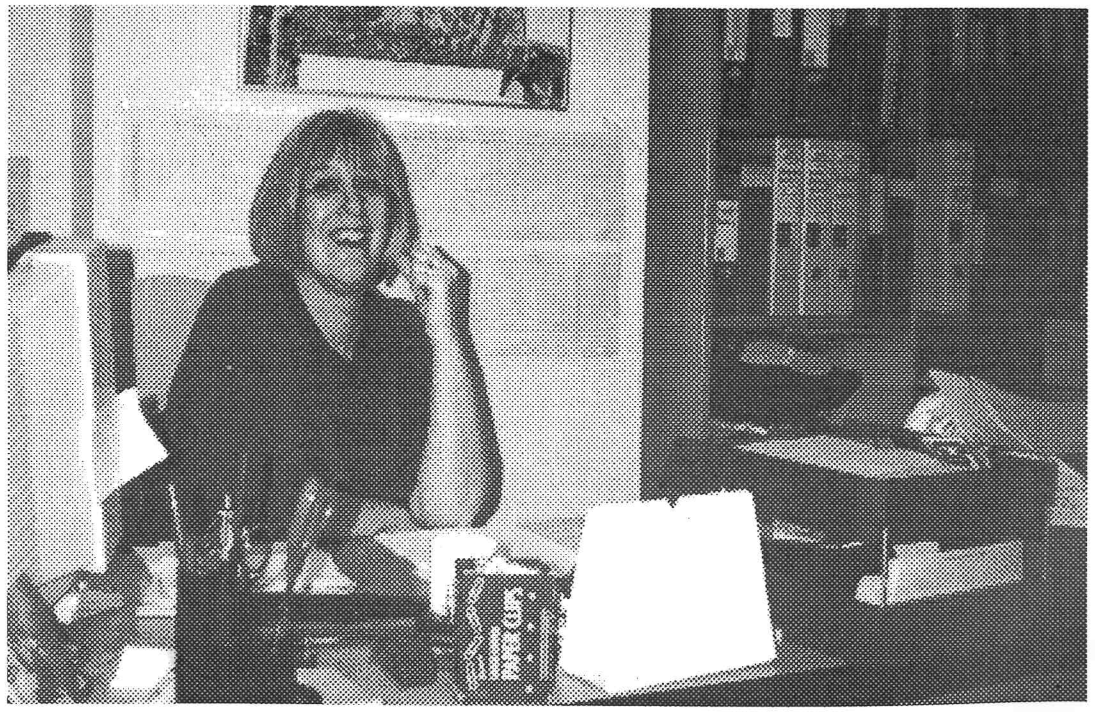
Enclosed with this Newsletter are application forms for the MPS Annual Weekend Conference, MPS Family Holiday and the MPS Teenage/ Adult Activity Holiday. Take up for all these events is expected to be high so please do apply early so as not to be disappointed.

We should also be very grateful if you would complete the questionnaire seeking confirmation of your correct address and telephone number. In some areas of the country the county has changed to a unitary authority and many of you have changed you telephone numbers. Even if there are no changes please complete the form.

In a majority of families, grandparents, aunts, uncles, brothers, sisters and friends play a big part in the lives of MPS children. We are offering the MPS Newsletter to these special people at a first year annual subscription of £7.50 (UK only). Please do pass this information to interested friends and relatives in your family. The forms can be photocopied or ask the MPS office for more.

Wishing each and everyone of our readers a peaceful New Year.

Christine Lavery
Director



Pictured here is Sue Balmforth whose happy smile we miss already.

CHAIRMAN'S REPORT

At the last Trustees meeting, I agreed to be the new MPS Chairman. Having been helped by many people who support the aims of the society, I wished to try and repay my debt of gratitude. Both my two children were diagnosed with an MPS disease and so I know some of the problems and challenges that need to be faced. Here are my initial thoughts about what has recently happened or is happening to some of the Trustees and the MPS Society: -

Alf King
Alf has been a leading light in the MPS Society since its early days. As Chairman, he has guided the Society through many changes and been an enthusiast of the Area Training Weekends. Having taken early retirement from his employer's, the Inland Revenue, it must be right that he should spend some time with his wife, Judy while they catch up with the sight seeing and visiting - the way I'm sure, many of us always promise to do. As a Trustee during the last twelve months, I was always struck during Trustee meetings by the way that Alf wanted to hear the views of all the Trustees and to do his best for MPS. I wish him and Judy, all the best for the future.

The MPS Accounts
I also want to take the opportunity of thanking Pauline and Sean Mahon for their efforts of dealing with the MPS Accounts. This is probably the most difficult job that Trustees can undertake and so I offer a big THANK YOU to them too and wish them well. Their expertise will be greatly missed as they have understood how to maximise our income and spend the money wisely. I hope they will be able to support the Society in other ways.

Lynne Grandidge has taken over the daunting task of Treasurer. I'm sure with the help of MPS staff, Trustees and members, she'll be very good.

The future of MPS
Before I offered to help the society as Chairman, I visited the main MPS Office at Amersham to learn more about the job. I was very impressed with the dedication of all the staff and amazed at their workload. Over the last three years, the society has grown enormously. In 1993, there were 650 families and in 1996 over 800 families. While the aims of MPS continue to be appropriate, the balance and amount of the work has changed enormously. Fund raising has been hit by the National Lottery and the number of newly diagnosed MPS cases is currently running at one per week.

Your Director, Christine Lavery, has been very good at obtaining grants to help specific families, both Christine and Mary Pagett, in the MPS Northern Office put in a large number of hours at the medical clinics and on home visits helping MPS suffers and their families. Ideally, the Society would benefit from greater resources both financially and from additional staff. At the Trustees meeting in March, one of the items I hope we will be discussing will be 'Beyond 2000 - A Strategy for the Future'. If you feel strongly about how you think MPS should be operating in the future, please write to me or Christine Lavery, at the MPS Office. Your views will be most welcome and could help in the formulation of our priorities.

I look forward to serving you.

Paul Leonard
Chairman

MILESTONES

New Families

Mr and Mrs Shields from County Down whose son Kyle aged 4 years old has recently been diagnosed with Sanfilippo Disease.

Mr and Mrs Fasey from Long Eaton whose daughter, Megan aged 2 years has recently been diagnosed with Hurler Disease.

Mr and Mrs Drayne from Lisburn whose daughter Roma aged 4 years old has recently been diagnosed with Morquio Disease.

Mr and Mrs Smeaton from West Yorkshire whose 11 year old son Christopher has been diagnosed with Morquio Disease.

Mr and Mrs Devine from Coventry whose daughter Katie, aged 14 months has recently been diagnosed with Hurler Disease.

Mr and Mrs Browning from St Albans whose 9 month old son, Lewis has been diagnosed with Hurler Disease.

Deaths

Sadly Michelle Brennan's 6 year old daughter, Kellie died on the 18th October 1996. Kellie who lived in Huddersfield suffered from MLII.

Sadly Mr and Mrs Obad's son, Ahmed, aged 6 years old died on the 1st of December 1996. Obad suffered from Hunter Disease lived in Liverpool.

Val and Dennis Mort's son, Richard died on the 25th of November 1996. Fifteen year old Richard from Swansea suffered from Sanfilippo.

Suddenly on the 2nd of January 1997 Jackie Edward's' son James died. James from Cardiff was 4 years old and suffered from Hunter Disease.

Change of Name

Thomas and Joe Fuzzard will be known in the future as Thomas Fuzzard-Tucker and Joe Tucker.

Congratulations

Many happy returns of the day on the 2nd December to Bill Blackburn who was sixty. Hope you had a lovely day, Bill. Bill and his wife Sylvia have been an area family and the society is grateful for all the work they do.

AREA FAMILY SUPPORT

Dawn and Ted Nelson kindly allowed the use of their garden for the South East Area Family Barbecue on the 22nd of June 1996.

Six families attended the barbecue and they had a wonderful day which ended with strawberries and cream.

Pictured below are some of the families who attended the barbecue.

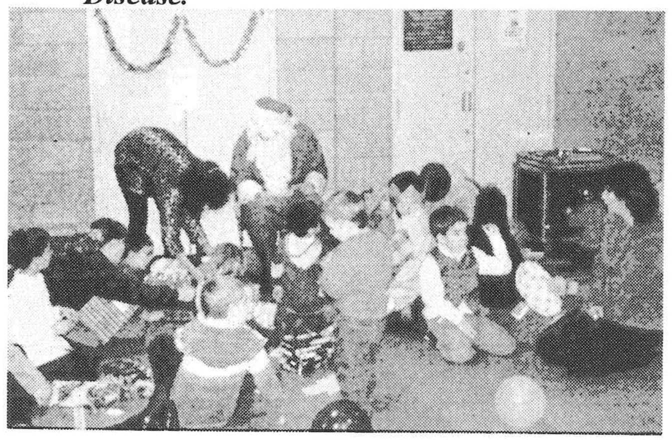
AREA FAMILY NEWS

MPS London and the Home Counties Xmas Party - 1st December 1996.

On Sunday, December the first, the London and Home Counties Xmas Party took place at the Loddon Leisure Centre, Reading. Gavin, Denise, Rachel and I had only had two months to arrange everything and hope it was not too obvious to thosewho attended. Due to the short period in which to organise the event we were very pleased with the amount of people who managed to make it to the party.

Pictured above is Jessica Stuart with her sister, Annie. Jessica is nearly six years old and suffers from Hurler Disease.

The party started at 2pm and we had a visit from Uncle Charlie the ***Children's*** Entertainer, although it was there for all to see that the adults were also entertained. Father Christmas also paid a visit to the delight of the children, I would like to thank Father Christmas for coming along as he appears to have lost some weight and is obviously under a great deal of stress. There was also a raffle which raised £90.00 with prizes going to Jenny Broom, Denise Brown and Peter Stuart.



Pictured above and below are some of the children with their gifts.

The children's gifts were very kindly donated by Mattell, Knex and Tomy we also received support from Walkers Crisps, Mars and Savacentre. It was great fun arranging this event and hopefully we can make it more fun next year. We are planning a BBQ this summer and we will inform all members of the date when this has been arranged. We would however appreciate our members input, so if you have any ideas please let us know.

Hope you all enjoyed the day as much as we did and hope to see you all again soon.

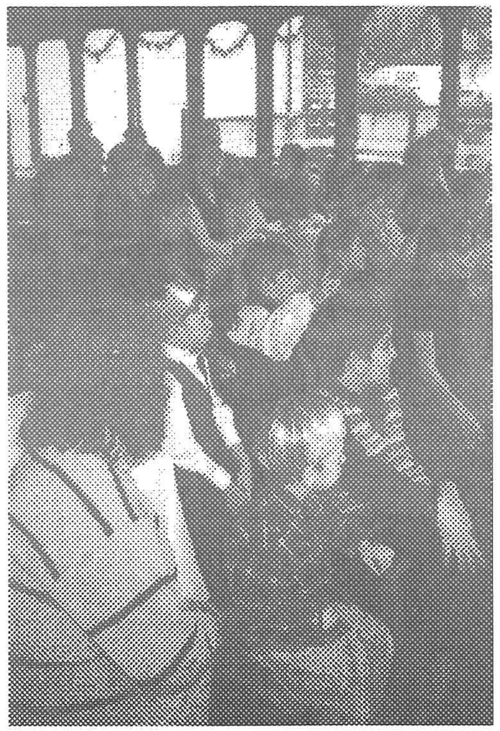
MARK WHEELER

AREA FAMILY NEWS

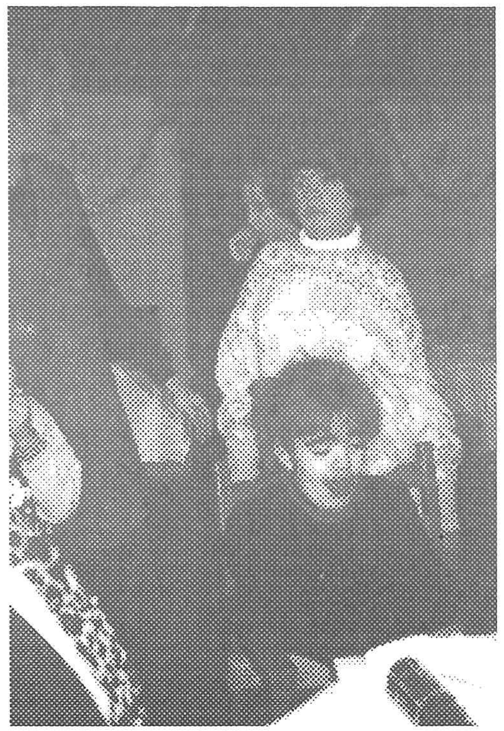
Pictured above are Josephine and Francesca Kembrey, aged 5 years old (Sanfilippo). Hannah Chisling aged 4 years (Sanfilippo) and Jamie and Jason George aged 7 years (Sanfilippo).

South West Christmas Party - 2nd December 1996

This Christmas Party was held at the Stakis Hotel in Bath where all the families had a lovely day. The day was the first event organised by Fer Pidden and Jackie Chisling and we are sure that all the families who attended appreciated their efforts.



Families enjoying the entertainment.



Louise Hill and Natalie Pidden who are both teenagers who suffer from Sanfilippo Disease.

INFORMATION

SLEEP PROBLEMS IN CHILDREN WIIH SANFILIPPO SYNDROME

Gillian A Colville, John P Watters, William Yule and Martin Bax

Sanfilippo syndrome is a rare inherited lysosomal storage disorder, first described by Sanfilippo et al. (1963). It belongs to a group of disorders called the Mucopolysaccharidoses, in which there is a deficiency of specific enzymes involved in the degradation of Mucopolysaccharides. At present there is no cure. Clinically, Sanfilippo syndrome is characterised by progressive mental degeneration leading eventually to death usually in the second or third decade of life, after two or more years of normal development. Somatic manifestations are considerably fewer than in the other Mucopolysaccharidoses - there is, for example, no corneal clouding. although hair and facial features become increasingly coarse. There are four recognised subtypes of the condition (A. B. C and D), and the highest overall incidence reported is I in 24,000 (van de Kamp er al. 1981).

As well as the physical changes, behavioural changes and sleep disturbances have been reported. Sleep problems are not unusual in young children (14% of 3-year-olds in a large epidemiological survey were found to have some kind of sleep disturbance (Richman et al. 1975). but the prevalence of sleep problems in children with mental disabilities appears to be higher. Clements et al. (1986) reported a rate of 33% for a group of
O to 15 year old children with mental retardation and noted a strong association with difficult behaviours occurring during the day. The rates in specific syndromes. including Sanfilippo syndrome, appear to be even higher. (Table 1)

According to Bax and Colville (1995). the prevalence of sleep problems in the Mucopolysaccharidoses is high (71%) and for Sanfilippo syndrome alone still higher (87%). Both van de Kamp (1981) and Nidifer and Kelly (1983) also reported high rates (56% and 86%, respectively). Other authors have noted that sleep difficulties can be a characteristic presenting sign (Leroy and Crocker 1966, Spranger 1972).
The aims of this study were to gather detailed information on the incidence, nature and development of sleep problems in Sanfilippo syndrome, by means of a parental questionnaire, and to explore the possibility that there may be differences in the sleep behaviour profiles of the four main subtypes of Sanfilippo syndrome.
A smaller group was investigated in more detail in order to establish whether standard behavioural techniques such as those commonly used with under 5's by psychologists and health visitors in primary health care settings could help to reduce the heavy burden on these families.

INFORMATION

TABLE I
Sleep problems in children with disabilities

Type of Disability	Percentage with sleep problems	Reference
Severe retardation	33	Clements et al.(1986)
Infantile hypercalcamia	46	Udwin et al. (1988)
Rett syndrome	74	Coleman et al. (1988)
Sanfilippo syndrome	87	Bax and Coleville (1995)
Sanfilippo syndrome	86	Nidifer and Kelly (1983)
Sanfilippo syndrome	56	Van de Kamp (1981)

Method

Subjects were recruited through the Society for Mucopolysaccharide Diseases, a parent support group set up in 1982 in the UK to increase awareness of these conditions and to raise money for research.
Questionnaires were distributed to families both in the UK and abroad. The response rate, which was only available for the UK cases. was 75%. A total of 80 cases were surveyed. In 58 of these there was only one affected child in the family, but there were seven families containing two affected children and one family with three children with the same condition (this information was not available for five cases). The subjects' mean age was 10 years 2 months (range 4.4 to 25.6. SD 4.9). Further demographic information is supplied in Table II.

Parental Questionnaire

A questionnaire, based on the Great Ormond Street sleep questionnaire (Douglas and Richman 1982), was adapted to include questions relating to particular aspects of Sanfilippo syndrome such as epilepsy and current medication.

TABLE II
Demographic characteristics of sample

	N	(%)
Sex		
Male	40	(50)
Female	40	(50)
Specific enzyme subtype (N=63)		
A	43	(68)
B	17	(27)
C	2	(3)
D	1	(2)
Country of origin (N=80)		
UK	38	(48)
Other European country	30	(38)
USA	9	(3)
Australia		

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Results

Current Problems

Sixty-two of the children surveyed were reported to have sleep problems (78%). and in nearly half of these cases (N=29) the problems were described as severe. Given the mean age of the sample, a surprisingly high number of children were sleeping in their parents' room (N=19). However, it seems likely that these sleeping arrangements came about through the necessity for close supervision. As Table III shows, these children exhibited a number of unusual behaviours at night, with major implications for management - particularly where other members of the family were regularly having their sleep disturbed.

TABLE III

Types of sleep difficulty and night behaviours

	N	(%)
Settling difficulties	45	(56)
Night walking	47	(59)
Early waking	22	(28)
Sometimes awake all night	36	(45)
Crying out	30	(38)
Wandering around house	30	(38)
Entering parents' bed	24	(30)
Talking in sleep	18	(23)
Body rocking	14	(18)
Chewing bedclothes	20	(25)

TABLE IV

Comparison between Sanfilippo types A and B

	Type A (N=43)	Type B (N=17)	p*
Sleep problems	33	13	NS
Settling difficulties	23	10	NS
Night waking	24	13	NS
Early waking	8	10	<0.02
Number of night wakings			
0-1	13	0	
2-4	30	17	<0.05
Number of nights affected weekly			
0-1	16	2	
4-7	27	13	NS

*NS = not significant. *Two-tailed X. incorporating Yates correction.*

Some parents spontaneously described their children singing (N=9) and laughing (N=3). Several children made a considerable amount of noise on waking in the night and one little girl had actually started two fires by playing with gas appliances.

Changes Over Time

Although the vast majority of children were said to have had a sleep problem in the past (92%), in only nine cases was it felt to have always been a difficulty.

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Age was not significantly associated with number of hours asleep, number of night wakings or a general measure of sleep difficulties, although in some individual cases parental reports did suggest changes over time.

Some parents felt that there was a temporal relationship between sleep problems and epileptic fits, in that the child's sleep was reported to become more disturbed a day or so before an attack. There was no significant association between presence of epilepsy and overall sleep difficulty.

Comparisons Between Subtypes A and B

Given the small numbers of children with Sanfilippo types C and D. it was only possible to examine differences in sleep pattern between those with types A and B. Although there was no difference in the level of reported sleep problems in the two groups. children with type B syndrome had more severe problems (Table IV). In particular the rates of early waking and night waking were higher.

Coping with Sleep Problems

Two-thirds of parents had given their children prescribed sleep-inducing medication at some stage (N=51), but with mixed effects. In 22 cases the medicine was thought to have been useful as a last resort, for short periods and usually at times of severe disruption. Other parents. However, discontinued drug use because of alarm at side-effects. These were sometimes paradoxical in that the child's arousal level seemed to be increased, but some parents were also concerned if their child became subdued on medication.

The night-time behaviours outlined above were described as very disruptive. In 36 cases parents felt they were not getting enough sleep themselves, and 28 families reported that siblings' sleep was suffering too. Several parents, when asked what changes they would like to see in their child's sleep pattern, commented that they wished their child would sleep in their own room.

Intervention

Five families were recruited for a brief behavioural intervention This involved the collection of baseline data on the child's sleep pattern for two weeks. and home visits by a clinical psychologist before and during the intervention period to negotiate the treatment plan. Weekly telephone contact was maintained throughout the six-week treatment phase, during which families continued to keep records. Follow-up data were collected 1 month after the intervention had started. A summary of the results are given in Table V and a case history is described over. A detailed description of the intervention is available elsewhere (Watters 1988).

In all but one of the five single-case interventions, clinically significant improvements were made during the treatment phase. Follow-up data available for three cases, indicated that there was some reduction in the extent of improvement over time, but in each case a degree of improvement was maintained without any further intervention.

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TABLE V
Results of Brief Intervention

Case	Sex	Age (yrs:Mths)	Sanfilippo Type	Treatment Goals	Goals achieved	Goals maintained at follow-up
1	F	6:10	A	1: Reduce settling time.	Yes	No
				2: Decrease number of night wakings	Yes	No
2	F	5:1	A	1: Decrease rate of early waking	Yes	No
				2:Reduce lengths of night waking	Yes	Yes
3	M	7:8	A	1:Reduce number of night wakings	Yes	Yes
				2: Reduce disruption at night	Yes	Yes
4	M	6:7	B	1:Reduce settling time	No	NA
				2: Decrease number of night wakings	No	NA
5	F	6:0	A	1: Decrease number of night wakings	No	NA
				2: Reduce settling time	Yes	NA
				3: Move back into own room	Yes	NA

Case History

Case 2, was in a girl aged 5 years and 1month with Sanfilippo type A. Two aspects of her sleep were causing disruption in the Family, particularly to her sibling with whom she shared a room. The first of these was early waking, and the second was noisy and disruptive behaviour on waking during the night. The parents reported that their daughter would sit on her bed talking to imaginary people, laughing, giggling and reciting nursery rhymes. She would sometimes wander round the house at night, turning on taps and disturbing her parents.

The planned intervention agreed was as follows: (1) a regular bedtime of 8p.m. was established, at which time the usual night routine (which included a story and a drink) was instituted. (2) On waking at night, the child was to be put back to bed firmly and immediately with the minimum of interaction, although she was allowed toys in her bed if she wished. (3) She was allowed up for the day only after 6a.m.

The frequency of early waking was reduced in the treatment period, although this was not maintained on follow-up (Fig. 1). However, the length of night wakings (which numbered on average one or two a night throughout the period records were kept), diminished from an average of over 40 minutes to under 15 minutes. This effect was maintained at follow-up some four months later. In summary, although the treatment goal of reducing the number of night wakings and early morning wakings was not successful overall, there were important positive gains in terms of lengths of nightwaking episodes and also in settling, incidentally, although this had not been a targeted behaviour.

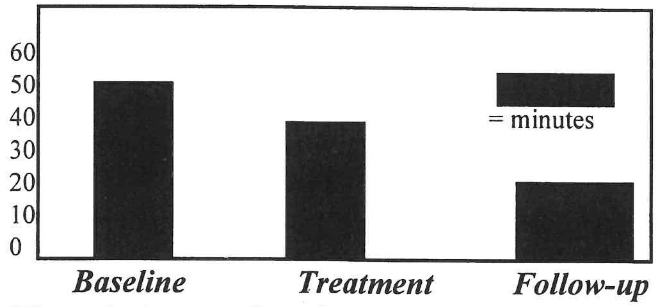

Figure1. Average length of night waking in a 5 year 1m girl.

INFORMATION

Discussion

The results of this questionnaire study confirm the findings of other authors that children with disabilities in general, and particularly children affected with Sanfilippo syndrome, have a very high prevalence of sleep problems. Furthermore, analysis of the detail in parental reports has provided a rich source of information about the nature and extent of these problems. Of particular interest are the bizarre behaviours reported, such as inappropriate laughing and singing at night. together with rapid waking and occasional periods of staying awake all night. It is possible that these are indicative of a particular form of disordered arousal as the degeneration of nervous tissue proceeds, which may form part of a behavioural phenotype (O'Brien 1982).

The extent of the problems described, and the evident strain on families, prompted us to examine a few individual cases in more detail to establish whether standard behavioural techniques would ameliorate their situations. The results of these interventions were encouraging and we hope they will increase awareness of the problems faced by families of children with Sanfilippo syndrome and stimulate further research into the effectiveness of behavioural intervention in this condition. The presence of such severe sleep disorders places additional pressure on families who already have to manage behaviour problems during the day and may still be coming to terms with the fact that their child's condition is deteriorating and incurable. Although tackling these sleep problems will have no direct benefit in terms of prolonging length of life, it will improve the quality of life for all the family and thereby facilitate better coping. We hope that on the basis of these results professionals will be less hesitant about applying social learning to the management of children with degenerative conditions. It is important not to become overwhelmed by the inevitable sense of impotence which arises from being unable to provide a cure, but instead to recognise the value of the application of strategies such as these which can have a significant positive effect on the day-to-day lives of the families.

Summary

Sanfilippo syndrome is a rare degenerative disorder which has severe intellectual and behavioural sequelae, commonly including sleep problems. A parental questionnaire was used to gather information on the sleep patterns of 80 children with Sanfilippo syndrome (mean age 10 years 2 months). The majority were found to have sleep problems (78%). Many also exhibited other distressing and unusual night time behaviours (staying up all night, chewing the bedclothes or crying out suddenly), and a few laughed or sang. Such problems may have been more severe in those with Sanfilippo syndrome type B. In four of the families offered individually tailored behaviour management advice there was immediate improvement, which was maintained at follow-up in two cases. These results demonstrate the usefulness of even such a minimal intervention, even in a very difficult population such as this.

INFORMATION

NEW ASPECTS ON THE MUCOLIPIDOSES

M. Cantz from the Institut fur Pathochemie und Allgemeine Neurochemie, Universitat, Heidelberg, Germany ***[reprinted from Courage]***

The Mucolipidoses (types I - IV) are genetic lysosomal storage disorders exhibiting clinical features of both the Mucopolysaccharidoses and Sphingolipidoses. In Mucolipidoses I - III, the underlying biochemical deficit has been elucidated, providing a basis for their diagnosis and genetic counselling, for a better understanding of their pathogenesis, and for future attempts at enzyme replacement or gene therapy.

Mucolipidoses I is a sialidosis due to the deficiency of a lysosomal sialidase (neuraminidase). The enzymatic defect leads to an abnormal accumulation in tissues and excretion in the urine of sialic acid containing carbohydrate chains, called sialyloligosaccharides. More recent studies have also shown an excessive deposition of sialic acid containing glycolipids in a variety of tissues. The clinical consequences may vary considerably from patient to patient and include motor and mental retardation, impaired vision, seizures, and skeletal changes. A clinically and biochemically related disorder is Galactosialidosis, where the simultaneous deficiency of sialidase and beta-galactosidase activities is caused by the genetic defect of a so-called protective protein which is itself an enzyme (carboxypeptidase) that is needed for their stabilisation.

In Mucolipidosis II (or I-cell disease) and Mucolipidosis III (or pseudo-Hurler polydystrophy) there is a simultaneous defect of most lysosomal enzymes in many cells and tissues. This is due to a genetic defect in the formation of a common signal on lysosomal enzymes (a mannose-6-phosphate residue) needed for their intracellular transport from the site of syntheses to the lysosome. Formation of the signal is accomplished by the successive action of two enzymes the first of which is a phosphotransferase, and it is this transferase that is primarily deficient in Mucolipidoses II and III. As a consequence, the lysosomal enzymes do not reach their proper destination but are exported into the extracellular space and are greatly increased in extracellular fluids. The multiple lysosomal enzyme defect causes the storage of mucopolysaccharides, complex lipids and oligosaccharides in the lysosome; recent results also indicated a severely compromised lysosomal protein degradation. The distinction between Mucolipidoses II and III is on clinical grounds only and cannot be made by biochemical tests. Mucolipidosis II is clinically more severe, the major systems consisting of psychomotor retardation, coarse facial features, enlargement of liver and spleen, and death usually in the first decade. Mucolipidosis III runs a milder course with a later onset, and survival into adulthood is possible.

The main clinical features of Mucolipidosis IV consist of progressive psychomotor retardation and corneal clouding. This is lysosomal accumulation of gangliosides, phospholipids and mucopolysaccharides previously thought to be due to the defect of a ganglioside sialidase. While it is now clear that the ganglioside sialidase is not primarily involved, the basic defect remains to be discovered.

INFORMATION

We have been asked by Ann Worthington of the In Touch Trust to pass on details of a new publication of "Useful Addresses", which may be of interest to some of our readers.

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FINANCE
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THE: IN TOUCH SERVICE

In Touch was founded by Ann Worthington in 1968. The aim was to promote informal contact between parents of children with special needs, particularly those affected by rare disorders, so that they could support each other by the exchange of ideas, experiences and encouragement.

From the outset, the membership grew steadily and so did the amount of 'feedback' from families and professionals. This led to the first one-page newsletter being sent to 200 members at the end of 1968. Currently, some 1,400 members receive a 32 page newsletter, three times a year. Two thirds of the In Touch membership is made up of parents. The remainder consists of professionals and organisations catering for special needs in children. Several national self-help groups for specific rare disorders have been formed by parents originally linked up by In Touch.

One of the most successful aspects of In Touch is the provision of contacts for parents of children affected by rare disorders. These families are unlikely to meet anyone in their own area whose child has exactly the same condition and, as a result, they feel especially isolated. Because In Touch has a national network, such families can often be linked up with others in which there is a similarly affected child.

In Touch also provides information about specific uncommon disorders and explanation of medical terminology to professionals and will refer enquirers to other support groups, where appropriate. In Touch became a registered charity in 1982.

For a copy of Useful Addresses please contact :
Mrs Ann Worthington, 10 Norman Road, Sale, Cheshire M33 3DF
Telephone - 0161 905 2440

INFORMATION

Whizz-Kidz

The Movement for Non-Mobile Children - Whizz-Kidz is a registered charity which aims to enable all disabled children to have improved mobility, increased independence, and a greater Quality of life. This is achieved by providing them with lightweight, powered and sports wheelchairs or any other mobility aid they may require. Mobility may seem unexciting but for a child or young person it can mean the difference between being able to move to where someone else puts you and moving independently around a school or home, or going to the local shops. The improvement in a child's self-confidence, development and enjoyment of life is immediate and remarkable. The whole ethos of Whizz-Kidz is to encourage individuals, schools, companies, and any other groups to raise money for a specific Young person, therefore making the fundraising instantly tangible and personal.

Whizz-Kidz:

- supplies mobility aids to any disabled child in the UK under the age of 18 years.
- has raised over £2.5 million for mobility aids since it began in 1990
- makes sure that every child is assessed individually for their mobility and seating by a qualified Occupational or Physio-Therapist.
- purchases mobility aids to meet each child's needs fully, regardless of cost. Equipment costs range from £500 to £8,000.
- helps children for whom NHS provision is inadequate.

Whizz-Kidz developed as a result of the London Marathon in 1989 so there is often an energetic slant to fundraising. Whilst marathons are a significant part of their events, people also climb mountains, cycle across continents, and brave the high seas - anything is possible. If your child needs a mobility aid, please contact Whizz-Kidz at 215 Vauxhall Bridge Road, London SWI IEN Tel. (0171) 233 6600.

"The Loss of a Child"

This article is reproduced from the Harrogate CVS Newsletter.

"Following a seminar of the Under Eights (Children and Families) forum held last October on "The Loss of a Child" a leaflet has been produced by a social work student, Julie Wilkinson, based at Harrogate and Area CVS. Entitled " When Your Child Dies", the leaflet describes a number of local organisations, helplines and national office numbers that can be contacted at a time when people may be finding it difficult to cope following the loss of a child. Being able to talk to others who have been through the same experience may be a way of finding comfort. Leaflets will be available from a number of sources including local GPs, hospitals, health centres and the CVS office.

For further details Tel: 012423 504074

INFORMATION

The Jessie May Trust,
founded by the parents of Jessie May Purrington, began a new service on April 1st 1996 aiming to provide respite care for children with terminal and life-limiting illnesses. The Trust's nurses are able to assist the family in the nursing of the child in the family home and in so doing give parents the confidence to take a much needed break. It can help where children are aged between 0 and 16 years and those aged up to 19 years will be considered for terminal care.

The Jessie May Trust operates across South Gloucestershire, Bristol and North Somerset.

Contact Tel: 0117 950 7580 Between 9am and 5pm or leave a message on the answerphone.

Phoenix Lodge
aims to be the first **adolescent** hospice and respite care centre in the country. The main aims are to provide a specialist respite care home with hospice facilities for young people aged between 15 and 30 years who have a life-threatening illness or disability.

Contact Tel: 01908 314092.
Phoenix Lodge, 33 Stratford Road, Wolverton, Milton Keynes K12 5LW

Children in Focus
The death of a child, whether it be your son or daughter, a grandchild, or your brother or sister is one of the most upsetting events for their family and friends.
Bereavement is 'mind-numbing'. It is the last thing you expected to experience at this time. You may feel confused and angry by what has just happened and the last thing you want to do is think about the painful decisions that lie ahead.
Children in focus is a new scheme which has been set up by funeral Directors who care about the choices and decisions that you will need to make in the coming days.

If you would like further information or a brochure please ring David Chadwick, Project Manager Tel: 01628 523253

There are no charges for the above services for babies and children's funerals.

INFORMATION

The book pictured below
may be helpful to you
in teaching your
children about tube feeding.

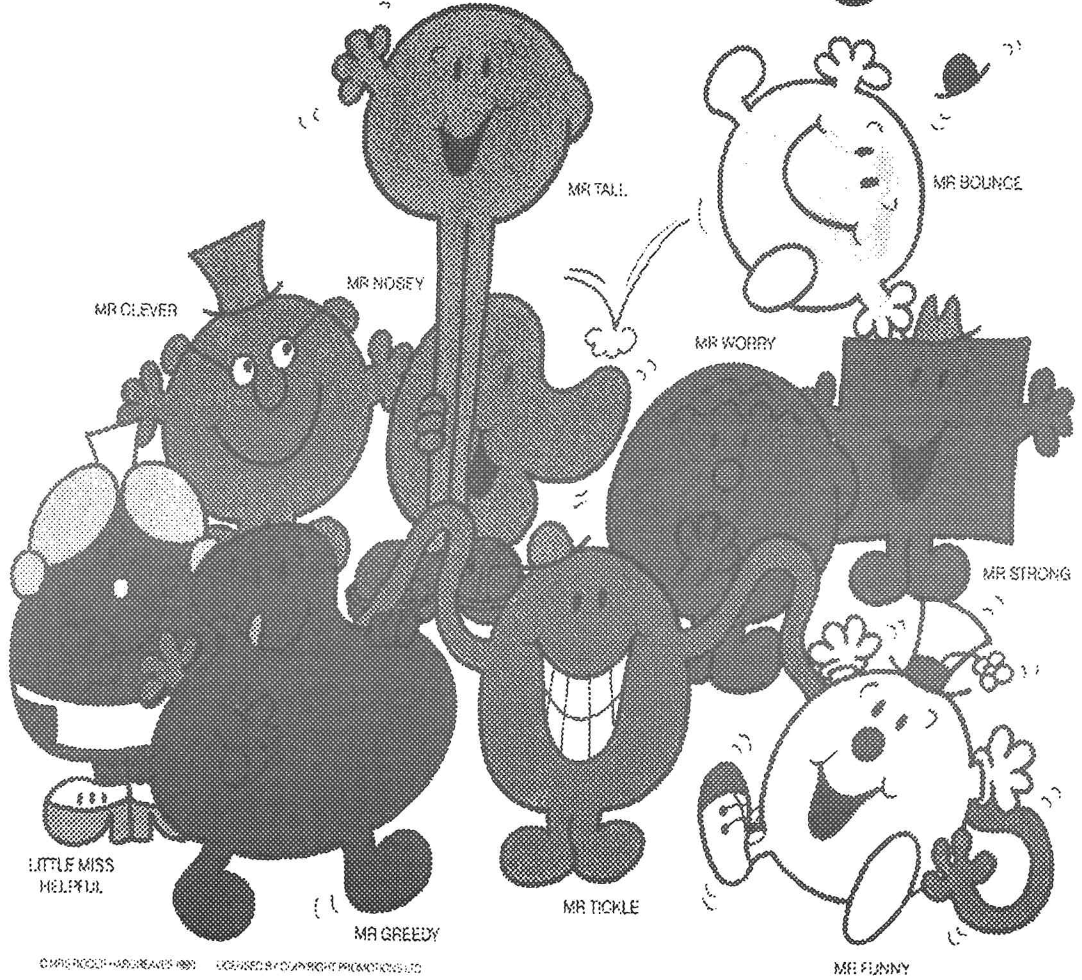
If you are interested in obtaining a copy
Please contact: Cow and Gate Nutricia Ltd.
Whitehorse Business Park, Trowbridge
Wiltshire BA14 0XQ

The book opposite is titled
"Tube Feeding at Home"
and may be obtained from;

Sherwood Medical Industries Ltd.
County Oak Way, Crawley
West Sussex RH11 7YQ

Tel: 01293 534501

The Mr Men tell you about tube feeding



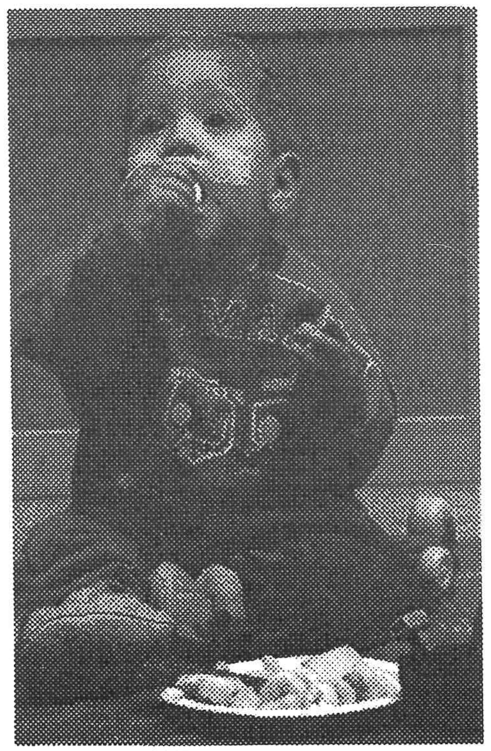
INFORMATION

How will my child learn to eat again?

Children who have been fed by a tube for a long time are often variable or poor feeders when they are first introduced to oral feeding. This is not surprising, particularly if they have had long periods of being unwell. Progress towards normal feeding may be up and down and can take a long time. It is very important not to try and rush this process as this may lead to other problems. However, even when your child is not in a good phase, it is still important you try to include him/her in the normal routine of family mealtimes.

What do I do if he hates mealtimes?

Mealtimes should be sociable occasions which are enjoyable and fun. Try not to start off with either of you in a bad mood! If your child is grizzly try to cheer him/her up with a quick play or cuddle before you start. Include your child in your mealtimes e.g. have him/her sitting at the table with you and give them a bowl, spoon, cups etc. to play with while you eat.



How can I help my child learn to eat?

Make sure your child is sitting comfortably in the right kind of chair with a table or tray in front of it. Sit in front of him/her so that they can see your face. Put a few toys to do with eating on the tray so that they learn to join in (e.g. plastic spoon, cup, plate and small doll). Make sure you are there to watch, encourage and be very pleased when they put things in their mouth, take food off a spoon and swallow. Make appropriate noises and mouth movements in order to encourage your child to copy you.

INFORMATION

MEDICAL ARTICLES - PEDIATRIC DENTISTRY AND MPS CHILDREN

Steven M. Baylin, D.D.S., MSc.D Calgary, Alberta, Canada

There is a lack of public information relating the Mucopolysaccharidosis (MPS) and paediatric dentistry. Hopefully, this article will provide information about these children's mouths and teeth, and how to care for them. The intention here is to heighten dental awareness in areas of growth and development and preventive oral health programming.

Paediatric patients diagnosed with MPS I, II, and VI (Hurler, Scheie, Hunter and Maroteaux-Lamy Syndromes) show similar dental characteristics which play a role in diagnosis, prevention and treatment. These children usually have widely spaced teeth which can appear to be ground down from grinding. Their tongues may be very large (macroglossia) and protrude. This, along with large adenoid tissue, can create the problem of an open bite along with mouth breathing.
It is not uncommon for these children to exhibit delayed and unusual eruption of the permanent 6year molars and subsequent permanent teeth. Also common are localised areas of bone destruction around unerupted second primary molars and permanent molars. These dentigerous cysts can be diagnosed as early as ages 3 to 5 years. Finally, the children in this group may exhibit underdeveloped lower jaw condyles (the "ball" of the ball-and-socket joint of the TMJ), thus limiting lower jaw movement.

MPS III (Sanfilippo A,B,C and D) patients may have multiple dental abscesses in late stages of the disorder. Usually they do not show macroglossia, but their tongues may enlarge (again, in later stages). MPS IVA (Morquio A) children have primary and/or permanent teeth with pitted or grey enamel which can chip off.

All MPS patients deserve and require a meticulous dental care program. This includes at least twice yearly or quarterly visits to a dental sugeon willing to take the time to follow the course of the disorder and its possible oral manifestations. Prevention includes: preventive diagnoses (radiographs to check eruption, cysts, etc.), preventive oral health care (tooth polishing, soft tissue management, scaling, flossing, pit and fissure sealants), management of growth and development, and control of caries and extraction/eruption sequencing. Treatment includes all of the above combined with a good home preventive program of daily brushing and healthy nutrition.

MPS patients often have difficulty chewing hard foods. As a result, the gums are not stimulated and are quite fragile. With associated mouthbreathing, it is common to find swollen, red and sometimes bleeding gums. Therefore, efficient care of all gum tissue with good brushing both at home and at the dental office is paramount. Plaque and calculus (tarter) build-up can then be minimised and periodontal disease effectively reduced or eliminated. Caries can be prevented by avoiding sticky and sugary carbohydrate foods. Limiting these types of snack foods will reduce the acid buildup that causes decay.

Paediatric MPS patients should be seen by a dental surgeon as early as age 2 or 3. Usually, they can be easily managed in the dental environment but later on may need general anaesthesia to effect a more efficient job of diagnoses and treatment. Consistency and thoroughness is the key to sustaining effective dental care for children with MPS disorders.

The Canadian Society For Mucopolysaccharide & Related Diseases Inc.

INFORMATION

CARE IN SCHOOLS OUESTIONNAIRE

Enclosed with this Newsletter is a questionnaire asking you about your child's school and the care input while in school. The reason behind this is to try and find out as much information as possible in order to address the need for palliative care in schools for children with life threatening conditions.

At present this seems to be disproportionate and it would appear that it all depends on which authority you live in as to the level of care your child receives.

As MPS is a well documented group of conditions, the level of disability is reasonably well known for the majority of children. Therefore, the information on the course of the illness is available on entering school or at diagnosis and, in theory, future provision should be made. However, parents and organisations like ourselves are continually having to battle for resources at practically every stage and especially when addressing palliative care and the wishes of the parents.

We are aware of the cutbacks in health and education and these have hit hard at resources like nursery nurses, speech therapists, physiotherapists and registered paediatric nurses. All ultimately would be responsible for activating and maintaining palliative care within your child's school.

In some schools which our children attend there are no qualified nurses and other therapists are limited, often sharing time with other schools, which means much is left to the good will of the school and teachers.

Many argue that if children are so ill that they need palliative care then they should not be in school. As MPS is a slow degenerative condition many of our children benefit enormously and enjoy the stimulus of school life, even when their skills are lost and much care is needed.

It is the issues surrounding the care component and palliative care plans for children with life threatening conditions which the MPS Society and other organisations are concerned about. We need to "know" what is actually happening, not just in one school but in every school that an MPS child attends.

There will be many of you with younger children who probably will not have thought along these lines yet, we do urge you to complete the questionnaire and any added information that you can give will be useful.

In closing, there are schools who have excellent practice and who endeavour to offer the very best for our children, we want to build on this for every child and to protect what has already been achieved.

Mary Pagett
Director of Family Support Services North

FAMILY NEWS

This article was taken from the Contact a Family Newsletter. Siblings workshops are still in great demand. In July, working in conjunction with the Network of Parents of Children with Special Weeds, we held another one, attended this time by 40 parents and professionals from East Sussex. Susan King, now 23, shared with participants her childhood memories of her disabled brother:

My brother, Michael

" My brother was diagnosed as having Hunter Syndrome when he was three and I was five years old. It is a rare condition and in 1978, when diagnosed, there were only about thirty other known children in the UK with this condition. He had a maximum ability equivalent to that of a two year old, was deaf and incontinent all his life. The illness is a progressive one - from running around to being unable to stand - we went from buggy to wheelchair. My brother Michael died in 1985 when he was nearly eleven: I was then twelve years old.

"What was important to me was being told at every stage what was going on and being consulted by my parents on what we should all do. People think that brothers and sisters are not aware of things - **this is not so**. I was aware if something was wrong with Michael before my parents were. Part of the syndrome is that the hands become clawed - I knew this before my parents did!

"I was involved with everything. When Michael had to be tube fed I did this - I would help the nurses with what they did. We used to go out a lot in the car and despite Michael's handicap we went to many places. Lots of parents I met were reluctant to take their children out - this is a mistake. We went most years in our caravan to France. We went down caves, my Dad would carry Michael; my Mum and myself carried the buggy or wheelchair and the various bits and pieces. If Mum was struggling often a complete stranger would offer to help carry things. If anything, I gained as Michael was going to have a short life and my parents tried to cram in as much into the years as was possible.

"It is probably easier to give attention to the disabled child and parents can, I suppose, ignore the fit child - but this did not happen to me. It is important I think that you do not spend all your affection on the disabled child to the detriment of the other children. I also had my own activities such as dancing, Brownies and riding. Michael went to Riding for the Disabled and I could help with this - it was something we shared without our parents being involved -just like a normal family, if there is such a thing!

"Although we usually went out as a family there were certain places we could not take Michael - for example to the top of Tower Bridge. My parents took me out on my own, probably once a month or every six weeks, for a day. Michael would go into respite care for the weekend to a place nearby. This was important as we could do everything we wanted to much more easily. It was good for my parents as well as for me.

FAMILY NEWS

"People have said to me that my childhood was not normal - I don't argue. My friends all loved Michael and he liked seeing lots of people - no one would be rude to or about Michael because they were my friends. When we were abroad once someone was staring so I ran into them on purpose with the buggy then apologised - they didn't stare anymore!

"My Dad said that being a girl made it easier for me because girls are more caring than boys - I hate to admit it but he is probably right! Having had a handicapped brother has certainly made me a more caring person. I also do not get involved in trivia - people seem to get bothered about minor things - but not me!

"I was always protective towards him - as I said my friends all liked Michael and if they didn't they were not my friends. What I think is important above all is to be consulted - I was treated as an equal and not as a child. I think I matured quicker but I did not lose my childhood. The consultation went on to the very end - Michael died with me holding his hand. The funeral service and the burial plot were chosen by us all - not just by my parents. After Michael died my father was asked to speak on how he and we as a family had coped. He asked me for my comments and feelings and what I said then does sum it up:

"A brother is someone you love, who loves you, and someone you have a lot of fun with and have in fact, shared your entire childhood with. "

"My initial reaction to Michael's death was one of shock but prior knowledge helped to lessen it. The loss changed the pattern of my life - there was no longer a continued commitment to his routine of feeding and care. My conversations with Michael were replaced with talking to my toys and pets - the games I played had always included him and so these had to change. I liked continuing to mix with children with similar difficulties as it helped me to be able to use my experiences positively and I could look back with pride and pleasure over times I had shared with Michael. Having friends is important and my true friends were friends I could talk to openly and not feel embarrassed with. I did not cut myself off from other siblings although I realise this could be upsetting for them. I did momentarily resent that Michael died when others were still alive but this quickly changed into a need to help others especially when they were facing the same difficult time.

"My father said that having Michael as a brother had made me more mature though not too mature. I was proud of him and could return, when necessary, people' s stares with smiles. To be included in discussions and plans for the funeral was most important as although, when my brother was buried I lost part of myself, it was easier for me having been included in all the decisions."

Susan King

FAMILY NEWS

Free - Good Home Wanted
For
Turbo Electric Wheelchair
10 years old.

If you are interested please contact:
Valerie Grant **Tel: 01303 252829**

Information Wanted

Do you have any solutions to Managing screaming in a Sanfilippo child?

If you have experience of this or if you have any advice at all about this please contact:

Hilary Stewart,
Community
Paediatric Sister
Tel: 01642 617617

Address and Telephone Update

As you will see we have included a Form for completion by any families who have moved house or changed their telephone number. We have been having a few problems in contacting members due to the fact that addresses and telephone numbers have changed.
We would therefore be grateful if **everyone** would complete the enclosed form and return it to the Amersham office **whether your details have changed or not.**

Thank You.

In Appreciation

I want to take this opportunity to thank Dr Ed Wraith for assisting me with TJ's care during 1996.

Any problem I encountered with TJ's condition and referred to Dr Wraith was immediately handled with the necessary expertise and diligence, and dealt with in a speedy reply, all very reassuring and of great comfort to me. My heartfelt thanks for your professionalism and concern.

Regards *Lynn Piennar*
(South Africa)

FAMILY NEWS

Poem by Denise Brown - Mum of Adam aged 5 (Sanfilippo)

ODE TO THE INCONTINENCE SERVICE
FROM A SANFILIPPO FAMILY

We're Running So Low On These Nappies
I Don't Know What We're To Do
Cos There's No Way We Can Stop Adam
From Needing To Go To The Loo

6 A Day You Say Is The Max
But We Find That Incredibly Lax
We Definitely Need More Maybe Three Maybe Four
We're Not Lying Just Stating The Facts

And With That We Had An Enquiry
Into Adam's Toileting Needs
And The Lady Proceeded To Ask Us
How Much He Drinks And He Feeds

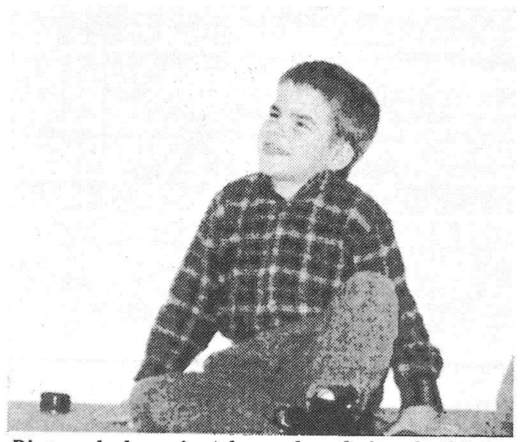
We Told About Adam's Habits
How Many times He Went A Day
The Lady Put Pen To Paper
We Thought We Were Doing O.K.
With That She Said "We Should Weigh Them"
Each Nappy Should Hold So Much Wee
With That Adam Stooped ***(We Knew What This Meant)***
Adam Proceeded To Giggle With Glee

We Weighed The Nappy In Question
The Lady Said This Is Too Light
We Looked At Each Other And Both Said At Once
Add The Trousers That'll Make It Alright

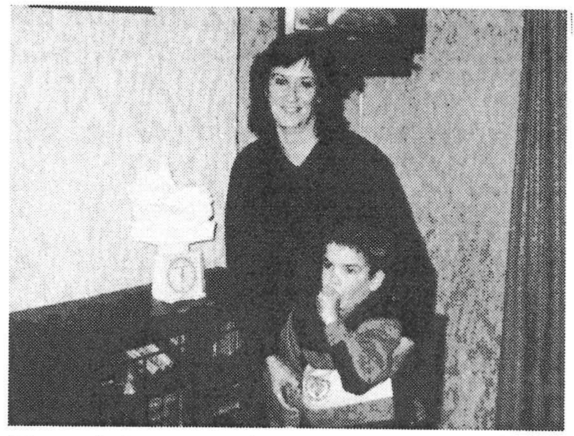
We Hope You Enjoyed Our Story
Which Many Will Know Very Well
Persevere With Your Quest
You Will Come Off The Best
As Gavin And I Live To Tell

We have since heard from the service and they agreed to an extra pack each delivery. Also, if it seems we are still running short to give them a ring and they will supply an emergency pack. I hope this will help with everybody having the same problems.

Denise.



Pictured above is Adam who obviously thinks his Mum is funny.



Pictured above are Adam, his mum, Denise and the guilty underweight nappy.

FAMILY NEWS

June's Story

My name is June Elliott and I would like to talk to you about living with Morquio. I am 11 years old. I have one brother, Mark who is 7 and one sister, Karen who is 13.

SCHOOL: My chair at school is to small for my table and so I have a larger chair than the rest of my class so I can reach the table. I don't find school too bad. Except when the new Junior infants come in it's a bit tough but once they get used to me it's fine, they understand who's older. I don't find school work too hard (except the usual like Maths and Irish)which is a help. I love doing P.E but I can't do most of the things the other kids can do like football and Basketball, which I really would love to do.

MEDICAL: I was first diagnosed when I was 5 years old. Before I was diagnosed I was thought to have scoliosis until I went to a conference in Limerick and then I was diagnosed. Shortly after that I visited Temple St. Hospital to see Dr. Naughten who works in the metabolic unit of the hospital. I also attend the eye and ear specialists and Mr. McManus, the orthopaedic surgeon.

The worst thing that I remember about going up to the hospital was when I was 9 and I received news about having an operation on my knees to straighten them. I knew they had to be operated on but I still didn't want to have it done.

I got my first operation on my right leg and I stayed in hospital for a while. Then I came home and I had a hard plaster on for 6 months. This meant that Mammy had to come to school with me and I had a wheelchair. When I got used to the plaster I could walk on it. A while after that healed up I had the same thing done with the left leg. This operation was not really successful because my legs are more or less back the way they were.

HOME: At home the handles of the doors are quite high and difficult to open. The light switches are also too high but Mammy and Daddy got cords installed in my bedroom and in the bathroom. I never really have showers so I settle for a bath. All the sinks are too high but I have a stool in the bathroom so I can reach the taps. The top shelves in the kitchen are also too high. I can't reach the table with the chairs properly and so I have a big cushion which means I can reach the table.

GOING OUT: When I go out in the car I bring my cushion with me so I can see out the window. I use my buggy when I go out shopping and on my holidays but I hope to get a wheelchair soon.

HOBBIES: I love swimming and I may be moving into the big pool which will be a great achievement for me. I also enjoy reading, writing, collecting stamps, I love Boyzone, and obviously, listening to music.

FAMILY NEWS

PHYSIOTHERAPY: The Physiotherapist came a few times this year and I had to do a few exercises. She is coming back next year to do more exercises and to check up on me.

HEALTH: This is the worst part because I get a lot of chest infections and when I get sick, I really do get sick and I have to take antibiotics. but then I bounce back.

Despite all this going to hospital, visiting doctors and having operations I do have my own life and I live it the best I can and enjoy it.

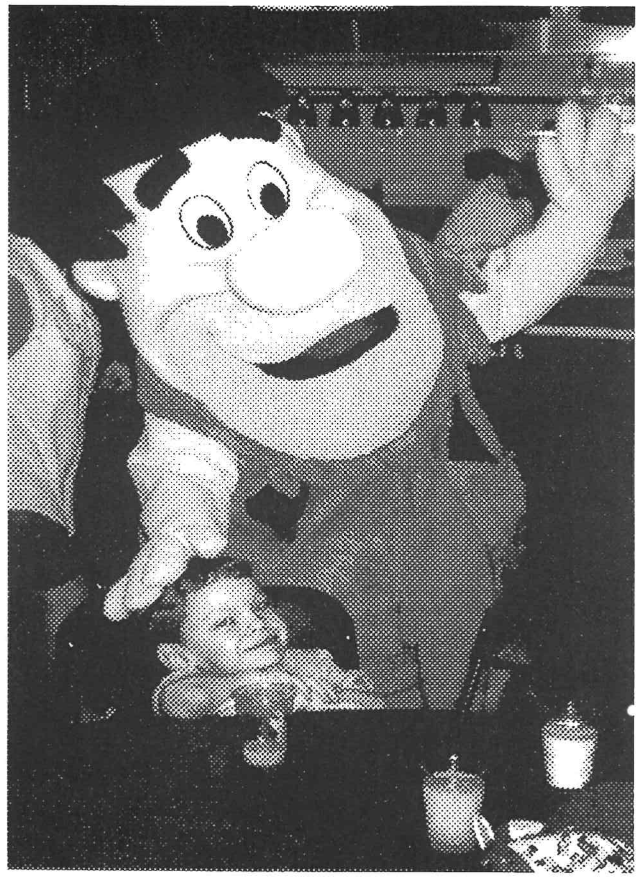
June Elliot

Pictured above are Karen, June and Mark.

FAMILY NEWS

It's been a great year for Henry Sutcliffe aged 9 years.

In April he was lucky enough to go to Florida with the National Holiday Fund and had lots of fun and different experiences. At the end of February he also received an electric wheelchair from Whizz Kids which has really improved his mobility
Now his friends have to chase him!.



Henry aged 9 years who suffers from Morquio Disease is pictured here in his new wheelchair and at Disney World with Fred Flintstone.

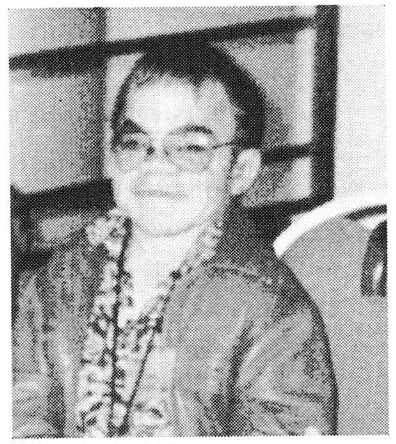
Photographed opposite holding the large cheque is Jonathan Armstrong brother of Michael aged 7 years who suffers from Hunter Disease.

Jonathan aged 9 years attends the R.C. Primary School, Hartlepool where the children and staff raised £115 for MPS by holding a 'No School Uniform Day'.

OVERSEAS NEWS

MPS Australian Conference 1996

During the conference Dr Wraith and myself were given the opportunity to facilitate an MPS adult workshop, looking at issues surrounding the problems adult sufferers encounter on a daily basis. The Workshop, to say the least, was enlightening even to Dr Wraith and myself. We learnt a great deal from these young people.
The following is a record of the workshop kindly written by Colin Thompson, an adult Hunter sufferer and our grateful thanks are extended to him and all those who took part. ***Mary Pagett***



Colin Thompson

ISSUES FACED BY ADULTS WITH A DISABILITY

Being an adult in the 1990's is difficult enough for most able-bodied people. The issues are even more pronounced for people with any kind of disability.

Without seeking to list all the various disabilities it is the intention of the author to take an inclusive rather than exclusive approach by not specifying particular conditions. It is enough to describe someone as, "disabled".

The most pressing issue that came up related to sex. Sex and its associated hang-ups is a topic that tends to confront all adults be they disabled or otherwise. Unfortunately the issue of sex tends to be taboo.

Sex and the disabled has always been a subject either ignored with embarrassment or discussed hesitantly and unwillingly. In 1977 Alan Marshall, a famous Australian author and person with some first-hand experience of the problems of the disabled, having suffered the effects of polio in childhood, made a submission to the advisory committee on the sexual rights of the handicapped. He wrote, "The world is crueller in its judgement on handicapped people than it is on those who are not handicapped. A handicapped person is not supposed to need physical contact or any sex whatsoever."

The problem is that parents, the public and the medical profession alike tend to regard disabled children as perpetual Peter Pans who never grow up to be men and women with sexual feelings.

The issues in relation to sex is even more complicated for adults with the various types of MPS. With the Hunter condition in particular, puberty and all the natural changes normally experienced in early adolescence is delayed. By the time you begin to

OVERSEAS NEWS

experience all the natural desires and urges any opportunity for shared experiences or questioning has passed.

Amongst the delegates there was unanimous agreement that the issue of sex needed to be acknowledged more by parents and the wider community. There seems to be a lack of general knowledge on the part of parents about the sexual needs of the disabled adult.

Interestingly enough I was asked whether I was going to Kings Cross, a red-light district of Sydney. As it was I wasn't, but the question served to highlight the lack of understanding about sex held by parents. I had the same sexual desires as the father.

Another prominent concern expressed by the adults related to the issue of workplace discrimination and the lack of employment opportunities for the disabled. Like anyone the sense of satisfaction and feeling of accomplishment that comes from a job was echoed. While our individual disabilities may be a barrier to some jobs everyone felt that they could contribute something if only they were given the opportunity.

The issue of body image is something that affects everyone at one stage in their lives. This problem is compounded doubly when you have a disability. When you don't seem to fit in or people stare or point, your self confidence tends to take a battering. It's something we all have to come to terms with. Usually people who make a fuss after they have got to know you, are not worth knowing.

The hardest thing to deal with are parental expectations. Having a disability in a way makes it easier to satisfy parental hopes and dreams. Once parents realise that their offspring has a disability they will hopefully adjust their expectations accordingly. In that case your outlook and level of personal achievement is up to you.

Another issue of particular importance raised by some delegates was the need for honesty on the part of doctors and other medical personnel. Having had many years to contemplate the extent of our own conditions, whether pleasant or not it is good to get a doctor to tell you how it is, minus the "bullshit" (colloquial Australian term meaning crap)

It is important for us to have all the facts before we go into an operation so we will know what to expect. It is also good, though frequently rare, to find a doctor who talks to you on a human face to face level, not from way up on their pedestals.

Finally a word should be said about the topic of loneliness and the feeling of isolation experienced by people with disabilities. Feeling as though you are alone and that no one else is aware of your difficulties can be a common problem. I'm afraid there is no simple answer. We all are aware of these problems. It's a bit of a relief to know that you are not alone and to take heart from the knowledge that everyone has a "downer" so don't despair.

This article has been written as a record of discussions by adult MPS patients at the 4th international Conference on MPS and Related Diseases in Wollongong, Australia in May,1996.

If you have any problems or other issues relating to any of the above issues you can write to: Colin Thompson 224 Foote Street, Templestowe, 3106 Australia

OVERSEAS NEWS

Living With MPS
Life With MPS VI Has Meant Conquering a Number of Hurdles

Introduction
For those of you who I have not had the pleasure to meet as yet, my name is Catherine Hartcher and I have MPS VI, Maroteaux-Lamy Syndrome. On February 9th 1996, I celebrated my 38th birthday. I am one of three children born to Arthur and Colleen Hartcher. Neither of my siblings have MPS, nor do any of their children.

Hurdles
Hurdle No.1, In 1967, my parents received an initial diagnosis of MPS when I was approximately 7 years of age. At this time my parents were told I had Hurler Syndrome. The diagnosis was later changed to Hurler-Scheie when I was examined by a visiting overseas specialist. Along with these diagnoses came the notion of "life-threatening" illness which had an enormous impact upon myself and my parents. This initial hurdle was overcome by not looking beyond the day at hand, that is to say, my parents (and relatives) instilled in me the idea that textbook diagnosis can be wrong and that it was essential for us to take each day as it came. This ensured that rather than take a negative or pessimistic view of what I would not be able to do my parents, and consequently myself, lived our lives with a more positive, optimistic view of accepting what I could manage.

Hurdle No.2 During my childhood I achieved age appropriate milestones and, although I contracted chickenpox and measles and occasionally experienced a bout of tonsillitis, my overall general health was good. My development during this period I consider to be fairly normal with the exception that the mobility of my limbs was mildly restricted. This immobility lead to the realisation that I would need to wear clothing that was easy for me to get in and out of without the assistance of another person. In order to achieve this Mum either made my clothes with openings down the front, or purchased garments that accommodated the inability to raise my arms.

Hurdle No.3 I began school at the age of 5 years and continued my schooling until I completed the School Certificate in 1973. Memories of my primary school years are happy ones of joining in, and enjoying, all school activities along with my peers. It is only when I recall my years in secondary school that some unpleasant memories surface of incidents which, at the time, resulted in me developing a rather cynical view of people and how they relate to one another. This cynical outlook remained with me, influencing my relationships, for some years.

One important thing that I, and my parents, learnt from my high school experiences was that we needed to discover all we could about MPS and its possible effects on me. We soon realised that, as not a great deal was known about the disorder at this time, what information we did gather needed to be passed on to any treating medical staff who we encountered.

Having finished Year 10, I left school to attend business college with a view

OVERSEAS NEWS

to gaining employment in some form of secretarial work. In 1975 I gained employment with an insurance firm and later in 1979 moved to a position with a finance company. Life was great and I certainly lived it to the fullest, going out and on trips, etc.. and, although I knew I didn't always do things exactly the same way as other people, having MPS wasn't causing too many problems. YET!!!

Hurdle No. 4 In 1982 problems with my health first appeared. I was experiencing difficulties with constriction of nerves in my neck. This eventually led to my leaving work in March 1983 and undergoing a Cervical Laminectomy (C 1-C6) in November of that year. At this time it was also discovered that I had cardiovascular irregularities. My mitral valve was not functioning properly. In 1984 I underwent open-heart surgery to rectify this problem. As a result of the pathology report of tissue removed from the mitral value, in 1984 the diagnosis of Hurler-Scheie was altered to Maroteaux-Lamy Syndrome. Both operations were extremely successful and alleviated the discomfort I had been experiencing However, the anaesthetists involved in both procedures reported experiencing difficulties with intubation.

As a consequence of the heart surgery, along with expected medical difficulties relating to MPS, I was advised to seriously consider tubal ligation. As I had no intention of passing MPS, in any way, shape or form, to any possible offspring, I underwent this procedure in 1985. Unfortunately my experience of what should have been, under normal circumstances, a simple operation proved to be rather complicated. The difficulties I experienced related directly to my having MPS. Firstly, due to the narrowing of my throat and dislocation of my larynx due to a build up of MPS matter, I had to be awake during intubation in order to be anaesthetised - a very frightening and somewhat uncomfortable experience. Secondly, since having had the mitral value replacement, I must now undergo a rather arduous process of heparinisation whenever any surgical procedure is performed. This meant that instead of being in and out of hospital the same day I was required to stay approximately 14 days.

I must admit, of the three operations, this was the most difficult for me to get through in a psychological sense. Not only was I still getting over two bouts of major surgery but in the period between the two operations I had suffered the death of my fiancé.

Hurdle No.5 I was now approximately 28 years of age and was having some difficulties coming to terms with the idea that I would not be able to return to work involving office procedures but certainly had no desire to reach the age of 30 years knowing I was to

OVERSEAS NEWS

remain an invalid pensioner. I was becoming more and more depressed as each week passed.

It was at this time that I first heard of the MPS Society and met Ros Smith and a number of the other families. I attended my first Family Conference held at Albury and consider this to be the point in my life when I overcame a crucial hurdle and gained a goal which I could strive for. On the journey home from the conference I had a sudden revelation of what it was I wanted to do with the rest of my life. I wanted to gain sufficient skills to enable me to assist families in some way. I wasn't sure yet how I could do this but I was willing to give it a try, after all, I still had sufficient mental skills even if my physical health was not 100%. I now had a goal, a light at the end of the tunnel.

In 1986, in an effort to gain the appropriate skills needed to assist families experiencing difficulties, I commencing university studies, majoring in psychology and graduated in 1991 with a BA(Psych). Having graduated, I married in November 1991. However, the relationship faltered and we divorced some three years later.

Fortunately for me, in 1992, I had gained employment as a family worker with Cessnock Family Support Services Inc., (a non government community-based organisation funded through the Department of Community Services) and was able to put all my efforts into my work, thereby alleviating some of the distress at the break-up of my marriage.

Hurdle No.6 In 1994 I again received confirmation of the diagnosis of MPS VI, under somewhat reluctant circumstances, when a procedure was performed to remove a build-up of tissue on the cornea of my left eye. The surgeon who performed the procedures ordered a pathology report because he did not believe I suffered from MPS VI. I must admit that I found his attitude a little disconcerting as I felt it not only called into question my understanding and experience of the disorder but also questioned the ability of the specialists I had grown to admire and trust. In many ways I feel as a result of this experience I am better able to relate to parents who are told nothing is wrong with a child when all the evidence before them clearly points to some problem, health or otherwise, existing.

Luckily for me, since joining Family Support, I have not only participated in but have also facilitated a number of assertiveness and self awareness programs and was not intimidated by such an attitude and was able to overcome this particular surgeon's disbelief. This unfortunately cannot be said for many of the parents I see at work who are easily intimidated by professionals and others who are perceived to be all-knowing.

Hurdle No.7 In December, 1995 I underwent a second heart operation, this time to replace a malfunctioning aortic valve. The heart surgery itself

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went well, however, I did experience some difficulties resulting in my being maintained on a respirator for 7 days. The senior anaesthetist had recognised my considerable fear at the thought of being intubated again whilst still awake and had determined to do the intubation procedure under general anaesthetic. Unfortunately, a misunderstanding arose and a larger diameter tube was inserted than had been intended. The consequence was that the lining of my throat was torn. When removal of the tube was attempted I bled into the left lung, the original error having been exacerbated by a failure to check the clotting factor of my blood, prior to my being given an inappropriate amount of warfarin on top of the already administered heparin. This experience was very frightening, not only for me but also for my family, and has clearly demonstrated to me that in future I should be intubated awake. I have made every effort to ensure that all doctors who deal with my medical needs are aware of this and I also carry with me the necessary information to ensure the correct procedure is carried out in the event of an emergency. Hopefully this type of error will never be repeated.

Conclusion

To date I have managed to meet, and conquer, the hurdles that Maroteaux-Lamy Syndrome, as well as other life experiences, have placed before me. At times it has been difficult but the continuing care and support from expert medical teams along with emotional and physical support from family and friends, including members of the MPS Society (past and present) and work colleagues has made the whole experience a little easier for me.

I feel that in my capacity as a Family Worker I have reached my intended goal. I now work in a position which allows me to utilise my counselling training along with my life experiences to assist families who are experiencing difficulties in a number of areas, for example, difficulties with parenting, life skills, separation, bereavement etc... Such difficulties may arise because of a number of reasons such as isolation, economic circumstances, domestic violence, to name but a few. Although this work can be physically taxing at times, on the whole I have found it to be very rewarding.

I consider there to be only two hurdles facing me as I look to the future. Firstly the impact of the uncertainty factor inherent in all MPS syndromes, that is to say, not quite knowing what will happen next. If some difficulties arise will they be operable? I am currently awaiting an appointment with a neurologist to investigate what appears to be wasting in the muscles of my hand. Also, continuing degeneration of my hips is currently being monitored by an orthopaedic surgeon with a view to possible hip replacements at a later stage.

Secondly I still have to search out various specialists who only treat the individual aspects of my condition. It has long been my hope to one day locate a physician who knows enough about MPS VI, and the various aspects of the syndrome, to oversee the ongoing management of my medical

OVERSEAS NEWS

needs, although I have had no success to date. As a result of a recent scare involving a car accident my cardiologist has offered to have his name and contact number engraved on my medi-alert identity bracelet, along with those of the Royal Prince Alfred Hospital emergency numbers.

It is not my intention to portray my future living with MPS as bleak. On the contrary, in the very near future I will be branching out into private practice as a registered psychologist and feel that my experiences as a result of having MPS have helped me develop as a therapist and will certainly be of benefit to me in relating to, and consequently assisting, anyone who comes to me for counselling.

Catherine Hartcher, Australia

(We will have a another life story from Australia in our next newsletter)

As you will see at the front of Newsletter we have noted our Home Page on the World Wide Page.

Address : ***http://www.vois.org.uk/mps***

If you have the opportunity please browse through our pages and let us know what you think of the information or tell us of any ideas you have.

OVERSEAS NEWS

A Mother's Plea (from Lithuania)

I attended the Genetic Centre, in Vilnius with my daughter in 1977. they assured me then that I would not have another child with such a disease. there were no other known cases of the disease in our family, although there were no thorough genetic tests carried out.
I can see that my boy is developing in the same way as his sister. The difference is that he is stronger and can be more aggressive.

I enclose Romas's photograph hoping that it will help find the precise diagnosis.
My second son is healthy, he was born in January 1979.

My son Romas was born on the 22nd of September 1986. He is the third child in the family. The delivery/childbirth was normal. I noticed that my child was not keeping up with the normal development patterns at the age of 18 months.

Mucopolysaccharide was diagnosed, the same diagnosis that had been given to his sister, Ilone, earlier. There were no bio-chemical tests carried out.

Ilone was born on the 28th January 1972 and died at the age of 12 years in 1984.

Until the age of 7 years Romas developed some language, he could say his name, surname and other people's names. He could express some thoughts using separate words, usually about his brother on a motorcycle. He then began to regress. he now cannot eat independently and he has no speech, not even Mum. There are a lot of behavioural difficulties, sometimes he becomes very angry and aggressive, I think this may be due to some pain he suffers. Sometimes he experiences, what appears to be like a fit of epilepsy, when his body becomes flexed, he rolls his eyes upwards and makes crying noises. His sleep patterns are usually disturbed, with him waking frequently, medication on irregular basis. His head is hydrocephalic (57cm). His gait is awkward and spastic at times. He does not indicate his need to use the toilet and urinates frequently. He still has some general understanding. He responds to the attention of being called by turning to his name or smiling. Generally you can take him by the hand and lead him to the place you want to go.

Mrs Naluzis.

As you will have gathered from Romas's mother's story she still is unsure of a diagnosis but she would be very grateful if parents, who recognise these symptoms and who have experience in dealing with their own children, would write to her. She feels very isolated and would welcome any replies to her story.

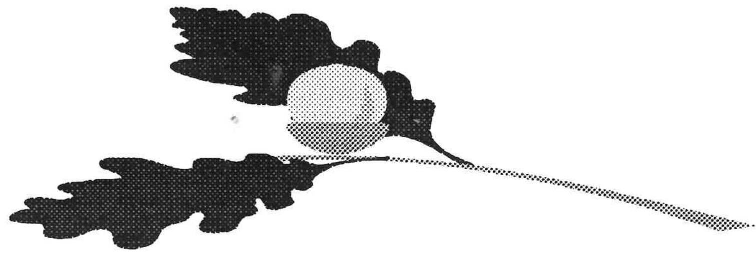
Her address is Mrs Naluzis, Poising.41-24, 5804 Klaipeda, Lithuania.

CHILDHOOD WOOD

MPS TREE PLANTING

SHERWOOD PINES

25th OCTOBER 1996



DANIEL JOHN ANTHONY

PAUL BEAN

OLIVE DIANE BULLOCH

ROBERT ANDREW BUTLER

LOUISE BYRNE

AMANDA CORCORAN

WILLIAM CORCORAN

JAMES GOOCH

MARK OLIVER HARVEY

VICTORIA SUSAN JOHNSON

SARAH ANNE KILVERT

ASHLEY MARTIN

BERNICE MARIE-FRANCE POUECH-SHOOTER

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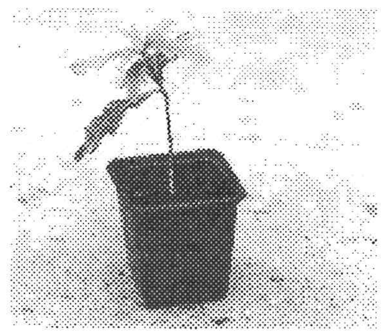
ALISTAIR JAMES REID

DARREN EDWARD TAILFORD

RHIANNETH LOUISE WHEELER

KATIE HEADLAND



FUNDRAISING

On Friday the 12th of July 1996 twelve friends and I took part in a sponsored Granny Night. Our outfits came from charity shops and we really did look the part. We visited two pubs and one night-club and managed to raise £335.50 through sponsorship alone. We thoroughly enjoyed ourselves by dancing, talking and acting like old grannies.

We chose MPS because we work with Mrs Zelda Hilton who has a 9 year old son, Shane who suffers from Hunter Disease.

May I take this opportunity to wish you the best in all the work you do.

Sarah Carter

Above: Some of the girls presenting the cheque to Mrs Zelda Hilton

Opposite: Sarah Carter with Shane Dickson aged 9 years who suffers from Hunter Disease.

Below: The Grannies in all their splendour.

FUNDRAISING

RAISING MONEY FOR THE MPS SOCIETY THROUGH MY EMPLOYER

I have worked for 3i plc, an investment capital company, for 6 years and last year I found out that the company administers something called Give As You Earn for those employees wishing to use it. Give As you Earn or 'G.A.Y.E' is run by the Charities Aid Foundation and is a tax free way of giving an amount of your choice to a charity of your choice straight from your salary. I know that not many people in my company are aware of this service and I guess people in general all over the country are not aware of it.

My company also has a charities committee called 3i Charitable Trust from which donations are made to a variety of causes. The 3i Charities Committee has a policy of matching the amounts donated through G.A.Y.E and therefore the £20 I donate out of my salary each month to the MPS Society is doubled to £40! I only wish I had known about G,A.Y.E earlier to take advantage of it, especially as my company is so generous.

Earlier this year I appealed to the 3i Charitable Trust for a donation to the MPS Society. I sent a memo explaining all about the Society and the work it does and how it greatly needs more money to support the families and for vital research into the diseases. I later received a cheque for £500 from the Committee for the Society.
My colleague, Natalie Reeve, plays in a Ladies Bowling League which holds a sponsored bowls match every year. The money raised is shared between 3 charities. Thanks to Natalie the MPS Society has been one of the recipients for a few years in succession. Natalie goes to great effort to raise as much sponsorship as possible to increase the amount that is donated to the MPS Society. This year Natalie appealed to 3i Charitable Trust to match the £300 she raised. They agreed which meant that a total of £600 was raised by Natalie's efforts alone.

Sharon, Daniel's mother, told me about the topics discussed at this year's MPS annual conference. Following this I realised that it is not only important to raise money for MPS families now but for families and MPS children of the future.

Everyone who meets Daniel falls in love with his sparkling personality. He brings so much love and happiness to our family and I feel grateful for all the time we have with him.

Joanne Coleman, Birmingham

I am Daniel Allen's Auntie from Birmingham. Daniel suffers from Sanfilippo A and is now 7 years old.

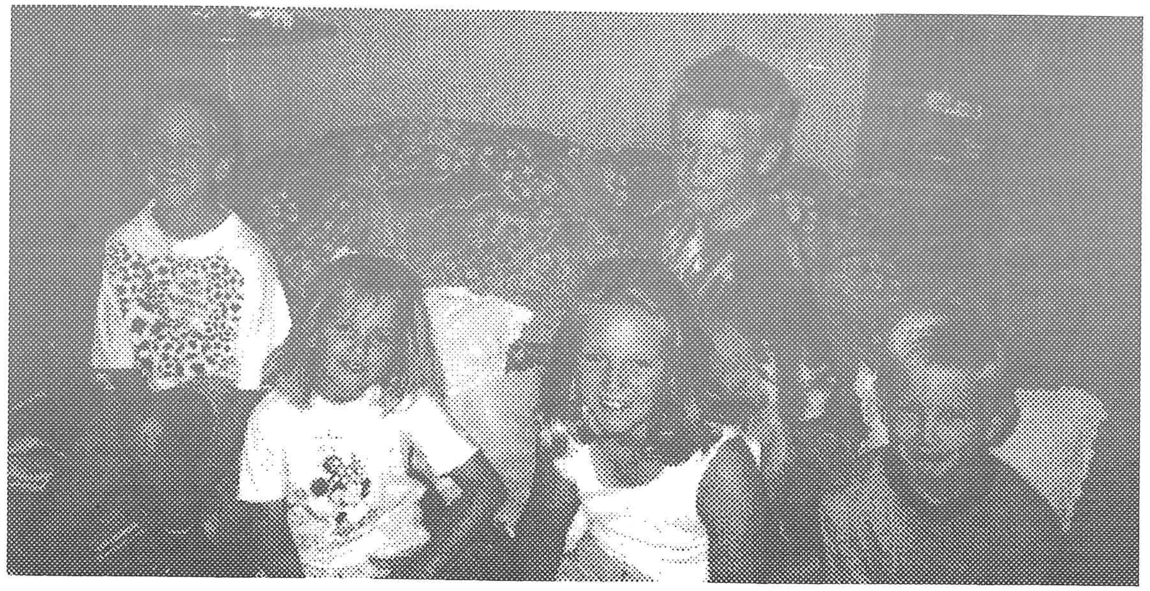
If you are interested in Give As You Earn, they can be contacted on -0891 42 42 44
Give As You Earn, Foundation House , Coach and Horses Passage, The Pantiles, Tunbridge Wells, Kent TN2 5TZ

FUNDRAISING

Pictured opposite is Deborah Thorburn who raised £75.00 especially for the MPS Society by holding her own Jeans for Genes Day.
Deborah who suffers from Sanfilippo Disease is 17 years old.

Pictured below are James, George, Kathryn, Alice and Kristy who are all friends of Toby and Hannah Parfitt. Toby who is ten years old suffers from Hurler/Scheie Disease.

The children collected £20.00 for MPS at Halloween when they went Trick or Treating. We would like to say a big thank you to this group of children who have donated their collection to the MPS Society for the second year.



FUNDRAISING

Holly and Edward Nowell assisted by their daughter Mary Ellen and their son, Edward organised a Charity Gala Recital in aid of the MPS Society. The Recital was held in Wells Cathedral on the 2nd of November 1996.

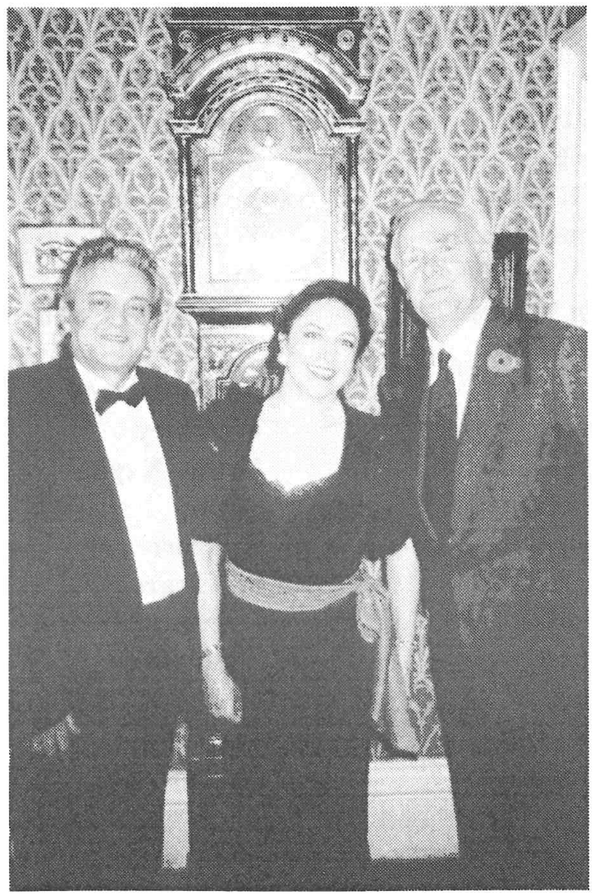
Susan Daniel, an international singer living in London and Milan and has sung leading roles at the Deutche Opera, Berlin, the Munich Stattsoper, the Vienna State Opera, La Scàla, Milan and the Paris Opera generously supported MPS by giving this special Gala Recital.

She was accompanied by **Roberto Negri**, from Milan who works regularly at La Scala and gives concerts throughout the world.

The Recital was followed by a candlelight supper. In the interval the famous author, **John Le Carre**, gave a talk on MPS.

We would like to thank the Nowell family, Susan Daniel, Roberto Negri and John Le Carre on behalf of the MPS Society for this wonderful fund raising event which to date has raised over £26,000.

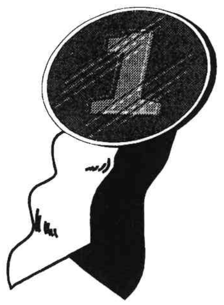
Pictured above is Susan Daniel taking a bow.



Opposite are Roberto Negri, Susan Daniel and John Le Carre.

Below are pictured some of the guests at the supper following the Recital.

FUNDRAISING



WINNERS LIST- NATIONAL DRAW 1996

1st Prize **5 Day P & O Crossing to Europe for 2 + £500 Cash**	**Philippa Power**	**London**
2nd Prize **Meal for 2 at The Grill Room-Cafe Royal Trust House Forte + £100 Cash**	**A Marshall**	**Caldicot**
3rd Prize **4 Tickets to Alton Towers + £50 Cash**	**Chris Kembrey**	**Bristol**
3 Family UCI Cinemas	**M. Davies**	**Pontyclud**
Kenwood Coffee Maker	**Mike K.**	**Steetley**
MacDonalds Meal vouchers x 6	**R.P. Gillam**	**Borehamwood**
Wall Clock	**Michelle Stanley**	**Whitney**
Silver Goblets	**Mrs V Brockle**	**Alcester**
Marks & Spencer - £5 voucher	**Dawn McNeill**	**Elmbridge Council**
Board Game	**Jean Poole**	**Sheffield**
Barbie Doll	**Peter Stuart**	**Borehamwood**
Barbie Doll	**Candice Powis**	**Wembley**
Sweatshirt	**Pauline Lawson**	**Milton Keynes**
Sweatshirt	**Derek Izzard**	
T-Shirt	**E Shore**	**Runcorn**
T-Shirt	**H McArther**	**St. Clears**
Hand Knotted Doll	**S Hood**	**Overmonnow**
Cuddly Toy	**B Elliott**	**Sheffield**
Ornament	**Gilbert**	
Pogo Stick	**Mrs Byrom**	**Sale**
Mills & Boon books	**C. Campbell**	**Greenford**

FUNDRAISING

DONATIONS

The Society is grateful to the following who made donations.

Mrs Moody
Mr S Morrison
Harrison & Clough
Gary and Joanne Adshead
Karen and Wayne Hoather
St John's Koala Club
John and Barbara Arrowsmith
Mr Stanmore
Miss E Jenkins
David Colclough
Mrs D Small
Mr De Matos
Dorothy Duckett
Mr and Mrs Taylor
Al Fayed Charitable Trust
Hilltop Church
Iris Weeks
Bakers Dolphin Holidays
Benham Charitable Settlement
Monmouth School
Dexion Ltd
Mrs Longhorn
Mr M Boudewijn
Mr B Minney
Express Newspapers plc
Gleeson Group
Fenwick
Mr and Mrs Knox
Natalie Pidden's Grandmother and Aunt
Mr and Mrs Humphreys
Rachel and Mark Wheeler
Michael R Barnett
Kings Heath Ladies Bowling League
The Astor Foundation
Mrs A Johnston
R.E.M.E. Workshops
Jackie and Barry Perfect
Japanese Women's club
Mrs Ward
Blair Foundation
Fitton Trust
Joseph Strong Frazer Trust
Mr and Mrs Hayward
High Wycombe Sports Association
Bellway homes Ltd
Mrs I K Bray
Clifford and Lella Fountaine

FUNDRAISING EVENTS

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

Fer Pidden - Sale of Jewellery
Haddenham Mummers - Play
Crosby Hotel, Scunthorpe - Bike Day
Paul and Tracey O'Garr and Staff, Scunthorpe - Lorry Pull
Mrs Brockie, Alcester - Raffle
John Byrom , Cheshire - North West Abseil

FUNDRAISING

Glynis Marshall, Newcastle upon Tyne - North West Abseil
Gary and Joanne Adshead, Manchester - Book Fair
Anne and Mike Kilvert, Wales - Clive Motorcycle Club Fundraiser
Mr and Mrs Westland, Reading - Bits and Pieces Sale
Clive and Jackie Chisling - South West Xmas Raffle
John and Barbara Arrowsmith - Xmas Raffle
131st Glasgow Boys Brigade, Glasgow - Coffee Morning and Jumble Sale
Tony Lockyer, Pontypridd - Raffle
Littlemoor Cycle Club, Monmouth - Coast to Coast Cycle Run
Edward Hurley, Newhaven - Collection at Sainsbury Store
Julie and Chris Kembrey, Kingswood - Second Severn Crossing
Mrs Dawn Nelson, Petersfield - Small Change Collection
Caroline Fisher, Saffron Walden - Sherry and Mince Pies Open House
Rachel Todd, Bushmills - Praise Service
The Perse School, Cambridge - Charity Christmas Sale
Holly and Edward Nowell and Family - Gala Recital in Wells Cathedral
Val and Mick Denyer, Cranleigh - Bazaar
Rachel and Mark Wheeler - Raffle
Cathy Flaig, Broadstairs - Sponsored Slim
Bill and Sylvia Blackburn - Xmas Raffle
Pam and Ken Ballard, Harrow - Sale of Goods at MPS Conference
Mrs A E Johnston, Tunbridge Wells - Knitting Bedsocks
Mrs K Jordan, Tunbridge Wells - Fundraising Lunch
Callagham Children, Somerset - Trick or Treating
Kieran and Bernie Houston, Stabane - Golf Tournament
Mrs V Brockie, Alcester - Sale of Pens
Christine Gooch and Pam Hodgkins, Tunbridge Wells - Sale of Baby Clothes
Andrew and Karen Weedall, Runcorn - Win on National Lottery
Mrs Goscombe, High Wycombe - Charity Auction
Carol and John Westland, Reading - Music Afternoon
Pauline and Sean Mahon, Sheffield - MPS Dinner
British Army, Andover - 3 Peaks Challenge
Lorna Young and Jean Ferguson, Glasgow - Glasgow Half Marathon
Mrs Foster and Mrs Baker, Bristol - Car Boot Sales
Mr G Laing, Dundee - West Highland Walk
Stonelaw High School PTA, Glasgow - Raffle

CHARITY BOXES

Mrs Brockie. Oversley Mill Service Staion, AlcesterSid Shiff and Family, Hunts Cross

Mrs Kirkpatrick, Antrim Daniel Croghan's Grandparents, Stockport

FUNDRAISING

DONATIONS IN MEMORY

The Society is grateful to the friends and relatives of:

Neil Palmer Gethin Robins Richard Mort

Florence Mitchell Daniel Anthony Mrs Doreen Rock Joh

Margaret Barnes (Alexander Butler's Grandmother)

STAMPS

Molly Griggs Letitia Ricketts Croydon Vehicle Registration Office Bailey Family

Mr and Mrs Tiley

We would like to thank all those who sent us stamps but did not include their names.

We have included various forms with this Newsletter and would really appreciate the prompt return of completed forms to the Amersham office as soon as possible.

We hope you all enjoy the new package for the conference and hope that the information will encourage families who have not previously attended the conference to consider coming this year.

Area Support Families

East Anglia

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

South East

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

Michael and Karen Wheeler — Tel: 01483 826135
'Gildon', Ballegers Lane, Horsell, Woking, Surrey GU21 4SB

Potteries

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

South West

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

Gordon and Anne Hill — Tel: 01395 567735
8 Hacker Close, Newton Poppleford, Nr. Sidmouth, Devon

Home Counties

Mark and Rachel Wheeler
26 Tamarisk Avenue, Reading, Berkshire RG2 8JB — Tel: 01734 861063

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

Wales

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

Jackie Edwards — Tel: 01222 756846
13 Heol-y-Carw, Thornhill, Cardiff, CF4 4EU

Area Support Families

Midlands

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

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

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

Yorkshire and East Coast

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

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

North West of England

Martine and John Brennan. — Tel: 01524 382164
105 Barley Cop Lane, Lancaster, Lancashire LA1 2PP

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

North East of England

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

Ann and Ron Thompson — Tel: 01325 489920
7 Sunningdale Green, Darlington, County Durham DL1 3SB

Scotland

Alan and Fiona Byrne — Tel:0141 5695376
3 Jedburgh Avenue, Rutherglen, Glasgow G7 3EN

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

Northern Ireland

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

Margaret Kearney (Secretary) — Tel: 0126 5762073
12 Coleraine Road, Ballycastle, County Antrim BT54 6DU