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SYNDROMES

EDITORIAL

GILL SMITH
Superintendent Paediatric Physiotherapist

This issue deals with case studies of children with unusual syndromes. We are aware that we are all being referred many more children with complex difficulties, having a diagnosis that we may only read about in a medical reference book with little or no text on physiotherapy management. Thank you to all the authors who have contributed. If any members are currently involved with children with other rare syndromes, please send us your thoughts, ideas and experiences to be included in future issues.

May I also take this opportunity to thank Jill Brownson and Fiona Corkhill on behalf of all the members for the excellent document produced in collaboration with SCOPE. If you have not already seen this document, please take the time to seek it out and use it.

We have welcomed to the Editorial Board, Angela Glyn-Davies (formerly APCP Treasurer) Judith Morrison and Sally Braithwaite, who is 'learning the ropes' before taking over from our current editor Lin Wakley, who deserves a well earned rest!!

Finally, please continue to send any letters, articles to be included in future issues.

Copy for the

DECEMBER 1999 JOURNAL

must be with the editor by

1st NOVEMBER 1999

The editorial board reserves the right to edit all material submitted

IF POSSIBLE, PLEASE SUBMIT COPY ON FLOPPY DISC IN WORD 6 FORMAT, TOGETHER WITH A PRINTED COPY.

LETTERS TO THE EDITOR

Carol McKay
Green Park Healthcare Trust
Mitchell House School
Marmont
Holywood Road
Belfast BT4 2GU

Dear Lin,

I am a Superintendent Physiotherapist working in a school for children whose physical disabilities prevent them from attending mainstream schools. These children are aged between 2 and 19 years.

Over the past few years, as part of a business plan for additional staff, I have been categorising the pupils in terms of their perceived Physiotherapeutic needs. This process has confirmed the gradual increase in the number of pupils who have complex disabilities, and whose treatment needs are significant and long term. I would be very grateful for information from any other Therapists who are devising a means of categorising the children in their caseload.

We are also carrying out a multi-disciplinary audit into our summer treatment arrangements and would be pleased to hear from other schools how continuity of treatment is maintained over the summer months.

Thank you for your help.

Yours sincerely

Carol McKay

Gill Smith
Superintendent Paediatric
Physiotherapist
Poole Hospital
Longfleet Road
Poole
Dorset BH15 2JB

Dear Lin

Please could anyone help us regarding the advice from Riding for The Disabled and the wearing of riding hats.

A microcephalic child who attends one of our local special schools is unable to wear a hat because of his head size. Our local RDA are asking (see enclosed protocols from RDA) for a written statement from the school physiotherapist, who does not attend the riding sessions.

We do not feel this is an appropriate request, surely the liability for a child attending Riding for the Disabled should be with the personnel concerned at the riding school and not with a school physiotherapist who is not present during these sessions or has any knowledge of the finer points of horse riding!

With many thanks,

Yours sincerely

Gill Smith

Pauline Bateman
Senior Physiotherapist
Bleasdale House School
27 Emesgate Lane
Silverdale
Nr Carnforth
Lancashire LA5 0RG

Dear Editor,

I am a Physiotherapist working in a school for children with profound and multiple problems.

Over the past four years many of our children have had Gastrostomy operations.

The placement of the Gastrotomy tube has caused difficulty with orthotic provision such as corsets, with prone positioning and with elevated post feed positioning.

I fear that at times the management of the gastrostomy may change our physical management of these children and perhaps hasten their physical deterioration.

I would be interested to hear from other centres who are managing this situation.

Yours sincerely

Pauline Bateman

Sarah Tomlinson
Lansdown Health Centre
34 Lansdowne Street
Winson Green
Birmingham B18 7EE

Dear Miss Wakley,

Re: Guidelines for levels of intervention in paediatric physiotherapy.

We are currently looking into developing guidelines for levels of intervention for use within our department. We want to ensure equity across the team and allow ourselves some flexibility to manage our caseloads in the light of increasing levels of inclusion and numbers of children receiving Botulinum Toxin therapy.

We are also considering conducting a client-satisfaction survey.

We would be very grateful to any colleagues prepared to share with us guidelines already in place, or questionnaire formats which have proved useful.

Please contact Sarah Tomlinson, Clinical Specialist, Paediatric Physiotherapy Department, Lansdowne Health Centre, 34 Lansdowne Street, Winson Green, Birmingham B18 7EE

Yours sincerely

Sarah Tomlinson

Kate Beattie and
Alison Carter
Physiotherapy Department
Mayday Hospital
London

Dear Lin

We have just returned from a two-week neonatal course/study trip in Brisbane, Australia. The course was led by Cathy Bagley who was one of the expert witnesses at the 'Inquiry into the provision of Chest Physiotherapy Treatment provided to pre-term babies at National Women's Hospital in Auckland, New Zealand between 1993 and 1994.

We were able to discuss with Physiotherapy colleagues from New Zealand and Australia the recommendations of the inquiry and the implications for our practice. The recommended lessons for all health professionals were :-

1. **Peer Review**
   All health professionals who have responsibility for patient care must have effective and regular peer review. This should include a regular review of their clinical and practical skills under direct observations by peers of equivalent or senior competence.
2. **Adequate Information of Outcome**
   Parents need to be informed of the risk of adverse events.

3. **Details Clinical Record Keeping**
4. **Parental Consent in Neonatal Intensive Care Units**
   A committee should confer to provide a standard approach to this.
5. **Consent to Training**
   The issue of staff training as different from student training needs to be clarified.
6. **Clarification of Publication of Internal Audits and Ethics**
7. **Patient Advocacy Services**
   This role should be strengthened.
8. **Research in Neonatology**
   Ongoing neonatal audit and research is to be encouraged and supported.

We feel that for Physiotherapists, especially those working in isolation, the key recommendation is that of peer review. There obviously needs to be further discussion nationally as to how this could best be established and we would be interested in hearing from anyone working in this field.

Yours sincerely

Kate Beattie and Alison Carter

Deryn Watts MCSP
Head Physiotherapist
Tadworth Court
Tadworth
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KT20 5RU

Dear Colleagues

I am interested to hear from anyone who has taken long term ventilated children swimming.

Although I have taken a variety of children with a tracheostomy for hydrothereapy, I have not yet taken a ventilated child into the pool. I would be interested to hear of other peoples' experiences in this area and know of the guidelines that have been set up for these two groups of children.

Yours sincerely

Deryn Watts

Clair Culligan
Senior Paediatric
Physiotherapist
Wirral Hospital
Physiotherapy Dept.,
Elleray Park School
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Merseyside
CH44 0LH

Dear Miss Wakley

I am currently planning a leaflet to distribute to new parents including a warning of the problems associated with the use of baby walkers and also the importance of playing with baby in the prone position. We are finding that many parents are not playing with their baby in prone believing they have to be on their backs when awake and asleep!

I wondered if anybody had knowledge of any such leaflets in circulation at present and if so would be grateful to hear from you.

Yours faithfully

Clair Culligan

LETTERS TO THE EDITOR

Rebecca Pearce
Senior Paediatric Physiotherapist
Wordsworth House
121-123 Tremona Road
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SO16 6HU

Dear Miss Wakley

Iwould be very grateful if this letter could be printed in the next available journal.

I am a physiotherapist working as part of the community paediatric physiotherapy team in Southampton. I am currently looking into the possibility of clinical use of the gait lab at Southampton General Hospital for some of our children with cerebral palsy. The lab is at present used as a research facility but has scope for clinical use. I would welcome any information anyone may have which could be useful in my quest. In particular I would like to hear from those physios whose children have received gait analysis to find out how they access this facility and how the process is funded.

If anyone has recently embarked on a similar exercise I would welcome any advice on how to do this successfully, especially from a business point of view.

Any guidance or information would be very gratefully received.

Yours sincerely

Rebecca Pearce

Gwyn Owen MSc MCSP
Professional Adviser
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Dear Editor

Motions 18 and 19, carried at ARC in May, ask the CSP to investigate the incidence and the impact of :

1. strain injuries to the joints of the hand (motion 18)
2. work-acquired infectious diseases (motion 19)

which physiotherapists may be at risk of in the course of their occupation.

Studies and local experience indicate that manual therapy techniques pose a risk to physiotherapists in terms of developing painful joints in the hand and wrist for example. Given the repetitive nature of other physiotherapy techniques, it is possible that this situation is not limited to other areas of practice (respiratory care, neurology and paediatrics for example).

Anecdotal evidence suggests that physiotherapists are coming into contact with increasingly diverse and resistant strains of infectious diseases as a result of their work. Infection control policies are in place, although these vary across the country, and not all workplaces will adhere to them.

In order to develop an indication of the extent of these issues within the profession, I am writing to ask your readers to contact me if they have experience of either of the above issues. The information obtained will be held in confidence and used to gain a picture of the situation, before considering what further work is indicated.

Yours sincerely

Gwyn Owen

LETTERS TO THE EDITOR

Gwyn Owen MSc MCSP
Professional Adviser
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Dear Editor,

Motion 32 carried at ARC earlier this year reads

Conference believes that the CSP should have a more proactive role in the national health education agenda. We call on Council to press the Government for formal inclusion of practical health education in the national curriculum with the appropriate part of the syllabus INITIATED by Chartered Physiotherapists.

This motion raises a number of further questions for consideration:

- is there evidence supporting the effectiveness of school health education in preventing illness and disability?
- what resources (staffing, time and training for example) will be needed to develop health education programmes in schools?
- are there any practical examples of the process of introducing health education progammes into schools at a local level, the difficulties experienced, and assessments of the outcome on children's behaviour?

In order to organise an effective campaign to lobby for change, viewpoints needs to be backed by evidence. I am therefore inviting any readers who have views on the above questions and/or experience of introducing practical health education into schools to contact me. The information gathered will be submitted to Professional Practice Committee in October. This committee can then consider what future action is indicated in light of this evidence.

Yours sincerely

Gwyn Owen MSc MCSP

Mrs Emma Isworth MCSP
Mrs Melanie Tilman MCSP
Senior Physiotherapists
(Paediatrics)
Esat Kent Hospitals
Physiotherapy Dept.,
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Dear Madam

We are a small team of Community Paediatric Physiotherapists covering a large geographical area of Kent.

We are seeking information from anyone who has experience in the treatment of children with Osteogenesis Imperfecta. We are especially interested in the management of these children from a prophilactic point of view. Is there any evidence that exercise programme with muscle strengthening and joint mobility reduces the incidence of fracture and deformity??

We would welcome any information on this subject.

Yours faithfully

Mrs Emma Isworth MCSP

Mrs Melanie Tilman MCSP

LETTERS TO THE EDITOR

Sue Booth
Senior Paediatric Physiotherapist
129 Rutherford Drive
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Dear Lin,

I was wondering if any of your members could provide insights and answers on a problem which I am sure plagues many of our paediatric physiotherapy services . . . namely 'D.N.A.'s. My colleagues and I have just spent another trying summer sometimes sat waiting for patients at our community base who have failed to turn up; fortunately there is always paperwork to be done, but this phenomenon does dog the efforts of all those wanting to run an efficient service, it is also very unfair to families who do attend and lastly those slots missed could have been allotted to others on the waiting list.

I know that in many adult services 3 consecutive D.N.A's result in a discharge, but can this be carried over into the paediatric setting?, it does seem rather a case then of 'visiting the sins of the fathers on the children' . . .

I look forward to hearing from any of your members on this matter.

Yours sincerely

Sue Booth

CHARCOT-MARIE-TOOTH SYNDROME

MARIAN MULCAHY PhD
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Charcot-Marie-Tooth (CMT) neuropathy, also known as heriditary motor and sensory neuropathy (HMSN), is a heterogeneous group of inherited diseases of the peripheral nervous system. It is a common disorder, thought to affect 1 in 2,500 individuals, although precise prevelance rates are difficult to determine because of the heterogeneity of the syndromes. Phenotypically, patients with this disorder may present with pes cavus, tendon areflexia, distal limb weakness and distal sensory loss.

This range of neuropathies can be categorized by measuring the nerve conductance velocities (NCVs).

CMT 1 refers to individuals with a hypertrophic demyelinating neuropathy, detected by 'onion bulb' formation, and with reduced NCV.

Individuals with CMT 2 typically have normal NCVs but have an axonal abnormality.

There are two other related neuropathies, Deferine-Sottas disease (HMSN III) and hereditary neuropathy with pressure palsies, HNPP).

For many neuromuscular disorders the chromosomal location, and causal gene have been identified. Studies of mice with the trembler (Tr) mutation, which appeared to cause peripheral demyelination served as a useful model for Charcot-Marie-Tooth type 1. Mapping studies revealed that in approximately 70% of CMT 1 individuals there was a duplicated of a region of chromosome 17 (17p 11.2), and that this contained the gene which encodes peripheral myelin protein 22 *(pmp-22)*, (See Table 1).

Table 1.

Charcot-Marie-Tooth Neuropathy Type 1 - Genetic Information

	LOCUS	GENE	MUTATION
CMT 1A	17p11.2-12	*pmp22*	Duplication/Point
CMT 1B	1q22-23	*po*	Point
CMT 1C	??	??	??
CMT X	Xq13.1	Connexin 32	Point
CMT 4A	8q	??	??

Interestingly, CMT 1A is caused primarily by an excess of PMP22. Recent studies of both the duplication and the point mutation in the *pmp22*, have demonstrated an accumulation of PMP22 intracellularly. Thus these cells have an impaired trafficking mechanism for this protein. Individuals with a 17p11.2 deletion suffer from hereditary neuropathy with liability to pressure palsies, HNPP. 84% of HNPP patients carry this deletion. It would appear therefore, that the *pmp22* gene dse is critical to healthy nerve development.

The underlying genetic defects are now known for the many neuropathies, including those discussed here. This has important implications for both diagnosis, and genetic counselling for individuals at risk from these disorders.

A Personal Case Study

I suffer from CMT 1A. As a baby I was described as 'floppy', lacking in muscle tone. At 2.5 years a diagnosis of 'muscle amyotonia' was reached. My younger brothers helped teach me to walk unaided, something I achieved at 4.5 years old. Two years later I underwent surgery to correct both dislocated hips. This was repeated on the right hip after another two years. Luckily for me, I went to a school for children with disabilities, so trips into hospital, to physio, to hydrotherapy etc were very much part of normal school life for all of us. We were always encouraged to be both as independent as possible, and also mindful of others' needs. It was a wonderfully supportive and enabling environment to grow up in.

Shortly after entering mainstream secondary education, I again went into hospital - this time for spinal fusion to treat scoliosis. Further treatment proved necessary after another three years, when I was fitted with a Harrington rod. Again I was very lucky to have had lots of academic support from my family and from the school. So although I missed a considerable amount of time from the classroom, I never fell behind my contempories - something which was very important to me.

As an adult I enjoyed a successful career as a research scientist until I gave up to work to take care of my family. My husband and I have 4 children, and as befits a geneticist, we have 2 girls and 2 boys, and 2 have the 17p11.2 duplication, the dominant mutation being inherited as one would predict!! Thankfully both children are fit and able, much more so than I was at their age. They are under the watchful eye of a pedriatician and of a physiotherapist who visits their schools.

I am confident that all of our children will be able to live full and active lives, regardless of the number of *pmp22* genes they have!!

HEREDITARY MOTOR & SENSORY NEUROPATHY

A CASE REPORT

NUALA BYRNE
Senior Paediatric
Physiotherapist (PICU)
St. Mary's Hospital, London

Nuala is now working as Clinical Specialist for Cystic Fibrosis at Newcastle Royal Infirmary.

A set of six-month old twins was referred to the PICU retrieval team with respiratory distress. They had a one-week history of coryzeal symptoms, irritable weak cough and poor feeding. Their GP prescribed oral amoxycillen but no improvement occurred so he referred them to the local hospital, to which they were admitted. Initially they were self-ventilating with nasal cannula oxygen, but over the course of the day developed increasing respiratory distress and oxygen requirements. They were retrieved to PICU at a specialist centre and both required intubation and ventilation.

Past medical history:

They were born at 38 weeks gestation by elective Caesarian Section for intra-uterine growth retardation. Birth weights were approximately 2kg. Neither required SCBU and so were discharged home. From a developmental point of view, mother reported that they had good head control, could roll side to side and sit with some support.

On admission naso pharyngeal aspirates confirmed that both twins had RSV+ bronchiolitis. It was also noted that they were significantly underweight (5kg), had bilateral planterflexed feet and flexion contractures of the 3rd, 4th and 5th digits of both hands. It was not possible to carry out any further assessment of their development at this stage as they were sedated and ventilated.

A broncho alveolar lavage was carried out but no abnormal growth was detected. One of the twins was initially extubated and transferred to the paediatric ward but over the course of two weeks required re-intubation on two occasions. The other twin failed extubation on several occasions. This was due to increasing respiratory distress despite non-invasive support with CPAP and negative pressure ventilation. Developmentally, they had regressed and were no longer able to roll or sit with support. Muscle power was in the range of grade 2-3 on the Oxford scale.

Mother then revealed that she had a previous baby who died in Nigeria aged 9 months from a chest infection. This child had the same hand and foot deformities as the twins.

Both twins had tracheostomies formed to facilitate long term ventilation with BIPAP. Nerve and muscle biopsies confirmed a diagnosis of Hereditary Motor Sensory Neuropathy with a probable classification of Type III (see Appendix). The prognosis was felt to be 3 years but that possibly they would not survive to 1 year of age. They were transferred to the general paediatric ward for long term care.

Physiotherapy input

Physiotherapy input for these patients mainly involved respiratory assessment and treatment in early stages. Long term, they intermittently

developed lobar collapse and required treatment. Input was also focused on developmental therapy. They were regularly assessed to monitor level of skills and any deterioration that had occurred. Appropriate seating was supplied and hand splints were made in order to maintain and improve range of movement of their contracted fingers.

Conclusion

In conclusion, these children were admitted to PICU with a viral respiratory condition which then exacerbated and brought to light an underlying more serious disease process. Their mother is a refugee and currently has no permanent housing, therefore making it difficult to discharge them into the community. Their prognosis is poor and they continue to deteriorate slowly.

Appendix

Literary searches reveal a large number of recorded supposed new variants within the group of conditions broadly classified as hereditary motor and sensory neuropathy.

Hereditary motor and sensory neuropathy (HMSN) – sometimes known as Peroneal Muscular Atrophy or Charcot-Marie-Tooth Disease.

This describes a group of conditions which are superficially similar, but may be otherwise classified by Type I/II /III.
Types I /II are inherited as autosomal dominant conditions.
Type III is inherited as autosomal recessive; it is less common and develops very early in life, sometimes making babies floppy in infancy and giving rise to delayed motor development. In this condition there is almost no myelin wrapped around the peripheral nerve fibres and conduction of electrical impulses in the nerves is extremely slow.
(Harding, 1993)

Dejerine–Sottas disease (DSD), also called Hereditary motor and sensory neuropathy type III, is a severe infantile-onset demyelinating polyneuropathy syndrome. (Keller & Chance, 1999).

An article by Thomas, Claus & King (1999) demonstrates that there may be further variations. They describe what might be a new variant of autosomal recessive HMSN II, presenting as limb weakness and severe distal sensory loss, leading to prominent mutilating changes. Electrophysiological and nerve biopsy findings indicate an axonopathy.

References

Harding, A. (1993). Hereditary Motor and Sensory Neuropathies. *Muscular Dystrophy Group Factsheet.* HE1, 1-6.

Keller M.P., Chance P.F., (1999) Inherited Neuropathies : From Gene To Disease. *Brain Pathology* 9(2), 327-41.

Thomas, P.K., Claus, D., King, R.H. (1999) Autosomal recessive type II hereditary motor and sensory neuropathy with acrodystrophy. *Journal of Neurology.* 246(2),107-12.

A CASE STUDY

TESSA SHUTTLEWORTH
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The physical signs of this syndrome are very apparent in the distinctive features shown in the child's face and the generalised low tone, that is combined with normal postural reactions. This gives us an unusual challenge in developing and retaining the child's physical skills.

Clinical Features

The facial features are apparent from birth, but become more pronounced with age. The forehead is high and rounded. The occiput flattened. The hair is sparse, with areas of alopecia that increase as the child gets older. The ears are low set. Ophthalmic malformation can be present. There is a high flat bridge to the nose with flaring of the nostrils and a prominent upper lip. The mouth is prominent and the tongue size increases the protrusion in time, there is a high arched palate.

The neck is short with a symmetrical trunk and oval chest. The limbs can be disproportionately short.

A characteristic is the coarse skin which shows patches of depigmention.

Hypotonia, epilepsy and profound learning difficulties are particular features.

These are features that we need to be aware of in surviving children with Tetrasomy 12p Pallister- Killian Syndrome.

Children with diaphragmatic defects do not survive.

History

A Ceasarian section was performed after spontaneous labour stopped. Birth weight was 9lb.1oz. Respiration did not start for 4 minutes. Fits started immediately and were controlled. Medication is still required. He has drop fits.

He was a very quiet and still baby who at three months was not showing movement. At six months he had meningitis which caused a fifty three minute fit. A CT scan showed a small brain for his age.

His physical development has been slowly achieved with plenty of encouragement.

Physical development

At nine years of age this boy has normal movement that is profoundly affected by his low tone. He has always had to be encouraged to stretch his abilities. His development was delayed, but followed a normal pattern,

slowly achieved. He shows appropriate saving reactions, no tactile defensiveness and or startle reflex. His movements are symmetrical and he is hypermobile, which is consistent with his low tone.

His chest is oval with straight spine. He has no discrepancy in leg length, but the left hip shows signs of subluxation on X ray.

Head control. His head control is good, in spite of tipping his head back to look at objects and to assist his reduced swallowing ability.

Sitting. Good sitting balance is well established. He sits on a stool totally unsupported. This made it a difficult skill to acquire because he searched for anything to lean on. When using a bench with pelvic and knee support he leant over to put his head on to the bench.

Standing. This has needed encouragement to maintain an upright posture when standing beside an adult. It has helped in his management that he has been able to stand when getting out of a car or transferring to a chair. A standing box has been used to encourage more extensive active use of his legs. Recently he has begun to use gaiters to ensure correct weight bearing and appropriate knee extension. If there is deterioration a prone stander will need to be used.

Walking. His walking with a frame, supervised by adult, was achieved for two years, but this has not been continued. The physical effort is better used now for more effective standing and transfers.

Sight. He is making increasing use of his sight. He tilts his head back to study an object. The pupils react to light. He fixes on a bright object following it from side to side and up and down. He wears glasses though he is distracted by them and takes them off. He shows strabismus and ptosis.

Hearing. He wore hearing aids for some time, but found them distracting, causing him to vocalise loudly.

Sounds. He is at times absorbed in being very noisy for a prolonged period and cannot be distracted. This is very difficult to manage at home.

Feeding. His trunk needs to be well supported when eating, because he will avoid the food presented to him by an adult. He has made no attempt to feed himself though he has the ability to and often puts his hands in his mouth. His low tone is very apparent when eating. He accepts mashed food from a spoon and takes it to the back of his mouth. It can be seen pooling, before he lets it slide down with a barely perceptible swallow, assisted by tipping his head backwards.

He holds a feeder cup and drink appropriately.
Any attempt to increase his diet is not absorbed.
Care has to be taken particularly if he is unwell that he does not choke due to his low tone.

Lung function. He has an effective cough and a clear chest. Chest infections have not been a particular problem.

Conclusion

The difficulty now is that this nine year old boy, having achieved this level of ability over the last three years, is beginning to be affected by his increase in height and his lack of development of muscle and bone to sustain him. The balance in sitting does not seen to have been affected and hopefully this valuable skill will be maintained. He remains active on the floor, getting up to high kneeling and moving across the floor.
He is, I understand far more physically able than other children with this syndrome.

References

Schinzel Albert. Medical Genetics 1991: 28: 122-125.
www.cpdx.com/cpdx/palliste.htm

WOLF-HIRSCHHORN SYNDROME

A SINGLE CASE STUDY

SALLY BRAITHWAITE
Physiotherapy Inclusion
Co-ordinator
Birmingham Children's
Hospital NHS Trust

This syndrome also known as 4p syndrome is an abnormality of chromosome 4, it usually comes in the form of a partial deletion (part of the short arm of the chromosome is missing). However, on occasions there may be a ring formation or a translocation. The condition is relatively rare, and is usually as a result of a balanced translocation in one of the parents.

The syndrome is characterised by multiple components. They are unlikely to all be present in a single child at the same time. They are:-

microcephaly	cleft lip and / or palate
skull asymmetry	low set ears
seizures	short upper lip
epicanthic folds	hypotonia
developmental delay	down turned corners of mouth

marked growth deficiency (of pre-natal onset)

Other possible defects may include:-

cryptorchidism (undescended testicles)
hypospadias
cardiac defects

Frequent respiratory infections are often a major problem.

History

I first took over the physiotherapy care of Sandie nearly four years ago, when she was approximately six years of age. Much hard work had been done with her in the past to maintain muscle length, prevent contracture and deformity, and allow her to have a good postural position for supported sitting, but despite this her head control was poor, she appeared to have no idea of body image, or position in space and her favorite occupation was to lie on the floor and bang her head. There was also a major problem with recurrent chest infections, which were often difficult to manage because of gastro-intestinal reflux and the need for feeding via a gastrostomy tube.

We knew for certain that Sandie had no functional vision or hearing, and was markedly hypotonic, had a slight scoliosis and had neither an effective cough or swallowing reflex. She only just accepted being handled for a short while, and did not seem to really like this, and she had no motor skills on which to build.

Treatment

Our first task was to provide a basic sensory awareness. We started out over the first six months or so to try and develop a situation where handling

was more pleasant for her, and tolerated more readily. This was done by introducing tactile stimulation for the whole body very slowly and gently, always starting with her hands, and following the same pattern of limbs and trunk. Different textures of materials, brushes, oils and lotions were used at first, giving lots of time for the body to adapt to the new sensations, and withdrawing if she became too distressed. Gradually new sensations were accepted, and along with this so were the adults who provided them. It seemed that adults were being recognised by their touch, and as this became more apparent, handling and changing position as long as they were done slowly and firmly and always with maximum support were being more readily accepted, and even welcomed.

Progress

Eight months on into my aquaintance with Sandie, she still only accepted sitting in a Snug Seat, which she was fast growing out of. Postural tone was still on the low side which still left the possibility of continuing to developing postural deformity. Therefore a foam fit type seating system was provided and proved to be an excellent way forward, giving both postural stability and being readily accepted as a primary seating position. Our next goal was to introduce the possibility of sitting unsupported, and this was to take the best part of eighteen months,and was difficult to achieve due to the lack of hearing and vision. A variety of sitting positions both on the floor and with various seats were gradually introduced into a daily routine, always providing necessary although gradually reducing support. Sandie always had to know that somebody was near, a very light touch gradually became sufficient for this. It was necessary to let her learn about her body position in space by experiencing what it was like to move backwards and forwards or side to side and what happened when she moved to far. This is almost certainly experiential learning taken to the highest degree but there was little else to use, she only ever tumbled once in any direction, and learnt her body position in space from this.

By the end of the first twenty months, it was possible to place Sandie in an unsupported sitting position both on a stool with feet flat on the floor and in long sitting. Using basic neuro-developmental facilitation we started to extend these skills over the next two years. Rolling to both left and right and getting from lying to sitting is now possible with just minor facilitation to indicate what is required, and if she is sitting on the floor she can also in a controlled way get herself into lying if she wishes to do so.

Over the last two years, whilst working on spatial awareness in sitting we have also introduced the idea of standing, firstly in a prone stander and several months later in an upright stander. It became noticable after working on supported standing for eight or nine months that Sandie was able, when she was supported from the pelvis down, to balance and correct the position of her trunk on her pelvis. A lightweight pair of long leg calipers were

introduced and work began to maximise the function of trunk on pelvis; after a while it was possible to take the knee pieces from the calipers and just use the side steels into the boots and the thigh cuffs allowing for some active knee control, but with the calipers providing a prompt for knowing the position of the body as a whole. After a lot of patience, facilitatating a reciprocal gait pattern has become possible with the calipers on and free standing balance has been developed for several seconds.

Present and Future

Sandie now likes hydrotherapy and enjoys being able to float hands off with floatation aids. She is able to communicate some of her basic needs, especially likes and dislikes. After four years she obviously likes to be handled and will now snuggle up to a known adult if they are sitting near by. She has become confident that she will not be placed in a position of danger and is willing, if a little wary at first to try new things. Her hand function remains poor and there are still plenty of areas of motor development to work on. However, she is a delight to her adoptive family who enjoy all her new achievements - even the very small ones - and we will continue with therapy, school activities and a home programme to develop her skills to their full potential, although we do not really have any idea what this will be.

There is a Wolf-Hirschhorn Syndrome Support Group with the following address:-

Mrs Chris Hilder
Wolf-Hirschhorn Syndrome Support Group
2b Harvesters Close
Rainham
Gillingham
Kent ME8 8PA
tel / fax 01634 264816

SEVERE COMBINED IMMUNODEFICIENCY

LIZ HARDY
Paediatric Superintendent Physiotherapist
Newcastle-upon-Tyne

Severe combined immunodeficiency (SCID) is a broad description for a group of rare conditions which affect the immune system, occurring in around 1 in 40,000 live births in the UK. Affected infants are usually well for the first 3 months of life, whilst protection from infection is offered by maternal antibodies acquired through the placenta. From then onwards life is a downward spiral of failure-to-thrive, frequent episodes of multi-system infection and developmental delay. Without treatment affected infants will invariably die within the first year of life. The only treatment which provides long-term survival is a successful bone marrow transplant (BMT).

Newcastle-upon-Tyne has one of only two specialist units within the UK for treating this disorder. Families come from as far away as Birmingham, Scotland, N. Ireland and Eire. Accommodation is provided and family life is transferred to the North East.

Very basic immunology

The function of the immune system is, in its simplest form, to distinguish self from nonself in order to provide defence mechanisms to protect the body against attack by 'foreign' invaders or antigens. These foreign bodies may be bacteria, viruses, or tissue from another person (donor heart, kidney, bone marrow, etc.). Occasionally in auto-immune disease 'self' is incorrectly recognised as 'foreign'.

In the normal immune system, stem cells (immature and undifferentiated white blood cells) are formed in the bone marrow. Some remain there and differentiate into B-leukocytes whose function is to produce immunoglobulins or antibodies . Others migrate to the thymus gland where they multiply and develop different characteristics, becoming known as T-lymphocytes. There are several types of T-cell each with a different function, e.g. 'Helper T-cells' stimulate B-cells to produce specific antibodies, 'Killer T-cells' directly attack viral tissue, whilst 'Suppressor T-cells' switch off the action of B- or T-cells. Both types of immunologically active leukocytes are stored in the lymph nodes and spleen, ready for release into the circulatory system as needed.

Other cells which play an important role are macrophages, phagocytes (granulocytes, neutrophils, basophils, eosinophils and mast cells), granular lymphocytes and the complement system.

Laboratory findings in SCID demonstrate almost total absence of T-lymphocytes usually, although not always, with a reduction in the number of B-lymphocytes.

Clinical features of SCID

Affected infants are super-susceptible to infections, especially of skin, gut, lung etc. They are usually admitted to the specialist unit in an extremely poor condition, often with profound and intractable diarrhoea, vomiting,

skin rashes and sore mouths due to candida, and with significant chest infection or pneumonitis. Some, aged 3-6 months will weigh less than their birth weight.

General management

Both before and after BMT, infants with SCID require high dependency in-patient care, which usually continues for around 6 months. They are cared for in strict isolation in order to minimise the risk of infection. This takes place in large cubicles equipped to intensive care specifications, with access to the child restricted to parents and essential staff. The cubicles contain an area, only large enough to include cot, chair and essential equipment, which has a laminar down flow of 99.99% sterile air, changed 200 times every hour. Other healthy visitors and staff may enter the cubicle, as long as they remain outside the laminar flow. Reverse barrier nursing takes place for all procedures, and includes a rigorous pre-entry preparation of scrubbing to the elbows with chlorhexidine or betadine for a minimum of 2 minutes. Hats and gowns are worn to facilitate comfortable and safe handling of the infant. Infants are bathed in sterile water, fed sterile food (until inevitably intravenous nutrition is required), and play with toys which can be frequently and thoroughly cleaned. Parents are strongly encouraged to spend as long as possible within the cubicle and to take a lead role in their child's care, with the exception of carrying out painful or invasive procedures.

All infants have double or triple lumen central venous catheters inserted, through which chemotherapy, other drugs and bone marrow are given and blood samples can be taken.

Nursing staff monitor for changes in vital signs and weight, whilst daily assessment by medical staff takes place, including bloods. All infants routinely have cultures of naso-pharyngeal aspirate, stool, urine and skin swabs twice weekly.

In our unit, the nursery nurse, occupational and physiotherapist work together in playing a vital role in assessing the infant's developmental status, and providing an appropriate programme of activity and positioning. Unfortunately we have only limited access to SLT, despite the fact that delayed speech and oral feeding are, unsurprisingly, significantly delayed.

Bone marrow transplantation

A successful BMT will replace the infant's defective immune system with a normal one, thus restoring normal immune function. The first BMT for SCID was carried out in the USA in 1968 and the patient continues to survive. The most important factors ensuring success are the health of the recipient at the time of transplant and the quality of the donor tissue match. The tissue of choice is always a matched sibling transplantation, since these have an engraftment success rate of over 90%. Recipients do not require conditioning with chemotherapy first, and whole bone marrow

can be infused. Engraftment occurs quickly and the whole 'new' immune system can be active in as little as 6 weeks.

However, about 80% of infants with SCID will not have a sibling with identical tissue. In these cases there are two options :

- A parental but non-identical transplant from which the T-cells have been removed can be infused, following intensive conditioning of the recipient with busulphan and cyclophosphamide. When the graft has been T-cell depleted, the 'new' immune system takes much longer to develop in the recipient, with T-cell function beginning at around 100 days and B-cell function between 200 and 250 days.

- Alternatively, a matched, but unrelated, donor BMT is required. It can take up to 6 months to identify and recruit a suitable donor, and this is, sadly, sometimes too long for the very sick SCID infant to wait. Several days of cytoreductive conditioning is required pre-transplant, as mentioned above, to a patient-specific protocol.

Severe graft versus host disease or rejection of the graft are more common in both these types of BMT than in the matched sibling transplant.

Once the BMT protocol has commenced the risk of infection in the infant increases even further due to the conditioning treatment. Infants are very debilitated by this stage with chronic carriage of infection and malnourishment. Depite good engraftment, survival is not guaranteed

Common respiratory problems in SCID

1. Bacterial pneumonia usually caused by pneumococcus / staphylococcus.
2. Pneumocystis carinii pneumonia (PCP) - ? fungus / protozoan. This is a very serious infection, signs of which are tachypnoea and hypoxia. On auscultation few crepitations may be heard. A consolidation with a 'cotton wool' appearance can be seen on chest X-ray. Infants are routinely given prophylactic anti-PCP antibiotics, such as cotrimoxazole or trimethoprim, but many still succumb to this infection.
3. Fungal infections, especially candida and aspergillus.
4. Respiratory viruses are common and especially difficult to manage effectively. Those frequently seen include RSV, parainfluenza, influenza and adenovirus. All can produce a severe pneumonia or pneumonitis. Clinical signs are tachypnoea, recession, nasal flaring, cough, hypoxia and copious secretions. Associated secondary bacterial infection frequently occurs.

All conditions are frequently managed with unconventional drug therapy.

Physiotherapy

There are two facets to the physiotherapy imput for infants with SCID - both of equal importance.

1. *Respiratory*

Assessment is carried out daily on all vulnerable infants, especially during conditioning and engraftment, and otherwise at the request of medical or nursing staff. Treatment is not carried out routinely, but vigorous, aggressive management of apparently minor respiratory symptoms is required. This will often begin on the evidence of altered baseline measurements for respiratory rate and oxygen saturation, without waiting for the development of altered chest sounds on auscultation. This is because delay will allow infection to spread and develop quickly, with possibly fatal consequences. In addition, usual 'rules' often need bending! For example, there is a constant 'juggling' of extremely low platelet count, which would normally contraindicate manual physiotherapy, versus the need to clear secretions quickly and effectively. Also abdominal distension does not contraindicate prone position, if indicated from the respiratory aspect, as long as rolled towels and small pillows can be used to relieve pressure whilst in prone.

Pneumonia in SCID infants improves very slowly and requires treatment, as expected, with positioning, postural drainage and manual techniques as indicated.

Pneumocystis can cause severe respiratory symptoms very quickly, and often the infant needs mechanically ventilating. Copious secretions are produced when the illness is resolving, requiring frequent endotracheal and nasal suction. There is a high risk of secondary infection.

Viral pneumonitis, once present, won't clear until after the BMT. Persisting secretions and ARDS are a significant threat and there is a great potential for lung damage to occur.

The nursing staff are all capable of carrying out effective treatment on both non ventilated and ventilated infants, and a plan is formulated at least once daily. However, the physiotherapist will carry out most treatment sessions during the day.

2. *Developmental intervention*

Multi-disciplinary assessment takes place on admission and then at regular intervals during the infant's admission to the unit, and a video record is made. A plan for daily activity is constructed, appropriate to the infant's age and current health status, and aspects of this will be carried out by everyone who comes into contact with the child - parents, medical and nursing staff and therapists alike!

Floor activities can be carried out on a clean blanket, but should a toy roll off - it is 'lost' for that session - very frustrating for both child and therapist. It can be especially difficult to encourage gross motor skills within the confines of the cubicle, especially with very limited resources available. For example, encouraging upright play for an extremely small child, without using velcro or foam on a standing frame or wooden seat (none of which can be cleaned effectively to 'go under the flow') sometimes provides great challenges. No less a challenge is getting oneself into the appropriate position to facilitate a movement without allowing clothing, overshoes etc. to contact the blanket over which the child will move and therefore introducing the possibility of infection!

During conditioning and engraftment, as well as when the child has an infective exacerbation, are difficult times for encouraging movement as the infants may be extremely unwell and in pain, so modifications are made and treatment or 'play' sessions can be very short. The child is allowed to dictate the pace to a large extent.

In contrast to the respiratory indication for the use of the prone position, an infant with abdominal distension would not be positioned in prone for play.

Once engraftment has occurred and the 'new' immune system is beginning to function there is a general trend of good developmental progress, and many infants will leave the unit with milestones within the expected range.

Discharge

Infants who have undergone BMT cannot be discharged home until they have active T-lymphocyte function. Intravenous immunoglobulin therapy will be required every 3 weeks until B-cell function returns. Antifungal cream and broad-spectrum antibiotics will also continue post-discharge.

Discharge is a gradual process, requiring careful planning. Families and their recovering child stay locally in a 'half-way house' for between 2-4 weeks after leaving the unit, in order that support in re-learning to care for their child can be given. Most parents welcome this arrangement, as there is a high level of anxiety at this time.

The continuing care of the child takes place in their local hospital, so a detailed discharge summary is sent to all personnel who will be involved. Handover phone calls and occasionally visits are also carried out. The Paediatric Immunology Liaison Sister fulfils an invaluable role in ensuring continuity and support for everyone.

Conclusion

Infants with severe combined immunodeficiency provide a management challenge for all members of the multidisciplinary team. It is impossible to achieve a 'normal' environment to encourage the growth and

development of these very sick children, since their survival is dependent upon isolation from harmful organisms. The treatments surrounding bone marrow transplantation carry significant risk, and also often make the child even more unwell. Infants are often highly dependent upon unconventional therapy.

Hovever, survivors of the process have normal immune function and go on to lead full and active lives. Development of all areas soon returns to normal once infants are out of the isolation cubicle, although at follow up a significant number would seem to display some features of motor incoordination. This, and many other areas associated with improving knowledge and management of SCID are currently being studied.

Liz Hardy wishes to acknowledge the help and support given by all members of the team, whilst working on the unit and writing this article.

VISIT TO THE COPENHAGEN CYSTIC FIBROSIS CENTRE

DAVE THRELFALL
(Senior Physiotherapist, Sheffield Childrens Hospital)

I was very fortunate to visit the Cystic Fibrosis Centre at the Rigshospitalet University Hospital in Copenagen in May 1998.

The Centre is very well known for its excellent work with CF children and adults, its numberous publications and its aggressive approach to the treatment of pseudomonas aeruginosa (PA) with the regime of 3 monthly intravenous (i.v.) antibiotic treatment.

The centre at Rigshospitalet cares for around 300 patients, representing 75% of all CF patients in Denmark, the rest attending a second centre at Aarhus in the north of Denmark. The incidence of CF in Denmark is 1:4,700 births, with a carrier rate of 1:37. 80% of the patients are homozygous and 19% heterozygous for the dF508 mutation. There is no neonatal screening programme and most infants are diagnosed early and referred to the centre.

I attended the centre on two days, during which time I was able to observe this clinic, visit the wards, meet the clinic staff and talk to several patients and their families.

Although the purpose of my visit was primarily to meet Mette Kelstrup, who along with her colleague Merette Falk, pioneered the PEP mask technique of airway clearance, I also had the opportunity to discuss many other issues with Dr Christian Koch (the Clinic Director), other medics who supported him in clinic, Annalisa the CF nurse specialist and others.

Fortunately they all spoke excellent English and during clinic examinations would speak English deliberately for my benefit. The only slight hiccup the whol time I was there was when I commented that the young boy in clinic "looked like he was cheeky" but his mum thought I had said he looked like a chicken! She was a bit offended at first but the misinterpretation was quickly explained.

Clinics

There was a clinic every day, Monday to Friday. The patients attended clinic every month. Different days were assigned to different groups of patients according to their microbiological status. Some clinic days were for PA free patients, other days for PA colonised patients and yet other clinic days for patients with Cepacia, MRSA or other resistant/infectious organisms. The centre is very strict on segregation and reports a fall in the mean annual incidence of patients becoming chronically infected with PA from 17% to 3% following the introduction of segregation.

The clinic was extremely friendly, Christian Koch obviously has a very good rapport with his staff and patients. The clinic accommodation was spacious, light and modern looking with jars of sweets everywhere. Their CF may be well managed but they've all got rotten teeth!

The patients arrive in clinic and are weighed and heighted by the physiotherapist who then records their lung function on a Jaegger spirometer. The results are computerised and a print out with a graphic record of the present and past values are made available to the clinician.

The physiotherapist then quickly checks their PEP mask technique and makes a note of the resistor used.

If a sputum sample is not obtained during the spirometer or PEP mask check, Annalisa the clinic nurse, obtains one via nasopharyngeal suction. This is performed with a fine bore rubber tube, which is inserted, suction applied, usually stimulating a cough and then withdrawn very quickly. This procedure is performed on any patient who doesn't spontaneously expectorate sputum, no matter what age! I saw the procedure performed on several patients of different ages, all but the very young ones seemed to tolerate it without any trouble, and of course they all get a sweet afterwards (including the staff). Cough swabs have never been used in the Copenhagen clinic.

On further discussion Christian Koch was not convincing that NPA's were any better than cough swabs as they did not always grow anything when clearly the patient was growing organisms in his lower airway.

The microbiology results were available to the clinician within two days of the sample being taken and then a prescription was faxed to the pharmacy nearest to where the patient lived for collection.

Apart from the medics, the CF nurse and physiotherapist, no other professionals were available in the clinic. The dietitian was situated in the same building but only saw patients when specifically referred to by the medics. A clinic psychologist was also available by referral. There didn't appear to be any need for social work input.

Observing several patients in clinic Dr Koch took the usual history concerning respiratory and gastrointestinal symptoms, discussed their lung function and sputum culture results. The examination was limited to listening to the chest, no abdominal examinatiin was witnessed (and of course more sweets were eaten!)

Children and adults were seen together in the same clinic, there was no adult physician involved in the patients care, although Christian Koch commented that he hoped that would change. All the medics wore white coats and the rest of the staff white uniforms.

CF Ward

I visited the main CF ward which had two four bedded bays and several cubicles, sitting room, kitchen and treatment rooms. TV's were suspended

from the ceiling at the end of each bed. The patients had to purchase a card to allow them to operate their TV, although there was one available for free in the sitting room.

The patients seemed to mix freely in the sitting room, kitchen or corridor and managed most of their own treatment. They also seemed to be wearing white gowns or overalls provided by the hospital.

Physiotherapy

All patients use the PEP mask to the exclusion of any other airway clearance technique. PEP is started in babies as diagnosis with a small mask and aiming for expiratory pressures of around 8 cm H_2O during tidal volume breathing for one minute periods. I got the impression that PEP in babies and small children was not particularly well tolerated because to work properly the mask has to have a complete seal with the face. One 18 month old child I saw using the PEP screamed and screamed. The advice given by Mette in such circumstances was to use the PEP when the baby was asleep, which she admitted was not ideal, preferring the child to be awake and upright.

In the older child and adults the PEP was used, aiming for pressures of 15 - 20 cm H_2O, for approximately 10 minutes at a time, twice daily, increased when necessary. The resistor was checked at clinic regularly but rarely changed. All the patients I spoke to reported that they found the PEP mask to be effective and easy to use. None of the patients I spoke to were properly aware of any other form of airway clearance technique.

The PEP masks used were those produced by Astra, at approximately £50 each, although with a reduction for bulk purchases.

There was no physiotherapy input on the wards, it was considered that all the patients were independent with their PEP masks and knew to increase the amount of time using the PEP during a respiratory exacerbation. Exercise was not prescribed, there were no exercise facilities in the clinic and exercise testing was never performed, not even as part of the annual review.

Antibiotics

The first growth of PA is treated with 3 months of oral Ciprofloxacin and nebulised Colistin, 2 megaunits three times a day. This regime has apparently increased the mean time to recurrence of PA after treatment of the positive culture from 9-18 months. There is no lower age for the use of Ciprofloxacin.

If PA is persistently cultured in the sputum, or there is an increase of precipitins to PA the patient is considered to be colonised with PA and as such will start 3 monthly i.v's, irrespective of their symptomology. They may have courses or oral Ciprofloxacin between i.v. courses and all the

patients colonised with PA are on nebulised Colistin. It is the very firm belief of the centre that the use of 3 monthly i.v antibiotics is central to improved prognosis. The accepted 3% annual fall of FEV1, reported in chronically infected patients does not occur in the majority of the Copenhagen 3 monthly treated patients, nor in their patients who are prevented from acquiring chronic PA infection by early treatment with Ciprofloxacin and Colistin. (Federikson et al 1997).

PA precipitins are checked every 3 months and 3 months i.v's are only stopped if they are consistently zero.

Many patients have their course of 2 weekly i.v antibiotics at home, but the first dose is always given in hospital and then a plan made for the next 14 days. Lung function tests are done at the start, half way and at the end of the 2 week course. Patients also return for Tobramycin levels. The i.v's are usually given via a canula, sometimes a longline but only 8 patients have a porta-cath.

Two patients I saw in clinic were starting a course of i.v's, one for her regular 3 monthly PA treatment and the other to treat his Cepacia. This young man was having iv imepenam via a canula and he was on a regime of 2 weekly i.v's every month. Annalisa told me they had approximately 9 patients out of 300 with Cepacia.

IV Tobramycin with an betalactam seemed to be the first preferred choice of treatment for PA. The dose of tobra was 10 mg/kg/day aiming for a peak dose of 15 - 17 and a trough of 1 - 2. Chromium DTA clearance tests are performed early when commenced on 3 monthly i.v's to check for toxicity. Antibiotic resistance, although quite common, is not regarded as a major problem. Between 10 and 20% of PA organisms are resistant to aminoglycosides. Christian Koch commented that they had more problems with hyperresponsiveness than with resistance.

Nebulised Tobramycin is only rarely used instead of the regular 3 monthly i.v's but is often used in addition to i.v's and as well as nebulised Colistin. The nebulised Colistin is not stopped while the patient also has nebulised Tobramycin.

All patients appeared to use a Medic Aid CR50 with a durable sidestream for Colistin and a CR50 with ventstream for Tobramycin. They do not use a filter with Colistin but use elephant tubing to vent the exhaled gases out of the room when using Tobramycin.

Christian Koch mentioned that they had trialed the used of high dose oral Ciprofloxacin instead of the regular 3 monthly i.v's but it was not found to be as effective (unpublished data).

Pulmozyme

Every patient chronically colonised with PA is on DNase, via a CR50 compressor and durable sidestream nebuliser. Some non-PA patients are also on DNase. Christian Koch commented that 1/3 of patients on DNase improve significantly, 1/3 improve moderately and another 1/3 on DNase do not seem to gain any obvious benefit. When I asked him if this 1/3 of patients therefore discontinued using DNase he said no they wouldn't unless the patient themselves requested it.

They have never used any other nebulised mucolytic e.g. amiloride or hypertonic saline in Copenhagen. They did trial oral n-acetylcysteine pre DNase but did not continue with it.

According to Annalisa the CF nurse, each patient would have ventolin, then do their PEP, then have nebulised Colistin followed by DNase. According to Mette, the CF physio, the order was ventolin, DNase, PEP and finally Colistin. If DNase is taken before using the PEP a time period was not advised for the patient to wait in between.

They are currently involved in two DNase trials in Copenhagen. One large 'in house' trial involving 70 patients simply looking at the number of bacterial isolates over 2 years compared to a non-DNase group and a smaller multi-centre trial wth 10 children aged between 8 - 10 years assessing the effect of lung function over a 2 year period.

According to Annalisa the patients used the same nebuliser for their Colistin and their DNase.

Nutrition

"If the child is not growing properly it is because the chest disease is not being effectively managed" - Christian Koch.

There appeared to be much less emphasis placed on nutritional management in Copenhagen than here in the UK. All the patients were known to the dietitian from diagnosis, but following that, her involvement was only on an 'as necessary' basis.

High strength enzymes are no longer used following "a scare" some time ago, when some patients developed colonopathy, all patients since then have been managed on Creon 6,000. While I was there the patients were all being changed to Creon 10,000. Their aim was to achieve 6 - 12,000 units of lipase/kg/day.

Only one patient has had a PEG, N/G feeding is used only occasionally and the only supplements mentioned while I was there was that some patients used Scandishakes. All the dozen or so patients I saw over the 2 days looked well nourished.

Socio Economics

Denmark introduced one of the first welfare state systems in the 1930's and health insurance now covers more than 96% of the population, providing free medical care and hospitalisation. There is no limit to what the doctor can prescribe and if they consider a drug necessary it is provided. To support this system the Dane's pay a very high rate of tax.

Another consequence of this is that the parents of a child with a chronic illness such as CF an take time off work, fully paid for as long as they want. It is only eligible to one parent at a time and it is reviewed every 6 months by the government but if the parents can show that their child is still dependent on them for their care they can continue to be paid on full salary while off work.

Conclusion

The CF centre at the Rigshospitalet is an extremely friendly and enthusiastic place. The team seem to work very well together with each member very proficient in their different and well defined roles.

Their dogmatic and aggressive treatment of PA has borne huge dividends for their CF population. For example there has been an increase in age of acquisition of chronic PA infection in their clinic from 10 - 20 years, the prevalence of chronic PA infection in the under 10 year olds is only 5% and the fact that 50% of their patients are still not chronically infected with PA 7 years after the first positive culture.

The aggressive treatment with 3 months IV's and Pulmozyme is obviously made a lot easier for them when they do not have to consider the cost unlike in the UK!

"GO FOR IT"

Guidance on physical activity and sports for people with haemophilia and related disorder

Available from The Haemophilia Society : Price £3.50

Don't be fooled into thinking that this small, spiral bound, card book is a quick five minute guide for children - it isn't. Packed into it's seventy two pages is detailed information on a variety of sports and activities for people with haemophilia; guidance for teachers, trainers and coaches; sections on drugs in sport and sports injuries, and much, much more.

The book is written to encourage people with haemophilia to take part in sport and to put their disease in second place. It is not written for young children but has 'you' sections for teenagers throughout which include general information about the benefits of sport; which one to choose, the importance of a good warm-up and a fitness programme for swimmers etc. Other parts give advice to older people, coaches, parents and teachers. The book does not seek to minimise the impact of the disease but gives what appears to be full and measured information on prevention of injury, first aid, cross infection, and haemophilia itself. The preface by Duncan Goodhew and a safe warm-up routine from olympic athlete Kirsty Wade add the glamour element to inspire youngsters.

My only quibble with the book (apart from the fact that I thought it was only going to take five minutes to read) is that the early section on 'severity and sport' and the accompanying table, assumes an in-depth understanding of the condition and is quite technical at this stage. It would sit better with the later section explaining the disease for coaches and teachers.

The text is underprinted together with colour washes and a mixture of line drawings or hazy prints which depending on your preference add or distract from the written word. I felt it worked well most of the time, but perhaps not on the page where coloured highlighters are used to categorise sports into recommended / not recommended / others.

But these are very minor points. Overall, this is an excellent, inexpensive and invaluable guide for those of us working with children with haemophilia. So 'go for it', buy the book and see what you think.

Carrie Jackson MCSP

PAEDIATRIC CHIROPRACTIC 1998

Editors : Claudia Anrig and Gregory Plaugher
Publishers : Williams and Wilkins, Baltimore, USA
ISBN 0-683-00136-1 789 pages

The book has drawn chapters from international chiropractic authors on various paediatric specialities, and presents as a dauntingly large but eminent manageable tome, written with enthusiasm in readable English, and incorporating diagrams, tables and plates. It is aimed at students and practitioners of chiropractic, and there is the assumption that the reader will understand chiropractic terminology.

Mentions of current research activities are presented for various conditions of childhood, and all seem to beg more vigorous protocols and conclusive results.

The editors have included chapters with discussion of spinal stresses, examination and adjustments for mother and foetus, exerises during pregnancy, and diagnostic imaging. This last is presented in words, chiropractic codes, and numerous plates, tables and diagrams. With 40 potted case histories and clinical courses, tracking relevant x-rays was, at times, confusing. Further chapters include Active Immunisation (377 references seem to have led to a sceptical attitude to the value of this) and Motor Vehicle Collisions (very clear presentations of mechanisms of injury). The section on Dis-ease Prevention covers paediatric nutrition and spinal examinations, with links from testing and X-ray to adjustment. Also in this section if a discussion on the relavance of Craniosacral Therapy as a response tool in the 1st year of life, to foetal and birth trauma. Clinical Neurology is exhaustively and clearly presented, while Orthopaedics has an equally extensive cover, with less emphasis on effective

chiropractic intervention. Intervention is also included in the chapter on growth and sexual development of adolescents.

All chapters have extensive reference lists.

This would be a useful resource in a Physiotherapy Paediatric Library. Readers should be prepared to become absorbed.

Elspeth O'Donnell, BA, MCSP

VIDEO REVIEW

ARTHROGRYPOSIS: A Short Guide to the Physiotherapy Management of the Disability

Cost : £20 (including postage and packing) and Information Pack

Running time : 9 minutes

Order from : The Arthrogryposis Group
1 The Oaks
Gillingham
Dorset SP8 4SW

The purpose of this video is to explain the group of conditions under the umbrella term 'Arthrogryposis Multiplex Congenita' and to give an introduction to its management. It describes possible causes and basic classification of the condition with a suggested list of investigations needed to reach an accurate diagnosis.

Early physiotherapy intervention and parent participation are stressed, as is the excellent long term prognosis for many of the children.

Basic physiotherapy techniques including daily stretching and mobilisation, splinting and orthotics are shown with reference to surgery at a later stage to improve function rather than appearance.

There are some nice sequences of children participating in a variety of physical activities and mention of adult independence and employment.

This is a well produced, if short, video which would be a valuable addition to Paediatric Physiotherapy Service libraries.

It would be suitable for Junior Physiotherapists, or those new to the condition, and would give an overview of the condition and its treatment to GPs, HVs and families.

Liz Roylance
Senior Paediatric Physiotherapist

TOOBERS AND ZOTS
by Imperial Games

This is a new range of bendy foam pieces and shapes that link together to make all sorts of different shapes – from really simple to very complicated constructions. All the children who have tried my set have really enjoyed them. Children with a hemiplegia seem to be encouraged to use both hands, as much as possible. The bendy Toobers can be formed into all sorts of shapes and can be 'played with' by all ages and used as simple craft toy, with or without the Zots. Parents just like the feel of them and become creative with their children. The little Zots can be used to make flowers, dinosaurs and all kinds of beasties. They are very light weight, so can be managed by most children. Some of the pieces are tiny, so care would be needed in selecting the appropriate set for younger children.

Sue Whitby

'FREEBIES'

Tying shoelaces may never be a problem again

The toggles on each little bag found in a box of washing detergent tablets can be used on shoelaces to hold them firmly. The shoelaces can be then be tied as well as the child can manage, or simply tucked inside the shoe or trainer. They look really stylish and have street cred! They are usually blue, so if you need another colour you may have to get the toggles from an old rucksack or anorak.

Sue Whitby

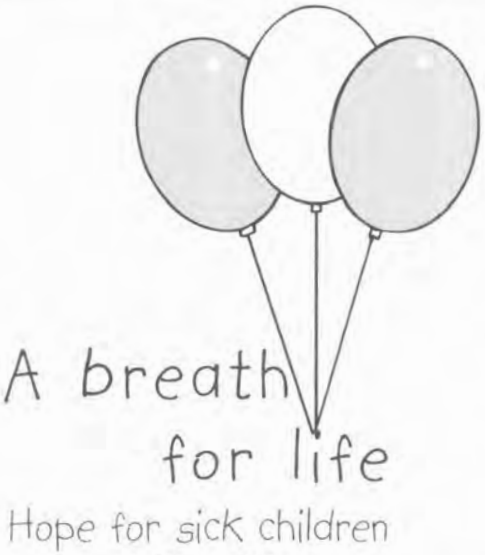
AN ALTERNATIVE USE OF A FLEXISTAND

I thought that others might be interested we have solved the problem of secure floor seating for James, a three year old boy with profound dystonic cerebral palsy affecting his whole body.

James in an intelligent and responsive person who likes to play, but until we were able to devise this adapted flexistand he always had to be supported by his mother,wh sat behind him and unconsciously corrected his postural variations. In this position she was not able to make eye contact or encourage him by visual clues. As you can imagine we tried the usual options of floor seating without a good result.

As can be seen in the enclosed photograph, James sits on the foot board of the stand with a dycem mat to prevent slipping. He uses his roll to stabilise him and to produce a degree of lumber lordosis and is held in place by the double strap. He now corrects his own postural variations and his mother can be seen in front of him.

Andrea Walton
Senior Paediatric Physiotherapist
Child and Family Health Centre
Ashurst Hospital
Lyndhurst Road
Ashurst,
Southampton SO40 7AR



Registered Office :
Waterview, Unit 1a, White Cross, Lancaster LA1 4XQ
Tel: 01524 380363 - Fax: 01524 844757

Now is the Time

A breath for life was born out of a free childrens' clinic set up in 1996 in Lancaster by Jane Dean, registered naturopath and Charles Tisdall, registered osteopath. Their idea was to offer a free clinic on the first Saturday of every month. This would allow alternative therapies to be available to all children in the area, not simply to those families who could afford such treatment. The clinic would offer individual programmes to stimulate healing and immune activity in each child.

Vision Statement

To build a reputation as a centre of excellence in the treatment of sick and brain injured children through the provision of services and facilities to promote and protect the health of children. The centre will be in Lancaster with a view to encouraging the establishment of such centres throughout the UK.

Mission Statement

With the focus on the children, *A breath for life* will offer a programme of alternative treatments that could enable sick and especially brain injured children and their carers to enjoy a better quality of life.

A breath for life aims to be a resource of information and provide a network of empathetic personnal, including parents, to offer some light at the end of a dark tunnel.

Aims and Objectives

1 To promote the rehabilitation of children who have suffered from a debilitating illness by providing individual programmes of care.
2 To offer a comprehensive selection of treatments which do not take recourse to drugs or surgery.
3 To increase understanding and awareness of the principles and practices of health, rather than disease.
4 To promote health by using Nature Cure which encourages the body's own mechanisms for self-healing.
5 To develop a supportive partnership with parents and children in order that they can take an active part in the management of their health.
6 To provide a resource centre for all interested parties.
7 To seek out new and rekindle old avenues of health care compatible with the charity's philosophy and relevant to the health of our children.
8 To work in conjuction with other health care providers in the community.
9 To conduct and publish research information from the charity's work and practices in order to evaluate our therapeutic modalities with a view to establishing them within the health care market.
10 To arrange and host study days and seminars.
11 To make available food and dietary supplements beneficial to the continued good health of the children and their carers.
12 To educate the public about A breath for life and in doing so raise the profile and build the brand in order to raise funds and sponsorship to support and grow the charity.

Treatments

Naturopathy - Osteopathy - Hyperbaric Oxygen Therapy - Exercise Programmes - Homeopathy - Craniosacral Therapy

The UK Federation for Conductive Education

The UK Federation for Conductive Education is an association which includes

- **trained conductors**
- **other professionals**
- **parents**
- **other interested parties**

All of whom have a direct or indirect interest in The System for Conductive Education.

Benefits for members include :

- A list of members and contact numbers
- A publication - Understanding Conductive Education
- A quarterly newsletter
- Practical Workshops
- Representation on the International Peto Association
- A forum for exchanging professional information
- Information on relevant publications
- Information on employment opportunities in this field
- Information on Conferences and Training Courses

Contact : Ann Loton (Hon. Secretary) c/o Horton Lodge School, Rudyard, Leek, Staffs ST13 8RB
Tel: 01538 306214 Fax: 01538 306006

Have you any useful ideas or tips?

Do you know about any useful Support Groups or Organisations?

HERE AND THERE

is the place you can share information with others.

We are always interested in hearing about new ideas, innovations etc. for inclusion in this section of the journal. What has worked for you, however simple, may help solve someone else's problem. Why not share it?

SUMMARY OF ISSUES DISCUSSED AT THE APCP NATIONAL COMMITTEE MEETING HELD AT THE CHARTERED SOCIETY OF PHYSIOTHERAPY ON 9TH JULY 1999

1. **C.S.P**

The Chairman welcomed Gwyn Owen to the meeting to discuss motions at ARC which have implications for paediatric physiotherapy and also to update us on the proposed changes to the SEN Code of Practice. (see Letters to the Editor).

2. **ARC**

Vice Chairman, Di Coggings, successfully seconded the BABTT Motion put forward to ARC in Glasgow in May 1999. ARC next year will be held in Eastbourne on the same dates as the APCP Conference in Bristol. Members from the South regions will be given the opportunity to be the nominated representatives of APCP at ARC.

3. **Journal**

The Editor has produced a leaflet giving guidance on how to write articles for the Journal. A copy will go out with each membership pack..

4. **Public Relations**

Concerns about gaining access to children in mainstream schools who require physiotherapy continue to be raised by the membership. All paediatric managers in England and Wales should now have received a copy of Scope's publication "Working Together, Parents and Physiotherapists".

5. **Education**

There have been successful meetings about the production of Guidelines. A draft will be presented at conference and completion by October 2000 The MSc Paediatric Module is on course to commence at Queen Margaret University college, Edinburgh in September 2000 .

6. **Membership**

There are now 1464 members of APCP.

7. **Publications**

The publication "Manual Handling in Paediatrics" is now available at a cost of £10.00.

8. **Research**

The Research Officer highlighted groups who make funding awards for research projects. Please contact Carrie Jackson for more information.

9. **Conferences**

The Conference and Exhibition of the Chartered Society of Physiotherapy of which APCP is a part, will be held in Birmingham on 8th - 10th October 1999.

APCP Conference 2000 and AGM will be held in Bristol in May. The AGM will be on 5th May.

10. **Next Meeting**

The next meeting of the APCP National Committee will be held on Friday 8th October in Birmingham.

A full copy of the minutes of the national committee meeting can be obtained from your regional representative.

PROFILE

Honorary Treasurer

JULIA GRAHAM MCSP BSc (Hons) Physiotherapy

Julia trained in Newcastle upon Tyne, qualifying in 1983. She began her career working in Gateshead carrying out junior rotations in a large general hospital. Eighteen months later she took up a Senior II post in rheumatology and orthopaedics but six months later moved in to paediatrics, an area of keen interest since student training.

She spent five years working on community paediatrics in Newcastle before moving south to take up the Head Physiotherapist post at Lord Mayor Treloar School and College, in Hampshire.

She remained there for six years before taking a community paediatrics post with Loddon NGS Trust where she works today.

She has had a special interest in paediatric manual handling for some years now and she is a cascade trainer within the Trust and goes out to other areas to facilitate manual handling training amongst other paediatric therapists.

She upgraded my original Diploma qualification in 1995 at Portsmouth University and she is currently carrying out an MSc in Disability Studies at King's College, London.

She has involved in APCP for as long as she has worked in paediatrics, originally in Newcastle but more recently as South West Regional Treasurer and then Regional Representatives and for the past year as National Treasurer.

In what little spare time she has, she enjoys her garden, circuit training and other activities in her village, walking the dogs, being on holiday in the sun and ski-ing in the winter!

PRO REPORT

In Birmingham, this year, from the 8th to the 10th of October, I will have an APCP stand, at the CSP conference, where we are holding our APCP conference. Please let me have any information you want to share with all the delegates.

From time to time I get request for APCP clothing - usually polo shirts or sweat shirts, for paediatric physios to wear whilst working with children. We often have conference specials but we have not, so far, had any

APCP clothing. After my comments on this in the last issue I received information about several schemes throughout the country. The National Committee has decided to make the Conference 2000 clothing double as the new APCP clothing, so this will be launched in Bristol next May.

I hope that you have all received the SCOPE document, which Jill Brownson has written about elsewhere in this issue. Please contact me if there are any more people who have not received their copy. With the advent of clinical governance, more of you are writing leaflets and information sheets for your patients. If you have written anything, which would be useful to all of us out there, please share it with us. I am grateful for documents that I have already received and look forward to some more!

Hopefully we will be able to produce more APCP publications in the future.

My secretary at work will always take a message for me. Please phone 01480 415203.

Mrs Sue Whitby

RESEARCH OFFICER'S REPORT

NEWS AND THINGS

Funding :

Details of three funding opportunities have reached APCP via the CSP.
If you are presenting a paper at an interdisciplinary conference, the UK Presentation Fund is the one for you.

For help with your research project try the Physiotherapy Research Foundation - reputed to be very generous (source - APCP committee) - or the H.S.A. Physiotherapy Research Degree Scholarship. Details are enclosed and for an application form, contact Marion Attew at the CSP.

CSP Information Papers :

There are a wide variety of papers available free from the CSP which some of you will be familiar with. A recent discovery was the IRC (Information Resource Centre) No. 4, which is a list of the Theses and Dissertations held in the IRC. A brief glance shows that it contains 11 paediatric entries dating from 1976 to 1991 - I feel sure there are many, more recent, works out there somewhere! If you have one, or know of a colleague's, please consider sharing it by offering a copy to the CSP. They are an extremely useful resource for researchers.

I would also recommend the IAC 6 - Using the CSP Current Awareness Bulletins. It doesn't take long to read and will reward the effort with practical suggestions and encouragement re implementing evidence based changes and CPD.

Statistics

- A dirty word to most of us - but a little knowledge may not always be a bad thing!

One recently discovered 'gem' relates to the dreaded 'p value' that crops up in all research.

It simply indicates the level of confidence the researcher has that his/her findings are not due to chance.

Hence a p value of 0.05 means that the researcher is 95% confident and in most cases this means the findings are significant. The lower the p value the higher the confidence/significance; e.g.0. 2 = only 80% confidence while 0.001 = 99.9%.

Literature Searching: - Databases

Our hospital library offers tutorials - approx. 1 hour long - in how to use the MEDLINE and CINAHL databases. These are supported by excellent hand-outs with photocopies of 'the screen as you see it' as a wonderful 'idiot's guide'. I recommend you to ask your library if it does, or could, offer the same service but remember, as in most computer programmes, you may still need the 'kick-now' button.

TRANSPORTING CHILDREN IN WHEELCHAIRS

- Notes from a Phone Call -

Enquiries are coming in regularly about this complex issue but succinct advice is not easily obtained. However, help is available from the Medical Devices Agency in the person of Alan Lynch. Alan is currently working with the Department of Transport to produce a Guidance Document on 'the major points to be considered when transporting people in wheelchairs'. Initially this was to be one side of A4 but it's grown into a much bigger production which will, I believe, update the current Code of Practice VSE 8.7/1.

Alan's main recommendation will be/is that the three agencies involved, namely: Transport (e.g. Minibus company), Suppliers (e.g. Wheelchair Centres), and Commissioners (e.g. Education department, Social Services) should talk to each other and agree joint responsibilities. At present a wheelchair is often/usually supplied without prior discussion between the agencies and in some areas a culture of non-co-operation appears to exist. Resource constraints and pressures are the obvious causes but things will have to change.

The child/client's physiotherapist, as the professional who recommended the wheelchair, is often the person to raise issues of safety, such as, 'have the chair and restraits been *crash tested'*. But this question is only part of the problem; a problem which needs tackling at its source. The term *crash testing* is misleading too. Are all crash tested chairs/restraints 100% safe? Are all untested systems *unsafe?*

So what can be done? We need to raise awareness of the problem; talk to the agencies concerned; get them to talk to each other, and look out for the Guidance Document. It is due out - er - soon!

In the meantime, Alan Lynch is happy for you to 'phone him on 01253 596000. Good Luck!

APPLICATION FORM FOR APCP PUBLICATIONS

TITLE	PRICE	QUANTITY
Serial Splinting in Hemiplegic Cerebral Palsy by Margaret Jones (2nd Edition)	£3.50	
The Children Act 1989 'A Synopsis for Paediatric Physiotherapists'	£2.50	
Paediatric Physiotherapy Guidelines for Good Practice	£2.50	
Dyspraxia - A Handbook for Therapists by Michelle Lee and Jenny French	£5.50	
Guidelines for Calculating Caseloads	£1.00	
Baby Massage	£1.00	
Standards of Practice for Paediatric Physiotherapy	£2.50	
Statutory Assessment of Children and Special Educational Needs	£4.00	
Tests and Measures Resources Pack (2nd Edition)	£3.50	
Haemophilia Booklet	£3.50	
Human Postural Reactions - Lessons from Purdon Martin by Dr. John Foley	£5.00 (incl. of P&P)	
Manual Handling Booklet	£10.00 (incl. of P&P)	

			TOTAL BOOK ORDER	£
* Post and Packing	Single Copies	£0.50	*POST AND PACKING	£
	2 - 5 Copies	£1.00		
	6 - 10 Copies	£2.50	TOTAL :	£
	over 10 copies on request			

TERMS: **STRICTLY CASH WITH ORDER**
**Cheques and postal orders should be made out to "APCP Publications" and included with order.
(International Money Orders accepted)

SEND ORDERS - WITH PAYMENT to :
Eileen Kinley, Superintendent Physiotherapist,
Royal Liverpool Children's NHS Trust, Alder Hey Hospital,
Child Development Centre - Physiotherapy Department, Eaton Road, Liverpool L12 2AP

Name and Address for delivery: .....

.....

.....

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OVERSEAS

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Huntingdon PE19 4JG

NORTHERN IRELAND

The committee would like to thank Adare Brady very much for all the hard work she has put in as chairperson, and wish her all the best in her new appointment on National Committee.

We have been very busy organising the evening meetings for the incoming year.

The following dates have been confirmed:

Monday 20th September 1999 - ' Update in Genetics' with Dr Fiona Stewart.

Monday 8th November 1999 - ' Seating ' with Peter Watson

Monday 7th February 2000 - ' Head Injuries ' with Carrie Spence.

The last meeting is still to be confirmed.

Hopefully, we will be able to follow up the 'Introduction to Bobath Course', with a study day on the Neurophysiological basis of the Bobath Concept, with Margaret Mayston possibly in early February

JUDITH MORRISON

NORTH EAST

I would like to welcome Sue Rumbold and Sam Arch onto the Committee as co-opted members .

We had excellent speakers for the 'Coping with loss' Study Day in May. Unfortunately, the course was poorly attended which must have been disappointing for the speakers.

The re-run of the 'DCD' Study Day in July, due to the overwhelming interest in February, was again very successful and provided many ideas for treatment sessions.

We have not had any response to our plea for suggestions for Study Day topics or venues. The N.E. members will be receiving a questionnaire with the newsletter regarding this issue. Do please fill them in and return to me a.s.a.p. The replies will hopefully influence our future plans. We intend holding two consecutive Study Days midweek on Neurophysiology/Bobath updates - dates and speakers are being negotiated at present. It has also been suggested that we hold a Study Day on the different approaches of alternative therapy.

N.B. Please remember to apply for our next Study Day, CHRONIC FATIGUE SYNDROME, on Saturday, 18th September 1999 at the Postgraduate Centre at the York District Hospital.

MARY HARRISON

EAST ANGLIA

A Study day was held in Bedford on 'Postural/ Positioning' at night for children. It was run by Symmrtrikit. It was an excellent day with very good attendance. The speakers made it a lively and interesting session. Many thanks to Liz Waugh and her team for all the organisation.

Organisation for the Study day at Cambridge on 9th October on Gait Analysis given by Dr. J.P.Lin from Guy's Hospital is finalised. Information has been sent to C.D.C. bases and paediatric centres in the area. If you have not received any information, contact Kishan Kooner, sec.(01480 415266-work-). Numbers are limited to 50.

Fiona Down is preparing the study day in November at Huntingdon on the treatment of the pre-school child, given by Colin Stevens. If you have a child of this age who would be suitable for Colin to work with, do contact Fiona (01480 415203 work).We are hoping to make a video of the sessions for in-service training sessions in physiotherapy departments.

Finally, the charity, 'Kids in Chairs' are happy to come to the area to give a session on road-handling and wheel-chair usage for children in wheelchairs. If you are interested in having a session, contact Fiona Down for more information.

TRICIA BROSNAN

SCOTLAND

Firstly let me thank on your behalf Christine Shaw for her sterling work and stimulating reports over the last two years. Chris as you know has moved on to the giddy heights of being National Secretary and I am happy to confirm that she made an excellent start when we attended the CSP on the 9th of July.

Hello again from me! I am pleased to be able to start my new term as Regional Rep.on a very positive note following "The Introduction to Paediatrics Course". Glasgow Caledonian University was an excellent venue for a very well received course. The 37 participants and 2 MSc. students from Wick to Wales, the south of England to Bahrain were very flattering in their course evaluation as to the content and the high standard of the lectures. The scoring in the multi-choice questions was excellent being between 67% and 84%.

The Scottish membership partly as a result of the course and partly I am sure from Christine's "stern letter" is now a very healthy 134. The committee hope to have your continued interest.

Course plans for the Autumn are:

- Shared Adult / Paediatric ITU Respiratory Day on Saturday 2nd October at Yorkhill Trust Glasgow.
- An overview to DCD multidisciplinary working on Friday 19th November- venue Edinburgh. It will be led by Catherine McDerment. Head OT at RHSC Edinburgh.

Watch notice boards for further information or please contact your local rep. directly.

LESLEY SMITH

OVERSEAS

I am delighted to say that we now have 54 overseas members. I will risk being boring and repeat some of my last report. Although we probably will not meet very often we can keep in touch. Several members have written to me asking for help and information. I think that we are a wonderful network of paediatric physiotherapists, working throughout the world. As the world gets 'smaller' now that the Internet has spread so far and wide, we have even better opportunities to keep in touch. I would like to use e-mail more. If you have an e-mail address, please let me have it. So far no one has admitted to having an e-mail address but someone must have one! I now have the facility to use e-mail and only need to get myself organised to make use of it. If enough people have one, maybe we could have the occasional electronic newsletter.

In October our APCP conference is being held alongside the Chartered Society of Physiotherapy conference in Birmingham, which is in the centre of England. It's really easy to get to! If anyone is going to be there, please let me know and we can meet. See the centre of the March APCP Journal for all the details.

If you have any questions or ideas you want to share with the rest of the members, as well as writing to me, you could post a letter for publication to the editor of this journal. We are all interested to know what is happening in different parts of the world, so please try and write a newsy item for all to share. You can include pictures if you like. I am often asked for pictures of children having physiotherapy, so if you have any spare, please send them to me.

This year nearly half the overseas members did not rejoin until a reminder was sent. You should all get a letter to remind you about next year's payment, in the Autumn this year, so I hope that this will encourage you to renew early and to pay by UK direct debit if you have access to a UK bank account.

SUE WHITBY

WEST MIDLANDS

On Wednesday 14 July Mr Glithero spoke to us about botulinum wiith a well supported evening lecture entitled 'Botulinum - Poison or Panecea'? For this we tried The Royal Orthopaedic Hospital as a new venue. Please if anybody has any ideas on other venues we can use around the venue, your committee would be glad to hear from you.

Your committee are busy finalising the details of our autumn evening lecture programme which will include topics based around the use of intra-thecal bacclofen and spinal rhisotomy, functional electrical stimulation and forensic pathology. Details of these will be circulated locally, so keep your eyes open. If you do not see or hear what is going on around the region in the next week or two, contact any committee member who will be happy to up date you, but please remember to spread this information around to your collegues.

Remember that if you have any thoughts on subjects for study days or evening lectures we would be glad to hear from you.

SALLY BRAITHWAITE

TRENT

Having recently taken over from Ann Peters I would like to thank her for all her hard work as Trent's previous representative.

The committee are at present working hard planning the APCP conference for 2001. Anyone interested in assisting with the organising of this, please get in touch.

In May we held a very interesting brain storming session relating to the management of children in schools. It was lovely to see so many people attend, which merely emphasises what a thought provoking area this is for all of us. Following this meeting a working party has been formed concentrating initially on manual handling. We will keep you informed of their progress.

The committee are keen to run more study sessions around the region including a Halliwick course provisionally set for January, and a Medico-Legal day.

The Trent region continues to grow and I would like to welcome all new members who have recently joined us.

LOUISE KELLEHER

SOUTH WEST

The Study Day on 26 June at Poole General Hospital on Orthotics was well attended and provided food for thought on the provision and application of various orthotics.

The next Study Day is provisionally on Saturday 20 November 1999 at Taunton, on Orthopaedics, including physiotherapy management and surgery for scoliosis and the management of paediatric sports injuries and adolescent knee problems. There will be further details nearer the time in the SIGs section of Frontline or contact me.

The Study Day / AGM in March 2000 is planned on Syndromes (eg Rett and Angleman) but no details have been finalised.

The Wessex CP Workshop on the older child, held at Lord Mayor Treloar College sparked a lot of interest and would perhaps provide the basis for a future Study Day?

Future workshops are planned on Sports for Children with Disabilities at LMTC and possibly one on Wheelchair Training at Dorchester. So far it has not been possible to arrange one on Conductive Education as planned.

Please continue to send me your news and views.

PAM EVANS

NORTH WEST

The Erbs Palsy study day held in May was well attended. It is hoped to do an article for the Journal summarizing the day's key points.

Pat Alexander is to run a Paediatric Manual Handling seminar in September at Booth Hall (see June Journal) for details.

The 2000 AGM is planned for early March - venue Trafford General Hospital. Topic - Lycra Suiting.

Further Courses are planned for 2000 June - The principles of the approach to Neurological Treatment and November - Premature Babies.

Please keep sending your ideas for other topics. Don't forget - study Bursaries are available to N. West members - contact Lorna Stybelska at R.M.C.H.

The Video Library continues to expand - if you know of any suitable video's we can consider buying - contact Gill Holmes at Alder Hey.

SUE WALMSLEY

WALES

It was lovely to see that we have four new members from West, Mid and North Wales. Croeso! Our membership is up to 45 now but I'm sure there's plenty more out there to keep on with the gentle encouragement!

In September, we're hoping that there will be a discussion afternoon on "Spines", following the very useful and interesting one we had with Lyn Horrocks in May on normal variations in Orthopaedics. Details yet to be confirmed but possibly on Wednesday 29th September at Caerphilly Children Centre again, with an informal lunch beforehand.

On October 29th there will be a Study/Discussion Day on Cerebral Palsy with David Scrutton at Hensol Conference Centre. It should be an excellent day so make sure you apply now! Details in the courses section.

The Serial Splinting Course will hopefully be later on in the Autumn and will be run by the Swansea gang! Looking ahead to next year's programme.

March :	AGM/Paediatric Manual Handling with Pat Alexander
May :	Muscular Dystrophy => to be arranged at a venue in Mid Wales for easier access to our friends from around there and further North.
July :	Care of the Terminally Ill Child
Oct./Nov. :	PMLD

Ideas for the future keep coming in and they include:

• Sensory Integration (apparently we're due this one!)

• Gross Motor Function

Few other things to mention are that we're hoping to set up a video library so if you know of any good ones that we can buy, let me know. Also, I'm looking into restarting a newsletter which will be sent to all members, possibly every April and September, with all the latest news, gossip etc!

Finally, on behalf of APCP WALES, I'd like to wish Rachel all the best on her exciting new life down under in Sydney. She will be missed by the Committee but Rachel, perhaps we can have a meeting at your new house?!!! Good Luck!

SIAN HOWELLS

NORTH EAST

CHRONIC FATIGUE SYNDROME

Saturday 18th September 1999
10.00 - 15.00
Postgraduate Centre, York District Hospital
Course Tutor : Various speakers.
Fee : APCP members £20.00, non-members £25
Please make cheques payable to APCP-NE.
Send to :
Jane Howland - Treasurer
8 Piper Road,
Hutton,
Driffield,
East Yorkshire
YO25 9YY
Tel: 01377 270149

WALES

STUDY/DISCUSSION DAY ON CP

Date : **Friday 29th October 1999**
Venue : **Hensol Conference Centre, Mid Glamorgan**
Speaker : David Scrutton
Senior Lecturer in Physiotherapy and Bioengineering
The Institute of Child Health
Cost : £45 APCP Members
£60 Non Members
Closing Date : 30.9.99
For further information, please contact :
Julie Williams
Paediatric Physiotherapy Dept.
Carnegie Clinic
Trealaw, Rhondda
Tel: (01443) 682130

APCP National Conference 2000

·2000·

University of the West of England
Bristol

Thursday 4th May to
Saturday 6th May 2000

'Partnership for Change'

Call for Poster Presentations

Anyone wishing to present a 'Poster' should contact Fiona Corkhill at :

Child Health Services
Loddon NHS Trust
'G' Floor
The North Hampshire Hospital
Aldermaston Road
Basingstoke
RG24 9LZ

Concept Training present "A MULTI-SENSORY ENVIRONMENT FOR SPECIAL CHILDREN"

The Kegworth Hotel, Kegworth, Derbyshire
13th & 14th October 1999
Keynote Speaker : **Flo Longhorn**
Author of Nine Books on Special Education

- Flo Longhorn: "Planning for Effective Learning, "Using Ultra Violet Light to Enhance Learning", "Auditing Happiness - what makes the individual child tick?
- Marion Cornick: Headteacher The Loddon School, "A Sense of Freedom for Children with problems Associated with Autism."
- Judy Denziloe: "Action for Leisure" Author of "Fun & Games": "Sensory on a Shoestring"
- Christina House: "Music Making with PMLD Children"
- Carol Penrice: "Introducing Aromatherapy & Massage"
- Martin Battye: "Approach to Colour"
- Tim Naylor: "Why Switch?" "South & Technology"
- Concept Training "Effective Use of your Effects Projector"

Concept Training, 19 Church Brow, Halton, Lancaster
Tel: 01524 832828 Fax: 01524 832852

ASSISTANTS COURSE

12th November 1999
Chailey Heritage Clinical Services
One day basic paediatric course - 9.30 a.m. - 3.30 p.m. including lectures covering posture management, common conditions and different approaches and treatments.
Various practical workshops in the afternoon.
Fee : £25 including a light lunch.
For further information : Physiotherapy Department, Chailey Heritage, Clinical Services, Beggars Wood Road, North Chailey, Near Lewes, East Sussex, BN8 4JN. Tel: (01825) 724703

HARE ASSOCIATION FOR PHYSICAL ABILITY

Date : Saturday 23rd October 1999
Time : 9.00 a.m. - 3.00 p.m.
Venue : The Church Centre, Bramcote, Nottingham
A workshop to explore posture and postural problems in adults and children with neurological disability. Using video case studies we will look at the analysis of posture, predicting problems, preventive therapy, and outcomes. Open to people from all disciplines with an interest in posture and movement.
Workshop Leaders : Pauline Pope, Nick Williamson
With contributions from other HAFPA members.
Fee : Members £30 Non-members £35
Applications to : Sue Newman, 12 Coval Road, East Sheen, London SW14 7RL. Tel: 0181 876 5086

EUROPEAN ACADEMY OF CHILDHOOD DISABILITY

11th Annual Meeting

Date : 21 - 23 October 1999
Venue : Senate House, London
Please contact Samantha Greshoff on 0181 333 6311 if you would like a copy of the conference programme and registration form.

Enhance and develop your skills - it's childs play

SENIOR I PHYSIOTHERAPIST

with an interest in Paediatrics

Term-time only **£19,640 - £23,130 pro rata** **Merseyside**

Linked to Alder Hey RLC NHS Trust, we are based at a school for children with physical difficulties situated just off the M57/M58 in a pleasant residential area, with exceptional facilities including a multi-sensory unit and a hydrotherapy pool. We are now looking for a Senior I Physiotherapist, to work term-time only in a warm and friendly atmosphere where experience with children is not essential but a lively approach and the enthusiasm to motivate is.

Providing a comprehensive physiotherapy service, your caseload will primarily involve patients with neurological conditions. Managing, treating and caring for children in a range of settings - special schools, nurseries, mainstream schools and the child's own home, you'll build strong relationships by offering advice and support to parents, carers and teachers.

With a minimum of 4 years' general experience and an interest in Paediatrics, you'll be a pragmatic problem solver with excellent organisational skills and the drive and initiative to develop your role and skills in a team orientated environment. In return we'll offer you comprehensive training where required and the assistance of a well established team including opportunities to work alongside other physiotherapists, OTs and speech and language therapists.

Working hours are 36 per week with a degree of flexibility.

For an informal chat and more information contact Carole Lovatt, Superintendent Physiotherapist or Sue Saville, Senior I Physiotherapist on 0151 928 0307.

For an application form, job description and person specification, please contact the Human Resources Department, Alder Hey - RLC NHS Trust, Eaton Road, Liverpool L12 2AP. Phone 0151 252 5339. Please quote reference: LCPC 996.

Closing date: 30th September 1999.

The Trust is committed to carefully screening all job applicants to ensure the safeguard of children.

Alder Hey

ROYAL LIVERPOOL CHILDREN'S N.H.S. TRUST

Open to Job Share • Working towards equal opportunities • We operate a restricted smoking policy

Child Health Directorate

Development Opportunity

• Are you an experienced Paediatric Physiotherapist?

• Are you passionate about the need to improve services to children?

• Are you a good communicator?

• Are you ready to develop your management skills?

If your answer is yes, then you are the person we need as our

SUPERINTENDENT IV
PAEDIATRIC PHYSIOTHERAPIST

Salary on point between £19,640 and £23,130 p.a.
based on 36 hours per week

Based at the Child Development Unit this post carries responsibility for the clinical physiotherapy input to the unit and we would expect you to become involved in the plans for implementing the recommendations following a recent review of the CDU.

You will support the Superintendent in the day-to-day management of the service and act as deputy in her absence. There is a move towards better communications between services to children in Tameside, with an under 8's project underway and plans for the inclusion of children with special needs into local schools. An interest in other paediatric specialist areas would also be welcome.

We can offer you:

- A supportive, friendly multi-disciplinary team.
- Personal Development opportunities through appraisals and in service training.
- Some funding towards training.
- Flexibility in working arrangements.
- Online access for up-to-date information.
- In-house management training.

For further information, informal visits etc., please contact Mrs Linda Whitaker, Supt. Physiotherapist, on 0161 368 4242.

For a job description and application form please contact: The Personnel Department, Tameside & Glossop Community and Priority Services NHS Trust, Tameside General Hospital, Fountain Street, Ashton-under-Lyne, Lancashire OL6 9RW. Tel: 0161 331 5111 (24 hour ansaphone), minicom 0161 331 5371 quoting reference number P129/99J. Closing date: 30th September 1999.

The Trust is working towards being an equal opportunities employer.

Tameside & Glossop

SENIOR I AND II PHYSIOTHERAPISTS

Directorate of Child Health

SENIOR I PHYSIOTHERAPIST

Child Development Centre, Plymouth

£19,640 - £23,130 p.a.

Due to promotion, we now have a full-time vacancy for a Senior I Physiothe.rapist.

You will provide a service for children who have a variety of conditions including cerebral palsy, syndromes, juvenile arthritis and dyspraxia. You will be working within the Child Development Centre at Scott Hospital - part of Plymouth Hospitals NHS Trust, in mainstream schools, special schools and outlying clinics and you will be liaising closely with education and social services.

You will act as a lead person within the team which includes Senior I's and II's and Therapy Assistants. You will have a minimum of 2 years' post graduate experience and 18 months' experience as a Senior II in Paediatrics.

Continuing professional development is actively encouraged. The ability to travel by car to various local locations is essential.

For an informal discussion about the position, please contact Mrs Heather Warner, Support Manager on 01752 763464 or the Physiotherapy Department on 01752 284341, quoting reference CH-57.

SENIOR II PHYSIOTHERAPIST

Woodlands School, Plymouth

£16,525 - £19,640 p.a.

You will be providing a physiotherapy service as part of a supportive, multi-disciplinary team for a range of mixed ability children with neuromuscular disorders, aged 3 - 17 years, in both a special school and mainstream setting. The physiotherapy team is managed by a Bobath-trained Senior I. The department takes students on clinical placement. Experience is also offered in orthopaedic and wheelchair services, hydrotherapy (on-site), orthotics and splinting. In addition, you will work with families on home visits and at the local child development centre in the school holidays. Regular in-service training is provided and close links maintained with local physiotherapy departments (acute and community).

This is an ideal position for someone wishing to gain a wide range of experience in the paediatric setting. An ability to relate to children and good communication skills are vital, although previous paediatric experience is not essential.

For further information regarding this position, please contact Ms Sue Bearne on 01752 785416 or Ms Janet Clarke on 01752 272414, quoting reference CH-50a.

SENIOR II PHYSIOTHERAPIST

Child Development Centre, Plymouth

£16,525 - £19,640 p.a.

You will be providing a physiotherapy service as part of a supportive team. The service is for children who may have a variety of conditions including cerebral palsy, syndromes, juvenile chronic arthritis and dyspraxia. You will play a vital part of the multi-disciplinary team with opportunities for joint working and shared practice.

You will need to have a genuine interest in working with children and the ability to learn new skills under the supervision of experienced Senior I Physiotherapists. We would welcome applicants who have paediatric experience or who wish to take their first step in this rewarding specialist area. The role will provide experience in hydrotherapy, orthotics and splinting and will provide the opportunity to work with children in mainstream schools and nurseries, as well as special schools.

We have an in-service training programme with a commitment to continuing professional development.

For further information regarding this position, please contact Mrs Heather Warner, Support Manager on 01752 763464 or the Physiotherapy Department on 01752 284341, quoting reference CH-51a.

For both Senior II positions, you will be a Chartered Physiotherapist with a minimum of 2 years' post graduate experience. The ability to travel by car to various local locations is essential.

Our facilities for staff working within the Trust include a health and leisure centre, social club, day nursery and extensive library. And with Plymouth, you'll soon appreciate the coastline, countryside and lively city atmosphere.

An application form and further details for any of the positions can be obtained from Michelle Thorburn on 01752 792121, quoting the relevant reference.

The closing date for receipt of completed application forms for all positions is 27 September 1999.
Late applications will be considered.

The selection process will consist of an informal visit and interview.

Plymouth Hospitals NHS Trust is an equal opportunities employer and is actively working towards a smoke-free working environment. The Trust operates a Green Commuter Strategy.

PLYMOUTH HOSPITALS NHS TRUST

WALSALL COMMUNITY HEALTH TRUST

PHYSIOTHERAPY DEPARTMENT

SENIOR 1 - PAEDIATRICS (SCHOOL HEALTH)
36 HOURS PER WEEK

The Paediatric Team provides physiotherapy for a wide variety of children with special needs within various community settings, mainstream schools, special schools, clinics and domicillary.

We are looking for an experienced and enthusiastic physiotherapist to join our well established team of 8 physiotherapists and 5 physiotherapy assistants.

You will have a varied caseload of conditions and ages and work closely with other members of the MDT. IPR's, Peer Review and CPD are all actively encouraged.

A car owner/driver with a current, clean licence is essential. If you feel you are suitably qualified and would like other further information or to arrange an informal visit please contact:

Suzanne Rimmer (Superintendent III PT) or Susan Morris (Physiotherapy Services Manager) on 0121 480 5953.

For an application form and job description please write to the Physiotherapy Department, 5 Lakeview Close, Queslett Road, Great Barr, Birmingham, B43 7EZ, quoting job reference number 32/99.

Closing date for applications: 17 September 1999
Interviews are expected to take place: commencing from 1st October 1999

AN NHS TRUST COMMITTED TO EQUAL OPPORTUNITIES
WE OPERATE A NO SMOKING POLICY

NOTICE

The charges for Recruitment Advertisements are :

FULL PAGE: 15 cms x 19 cms £150

HALF PAGE: 15 cms x 9 cms £75

Advertisements should be submitted complete with typesetting and artwork, if possible.

Notes for Contributors

The Editorial Board welcomes research material; referenced articles and evaluations of physiotherapy practice; informal articles.

Manuscripts should be sent to Lin Wakley, 2 Ash Bank, Pipers Ash, Chester, Cheshire, CH4 7EH, U.K.

Copy to be submitted should be typed on one side of the paper, double spaced and with ample margins. All pages should be numbered consecutively.

Manuscripts should provide the title of the article and the author(s) name(s) and full postal address for correspondence.

References should be given in the Harvard System.

In text	Author(s) name and initials followed by the date of publication. Use a,b, to indicate more than one publication in the same year. Where there are 3 or more authors use first name followed by et al.
For books	Laszlo, J. & Bairstow, P. (1985) *Perpetual Motor Behaviour* (Rinehart and Winston)
For chapters within books	Morley, T.R. (1992) Spinal deformity in the physically handicapped child, in : G.T. McCarthy (Ed). *Physical Disability in Childhood* (Churchill Livingstone)
For articles	Scott O.M., Hyde S.A., Goddard C.M., Dubowitz V., (1981a) Prevention of deformity in Duchenne muscular dystrophy. *Physiotherapy* 67(6), 177-80.

Tables and Figures

The approximate position of the tables and figures should be indicated in the manuscript.

Keys to symbols should be included.

Tables should be numbered by Roman numerals and figures by Arabic numerals.

Figures should be supplied in a finished form, suitable for reproduction. Figures will not normally be redrawn.

Proofs will be sent to authors if major alterations have been made to the text.

The Editorial Board reserves the right to edit material submitted for publication.

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In this issue

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Severe Combined Immunodeficiency

plus

A Visit to a Danish CF Centre