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# Problematic clinical features of children and adults with cerebral palsy who use electric powered indoor/outdoor wheelchairs: A cross-sectional study

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**ABSTRACT**
This article aims to describe the clinical features of electric powered indoor/outdoor wheelchair (EPIOC) users with cerebral palsy (CP) that are problematic to optimal prescription and to explore comorbidities, features of CP, and conditions secondary to disability impacting on equipment provision for children and adults. The method is a cross-sectional study of EPIOC users (*n* = 102) with a primary diagnosis of CP. This is a retrospective review of electronic and case note records of EPIOC recipients attending a specialist wheelchair service in 2007–2008. Records were reviewed by a rehabilitation consultant. Data were extracted under three themes; demographic, diagnostic/clinical and wheelchair factors. There were 48 males mean age 27.5 (range 8–70, *SD* 13.9) years and 54 females, mean age 29.5 (range 7–68, *SD* 14.6) years with CP. Sixteen comorbidities, nine features of CP, and five features of disability influenced wheelchair prescription. Sixty-four users were provided with specialized seating (SS) and 47 with tilt-in-space (TIS) seats. Complex controls were provided to 16 users, 12 tray-mounted. The majority of users had both SS and TIS. Powered wheelchair prescription has important therapeutic roles in clinical management in addition to enhancing mobility, independence and participation. Clinical features such as spasticity and problematic pain appeared less well managed in adults than in children.

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**KEYWORDS**
adaptive seating; aging; cerebral palsy; comorbidity; powered wheelchairs

## Introduction

Cerebral palsy (CP) is an umbrella term for a group of long term non-progressive but often changing neurological conditions (Aisen et al., 2011) affecting approximately 2.0 per 1,000 live births in developed countries (Rosenbaum, 2014). It occurs due to brain damage either before, during, or after birth (Parkes, Donnelly, & Hill, 2001) and is thought to include a genetic component (Tollanes, Wilcox, Lie, & Moster, 2014). Most people with CP now have a normal life span (Aisen et al., 2011; Parkes et al., 2001) and the number of adults is increasing due to improved survival of premature infants and increased age in the general population (Haak, Lenski, Hidecker, Li, & Paneth, 2009; Zaffuto-Sforza, 2005).

Approximately one third of children with CP are non-ambulatory (Novak, Hines, Goldsmith, & Barclay, 2012) and 37% of adults with CP use powered wheelchairs (Andersson & Mattsson, 2001). The benefits offered by powered mobility such as enhanced independence (Davies, De Souza, & Frank, 2003) and increased participation in education, (Evans, Neophytou, De Souza, & Frank, 2007) employment, and social life (Zaffuto-Sforza, 2005) are well established. Even young children provided with powered mobility gain benefits in social, language, and play skills (Rosenbaum, 2003) while the benefits for adults are mainly the maintenance of independence and self-care (Zaffuto-Sforza, 2005).

For those with severely disabling CP, these benefits may be achieved by the additional use of specialized seating (SS) systems (adaptive seating) (Angsupaisal, Maathuis, & Hadders-Algra, 2015) which may incorporate enhanced features, e.g., tilt-in-space (TIS) (Dicianno et al., 2015). It has been proposed that the primary focus of provision of assistive technologies (ATs) is not only the maintenance of skeletal integrity, the therapeutic value of powered chairs (e.g., for pain relief; Dicianno et al., 2015) but also to improve functional ability and participation (Angsupaisal et al., 2015). A combination of powered mobility devices, enhanced wheelchair features, SS, and complex control systems may be needed to achieve these aims (Zupan & Jenko, 2012).

The systematic review of Novak et al. (2012) provided clinical recommendations for mobility assessment and provision in adolescents with CP. This review also identified a range of “co-occurring impairments, diseases and functional limitations” experienced by individuals with CP that impacted on their disability (Novak et al., 2012). These co-occurring clinical conditions are likely to change over time influencing the wellbeing of individuals and their rehabilitation needs (Novak et al., 2012). The long-term consequences of CP are further complicated by growth and aging (Kembhavi, Darrah, Payne, & Plesuk, 2011). CP is now considered a lifespan condition with associated health factors, e.g., musculoskeletal

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impairments, medical complications, speech impairments, pain, and fatigue (Kembhavi et al., 2011). It is unclear if these are part of the natural course of CP, a consequence of long-term disability, or unrelated comorbidity.

Comorbidity has been defined as “any distinct additional entity that has existed or may occur during the clinical course of a patient who has the index disease under study” (Feinstein, 1970). Complication is considered to be a condition that co-exists or follows the index condition (Valderas, Starfield, Sibbald, Salisbury, & Roland, 2009). A clear differentiation between comorbidities, consequences of long-term disablement, and clinical features associated with CP may be complex and imprecise as reported for multiple sclerosis (De Souza & Frank, 2015) and rare diseases (De Souza & Frank, 2016). For example, epilepsy in those with CP may be considered a “co-occurring disease” (Novak et al., 2012), a co-morbidity (Govender, Hepworth, Bagwandeen, & Chetty, 2015), or a common symptom of CP (Zenczak-Praga, Zgorzalewicz-Stachowiak, & Cesar, 2015). Consequently it has been suggested that these clinical factors may be considered collectively as additional clinical features (ACFs) (De Souza & Frank, 2015).

In the United Kingdom, the National Health Service (NHS) provides electric powered indoor/outdoor wheelchairs (EPIOCs) to people with severe/complex disabilities fulfilling strict criteria (Frank, Ward, Orwell, McCullagh, & Belcher, 2000).Those eligible for provision of an EPIOC by the NHS must be are unable to walk around their home unaided, cannot self-propel a manual wheelchair, but are able to operate the powered chair independently and safely. These criteria for EPIOC provision in the NHS are most likely to relate to individuals with CP having Gross Motor Function Classification System (GMFCS) level IV, defined as “self-mobility with limitations; children are transported or use power mobility outdoors and in the community” (Palisano et al., 1997). Some individuals defined as level V who “achieve self-mobility using a power wheelchair with extensive adaptations” (Palisano et al., 1997) may be provided with an EPIOC. Although the GMFCS was developed to describe children, they have been found to be helpful for adults with CP (McCormick et al., 2007).

Many body systems are affected during the growth of a child with CP with a high risk of developing secondary conditions (Kembhavi et al., 2011). The interaction between the aging process and CP is poorly understood, particularly for those unable to walk (Haak et al., 2009). The challenge for powered mobility service providers is to prescribe equipment that maximizes functional potential yet simultaneously manages the risk of developing secondary conditions.

Evidence on the management of those with comorbidity is limited (Smith, Soubhi, Fortin, Hudon, & O’Dowd, 2012).Thus the aims of this study are to describe the clinical features of EPIOC users with CP of all ages and explore the complexities of comorbidities, CP features, and conditions secondary to disability impacting on powered wheelchair provision and clinical management.

## Methods

This is a cross-sectional study of a NHS clinic population and was approved by the National Research Ethics Service.

### *The service*

A specialist NHS service was established for the provision of EPIOCs in 1997 to co-exist with the pre-existing Special Seating Service at the hospital. This Specialist Wheelchair Service (SWS) served a population of around 3.1 million people from both rural and inner city areas. All the therapists and engineers were full-time wheelchair professionals and were supported by three part-time rehabilitation medicine consultants. Users were referred by the local NHS wheelchair services for assessment of suitability for an EPIOC which involved:

(1) Assessment by the locality-based occupational therapist for the suitability of the home environment for the EPIOC prescription and that eligibility criteria (Frank et al., 2000) would be fulfilled. Referral to the SWS included a full medical history and details of the home environment.
(2) Children (those under 19 years) were also assessed by their paediatric therapist to provide details of current management and an evaluation of cognitive, emotional, visuospatial and physical development relating to their suitability for EPIOC driving.
(3) The clinic assessment involved a review of the medical history and relevant social situation (including the home environment suitability for EPIOC use). Visual fields were examined by confrontation, the individual was weighed and physical examinations (in sitting and lying) were performed to identify potential problems with posture/seating or controlling an EPIOC. Transfers were observed for safety and for pain and/or spasticity.
(4) An EPIOC driving assessment included crossing a street and negotiating uneven surfaces, potholes, and kerbs. Consideration was given for the safety of the user and others whilst ensuring satisfactory control of the wheelchair. Age-appropriate supervision of children was accounted for. Following these assessments, an optimal EPIOC and cushion were prescribed.
(5) A rehabilitation engineer from the SWS delivered the EPIOC, usually to the user’s home, explained its features, checked seating, and that driving appeared satisfactory in the user’s home and outdoors.

Community living EPIOC users relevant to this research had a diagnosis of CP. Data consisting of demographic data, clinical issues, and wheelchair factors were entered into electronic records and reviewed between June 2007 and September 2008 by a consultant physician in rehabilitation medicine. They were systematically extracted and entered into a database. Further data were entered from clinical notes (charts) and anonymized by removing names, addresses, and any unique identifier, e.g., hospital identifying number.

Demographic profiles consisted of age at initial assessment and gender. The type of CP was classified into predominantly spastic, dyskinetic, and ataxic groups (Cans et al., 2007). Where a predominant type of CP was not identified, it was classified “unclear.” Where the database held no information, it was classified as “unknown.”

Clinical profiles included comorbidities, complications relating to CP, or disability. Due to the ambiguity which some clinical features may reflect either the CP itself, co-morbidity or a complication of disability, they were grouped as ACFs when referred to collectively (De Souza & Frank, 2015, 2016).

All impairments noted by Novak et al. (2012), in a systematic review with meta-analysis of 30 studies on CP, were categorized as disease related. We added spasticity as a feature of CP although omitted by Novak et al. (2012). Spasticity is a common feature of CP. In the context of this research, spasticity was noted as "problematic" when it interfered with the EPIOC prescription, e.g., stability in the chair (Lacoste, Therrien, & Prince, 2009), increased risk of contracture or pressure ulcers (Noonan, Jones, Pierson, Honkamp, & Leverson, 2004), and musculoskeletal deformity and dysfunction (Liptak, 2008). Users with pain requiring further investigation/management, or influencing the EPIOC prescription were recorded as "problematic pain," as noted by Novak et al. (2012). Those noted by others as being disability related were spinal deformities (Kembhavi et al., 2011; kyphus, scoliosis, and kyphoscoliosis were all noted as kyphoscoliosis), osteoporosis (Kim et al., 2015), oedema/cellulitis (Zaffuto-Sforza, 2005), thromboembolism (Ohmori et al., 2013), pelvic obliquity (Krautwurst et al., 2013), and pressure sores (Janicki, Henderson, & Rubin, 2008). Conditions classified as comorbidities were those considered unrelated to CP.

Wheelchair factors included information about SS, defined as "that which is needed by people who require a wheelchair but due to instability or deformity need additional support in order to function" (British Society of Rehabilitation Medicine, 2004, p. 7). Other data included TIS, cushions, and complex controls.

## Methods of analysis

This was a secondary data analysis of a sub-group of EPIOC users from a cohort reported elsewhere (Frank & De Souza, 2013). Descriptive statistics were used to analyze demographic and wheelchair data.

## Results

A diagnosis of CP was found for 102 users, mean age 28.6 (range 7–70, *SD* ±14.2) years (Table 1), comprising 48 males mean age 27.5 (range 8–70, *SD* ±13.9) years and 54 females, mean age 29.5 (range 7–68, *SD* ±14.6) years. Twenty-three were under 19 years of age (Table 1), two aged under 10, and two aged 65 or over. Users were reviewed a mean of 76 (range 0–133) months after EPIOC assessment. Partial data were available on the medical profiles of 13 users whilst TIS data were available for 90 users. Only 28 users had CP as a single diagnosis, whilst 36 had one ACF and 38 had two or more (Table 1).

The majority had spastic (*n* = 44) or dystonic CP (23 with observed athetoid movements, five referred with dystonia). Ten users were categorized "unclear;" six low-toned (two related to hip surgery), two hemiplegic (not spastic), and two "mixed—spastic and athetoid." There were insufficient data to classify 21 users (Table 2).

Table 1. Features of children aged 0–18 and adults aged 19+ with CP.

| | Age 1–18 | Age 19+ | Total |
|---|---|---|---|
| **Number (Number of males)** | 23 (11) | 79 (37) | 102 (48) |
| **Mean age (range)** | 14.7 (7–18) | 32.6 (19–70) | 28.6 (7–70) |
| **Special seating** | 21 (91.3%) | 43 (54.4%) | 64 (62.7%) |
| **TIS*** | 12 (60%) | 35 (50%) | 47 (52.2%) |
| **Complex controls** | 5 (21.7%) | 11 (13.9%) | 16 (15.7%) |
| **Features of CP** | | | |
| **None** | 14 | 37 | 51 |
| **One** | 7 | 23 | 30 |
| **Two or more** | 2 | 19 | 21 |
| Athetosis | 7 | 16 | 23 |
| Problematic pain | 1 | 19 | 20 |
| Hip problems | 4 | 14 | 18 |
| Spasticity | 1 | 12 | 13 |
| **Comorbidities** | | | |
| **None** | 16 | 56 | 72 |
| **One** | 7 | 18 | 25 |
| **Two or more** | 0 | 5 | 5 |
| **Complications of disability** | | | |
| **None** | 16 | 61 | 77 |
| **One** | 7 | 16 | 23 |
| **Two or more** | 0 | 2 | 2 |
| (Kypho)scoliosis | 7 | 14 | 21 |
| **Total ACFs** | | | |
| **None** | 5 | 23 | 28 |
| **One** | 12 | 24 | 36 |
| **Two or more** | 6 | 32 | 38 |

*Note*. *No data on 12 users (three <18; nine 19+).

Two users were mothers. One, who had difficulty caring for her 9-year-old child, had dystonic CP with problematic pain and spasticity complicated by recurrent deep venous thromboses (on Warfarin) and asthma. The other, with small children, had problematic back pain associated with kyphoscoliosis, severe oedema, and chronic cellulitis.

### *Comorbidities*

Sixteen comorbidities were identified with asthma and osteoarthritis being the commonest (Table 3). Five users had congenital conditions (two hydrocephalus and one each with cystic fibrosis, congenital heart disease, and familial spastic paraplegia). Three adults had depression.

### *CP disease-related impairments*

We found nine features of CP, of which eight were reported by Novak et al. (2012), the most frequent being problematic pain (*n* = 20), hip problems (*n* = 18), and problematic spasticity (*n* = 15) (Table 3). Specified causes of problematic pain were spasticity (*n* = 7), (kypho)scoliosis (*n* = 6), back pain (*n* = 5), hip pain (*n* = 2), back pain, and spasticity (*n* = 2), no specified cause (*n* = 4), or more than one of the above. Nine users reported back pain thought to be treatable with standard approaches.

Thirteen users had problematic spasticity (12 aged 19 or older—Table 1), two had contractures complicating management and one had a large ischial pressure sore. We also found epilepsy such that driving was not contraindicated (*n* = 8). Fatigue was not reported.

### *Complications of disability*

(Kypho)scoliosis was the commonest complication of disability (*n* = 21 mean age 25.1, *SD* ±14.2, range 7–68 years; Table 1)

Table 2. Categories of 102 CP EPIOC users by age, gender, use of SS, and TIS; problematic pain, scoliosis, spasticity, and hip problems.

| CP category | *N* (*N* of men) | Mean age (±*SD*) years | *N* under age 19 years | SS *N* | TIS *N** | SS and TIS *N** | Scoliosis *N* | Problematic pain *N* | Hip problems *N* | Spasticity *N* |
|---|---|---|---|---|---|---|---|---|---|---|
| **Spastic** | 43 (23) | 26.6 (14.4) | 10 | 29 | 26 | 22 | 10 | 12 | 11 | 9 |
| **Dystonic** | 28 (13) | 27.2 (13.2) | 9 | 19 | 11 | 10 | 2 | 5 | 4 | 4 |
| **Unclear** | 10 (5) | 26.2 (17.8) | 3 | 8 | 4 | 3 | 5 | 0 | 2 | 0 |
| **Not known** | 21 (7) | 35.6 (12.2) | 1 | 8 | 6 | 5 | 4 | 3 | 1 | 0 |
| **Total** | **102 (48)** | **28.6 (14.2)** | **23** | **64 (63%)** | **47 (52%)** | **40 (44%)** | **21** | **20** | **18** | 13 |

*Note*. Number with unknown TIS use = 12.

Table 3. Features of CP and disease-related impairments, comorbidities and complications of disability of 102 EPIOC users compared to published impairments for CP.

| **Impairments noted by** Novak et al. (2012) | Disease-related impairments (*n*) | Disability (*n*) | Comorbidity (*n*) |
|---|---|---|---|
| Behavioral | | | |
| Constipation | | | |
| Dribbling | | | |
| Eating difficulties | | | |
| Sleep disorder | | | |
| | | (Kypho)scoliosis 21 | |
| For those able to walk, neck, back, and feet are high risk pain sites | Problematic pain 20<br>Back pain 11 | | |
| Hip dislocation | Hip dislocation/other hip 18 | | |
| | Problematic spasticity 13 and contractures 2 | | |
| | | | Asthma 11 |
| Epilepsy | Epilepsy 8 | | |
| | | | Osteoarthritis 5 |
| Intellectual impairment | Learning impairment 4 | | |
| Hearing impairments | | | |
| Talking | Communication impairments 3 | | |
| | | Pressure Sores 3 | |
| | | | Depression 3 |
| | | | Peptic ulcer 2 |
| | | | Hiatus hernia 2 |
| | | | Hydrocephalus 2 |
| High risk for malnutrition | Weight loss2 | | |
| | | Pelvic obliquity (not associated with scoliosis/hip | |
| | | Pain 1 | |
| | | Osteoporosis 1 | |
| | | Thromboembolic disease 1 | |
| Vision impairment | Vision impairment 1 | | |
| | | Oedema/cellulitis 1 | |
| | | | Cancer 1 |
| | | | Encephalopathy 1 |
| | | | Congenital heart disease 1 |
| | | | Tenosynovitis 1 |
| | | | Chronic fatigue syndrome 1 |
| | | | Migraine 1 |
| | | | Discoid Lupus 1 |
| | | | Familial spastic paraplegia 1 |
| Bladder control | Urinary incontinence 1 | | |
| | | | Cystic fibrosis 1 |
| | | | Knee arthrodesis 1 |

managed with SS ($n$ = 19) and TIS ($n$ = 12). Three users had pressure sores, one user had oedema and cellulitis, and another user had thromboembolic disease (see above and Table 3). Problematic ACFs requiring further medical management were found in 47 users who were referred to their family doctors. Of these, 18 required medical management for problematic pain, 16 for (kypho)scoliosis and three each for spasticity and hip problems.

## *Wheelchair factors*

Data on TIS were available for 90 users of whom 47 (52%) used TIS. Sixty-four (63%) users had SS and 40 (44%) had both SS and TIS (Table 2). Two users under 19 years, both having spastic CP without ACFs, did not have SS (Table 1). Sixteen users, five under 19 years, had complex control systems (Table 1) of which 12 were tray mounted, eight were non-standard, and nine had controls interfacing with other AT, e.g., environmental control systems. Nine users with complex controls were provided with both SS and TIS.

The most common SS provided were CAPS 11 (seating system for postural management which allows for growth; $n$ = 12; mean age 16, range 7–26, *SD* ±6.1 years), carved foam ($n$ = 10; mean age 29.5, range 20–45, *SD* ±9.4 years), and Scott Seating (a modular seating system; $n$ = 9; mean age 21.3, range 13–45, *SD* ±9.4 years). Two users were prescribed Matrix seating and two molded seat inserts. Users aged 18 or less were provided with SS more often than those aged 19 or more (Table 1).

The most common cushions provided were Qbitus (bespoke pressure reduction; $n$ = 29, mean age 32.2, range 14–70, *SD* ±16.3 years), Jay 2 ($n$ = 13; mean age 29.4, range

18–51, *SD* ±13.1 years) and standard cushions (*n* = 12: mean age 29.2, range 12–60, *SD* ±14.2 years).

## Discussion

This study is the first to describe a cohort of individuals with CP GMFCS levels IV and V, irrespective of age, prescribed an EPIOC. It highlights issues considered most relevant to aging with CP, namely, mobility, pain, fatigue, and comorbidity (Haak et al., 2009). Our cohort is atypical, mostly excluding those with learning/intellectual impairments and not including children of preschool age (Frank & De Souza, 2013). It excludes those able to walk and self- propel and thus includes a high proportion of users with spastic tetraplegia. Six users with predominant hypotonicity did not fit into the categorization of Cans et al. (2007) and were grouped "unclear" although others consider hypotonicity as a notable feature of CP (Aisen et al., 2011; Shevell, Dagenais, & Hall, 2009).

### Problematic features of CP

Problematic pain is a major issue for children, young people (Novak et al., 2012; Penner, Xie, Binepal, Switzer, & Fehlings, 2013) and adults (Opheim, Jahnsen, Olsson, & Stanghelle, 2009) with CP. Our finding that problematic pain was predominately in adults may reflect deterioration in the underlying CP (Opheim et al., 2009) inadequate management (Noonan et al., 2004; Zaffuto-Sforza, 2005) or both. Provision of an EPIOC can be used to reduce users' pain when first provided (Davies et al., 2003). Although a similar service was provided for both adults and children, children may have been seen more often due to their growth indicating wheelchair adjustments were needed. Problematic pain in EPIOC users may reflect pain relating to their underlying medical condition, their wheelchair and seating or a combination of the two (Frank, De Souza, Frank, & Neophytou, 2012). The problematic pain may also relate to spasticity (Aisen et al., 2011; Zaffuto-Sforza, 2005), dislocated hips contributing to back pain and/or spasticity, (Van Der Slot et al., 2012), scoliosis, and/or prolonged sitting (Frank et al., 2012).

Although problematic neck pain is a noted feature of CP (Jahnsen, Villien, Aamodt, Stanghelle, & Holm, 2004; Novak et al., 2012) our users were routinely provided with neck restraints to minimise any neck pain. The provision of TIS also facilitates resting the neck during tilting (Angsupaisal et al., 2015).

Most of our users had spasticity, which was problematic in 13 of them. Spasticity due to poor posture is managed through appropriate seating which probably explains the high use of SS in this study. Contractures are widespread in adults with CP but the majority of contractures can be "accommodated" through the chair and SS.

Hip problems are widespread in CP (Novak et al., 2012). In children, management relates to the underlying cause and EPIOCs with SS may help to prevent hip problems from developing/progressing (Pountney, Mandy, Green, & Gard, 2002) while allowing for growth. Consequently, all users, except one adult with hip problems, in this cohort were provided with SS. This indicates that hip assessment is a key clinical focus those with CP and is likely to partly explain the use of CAPS 11 seating systems in this cohort.

Athetosis is challenging for wheelchair providers as prevention of limb trauma through the athetoid movements is needed, e.g., by providing padding over projecting metal surfaces. Athetosis was not contraindicated for EPIOC provision for 23 users who completed the assessment safely.

Fatigue was not an issue for this cohort as EPIOCs eliminate energy expenditure through impaired walking or by self-propelling a manual chair. Just over half of this cohort was provided with TIS—a widely used fatigue management strategy (Dicianno et al., 2015).

### ACFs

Nearly two-thirds of our users had ACFs potentially influencing the EPIOC prescription which may be critical for service funders. The number of ACFs increases with CP GMFCS levels IV and V and in those with specific neurologic profiles such as spastic tetraplegia and dyskinesia (Shevell et al., 2009). We have shown there is similar impact in adults with CP, compounded by ACFs associated with aging, indicating that practitioners should be proactive in responding to age-associated features (Janicki et al., 2008).

There is ambiguity concerning what clinical features are associated with CP and what are comorbidities. Thus epilepsy is noted to be a feature of CP by Novak et al. (2012) but a comorbidity by Shevell et al. (2009). Likewise it is debateable if contractures are a feature of CP or if they are a consequence of long-term disability—dilemmas already noted in multiple sclerosis (De Souza & Frank, 2015). In determining which ACFs were associated with CP we used published clinical prognostic recommendations (Novak et al., 2012).

### Comorbidity

We used a straightforward count of comorbidity (Carmona, 2014) and confirm its increased occurrence in CP (Liptak, 2008). Similar to other reports, we found a few users with comorbid congenital anomalies (Rankin et al., 2010). Although we noted only one user with encephalopathy, it has been reported previously (Kyriakopoulos, Oskoui, Dagenais, & Shevell, 2013). Hydrocephalus was coded as a comorbidity as it has been considered a "chance finding" in CP (Rankin et al., 2010). We noted depression as a comorbidity (Sangha, Stucki, Liang, Fossel, & Katz, 2003) whilst recognizing it may relate to long term disablement.

### Features secondary to severe disability

A clinically significant (kypho)scoliosis was the commonest complication of disability found (Graham, Ang, Johnson, Torode, & Simpson, 2014). Most of our users were aged over 18 suggesting that higher levels of disability increase the risk of developing scoliosis (Persson-Bunke, Hagglund, Lauge-Pedersen, Ma, & Westbom, 2012). All but two users with a (kypho)scoliosis were provided with SS and whilst postural stability can be supported by SS (Angsupaisal et al., 2015), this may sacrifice upper trunk function which may not be the choice of the user.

Pressure sores, thromboembolic disease, oedema/cellulitis, and osteoporosis are all preventable complications of disability and the incidence in this cohort was low. This finding does not negate the seriousness of these complications and vigilance is needed to identify those at risk. Modest dependent oedema can be assisted with TIS, elevating leg rests, and users should elevate their legs at night.

### *Wheelchair features and seating*

CP users were provided with SS more frequently than any other diagnostic group from our main study (Frank & De Souza, 2013). The clinical reasoning stems from evidence that maintaining adequate seating and postural control is a priority for preventing deformity (Noonan et al., 2004), to facilitate activities of daily living (Lacoste et al., 2009), and optimize pain management (Noonan et al., 2004). CAPS II seats were often utilized for younger users to maintain optimal hip position, inhibit development of scoliosis, and allow for growth. Where CAPS II was inappropriate but bespoke pressure reduction was needed, carved foam seating was provided for those at risk of skin breakdown (Apatsidis, Solomonidis, & Michael, 2002). All but 12 users were provided with pressure reduction cushions/seating, thus minimizing the likelihood of pressure sores and maximizing comfort.

TIS was provided to just over half the users, irrespective of age, reflecting common practice (Frank & De Souza, 2013). TIS benefits users with poor head control, discomfort, fatigue, and/or pain (Dicianno et al., 2015; Frank et al., 2012).

We found that most EPIOC users drive safely using standard control systems but 16 were unable to do so. They were provided with bespoke controls to facilitate driving independently and interface with other AT, e.g., environmental controls. For those with CP needing complex controls, an assessment regarding additional AT is recommended.

Supporting individuals with CP is costly (Kruse et al., 2009). Our finding that a high proportion of users were given SS, TIS, and/or both, together with modifications to control systems, as part of condition management has cost implications for service payers.

### *Safety*

Safety issues exist for all EPIOC users irrespective of age or diagnosis (Evans et al., 2007; Frank, Neophytou, Frank, & De Souza, 2010). Assessments minimize risk for users and the public. Those with severe visual impairment, uncertain epilepsy control, and learning disabilities are unlikely to fulfill eligibility criteria. Our cohort had one with vision impairment, eight with epilepsy and four with learning disability, all of whom fulfilled eligibility criteria. For those with epilepsy, the criteria were identical as for driving cars (Driver and Vehicle Licensing Authority, 2014). For those with visual impairments, the clinic and driving assessments seemed adequate.

### *Strengths and limitations of the study*

A major strength of this study was the eligibility criteria, restriction of EPIOC provision to those with GMFCS levels IV and V. This cohort included all ages apart from pre-school age children—a limitation as a complete picture is not provided of EPIOC provision in those severely affected by CP. This, together with severe disability without intellectual impairment, does not permit comparison with other CP cohorts. Research on adults with CP is currently sparse and our older subgroup provides comparisons for future studies.

All users were seen by the same multidisciplinary team with expertise in wheelchair prescription for users with severe and/or multiple impairments. Thus expertise was consistently applied in prescribing EPIOCs.

This study is likely to under-report ACFs, as the data were mostly obtained via referral letters and patient histories; and as case note reviews are less comprehensive than prospective data. Nonetheless these data are more objective than that relying purely on patient self-report surveys.

Our sample does not include those who purchased wheelchairs privately or through charitable funding. This limitation is more likely to influence our findings in children/young adults as sources of charitable funding are greater for these users.

Findings were limited by missing TIS information and the medical profiles. Prospective studies are needed to obtain complete comorbidity information.

## Conclusion

Powered wheelchair prescription has an important therapeutic role in clinical management in addition to enhancing mobility, independence, and participation. It is influenced by the underlying neuromuscular features of CP, comorbidities and conditions related to long-term disability, and requires a model of rehabilitation which is holistic rather than a disease management orientated approach.

Clinical features such as spasticity and problematic pain appeared less well managed in adults than in children and have consequences for EPIOC provision especially with regard to type of chair and seating prescription. Children were more frequently provided with the additional wheelchairs features of TIS, SS, and complex controls. Conversely, adults experienced more problematic pain, more problematic spasticity, and more comorbidities. The impact on EPIOC provision of ACFs in adults with CP requires further study.

## References

Aisen, M. L., Kerkovich, D., Mast, J., Mulroy, S., Wren, T. A., Kay, R. M., & Rethlefsen, S. A. (2011). Cerebral palsy: Clinical care and neurological rehabilitation. *The Lancet Neurology*, *10*(9), 844–852. doi:10.1016/S1474-4422(11)70176-4

Andersson, C., & Mattsson, E. (2001). Adults with cerebral palsy: A survey describing problems, needs, and resources, with special emphasis on locomotion. *Developmental Medicine and Child Neurology*, *43*(2), 76–82. doi:10.1017/S0012162201

Angsupaisal, M., Maathuis, C. G. B., & Hadders-Algra, M. (2015). Adaptive seating systems in children with severe cerebral palsy across international classification of functioning, disability and health for children and youth version domains: A systematic review. *Developmental Medicine and Child Neurology*, *57*(10), 919–930. doi:10.1111/dmcn.12762

Apatsidis, D. P., Solomonidis, S. E., & Michael, S. M. (2002). Pressure distribution at the seating interface of custom-molded wheelchair seats: Effect of various materials. *Archives of Physical Medicine & Rehabilitation*, *83*(8), 1151–1156. doi:10.1053/apmr.2002.33987

British Society of Rehabilitation Medicine. (2004). *Specialised wheelchair seating national clinical guidelines* (Report of a multidisciplinary expert group). London: British Society of Rehabilitation Medicine.

Cans, C., Dolk, H., Platt, M. J., Colver, A., Prasauskiene, A., & Kragel-Oh-Mann, I. (2007). Recommendations from the SCPE collaborative group for defining and classifying cerebral palsy. *Developmental Medicine and Child Neurology*, *49*(Suppl. 2), 35–38. doi:10.1111/j.1469-8749.2007.tb12626.x

Carmona, L. (2014). Is there a best way to measure co-morbidity? *Rheumatology*, *53*, 963–964. doi:10.1093/rheumatology/ket493

Davies, A., De Souza, L. H., & Frank, A. O. (2003). Changes in the quality of life in severely disabled people following provision of powered indoor/outdoor chairs. *Disability and Rehabilitation*, *25*(6), 286–290. doi:10.1080/0963828021000043734

De Souza, L. H., & Frank, A. O. (2015). Problematic clinical features of powered wheelchair users with severely disabling multiple sclerosis. *Disability and Rehabilitation*, *37*(11), 990–996. doi:10.3109/09638288.2014.949356

De Souza, L. H., & Frank, A. O. (2016). Rare diseases: Matching wheelchair users with rare metabolic, neuromuscular or neurological disorders to electric powered indoor/outdoor wheelchairs. *Disability and Rehabilitation*, *38*(16), 1547–1556. doi:10.3109/09638288.2015.1106599

Dicianno, B. E., Lieberman, J., Schmeler, M., Souza, A., Cooper, R., Lange, M., … Jan, Y.-K. (2015). *RESNA position on the application of tilt, recline, and elevating leg rests for wheelchairs: 2015 current state of the literature*. Arlington, VA: RESNA.

Driver and Vehicle Licensing Authority. (2014). *Driving liciences—epilepsy: Group 1 (car + motorcycle) driving entitlement* (IN59.2014). Swansea, Wales: Driver and Vehicle Licensing Agency.

Evans, S., Neophytou, C., De Souza, L. H., & Frank, A. O. (2007). Young people's experiences using electric powered indoor-outdoor wheelchairs (EPIOCs): Potential for enhancing users' development? *Disability and Rehabilitation*, *19*(16), 1281–1294. doi:10.1080/09638280600964406

Feinstein, A. R. (1970). The pre-therapeutic classification of co-morbidity in chronic disease. *Journal of Chronic Diseases*, *23*(7), 455–468. doi:10.1016/0021-9681(70)90054-8

Frank, A. O., & De Souza, L. H. (2013). Recipients of electric-powered indoor/outdoor wheelchairs provided by a national health service: A cross-sectional study. *Archives of Physical Medicine and Rehabilitation*, *94*, 2403–2409. doi:10.1016/j.apmr.2013.07.010

Frank, A. O., De Souza, L. H., Frank, J. L., & Neophytou, C. (2012). The pain experiences of powered wheelchair users. *Disability and Rehabilitation*, *34*(9), 770–778. doi:10.3109/09638288.2011.619620

Frank, A. O., Neophytou, C., Frank, J., & De Souza, L. H. (2010). Electric powered indoor/outdoor wheelchairs (EPIOCs): Users views of influence on family, friends and carers. *Disability and Rehabilitation: Assistive Technology*, *5*(5), 327–338. doi:10.3109/17483101003746352

Frank, A. O., Ward, J. H., Orwell, N. J., McCullagh, C., & Belcher, M. (2000). Introduction of a new NHS electric-powered indoor/outdoor chair (EPIOC) service: Benefits, risks and implications for prescribers. *Clinical Rehabilitation*, *14*(6), 665–673. doi:10.1191/0269215500cr376oa

Govender, V., Hepworth, L., Bagwandeen, N., & Chetty, T. (2015). P6–2654: Clinical spectrum of cerebral palsy in Durban, South Africa. *European Journal of Paediatric Neurology*, *19*(Suppl 1), S96. doi:10.1016/S1090-3798(15)30319-6

Graham, H. K., Ang, S. G., Johnson, M. B., Torode, I. P., & Simpson, P. (2014). Scoliosis in cerebral palsy: A population-based study. *Developmental Medicine and Child Neurology*, *56*, 34.

Haak, P., Lenski, M., Hidecker, M. J. C., Li, M., & Paneth, N. (2009). Cerebral palsy and aging. *Developmental Medicine and Child Neurology*, *51*(Suppl 4), 16–23. doi:10.1111/j.1469-8749.2009.03428.x

Jahnsen, R., Villien, L., Aamodt, G., Stanghelle, J., & Holm, I. (2004). Musculoskeletal pain in adults with cerebral palsy compared with the general population. *Journal of Rehabilitation Medicine*, *36*(2), 78–84. doi:10.1080/16501970310018305

Janicki, M. P., Henderson, C. M., & Rubin, I. L. (2008). Neurodevelopmental conditions and aging: Report on the Atlanta study group charrette on neurodevelopmental conditions and aging. *Disability and Health Journal*, *1*(2), 116–124. doi:10.1016/j.dhjo.2008.02.004

Kembhavi, G., Darrah, J., Payne, K., & Plesuk, D. (2011). Adults with a diagnosis of cerebral palsy: A mapping review of long-term outcomes. *Developmental Medicine and Child Neurology*, *53*(7), 610–614. doi:10.1111/j.1469-8749.2011.03914.x

Kim, W., Lee, S. J., Yoon, Y.-K., Shin, Y.-K., Cho, S.-R., & Rhee, Y. (2015). Adults with spastic cerebral palsy have lower bone mass than those with dyskinetic cerebral palsy. *Bone*, *71*, 89–93. doi:10.1016/j.bone.2014.10.003

Krautwurst, B. K., Wolf, S. I., Heitzmann, D. W., Gantz, S., Braatz, F., & Dreher, T. (2013). The influence of hip abductor weakness on frontal plane motion of the trunk and pelvis in patients with cerebral palsy. *Research in Developmental Disabilities*, *34*(4), 1198–1203. doi:10.1016/j.ridd.2012.12.018

Kruse, M., Michelsen, S., Flachs, E. M., Bronnum-Hansen, H., Madsen, M., & Uldall, P. (2009). Lifetime costs of cerebral palsy. *Developmental Medicine and Child Neurology*, *51*(8), 622–628. doi:10.1111/j.1469-8749.2008.03190.x

Kyriakopoulos, P., Oskoui, M., Dagenais, L., & Shevell, M. I. (2013). Term neonatal encephalopathy antecedent cerebral palsy: A retrospective population-based study. *European Journal of Paediatric Neurology*, *17*(3), 269–273. doi:10.1016/j.ejpn.2012.11.001

Lacoste, M., Therrien, M., & Prince, F. (2009). Stability of children with cerebral palsy in their wheelchair seating: Perceptions of parents and therapists. *Disability and Rehabilitation: Assistive Technology*, *4*(3), 143–150. doi:10.1080/17483100802362036

Liptak, G. S. (2008). Health and well being of adults with cerebral palsy. *Current Opinion in Neurology*, *21*(2), 136–142. doi:10.1097/WCO.0b013e3282f6a499

McCormick, A., Brien, M., Plourde, J., Wood, E., Rosenbaum, P., & McLean, J. (2007). Stability of the gross motor function classification system in adults with cerebral palsy. *Developmental Medicine and Child Neurology*, *49*(4), 265–269. doi:10.1111/j.1469-8749.2007.00265.x

Noonan, K. J., Jones, J., Pierson, J., Honkamp, N. J., & Leverson, G. (2004). Hip function in adults with severe cerebral palsy. *Journal of Bone and Joint Surgery—Series A*, *86*(12), 2607–2613.

Novak, I., Hines, M., Goldsmith, S., & Barclay, R. (2012). Clinical prognostic messages from a systematic review on cerebral palsy. *Pediatrics*, *130*(5), e1285–e1312. doi:10.1542/peds.2012-0924

Ohmori, H., Ochi, F., Tanuma, N., Ohnuki, E., Yamasaki, M., Takesue, H., … Harada, A. (2013). Deep vein thrombosis in patients with severe motor and intellectual disabilities. *Annals of Vascular Diseases*, *6*(4), 694–701. doi:10.3400/avd.oa.13-00090

Opheim, A., Jahnsen, R., Olsson, E., & Stanghelle, J. K. (2009). Walking function, pain, and fatigue in adults with cerebral palsy: A 7-year follow-up study. *Developmental Medicine & Child Neurology*, *51*(5), 381–388. doi:10.1111/(ISSN)1469-8749

Palisano, R., Rosenbaum, W., Walter, S., Russell, D., Wood, E., & Galuppi, B. (1997). Development and reliability of a system to classify gross motor function in children with cerebral palsy. *Developmental Medicine and Child Neurology*, *39*(4), 214–223. doi:10.1111/j.1469-8749.1997.tb07414.x

Parkes, J., Donnelly, M., & Hill, N. (2001). *Focussing on cerebral palsy: Reviewing and communicating needs for services* (1st ed.). London: Scope.

Penner, M., Xie, W. Y., Binepal, N., Switzer, L., & Fehlings, D. (2013). Characteristics of pain in children and youth with cerebral palsy. *Pediatrics*, *132*(2), e407–e413. doi:10.1542/peds.2013-0224

Persson-Bunke, M., Hagglund, G., Lauge-Pedersen, H., Ma, P. W., & Westbom, L. (2012). Scoliosis in a total population of children with cerebral palsy. *Spine*, *37*(12), E708–E713. doi:10.1097/BRS.0b013e318246a962

Pountney, T., Mandy, A., Green, E., & Gard, P. (2002). Management of hip dislocation with postural management. *Child: Care, Health and Development*, *28*(2), 179–185.

Rankin, J., Cans, C., Garne, E., Colver, A., Dolk, H., Uldall, P., … Krageloh-Mann, I. (2010). Congenital anomalies in children with cerebral palsy: A population-based record linkage study. *Developmental Medicine & Child Neurology*, *52*(4), 345–351. doi:10.1111/dmcn.2010.52.issue-4

Rosenbaum, P. (2003). Cerebral palsy: What parents and doctors want to know. *British Medical Journal*, *326*(7396), 970–974. doi:10.1136/bmj.326.7396.970

Rosenbaum, P. (2014). What causes cerebral palsy? *British Medical Journal*, *349*, g4514. doi:10.1136/bmj.g4514

Sangha, O., Stucki, G., Liang, M. H., Fossel, A. H., & Katz, J. N. (2003). The self-administered comorbidity questionnaire: A new method to assess comorbidity for clinical and health services research. *Arthritis and Rheumatism*, *49*(2), 156–163. doi:10.1002/art.10993

Shevell, M. I., Dagenais, L., & Hall, N. (2009). Comorbidities in cerebral palsy and their relationship to neurologic subtype and GMFCS level. *Neurology*, *72*(24), 2090–2096. doi:10.1212/WNL.0b013e3181aa537b

Smith, S. M., Soubhi, H., Fortin, M., Hudon, C., & O'Dowd, T. (2012). Managing patients with multimorbidity: Systematic review of interventions in primary care and community settings. *British Medical Journal*, *345*, e5205. doi:10.1136/bmj.e5205

Tollanes, M. C., Wilcox, A. J., Lie, R. T., & Moster, D. (2014). Familial risk of cerebral palsy: Population based cohort study. *British Medical Journal*, *349*, g4294. doi:10.1136/bmj.g4294

Valderas, J. M., Starfield, B., Sibbald, B., Salisbury, C., & Roland, M. (2009). Defining comorbidity: Implications for understanding health and health services. *The Annals of Family Medicine*, *7*(4), 357–363. doi:10.1370/afm.983

Van Der Slot, W. M., Nieuwenhuijsen, C., Van Den Berg-Emons, R., Bergen, M., Hilberink, S., Stam, H., & Roebroecks, M. (2012). Pain in adults with spastic bilateral cerebral palsy. *Developmental Medicine and Child Neurology*, *54*, 69–70. doi:10.1111/j.1469-8749.2012.04371.x

Zaffuto-Sforza, C. (2005). Aging with cerebral palsy. *Physical Medicine and Rehabilitation Clinics of North America*, *16*(1), 235–249. doi:10.1016/j.pmr.2004.06.014

Zenczak-Praga, K., Zgorzalewicz-Stachowiak, M., & Cesar, K. (2015). Analysis of using assistive devices by patients suffering from cerebral palsy—preliminary report. *Polski Merkuriusz Lekarski: Organ Polskiego Towarzystwa Lekarskiego*, *39*(233), 311–315.

Zupan, A., & Jenko, M. (2012). Assistive technology for people with cerebral palsy. *Eastern Journal of Medicine*, *17*(4), 194–197.