JAN RESEARCH PROTOCOL

# Lifestyle limitations of children and young people with severe cerebral palsy: a population study protocol

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## Abstract

**Title. Lifestyle limitations of children and young people with severe cerebral palsy: a population study protocol**

**Aim.** This paper is a presentation of a study protocol to establish the prevalence of orthopaedic problems (hip dislocation, pelvic obliquity, spinal deformity and contractures) and their impact on pain, function, participation and health in a population of children and young people with severe cerebral palsy.

**Background.** Cerebral palsy is the commonest cause of motor impairment in childhood and is associated with life-long disability. An estimated 30% of people with cerebral palsy have severe forms and are non-ambulant. Although the underlying neurological damage is not amenable to correction, many health services are dedicated to providing therapeutic and adaptive support to help people with the condition reach their potential.

**Method.** A cross-sectional survey of children and young people, aged 4–25 years with severe, non-ambulant cerebral palsy as defined using the Gross Motor Function Classification System (Levels IV and V). Study participants will be identified from a pre-existing, geographically defined case register and recruited via a healthcare professional known to them. Two assessments will be undertaken: one involving parents/carers at home and using questionnaires; the other involving the child/young person ideally in one of three settings and including X-rays if clinically indicated.

**Discussion.** This study will contribute to our knowledge of the history and epidemiology of orthopaedic problems in children and young people with cerebral palsy and how these problems accumulate and impact on participation, health and well-being. The study will also identify unmet need and make recommendations for good practice in relation to the orthopaedic care and management for people with severe cerebral palsy.

**Keywords:** cerebral palsy, child nursing, disability, epidemiology, orthopaedics, pain, study protocol

## Introduction

Cerebral palsy (CP) is a disorder of voluntary movement and posture caused by damage to the developing brain and affects 2–2·5 per 1000 of the population. CP is the commonest cause of motor impairment in childhood and associated with life-long disability (Stanley *et al.* 2000). It is caused by damage to the developing brain sometime before, during or soon after birth. While the damage to the brain is static, the clinical manifestation of the condition can change over time and lead to the development of muscle contractures, bony deformities and pain. Although the underlying neurological damage is not amenable to correction, there is a major expenditure of resources within the health service providing therapeutic and adaptive support to help people with CP to fulfil their potential. Finally, very little is known about the health needs and services received by the older population with CP after they have moved from paediatric to adult services.

The revised International Classification of Functioning, Disability and Health or ICF (World Health Organization 2001) provides a useful conceptual framework and important context for this study. The term 'disability' is now used as an umbrella term to encompass impairments, activity limitations (previously called 'disability') and participation restrictions (previously called 'handicap'). Activity limitations refer to difficulties in the activities of daily living and participation restrictions to limitations in life situations. In this study, we are investigating the relationship between impairments (neurological and orthopaedic), activity limitations (in functional abilities) and participation restrictions (affecting education, social life and recreation among other things) in children and young people with severe CP. In addition, we will also study health status and health service use considered here under 'personal factors' in the ICF model and relating to the particular background of an individual's life which may play a role in the person's experience of disability (World Health Organization 2001). There is an emerging literature on the relevance of the ICF to nursing practice, education and research (Kearney & Pryor 2004, Heinen *et al.* 2005, Parkes & Clarke 2006). In particular, the ICF provides a conceptual framework to link 'patient problems' (or impairments), to outcomes and social factors which may affect their recovery, rehabilitation and participation. Kearney and Pryor (2004) illustrate the utility of the ICF specifically in relation to teaching students about the needs of people with CP (p.166).

This study will make recommendations for nursing practice in relation to orthopaedic care and management of people with severe CP. As this study is based on a representative case series of people with CP, we anticipate that the findings will have relevance internationally to nurses and other practitioners working in the area.

## Background

There have been few attempts to clarify the clinical and functional progression of children and adolescents with CP using a population-based approach. Other studies (Drummond *et al.* 1979, Moreau *et al.* 1979, McCarthy *et al.* 1988, Bagg *et al.* 1993) have relied on clinic or institution-based populations and are likely to have been biased in terms of the clinical profiles of children included (see Parkes *et al.* 2006). For the purposes of this study, we have defined the severity of CP according to the Gross Motor Function Classification System (GMFCS) (Palisano *et al.* 1997), a standardized, age-specific, five level system ranging from Level I (describing the functionally most able children) to Level V (the least able). For this study, 'severe CP' was defined as Levels IV and V of the GMFCS – the two most severe categories of motor function and accounting for approximately 30% of cases in population-based studies (Beckung & Hagberg 2002, Parkes *et al.* 2005).

The incidence of hip dislocation, pelvic obliquity and scoliosis in CP is related to severity of motor involvement (Cooperman *et al.* 1987), but earlier reports vary and are uncertain. For example, hip subluxation or dislocation has been estimated to occur from 2.6% to 35% of the population with quadriplegic or bilateral spastic CP (Cooperman *et al.* 1987, Widmann *et al.* 1999, Scrutton *et al.* 2001, Morton *et al.* 2006); and scoliosis from 26% (Samilson 1981, Thomson & Banta 2001) to 64% (Comstock *et al.* 1998) although it is known that the presence of these complications varies by age. Pain as a direct consequence of hip instability has been estimated to occur in 70% of cases (Widmann *et al.* 1999, Hodgkinson *et al.* 2001), but the reliability of pain report among those with cognitive and communication impairments is difficult to ascertain. The existence of these problems is likely to lead to increasing strain on the child and caregiver as positioning, seating, transferring and maintaining hygiene and comfort become increasingly difficult and impact negatively on quality of life, health and well-being. Previous studies have not investigated the relationship between orthopaedic complications and lifestyle factors in a population series.

### Orthopaedic problems in CP

Hip abnormalities in CP rank second only to heel cord tightness (Kalen & Bleck 1985). The hip is considered subluxed when more than one-third of the femoral head is

uncovered by the acetabulum; and dislocated when the femoral head loses contact with the acetabulum (Moreau *et al.* 1979, Kalen & Bleck 1985, Lonstein & Beck 1986, Bagg *et al.* 1993). Hip dislocation is most common in those with spasticity; in those with non-ambulant CP compared to ambulant forms (25% vs. 4%) (Drummond *et al.* 1979) and in children aged around 7–9 years (Samilson *et al.* 1972, Letts *et al.* 1984, Hoffer 1986, Lonstein & Beck 1986) with 39% of those in GMFCS Levels IV and V having a dislocated hip by 15 years (Morton *et al.* 2006). In a prospective study of the development of hip problems in children with CP (Scrutton *et al.* 2001), inability to walk at age five was associated with a 46% likelihood that hips would require treatment in the future. The causes of hip instability are multifactorial, but have been hypothesized to include muscle imbalance causing the centre of rotation of the femoral head to be forced out of the acetabulum (Hoffer 1986); the presence of strong extensor tone and the existence of primitive reflexes which can result in the hips being 'wind-swept' with one hip adducted and likely to dislocate (Samilson *et al.* 1972, Sherk *et al.* 1983); and damage to the lateral margin of the acetabulum caused by pathologic tone in the surrounding musculature (McCarthy *et al.* 1988, Heinrich *et al.* 1991).

Pelvic obliquity can be caused by problems related to the hip (infra-pelvic phenomena) and/or the spine (suprapelvic phenomena) and is associated with increasing age in the CP population (Terjensen *et al.* 2000). The sequelae of persistent fixed pelvic obliquity include poor pressure distribution over the ischial tuberosities with an increased risk of ulceration (Drummond *et al.* 1979). Unfortunately, there is little agreement in the literature about the definition of pelvic obliquity making its true prevalence uncertain and comparisons between studies limited. If the cause of pelvic obliquity is assumed to be infra-pelvic, then pelvic obliquity has been assessed by considering the relationship of the pelvic ilia to the horizontal (Lonstein & Beck 1986, Cooperman *et al.* 1987, Terjensen *et al.* 2000). If the obliquity is considered suprapelvic, then the relationship of the pelvic ilia to the spine should be considered (Drummond *et al.* 1979, Moloney *et al.* 1990, Dias *et al.* 1996). Neither of these methods has been the subject of reliability testing and this will form part of this investigation. However, there is no certainty as to which measurement should be used in any given clinical situation.

Scoliosis, like hip dislocation, is more often associated with spasticity than dyskinetic forms of CP and is more common in non-ambulant forms of CP than ambulant (39% vs. 7%) (Drummond *et al.* 1979, Cooperman *et al.* 1987). There are some contradictory reports about age of onset (5–26 years), but early onset is associated with a poorer prognosis (Thometz & Sheldon 2001). The aetiology of scoliosis has been hypothesized to include the absence of righting and equilibrium reactions commonly seen in CP, which may contribute to the development of curvature as may retention of primitive reflexes (e.g. Galant reflex) (Samilson *et al.* 1972). Sensory defects including ophthalmic defects have also been implicated (Drummond *et al.* 1979). The most common form of scoliosis seen in CP is the long 'C' curve that extends down to the pelvis. It is progressive and frequently becomes rigid in adolescence. It is these curves that are most often associated with fixed pelvic obliquity and dislocation (Samilson *et al.* 1972). In particular, curves $>50°$ at skeletal maturity are more vulnerable to progression and at twice the rate of curves $<50°$ (Thometz & Sheldon 2001). While it is acknowledged that there is a relationship between hip instability, scoliosis and pelvic obliquity (Letts *et al.* 1984), the exact nature of this is unknown.

## The study

### Research objectives

The study has two primary objectives: firstly, to establish the prevalence of orthopaedic problems (hip dislocation, pelvic obliquity, spinal deformity, pain, history of fractures and contractures) in a population of children and young people with severe CP; and secondly, to investigate the relationship between the severity of CP (including CP subtype and the presence and severity of orthopaedic problems), functional ability, caregiver assistance, participation and health status in a geographically defined population of children with severe CP.

Secondary research objectives include establishing baseline data for future research, which would assess these children and young people over time. More specifically, this would make it possible to monitor the natural progression of the condition and assess the effect of any changes in orthopaedic management. A study into the locomotor abilities and health status of the population of children with CP in GMFCS Levels I–III is also underway (Cosgrove *et al.*). By combining this study and LiLaC would lead to the first-ever population-based study of functional abilities in children and young people with CP recruited from a geographically defined case register.

### Design

This is a descriptive, cross-sectional study involving both a community- and hospital-based, clinical assessment of children and young people presenting with severe forms of

 

CP who are resident in the UK. A geographically defined case register [the Northern Ireland Cerebral Palsy Register (or NICPR), the method of which is described elsewhere (Parkes *et al.* 2001], will be used to identify the population. The NICPR is considered a highly ascertained source of information about people CP and the prevalence of the condition in Northern Ireland is 2.2 per 1000 livebirths (95% CI: 2.07–2.34) (Parkes *et al.* 2005).

*Study hypotheses*

The following null hypotheses will be investigated:

- There is no correlation between the presence of hip migration, the extent of hip contracture, pelvic obliquity, pain and the presence of spinal deformity (scoliosis, lordosis and kyphosis).
- There is no correlation between the retention of primitive reflexes and spinal deformity (scoliosis, lordosis and kyphosis).
- There is no correlation between orthopaedic problems, pain, functional ability, participation, health status and service use.
- There is no correlation between upper limb function, child health status and caregiver assistance.

*Definitions*

The definition and classification of CP used here is consistent with that published by the Surveillance of Cerebral Palsy in Europe project (SCPE 2000). There are three CP subtypes including spastic, dyskinetic and ataxic. Spastic subtypes are further divided into unilateral (affecting one side of the body) and bilateral (affecting two sides of the body). Intellectual impairment is considered present where the IQ is $< 70$; and considered severe where the IQ is $< 50$. Visual impairment is considered severe or profound where visual acuity was $< 6/60$ in the better eye. Hearing impairment is considered severe or profound where there was a $> 70$ dB loss without the use of hearing aids. Quintile of deprivation was established using the Noble Index deprivation score (Noble *et al.* 2005), which was assigned to each case on the NICPR based on their electoral ward of residence (identified from postcode). The data were then sorted by deprivation score and divided into fifths ranging from the most affluent quintile (1) to the most deprived quintile (5) of child population.

*Timescale*

This is a 4-year study that commenced in September 2006. Recruitment and data collection will start in summer 2007, data analysis will in 2008/09 and reporting and dissemination will be completed in 2010.

**Participants**

This study will include children and young people with CP from the birth cohort 1st September 1981 to 1st September 2002 (4–25 years by 1st September 2007) with all clinical subtypes of CP, but restricted to those in Levels IV and V of the GMFCS. Potential participants will be excluded if they have had orthopaedic surgery in the last 6 months; Botulinum toxin therapy in the last 3 months; those too unwell to participate (although every effort will be made to accommodate these participants at a later date). Children who have late onset CP, i.e. acquired after the neonatal period but before their fifth birthday, will also be included although it is recognized that this group is clinically distinct and sometimes atypical.

*Sample size*

This is a single group prevalence study about a known population and thus there is no requirement for a formal power calculation. However, as aspects of the study will examine associations and correlations, we assumed that if the correlation coefficient ($r$) is of the order of 0.28 then to detect a correlation at the 5% significance level with the power of 90% would require a sample of 130 participants (Bland & Altman 1996). Figure 1 shows that, after excluding children and young people who are known to have died or moved out of the area, there will be 1245 people aged 4–25 years by 1st September 2007. Of these, sufficient data are available to ascertain that 251 are eligible to take part in the LiLaC Study. A further 189 participants will be followed up to check their eligibility. Allowing for an attrition rate of 40%, particularly among the adults, would lead to a possible sample size of 151 out of 251 participants.

*Identification, approach and recruitment of participants*

As well as identification of potential participants via NICPR, additional checks and searches will be carried out via the orthopaedic surgeons working within the regional orthopaedic service. This will ensure complete case coverage. The characteristics of the 251 children and young people currently eligible to take part are shown in Table 1.

Initially, all families, young adults and/or their carers will be approached by a healthcare professional known to them. This will be carried out using an information pack containing all the relevant information sheets (parent/child/young adult and/or carer) and seeking their permission for the release of their contact details to the research team. Where permitted, a direct approach by the researcher (first author) will be made to obtain informed consent to participate in the study. It will be explained to families that they may wish to opt out of the

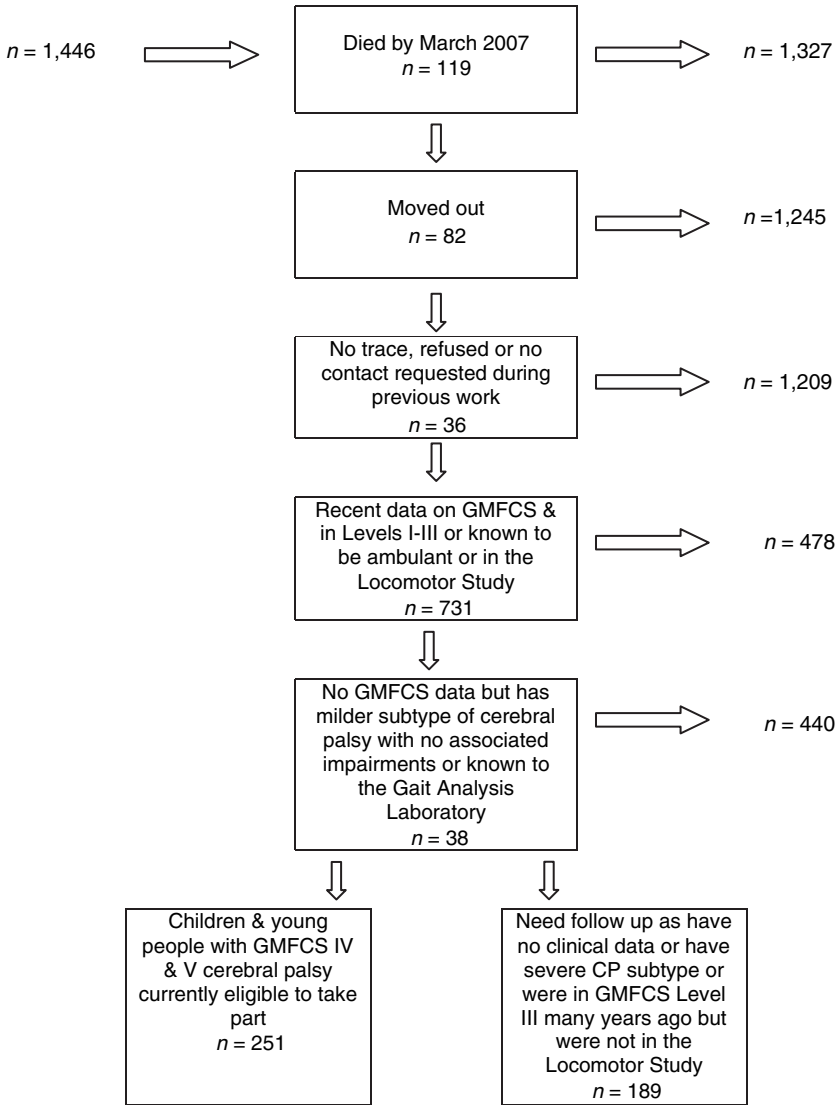


**Figure 1** The Northern Ireland Cerebral Palsy Register sampling frame of children and young people with cerebral palsy born 1/9/81 to 1/9/02.

study at any stage. The informed consent process will involve a personal visit ensuring families have had the opportunity to read the information sheet and consider their position. Families will also be given an opportunity to ask questions before completing the consent forms. Copies of the information sheet and consent forms are available on request from the first author.

Informed consent will be sought from all parents/carers for children under the age of 16 years and where possible, from the children themselves (informed assent). In the case of young people aged 16 years or over who are unable to give informed consent because of cognitive impairments process consent will be sought from parents/carers. This involves parents/carers giving a detailed explanation to the young person that has regard for their level of comprehension. The carer will be asked to sign a form to say that they have been consulted and agree for the young person to be included in the study.

## Data collection

*Measures*

Summaries of the published psychometric measures that will be included in this study are given in Table 2.

*Orthopaedic assessment.* A physical examination of the spine, hips, knees and ankles will be carried out using the Spinal Alignment and Range of Motion Measure (SA-ROMM), a measurement tool designed to detect the presence or absence of deformity at all these levels (Barlett & Purdie 2005). Additional checks/X-rays will be carried out as follows.

 

**Table 1** Characteristics of the children and young people eligible to take part in the study ($n$ = 251) (percentages may not sum to 100 because of rounding)

| Characteristics | *n* (%) |
|---|---|
| *Age (years) on 1/9/07* | |
| 4–9 | 44 (17) |
| 10–14 | 52 (21) |
| 15–19 | 73 (29) |
| 20–25 | 82 (33) |
| *Age at assessment for NICPR* | |
| < 3 | 12 (5) |
| 3–5 | 16 (6) |
| 5–9 | 174 (69) |
| 10–14 | 44 (17) |
| 15–19 | 5 (2) |
| *Sex* | |
| Male | 150 (60) |
| Female | 101 (40) |
| *Clinical subtype of cerebral palsy* | |
| Unilateral spastic | 12 (5) |
| Bilateral spastic | 214 (85) |
| Dyskinetic | 19 (8) |
| Ataxic | 3 (1) |
| Unclassifiable | 3 (1) |
| *GMFCS Levels* | |
| Level IV | 68 (27) |
| Level V | 176 (70) |
| Level IV or V | 7 (3) |
| *Upper limb function* | |
| Normal | 12 (5) |
| Some restrictions | 10 (4) |
| Moderate | 72 (29) |
| Severe | 142 (57) |
| Missing | 15 (6) |
| *Intellectual impairment* | |
| IQ 70+ | 35 (14) |
| IQ 50–70 | 40 (16) |
| IQ < 50 | 154 (62) |
| Uncertain | 21 (8) |
| Missing | 1 (< 1) |
| *Visual impairment* | |
| None, mild, moderate | 183 (73) |
| Severe, no vision | 53 (21) |
| Uncertain | 11 (4) |
| Missing | 3 (1) |
| *Hearing impairment* | |
| None, mild, moderate | 224 (89) |
| Severe, no hearing | 9 (4) |
| Uncertain | 13 (5) |
| Missing | 5 (2) |
| *Seizures* | |
| None (ever) | 88 (35) |
| None last 12 months | 50 (20) |
| Seizures (last 12 months) | 107 (43) |
| Uncertain | 3 (1) |
| Missing | 3 (1) |
| Deprivation quintile | |
| 1 (most affluent) | 20 (8) |
| 2 | 20 (8) |
| 3 | 22 (9) |
| 4 | 50 (20) |
| 5 (most deprived) | 132 (53) |
| Missing | 7 (3) |

NICPR, Northern Ireland Cerebral Palsy Register.
GMFCS, Gross Motor Function Classification System.

*Hips* Each participant will be screened by X-ray for hip migration if clinically justified i.e. if no standardized X-ray has been taken in the previous 6 months. For ethical reasons, recruitment and subsequent assessment will be timed to coincide with routine X-ray examination. X-rays will be standardized and measured using a modified light box as recommended elsewhere (Scrutton & Baird 2001). Hip X-rays will be assessed using Reimer's migration index (Reimers 1980, Moreau *et al.* 1979, Sherk *et al.* 1983, Cooke *et al.* 1989, Heinrich *et al.* 1991; Scrutton *et al.* 2001) and the acetabular index (Lonstein & Beck 1986, Cooperman *et al.* 1987, Comstock *et al.* 1998, Terjensen *et al.* 2000, Scrutton *et al.* 2001) both of which can be measured reliably (Parrot *et al.* 2002).

*Spine* All participants will be referred to a Spinal Orthopaedic Surgeon and if scoliosis, lordosis or kyphosis deformity is suspected and where no standardized X-ray has been taken in the previous 6 months, participants will be referred for coronal and sagittal plane X-ray as appropriate. Scoliosis and sagittal plane spinal deformity will be assessed using Cobb angles (Cobb 1948), the accepted gold standard for the radiological definition of scoliosis (Herring 2002).

*Pelvis* Pelvic obliquity will be assessed clinically while sitting and radiologically using the standardized pelvic anterior-posterior X-ray taken for hip measurements. A protocol for the assessment of pelvic obliquity will be developed in the first 6 months of the project.

*Pain assessment.* The paediatric pain profile (PPP) will be used for assessing pain and is a valid and reliable instrument developed for children with communication difficulties who are unable to self-report (Hunt *et al.* 2004). It will also be used with adults who are unable to report and its validity reported on. For those able to self-report, two health status

**Table 2** Outcome measures and instruments

| Instrument | Reference | Measures | Domains | Type of variable | Validity and reliability reported |
|---|---|---|---|---|---|
| *Impairments* | | | | | |
| Spinal Alignment and Range of Motion Measure (SAROMM) | Barlett & Purdie 2005 | Spinal deformity and range of motion of joints | Two parts: four items for spinal alignment; 11 items for range of motion | Primary outcome; dependent variable (ordinal) | Yes |
| Cobb angle | Cobb 1948 | A calculation of the angle of deformity of the spine | – | Primary outcome; independent variable (continuous) | Yes |
| Reimers' Migration Index and Acetabular Index | Reimers 1980 | Measures the extent of subluxation of the hip | – | Primary outcome; independent variable (continuous) | Yes |
| Goniometry | Norkin *et al.* 2003 | Measurement of joint motion | – | Primary outcome; independent variable (continuous) | Yes |
| Radiographic measures | Abel *et al.* 1999 | To measure pelvic obliquity both infra pelvic and supra pelvic | – | Primary outcome; independent variable (continuous) | No |
| Neurological deficits | Evans & Alberman (1985) | Standardized recording form for neurological deficits of central origin | Motor and associated impairments (cerebral palsy subtype) | Secondary Northern Ireland Cerebral Palsy Register data; independent variables (ordinal/ categorical) | No |
| Paediatric Pain Profile (PPP) | Hunt *et al.* 2004 | Pain behaviours and pain history | 20 item behaviour rating scale | Primary outcome; independent variable (ordinal) | Yes |
| Braden Scale | Braden *et al.* 2005 | Pressure sore risk assessment tool | Six item scale assessing sensory perception, moisture, activity, mobility, nutrition, friction and shear | Secondary outcome; independent variable (ordinal) | Yes |
| *Activity limitations* | | | | | |
| Gross Motor Function Classification System (GMFCS) | Palisano *et al.* 1997 | Gross motor function | Five level classification system ranging from Level I (most able) to Level V (least able) | Classification system; independent variable (ordinal) | Yes |
| Manual Ability Classification System (MACS) | Eliasson *et al.* 2006 | Classification of manual ability and dexterity | Five level classification ranging from Level I (most able) to Level V (least able) | Classification system; independent variable (ordinal) | Yes |
| Pediatric Evaluation Disability Inventory (PEDI) | Haley *et al.* 1992 | Measure of general functional ability including caregiver assistance | Functional skills, caregiver assistance and modifications in self care, mobility and social function | Primary outcome; independent variable (ordinal) | Yes |

 

**Table 2** (*Continued*)

| Instrument | Reference | Measures | Domains | Type of variable | Validity and reliability reported |
|---|---|---|---|---|---|
| *Participation restrictions* | | | | | |
| Life-Habits Questionnaire (Life-H) | Fougeyrollas *et al.* (1998) | Quality of social participation of individuals with disabilities | 64 items communication, personal care around the house, feeding, mobility fitness, responsibility, recreation, education, community life and interpersonal work. | Primary outcome; dependent variable (ordinal). | Yes |
| Personal factors (including health status) | | | | | |
| Child Health Questionnaire (CHQ) (two items only) | Landgraf *et al.* 1998 | Health status and well-being of children and adolescents | two items scales – global health, pain. | Primary outcome; independent variable (categorical + ordinal). | Yes |
| Measure of Process of Care (MPOC) | King *et al.* (1995); McConachie & Logan (2003) | Family Centredness of Services | 20 items 5 scales - enabling and partnership, providing general information, providing specific information about the child, co-ordinated and comprehensive care, respectful and supportive care. | Primary outcome (categorical). | Yes |
| Background proforma | Developed within the study | Descriptive | Reflexes, medical history, hospital admission, schooling, aids and adaptations, services, expectations of surgery. | Secondary outcome; independent variables (categorical). | No |

measures described below include questions about bodily pain for those able to self-report.

*Functional assessment.* The Paediatric Evaluation of Disability Inventory (PEDI) is an interview style questionnaire which will assess general functional ability and caregiver assistance and is a valid and reliable measure (Nichols & Case-Smith 1996). Upper limb function will be classified using the Manual Assessment Classification System (MACS) (Eliasson *et al.* 2006) and lower limb function using the GMFCS (also being used as a screening tool). While these classification systems have primarily been designed for children, this study will make a contribution to the ongoing work on with their use with teenagers and young adults.

*Participation and care health.* The LIFE–Habits Questionnaire (Life-H) (Fougeyrollas *et al.* 1998) will be used to assess the quality of social participation by assessing the participants' performance in daily activities and social roles (life habits). Two items from the Child Health Questionnaire (CHQ-PF50) will be used to assess global health and pain and is a parent or carer, and completed measure appropriate for use with children aged 4–18 years and has evidence of

internal consistency (Langraf & Abetz 1997) and concurrent validity (Landgraf *et al.* 1998). The measure of Process of Care (MPOC; King *et al.* 1995) is a questionnaire developed in Canada to assess the family-centredness of service delivery. It has been modified for use in the UK (McConachie and Logan 2003). Other information on service use will also be collected (see below).

*Other assessments.* Braden Scale will be used to assess and document pressure sore areas. A data extraction proforma has now been developed and will be used for collating information on associated impairments; schooling; services; aids for living, current/past medical and surgical history, and medication.

*Data collection*

Two assessments will be undertaken by the researcher. The first will be an interview with parents/carers. As part of the interview questionnaires assessing pain, function, participation and health will be completed in a systematic way and according to a standardized protocol. The second assessment will be an orthopaedic assessment of the child/young person conducted at one of three possible places (hospital, school or home), depending on their preference. The researcher will try and coincide this with a hospital visit to minimize the impact of the research on the participant's home or school life. The orthopaedic and pressure area assessment will be carried out at this stage. When clinically justified, the child/young person may require X-rays of their spine and pelvis. Each orthopaedic measure will have a protocol to standardize the positioning of participants for X-ray.

*Training for data collection*

In relation to the orthopaedic assessment, the researcher is currently receiving clinical training in identifying the spinal, pelvic and hip deformity and in the range of motion measurements necessary for the SAROMM. This is being provided by specialist orthopaedic surgeons and senior physiotherapists based within a National Health Services Trust. Assessment of manual ability has involved training with the Occupational Therapy Department although in practice it will require information from carers. To check the consistency of technique, interpretation and report of the radiological measurements taken by the researcher (when compared with the Consultant Orthopaedic Surgeon), several sub-studies of inter-rater reliability will be carried out on approximately 20 X-rays. A sub-study of intra- and inter-rater reliability will also be undertaken with regard to pelvic obliquity. Some preliminary work will be undertaken with a small number of parents and young people to pilot the other instruments and check the interview protocol.

### Ethical considerations

The study has been approved by the Office of Research Ethics Northern Ireland; REC reference 06/NIR02/137. Information leaflets have been designed (and approved by the above Committee) for (i) parents/carers, (ii) children under the age of 16 years and (ii) young adults. Where a potential participant has significant cognitive impairments, the parent/carer will read the information to them in a manner consistent with their level of comprehension. As part of the informed consent process, it will be explained to potential participants that taking part in the study is voluntary, that they have the right to refuse or opt out at any time and without giving a reason (and that this will not affect the services they receive), that their information will be treated as confidential and stored according to the principles of the Data Protection Act (1998), and finally that they will never be identified in any part of the dissemination process. This study will offer some direct benefits to participants as all participants will be screened for orthopaedic problems and offered a referral to an orthopaedic spinal surgeon.

### Data analysis

Causes of attrition might include non-response, non-traceability, recently deceased, moved away or refusal to take part and will be noted and an analysis at the end of the study will determine any differences between the recruited sample and the total eligible population. The use of a comprehensive case register makes this feasible.

The main hypotheses will be tested using correlation and regression (for continuous variables); independent *t*-tests (for continuous by dichotomous variables); contingency tables and chi-square (for dichotomous variables only) and some non-parametric tests such as the Spearman's rank coefficient where the relationship between ranked or ordinal variables is being investigated. Adjustments for covariates will be made.

## Discussion

The publication of study protocols has been traditional practice in systematic reviews and randomized-controlled trials but not epidemiological or observational research until more recently (Colver and the Sparcle Group 2006). There are certain benefits to publishing a study protocol which includes the specification of the hypotheses for testing

 

*a priori*. This helps avoid the known pitfalls of fishing in the data and *post hoc* analyses giving more confidence in the conclusions drawn from the study (Colver and the Sparcle Group 2006). A secondary benefit is early dissemination of the research and this is useful for clinicians working in the field, service planners, policy makers, lay users as well as other researchers with whom it may be possible to form subsequent collaborations.

This protocol is an outline of a study into the orthopaedic and health-related problems of a representative case series of people with severe CP including young adults who have seldom been followed up until now. Using a case register approach to the identification of potential participants offers a number of methodological strengths to the research but is an approach seldom used by nurse researchers. A number of dedicated disease registers exist in the United Kingdom and Europe and include conditions like CP, cancer, diabetes and congenital malformations. Many registers have an established process whereby external researchers may apply to use the register as a sampling frame. In the case of the NICPR, researchers apply to the Advisory Committee and their application is also contingent upon ethical approval. This process never involves the release of identifiable patient information from the Register but rather, a system of contact with notifying clinicians is undertaken by the data manager on behalf of the researchers for a small fee. This process works relatively well and examples have been reported in the literature (Dickinson *et al.* 2006, McClelland *et al.* 2006, Parkes *et al.* 2006).

We are aware of the limitations of undertaking cross-sectional research. This includes the inability to make inferences about causation or the sequencing of events in relation to the orthopaedic problems under study. Furthermore, we will be unable to determine whether any differences in the health or orthopaedic status between children and young adults are due to age, the aetiology of their clinical condition or cohort effects. Nonetheless, this study will provide population-based information on the clinical status of the severe subgroup of people with CP for the first time and will enable further hypotheses to be developed in preparation for longitudinal work that will include study participants.

This study builds on a number of important developments in the management of people with CP. Firstly, in relation to function, two robust, standardized, classification systems now exist to describe the functional abilities of people with CP (the GMFCS and the MACS). These systems have been incorporated increasingly by researchers working in the field (Morris & Bartlett 2004), but their uptake and use in clinical practice is much slower. This may be partly because of problems of dissemination and training. Advantages of both of these systems include their ease of use including parent completion, their applicability across a range of age groups and in relation to the GMFCS some evidence to support its use as a prognostic tool in relation to gross motor function in later childhood (Wood & Rosenbaum 2000). If these classification systems were routinely incorporated into nursing, physiotherapy and medical assessment, they would help introduce a common language that parents could also understand when describing abilities and changes in abilities in CP. Secondly, in relation to pain, a relatively new measure has been introduced by Hunt *et al.* (2004), the PPP which is suited to children (and potentially young adults) with neurological deficits who are also unable to communicate. One of the advantages of this assessment is the systematic and comprehensive approach to making structured observations of a child's 'pain behaviours', which includes extensive input from the parents. The PPP can also form part of the patient's record and therefore provides useful, comparable baseline data regarding the child's behaviour during hospitalization.

Detailed assessment of the orthopaedic problems in people with severe CP is seldom undertaken outside of orthopaedic settings, yet these problems are likely to be clinically very significant in the person's lifetime. It is very important that careful observations should be made of joints, contractures, abnormal body postures to detect and act promptly to help prevent further complications. The clinical training described here demonstrates that there are a number of relatively simply, inexpensive techniques that are easily taught and managed by nurses at little cost to the person with CP. Some of the techniques described here may not be applicable to children with mild-to-moderate CP although they could be used in relation to people with other severe types of physical disability.

Finally, this study will highlight the importance of multi-disciplinary working in the management and research of people with severe CP. As nurses, physiotherapists, surgeons and physicians we share common goals in the desired outcomes of care which are ultimately to improve function, prevent deterioration and problems, promote health and well-being leading to improved participation and quality of life.

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## What is already known about this topic

- People with severe cerebral palsy are at increased risk of orthopaedic problems, pain and poorer health.
- The orthopaedic problems of hip dislocation, pelvic obliquity and spinal deformity are related to one another.
- People with severe physical disabilities have fewer opportunities to participate in everyday life situations.

## What this paper adds

- The prevalence of orthopaedic problems and pain in a representative case series of children and young people with severe cerebral palsy.
- The strength and nature of the relationship between orthopaedic problems (hip dislocation, pelvic obliquity and spinal deformity) and their relationship with function, pain, participation and health.
- The extent of participation among children and young people with severe cerebral palsy and how their participation varies according to the presence of impairments, functional ability and health.

EAT/3178/05) and supervised by the co-authors of this paper.

## Author contributions

BM, JP and CDu were responsible for the study conception and design and CD, JP, BM and CDu were responsible for the drafting of the manuscript. CD, JP, BM and CDu obtained funding. CD, JP, BM and CDu made critical revisions to the paper. JP and BM provided statistical expertise. JP, BM and CDu supervised the study.

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