Articles in this issue ...

Assisted standing - experiences of children with cerebral palsy and their physiotherapists

The role of hypertonic saline in airway clearance in pre-adolescent and adolescent CF patients

Selective Dorsal Rhizotomy – an overview

ASSOCIATION OF PAEDIATRIC CHARTERED PHYSIOTHERAPISTS
JOURNAL

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Aims and scope: the APCP Journal aims to publish original research and other scholarly work related to paediatric physiotherapist – its scientific basis and clinical application, education of practitioners, management of services and policy.

For enquiries relating to the submission of articles contact:

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We should like to thank the following people who kindly reviewed articles for us:

Di Coggins  Chris Morris
Joy Conway  Cate Naylor
Sarah Crombie  Diana Oford
Denise Hart  Terry Pountney
Jeanne Hartley  Jane Reid
Lesley Katchburian  Amanda Trees
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**Submission to the APCP Journal**
EDITORIAL

Eva Bower – APCP Editor

Disability need not hold individuals back and we should strive to eradicate stigmatisation. The polymath Sir Isaac Newton was born to farming folk in Woolsthorpe, near Grantham, on Christmas day, 1642. His mother was widowed during the pregnancy and Isaac was born prematurely and underdeveloped. Legend has it that he was so small that he could be put into a quart mug (1.1 litres) and had to wear a supporting device around his neck to hold his head upright. He was not a sociable child and his early years were rather lonely. Nevertheless, none of this prevented him from becoming the leading mathematician, physicist and natural philosopher of the enlightenment as well as allegedly an alchemist. He was a secretive man who never married and had many problems communicating with people. However, he did acknowledge the role of other scientists in his success; he wrote to Robert Hooke in 1676 “if I have seen further it is by standing on the shoulders of giants”. Maybe in modern times he would have been labelled with Asperger’s syndrome (Bixby W, 1966).

Selective dorsal rhizotomy (SDR) is again in vogue. The last time round was in the late 1980’s when Warren Peacock talked to audiences at Great Ormond Street Hospital, London. Fashion in medicine, as other fields, seems to come round in a circular pattern, our readers may like to read Grunt, Becher and Vermeulen’s systematic review (2011), along with the articles by MacWilliams et al (2011) and Tedroff et al (2011) describing their original research. In this issue there is both a review of SDR by Burslem and a precis of the NICE guidelines by Coombe.

In this edition we also publish two clinical policy articles from physiotherapy community services that suggest that health services working collaboratively with those working in educational services may improve the services to children - in the first case, for children with developmental co-ordination disorder (DCD) and in the second article, for children with profound and multiple learning difficulties (PMLD). These findings would seem to make good economic sense in these times of cuts and budget restraints. We also have a paper from an acute physiotherapy service advocating the replacement of a generalist who undertakes many duties, with the services of a specialist in screening for the detection of developmental dysplasia of the hip (DDH). I wonder if this is economically viable in the current financial climate and what the cost implications and difference in long term outcomes really are.

Last but certainly not least, I would like to thank all those people who have responded to our call for submissions of manuscripts to the Journal and those who have so generously given up their time to review articles. Without them there would be no peer-reviewed journal. Please submit your papers for the spring edition of the Journal by email to va@apcp.org.uk by 1st January 2012.

References


Assisted Standing
Experiences of children with cerebral palsy and their physiotherapists

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ABSTRACT

Background
Standing frames or supports are used to help children who cannot maintain a well aligned standing position independently. Physiotherapists cite physiological and psychosocial benefits from the use of such equipment. The research aimed to explore the experience of assisted standing of individual children with cerebral palsy (CP) and the factors which influenced their physiotherapists’ prescription and delivery of their standing programmes.

Method
Semi-structured interviews were conducted with nine children with CP who had used, or were currently using, standing frames and with the children’s paediatric physiotherapists. Data from interviews of children and physiotherapists was analysed.

Results
The children perceived little, if any, immediate benefit from standing and generally could not identify any activities which they performed better in standing. Some found standing in standing frames uncomfortable, others experienced discomfort after standing. Despite this most children continued to stand regularly and cited benefits such as strengthening or straightening their legs which they hoped to achieve by standing. The physiotherapists were convinced that assisted standing was a valuable adjunct to treatment.

Conclusion
Both the children and physiotherapists interviewed thought that use of a standing frame would confer improvement in body structure and function. The therapists’ opinions that there are also psychosocial benefits are not born out in the children’s accounts. Prescription of standing equipment relies more on pragmatic issues such as what is readily available than on what is the optimum for the individual child.

Introduction
Children with cerebral palsy (CP) who cannot maintain a well-aligned, symmetrical standing position independently often use standing frames or supports to help them stand. In this way they can experience the upright position and begin to learn appropriate postural mechanisms to maintain it (Green et al 1993). Physiotherapists cite many goals for assisted standing: improvement in function; maintenance or improvement of bone mineral density; maintenance or improvement of the integrity of the developing hip joint; maintenance of soft tissue length; reduction of muscle tone; physiological improvements in bladder, bowel and respiratory function; and positive psychosocial effects (Wintergold, Pountney and Cowan 2008). Where the main aim of such assisted standing is to improve function, the physiotherapist may use the equipment to position the child, freeing the child to facilitate tasks requiring a higher level of postural ability than the child can attain unaided. Where the goals involve maintenance of bone mineral density, improving or maintaining the integrity of the hip joint or soft tissue length, the standing is more likely to be carried out as part of a programme of postural management. A combination of these goals for standing may be used within home or classroom based activities administered by a parent, teacher or learning support assistant. There is some, albeit limited, research evidence to support the various goals of standing. Wren et al (2011) measured volumetric cancellous bone mineral density (BMD) using quantitative computed tomography in 37 ambulatory children with CP, Gross Motor Function Classification System (GMFCS) levels I-IV. They found that children with CP of all levels have less bone in their tibias than normally developing controls and that vertebral BMD was decreased in children at GMFCS levels III-IV. The authors concluded that increased weight bearing and physical activity would benefit these children by aiding BMD accrual. Pin (2007) in a review of the
effectiveness of static weight-bearing exercise in CP concluded that there was ‘good evidence’ to support the use of weight bearing activities (including standing frame use) to increase vertebral BMD (Cautlon et al 2004) and femoral BMD (Chad 1999). Herman (2007) and Kecskemethy (2008) showed significant variations in weight bearing during standing in passive standing devices varying from 37% to 101% of body weight, according to type and inclination of standing support. A review of the evidence for assisted standing in CP (Bush et al 2010) found that while static standing may be useful in maintaining BMD in CP the exact duration and frequency remained open to debate and that the angle of stand needed to be carefully considered if weight bearing was a goal of standing. For maintaining the integrity of the hip joint, properly directed weight bearing is crucial (Gudjonsdottir 1997). Early changes to hip architecture have been identified by Scrutton and Baird (1997, 2001) who highlighted the need for early surveillance and intervention to prevent hip subluxation. In a retrospective study of 59 children with CP, 24 hour postural management was demonstrated to help to maintain integrity of the hip joint (Pountney 2002). Morton et al (2006) studied 110 individuals with CP retrospectively and concluded that postural management alone would not be sufficient for many children but should be seen as complimentary to surgery. Hagglund et al (2011) introduced a successful screening programme, with early intervention to prevent dislocation of the hip in children with CP. Orthopaedic surgery, in some cases in conjunction with intrathecal baclofen or selective dorsal rhizotomy, was used to manage hip subluxation. The place for assisted standing was felt to be in the postoperative management of these children. Pin (2006) reviewing the evidence for the effectiveness of passive stretch in children with CP found that there was evidence for improvement in range of movement with sustained stretching (such as standing frame use) but the effect size was small. Two studies, Tremblay et al (1990) and Richards, Malouin and Dumas (1991) which involved only small numbers of subjects and one session of muscle stretch, showed lowered EMG and improved muscle activation after a 30 minute stretch to triceps surae using a modified tilt table. The effects did not translate to functional improvement and the duration was not measured after 30 minutes so the significance of this reduction in tone is uncertain. No published evidence could be found for anecdotally reported effects on bladder and bowel function in CP, nor for psychosocial effects. Within clinical practice, it is evident that some children do not like standing and refuse to stand despite every encouragement. There is no published research eliciting the views of children about the standing supports they use. The aim of this research was firstly to understand the children’s individual experience of standing using a standing frame. Secondly it aimed to explore the clinical reasoning of their physiotherapists and the way in which standing programmes are implemented.

Method

The research proposal was approved by Brent Medical Research Ethics Committee and St Marys Hospital Ethics committee (REC reference: 06/Q0408/7). Potential participants were approached by their paediatric physiotherapist and the research explained to them. If they expressed an interest in taking part, an information pack was sent home and written informed consent was obtained from the parent. Each child signed a consent form or had their assent documented.

Children with any presentation of CP, who were able to communicate their feelings, and who were using, or had used, standing frames or supports were included in the study. Children were excluded where either parental or their own consent was not given.

The children were interviewed individually within their school settings or at home, using semi structured interviews with open questions to allow them to introduce their own thoughts and concerns. The interviewer for all children was a senior paediatric physiotherapist (SB), who was not the child’s named physiotherapist at the time of interview. Interviews were digitally recorded and transcribed verbatim. Eight children were also videoed to record non-verbal communication. After reviewing the videos, non verbal communication, such as nodding, shrugging or smiling, was recorded in the transcription. The children’s age, school placement, clinical presentation and GMFCS level (Palisano et al 1997) were also recorded.

The responses of the first two participants (one in mainstream and one in special education) were used to inform the structure for the discussion in two focus groups with paediatric physiotherapists working in either mainstream or special education. The focus groups were recorded and transcribed verbatim for analysis.

Using information generated by the first two interviews and the focus groups, the child interview format was refined and an interview schedule composed for the individual physiotherapists who treated each child.
The individual physiotherapists treating each child were approached after the child had consented to participate and asked if they would agree to be interviewed. Interviews took place at a time and place convenient to the participant, were digitally recorded and transcribed verbatim for analysis. All interviews were conducted by SB.

Data analysis
Thematic analysis of the interview data was carried out from a critical realist perspective using the framework set out by Pope and Ziebland (2000) which was developed for research in which the aims of the research are set out in advance. The framework is grounded and inductive as it reflects the children’s accounts, but starts deductively from its preset aims.

The first stage of data analysis identified key issues, concepts and themes and formed an index from line by line analysis of the transcribed interviews. The index was then systematically applied to the data within the interview transcripts. Some passages of data encompassed more than one theme. The transcripts were annotated using colour coding of themes and concepts, and additional scribed comments.

In order to guard against researcher bias and ensure the trustworthiness of the analysis, the un-coded transcripts of all interviews were given to a colleague (RB) experienced both in qualitative research and in working with children with CP. A comparison between the interpretations of the data was made, and additional themes or differing interpretations were explored.

In order to minimise the possibility of bias in the analysis of the results a summary of their interview was returned to each of the participants to allow them to validate the interpretation of what they had said. The children were asked by a carer or the interviewer whether or not the summary reflected their expressed views. One child was unavailable to participate in this process, so the child was sent the summary and asked to contact the interviewer if she disagreed with the interpretation. Physiotherapists sent their comments to the interviewer by email. Four children were revisited within a year of their first interview, to explore themes further. Data from these interviews was analysed and any new themes integrated into the overall analysis.

Results
Nine children agreed to be interviewed but only seven yielded usable data and were included in the analysis. Two preschool children who agreed to be interviewed were either unable or unwilling to express their views on standing. The mothers were present at the interviews and offered interpretations but this data was excluded to avoid bias. The demographics of the child participants are shown in Table 1 and the five participating physiotherapists in Table 2.

<table>
<thead>
<tr>
<th>Child</th>
<th>Age (yrs)</th>
<th>gender</th>
<th>School</th>
<th>Presentation</th>
<th>GMFCS level</th>
<th>Interview site</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>9</td>
<td>Female</td>
<td>S</td>
<td>Hypotonia, mild dystonic athetosis</td>
<td>IV</td>
<td>School</td>
</tr>
<tr>
<td>2</td>
<td>13</td>
<td>Female</td>
<td>M</td>
<td>Spastic diplegia</td>
<td>II</td>
<td>School</td>
</tr>
<tr>
<td>3</td>
<td>13</td>
<td>Male</td>
<td>S</td>
<td>Athetosis with spasticity</td>
<td>III</td>
<td>School</td>
</tr>
<tr>
<td>4</td>
<td>10</td>
<td>Male</td>
<td>S</td>
<td>Spastic diplegia</td>
<td>III</td>
<td>Home</td>
</tr>
<tr>
<td>5</td>
<td>14</td>
<td>Male</td>
<td>S</td>
<td>Athetosis with spasticity</td>
<td>III</td>
<td>School</td>
</tr>
<tr>
<td>6</td>
<td>9</td>
<td>Male</td>
<td>M</td>
<td>Diplegia</td>
<td>II</td>
<td>School</td>
</tr>
<tr>
<td>7</td>
<td>11</td>
<td>Female</td>
<td>S</td>
<td>Spastic quadriplegia</td>
<td>IV</td>
<td>School</td>
</tr>
</tbody>
</table>

(Abbreviations: M = mainstream school; S = special school; GMFCS = Gross Motor Function Classification System)

<table>
<thead>
<tr>
<th>Physiotherapist</th>
<th>Age (yrs)</th>
<th>Gender</th>
<th>Where trained</th>
<th>Experience (yrs)</th>
<th>Caseload</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>33</td>
<td>Female</td>
<td>Holland</td>
<td>10</td>
<td>Community</td>
</tr>
<tr>
<td>2</td>
<td>48</td>
<td>Female</td>
<td>England</td>
<td>26</td>
<td>Community</td>
</tr>
<tr>
<td>3</td>
<td>64</td>
<td>Female</td>
<td>England</td>
<td>21</td>
<td>Special school</td>
</tr>
<tr>
<td>4</td>
<td>52</td>
<td>Female</td>
<td>England</td>
<td>21</td>
<td>Special school</td>
</tr>
<tr>
<td>5</td>
<td>34</td>
<td>Male</td>
<td>England</td>
<td>11</td>
<td>Community</td>
</tr>
</tbody>
</table>
Child seven used a communication book, with gesture and some Makaton signing to communicate. At her initial interview it was impossible to ascertain her views using her communication book, despite the presence of her mother and her speech therapist. This was partly due to lack of vocabulary and the need to flick from page to page for different categories. Her speech therapist introduced her to the talking mats system (Murphy 2002) (see fig.1) and a second interview was successfully conducted using this system.

The initial themes that emerged from the children’s interviews were:
1. reasons for standing
2. effects of standing (both positive and negative)
3. desire for change
4. control and acceptance.

The themes from the physiotherapists’ interviews were:
1. why standing is important
2. criteria for choosing equipment
3. implementing and monitoring standing
4. perceptions of children’s feelings about standing
5. reactions to children’s reluctance to stand

**Reasons for Standing**

The children’s reasons for standing focused on hoped for improvement in strength or alignment of their lower limbs - “It helps you stand totally up, makes your legs better” (child 1), “Makes me more stretched” (child 2), “To keep my legs straight and make my legs grow stronger” (child 6). Child 7 indicated a perceived benefit in getting out of her chair and stretching. These views are reflections of the paediatric physiotherapist’s perceptions of benefits of assisted standing, such as prevention of lower limb contractures and deformities and postural management and suggest that the children have accepted what they are told by their therapists about standing. Physiotherapists cited broader benefits to the child, including physiological (e.g. improved bone density), and psychological (e.g. motivation and social communication). “Motivation to do things … alertness … muscle length, joint range, improve posture … head control … bone density … hips” (physio 3).

The physiotherapists all clearly expressed that assisted standing was important to achieve physiological and functional short and long term goals and promote participation. ‘Change of position … postural management … prevention of contractures and deformities … (makes them) more alert … (improves) fine motor skills’ (physio 2). All physiotherapists acknowledged that there is limited research evidence to support their views, and cited their clinical experiences as supporting their use of standing supports. “I think it’s difficult because there’s not actually that much (evidence)” (physio 5); “Only from what you see yourself, you can see the ones who haven’t been stood” (physio 2).

**Children’s Experiences of Standing**

Overall the children reported negative experiences of supported standing, including pain, isolation and loss of participation. Only child 7 did not complain of pain during or after standing. The others all mentioned pain or discomfort which resulted in impaired participation or function. “(I get) a bit sore … It’s hard to do my work” (child 3), “When I come out … my legs are floppy … I can’t even stand up … it really hurts.” (child 6). Although standing was often uncomfortable all the children interviewed except child 3 were prepared to continue using a standing support. “It’s fine… it’s just normal” (child 2); “I can manage it … when I sit down (the pain) goes away” (child 6). Child 3 said “It’s hard to do my work … I don’t want to stand any more”. Although the children were of varying levels of motor ability, as reflected by their GMFCS scores, this was not seen to
influence their experience of standing. The more able children did not complain about being made to stand still, nor did the less able perceive benefit from the added postural control.

Some of the children reported feeling isolated, lonely or self conscious when using their standing frames. “I feel a bit left out” (child 7); “It’s only me standing” (child 3). Child 2 commented that she preferred to stand at the back of the class or she felt that everyone was looking at her. It would seem that the physiotherapist’s views of supporting children’s function and participation were not borne out by the majority of the children’s reported experiences. Child 4 had stopped standing at home, as he preferred to be able to play on the floor with his younger brother. The exceptions to this were child 1 who reported one benefit when in the standing frame “It helps me with the lightwriter” (keyboard operated speech output device) and child 7 indicated, using the talking mat, that it helped her legs, back and upper limbs.

Four of the five physiotherapists were aware that many children do not enjoy standing “There’s a small group who really enjoy it, the rest put up with it” (physio 2). This resonates with the children’s apparent passive acceptance of standing being part of their daily experience; an uncomfortable experience to be tolerated. “I think a lot of our children just do it because they have a lot of things that happen to them and that’s just one of those things that they do …” (physio 3). There is a sense that the children have used standing supports from their early years and have not developed into active participants in their physical management programmes.

However, there was evidence that the children had definite views on standing. Child 3 said “I don’t want to stand any more” and asked that his physiotherapist be told this. He reported that he had not asked her himself as he was worried she might be upset or annoyed with him. Child 6 said that when he wants to stop, but is told he has to go on “I get frustrated, very frustrated”. Others mentioned being made to continue but accepted this and were not troubled by it.

The physiotherapists recognised that there are some children who may refuse or be reluctant to use a standing frame. The physiotherapists’ reactions to this situation were guided by the child’s age and understanding and principles of consent. “If you’ve got a young person who’s 14, knows exactly why they’re standing and they really, really, find it hard and want to stop you’d listen to them … if you’ve got a three year old who cries when you do anything then I’d maybe push a bit longer because it’s more important for a three year old” (physio 3). It would seem that the physiotherapists considered a long term perspective of the perceived benefits of standing “... how much pain would they be in if they don’t stand ... a few years further down the line?” (physio 1).

Children’s Choice

The children had definite ideas about how they would like to change their standing supports. These proposed changes all indicated that they wanted more control over aspects of standing to improve their daily experience e.g. when, where, the length of time and the type of standing support used. “I’d like to stand after break ... I want a standing frame at home ... I want another one (electric) ... (so I can) press the button” (child 1). Child 6 said of his frame “I don’t like those bits at the side … they get in the way when I’m trying to get up.” Child 5 described difficulty getting his feet into the frame and child 2 wanted a lower frame. None of the children commented upon the overall design and appearance of the standing supports.

Most of the children in special schools were given some choice over which lessons they stood in, but those in mainstream schools stood when told to by classroom staff, and seemed to accept this as another part of the school timetable. Physiotherapists stated that they did intervene if a child did not like to stand in a particular lesson and negotiated with staff on their behalf.

Physiotherapist’s Choice

The physiotherapists had clinical reasons for choosing upright, prone or supine standers related to the child’s abilities, e.g. degree of head control, presence of contractures or deformities. Individual models of frame were chosen largely for pragmatic reasons. The first choice was usually from what was already available and/or known to the therapist “Availability is important … that does have to come top, I wish it didn’t but it does… You tend to use the same ones because you’re used to them and you’ve got them so you don’t have to ask for funding” (physio 3).

The physiotherapists all set up the equipment and check that the child was comfortable, but the actual process of standing was performed and timetabled by classroom staff. “The teachers say when they stand ... we have books that say when they’re standing and how long ... we have to rely on staff to tell us things” (physio 3). All physiotherapists regularly monitored
progress and checked equipment, usually on a half-termly basis, relying on school staff to report any difficulties in the interim. This consultative approach to delivering physiotherapy services, driven by resource limitations, may place distance between the child and physiotherapist, reducing the child’s opportunity to express their views and participate in decision making in relation to their standing supports.

Discussion
The World Health Organisation’s International Classification of Functioning, Disability and Health (ICF) (WHO 2001) (Fig 2) is increasingly used as a framework for evaluating goals and interventions (Wright et al 2008). Both the children’s reasons and the physiotherapists’ goals for standing relate mainly to the body function and structure arm of the model. Children and physiotherapists have differing perceptions about the use of standing support equipment. Physiotherapists articulated perceived benefits to impairment and participation through standing. For the children, although they acknowledged a hope of improvement to impairments, their lived out reality was different. Children reported pain, discomfort, reduced abilities to participate and for some a sense of isolation when standing.

Figure 2 - International Classification of Functioning, Disability and Health (ICF)

Choice of equipment was dictated by knowledge of a few commonly used frames. All the physiotherapists interviewed mentioned the same equipment models and manufacturers as a starting point for standing children with CP. Only if these did not meet the child’s needs would they consider other makes of equipment. In research for the MHRA, (Daniels et al, 2005) found that many participants preferred their trial frame to the one they usually stood in. Although this could relate to some children’s preferences for novelty, and could have been based on a non-therapeutic aspect such as the colour of the equipment, it does suggest that allowing children to try more than one piece of equipment at assessment might result in better outcomes. The four participants (child 1, 3, 4 and 5) with whom a second interview was conducted had all trialled a standing frame/balance trainer which allowed movement whilst being held in standing and all preferred this to static standing and reported less discomfort. In view of animal studies showing that bone density improves in response to dynamic loading, (Lanyon and Rubin 1984, Lanyon 1996) the development of dynamic standing equipment would be desirable.
The use and choice of equipment should be a collaborative decision between child, family and physiotherapist, based on a systematic assessment considering the environment, child’s functional abilities, and anticipated benefits in terms of impairment, activity and participation. This study suggests that the child’s abilities, resource availability and physiotherapists’ familiarity with specific equipment primarily determines the choices made.

This study has a number of limitations. All the children were enthusiastic about taking part in the interviews; however their responses were often limited to answering the questions asked. Some replies were monosyllabic, possibly reflecting the difficulty that many of the children had with the articulation of speech. The interviewer often needed to lead the conversation, assisting the children to elaborate on their responses, to elucidate their meaning. Care was taken to give choices that would lead to clarification of the original response rather than impose the interviewer’s idea of what the child meant. Krähenbühl and Blades (2001) reviewing the literature on techniques for interviewing children, reported that researchers found 40% of information gained during interviews came from option posing and suggestive prompts.

The interviewer was either a current or former colleague of the physiotherapists who participated, potentially limiting the scope for diverse views to be gained at interview. The study numbers are small and the study does not include parental or carer perspectives, which could add further concepts.

Conclusion

Assisted standing was at best uncomfortable for the majority of the children included in this study. They were willing to endure this discomfort in order to gain the reported benefits. They could not identify any immediate benefits themselves. All the physiotherapists acknowledged the lack of evidence to back their beliefs that standing was beneficial. Research is needed to identify the effects of assisted standing and to improve the design of standing equipment to minimise discomfort. Better funding for equipment would enable therapists to choose from a wider range of standing frames and supports to find the ‘best fit’ for each child. Physiotherapists should actively seek children’s views and be prepared to act upon them, in order to help them develop into active partners in their own health care.

1. Both children with CP and their physiotherapists expect improvement in body function and alignment from use of assisted standing equipment despite little supportive evidence.
2. This study found that most of the children reported pain or discomfort while using standing equipment.
3. The children interviewed did not experience the psychosocial benefits from standing that their therapists cite.
4. Prescription of equipment is influenced more by pragmatic issues such as availability than by clinical criteria for optimal positioning.
5. All children except one reported pain during or after standing; standing hinders participation; standing programmes take time and effort on the part of physiotherapists, school staff and parent/carers - further research is therefore required to verify the benefits or otherwise of the use of assisted standing within paediatric physiotherapy.

References


A Systematic Review

The role of hypertonic saline in airway clearance in pre-adolescent and adolescent Cystic Fibrosis patients

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ABSTRACT

Background
Cystic fibrosis is one of the most common life-threatening inherited diseases in the UK. It is characterised by a continuous circle of sputum retention, infection and inflammation. One current treatment option is nebulised hypertonic saline inhalation, which produces known improvements in lung function and sputum clearance in cystic fibrosis adults. There is limited knowledge about the effect of hypertonic saline inhalation during the early years of the disease.

Objective
A critical review was undertaken to investigate the role of hypertonic saline in airway clearance in pre-adolescent and adolescent cystic fibrosis patients.

Method
A literature search was conducted using the key words and a combination of key words including hypertonic saline, cystic fibrosis, airway clearance and sputum clearance. This included a structured search of the Cochrane Library, National Guideline Clearinghouse and Scottish Intercollegiate Guidelines Network, EBSCOhost, CINAHL and MEDLINE. A hand search of key journals and conference papers to locate papers was performed. Eight studies met the inclusion criteria and were examined thematically to determine the effect of hypertonic saline on lung function, sputum clearance and patient treatment compliance.

Results
Lung function improved following a multi-dose trial of 10mL of 5.85% hypertonic saline. There was an increase in weight of sputum cleared showed after a single-dose 10 minutes nebulisation of 6% hypertonic saline in conjunction with a session of chest physiotherapy. Pre-adolescent and adolescent participants showed high treatment compliance to hypertonic saline when compared to other sputum clearance solutions.

Conclusion
In conclusion, to some extent, these findings suggest that short-term hypertonic saline inhalation has a beneficial role in improving airway clearance in pre-adolescent and adolescent cystic fibrosis patients. Variables exist and further research is required to determine the optimal length, concentration and volume of solution required.

Introduction
The health topic selected for investigation is the role of hypertonic saline in airway clearance in pre-adolescent and adolescent cystic fibrosis patients.

The Cystic Fibrosis Trust (2009) describes cystic fibrosis (CF) as one of the most common life-limiting inherited diseases in the UK. In 1989 the defective CF gene was proposed (Riordan et al, 1989). This abnormality leads to a malfunctioning or absent CFTR protein, resulting in a reduced regulation of sodium absorption and chloride secretion, which is thought to increase water re-absorption, decrease mucus clearance and possibly lead to a continuous circle of sputum retention, infection and inflammation (Matsui et al, 1998; Ratjen, 2009). The worldwide prevalence of CF sufferers is an estimated 70,000 (Cystic Fibrosis Foundation, 2010) with a recent study by Farrell (2008) showing approximately 8284 CF patients in the UK. The estimated incidence for newborns affected by CF is 1 in 2500 worldwide (Cystic Fibrosis Worldwide, 2009) and 1 in 2750 in the UK (Southern et al, 2007). An approximate £308million per year is necessary for adequate care of CF patients in the UK (PR Newswire Europe Ltd., 2004).

Previously few CF patients lived to adolescence but today many CF suffers live into their mid-30s (Cystic Fibrosis Foundation, 2010). Various treatments are available to CF patients including chest physiotherapy, nutrition support,
psychological counselling and medication. One therapy option becoming more relevant in CF management is the use of hypertonic saline (HS). HS refers to any saline solution with a sodium chloride concentration greater than physiologic (0.9%) (Schretzman, Mortimer, & Jancik, 2006). Coakley and Boucher (2007) suggested that through nebulised inhalation HS breaks the ionic bonds in the mucus solution lowering viscosity, increasing rheology and inducing osmotic flow into the mucus layer. The current physiotherapy HS guidelines state three recommendations:

1. its use in enhancing the effectiveness of other techniques
2. the use of a pre-dose bronchodilator
3. a bronchoconstriction trial before the initial inhalation (Bott et al, 2009)

The positive role of HS for adult CF patients has been well documented (Wark & McDonald, 2010). Its role in the airway clearance of pre-adolescent and adolescent CF patients (<21 years) (Hagen et al, 2007) has yet to be fully investigated. It has been hypothesised that an early diagnosis with effective treatment could reduce lung damage in young CF sufferers (Dankert-Roelse & te Meerman, 1995; Accurso, 1997; Düsing & Hoiby, 2004). Considering the suggested importance of correct treatment during pre-adolescence and adolescence, the role of HS in airway clearance to this age range is of interest. A recent Cochrane review (Wark & McDonald, 2008) stated that future assessment is needed to further define which patient groups will respond better to hypertonic saline or other mucociliary clearance enhancing treatments.

Method

A literature search was conducted using key words and a combination of key words, including hypertonic saline, cystic fibrosis, airway clearance and sputum clearance.

An electronic search was directed through filtered resources of the Cochrane Library, National Guideline Clearinghouse and Scottish Intercollegiate Guidelines Network (SIGN); and unfiltered resources of EBSCOhost, PsycINFO, CINAHL and MEDLINE.

Further hand-searching was performed of the Journal of Cystic Fibrosis, European Respiratory Journal and Pediatric Pulmonology. A search of the European Cystic Fibrosis Conference and the North American Cystic Fibrosis Conference was conducted to identify any unpublished work.

The abstract of studies identified during the initial search were screened, with 31 studies selected for further investigation. Of these the type of study, participants, interventions and primary/secondary outcome measures were examined applying a strict inclusion and exclusion criteria (Table 1).

Table 1: Inclusion and exclusion criteria

<table>
<thead>
<tr>
<th>Inclusion</th>
<th>Exclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Articles</td>
<td>Non-analytic studies, Expert opinions.</td>
</tr>
<tr>
<td>• Systematic reviews, meta-analyses, randomised-control trials (RCT), non-randomised control trials (CT), quasi-experimental trials (QE).</td>
<td>• Studies outside English language.</td>
</tr>
<tr>
<td>• No time frame restrictions, however data published in the preceding five years were scrutinised for relevancy to current practice (Patsopoulos &amp; Ioannidis, 2009).</td>
<td></td>
</tr>
<tr>
<td>Participants</td>
<td>Adults (&gt;21 years).</td>
</tr>
<tr>
<td>• Pre-adolescent and adolescents (&lt;21 years) (Hagan et al, 2007).</td>
<td>• Not CF diagnosed by clinical method (Farrell et al, 2008).</td>
</tr>
<tr>
<td>• Both male and female.</td>
<td></td>
</tr>
<tr>
<td>• Clinically diagnosed CF (Farrell et al, 2008).</td>
<td></td>
</tr>
<tr>
<td>Interventions</td>
<td>Studies that combined HS with another medication.</td>
</tr>
<tr>
<td>• Studies that examined the role of nebulised HS on mucus mobilisation in airway clearance.</td>
<td>• Exercise and HS investigated.</td>
</tr>
<tr>
<td>• Minimum trail length of one dose.</td>
<td></td>
</tr>
<tr>
<td>Outcomes</td>
<td>Changes in luminal chloride and sodium conductance.</td>
</tr>
<tr>
<td>• Lung function (forced expiratory volume at one or 0.5 seconds (FEV1, FEV0.5); forced vital capacity (FVC); forced expiratory flow 25-75% (FEF 25-75%) assessed through spirometry or raised volume rapid thoracoabdominal compression technique.</td>
<td>• Measurement of nasal potential difference.</td>
</tr>
<tr>
<td>• Sputum expectoration collected.</td>
<td>• Modified shuttle test.</td>
</tr>
<tr>
<td>• Treatment compliance recorded.</td>
<td></td>
</tr>
</tbody>
</table>
Following this process, eight papers met the inclusion criteria (Figure 1) (Table 2).

**Figure 1** Identification and selection of Studies for review

<table>
<thead>
<tr>
<th>Titles and abstracts screened by reviewer</th>
<th>Excluded papers</th>
</tr>
</thead>
<tbody>
<tr>
<td>133</td>
<td>103</td>
</tr>
</tbody>
</table>

**Table 2: Table of included studies**

<table>
<thead>
<tr>
<th>Title</th>
<th>Study Type</th>
<th>Subjects</th>
<th>Intervention</th>
<th>Saline</th>
<th>Outcome Measures</th>
<th>Results</th>
<th>Evidence Level</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amin et al (2010)</td>
<td>Randomized Control Trial</td>
<td>Female/Male: 12/7</td>
<td>12 week crossover trial (two four week treatment periods, separated by a four week washout period). Hypertonic saline vs Normal saline. HS: 4mL X 2 daily, NS: 4mL X 2 daily Pre-inhale 2 puffs salbutamol.</td>
<td>HS 7% NS 9%</td>
<td>Lung function: Lung clearance index (LCI), FEV1, FEF25-75, FVC. Treatment compliance: Returned used/unused ampoules. Treatment acceptability: CFQ-R.</td>
<td>Lung function: FEV1 increased 0.7% with HS &amp; 0.3% with NS over baseline. The LCI was significantly lower after 4 weeks of HS vs NS inhalation. No significant change in FEF25-75 or FVC from baseline following either solution. Treatment compliance: Returned ampoules showed a 93.3% HS versus 84.5% NS. Treatment acceptability: Participants found both therapies acceptable; HS = 84/100 &amp; NS = 80/100.</td>
<td>1++</td>
<td>This study focuses more on the use of LCI as an outcome measure rather than HS as a more beneficial Rx. Most other studies use lung function as an outcome measure.</td>
</tr>
<tr>
<td>B. Culhane et al. 2011</td>
<td>Randomized Control Trial</td>
<td>F/M: 6/8</td>
<td>9 week crossover trial (two three week treatment periods, separated by a three week washout period). Hypertonic saline vs rhDNase. HS: 10mL X 1 daily, rhDNase: 2mL X 1 daily Pre-inhale 2 puffs salbutamol.</td>
<td>HS - 5.85%</td>
<td>Lung function: FEV1 Treatment compliance: General acceptance score, 1-6 score very good to unacceptable.</td>
<td>Lung function: FEV1 increased 7.7% with HS &amp; 9.3% with rhDNase over baseline. Clinically relevant increase ≥ 10% in approximately 30% of participants with both rhDNase. Treatment acceptability: Median range rhDNase 2 vs. HS: 5. Strong correlation between inhalation time and acceptence.</td>
<td>1+</td>
<td>First evidence for a comparable increase in FEV1 with rhDNase and HS. Argues that HS is a cheaper and more accessible treatment option than rhDNase to patients living in poverty.</td>
</tr>
<tr>
<td>De Boeck et al. (2000)</td>
<td>Control Trial</td>
<td>F/M: 9/10</td>
<td>Multi-dose cross-over trial on same day. Hypertonic saline vs Normal saline. HS: 5mL X 5mL NS: 5mL X 5mL Pre-inhale 4 puffs salbutamol Pre-inhale bronchodilator; inhaled normal saline. 5 minute break. Increasing doses HS separated by 5 minute breaks between inhalations.</td>
<td>HS - 3%, 45%, 6% NS - 0.9%</td>
<td>Lung function: FEV1, SaO2 Sputum expectoration: number of participants expectorating post-HS. Treatment compliance: VAS scale: -7 to +7, - very unpleasant to very pleasant. Chest auscultation.</td>
<td>Lung function: Overall FEV1 decrease of 7% 2 participants had a clinically significant decrease ≥20%. 35% of participants had an FEV1 increase with 6% HS. SpO2 no significant change following either intervention Sputum expectoration: 8 of the 19 participants expectorated post-HS inhalation 1 participant expectorated post-NS 1 post-3% HS. 1 post-4.5% HS. 11 post-6% HS. Treatment acceptability: Majority of Takers scored HS more unpleasant compared to NS.</td>
<td>2-</td>
<td>Showed sputum induction using HS is successful, safe and acceptable in pre-adolescent and adolescent cystic fibrosis patients who do not expectorate spontaneously. It can be performed from the age of 4 yrs on.</td>
</tr>
<tr>
<td>Title</td>
<td>Study Type</td>
<td>Subjects</td>
<td>Intervention</td>
<td>Saline</td>
<td>Outcome Measures</td>
<td>Results</td>
<td>Evidence Level</td>
<td>Comments</td>
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</tr>
<tr>
<td>Dellon et al. (2008)</td>
<td>Control Trial</td>
<td>Preschool children</td>
<td>Single-dose trial. Hypertonic saline vs. Normal saline. HS 5mL X 1 NS 5mL X 1</td>
<td>HS - 3% and 7% NS- 0.9%</td>
<td>Lung function: FEV0.5, FEV1, FVC, respiratory rate.</td>
<td>Preschool children Lung function: 3% HS: no significant difference for FVC, FEV 0.5 or FEV25-75. 7% HS: Increase in FEV 0.5, FEV25-75 and A clinically significant decrease in FEV1 of 20%. No difference of resp. Rate or SpO2 with either conc. of HS. Infants Lung function: No clinically or statistically significant changes in any parameter occurred with either 3% or 7% HS.</td>
<td>2-</td>
<td>This study shows safety and tolerability of 3% and 7% HS in both groups. Early intervention therapies may preserve lung function in improve prognosis. The 20% drop in FEV1 occurred in only 1 child (History of reactive airway disease) and was asymptomatic and resolved without intervention.</td>
</tr>
<tr>
<td>Ho et al. (2004)</td>
<td>Quasi-experimental study</td>
<td>F/M: 27/16 Sample Size: 45 Age: 1.8-12.9 Mean Age: 7.2</td>
<td>Single dose trial. Hypertonic saline only. HS 4mL X 10mins Pre-inhale 2 puffs salbutamol (2.5 mg per 5 years of age, or 5 mg &gt;5 years of age).</td>
<td>HS - 6%</td>
<td>Sputum expectoration: Number of participants expectorating post-HS</td>
<td>Sputum expectoration: 19 of 43 of participants who were unable to produce pre-HS could expectorate sputum following.</td>
<td>2-</td>
<td>This study shows that sputum induction was enhanced by HS inhalation and acceptable to the majority. The role of HS in airway clearance here relays more to the production of sputum for analysis proposes rather than as a long-term treatment option.</td>
</tr>
<tr>
<td>Riedler et al. (1996)</td>
<td>Randomized Control Trial</td>
<td>F/M: 7/3 Sample Size: 10 Age: 13-20 Mean Age: 16.5</td>
<td>Single-dose cross-over trial. Hypertonic saline vs. Normal saline. HS 10mins inhalation with a nebuliser output of 1.6-2.5mL per minute. NS 10mins inhalation with a nebuliser output of 1.6-2.5mL per minute. Pre-inhaled 5mg salbutamol. Following inhalation patients received physiotherapy – postural drainage and percussion or positive expiratory pressure mask. 7 patients also completed a second block of trial within 1-5 days.</td>
<td>HS - 6% NS - 0.9%</td>
<td>Lung function: FEV1, FEV25-75, FVC. Sputum expectoration: Weight of sputum and numbers of participants expectorating post-HS Treatment acceptability: VAS measuring subjective feeling of cleared chest post-physiotherapy -5 to +5; tight chest to clear chest.</td>
<td>Lung function: No changes in any lung function parameters (FEV1, FVC or FEV25-75) following either intervention in either block. Sputum expectoration: Greater sputum weight cleared after HS vs. IS – 17.2g vs. 11.3g. In second block, sputum weight was HS 16.4g vs. NS 8.5g. Small participant numbers reducing power of the second block results. Treatment acceptability: Overall median VAS score significantly higher HS vs. IS – 2 vs. 0.5. No significant difference in VAS following second block &gt; HS 4 vs. NS 1 H.</td>
<td>1+</td>
<td>This study investigates the role HS plays in increasing sputum expectoration during chest physiotherapy session. Single-dose short-term study. HS increased cough but this unlikely to significantly enhance sputum expectoration.</td>
</tr>
</tbody>
</table>
Methodological Quality

The methodological quality of each RCT and CT was evaluated using the PEDro appraisal tool (The George Institute for Global Health, 1999) and QE studies using the critical thinking tool (Social Exclusion Task Force, 2008). The maximum PEDro score possible is 10 - an RCT or CT scoring 9 or 10 on the PEDro scale is graded as of ‘excellent’ methodological quality; studies with scores from 6 to 8, ‘good’; studies 4 or 5, ‘fair’; and those <4, ‘poor’. With the critical thinking tool, studies scoring 9 or 10 are considered of ‘good’ quality; 6-8, ‘fair’; and <5 ‘poor’. Each paper was assessed for bias using the Jadad scale (Jadad et al, 1996) (Table 3).

Table 3  Bias, methodological quality and level of evidence.

<table>
<thead>
<tr>
<th>Title</th>
<th>Risk of bias</th>
<th>Methodological quality</th>
<th>Evidence level (SIGN)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amin (RCT)</td>
<td>3/5</td>
<td>Low</td>
<td>Excellent</td>
</tr>
<tr>
<td>Ballmann (RCT)</td>
<td>3/5</td>
<td>Low</td>
<td>Good</td>
</tr>
<tr>
<td>Riedler (RCT)</td>
<td>3/5</td>
<td>Low</td>
<td>Good</td>
</tr>
<tr>
<td>Suri (RCT)</td>
<td>3/5</td>
<td>Low</td>
<td>Good</td>
</tr>
<tr>
<td>De Boeck (CT)</td>
<td>1/5</td>
<td>High</td>
<td>Fair</td>
</tr>
<tr>
<td>Dellon (CT)</td>
<td>1/5</td>
<td>High</td>
<td>Fair</td>
</tr>
<tr>
<td>Subbaro (Q-E)</td>
<td>1/5</td>
<td>High</td>
<td>Fair</td>
</tr>
<tr>
<td>Ho (Q-E)</td>
<td>1/5</td>
<td>High</td>
<td>Fair</td>
</tr>
</tbody>
</table>

All studies were subsequently attributed a level of evidence using SIGN guidelines (Table 4).
Annex B: KEY TO EVIDENCE STATEMENTS AND GRADES OF RECOMMENDATIONS

1++ High quality meta-analyses, systematic reviews of RCTs, or RCTs with a very low risk of bias
1+ Well-conducted meta-analyses, systematic reviews, or RCTs with a low risk of bias
1- Meta-analyses, systematic reviews, or RCTs with a high risk of bias
2++ High quality systematic reviews of case control or cohort or studies
   High quality case control or cohort studies with a very low risk of confounding or bias and a high
   probability that the relationship is causal
2+ Well-conducted case control or cohort studies with a low risk of confounding or bias and a moderate
   probability that the relationship is causal
2- Case control or cohort studies with a high risk of confounding or bias and a significant risk that the
   relationship is not causal
3 Non-analytic studies, e.g. case reports, case series
4 Expert opinion

GRADES OF RECOMMENDATIONS

A At least one meta-analysis, systematic review, or RCT rated as 1++, and directly applicable to the target
   population; or a body of evidence consisting principally of studies rated as 1+, directly applicable to
   the target population, and demonstrating overall consistency of results.

B A body of evidence including studies rated as 2++, directly applicable to the target population, and
   demonstrating overall consistency of results; or extrapolated evidence from studies rated as 1++ or 1+.

C A body of evidence including studies rated as 2+, directly applicable to the target population and
   demonstrating overall consistency of results; or extrapolated evidence from studies rated as 2++.

D Evidence level 3 or 4; or extrapolated evidence from studies rated as 2+.

Results

To determine the effect HS has in airway clearance of pre-adolescent and adolescent CF patients, a
    critical evaluation of the eight studies was conducted. The key aspects of each study are
    summarised (Table 2). The findings were appraised and for the purposes of this review will
    be analysed thematically by outcomes measure:
    1. Lung function
    2. Sputum expectoration
    3. Treatment compliance

Lung function

Seven reviewed papers (Table 5) investigated spirometry as an outcome measure. Spirometry is
    a valid and reliable measure of lung function in both pre-adolescent (Gangell et al, 2010) and
    adolescent patients (Perez-Padilla, 2003). A paper’s participant sample must be of sufficient
    size to warrant any results presented. Previous research calculated the minimum sample size
    required to detect the treatment effect of HS on FEV1 as 16 participants (Donaldson et al, 2006).

In reviewing the literature, three studies (Suri et al, 2001; Ballmann & von der Hardt, 2002; Amin et al,
    2010) reported an increase in FEV1 following HS intervention. Ballmann and von der Hardt (2002)
    and Suri et al (2001) aimed to examine HS compared to rhDNase therapy. Ballmann and von der Hardt
    (2002) following a nine-week cross-over trial had substantial FEV1 increases of 9.3% post-rhDNase
    therapy and 7.7% with 10 millilitres (mL) 5.85% HS inhalation. Suri et al (2001) after a 40-week cross-
    over study reported FEV1 increases of 16% for daily rhDNase and 3% from baseline for 5mL 7%HS.
    Participants in Suri et al (2001) demonstrated a much lower FEV1 increase post-HS treatment than seen
    in Ballmann and von der Hardt’s trial (2002). Overall participants in the two studies showed greatest
    improvement in lung function with rhDNase and it should be considered the optimal treatment option.
    Ballmann and von der Hardt (2002) argue, supported by Suri et al (2001), that HS could be more accessible
    to CF patients worldwide, especially in third world countries, as it is significantly cheaper and more
    easily produced when compared to rhDNase. The results gained from HS inhalation should be
    considered as it is potentially a more viable treatment
option for many patients. Amin and associates (2010) compared 4mL 7% HS with 0.9% normal saline (NS) in a 12-week cross-over study and also reported a percentage rise in FEV1. In contrast to previous studies, it revealed a significantly lower FEV1 increase of 0.7% post-HS. All three RCTs are of low bias and good methodological quality (Table 3). Ballmann and von der Hardt’s (2002) sample registered as just below the necessary 16. Suri et al (2010) had sufficient sample sizes and both included a power calculation, minimising possibility of type II error. The results of these samples could be generalised to the wider pre-adolescent and adolescent CF population (Donaldson et al, 2006). Strong consideration must be given to these studies’ findings on treatment length, percentage HS and volume.

Conversely, two studies (Dellon et al, 2000; De Boeck et al, 2008) suggested a FEV1 decrease post-HS intervention. De Boeck and colleagues (2008) aimed to study the safety of HS and compared a 5mL single-dose of NS, 3%, 4.5% and 6% HS. Findings showed an overall 7% FEV1 drop, with two participants showing a clinically significant ≥20% fall. Dellon et al (2000) observed the effect of a single-dose of 5mL 3% and 7% HS on preschool and infant CF patients. The preschool participants had a substantial reduction in FEV1 post-7% HS. Dellon et al (2000) used a sample size, in all groups, which is notably below that necessary to detect effect between HS and FEV1. De Boeck and colleagues (2008) used a satisfactory sample size. Both studies are of high bias and poor methodological quality and caution must be taken when reviewing their results (Table 3).

Interestingly, three studies (Riedler, 1996; Dellon et al, 2000; Subbaro, 2007) that used spirometry as an outcome measure did not demonstrate a FEV1 percentage change. Riedler and colleagues (1996) applied a single-dose cross-over trial, comparing 10 minutes of 6% HS to NS prior to a session of chest physiotherapy. After five days, a second block of seven participants undertook the same trial. No change in FEV1 in either block was reported following intervention. Again, Dellon et al (2000) observed no FEV1 percentage change following 3% and 7% HS with infants and 3% HS with pre-school children. Similarly, Subbaro et al (2007) using a 5mL single-dose of 7% HS, failed to show any considerable FEV1 changes from baseline. All studies (Riedler et al, 1996; Dellon et al, 2000; Subbaro et al, 2007) were single-dose and used insufficient samples. Furthermore, both Dellon et al (2000) and Subbaro and colleagues (2007) are limited by the lack of a control group or randomisation. The study participants in Subbaro et al (2007) are the youngest of all the available literature (mean age 80 weeks) and it is important to examine this paper’s result on the effects and safety of HS on this age range, regardless of study flaws. Four papers (Amin et al, 2010; Dellon et al, 2000; Subbaro et al, 2007; Riedler et al, 1996) recorded FEF25-75 and three studies (Amin et al, 2010; Suri et al, 2001; Subbaro et al, 2007) documented FVC post-HS, with no significant changes found.

By analysing the seven papers (Table 5), conclusions can be made that a multi-dose trial of HS improves lung function. Variables in volume and percentage HS exist with further investigation required to determine the correct therapy parameters.

### Table 5 Studies that examined lung function

<table>
<thead>
<tr>
<th>Study</th>
<th>Volume</th>
<th>Percentage HS</th>
<th>Study Duration</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amin et al (2010) 1++</td>
<td>4mL twice daily</td>
<td>7%</td>
<td>12 week cross-over trial: HS treatment period lasting 4 weeks.</td>
<td>FEV1 increased by 0.7%. No significant change in FEF 25-75 or FVC.</td>
</tr>
<tr>
<td>Ballmann &amp; von der Hardt (2002) 1+</td>
<td>10mL once a day</td>
<td>5.83%</td>
<td>9 week cross-over trial: HS treatment period lasting 3 weeks.</td>
<td>FEV1 increased by 7.7%.</td>
</tr>
<tr>
<td>Suri et al (2001) 1+</td>
<td>5mL twice daily</td>
<td>7%</td>
<td>40 week cross-over trial: HS treatment period lasting 12 weeks.</td>
<td>FEV1 increased by 3%. No significant change in FVC.</td>
</tr>
<tr>
<td>Riedler et al (1996) 1+</td>
<td>One 10 minute inhalation with a nebuliser outputting 1.6-2.5mL/minute</td>
<td>6%</td>
<td>2 blocks of a single-dose trial.</td>
<td>No significant changes in FEV1, FVC or FEF25-75 in either of the two blocks.</td>
</tr>
<tr>
<td>De Boeck et al (2000) 2-</td>
<td>5mL for a 5 minute inhalation period.</td>
<td>3%, 4.5%, 6%</td>
<td>All doses taken once in the same day 5 minute period separating each dose.</td>
<td>Overall FEV1 decrease of 7%.</td>
</tr>
<tr>
<td>Dellon et al (2008) 2-</td>
<td>One 5mL inhalation.</td>
<td>3% &amp; 7%</td>
<td>4 groups (2 preschool children + 2 infants) Each group single inhalation of allocated percentage.</td>
<td>No significant change in FEV1 with 3% in either group and 7% in the infant group. A significant FEV1 decrease of 20% following 7% inhalation in preschool group.</td>
</tr>
<tr>
<td>Subbaro et al (2007) 2-</td>
<td>4mL for a 15 minute inhalation period.</td>
<td>7%</td>
<td>Single-dose trial.</td>
<td>No significant changes in FEV1, FVC or FEF25-75.</td>
</tr>
</tbody>
</table>
### Sputum expectoration

Three studies (Riedler et al, 1996; De Boeck, 2000; Ho, 2004) reported the role of HS in sputum clearance (Table 6). All demonstrated that HS increases sputum expectoration in pre-adolescent and adolescent CF patients.

In Riedler and colleagues (1996) controlled crossover study, all 10 participants from the first block cleared sputum after inhalation, with significantly greater weight cleared by HS in conjunction with physiotherapy compared to NS, 17.2 grams (g) versus 11.3g. The second block reported similar increase in weight, 16.4g cleared with HS versus 8.5g with NS but had insufficient participant numbers, reducing applicability to the wider population. De Boeck et al (2000) reported 18 of 19 participants expectorated post-HS inhalation and 1 from 19 post-NS. Similar to Riedler and associates findings (1996), 6% HS inhalation caused 11 of 19 participants to expectorate. De Boeck and colleagues’ (2000) method is limited as a multi-dose single day trial was used and the possible cumulative effect of four HS doses may have caused 6% HS (the final solution inhaled) to produce optimal sputum clearance. Like previous trials, the effect of a 10 minute single-dose 6% HS inhalation in pre-adolescent CF patients was studied by Ho and associates (2004). Of the 43 participants unable to spontaneously expectorate, 19 were able to clear sputum post-HS intervention.

Both De Boeck et al (2000) and Ho et al (2004) had adequate samples, increasing the ability to generalise their results beyond the study population. Caution is taken when considering the research of either study due to the lack of blinding or randomisation. Additionally, Ho and colleagues (2004) study did not include a control. Comparatively, stronger weight may be given to Riedler et al (1996) due to the study’s high evidence level (Table 3) and clinical relevance of the participation group, hospital in-patients suffering a pulmonary exacerbation.

All three studies showed an increased weight of sputum cleared post-HS. Homogeneity between studies of single dose 6% HS is evident. Future research is required to consider the effect of a multi-dose HS trial over a longer time period.

### Compliance with treatment intervention

Suri et al (2001) and Amin et al (2010) calculated treatment compliance by collecting returned used ampoules (Table 7). Amin and colleagues (2010) demonstrated 93.3% HS versus 84.5% NS returned ampoules. Similarly, Suri et al (2001) had 93% HS versus 84% rhDNase. Both studies were multi-dose and of high evidence level (Table 3). This degree of compliance to HS compared to other solutions reflects the importance CF patients place on its role in counter-acting their disease.

### Table 6  Studies that examined sputum expectoration

<table>
<thead>
<tr>
<th>Study</th>
<th>Volume</th>
<th>Percentage HS</th>
<th>Study Duration</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Riedler et al (1996) 1+</td>
<td>A single 10 minute inhalation with a nebuliser outputting 1.6-2.5mL/minute.</td>
<td>6%</td>
<td>Two blocks of a single-dose trial.</td>
<td>17.2g weight of sputum cleared in the 1st block. 16.4g weight cleared in the 2nd block.</td>
</tr>
<tr>
<td>De Boeck et al (2000) 2-</td>
<td>5mL for a 5 minute inhalation period.</td>
<td>3%, 4.5%, 6%</td>
<td>All doses taken once in the same day 5 minute period separating each dose.</td>
<td>18 of the 19 participants cleared sputum after HS inhalation. 6 expectorated after 3%, 1 after 4.5% and 11 after 6%.</td>
</tr>
<tr>
<td>Ho et al (2004) 2-</td>
<td>4mL for a 10 minute inhalation period.</td>
<td>6%</td>
<td>Single-dose trial.</td>
<td>19 of the 43 patients cleared sputum after HS inhalation.</td>
</tr>
</tbody>
</table>

### Table 7  Studies that examined treatment compliance

<table>
<thead>
<tr>
<th>Study</th>
<th>Volume</th>
<th>Percentage HS</th>
<th>Study Duration</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amin et al (2010) 1++</td>
<td>4mL twice daily</td>
<td>7%</td>
<td>12 week cross-over trial: HS treatment period lasting 4 weeks.</td>
<td>93.3% of returned ampoules were used. Participants showed good adherence to treatment.</td>
</tr>
<tr>
<td>Suri et al (2001) 1+</td>
<td>5mL twice daily</td>
<td>7%</td>
<td>40 week cross-over trial: HS treatment period lasting 12 weeks.</td>
<td>93% of returned ampoules were used. Participants show good treatment compliance.</td>
</tr>
</tbody>
</table>
Discussion

The purpose of this critical review was to investigate the role of HS in airway clearance in pre-adolescent and adolescent CF patients. To suggest a conclusion on HS treatment, it is necessary to interpret its effect on lung function, sputum clearance and patient compliance.

To summarise the thematic findings: lung function - improvement following a multi-dose HS treatment (Suri et al, 2001; Ballmann & von der Hardt, 2002; Amin et al, 2010); sputum expectoration - increase after a single-dose 6% HS inhalation (Riedler et al, 1996; De Boeck, 2000; Ho, 2004); high patient compliance with HS treatment (Amin et al, 2010; Suri et al, 2001).

It is essential to further consider these short conclusions before forming implications for practice. This review suggests that HS improves lung function, with the greatest effect on FEV1. Liou et al (2010) reported that CF-adolescents have the greatest FEV1 decline, with Kerem et al (1992) suggesting FEV1 to be a significant predictor of mortality in CF patients. By increasing the FEV1 of a pre-adolescent or adolescent CF patient, it is argued that quality of life and life expectancy may improve as a consequence (Abbott & Gee, 2003) and thus, any therapy to improve/maintain lung function could have an important treatment role. All significant FEV1 increases were recorded by multi-dose treatment studies, suggesting a single HS inhalation may not improve lung function. Of the three studies that showed a FEV1 increase, both Ballmann and von der Hardt (2002) and Amin et al (2010) had a shorter length study, three and four weeks, in comparison to Suri and colleague’s (2001) 12 week trial. These results propose that any improvement in lung function cannot be sustained over a longer period. This suggestion is undermined by other variables, as both Ballmann and von der Hardt (2002) and Amin et al (2001) despite similar length trials, reported hugely different FEV1 improvements. A possible reason for this discrepancy could be the large 10mL nebulised during the three week trial by Ballmann and von der Hardt (2002), while Amin et al (2010) and Suri et al (2002) used smaller volumes (4mL; 5mL) which may account for the lower effect size observed. This 10mL HS volume produced the greatest increase in FEV1, yet was met with a low patient tolerance which may have implications should it become regular clinical practice. To overcome this trend of poor patient tolerance to large HS treatment inhalation volumes, Elkins and Bye (2006) suggested that using a more efficient nebuliser could reduce inhalation time and improve acceptability.

It is also noted that Amin et al (2010) had a narrow inclusion criterion, with participants having a predicted FEV1≥80%. Trailists in Suri et al (2001) and Ballmann and van der Hardt (2002) had a FEV1≥75%. The possibility is presented that CF patients with mild-severe lung disease (Cooper & Mitchell, 2003) may gain greatest benefit from HS inhalation treatment.

Sputum clearance was found to improve post-HS. Further interpretation is necessary to determine what clinical benefits can be gained from these findings. Homogeneity between all three studies was established for single-dose 6% HS inhalation. Pre-adolescents and adolescents who were initially non-productive, were able to expectorate post-HS (Ho et al, 2004; De Boeck et al, 2000) and more importantly, as suggested by Elkin and Bye (2006), also clear trapped bacteria which may be contributing to infection and inflammation (Tunney, 2008). It is possible through removing the threat of infection and potential lung damage, lung function and life expectancy could improve. Optimal mucus clearance occurred during and after a session of physiotherapy, implying the possibility that HS treatment just before a physiotherapy session is the most advantageous inhalation time for young CF patients.

While the younger patients in these research studies showed a high compliance with HS treatment, this remains a major problem in the management of adult CF patients (Dodd & Webb, 2000). If pre-adolescent and adolescent patients can be encouraged to make HS inhalation part of their daily routine, it could change these poor-compliance attitudes to treatment as they reach adulthood.

Between the seven included studies there is heterogeneity on the optimal length of HS to be inhaled. Those studies (Dellon et al, 2000; Suri et al, 2001; Ballmann & von der Hardt, 2002; De Boeck et al, 2008; Amin et al, 2010) examining the effect of HS on lung function use a HS concentration ranging 3%-7%; while conversely studies (Riedler et al, 1996; De Boeck, 2000; Ho, 2004) investigating the role of HS on mucus clearance had a homogeneity of 6% HS. In a previous study on the role of HS in the airway clearance of adult CF patients, Eng et al (1996) further advised that lower concentration HS
inhaled should only be considered with patients who show poor tolerance to higher strength HS. This was supported by Dellon et al (2000) who examined a lower range 3% HS and also 7% HS. Dellon and colleagues (2000) recommended that a 7% HS be used where possible. Due to the heterogeneity between papers this review is unable to make a definitive HS strength recommendation.

Having synthesised the results of all seven papers, a tentative conclusion can be suggested that a multi-dose treatment of 10mL 5.85% – 7% HS may increase lung function and sputum clearance.

**Clinical Implications**

The results of this appraisal were viewed in the wider context of CF patient management. A search of relevant literature identified a recent Cochrane review (Wark & McDonald, 2008), which concluded similar treatment recommendations. The findings of this critical review may have practice implications for the treatment of pre-adolescent and adolescent CF patients, including implications for physiotherapy practice. Current physiotherapy practice guidelines on HS use (Bott et al, 2009), were formed principally using the Cochrane review (Wark & McDonald, 2008) which focuses primarily on the role of HS in airway clearance of adult CF patients. These guidelines are therefore not directly transferrable to pre-adolescent and adolescent patients. Conversely, this review using strict inclusion criteria details the role of HS therapy in pre-adolescent and adolescent CF patients and, to some extent at least, shows improvements in airway clearance and lung function. With further research it could be determined whether or not this patient group would gain benefit from continued HS inhalation as an adjunct treatment to physiotherapy.

This review is fundamentally restricted by the limitations of the papers themselves. Of the eight studies reviewed, four have an evidence level rating of 2 (Table 3). Using the SIGN grading system (SIGN, 2008) the recommendations can be recognised as a grade D (Table 4). A narrow amount of literature investigating the role of HS in the relevant age group and restricted access to relevant articles has limited this review. Although several high quality papers such as Tarran et al (2007) and Donaldson (2006) produced relevant recommendations, the age of study participants did not meet the inclusion criteria of this review.

It has been suggested that a multi-dose treatment of 10mL 5.85% – 7% HS could increase FEV1 and sputum expectoration in pre-adolescent and adolescent CF patients. Due to study limitations there is a large scope for further research. Patients were less tolerant of 10mL HS volume due to a long inhalation time caused by the use of a jet nebuliser. Future research is needed into the use of a quicker nebuliser. Only one single-dose study (Riedler et al, 1996) observed the combination of chest physiotherapy and HS inhalation, with very positive results. Further investigation is required to establish the effect of HS and chest physiotherapy on lung function and sputum expectoration over a multi-dose clinical trial.

**Conclusion**

This review has found that short-term HS inhalation provides a modest improvement in lung function, increase in sputum clearance and is acceptable to pre-adolescent and adolescent CF patients. HS inhalation has a beneficial role in improving airway clearance in this patient group.

**Reference List**


<table>
<thead>
<tr>
<th>Study – Randomised Control Trial</th>
<th>Sample size – 16 necessary to detect treatment effect (Donaldson et al, 2006)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amin et al (2010)</td>
<td>Y</td>
</tr>
<tr>
<td>Ballmann and von der Hardt (2002)</td>
<td>N</td>
</tr>
<tr>
<td>Riedler et al (1996)</td>
<td>Y</td>
</tr>
<tr>
<td>Suri et al (2001)</td>
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</table>

<table>
<thead>
<tr>
<th>Study – Quasi-experimental</th>
<th>Sample size – 16 necessary</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sultar et al (2007)</td>
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**Appendix 1 Strengths and weaknesses of papers**

<table>
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<th>Strengths and weaknesses of papers</th>
<th>Study – Randomised Control Trial</th>
<th>Sample size – 16 necessary</th>
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<tbody>
<tr>
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<tr>
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<tr>
<td>Concealed allocation (allocation similar between groups)</td>
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<td>Y</td>
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<tr>
<td>Both groups similar at baseline</td>
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<td>Y</td>
</tr>
<tr>
<td>Random allocation of similarities of analytical variables</td>
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<td>Y</td>
</tr>
<tr>
<td>Therapist administering treatment blinded</td>
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</tr>
<tr>
<td>Participant blinding</td>
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</tr>
<tr>
<td>Outcome assessor blinded</td>
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<td>Y</td>
</tr>
<tr>
<td>Key outcome measure obtained from 85% of group</td>
<td>Y</td>
<td>Y</td>
</tr>
<tr>
<td>Analysis by intention to treat</td>
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<td>Y</td>
</tr>
<tr>
<td>Between group statistical comparison reported</td>
<td>Y</td>
<td>Y</td>
</tr>
<tr>
<td>Point measures +/or measures of variability provided</td>
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<td>Y</td>
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<th>Weaknesses of papers</th>
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<th>Sample size – 16 necessary</th>
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<tbody>
<tr>
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<td>No (N)</td>
</tr>
<tr>
<td>Ballmann and von der Hardt (2002)</td>
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<td>N</td>
</tr>
<tr>
<td>Dellon et al (2008)</td>
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<td>N</td>
</tr>
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<td>Riedler et al (1996)</td>
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<td>Y</td>
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<tr>
<td>Suri et al (2001)</td>
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</table>
Appendix 2 - Glossary of Terms

ANCOVA – Analysis of covariance is the initial step in identifying factors that are influencing a given data set. Generally the variables in an ANOVA analysis are categorical, not continuous.

Anderson-Darling test – normality test, designed to detect all departures from normal distribution. It uses a sample’s “P-value” to measure whether the data collected is of “normal” distribution.

Confidence interval – explains the range of possible effect sizes compatible with the data. Confidence intervals aid interpretation of clinical trial data by placing upper and lower bounds on the likely size of any true effect.

CFTR protein – cystic fibrosis transmembrane conductance regulator (CFTR), this protein functions as a channel across the membrane of cells that produce mucus, sweat, saliva, tears, and digestive enzymes.

Cystic fibrosis – common hereditary disease which affects the entire body, causing progressive disability and often early death

FEF25-75 – A clinical test that measures the forced expiratory flow from 25% to 75% of vital capacity.

FEV1 – Forced Expiratory Volume in the first second. The volume of air that can be forced out in one second after taking a deep breath, an important measure of pulmonary function.

FVC – forced vital capacity vital capacity measured when the patient is exhaling with maximal speed and effort.

Homogeneity – the quality of being of uniform throughout in composition or structure.

Hypertonic Saline – saline solution with a sodium chloride concentration greater than 0.9%.

Non-Parametric statistics – is a branch of statistics dealing with variables without making assumptions about the form or the parameters of their distribution.

Normal Saline – a solution of common salt in saline, of a strength of 0.9%.

P-values – calculated to assess whether trial results are likely to have occurred simply through chance. A p-value of <0.05 is necessary for results to be statistically significant.

Paired t-test – assesses whether the means of two groups are statistically different from each other. This analysis is appropriate whenever you want to compare the means of two groups.

Parametric statistics – is a branch of statistics that assumes data come from a type of probability distribution and makes inferences about the parameters of the distribution.

Raised volume rapid thoracoabdominal compression technique – technique for lung function testing in infants.

rhDNase – is a synthetic enzyme that removes neutrophil derived DNA in sputum to reduce viscosity and thus in theory to aid sputum removal.

Rheology – is the flow of matter, principally liquids.

Spirometry – is the most common of the lung function tests. These tests look at how well your lungs work. Spirometry is also used to monitor the severity of some other lung conditions, and their response to treatment.

Washout period – is necessary if treatments might interact in an adverse way. If two drugs are being compared, it is used to prevent both of them presenting at the same time.

Wilcoxon single-rank test – a non-parametric test of the null hypothesis. It requires the knowledge that the distribution being sampled is symmetric (achieved using a normality test). It is based on ranking the observations by their distance above and below the median and comparing the total of the rankings above and below.
Selective Dorsal Rhizotomy – An Overview

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Introduction
This article examines the evidence to date regarding the neurosurgical technique Selective Dorsal Rhizotomy (SDR) as a treatment option in the management of spasticity and its impact for paediatric physiotherapists. SDR is a surgical procedure dividing the posterior nerve rootlets from spinal cord segments L1 and L2. An incision is made along the lower back and a laminectomy of one or more vertebrae is made to expose and test small nerve rootlets consisting of spinal sensory nerves. 3-5 rootlets are normally identified. Rootlets with an abnormal electromyography response are selectively cut, the motor nerve rootlets remain untouched and leg movement is preserved (NICE guidelines 2006).

Spasticity is a positive sign of an upper motor neurone syndrome and the focus of many treatment interventions, it is also a clinical feature in over 75% of cases of children with Cerebral Palsy (CP) and is considered to be the major cause of discomfort affecting gait and function in children with CP (McLaughlin et al 2002). Although SDR is the focus of this paper due to the recent increase in publicity surrounding it, paediatric physiotherapists should be cognisant with the other ways to manage spasticity. Pountney (2007) identifies physical treatment techniques, e.g. neuro-developmental therapy or conductive education; medication intervention, e.g. baclofen and botulinum toxin; and also other surgical/neurosurgical management approaches to spasticity.

Background
Dorsal rhizotomy was first documented by Sherrington (1898); he noted that extensor rigidity was eliminated in some de-cerebrate cats by sectioning posterior rootlets. Fifteen years later Foerster (1913) used this technique in patients with congenital spastic paraplegia to reduce their tone, but did not recommend it for improving function due to the negative side effects noted by sensory loss. Fasano et al (1978) later modified the technique to be more ‘selective’ in its approach by identifying and sectioning aberrant posterior rootlets that reduced sensory loss. By electrically stimulating rootlets they were able to identify the ones showing abnormal tonic contraction in the muscles innervated by the stimulated root as well as in distant muscles. Fasano et al (1980) claimed that by selective interruption of the abnormal circuits, spasticity could be reduced and sensation preserved by sectioning only the aberrant rootlets. Within North America in the 1980’s the procedure was further modified by changing the surgical site from the conus medullaris to the cauda equina, preserving the sacral nerve roots innervating the bowel and bladder (Peacock et al 1991, Giuliani 1991).

Neurophysiological evidence supported the view that spasticity was due to the decreased inhibition from multiple upper motor neuron and interneuron inputs and could increase excitability of alpha motor neurons (Young 1994). Sensory afferents from muscles have a mainly excitatory effect on alpha motor neurons and the developers of SDR (McLaughlin et al 1998) surmised that excitatory sensory input to the anterior horn could be reduced without impairing the sensory function. Electrophysiological measurement criteria were introduced by Fasano and modified by Peacock (Staudt et al 1995) to optimise the ‘selectivity’ of the rhizotomy. Opinion on the technique has alternated between enthusiasm (Park and Owen 1992) to varying levels of scepticism (McLaughlin et al 1998).

Literature Review
A review of the literature pertaining to SDR was conducted between April and July 2011. The search terms used were: SDR, neuro-surgery, cerebral palsy and diplegia. The following databases were accessed: Medline, CINHAL, EBOS, Pub Med and the Cochrane Collaboration data bases. 30 publications were retrieved and of these 10 were deemed suitable.

The majority of the evidence to date originates from North America and Europe which includes three randomised control trials (RCTs) (Steinbock et al
1997, McLaughlin et al 1998, Wright et al 1998) and a meta-analysis (McLaughlin et al 2002). A brief overview of these studies follows to contextualise the impact of SDR for paediatric physiotherapists.

Steinbock et al (1997) compared selective posterior rhizotomy plus physiotherapy with physiotherapy alone in children with spastic diplegic CP. A total of 15 patients were included within this RCT, with one child in each group dropping out after randomisation. Although numbers are small it reflects the relatively few numbers of children undergoing this procedure. Patients were randomly assigned to one of two treatment modalities:

1. SDR followed by 9 months of intensive outpatient physiotherapy;
2. 9 months of intensive outpatient physiotherapy (3 x week for 3 months, 2 x week for 6 months – passive ROM, strengthening exercises to hip abductors and extensors, knee extensors and ankle dorsiflexors, plus practice of normal patterns of movement based on neurodevelopmental theories).

Steinbock et al (1997) found a statistically significant and clinically important difference in improvement in motor function in favour of the SDR group.

In the second RCT McLaughlin et al (1998) looked at the efficacy and safety in an investigator-masked RCT. This was a larger study where 43 children with spastic diplegia were randomly assigned to receive SDR plus physiotherapy or physiotherapy alone. A total of 38 children completed follow up through 24 months with 21 children in the treatment group and 17 in the control group. McLaughlin et al (1998) concluded that SDR is safe and reduces spasticity in children with spastic diplegia. Unlike Steinbock et al (1997) the results of the McLaughlin et al study (1998) reported an equal improvement in the independent mobility at 24 months in the treatment and control group. They went a step further and stated that SDR may not be an ‘efficacious’ treatment for children with mild spastic diplegia.

The final RCT (Wright et al 1998) evaluated the efficacy of SDR for the reduction of spasticity in cerebral palsy. This study included 24 children who were randomly assigned to one of 2 groups:

1. SDR with physiotherapy and occupational therapy as an in-patient for 6 weeks;
2. equivalent physiotherapy and occupational therapy (each child received a 45 minute physiotherapy session and a 45 minute occupational therapy session twice a week, reduced to 2 sessions a week - 2 hours in total - for the remainder of the study period).

One year post SDR, changes with gross motor function were measured - findings concluded that SDR combined with physiotherapy and occupational therapy produced a greater functional motor improvement after 1 year than just therapy alone.

A meta-analysis of the results showed only a small functional improvement in the client (McLaughlin et al 1998, Cole et al 2007). Studies have been plagued by bias, lack of controls, variable surgical techniques and subjective outcome measures (Arens et al 1989, Abot et al 1993, Cohen et al 1991). The absence of solid evidence to support efficacy and the lack of information regarding safety aspects and long term implications has resulted in some controversy over this technique and some doctors have had concerns regarding the invasive and irreversible nature of the procedure (NINDS 2009). There is controversy regarding how selective SDR is and the fact that there might only be a small improvement in function for such an extensive operation (NINDS 2009).

McLaughlin’s meta-analysis (2002) highlighted a small, but statistically significant, advantage of SDR and physiotherapy over physiotherapy alone. The study also identified a direct causal relationship between the percentage of dorsal root tissue cut and the level of gain in function, however there is marked variability between studies on percentage of rootlets cut. McLaughlin (2002) also noted that although the improvements in gross motor function were disappointing the results did not take into account the longer term outcomes of the surgery; positive and negative.

Although there are limitations in the research evidence to date it is possible to conclude that SDR reduces spasticity in children with CP. There is no conclusive evidence that SDR leads to long term functional benefit. This needs to be contextualised with the risks of any surgical intervention and also in the cost benefit analysis for all children in the decision making process prior to further investment. There has been an acknowledgment in many centres of the need for selection criteria, as variations have been noted in patient selection, patient numbers, intra operative techniques, post operative therapeutic intervention and outcome measures for this operation (Cole et al 2007). Further research
would be beneficial to enable an evidence base to be established.

Current Practice
SDR is only practiced in a few centres in the UK, one of which is Oswestry. The Oswestry experience (Cole et al 2007) is the first UK cohort studied consisting of 19 patients. Their findings supported previous studies whereby SDR reduced lower limb spasticity and also demonstrated a small functional benefit on children with CP when adhering to strict selection criteria (Table 1). Cole et al (2007) further purported that SDR is an excellent procedure for overcoming crouch gait and tightness at the hamstrings if strict selection criteria are maintained. The outcome measure to support this was gait analysis.

Frenchay hospital in Bristol has just become the first hospital in the UK to carry out a pioneering neurosurgical procedure offering a new version of SDR which concentrates on the lower part of the spinal cord, increasing its effectiveness and reducing risk (North Bristol NHS Trust May 2011). A programme following the procedure was broadcast on BBC Inside Out West on the 20th June 2011 linking with another child having the operation at St Louis, USA.

<table>
<thead>
<tr>
<th>History</th>
<th>Examination</th>
<th>Investigations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age range 5-10</td>
<td>Diagnosis of diplegia, severe hemiplegia, HSP</td>
<td>No hip dysplasia</td>
</tr>
<tr>
<td>Absence of chronic conditions</td>
<td>Spasticity moderate to severe</td>
<td>No basal ganglia change on MRI</td>
</tr>
<tr>
<td>Cognitive ability IQ 70 or above</td>
<td>Mean lower limb power&gt;3 on MRC scale</td>
<td>Weight not disproportionately greater than height</td>
</tr>
<tr>
<td>Well motivated, emotionally robust</td>
<td>Movement control at least moderate</td>
<td></td>
</tr>
<tr>
<td>No previous multi-level surgery</td>
<td>Balance at least moderate</td>
<td></td>
</tr>
<tr>
<td>Good family/social support</td>
<td>Absence of severe fixed joint deformity</td>
<td></td>
</tr>
<tr>
<td></td>
<td>No involuntary movements or dystonia</td>
<td></td>
</tr>
</tbody>
</table>

(BPD – broncho pulmonary dysplasia; HSP – hereditary spastic paraparesis)

Table 1 Criteria used to select patients for SDR – Oswestry (Cole et al 2007)

There is a growing body of interest in SDR as a treatment option and doctors from hospitals such as Liverpool and Bristol have begun to develop links with doctors in USA who have significantly more experience. A study day held in Great Ormond Street Hospital in 2011 was facilitated by Dr Park and his St Louis Hospital team. Dr Park and his team state that of all the surgical techniques currently performed on patients with CP, SDR has undergone more thorough scientific scrutiny than any other neurosurgical technique (video footage 2011 website www.stlouischildrens.org). Accumulated evidence and experience indicate that SDR is an excellent option for patients with spastic CP (St Louis 2010). Dr Park and his team in USA have performed the SDR procedure on over 2000 children between the ages of 2 and 18 years with 66% under the age of 5 years, 44% of whom were born at less than 29 weeks (St Louis Hospital Brochure p.16 2010). Dr Park and his team recommend early surgery between 2-5years to avoid potential problems with deformities of the legs. 79% of the patients had spastic diplegia with 83% walking with either a walker, crutches or independently – all improved their walking ability post operatively (St Louis SDR Brochure 2010).

Table 2 Criteria used to select patients for SDR - St Louis, USA

Factors to be considered in italics

<table>
<thead>
<tr>
<th>History</th>
<th>Examination</th>
<th>Investigations</th>
</tr>
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<tr>
<td>At least 2 yrs of age</td>
<td>Diagnosis of spastic diplegia, triplegia, quadriplegia or spastic hemiplegia</td>
<td>No severe damage to basal ganglia on MRI</td>
</tr>
<tr>
<td>Motivation and ability to co-operate in therapy</td>
<td>Some form of mobility – e.g. crawling, walking with/without assistance</td>
<td></td>
</tr>
<tr>
<td>Commitment to rehabilitation and follow up</td>
<td>History of premature birth – if born full term must have signs of spastic diplegia</td>
<td></td>
</tr>
<tr>
<td>Wait at least 1 yr post orthopedic surgery</td>
<td>Patient exhibits potential for improvement in functional skills after SDR</td>
<td></td>
</tr>
<tr>
<td>Good muscle strength in legs and trunk</td>
<td>Evidence of adequate motor control</td>
<td></td>
</tr>
</tbody>
</table>
Discussion

There are still considerable variations in the way that SDR is conducted and how patients are selected, most notably the disparity in ages between St Louis (<2 years) and Oswestry (5-10 years). It could be argued that 2 years of age is too young to have such a major intervention when the clinical picture of spasticity and movement is changing so much and the full picture will not have evolved with regard to functional mobility or conservative management. It does mean that it precedes the development of deformities and contractures that may have started and all the postural issues that ensue. The presentation of spasticity in children with CP makes it very challenging to have defined criteria which does explain the variances. There are enough similarities that it would facilitate discussions with parents who are considering the technique.

In any discussion with parents the potential complications of this procedure need to be highlighted as SDR is a long and complicated neurosurgical operation. Some reported and potential risks must be considered - paralysis of the legs and bladder, impotence and sensory loss are the most serious, but wound infections, and leakage of the spinal fluid through the wounds are also potential issues. Sensitivity of the skin on the feet and legs is quite common post SDR but usually resolves within 6 months. Urinary tract infections and pneumonia are also a possibility, as is hip subluxation. Vertebral prominence has been noted and the most frequent occurrence has been weight gain (St Louis 2010, Cole et al 2007). Dr Park’s team note that they have had no long term complications in any of their patients who have undergone SDR with data going back to 1987.

With the advent of more centres looking into SDR both abroad and in the UK, more studies and research should be forthcoming. A large study looking at patients with spastic diplegia from childhood into adulthood is required to see the long term outcome on gait, pain reduction, function, participation and quality of life as well as the rate of acquired musculoskeletal deformities (McLaughlin et al 2002, NINDS 2009). This is an area that is developing and evolving and members of the health profession need to be aware of current practice and the criteria for selection to enable parents to make an informed choice on whether to proceed. This year alone over 40 families from the UK have gone to USA for SDR treatment at their own expense, but more hospitals in the UK are showing an interest at using this technique and parental pressure to have the procedure available on the NHS is strengthening in force as can be seen on their website (www.support4sdr.org).

SDR is currently the only surgical procedure that can provide permanent reduction of spasticity in CP; it is not suitable for all types of CP but studies with children with diplegia are showing small but positive outcomes as long as set criteria are adhered to and all parties involved are committed. Given the lack of conclusive evidence of the impact on gross motor function it could also be argued that the commitment of these parents to the treatment if applied to physiotherapy alone could produce similar results. St Louis is now treating children with hemiplegia after several procedures showed significant improvement. It follows a similar technique but only the nerve roots on the affected side are severed.

Conclusion

SDR is one of a range of treatment options available to control spasticity; it is non-reversible and needs to be approached with all the facts and a clear view of all the potential issues that may arise. The procedure is now being performed in the UK but many parents may still want to go to USA as they feel that with over 2000 operations behind them they are the main centre of excellence for this procedure. This could have financial implications and also potential conflicts of interest regarding protocols and follow up which are given post surgery in the USA. Not all children are suitable for this technique and careful assessment and discussion is required to avoid false hope and non-effective surgery.

Another major issue that has been raised on the iCSP has been the amount of post operative physiotherapy recommended. Most NHS services do not have the capacity to offer the recommended sessions when considering the needs of all children with paediatric physiotherapy requirements. Therefore parents must be committed to carrying out the programme themselves or potentially employing private physiotherapists to provide the level of input they feel is necessary. This may be more difficult in remote and rural areas and will need close working relationships with NHS paediatric physiotherapists. Having spoken to a colleague who has just returned from USA observing Dr Park and his surgical technique she states that there is no hard evidence in favour of therapy 4-5 times per week as opposed to twice weekly. The different structures in healthcare may...
go some way to explain the historical presence of this input and this is likely to be reviewed.

In the longer term, if SDR becomes more mainstream and the expectation is for intensive rehabilitation post surgery then workforce planning will need to take this into consideration. In the short term where there is no conclusive evidence of long term or significant functional improvement then it would be very difficult for the NHS to justify the intensive rehabilitation at the expense of other children. It will be up to local resources and negotiation to decide upon appropriate level of input.

Ongoing research is essential and centres are continually developing and refining their techniques and criteria. Research is required into the long term implications of SDR, beyond 5 and 10 year studies and the success rate depending on the severity of the spasticity – how effective is it with milder cases, is it worth doing for the more severe, non mobile children focusing on the reduction of spasms and the beneficial aspects of positioning and toileting. Many more questions are out there and it will be interesting to monitor the evidence coming forward over the next few years.

**References:**


Foerster O (1913) On the indications and results of the excision of posterior spinal nerve roots in met. Surgery, Gynecology and Obstetrics 16:463-474


North Bristol NHS Trust publicity statement – UK’s first pioneering operation at Frenchay offers hope to children with cerebral palsy. 24th May 2011


St Louis Hospital Center for Cerebral palsy spasticity Aug 2010 - SDR Brochure www.stlouischildrens.org


www.support4sdr.org – parent led website in support of SDR in the UK


Young RR (1994) Spasticity: a review. Neurology 44 (suppl.) 12-20
An integrated pathway for children with coordination difficulties in Wakefield

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ABSTRACT

This article explains the development of an innovative service for children with coordination difficulties in the Wakefield district. The project, which was developed in partnership with the Local Education Authority, trains school staff to identify children with movement difficulties and to implement a tailor-made programme. Although in its infancy, the service has already eliminated waiting lists for physiotherapy. This paper explains why the service was introduced, summarises the research evidence underpinning the approach, and describes the integrated pathway devised in Wakefield for this group of children.

An integrated pathway was developed to address coordination difficulties in school-age children in Wakefield. Its purpose was to:

[1] encourage early identification by training education professionals to recognise coordination difficulties;
[2] provide a standardised plan of action;
[3] provide graduated group intervention in a familiar environment;
[4] reduce inappropriate referrals to therapy services.

Introduction

Coordination difficulties can have many causes including:

[1] increasing survival rates of premature babies;
[2] sensory and/or intellectual impairment;
[3] limited opportunities to experiment with movement in the early years;
[4] changes to the way babies are positioned and carried in recent years.

These have all had an effect on children’s ability to move confidently and with skill.

One specific movement difficulty is known as Developmental Coordination Disorder (DCD). This group of children provided the stimulus for this project. DCD is thought to affect approximately 6% of the population (Gaines et al 2008, Missiuna et al 2006). The disorder is defined in the American Psychiatric Association Diagnostic and Statistical Manual of Mental Disorders (DSM IV 2000) and considered together with the Leeds Consensus Statement 2006. There are at present 4 criteria:

[1] Performance in daily activities that require motor coordination is substantially reduced given the person’s chronological age and measured intelligence - this change may manifest as marked delays in achieving motor milestones (eg, walking, crawling, sitting) and as dropping things, clumsiness, poor performance in sports, or poor handwriting.

[2] The disturbance in criterion 1 substantially interferes with academic achievement or activities of daily living.

[3] The disturbance is not due to a general medical condition (e.g. cerebral palsy, muscular dystrophy), and it does not meet criteria for a pervasive developmental disorder.

[4] If learning difficulties are present the motor difficulties are in excess of those usually associated with it.

We know this is a life-long condition with associated mental health, secondary medical conditions, reduced independence and poor integration into society particularly when there is a co-occurring disorder, and it affects the way in which the whole family functions (Rasmussen & Gillberg 2000, Cairney & Hay 2005 Chen & Cohn 2003, Drew 2005).

Intervention options have been reviewed most recently in 2007 (Hillier S, 2007). The findings are that any intervention is better than none but that those that target specific skills are more successful. This fits in with current practice in the UK where goal orientated therapy is popular.
Historically, children with coordination difficulties had either been referred directly to therapy by their GP or, more commonly, by their school doctor. Recent reductions in routine screening by school doctors has meant that parents and teachers need to be more alert to children’s motor difficulties. Referrals were made to occupational or physiotherapists, who had tended to view these children as a low priority. Valuable time was then lost whilst children waited to be seen, and their school work and sociability continued to be adversely affected. (Dewey et al 2002, Missiuna & Moll 2006).

Health and Education in Wakefield wanted to provide a more unified approach that would become standard in all schools and sustainable across the district. Various researchers have described such joint working as the way forward. (Forsyth et al 2003; Salmon et al 2006). Group work is considered effective in offering an environment where children can persist with skills their peers already possess. (Forsyth et al 2008; Quigg et al 2003; Salmon et al 2006).

Government guidelines encourage agencies to work together in the following documents:

[1] The National Service Framework (NSF) for Children, Young People and Maternity Services (DoH) suggests that “multi-agency pathways and collaboration between services are essential”, therefore services should be “designed around the needs of the child to pick up problems and take preventative action”. By 2014 public services are expected to adhere to these recommendations.

[2] SENDA DfES 2001 recommends regular contact between practitioners in Education and Health to create a “culture of joint working”.

[3] Every Child Matters (Children Act) also encourages cross organisational teamwork to protect children and help them to achieve in the following areas:
   a. Be healthy
   b. Stay safe
   c. Enjoy & Achieve
   d. Make a positive contribution
   e. Achieve economic well-being

Background and development

2005 - educational staff in Wakefield were concerned by the lack of therapy intervention for children with coordination difficulties. They felt this was having an impact on their ability to access the curriculum. The Special Educational Needs Service therefore purchased a motor programme to provide intervention for these children. Some advisory teachers attended training in how to implement it in schools and began a pilot in 8 schools. Teaching assistants ran the groups that had an average of 7 children. Initially the programme was well received, but as it became more widely used, feedback suggested that it did not suit the needs of the Wakefield schools.

2006 - saw the appointment of a Clinical Specialist Physiotherapist to develop and coordinate a service for children with coordination difficulties across the Wakefield district.

The authors, H.A. and T. L. developed an alternative programme that was more flexible, sustainable and accessible throughout Key Stages 1 & 2 (i.e. for children aged 5 to 11 years). Alongside this the pathway was developed to make the referral process more equitable and understandable. This was done in consultation with other professionals and agreed with them.

2007 - the new programme, called “Fit to Learn”, was again introduced to a small number of pilot schools, primarily those who had used the previous programme. It was well received as being easy to assess the child’s ability, easy to use and enjoyable for staff as well as the children. The authors provided the training for school staff. A teacher and at least one teaching assistant from each school attended. Following the training, the assistant then took the lead in running the group in their school, with the teacher providing mentoring and managerial support.

The training aimed to:

[1] built on the observational abilities that teachers already possess;
[2] increased understanding of coordination disorders;
[3] introduced the pathway and the programme.

Further development training was held for those running groups to improve provision and skill in delivery.

2008 - a high level of interest in the training led to 90% of 125 infant and junior schools completing training by the end of 2010.
A secondary programme – “Fit to Learn Extra”, was piloted in 3 schools to cater for the children who had participated in their junior school but would benefit from the further input and support from a teaching assistant. The feedback was mixed and it seemed that the only feasible way to run the group was as an after school club once a week. This ensured that staff could access the space and equipment they required.

2009-11 - interest started to be received from other education authorities to purchase the training and the programme.

The Pathway

The aim of this pathway was to:
[1] improve the understanding of movement difficulties in schools (Stafford 2000, Missiuna & Moll);
[2] identify children earlier;
[3] provide an intervention within the familiar school environment (Pless & Carlsson 2000).

Only children who did not improve were to be referred for specific diagnosis and therapy. All individuals and services involved with the process

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**Figure 1**

The Pathway

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MULTI-AGENCY PATHWAY FOR CO-ORDINATION DISORDERS

**INCLUSIVE PRACTICE / WAVE 1**

e.g. Beam / Write Dance / Wake Up Shake Up

Identifiable motor need

**SCHOOL ACTION / WAVE 2**

Motor skills programme e.g. Fit to Learn for small groups

Pre-test; intervention for 1 term; post-test
Review evidence, including results of hearing & vision tests

Concerns remain

**SCHOOL ACTION PLUS / WAVE 3**

Seek advice from Cognition and Learning Advisory Teacher

Further programmes
Complete ABC Checklist / Early Years Movement Checklist
Parent Leaflet
Teacher Leaflet
Advice for IEP
Request appointment with Community Paediatrician via School Nurse
Referral to therapy
Therapy Assessment
Problems identified
Discussion between paediatricians and therapists about diagnosis

Concerns remain

**INTERVENTION**

Information leaflets
Groups
Individual therapy
Parent groups
Liaison with other agencies
had agreed to the stages in the pathway. The school is central to the process, and takes the main responsibility for identifying the child with movement difficulties in discussion with parents. Strategies are put in place to improve the child’s skills either through the use of the ‘Waves of Intervention’ (DfES 2005) in whole class (Inclusive Practice / Wave 1 – Figure 1) or through small group intervention (School Action / Wave 2 – Figure 1), in a non-threatening and fun environment.

‘Fit to Learn’ is a Wave 2 daily group activity of 15-30 minutes. The recommended group size is 6-8 children of similar ages. The programme initially continued for 2 half terms. This was so that children who might display coordination difficulties as a result of the onset of a neuro-muscular condition could be identified and referred to the Health Service without delay. The testing that was scheduled at the beginning and end of each half term was found to be too time consuming so this was revised in response to the feedback and will in future only be carried out once each term.

If improvement is seen between the assessments, then inclusion in the programme continues. The trained school support assistant carries out both the assessments. They take about 10 minutes per child and cover the 10 areas of the programme. To make it easier for the assessors 2 areas have been combined so assessments cover the following 8 categories:

[1] **Balance and Postural Control** *(2 sections combined)*  
Stand on preferred leg for 10 seconds.

[2] **Fine Motor**  
Using only the preferred hand, place 10 pegs, one at a time, into a pegboard.

[3] **Bilateral Skills**  
Cut between two lines, 2cm apart across an A4 piece of paper.

[4] **Body Awareness**  
Jump backwards five times. Feet do not have to land together.

[5] **Ball Skills**  
Bounce a large ball against the wall and catch after bouncing.

[6] **Listening, Planning and Sequencing** *(2 sections combined)*  
Carrying a beanbag, walk heel to toe along a one metre line, put the beanbag in the hoop and turn the bucket upside down.

Copy a circle, triangle and square.

[8] **Proprioception and Sensory Perception**  
With the child’s eyes closed, place one arm in a posture. Child copies position with their other arm.

The child is assessed before starting the programme and progress is monitored at the end of the term by the teaching assistants using this 4 point score:

- 0 – can’t do it
- 1 – can partially do it
- 2 – can complete
- 3 – completes and has improved from last time

If the child does not improve, the school will ask an advisory teacher to become involved (School Action Plus / Wave 3 – Figure 1). The advisory teacher will provide individual advice to the school, together with information leaflets for the parents and teacher. The leaflets introduce the concept of Developmental Coordination Disorder (DCD). A standardised and validated checklist (Movement Assessment Battery for Children 2nd edition, Pearson Assessment) is also completed. If the checklist reveals a problem, the school can refer to the community paediatrician. The school nurse checks that eyesight and hearing test results are satisfactory and passes on the referral.

The paediatrician will examine the child and exclude conditions such as muscular dystrophy, cerebral palsy or global delay. The therapy service will then assess the child using the Movement ABC2, and decide on the best course of action taking into account the perceived needs of the child, parents and school staff to set functional goals. Some children will receive a diagnosis of Developmental Coordination Disorder (DCD).

**Results and implication for future practice**

When the service began in 2007 there was a 2 year waiting list for physiotherapy. There is now no physiotherapy waiting list for this group of children. Referrals for occupational therapy have also fallen.

Collaboration between Health and Education has meant that there is adequate support and expertise to ensure sustainability of the ‘Fit to Learn’ programme and timely onward referrals. Use of the Pathway has reduced the incidence of inappropriate referrals.

The average assessment time that is taken up with a new referral is 3 hours. This includes the Movement ABC2 second edition (Pearson Assessment), a school...
or home visit, identification of functional goals and provision of a programme and report for all interested parties. The children, parents and teachers are asked what the functional difficulties are for the child and in which areas they would welcome improvement. These then become the priority for therapy.

Some children will be discharged at this point with individual programmes, whilst others are offered further support from a range of options such as small therapy groups, after school clubs, holiday activities e.g. cycling, drumming, parent groups, advice leaflets, etc. Children who are discharged can continue to attend a low level sports club set up by the physiotherapist but run by Leisure Services.

Coordination difficulties are commonplace in the classroom. This project seeks to increase the understanding of school staff so that they are equipped to address the difficulties as they arise.

The authors believe that this is the first service of its type in the UK, in which different agencies work together using a standard pathway to identify coordination difficulties.

As the service is still in its infancy, long-term effects are unknown. The early results are encouraging and consistent with findings of earlier research. The principles of the Pathway and the Programme are easily transferable to other situations and enquiries have been received from as far away as Australia.

It is believed that early intervention (Missiuna et al 2003) can reduce the impact of coordination difficulties on day-to-day life. It can increase the child’s ability to understand and cope when intervention is received in a familiar and supportive environment without the need to attend their local hospital or health centre (Sugden & Chambers 1998).

One of the authors has begun work on a similar project for use in early years settings. The results of a small pilot project appear encouraging.

A key factor in continued success will be schools’ ability to maintain momentum. Ongoing training should facilitate this, but it will need regular review. At the time of writing proposed cuts in public expenditure have yet to bite. It will be interesting to see if collaborative projects such as this can survive a more austere spending environment.

Acknowledgements
The authors would like to thank the Health and Education services who have funded this project, the Leisure Services and the Road Safety Team who have worked with us to deliver the service.

References
American Psychiatric Association Diagnostic and Statistical Manual of Mental Disorders (DSM IV 2000)
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ABSTRACT

In the UK, the routine examination of the newborn is usually performed by doctors who rotate through the post-natal wards. Part of this examination is to detect a dislocating or unstable hip: currently called developmental dysplasia of the hip (DDH). Early detection and accurate treatment are helpful for a normal outcome. If undetected until walking age, a dislocated hip can require corrective surgery and cause long-term disability. This article describes how a physiotherapist-led neonatal hip screening service was developed using examiners in conjunction with ultrasonography, ensuring early diagnosis and effective treatment.

Introduction

Developmental dysplasia of the hip (DDH) comprises a range of abnormalities of the neonatal hip, ranging from a minor self correcting abnormality which has no long term effect, to full hip dislocation which can lead to numerous surgical procedures as a child and total hip replacement in early adulthood due to osteoarthritis (Furnes et al, 2000). There is much debate about screening, especially how to detect cases and when to treat.

In 1937 Ortolani, an Italian orthopaedic surgeon, published a description of his test to detect hip instability (Ortolani 1937). During the examination the thighs are held together at 90 degrees of flexion then gently abducted. A dislocating hip will relocate with an entry jolt or clunk during abduction.

In 1961, Barlow described a test that recognised that some unstable hips subluxed rather than dislocated (Barlow, 1962). His test was performed on one hip at a time with the examiner cradling the pelvis in order to stabilise the sacrum. With the hips flexed to 90 degrees, gentle fingertip pressure is exerted along the length of the femur. The hip can be felt to slide or sublux posteriorly.

There are known risk factors such as a positive family history and extended breech position (Bache et al, 2002). Babies born to mothers who had DDH have a significantly increased chance of having the same condition. DDH is more common in first-born girls who are large and over-due and those babies who have lack of liquor (oligohydramnios) during the pregnancy. It occurs in females 4-8 times more frequently than males.

In 75% of cases of DDH there are no known risk factors. The main cause is thought to be intraterine restriction (Hinderaker et al, 1994). Most babies lie in the vertex position with the left leg against the mother’s spine, which is less yielding than the abdominal wall. The left leg is pushed into adduction, putting pressure on the posterior and upper aspects of the margin of the acetabulum, preventing its normal development and resulting in dysplasia. The adductors become tight in utero and postnatally the baby maintains the same ‘position of comfort’ (Chapple et al, 1941) leading to continued damage to the acetabulum (cartilaginous labrum) so that the hip eventually dislocates.

In the 1940’s, von Rosen devised treatment by use of an abduction device (von Rosen, 1956). He designed a rigid aluminium splint which was very effective. A soft fabric splint called the Pavlik harness is now more commonly used; it is thought to be more acceptable to parents and allows the baby to move almost normally within a safe range, preventing only hip adduction and extension (Mostert, 2000). This may help to minimise the incidence of avascular necrosis the most serious side effect of treatment.

The incidence of clinical instability was about 2.5 per 1,000 births in the 1950s (Jones, 1994) but DDH is now diagnosed in 5-10 per 1,000 births. This increase may be due to a number of factors, including the ‘Back to Sleep’ campaign which advises parents to sleep their baby supine to prevent cot death (supine babies can lie with legs adducted) (Gilbert et al, 2005), and the widespread use of ultrasound since the 1990s (Graf, 1980). Ultrasound may detect less severe cases not detectable clinically, though its critics claim that ultrasound has a high false positive rate

Clinical screening programmes are in place in most developed countries. An experienced person
examining every baby is the ideal but practice varies. Physiotherapists have been reported as performing this role in the UK (Fiddian et al, 1994). In the UK doctors generally perform this primary examination and secondary examinations by ‘experts’ is reserved for babies with risk factors or those babies in whom the primary examiner feels there may be a problem. Alternatively ultrasound examination can be used as the secondary test. The main problem with this approach is that over half the babies that have unstable hips have no risk factors, and therefore if missed at primary testing would not be referred for the secondary screening examination. In Germany and Austria whole population ultrasound screening is in place and treatment rates are as high as 22 per 1,000.

Method

There is a single maternity unit in Edinburgh which has a birth rate of between 6,000 and 7,000 per year. A Medical Assistant who specialised in examination of the neonatal hip examined all babies until 1998. A permanent replacement was not made until 2001 when a Physiotherapist was appointed.

The post developed into two defined roles:
1. to provide expert clinical examination of the neonatal hip;
2. to lead, co-ordinate and develop the neonatal hip screening service (Jones, 1994) ensuring early detection and accurate treatment of DDH.

Until November 2008, the physiotherapist worked single-handed until a second part-time physiotherapist was appointed. This part-time physiotherapist occasionally seeks advice on difficult cases from the lead physiotherapist.

Clinical examination of the neonatal hip requires many hours of practice (Jones, 1994). Due to the low incidence of DDH, a person needs to examine at least 1000 babies in order to become adept at this technique (Myers et al, 2009). Sensitivity of doctors who perform the newborn examination is as low as 40% (Godward et al, 1998).

In the first year of being in post, the physiotherapist examined as many babies each day as possible on the postnatal ward, gaining experience through constant review of practice and outcome (The Chartered Society of Physiotherapy, 2001). A database of all babies examined including risk factors, clinical findings, referral for hip ultrasound and those recalled for a further clinical examination was initiated and maintained. The new-born record of any baby presenting to Orthopaedic clinic with a late (>12 weeks) dislocation, was reviewed (Jones, 1994). Experience was gained by constant review and the physiotherapist observed hip ultrasound performed on babies whom she had referred, comparing clinical findings with hip morphology.

In the first three years of the programme, the physiotherapist examined 3798 newborns but failed to recognise abnormality in 6 babies. These children presented to orthopaedic clinic with a late dislocation at walking age. The number of cases missed by the physiotherapist declined with increasing experience over subsequent years.

An audit of babies treated in harness (2002-2004 n=18,000 births) showed that doctors failed to recognise half the cases that required treatment (n=86). The other half were diagnosed by the physiotherapist who re-examined the baby after the newborn examination had been performed by the doctor who had stated that the hips were normal (false negative). All cases of instability diagnosed by the physiotherapist were confirmed with hip ultrasound prior to commencement of treatment.

The audit also confirmed the significance of restricted movement in-utero and the associated features of moulding, especially calcaneovalgus foot deformity (Lloyd Roberts et al, 1965) as indicators of DDH. The percentage of babies presenting with risk factors is shown in Figure 1.

![Figure 1 Risk factors in 86 babies treated for DDH 2002-4](image-url)
Detection
A major role of the physiotherapist is to teach doctors, midwives and neonatal nurse practitioners about DDH and how to perform clinical examination of the neonatal hip in order to improve the detection of unstable hips at primary examination (Goss, 2002). There seems to be a trend of decreasing experience in doctors at the start of their neonatal attachment; of 10 doctors surveyed in 2003, 6 had examined one or two hips, 2 several hips and 2 none. Only one had experience of an unstable hip. Recently, five doctors about to start performing the newborn examination had never performed a neonatal hip examination.

All professionals who perform the newborn examination in Edinburgh are expected now to attend a theory and practical session. They are taught the known risk factors and the clinical indicators of restricted movement which puts a baby ‘at risk’ of DDH, particularly reduced hip abduction, a deep inguinal fold, calcaneovalgus foot deformity and asymmetric lying position. The doctors often have no knowledge of these indicators, but once shown will recognise them in future and refer the baby for a second ‘expert’ hip examination.

The referral criteria for a second ‘expert’ hip examination have been developed through experience and now include:
1. Reduced hip abduction
2. Positive family history of DDH
3. Extended breech presentation
4. Breech position after 35 weeks
5. Oligohydramnios
6. ‘Moulded baby’ - bat ear, torticollis, spinal asymmetry associated with reduced hip abduction or calcaneovalgus foot deformity
7. First-born, big, over-due girl with doubtful examination
8. Uncertainty or poor examination

A teaching model called a ‘Baby Hippy’ is used but simulators cannot mimic in vivo examinations complicated by a wide variety of abnormal ‘feels’ and unsettled babies. The correct method of examination of the hip is demonstrated and practised on the simulator. Until recently, the doctor only observed the physiotherapist performing the examination on a real baby but the teaching package has been altered so that all personnel must perform a neonatal hip examination observed by the physiotherapist.

As many babies as possible are examined on the post-natal ward by the physiotherapists. Although priority is given to babies with the risk factors listed above, the greatest number of babies examined, are those without ‘known’ risk factors since 75% of babies with DDH have none. Midwives and nursery nurses are educated in signs of DDH and also refer to the physiotherapist.

The true incidence of hip dysplasia is low (6-9/1,000) but doctors find that about 25% of babies have hips that are equivocal or abnormal. The main role of the physiotherapist on the postnatal ward is to screen out a large number of false-positive diagnoses by the doctor (Macnicol, 1990), referring only a small proportion of these babies for ultrasound. It is important to recognise that the physiotherapist acts with high specificity (i.e. excluding most normal babies from ultrasound examination) as well as high sensitivity (ensuring that all of the relatively few babies with abnormal hips are detected). Referral for ultrasound from the post-natal ward is restricted to the physiotherapist and only 2% of the total births undergo hip ultrasound. There is a lower threshold for any baby whose older sibling had DDH.

Due to early discharge and the physiotherapist not being present during certain periods, an outpatient service was set up in 2001. This service has developed and doctors are encouraged to refer. These babies are examined at 3-4 weeks of age so that any case of missed diagnosis (false negative) by the doctors can be picked up by the physiotherapist and referred urgently for hip ultrasound so that treatment can commence by 6 weeks of age.

Neonatal Hip Examination Technique
The resting position and posture during movement should be observed as the baby lies undisturbed in the cot (Macnicol, 1992). A normal newborn baby should lie supine with flexed and abducted hips indicating that the hips are in joint. Slight head preference to the right is normal. A skew or moulded baby may lie with adducted or crossed legs or to one side with the adducted hip(s) at risk. The leg may be flexed up onto the body or extended and adducted.

The nappy should then be removed. A deeper groin crease or an extended posterior gluteal crease indicates that the hip has been adducted in-utero. Features of moulding, especially calcaneovalgus foot deformity, should be looked for. The baby should be settled and not restless. The hips should...
then be tested one at a time. The pelvis is stabilised and cradled gently in the examiner’s hand. The hip is flexed to 90 degrees with both knees kept flexed by the examiner. Finger-tip pressure should be exerted along the length of the femur in a posterior direction to feel for posterior instability of the femoral head, and then the hip is gently abducted to detect any anterior movement as the femoral head relocates. Some examiners use too much force which does not allow the examiner to appreciate small degrees of movement of the femoral head. If the femoral head is subluxing (small degree of posterior movement) it may or may not relocate with a jolt (Hadlow, 1988) and this can be difficult to appreciate. If the femoral head is dislocating (greater degree of movement) the femoral head can be felt to relocate within the first few degrees of abduction as the femoral head moves over the cartilaginous labrum. Examiners who force the hip into abduction can fail to recognise a relocation clunk or jolt which occurs earlier in the range.

A ‘moulded baby’ often has a right calcaneovalgus foot deformity associated with an unstable hip on the left. They are wind swept with the affected hip extended and adducted with a deep inguinal fold. Hip flexion in abduction is reduced but the hip feels stable posteriorly. Movement of the femoral head is not recognised clinically because the hip is unstable superiorly. Neither the Barlow test nor the Ortolani manoeuvre detects this. It is important therefore to push the hip in a superior direction when the hip can be felt to slide or piston (Hadlow, 1988).

If a hip is diagnosed as being unstable the parents are counselled by the physiotherapist. A leaflet is given which describes the condition, shows a picture of the hip ultrasound being performed and a baby in harness. It informs the parents of the treatment plan and long term follow-up. The ultrasound is performed in 2-3 weeks. This allows time for any soft tissue laxity to resolve and treatment if required, to be commenced within the correct time period.

Babies with unstable hips often have an asymmetric lie and roll to one side with reduced flexion/abduction on the contra-lateral leg. In order to stretch the adductors, parents are shown how to place the baby prone in the frog position, when awake for a total of 1 hour per day (Macnicol, 1992). A muslin roll should be placed under the opposite hip (the side to which the baby rolls) when the baby is asleep or supine. If the need for harness treatment is confirmed at ultrasound, the positioning will have stretched the adductors on the affected side ensuring that the hip is not forced into joint during application of the harness as a result of tight adductors.

Babies examined on the post-natal ward by the physiotherapist are divided into 4 categories:
1. Babies who are clinically normal are discharged without follow-up.
2. Babies who have an unequivocally unstable hip are referred for hip ultrasound within 3 weeks - if the hip is still unstable a harness is fitted.
3. Babies who have a lax hip at birth are given positioning advice and are referred for hip ultrasound within 4-6 weeks.
4. Babies with equivocal findings cannot be re-examined on the postnatal ward due to the trend for early discharge, often within 6 hours of delivery - these babies return as outpatients within 4 weeks for reassessment in the physiotherapy department by the physiotherapist. If normal they are discharged, if there is continuing concern they are referred for ultrasound examination.

Neonatal Hip Ultrasound
Neonatal hip ultrasound was introduced in the 1980’s. The Graf Classification (Figure 2) is widely used throughout Europe (Graf, 1980). This was initially based on static images and angles, the most important being the angle the bony roof of the acetabulum makes with the baseline or lateral wall of the pelvis (alpha angle).

Another method of categorising neonatal hip ultrasound is to note how much of the femoral head is contained within the acetabulum (percentage femoral head cover) (Morin et al, 1985). More than 58% is normal, 33% to 58% equivocal and less than 33% abnormal.
In the late 1980’s dynamic or real-time ultrasound was introduced. This allows observation of movement of the femoral head within the acetabulum. Rotation of the head concentrically within the acetabulum is normal but sliding posteriorly or superiorly indicates instability. This may happen spontaneously as the baby wriggles or during stress testing similar to the Barlow test and should mirror clinical findings. The role of ultrasound in hip screening is controversial, but the consensus view is that ultrasound is useful for assessment of those babies thought to have clinical abnormality (Gray et al, 2005).

Treatment Decisions
Treatment decisions are based on several factors:
1. clinical instability;
2. ultrasound appearance (static and dynamic images);
3. age of the baby;
4. how the baby lies.

Since the clinical examination detects by feeling exactly the same phenomena that the ultrasound examination shows on images, the radiologist and the physiotherapist should agree which babies require treatment.

Some unstable hips at birth stabilise in the first few days or weeks of life so that delay in treatment decision to 3-6 weeks allows only cases with persistent abnormality to be treated. If parents comply with advice on prone lying during this period the need for formal treatment in Pavlik harness may be reduced.

Ideally application of the Pavlik harness should be before 6 weeks. A review of babies born elsewhere but treated in Edinburgh later than 8 weeks of age showed that only 66% had a normal pelvis x-ray at 12 months, compared to 97% for babies treated before 8 weeks.

Treatment
The Pavlik harness is often used although several alternative abduction devices (the von Rosen splint or the Craig splint) are used by other centres (Wilkinson et al, 2002). The Pavlik harness consists of a chest band with 4 straps; 2 on each leg. The leg straps attach to stirrups into which the feet are held by Velcro straps. The anterior straps flex the hip and knee and the posterior abduct the hips (Figure 3).

Correct application of the harness is essential for a good outcome (Song et al, 2000). The hip should be maintained in 90-120 degrees of flexion and 50-80 degrees of unforced abduction. Inexperience can lead to ineffective application of the abduction device with consequent failure of treatment (Wilkinson et al, 2002). It is important that the person fitting the Pavlik harness is experienced as the degree of instability, tightness of the adductors and the degree of instability of the hip varies between babies (Macnicol, 1992).

The person fitting the harness should be able to detect whether the hip is in or out of joint so that the hip is not held out of joint in the harness, as this can cause rapid destruction of the acetabulum. It is important to ensure that the hip stays in joint as the baby wriggles in the harness. If the adductors are tight, application in 120 degrees of hip flexion, combined with unforced abduction should be maintained for a week to stretch the adductors. Prone lying for a total of 1 hour per day when the baby is awake stretches the adductors. This allows the hip to reduce gently, with unforced abduction so as not to reduce the blood supply to the femoral head which can cause avascular necrosis of the femoral head (AVN). This is reported in up to 5% of babies treated in harnesses and 30% of those treated in plaster cast (Bearcroft et al, 1996). In Edinburgh there has been one case of AVN in 223 babies treated in harnesses. This occurred in a baby who had a severe congenital scoliosis so adjustment of the harness was complicated by the deformity.

The parents are shown how to care for the baby in the harness and given an advice leaflet. The harness is changed fortnightly in the Physiotherapy Department when the parents can bathe the baby before the new harness is applied.
Treatment progress is monitored by monthly ultrasound scans performed in the hip clinic. Failure to improve by at least one Graf grade is cause for concern: either the harness is poorly adjusted or there is incomplete compliance. Lack of compliance with treatment by Pavlik harness is rarely a problem but parental support and reassurance is vital (Gardner et al, 2005). Parental refusal to continue with treatment has occurred once in 10 years whilst over 360 treatments have been completed.

The harness is usually worn for 12 weeks allowing the acetabular margin to normalise in shape and ossify but in some cases early removal is allowed when the acetabulum is absolutely normal on ultrasound examination. Follow-up at 1 year of age consists of pelvic x-ray to assess acetabular development and clinical review to assess motor development.

Results
Several trends have been noted over the years:

1. With increasing experience the number of cases missed by the physiotherapist decreased from 3 cases in 2002 (2.2/1,000 babies examined) to a single case in the last 6 years (0.14/1,000) - nevertheless the number of late presentations remained greater than this because of cases not examined by the physiotherapist due to absence of risk factors and failure of primary screening to raise concerns (Figure 3).

2. The number of babies treated with Pavlik harnesses per year has increased from 15 in 2002 (2.6/1,000 births) to 66 in 2010 (9.4/1,000) (Fig.4).

This is due to the recognition that less severe degrees of dysplasia benefit from treatment and is reflected in fewer follow-up radiographic examinations at 12 months of age showing abnormalities.

3. The number of abnormal hips not recognised by doctors remains constant at approximately 50% of treated cases (Fig.4).

Discussion
The cost of neonatal screening programmes has long been debated (Dezateux et al, 2003) with the consensus that universal hip ultrasound screening is not cost effective despite evidence from a single centre suggesting that it is (Clegg et al, 1999). Most screening programmes in the UK have both clinical and ultrasound components and good communication between the clinical examiner and the radiologist is important so that each discipline can learn from the other (Clarke, 2004).

There continues to be controversy amongst professionals who are involved in neonatal hip screening in both detection methods and when to treat.
There are several outstanding problems:

1. Most centres continue with doctors performing the newborn examination who then refer babies for secondary hip ultrasound screening only if they have known risk factors (positive family history or breech presentation). This leaves 75% of babies who have no known risk factors but who may be at risk because they have been restricted in utero. With published sensitivities of 50% or less for inexperienced examiners many cases of hip dysplasia will present late- typically reported as about 0.7-1.0 per thousand births.

2. Teaching is often poor and performed by clinicians who are not themselves experienced in examination of the newborn hip. Doctors may have insufficient support on the postnatal ward.

3. Early discharge means that the baby is examined when it is still unsettled and physiologically flexed up; this can be challenging even to an experienced examiner.

4. Ultrasound of the neonatal hip is operator dependent, and some examiners may be less experienced leading to significant interobserver variation.

The ‘Baby Hip Screening Report’ (STEPS, 2009) showed that there was inconsistency across Acute Trusts in England regarding their policies on hip screening. Very few centres had a designated person in charge, with a lack of patient information, data collection and audit, and variations on treatment outcome.

In 2010, on behalf of the National Screening Committee (UKNSC), the NHS Newborn & Physical Examination Programme (NIPE) undertook an exercise to ‘systemise and standardise the newborn examination’. This recommended that “all babies should have a clinical examination of their hips and an ultrasound, if the clinical examination suggests it” (NIPE, 2010). The evidence is that doctors sometimes fail to recognise instability in 60% of cases so do not refer for ultrasound.

CONCLUSION
The screening system described here relies on emphasis of restricted intrauterine movement of the foetus being the cause of DDH and the expertise of the physiotherapist to minimising false-negative and false-positive results.

The role of the physiotherapist comprises four elements of activity:

1. clinical expertise in diagnosis and treatment.
2. teaching,
3. evaluation and
4. audit of the service and service development.

It has reduced the late presentation rate to only 10% of that before the physiotherapy service was started with only 2 late presentations during the last 2 years, neither of which had a risk factor or were examined by the physiotherapists.

The physiotherapy-led hip screening programme has been successful in reducing the late presentation rate of DDH to 0.14/1,000 which compares favourably with published figures of 0.19-0.78/1,000 (Godward et al, 1998; Holen et al, 2002; Ihme et al, 2008; Gunther et al, 1998; Chan et al, 1999).

References


Chartered Society of Physiotherapy (2001). Specialisms and specialists: Guidance for developing the clinical specialist role. PA 23


Ortalani M (1937). Un segno poco noto e sua importanza per la diagnosi precoce di prelussazione congenita dell’anca. Pediatria,45,129.


UK National Screening Committee. (2010) Screening Programs: Newborn and infant physical examination.


A Case Study Using the Transdisciplinary Approach to Develop an Individual Method for Measuring Progress for Children with Profound and Multiple Learning Difficulties

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ABSTRACT

Objective
To demonstrate how an in-house tool was developed to measure pupils who function at developmental levels between 0-9 months, and to support closer team working between physiotherapists and educational staff.

Background
Increased educational demands for reporting pupil progress highlighted the need for a more refined tool specific to this pupil population. Changes in health structures have reduced direct therapy time available within the school setting so that teaching staff often require advice and support for progressing or changing gross motor skill interventions when the physiotherapy team are not available.

Method
The in-house ImPACTS - Individualised Profile Assessment Curriculum Target Setting - was designed to cover 5 key skill areas: Communication, Cognitive Skills, Environmental Control Technology, Social and Emotional Well-being and Physical Skills (Gross and Fine Motor).

Participant
The pupil used to illustrate the use of ImPACTS is John who was born in 2002 with PMLD.

Results
1. The use of ImPACTS improved the quality of initial assessment and led to specifically targeted areas of development being achieved.
2. The use of ImPACTS demonstrated small step development between the standardised national framework grades used both in education and in health.
3. The use of ImPACTS provided shared ownership and responsibility for physical development between school staff and physiotherapists.

Conclusion
Correct use of the ImPACTs tool should enable school staff to assess and to make an informed decision alongside a physiotherapist about appropriate progression of interventions.

Introduction
In the current climate, both economically and politically, changes in how professionals work together across disciplines is under review. From the educational perspective the current Green Paper SEN (March 2011) re-iterates the need for greater joint working across boundaries of Health, Education and Social Care. The restructuring of many Primary Care Trusts challenges staffing and division of labour across all key areas of need. As professionals working with children we need to look at working practices that maintain standards, reflect current research and preserve our professional integrity.

For many years Rosewood School, a small non-maintained special school catering for children aged 2-19 years with PMLD, has adapted an inquiry based model for improving practice. With a full range of professionals working together it is essential that working practices allow professional expertise to be utilised most effectively for the benefit of the children and their families. The model adopted was a trans-disciplinary one, originally designed to serve high-risk infants (Hutchinson, 1978; United Cerebral Palsy, 1976). The model is characterised by a sharing of information and skills across traditional disciplinary boundaries. In contrast to the Multi- or Inter-disciplinary approaches the Trans-disciplinary model incorporates an indirect model of services. It involves Key Persons facilitating the services given to the child and other team members acting as consultants.
To develop this approach it is essential that professionals are confident in the Key Person’s abilities, share a common understanding of the child’s needs and do not allow the model to entirely replace direct intervention but accept limitations. The method is further supported by the underpinning belief, shared in the school by all professionals, that a thorough assessment of the child’s needs is essential prior to planning interventions.

Assessing a child with PMLD whose development in all areas is often very slow and unique in its features is a very real challenge.

Wolfe Schein (1998) wrote "it is important that individuals working with children who are severely disabled are given tools that enable them to address the relevant features of the child’s behaviour without trying to fit the behaviour into a pre-existing tool that was not developed for, or related to, the behaviour of someone with very special problems, i.e. unique abilities and patterns of growth”.

In education teachers are required to assess against National Standards of Performance, the ‘P’ Levels, having been devised for all children to show progress at working towards the National Curriculum. For pupils with PMLD 3 levels exist. The child may stay within those 3 levels for their whole school career making the ability to demonstrate progress or pick up on regression very difficult and hard for parents to feel a sense that their child is progressing. In physiotherapy, validated assessment tools exist for motor development i.e. the Gross Motor Function Measure 88 (GMFM) Russell et al (1993). We considered the research on Motor Development Curves specifically looking at Gross Motor Function Classification System (GMFCS) Level V (Rosenbaum et al, 2002) which indicates the limitations of expected motor development in these children. Interestingly the curves indicated that the significant motor development occurs before the age of 3 years. The pupil population at Rosewood School are all motor developmentally below the age of 3 years.

We wanted as a team to address our assessment approach and to influence our model of working, not only to demonstrate progress but also to raise professional standards. A working party consisting of 3 physiotherapists and the Deputy Head Teacher reviewed the following aims:

1. To devise an assessment approach valued both by physiotherapists and teaching staff.
2. To use assessment information to plan an intervention for an individual child that allowed progress to be charted and measured.
3. To devise a programme of training to raise skills, knowledge and understanding of motor development, intervention and handling techniques.

An existing model of Trans-disciplinary assessment and planning had already been devised in the school for communication, cognitive skill development and environmental control technology. Input from professionals included speech and language therapy, educational psychology and occupational therapy. It was decided that the ImPACTS Key Skill approach should be the working format and to develop it for motor development assessment and intervention planning.

ImPACTS is a developmentally sequenced assessment approach that leads to the development of an Individual Profile. The assessment reflects very small stages of progression and is divided into strands (i.e. areas of development e.g. in communication these include vision, hearing, interaction, etc.) It is on completion of the profile that a child’s strengths and areas of development can be identified and a linked curriculum offers further breakdown of progression, activity ideas, teaching points and other aspects to support the child. Once a year this assessment is reviewed and progress checked in each strand and in the area as a whole. Fine grading allows small progress to be measured and a pattern of learning unique to the child emerges.

Method

The physiotherapists in the school use the Gross Motor Function Classification System (GMFCS) (Wood 2000) as a basis for describing the levels of gross motor function in the pupils. The system was designed for use with children with cerebral palsy (CP) and Down’s Syndrome but the categories can be extrapolated to children who do not have CP using the levels (1-5) to differentiate.

The tool produced resulted in the development of a Profile covering 5 strands: supine lying, prone lying, sitting, standing, and sequencing of movement e.g. crawling. To meet educational requirements of assessment these were subdivided into three key developmental stages: Pre-Intentional, Intentional, and Formal that are cross referenced to the national ‘P’ levels (Figure 1).
By answering a series of questions, the boxes on the profile are etched to represent achievement on a criterion of 2 out of 3 positives equalling a developmental step. The etched boxes are totalled up to provide a score. For each strand area a total is recorded and then an overall physical skills development score calculated.

Once the profile is completed, the last etched box has an associated curriculum page with suggested objectives and therapy points. This allows further detailed analysis of the developmental stages and expectations of the child. It includes, where relevant, those aspects of intervention that should only be completed by physiotherapists or when a physiotherapist is present and working with the teaching team.

The curriculum allows the physiotherapist and teacher to agree the target to be added into the pupil’s Individual Education Plan (IEP). This target is often one of a number, others of which are met through direct physiotherapy intervention or through the routine activities in the class undertaken to prepare the child to learn e.g. wearing of ankle foot orthosis (AFOs).

**Case Scenario**

John (born in 2002) has a chromosomal anomaly, epilepsy, visual and hearing impairment. Physically he presents with hypotonia, hypermobile joints in his upper limbs, and has a scoliosis. John had a gastrostomy in September 2008. His gross function motor level correlates with a GMFCS Level V. He lives at home with his parents supported by his grandparents.

John’s equipment in school consists of:

1. a Kimba Spring Buggy
2. a James Leckey Advance chair
3. a raised Side lyer
4. an R82 supine standing frame
5. a brace
6. AFOs and Piedros
7. arm gaiters
8. thumb splint
9. Liftech sling and hoist.

John’s health has noticeably improved since his gastrostomy. His seizures, which include absences, jerks and tonic seizures have reduced over the last few months. He has also matured cognitively which has enabled his general development.

John was initially assessed using the ImPACTS tool in July 2008. He was reassessed a year later. The areas for development were indicated from the last box etched on each strand of the Profile: ‘Early Reflex Held’ (Figure 1)

The assessment questions from the sitting strand sub-section, Early Reflex are:

*With the child positioned on the floor and handled to achieve a wide base i.e. legs positioned out in front, adult seated and working from behind controlling with a high level of support.*
Can the child:

a) be positioned by placement in a sitting position without pushing/extending backwards?

b) from a curved forwards position, momentarily control the trunk upwards and then be fully supported to maintain the position?

c) hold a partially straightened trunk for increased brief periods with supports being lowered down the body?

It was found that John could manage to keep his trunk upright briefly while fully supported however he could not achieve ‘a’ and ‘c’ above therefore this was identified as an area that could be worked on.

Together the physiotherapist and teacher referred to the curriculum matched to this development area to agree the target (Figure 2).

Figure 2: Related area from curriculum.

<table>
<thead>
<tr>
<th>Sitting Strand – Early Reflex Held</th>
<th>Therapy points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Suggested Objectives</td>
<td></td>
</tr>
<tr>
<td>On the floor with a wide base of support and high level of support.</td>
<td>Ensure learner feels secure</td>
</tr>
<tr>
<td>To tolerate being positioned in a sitting position without pushing or extending back.</td>
<td>Check position - bottom back, neutral pelvis [severity of contractures or spinal deformity may affect this]</td>
</tr>
<tr>
<td>To momentarily join with sitting up slightly from a forward curved position</td>
<td>Ensure child is receiving positive but simple sensory messages.</td>
</tr>
<tr>
<td>To increase degree of sitting up trunk control by:</td>
<td>Tone may vary, specific levels of support required [may help to raise tone first by ball/roll activities or by joint compression]</td>
</tr>
<tr>
<td>a) increasing time of activity</td>
<td>Small pillow under bottom may prevent sacral sitting; small cushions under outer edge of knee if tone is low and legs turn out too freely some arm support on pillow on lap may be of value.</td>
</tr>
<tr>
<td>b) lowering level of trunk support down body.</td>
<td>Keep session short</td>
</tr>
<tr>
<td></td>
<td>Care with head and airway, support head as necessary [second carer for larger child]</td>
</tr>
<tr>
<td></td>
<td>Think about surfaces [mat/playmat]</td>
</tr>
</tbody>
</table>

John’s IEP target was set as the following:

To facilitate the raising of tone leading to head and trunk control by:

a) sitting on box with brace on, mid thoracic support from behind and with arm prop on the table, grasping the bar – to raise head for 5 seconds x 3.

b) in supported side sitting to prop with forearm on small roll with some head raise x 30 seconds.

This activity was shared with the class staff and incorporated into John’s daily routine. The ‘Therapy Points’ (Figure 2) are designed to act as prompts for the staff when they are working on the activity. John’s IEP target was practised regularly by the classroom staff, who notified the physiotherapist if there was any change.

The following year, John was reassessed and managed to graduate to the box ‘Sitting Awareness’ (Figure 1)

Discussion

The Transdisciplinary Model

There are usually three concerns cited by professionals concerning the movement towards a transdisciplinary approach:

[1] it is the therapists themselves who have the expertise and specialist skills and training that teachers have not acquired;

[2] with therapists adopting an indirect model, more input will happen in a classroom where there may be more distractions for the child;

[3] loss of professional identity as educational staff take on more of the therapy interventions.

It is worth discussing all three concerns as it shows further benefits to the model. As regards professional identity the method encourages greater awareness and understanding of different professionals’ roles and offers the potential for role transition and this can be a two way development.

In the school all therapy staff shared training in the educational beliefs and values of the teaching staff and regularly attend joint in-service training (INSET). Training by the therapists and specialists form part of the education staff’s INSET. This links in to the view of how can, or even should, teachers do the work of physiotherapists. The reality is that it is the educational staff have most access to the child and can offer more flexible timetabling to match the bio-behavioural states of the child.

Shared Assessment Approaches

It is important that time is given to a thorough assessment of a child’s need. The process of assessment is a time consuming one but arguably without it the impact of teaching would be restricted.

Vygotsky (1978) showed through his research on Zones of Proximal Learning that if you do not know which skills a child already has and utilise these to move to the next stage, progress will not be achieved. The assessment of children with PMLD
requires sustained observation periods and opportunities for the child to learn through daily routines. (Lacey, P, Summer 2008). For therapy to be effective it must be integrated and a shared assessment means that priorities can be agreed and a sense of the whole child developed. Regular multi professional meetings need to take place to support assessment and the model so that all professionals agree what the priorities are for a child at any given time, i.e. a post operative child may require additional physical support so that less emphasis is given to cognitive skills for a time; a different child might be attaining contingency awareness and sustained cognitive experiences might support the consolidation of this major learning milestone.

Conclusions
The ImPACTS model has allowed progress to be plotted and has facilitated greater shared working practices across the disciplines. The model has been applied to other children across the spectrum of PMLD. Successes have been found and it has also supported the early identification of regression patterns and triggered full team re-assessments.

The approach is about effective team work and teams take time to form (Hutchison 1978). The change in the school’s physiotherapy team has meant that time has been needed for new relationships to form and professional trust to be built. The model and the assessment approach have been useful to facilitate development and allowed the standards and working practices to remain. The approach is supportive of the new physiotherapist exploring and developing an understanding of the role required in the school.

Due to the number of pupils in the school it would be difficult, without further empirical data based research to say whether the tool alters the expected gross motor development potential for these Level V children. The tool allows real measurable progress to be shared between professionals and most importantly in a way parents/carers can understand.

References:


Hutchinson, DJ (1978) The transdisciplinary approach. In J>B> Curry & K.K Peep (Eds), Mental Retardation: Nursing approaches to care (pp.65-74). St. Louis: C.V. Mosby

Lacey, P (2009) Developing The Thinking of learners with PMLD. PMLD Link Vol 21; No 2 Issue 63


United Cerebral Palsy, National organised Collaborative Project to provide Comprehensive services for atypical infants and families (1976) Staff development handbook: A resource for the transdisciplinary process. New York: United cerebral palsy Association


**Case Report**

**Management of a Focal Dystonic Crisis in Cerebral Palsy: Use of Botulinum Toxin**

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**Introduction**

Dystonic Cerebral Palsy (CP) is the smallest classified group of CP, coming within the sub-group of athetosis, which results from a lesion primarily in the basal ganglia. Although the mechanism behind observed involuntary movements is unclear we do know that the basal ganglia are involved in the refinement of information being transferred to the cortex and motor areas, the scale and amplitude of movements as well as the control of automatic background activity. The reported incidence of dystonic CP varies from 2-15% of the CP population (Reid et al, 2010).

This case report focuses on a 5 year old boy, ‘E’, with a primary diagnosis of CP secondary to prematurity. He presents with a mixed tonal pattern, predominantly those of a child with dystonic CP. The management of a growing number of such children is increasingly complex, putting additional demands on therapy departments.

A child presenting with dystonic CP secondary to prematurity requires careful assessment and management. The child often presents with central hypotonia, poor dissociation between the pelvic and shoulder girdles, fluctuating tone and dystonic posturing, resulting in ever-increasing stresses on the musculoskeletal system.

Botulinum Toxin has been used to treat spasticity since the early 1990s. Its efficacy in reducing spasticity in children with CP has been validated in studies (Keewon et al, 2011; Love, Graham et al, 2010).

Signed written consent for publication was obtained from the child’s parents prior to writing this case report.

**History**

‘E’ presented with a 4 day history of severe left upper limb posturing and pain. Assessment indicated that it resulted from an upper respiratory tract infection. The community nursing team had previously advised medication to manage the pyrexia. Since becoming unwell, his general tone had increased, particularly in his left upper limb which had become fixed in an abnormal position.

‘E’ was born at 24 weeks and required 9 months of respiratory support. He was diagnosed with 4 limb CP (GMFCS level 5). All areas of his development have been and continue to be affected. Tone fluctuates throughout his body from hypo to severe hypertonia, exacerbated by effort, emotion and pain. Support is required to maintain symmetry in all lying sitting and standing positions. He had a past history of left shoulder posturing and possible subluxation. ‘E’ was already known to the tertiary movement disorder clinic prior to this episode. The clinic that had advised a trial of Trihexyphenidyl with positive effect.

**Acute intervention**

On initial examination at home, ‘E’ was unable to be placed in any position and was being held by his mother at all times. When placed on the bed, his whole body arched into extension with rotation to the left. His left upper limb was fixed in pronation and extension at the shoulder and elbow, with his hand fisted and thumb adducted.

On palpation his left humerus appeared subluxed and at risk of further injury. When held by his mother, with his left arm against her body, he settled for approximately 15 minutes at a time. Different positions were trialled including cradling him in a more flexed position. The left arm could be brought into neutral flexion/extension in this position, but remained in pronation and the child had difficulty settling.

Discussion between his mother, physiotherapist and paediatrician concluded that ‘E’s’ pain was worsening and he was becoming exhausted secondary to lack of sleep and possible infection. Admission to the local hospital was arranged via A&E.

**Inpatient Local Hospital Care**

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Following Morphine, two doses of Diazepam and Chloral Hydrate, ‘E’ slept and his shoulder position improved. Discharged the next day, within hours his symptoms reappeared. His overall posture was improved, the left arm had reverted to the same extreme position and he was in pain on any movement.
The physiotherapist trialled various positioning techniques, specifically for the left upper limb. Triceps was palpably rigid and attempts to massage it were clearly painful. By abducting the thumb, the physiotherapist was able to position and strap his left arm across the body in more flexion. ‘E’ could then be placed in a Tumbleform chair for short periods of time. Liaison with the local hospital ensured that the child could return to the ward for more medication if required.

Referral to a Tertiary Centre and Care
An urgent review was requested at the tertiary movement disorder clinic. This review highlighted ‘E’s’ significant pain levels, for which three analgesics and an increased dose of Trihexyphenidyl had already been prescribed. The treatment plan, following assessment by a neurologist and physiotherapist, was to inject Latissimus Dorsi and Triceps (both palpably rigid) with 50 units of Botulinum Toxin A (Dysport) (Btx A). ‘E’ was also provided with Diazepam and Chloral Hydrate to use at home if necessary and commenced on a small dose of Baclofen.

Goals for Btx A intervention
Short Term
1. To reduce pain levels, measured through monitoring analgesia levels
2. To increase length of time that ‘E’ was able to sleep at night
3. To allow ‘E’ to be placed in his postural management equipment (seating and lying support)

Long term
1. To allow a window of opportunity for ‘E’ to develop a preferred switch position to improve communication
2. To help determine suitable treatment strategy to manage ‘E’s’ left shoulder posturing and help reduce the risk of further dystonic crises.

Community Care
Two days later the local physiotherapist reviewed ‘E’ and advised the mother on:
1. The use of positions to maintain postural alignment
2. The maintenance of a flexed posture or a ‘nested’ position (hips and knees flexed) thus reducing tone and allowing the head to remain in midline.
3. The facilitation of shoulder protraction using pillows.
4. Encouraging hands to midline to touch his face and other activities
5. The use of a small soft object placed in his left hand to help maintain thumb abduction

Because of concern regarding possible ‘neglect’ of his left upper limb, his Mother was encouraged to slowly increase his awareness of that limb through massage, placing objects in that hand, looking at it, etc.

Outcomes
Improvement in symptoms following Btx A injection was observed a few days post-injection, with ongoing improvement over time. His left upper limb position improved dramatically and, when supported in his seating system, he was able to rest both hands on his knees comfortably. Physiotherapy focussed on facilitating the left upper limb into midline and flexion by reading books and playing with toys in various positions. Hydrotherapy in the early post-injection period facilitated rehabilitation goals, decreasing the influence of gravity and providing a gentler environment in which to exercise (Kelly et al 2005).

Orthotic Care
‘E’ was assessed a month later for a shoulder stability lycra garment to be used as tolerated. On further review it was decided to add sleeve length and paneling to facilitate external rotation and a thumb loop to maintain thumb abduction.

Discussion
This case report may be of interest to paediatric physiotherapists as it demonstrates the effective use of Btx A in the management of an acute focal dystonic crisis.

Dystonic CP can be difficult to manage. Fluctuating tone causing extreme posturing through range seems to lead to lower use of Btx A in the early years. Instead therapists and paediatricians cooperate to ensure that the child receives other medication and an integrated postural management/therapy approach to manage tone as efficiently as possible. Therapists’ main concern is often to create proximal stability. They focus on postural management systems to provide this and therapeutic handling to facilitate improved postural tone and the grading and control of movement (Bobath course notes 2009).

Use of medication in the management of tone is widespread. ‘E’ was already prescribed Trihexyphenidyl, an antimuscarinic acting on synapses, blocking receptor sites. He was subsequently prescribed Baclofen, a skeletal muscle
Lycra garments are increasingly used for children with CP. Some evidence suggests there is measurable improvement associated with their use (Angilley 2008, Knox 2003). They provide stability, both centrally and in larger limb joints. Shoulder stability garments provide specific support when the posture of the shoulder is affecting overall posture.

Multi-disciplinary teams regularly refer children with CP for Btx A assessment when there is stiffness and reduced movement related to spasticity. Btx A can give the team a window of opportunity to try to maximize length in the injected muscles whilst strengthening the opposing muscle group. As a result of ‘E’s’ fluctuating tone and previous management, he had relatively good muscle length throughout and had no lower limb pathology. He had not therefore previously been considered a good candidate for Btx A treatment.

This case report highlights that children experiencing a focal dystonic crisis can benefit from Btx A injections. Medication worked initially to sedate him but this was not found to be the most functional management strategy. By directly targeting the muscles causing the posturing, subluxation and pain, a positive effect was achieved. Not only did this give fairly rapid relief but there was also an opportunity for rehabilitation post-injection.

The positive effects of the Btx A lasted approximately 2 months, after which the tone in his left upper limb began to increase and the posturing returned, albeit less severe. This was obviously frustrating for him and his Mother as well as the team working with him. It is considered that repeat injections may be beneficial.

Once again the benefits of multi-disciplinary working with this client group is demonstrated in providing support and care at home, school, local hospital and in a tertiary centre. Through joint working and good relationships with acute partners the local team was able to support ‘E’ and his family through a focal dystonic crisis using the most appropriate intervention. With an increasing number of complex children to manage in primary care, it is vital to have good relationships with tertiary partners. This supports effective communication and a sharing of knowledge and skills between agencies. It also facilitates involvement of the whole team and family in the decision-making process and most importantly, helps to achieve the best possible outcomes for the child.

**Conclusion**

This case study describes one way to manage a child experiencing a focal dystonic crisis. Early identification of the main problems and working in partnership with the tertiary centre meant that this child had access to Btx A as an acute intervention to manage his dystonic posturing subluxation and pain.

The use of Btx A in this way is not widely publicised and may not be at the forefront of community therapists’ minds when they are clinically reasoning through a patient with this presentation. As anticipated, the effects of the Btx A wore off after 2 months and the symptoms began to return. The question requiring further investigation is whether regular Btx A could be a long term solution to the management of a dystonic upper limb in order to prevent crises occurring.

**Acknowledgements**

The authors would like to thank the family concerned for giving their consent to share this experience with other paediatric physiotherapists.

**References**

- British National Formulary No 61. March 2011
- Bobath Course Notes. Winter 2009
ABSTRACT

Background: There is a lack of evidence to support the amount of physiotherapy needed by children with cerebral palsy (CP), yet physiotherapists are increasingly requested to provide specific and quantified advice regarding amounts of intervention.

Objectives: This survey aimed to establish the amount of intervention physiotherapists in the National Health Service (NHS) are currently providing for children with CP at different severity levels and ages across the UK. This survey did not aim to establish whether the amount of time spent makes a measurable difference to outcome.

Method: A questionnaire was piloted, and paediatric physiotherapists working in the NHS across the UK were invited to participate. Questions asked included the age and severity level of each child, plus the amount of therapy provided each week over one year. Therapy time was defined as that spent in contact with a child or his/her carers by a qualified physiotherapist.

Results: A total of 245 physiotherapists completed the survey between January 2009 and December 2010, reporting intervention levels for 1425 children aged 0-18 over one year. The children receiving the most intervention over one year were those aged 0-6, GMFCS Level V (median 17.6 hours, interquartile range 11.5-23.5), and the children receiving the least intervention were aged 12-18, GMFCS Level I (2.3 hours median, interquartile range 1-4.6). Across all ages and GMFCS Levels, overall the most time in one year was spent on Body Functions and Structures (WHO, 2002) at 3.8 hours (median), with 2.9 hours (median) on Activity and 1 hour (median) on Participation.

Conclusions: This survey provides a national reference for the amount of physiotherapy that is actually being provided by physiotherapists in the NHS in the UK. This information may be useful for physiotherapists to use as a background against which they can set their own individual advice.
ABSTRACT

Background: Physiotherapists are required under the European Core Standards of Physiotherapy Practice to select valid, reliable and published outcome measures.

Objectives: This study examined the psychometric properties of the Functional Walking Test (FWT).

Method: 56 subjects with cerebral palsy (CP) (21 females and 35 males; mean age 9 years 6 months; SD 3 years 9 months; range 4-17 years) were assessed on two occasions, 6 months apart, using both the FWT and the Gross Motor Function Measure (GMFM).

Results: Generalisability correlation coefficients (GCC) for all 11 items were high (0.91-0.99). Inter-rater reliability was also high with excellent consensus in the scores given by the eight raters (intra-class correlation coefficient and GCC 0.99). Intra-rater reliability was equally high (GCC 0.99). The internal consistency of the FWT was estimated using Cronbach’s alpha as 0.95 and 0.94 at Time 1 and 2, respectively. The FWT had a high degree of correlation with the GMFM when total scores were compared at Time 1 and 2 (Pearson’s r 0.86 and 0.87, n=56, p<0.01). The FWT also found statistically significant difference in total scores between the 3 Gross Motor Function Classification System (GMFCS) levels. The correlation between the FWT scores and GMFCS was -0.70 at Time 1 and 0.76 at Time 2 (p<0.01) indicating the construct validity of the FWT.

Conclusions: The FWT was designed to be a quick, easy to use measure of functional walking ability at the level of Activity within the framework of the International Classification of Functioning, Disability and Health. This study has demonstrated that the FWT has sound psychometric properties and is valid and reliable in a sample population of ambulant children with CP.

Pschometric evaluation of the Functional Walking Test for children with cerebral palsy.

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ABSTRACT

**Background:** Instrumented 3-dimensional gait analysis is the gold standard in the management of gait in cerebral palsy (CP). However, the majority of children are managed clinically by physiotherapists using observational gait analysis. Less objective assessment and few relevant indicators for monitoring CP gait leads to management variation (Skaggs et al, 2000; Ballaz et al, 2010). Of UK paediatric physiotherapists 98.6% stated a need for a short, clear, reliable and valid tool (Toro et al, 2003). The Wee Glasgow Gait Index (WeeGGI) is scored, by selecting one of three parameters. Each parameter has clear descriptors and values, and has been identified by Westmarc neurobiomechanics team as key indicators of gait cycle breakdown in CP.

**Objectives:** The study aim was to assess initial validity by comparing the WeeGGI with the Edinburgh Gait Score (EGS) (Read et al, 2003) a validated, observational gait tool used in gait laboratories.

**Method:** 75 limbs of 42 children with CP were scored by both the WeeGGI, and EGS. The EGS produced a 0.91 Pearson Product Moment Correlation Coefficient (PPMCC). The WeeGGI scored ‘live’ and from video produced an equally high correlation of 0.91 PPMCC.

**Conclusions:** Initial results are encouraging for a clinically based, clear, short and easy to use gait tool.

**References**


Ballaz L, Plamondon S, Lemay M: Ankle ROM is Key to Gait Efficiency In Adolescents with Cerebral Palsy. Clinical Biomechanics, article in press 2010.


ABSTRACT

Background: Pre-term infants are at increased risk of behavioural, cognitive and physical difficulties (Woodward et al, 2009). Guidelines exist for the management of pre-term infants in the inpatient setting (Department of Health, 2009), however there is little guidance on the management of these infants once discharged.

Objectives: The aim of this study was to see if a consensus opinion could be found for the physiotherapy management of pre-term infants once discharged from the inpatient setting focusing on referral, management and discharge.

Method: An expert panel of physiotherapists experienced in the management of pre-term infants in the outpatient setting completed a series of questionnaires following the Delphi method. A 5-part Likert scale was used and consensus was set at 75% of participants strongly agreeing with the statement. Participants were asked to describe their ideal service without limitation in resources.

Results: 16 out of 18 participants completed the study each with an average of 9 years and 11 months’ experience of following up pre-term infants. A total of 56 out of a possible 126 statements reached consensus, including which infants should be followed up, who should follow them up, frequency of intervention, and at what point they should be discharged.

Conclusions: Expert physiotherapist opinion suggests children outside the recent Department of Health guidelines (2009) should be followed up and this should be multi-professional. A physiotherapist’s clinical reasoning and professional autonomy will ultimately guide best practice, however recommendations can be made to help manage equity across services and develop guidelines for this cohort of infants.

References
ABSTRACT

Background: Great Ormond Street Hospital Neuromuscular Physiotherapy Team were increasingly concerned by the incidence of sleep disturbance in childhood neuromuscular disorders (NMDs). In conditions such as Congenital Muscular Dystrophy, children can wake up to 12 times nightly. The negative implications of lack of sleep affect the children, parents/carers and siblings. No research has been found into the cause of sleep disturbance in children with NMDs and there are no studies to determine if sleep systems, splints, or profiling beds relieve or exacerbate the problem.

Objectives: To audit the causes and frequency of waking at night in children with neuromuscular conditions.

Method: A questionnaire has been added to the routine physiotherapy assessment to audit the cause and frequency of waking at night including factors such as splints, scoliosis, contractures, forced vital capacity. All children over the age of 3 years with a confirmed neuromuscular diagnosis will be included in the audit. The aim is data collection for 1 year, from June 2011 from approximately 300 ambulant and non-ambulant children.

Conclusions: Early findings show that for ambulant children the main cause of waking is to remove night splints. For non-ambulant children a variety of factors have been identified most notably, weakness and contractures preventing independent bed mobility.

Implications: Results from the audit should identify the causes of sleep disturbance. It is hoped that from this, better physiotherapy advice on night time management will be possible, however further research may be necessary into specific strategies leading to improved sleep for the whole family.
BOOK REVIEW
Physiotherapy and Occupational Therapy for People with Cerebral Palsy:
A Problem-Based Approach to Assessment and Management

Editors: Karen J. Dodd, Christine Imms, Nicholas F. Taylor

Reviewed by Eileen Kinley

I enjoyed reading this book. It uses theoretical models to provide the basis for planning physiotherapy and occupational therapy interventions for children and adults with cerebral palsy. To a very large extent it succeeds in this aim.

There is an initial scene setting foreword from Peter Rosenbaum. This provides a very helpful context for the chapters that follow and emphasises that current perspectives on terminology, functioning, and on understanding cerebral palsy should be the basis for the interventions which are used in practice.

The editors are two physiotherapists and an occupational therapist. In part I of the book they emphasise and describe the clinical reasoning process as the basis for the intervention process model (adapted from the Canadian Occupational Performance Process Model) which they advocate and use. In addition they incorporate the framework of the International Classification of Functioning, Disability, and Health (ICF), a family-centred approach to therapy and multidisciplinary practice models.

Chapter II provides an excellent overview of ‘What is cerebral palsy?’ in terms of its classification, aetiology and outcomes. I would have preferred a stronger emphasis on recent changes in terminology as well as its consistent use throughout the text.

Subsequent parts describe treatment chronologically with the use of case studies to illustrate the approach used, which is particularly helpful. Some statements provide cause for thought with respect to their accuracy. For example, chapter V on the infant with complex needs infers that the General Movements Assessment is an accepted indicator for informing a family that a young baby is “at risk of having cerebral palsy”. I would not regard that this is a sufficient criterion for such a diagnosis. The illustrations in this chapter are limited in their number and usefulness for therapists.

Chapter X dealing with physiotherapy following single event multilevel surgery is excellent and illustrates the strengths of the book. The chapter is comprehensive, clear and well illustrated. It lists a number of key clinical messages.

Given that the majority of people with cerebral palsy are adults, it is refreshing that both transition and adults with cerebral palsy are considered in detail with respect to their therapeutic needs. I thought that these chapters covered the issues well.

This book succeeds in its stated aims of providing a practical guide for physiotherapists and occupational therapists, as well as being of interest to student therapists and other health professionals.

I am happy to recommend it and I consider that it should be available in paediatric physiotherapy departments. Both experienced physiotherapists and students are likely to find it a helpful resource.
This is a welcome revision and extension of Sophie Levitt’s pragmatic and well used book for therapists. In collaboration with Dawn Pickering she has provided additional material on evidence based practise, research and updated her own writing including new evidence and current opinion. It is aimed at practising paediatric therapists involved in optimising the functional motor development of children with cerebral palsy and supporting their families and carers. It also has a broader appeal to other professionals in the field. Although easily readable the therapy language may make some sections less accessible for parents and teachers. Sophie Levitt herself suggests therapists provide additional interpretation to help non therapy readers.

The book has 13 chapters which I found logically ordered. I would recommend reading the preface in which she sets out her objectives. The contents page includes more detail of each chapter, allowing a quick overview. The references are included at the end of the book rather than at the end of each chapter, and are clearly laid out.

The first chapter describes the clinical picture of cerebral palsy (CP) and introduces The International Classification of Functioning, Disability and Health (WHO) and its relevance to therapy practice. The message of functional gain is a strong theme throughout subsequent chapters. This first chapter is limited by the absence of the more current movement disorder definitions and classifications now agreed and used internationally. Most therapists reading the detail will recognise the descriptors which are still useful in classifying children’s movement problems.

The chapter on a collaborative learning approach outlines concisely a family centred therapy approach which is now supported by research. This is followed by an outline of many different treatment approaches and theories which I think is one of the unique features of this book. Sophie Levitt brings her own wealth of personal experience, providing descriptions which are well balanced, and discussions that allow the reader to put them into context with scientific evidence. This would be particularly useful for less experienced therapists who may not have a working knowledge of some of the interventions on offer, especially at a time when many parents are considering alternative treatment modalities and looking outside local NHS provision for additional therapy support.

The chapter on evidence based practice emphasises the importance of clinician evaluation of available evidence and caution when applying this to clinical practice. I particularly support her statement, “It is still unwise to be dogmatic about a theoretical framework or about procedures” which arise from limited evidence. Different study designs, their merits and limitations are discussed and there are useful suggestions for getting the most out of published trials.

As an eclectic practitioner I found the chapter on synthesis of treatment systems readable and accessible. Her ability to bring original theory and current evidence together is an enviable skill. This and the subsequent chapter on motor learning are a ‘must read’ and lead on well to the rest of the book which is mainly practical.

The assessment chapter is current, well referenced and provides useful discussion on interpretation of assessment findings to help treatment planning. Following on from this is the largest chapter of the book - treatment procedures and management - which has photographs, simple clear line drawings and some useful developmental tables. This chapter is very practical and while not all encompassing, it is quite extensive.

The chapter on function and daily life brings the reader back to why children need movement, breaking down the requirements of functional skills and reminding us of how challenging it is to be truly holistic in therapy practise. The book then considers causes of deformity, how this affects function and options for management. There is also a good chapter on the older person with CP which considers wider aims and transition into adult life.

This very practical, portable and readable book has many strengths, not least the affordability. I am glad that this book has been updated. It retains Sophie Levitt’s strong practical, eclectic emphasis and with its updates remains an excellent resource for all paediatric therapists working in this field.
Descriptions of Rare Diseases Relevant to Paediatric Physiotherapy

A disease or disorder is defined as rare in Europe if it affects less than 1 in 2000. One rare disease may affect only a handful of patients in the EU, and another touch as many as 245,000. There are between 6000 and 8000 rare diseases. On the whole, rare diseases may affect 30 million European Union citizens. 80% of rare diseases are of genetic origin, and are often chronic and life-threatening.

European Alliance for Rare Diseases
www.eurordis.org

It is estimated that up to 4 million children and adults are affected by rare disorders in the UK according to Contact a Family
www.cafamily.org.uk

Pompe Disease

Pompe disease is a rare, progressive muscle wasting disease that can affect infants, children and adults. It is one of more than 40 genetic disorders that are known as Lysosomal Storage Disorders (LSD). It is also classed as a metabolic disease – Glycogen Storage Disorder (GSD) and, due to its progressive nature, a neuromuscular disorder.


There is a varied clinical spectrum. The infantile onset is characterised by hypertrophic cardiomyopathy and profound generalised weakness which presents in the first few months of life with rapid disease progression. Late onset disease (juvenile and adult) is characterised by onset of symptoms after one year of age, less severe to no cardiac involvement and a slower progression with symptoms primarily related to progressive dysfunction of skeletal and respiratory muscles.

Pompe Disease is an autosomal recessive disorder characterised by deficiency or dysfunction of the lysosomal enzyme, acid alpha-glucosidase (GAA). Without GGA the lysosomes can not break down any excessive amount of glycogen primarily found in muscle cells. The resultant build up of glycogen causes the lysosomes to expand until they take up so much space that the muscle cell is damaged. Glycogen begins to leak out of the lysosomes causing further damage to the surrounding muscles cells. Myopathy is a result as muscle fibres enlarge. Hydrolytic enzymes are believed to be released which also have a role in muscle destruction. Healthy myofibrils are damaged and muscle function is greatly impaired. Cardiac, skeletal and smooth muscle are affected. Most patients with infantile onset disease have minimal to undetectable enzyme level leading to massive glycogen accumulation and rapid, aggressive progression of the disease. In contrast, late onset patients tend to have limited, but detectable, residual GAA activity and in these cases organ damage is less pronounced and disease progression slower. Progressive muscle weakness is the most common symptom of all forms of Pompe Disease.

Incidence

Pompe is a rare disease, with an estimated incidence of 1 in 40,000 to 1 in 100,000.

Diagnosis

Clinical diagnosis is confirmed by the absence or virtual absence of the GAA enzyme.

Presentation

Infantile Pompe Disease is the most severe form. Typically symptoms appear in the first few months of life, with feeding problems poor weight gain, muscle weakness, floppiness, respiratory problems, cardiomyopathy and failure to achieve motor milestones. In late onset disease symptoms appear any time after the age of one year and there is a huge variation in clinical manifestation. Progression tends to be much slower. Progressive proximal muscle weakness is still a feature as are functional deficits, respiratory insufficiency and osteoporosis. Progression of weakness often leads to the use of mobility aids, from walking sticks to powered chairs.

Enzyme Replacement Therapy

Historically infantile Pompe Disease resulted in babies dying before their first birthday, most commonly due to cardiorespiratory failure. Clinical trials successfully showed that Enzyme Replacement Therapy (ERT) reduced mortality. Recombinant (genetically engineered) enzyme (Myozyme) has been commercially available since 2006. ERT has become a recognised treatment and timely
intervention has slowed disease progression and improved cardiomyopathy. Some children who have started treatment early in the disease progression are now surviving with none or very few signs of the disease. Some have not responded as would be hoped, and the disease has continued to progress.

**Clinical Presentation**

Patterns of weakness have been described in Pompe Disease which are worse proximally than distally and greater in the lower extremities than upper extremities. The muscles in the trunk are also weak. Usually it presents symmetrically but imbalanced across joints. In the infantile form progressive and profound weakness occurs in the neck, trunk, extremities and facial muscles. There tends to be marked cardiac involvement with hypertrophic cardiomyopathy. Respiratory weakness also occurs with diaphragmatic weakness along with the accessory muscles. Frequently babies with Pompe Disease have been developing problems for quite some time before a diagnosis is made and treatment started; ventilatory and cardiac support may be required to enable them to overcome an acute period of illness or until there is a response to treatment. In the late-onset form progressive proximal myopathy occurs with greater variation in distribution, extent and rate of progression. Cardiac involvement is not typical in this form but respiratory involvement can be severe also requiring ventilatory support.

**Physiotherapy Management**

Intervention is aimed at optimising and preserving motor function within the limits of the disease. There are a number of assessments that can be used including standardised measures and video but components should include: range of movement, muscle extensibility, posture and alignment, hip stability and motor development.

**Muscle Weakness**

Currently there are no national guidelines for strengthening programmes for children with Pompe Disease but literature suggests that exercise should be aerobic sub-maximal to avoid over fatigue and further damage to the muscles. Active assisted exercise and movements to encourage head and trunk control, weight shift and sequencing between positions in sitting, crawling and standing may be encouraged. Activities should be functional and avoid excessive resistance with incorporation of rest and recovery periods. Adaptive equipment and orthotic intervention can be used to support function. For late onset individuals and those with residual strength gentle sub-maximal and aerobic functional exercise are recommended. Swimming/cycling, etc. without excessive resistance and active assistance may be considered. Hydrotherapy may be beneficial but precautions with regard to medical stability and prolonged exposure to heat need to be taken into account. Specialist buggies and wheelchairs need to be considered for long term use if the child is severely affected. Some children may only need a buggy for long distances.

**Muscle Contractures**

Contracture formation and deformity can develop from chronic alteration in posture and positioning which results from weakness. It is important to use positioning, stretching and exercise programmes to counteract these forces. Wraps, pillows, seating systems, standing frames, splints, etc. may be used to provide adequate support for all joints in all positions. Two joint muscles are at increased risk of early contracture. Areas particularly at risk are muscles around the hips, knees, ankles and elbows.

**Respiratory Compromise**

Respiratory muscle weakness can cause complications. Monitor for signs of deterioration such as an increase in the work of breathing or thicker secretions which would require medical intervention and treatment.

**Conclusion**

Physiotherapy is considered an important component in the management of Pompe Disease and the advent of ERT has increased the importance of maximising the clinical and functional benefits that may now be achieved.

**References**

Case L; Kishnani P. Genetics in Medicine. Physical Management of Pompe Disease: May 2006 vol. 8 no. 5

Baethmann M, Straub V, Reuser A. Pompe Disease. 2005 A Clinicians Guide to Pompe Disease, Genzyme 2005

What’s Missing – A deeper understanding of Misfolded Enzymes and LSD’s, Amicus therapies 2008

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STRETCH FOR THE TREATMENT AND PREVENTION OF CONTRACTURES

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Many of us working with children with neurological or musculoskeletal conditions spend much of our time trying to maintain joint mobility and prevent contractures of muscles and joints. The commonest strategy used is stretching programmes of various kinds (passive and active stretches, positioning, splints and / or serial casting). Consensus as to how to apply stretch, how long to stretch, how frequently to stretch, or for how many repetitions, is lacking. This is illustrated by the differing advice given, even within one small physiotherapy department (for over 100 examples of techniques used to administer stretches see http://www.physiotherapyexercises.com/).

Contractures interfere with activities of daily living, can cause pain, sleep disturbance and pressure problems as well as severe deformity and increased burden of care. Importantly a large number of resources are allocated to the administration of stretch for the treatment and prevention of contractures. Although controversial and challenging, a review of the literature on the efficacy of stretch for the treatment and prevention of contractures is to be welcomed. The authors of this review looked at randomised controlled trials and controlled clinical trials of stretch applied with the purpose of preventing or treating contractures, with the primary outcomes identified as joint mobility and quality of life. Their secondary outcomes of interest were pain, spasticity, activity limitation and participation restriction. Thirty five studies with 1391 participants, were identified. Twenty four studies were with participants with neurological conditions (stroke, spinal cord injury, traumatic brain injury, Cerebral Palsy, Charcot-Marie-Tooth, Duchenne Muscular Dys trophy) and eleven studies with participants with non-neurological conditions (total knee replacement (TKR), anterior cruciate injury (ACL), ankle fracture, post-radiotherapy for breast cancer and jaw cancer, systemic sclerosis and fragility). All participants were adults.

The reviewers included studies that compared stretch with no stretch, stretch versus placebo or sham stretch, and stretch plus co-intervention versus co-intervention. To reduce complexity studies comparing stretch with another stretch or stretch with another intervention were not included.

Results showed that in the primary objective of this systematic review on the effects of stretching programmes on treating and preventing contractures, the moderate to high quality evidence available showed that stretch does not have a clinically important effect on joint mobility in people with neurological conditions in the short or long term, estimating that any possible treatment effect is not greater than 3°. Few would consider a treatment effect as small as 3° to be clinically important. Most studies investigated the use of stretch over short term periods (4-12 weeks) with none more than 7 months. Likewise there were no short term effects on joint mobility in people with non-neurological conditions. The effects on people with TKR and ankle fractures were small and clinically unimportant and the effects were unclear in those with ACL injuries.

There was medium quality evidence, needing to be interpreted with caution, suggesting that stretch causes immediate increases in pain in people with neurological conditions but no evidence to show a long term deleterious effect. There was no indication to show that stretch caused pain in the non-neurological group. Few studies included measurement of spasticity and those that did showed no clear effect of stretch on spasticity. Looking at quality of life issues there were no underlying changes at the impairment level so it is difficult to suggest that stretch has any clear benefit in improving quality of life or reducing activity limitation and participation restriction.

Implications for practice: The authors of this review have shown, from the literature available, that stretch does not have clinically important effects on joint mobility in people with or at risk of contractures and that, if performed for less than 7 months, the routine use of stretch in the management of contractures is of ‘no or little benefit over and above the usual care’. No studies used paediatric participants so the effects of stretching programmes on children to prevent or manage contractures is still unclear and largely anecdotal and particularly challenging during periods of rapid skeletal growth.
Selective dorsal rhizotomy is an intervention which aims to reduce sensory input to those reflex arcs responsible for increased muscle tone.

The purpose of the guidance is to make recommendations on the safety and efficacy of the procedure. It states that the procedure may be used, as there is adequate evidence for its efficacy. However, it does not make any recommendations as to whether it should be funded by the NHS.

The procedure, which is irreversible, involves a laminectomy of one or more vertebrae, allowing relevant sensory nerve rootlets to be identified and divided, while preserving some sensory input and motor roots responsible for voluntary movement. The procedure, which is irreversible, is not without risk, and there may be deterioration in bladder control or walking ability, as well as a risk of spinal deformity in the longer term. Patients will require prolonged physiotherapy and aftercare, and may need to learn to walk again. There may also be a need for additional surgery.

This guidance replaces that previously issued by NICE in 2006 (Intervention Procedure guidance 195), and follows the Intervention Procedure Overview carried out in July 2010. This was based on one meta-analysis of three Randomised Controlled Trials (RCTs) involving 1048 patients, six RCTs, and six case series, as well as opinions from a group of four nominated Specialist Advisers. It is noted that most of the evidence relates to children aged 4-10 years.

The importance of gaining informed consent is emphasised, as well as appropriate patient selection, which should be carried out by a multi-disciplinary team to include a physiotherapist, paediatrician, and surgeon, all of whom should have specialist training in the management of spasticity.

Finally, it is acknowledged that this is an evolving procedure, and further research is encouraged, with particular emphasis on long-term outcomes.

Reference

References for studies included in the overview:
Australian Medical Services Advisory Committee. (1-11-2006) Selective Dorsal Rhizotomy (SDR): Assessment for Nationally Funded Centre Status (A report by the Medical Services Advisory Committee to the Australian Health Ministers’ Advisory Council). 1-82.


NHS Quality Improvement Scotland (NHS QIS 2007)  
Evidence Note 16

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NHS Quality Improvement Scotland (NHS QIS), a special health board in Scotland, was established in 2003 and leads the use of knowledge to promote improvement in the quality of healthcare for the people of Scotland. It performs three key functions:

[1] providing advice and guidance on effective clinical practice including setting standards;

[2] driving and supporting implementation of improvements in quality;

[3] assessing the performance of the NHS, reporting and publishing the findings.

This clinical effectiveness summary was published as a result of the apparent increase in the incidence of deformational plagiocephaly (Aargenta et al 1996) to identify if the use of a cranial orthosis is an effective treatment option. The summary gives a brief overview of the orthotic device (aka helmet) and thereafter a description of the aetiology of infant deformational plagiocephaly.

NHS QIS provides an overview of their literature review prior to adopting the results of a systematic review conducted in 2005 (ECRI 2005). This identified six randomised controlled trials, comparing a cranial orthosis to comparator treatments or no treatment. The RCTs were found to be either not methodologically robust or else the quality of the outcome data was too low to be reliable (Clarren SK 1981, Mulliken et al 1999, Graham et al 2005a, Graham et al 2005b, Loveday BP& de Chalain TB 2001, Vles JS et al 2000).

It was therefore impossible to provide any evidence-based conclusions upon the effectiveness of a cranial orthosis. Economic implications for the provision of these devices were considered. Again there was not enough evidence to give guidance and the current provision has been agreed at a Health Board Level. A safety note was highlighted in the evidence note (NHS QIS) re. adverse incidents including skin irritation, infant distress and embarrassment of care givers. Physiotherapists must be aware of these when referring for orthotic provision.

In summary, NHS QIS were unable to find any evidence to support or disprove the effectiveness of a cranial orthosis, nor evidence of further research. The use of cranial orthosis is therefore likely to be on an individual and health board basis.

References


Submissions to the APCP Journal

The APCP Journal aims to disseminate original research, facilitate continuing medical education and to provide an opportunity to debate controversial issues in paediatric physiotherapy. Listed below are the different types of submissions that will be considered for publication in the APCP Journal with guidance on how to write and format your work to maximise the chances of your submission being successful. The Editorial Board also welcome pre-submission questions and will be happy to advise further. Please contact eva.bower@tinyworld.co.uk in the first instance for guidance or pre-submission advice.

Original Research Reports

Original research in many formats, including quantitative and qualitative research, case series and case reports. These articles should be 4500 words or less, excluding the references and abstract. All submissions in this category will be subject to anonymous peer review by 2 reviewers.

Research Papers

Research papers should generally follow the ‘IMRD’ pattern (Introduction, Methods, Results and Discussion). When writing your paper, it is usually best to start with the most important parts, the methods and results, before writing the discussion, introduction and conclusion.

The methods section should include sufficient information to enable other clinicians to reproduce your work. Any work involving human or animal subjects must have appropriate ethical approval from the relevant Research Ethics Committee. In addition, written permission from children, parents or guardians to publish photographs of individuals must be submitted. The methods section should also include details of statistical methods if they are used and state which software, if any, was used to obtain the results. Any apparatus used in the study should be reported in terms of manufacturer and location (city, county, country).

The results section should be clear and easily understood. Rather than presenting the reader with masses of data, it often helps to construct your results to tell a story, taking the reader step by step through your findings. Do not present data twice in both text and tables/figures, and do not include material that belongs in the discussion, i.e. present results only, not interpretation. Consider how statistical data is presented, ensure that descriptive and inferential statistics are used appropriately to provide meaning to the data collected.

Tables and figures should be numbered consecutively as they are referred to in the text, and placed after the references on a new sheet. Abbreviations should be explained in a footnote and only horizontal lines should be used. Table and figure captions/legends should be included on a separate sheet.

The discussion will allow you to succinctly summarise the major findings of your work and explain its relevance in terms of the available literature and current practice. It is helpful if the first paragraph briefly summarises the major findings. The discussion will also allow you to address any potential weaknesses in the methodology and justify why the research was performed in a particular way. It is important to keep the discussion relevant to the results obtained.

Ideally the introduction should be short and engage the reader, explaining why the paper is relevant to clinical practice. Often a brief summary of the existing literature highlighting the need for this particular research is useful, as it leads directly to the research question being asked.

Finally the title and abstract can be written. The abstract should be structured (limited to 300 words) consisting of ‘Background and Purpose’ (why the research done), ‘Method’ (what was done), ‘Results’ (what was found) and ‘Conclusion’ (what was concluded). The title itself should describe the contents of the paper succinctly and accurately.

Scholarly Papers

Discursive papers sharing ideas or experiences in specific areas of practice can be structured more freely but should still include an ‘Introduction’, ‘Discussion’, and ‘Conclusions’. Scholarly papers should be no longer than 4500 words.

Case Studies and Case Series

The format for case studies and case series differs from that given above and should start with an ‘Introduction’, followed by ‘Case Report’ (history, investigations, treatments, outcome), ‘Discussion’ and ‘Conclusion’. Case reports may be notable because they either focus on a rare condition or on a new method of treatment. The use of false names in case reports is encouraged but if a child is recognisable in the report (due to the condition or the specific nature of the treatment given), then written consent for publication should be obtained. Case reports should be no longer than 2500 words.
Audit Reports
Reports of clinical audit should include an ‘Introduction’, ‘Standard Setting’ (with appropriate reference to the available literature), ‘Method’, ‘Results’, ‘Discussion’, and ‘Conclusion’. These should be no longer than 3000 words.

Review Papers
Systematic reviews undertake specific methodology and focus on a specific question, perform a thorough literature search and critical appraisal of individual studies using strict criteria. Less formal review articles will summarise the current literature on a particular topic. The Cochrane Collaboration has published a handbook on conducting systematic reviews (http://www.cochrane-handbook.org/) and you should structure your review in terms of ‘Introduction’, ‘Objectives’, ‘Methods’, ‘Results’, ‘Discussion’, and ‘Conclusion’. There are published criteria that should be applied to the analysis of randomised controlled trials: the Delphi criteria (http://www.ncbi.nlm.nih.gov/pubmed/10086815) and the PEDro scale (http://www.pedro.org.au/scale_item.html). The MOOSE guidelines should be applied to the analysis of observational studies (http://www.consortstatement.org/mod_product/uploads/MOOSE%20Statement%202000.pdf). Review papers should be no more than 4500 words.

Technical Evaluation
Technical evaluations describe mechanical or technical devices used in clinical practice or education and should include an introduction, methods including the specifications of the equipment used and the means of the evaluation, the evaluation outcome, discussion and conclusions. Technical evaluations should be no longer than 2500 words.

Service Development Report
A service development report should describe changes in service delivery/management. The structure of the report can be less formal but should include an introduction, description of the service change(s), outcome and discussion on the implications for future practice. Service development reports should be no longer than 2500 words.

Abstracts of Theses and Dissertations
Abstracts of research projects, audits and presentations from undergraduate and postgraduate degrees should be no more than 300 words in length and structured as a standard abstract (introduction, methods, results, conclusion). However, the Editorial Board would strongly encourage those considering such a submission to formulate their work instead as one of the above peer reviewed articles. In such cases, a pre-submission enquiry to the editor may be helpful.

Other types of editorial material
The Journal will also consider the following submissions:

Letters to the editor
Letters to the editor can be on any issue pertinent to paediatric physiotherapy or to APCP. Letters should be no more than 500 words long.

Book reviews
Book reviews should be no more than 750 words long.

Referencing
All work submitted for peer review should be referenced in the Harvard style:

In text, cite only the author(s) surname(s) followed by the date of publication, e.g. (Robinson, 1994) or Robinson (1994). ‘a’, ‘b’, etc., is used to indicate more than one publication by the same author(s) in the same year, e.g. (1992a,b). For three or more authors of a cited paper, name the first followed by et al, e.g. (Smith et al, 1990).

In the reference list, include articles in journals and books alphabetically by author. For citations from journals, give the names and initials of all authors (year of publication), title of the article, full name of the journal, volume number, issue number and first and last page numbers, e.g. Brown A, Green B and Gold C (2001). ‘The value of exercise’, Physiotherapy, 87, 1, 77-79. Referring to books, give the names and initials of all authors/editors (year of publication), title, publisher, place of publication, and the chapter number or the page number of the citation or both, e.g. Gardner, M (2001). The Annotated Alice, Penguin Books, Harmondsworth, Chap 10, page 210.

The submission process
Email your submission in the first place in Word format to va@apcp.org.uk, clearly indicating the nature of the submission e.g. case series, research project.
Ensure that your copy includes information about the author/s (full name, qualifications, email address).

Once your submission has been received you will receive feedback from the reviewers indicating whether the copy has been accepted for publication.

There are a number of levels of acceptance:
• acceptance: no amendments required;
• acceptance: minor amendments required;
• acceptance major amendments required;
• rejection: not suitable for publication in the APCP Journal.
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JOURNAL

Vol 2 No 3 November 2011

www.apcp.org.uk