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ENERGY REQUIREMENTS IN CHILDREN WITH CEREBRAL PALSY

Bachelor's thesis

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ENERGY REQUIREMENTS IN CHILDREN WITH CEREBRAL PALSY

Abstract

Cerebral palsy (CP) is the most frequent cause of neurological impairment in childhood, with a prevalence of 2-2.5 per 1000 live births in developed countries. The main objective of this work is to review energy requirements in children with this condition in order to perform early nutritional management and to avoid complications such as undernutrition, overweight, or lack of growth. Results of different studies showed that energy requirements in children with CP are significantly different compared to typically developing children and depend upon the condition of the child. Therefore, equations based on healthy children tend to overestimate energy requirements in this population. There is literature demonstrating that fat-free mass, ambulatory status, motor type, and distribution of limbs influence energy requirements and should be considered in daily clinical practice. Generally, children with CP have a lower fat-free mass and a higher percentage of body fat in comparison with typically developing children leading to a decrease in energy requirements. This work proposes a decision tree considering the ambulatory status of the child to provide a starting point to estimate energy requirements. Further studies are required to develop more precise specific formulae in children with CP, taking into consideration the factors that may influence energy requirements.

Keywords: cerebral palsy, energy requirements, neurological impairment, children, formulae.

Resum

La paràlisi cerebral (PC) és la causa més freqüent de disfunció neurològica en la infància, amb una prevalença de 2-2.5 per cada 1000 naixements als països desenvolupats. El principal objectiu d'aquest treball és revisar els requeriments energètics en el grup d'infants que pateixien aquesta condició per tal de realitzar una intervenció nutricional primerenca, i així evitar complicacions com ara la desnutrició, el sobrepès o la falta de creixement. Els resultats de diferents estudis han demostrat que els requeriments energètics en nens amb PC són significativament diferents en comparació amb nens amb un desenvolupament típic i depenen de la condició de l'infant. Així doncs, les equacions basades en nens sans tendeixen a sobreestimar els requeriments energètics. S'ha observat que la massa lliure de greix, l'estat ambulatori, el tipus motor i la distribució de les extremitats afectades de l'infant influeixen en els requeriments energètics i s'han de tenir en consideració durant la pràctica clínica diària. Generalment, els nens amb PC tenen una menor massa lliure de greix i un percentatge més elevat de greix corporal en comparació amb els nens que tenen un desenvolupament típic,fet que condueix a una disminució dels requeriments energètics. Amb aquest treball es proposa un arbre de decisió tenint en compte l'estat ambulatori del nen amb l'objectiu de proporcionar un punt de partida per a l'estimació dels requeriments energètics. Es requereixen estudis addicionals per desenvolupar fórmules més precises pels infants amb PC, considerant els factors que poden influir en els requeriments energètics.

Paraules clau: Paràlisi cerebral, requeriments energètics, disfunció neurològica, nens, fórmules.

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List of Abbreviations

BIA bioelectrical impedance analysis. 15

CP cerebral palsy. 8

DLW doubly labeled water. 12

EC energy cost. 15

ER energy requirements. 11

ESPGHAN European Society for Paediatric Gastroenterology. 13

FFM fat-free mass. 14

GMFCS Gross Motor Function Classification System. 9

IC indirect calorimetry. 12

REE resting energy expenditure. 12

SCPE Surveillance of Cerebral Palsy in Europe. 8

TBW total body water. 12

TDC typically developing children. 11

TEE total energy expenditure. 12

WHO World Health Organization. 13

Chapter 1

Introduction

Cerebral palsy is considered the most common cause of neurological impairment in children and it encompasses a heterogeneous group of children who suffer from a non-progressive brain injury caused in the early years of life, specifically during the antenatal, perinatal or early postnatal period of time [1].

Malnutrition is common among children with cerebral palsy. Both under- and over- nutrition are associated with worsened quality of life, increased health care use, and heightened morbidity and mortality. On the one hand, if undernutrition is not treated, severe nutritional problems may occur, such as an impaired immune system, cognitive problems, and neuromuscular disabilities [2]. On the other hand, overweight may result because the caregiver perceives that the child is malnourished and may cause complications such as an increased incidence of sleep apnea [3].

Inadequate energy intake is the main cause of undernutrition, overweight, and growth failure in this group of children. Indeed, this population is often unable to communicate their hunger or satiety, food preferences, and self-feed between meals. This involves giving caregivers the responsibility to regulate their dietary intake. Besides, feeding issues are also frequent and hinder the meal time. As a consequence, the meal time is prolonged, slow and stressful for both the patient and caretaker and often results in weight loss that further increases energy requirements [4]. Caretakers often tend to overestimate the child's energy intake and underestimate the time spent feeding the child [5].

Prevention and treatment of malnutrition requires an understanding of their nutritional needs, especially energy requirements. Unfortunately, precise estimations in energy needs are challenging because of variations in energy requirements linked with the heterogeneity of the group, altered body composition and low physical activity levels. Thus, energy requirements are influenced by many factors that are not routinely found in typically developing children [6].

There is a need to reach a consensus regarding which are the specific recommendations for estimating energy requirements in children with cerebral palsy. Thus, this thesis examines what is currently known about energy needs through a literature review. Firstly, it analyzes the measured resting energy expenditure and total energy expenditure values in subjects with cerebral palsy in different studies using reliable methods like indirect calorimetry or doubly labeled water. Secondly, it examines the available equations for calculating the energy requirements in this group of children. Finally, it profiles energy assessment considering the factors which alter energy requirements.

1.1 Objectives

The main objective is to do a literature review of energy requirements in children with cerebral palsy and provide a tool to estimate them taking into account their conditions and factors that may influence them.

The secondary aims of this bibliographic research are:

1. Review the measured resting energy expenditure and total energy expenditure in subjects with cerebral palsy in different studies using reliable methods such as indirect calorimetry and doubly labeled water.

2. Identify and summarize the current available equations to estimate energy requirements in children with this condition considering their limitations.

3. Provide a guide to schools and education centers to estimate their energy requirements with a decision tree.

1.2 Methods

This thesis consists in a bibliographic research and has been based on research found through literature concerning energy requirements in people with cerebral palsy. First of all, a general overview of the topic was consulted in books and reviews from the field to write the background theory. The search strategy included these terms: ”cerebral palsy” [AND] ”history”,”cerebral palsy” [AND] ”definition”,”cerebral palsy” [AND] ”epidemiology”,”cerebral palsy” [AND] ”classification”, ”cerebral palsy” [AND] ”Gross Motor Function System”.

After that, research in clinical practice guidelines was conducted to investigate current nutritional recommendations in children with cerebral palsy. Furthermore, a search of the literature was performed using the following terms: ”energy requirements” [OR] ”energy needs”, ”energy intake”,”dietary intakes” [OR] ”nutrient intakes”, ”indirect calorimetry”,”doubly labeled water”, ”feeding difficulties”, ”fat-free mass”, ”ambulatory” [OR] ”ambulant”, ”nonambulatory” [OR] ”nonambulant”, ”Gross Motor Function System”, ”spastic cerebral palsy”, ”dyskinetic cerebral palsy” [OR] ”athetoid cerebral palsy” ”neurological impairment, ”cerebral palsy”, etc.

Although other bibliographic databases were used for the search (Cochrane Library, Scopus, Google Scholar and ”Cerca-bib” from the Universitat de Barcelona), the main used source was PubMed. Research criteria consisted on finding the most relevant articles giving priority to recent studies and considering the type of study. Thus, randomized clinical trials and prospective cohort studies were preferred despite examining other studies.

Chapter 2

Background theory

2.1 Definition and classification of cerebral palsy

In 2004, an International Workshop on Definition and Classification of Cerebral Palsy at the US National Institutes of Health was carried out in order to review the information about CP and reach an international consensus on the definition and classification of cerebral palsy (CP). In the consensus document, they defined CP as follows [7]:

”Cerebral palsy describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems.”

As shown in the previous definition, the therm CP is deeply heterogeneous due to many clinical presentations and different degrees of activity limitation [7]. Thus, a classification is useful to compare similar groups and achieve significant results in studies. There are different classification systems used in CP and these are mainly classified by motor type, distribution of limbs and functional level [8].

In the first place, the motor type is classified as spastic, diskinetic and ataxic [9]. Table 2.1 shows the definitions adopted by the Surveillance of Cerebral Palsy in Europe (SCPE) [9].

Furthermore, the SCPE suggests classifying the SCP subtipes based on the distribution of limb involvement as unilateral or bilateral (See Table 2.1) with a description of the motor impairment and the functional motor classification [7; 9; 11] (such as the Gross Motor Function System, which is explained later in this section). On the other hand, this group and other experts [7; 9; 11] recommend not to use the traditional classification of limb distribution (hemiplegia, diplegia and tetraplegia) because of the low inter-rater reliability and the disagreements between the different registers [9]. Nonetheless, this change has been questionable as there is plenty of literature based in these traditional terms [11; 12]. However, experts recommend that if this classification is used, a description of the motor types in all body parts is suggested [7]. Diplegia consists of gross motor problems marked predominantly in the lower limbs [13] while children with hemiplegia have limitations on one side of the body. Children with triplegia and quadriplegia have severe motor involvement, with three and four limbs affected, respectively [13].

Table 2.1: **Classification of motor type**

Definitions adopted for European classification of cerebral palsy
Spastic cerebral palsy is characterized by at least two of: Abnormal pattern of posture and/or movement Increased tone (not necessarily constant) Pathological reflexes (increased reflexes: hiperreflexia and/or pyramidal signs e.g. Babinski response) ***Spastic cerebral palsy may be either bilateral or unilateral:*** Spastic bilateral CP is diagnosed if: Limbs on both sides of the body are involved Spastic unilateral CP is diagnosed if: Limbs on one side of the body are involved
Ataxic cerebral palsy is characterized by both: Abnormal pattern of posture and/or movement. Loss of orderly muscular coordination so that movements are performed with abnormal force, rhythm, and accuracy
Dyskinetic cerebral palsy is dominated by both: Abnormal pattern of posture and/or movement Involuntary, uncontrolled, recurring, occasionally stereotyped movements ***Dyskinetic cerebral palsy may be either dystonic*** **or choreo-athetotic:** *Dystonic cerebral palsy is dominated by both:* Hypokinesia (reduced activity, i.e. stiff movement) Hypertonia (tone usually increased) ***Choreo-athetotic cerebral palsy is dominated by both*** Hyperkinesia (increased activity, i.e. stormy movement) Hypotonia (tone usually decreased)

Extracted from [10].

2.1.1 Gross Motor Function Classification System

The Gross Motor Function Classification System (GMFCS) [14] was a major step forward as it is an standardized system that reveals the severity of movement disability. In this classification system, we can distinguish five groups conforming to the children's level of mobility. Level I represents a person that has minimal disability and level V means a severe limitation based on the total dependence on a wheelchair to maintain posture (see table 2.2). A pictorial representation can be found in Appendix A, for ages 6 through 12 and for ages 12 through 18. The GMFCS generally remains stable during childhood [15]. Yet, sometimes improvements or deteriorations in the child's GMFCS level can occur.

Table 2.2: **GMFCS between 6th-18th birthday**

Level	Descriptors between 6th-12th birthday	Descriptors between 12th-18th birthday
GMFCS I	Children walk at home, school, outdoors and in the community. They can climb stairs without the use of a railing. Children perform gross motor skills such as running and jumping, but speed, balance and coordination are limited.	Youth walk at home, school, outdoors and in the community. Youth are able to climb curbs and stairs without physical assistance or railing. They perform gross motor skills such as running and jumping, but speed, balance and coordination are limited.
GMFCS II	Children walk in most settings and climb stairs holding onto a railing. They may experience difficulty walking long distances and balancing on uneven terrain, inclines, in crowded areas or confined spaces. Children may walk with physical assistance, a hand-held mobility device or used wheeled mobility over long distances. Children have only minimal ability to perform gross motor skills such as running and jumping.	Youth walk in most settings but environmental factors and personal choice influence mobility choices. At school or work they may require a hand held mobility device for safety and climb stairs holding onto a railing. Outdoors and in the community youth may use wheeled mobility when traveling long distances.
GMFCS III	Children walk using a hand-held mobility device in most indoor settings. They may climb stairs holding onto a railing with supervision or assistance. Children use wheeled mobility when traveling long distances and may self-propel for shorter distances.	Youth are capable of walking using a hand-held mobility device. Youth may climb a railing with supervision or assistance. At school they may self-propel a manual wheelchair or use powered mobility. Outdoors in the community youth are transported in a wheelchair or use powered mobility.
GMFCS IV	Children use methods of mobility that require physical assistance or powered mobility in most settings.They may walk for short distances at home with physical assistance or use powered mobility or a body support walker when positioned. At school, outdoors and in the community children are transported in a manual wheelchair or use powered mobility.	Youth use wheeled mobility in most settings. Physical assistance of 1-2 people is required for transfers. Indoors, youth may walk short distances GMFCS IV with physical assistance, use wheeled mobility or a body support walker when positioned. They may operate a powered chair, otherwise are transported in a manual wheelchair.
GMFCS V	Children are transported in a manual wheelchair in all settings. Children are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements.	Youth are transported in a manual wheelchair in all settings. Youth are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements. Self-mobility is severely limited, even with the use of assistive technology.

Extracted from [10].

Chapter 3

Estimating energy requirements

It is well known that people with cerebral palsy (CP) have different energy requirements (ER) compared to typically developing children (TDC). Thus, energy needs are challenging to assess due to the heterogeneity of the disease and currently there is no consensus regarding which method is the best for estimating them [16].

Poor nutrition and growth are commonly reported and many children with CP are shorter and thinner than similarly aged TDC [16]. Therefore, standard growth charts may not be useful in children with CP whose growth patterns are often notably different from the general population. CP-specific growth charts have also been developed, however, they describe growth which is not necessarily ideal as they also consider many children with health issues affecting growth, especially malnutrition [17].

Feeding and gastrointestinal difficulties (oropharyngeal dysfunction, drooling, foregut dysmotility, gastro-oesophageal dysmotility, retching, delayed gastric emptying and chronic constipation) are also frequent in children with CP and if they are not appropriately managed can result in a reduced quality of life, undernutrition and poor growth [1]. In this group of children, the feeding process is frequently prolonged, stressful for the child and the caregiver, and slow, causing weight loss that further increases ER [4].

Additionally, it has been reported a reduced quality of life in children with CP and their carers and family. The degree to which it is reduced is related to the severity of the disease [17; 18]. Sometimes this group of children are not able to communicate their hunger and satiety that is why caregivers and health professionals must be informed about their specific ER [4].

A multidisciplinary team experienced in the management of nutritional problems in children with CP including occupational therapists, psychologists, speech therapists, dietitians, physicians and nurses [17] is the best way to improve their quality of life and nutritional status. Therefore, the estimation of ER is the first step towards a personalized nutritional intervention.

3.1 Measured resting energy expenditure and total energy expenditure in subjects with cerebral palsy

Eight studies which measured resting energy expenditure (REE) and total energy expenditure (TEE) in children with CP have been reviewed. Methods employed for measuring REE and TEE were: doubly labeled water (DLW), indirect calorimetry (IC), and BIA QuadScan® 4000 equipment.

IC is the reference method for measuring REE and it uses the relationship between oxygen consumption and carbon dioxide production measured in expired breath samples to determine substrate oxidation and thereby REE [3]. Furthermore, the gold standard to assess TEE is the DLW method. This technique uses stable isotopes (tracers) of deuterium and labeled oxygen and provides accurate and reliable measurements of TEE and total body water (TBW) [19]. Finally, the BIA QuadScan® 4000 equipment has shown a good correlation between anthropometric indicators and body composition measurement in children with SCP.

Table 3.1 shows mean measured REE and/or TEE expressed in kilocalories. Results have demonstrated a lower REE and TEE in children with CP compared to healthy children (data not shown for TDC). The causes of these differences are explained further on (see section 3.2.1). Children with spastic quadriplegic CP had the lowest REE and TEE and ambulatory children with bilateral spastic CP had the highest REE and TEE.

Table 3.1: **Measured REE and TEE in subjects with SCP**

Author	Method	Subjects	Mean Measured REE and/or TEE (kcal)
Bell et al. (2010) [16]	IC (REE) DLW (TEE)	16 ambulatory children with BSCP aged 5-12y	REE = 1074.33±167.91 TEE= 1674.79±302.86
Walker et al. (2012) [6]	DLW	26 preschool children with SCP aged 2.9-4.3y	TEE= 1367 kcal±329.17
Iñíguez et al. (2018) [20]	BIA Quadscan® 4000 equipment	79 SCP subjects aged 24m-16y 9m	REE= 890.21 TEE= 1231.29
Penagini et al. (2018) [21]	IC	54 children with SQCP aged 24-18y	REE=876 (699;1229)
Lee et al. (2011) [3]	IC	15 children with SQCP	REE= 800.50±295, 70
Azcue et al. (1996) [22]	IC	13 nonambulatory children with SQCP aged 2-16y	REE=760±241
Stallings et al. (1996)[23].	IC (REE) DLW (TEE)	61 children with SQCP aged 2-18y	REE= 826.40±254.85 TEE=961.31 ±258.91

All values are means. Most of them are expressed as standard deviation (SD). Pegagini et al.'s means are expressed as interquartile range (IQR). BIA=Biolectrical Impedance Analysis; BSCP=Bilateral Spastic Cerebral Palsy; DLW= Doubly Labeled Water;GMFCS= Gross Motor Function System; IC= Indirect calorimetry; m=months; REE= Resting Energy Expenditure; SCP= Spastic Cerebral Palsy; SQCP= Spastic Quadriplegic Cerebral Palsy; TEE= Total Energy Expenditure; y= years.

Walker et al. [6] and Rieken et al. [24] measured TEE depending on Level of GMFCS (see Table 3.2). Both studies showed a decline in the TEE with increasing GMFCS level. These values demonstrate that as a child becomes more impaired in the overall gross motor function, their ER are decreased [6].

Table 3.2: **Measured TEE depending on Level of GMFCS**

Author	Subjects	Level of GMFCS	TEE (kcal)
Walker et al. [6]	32 children with CP aged 2.9-4.4y	GMFCS I and II (n=16) GMFCS III, IV and V (n=16)	1442.63 ±228 1041.13 ±239
Rieken et al. [24]	61 children with severe neurological impairment aged 2-19y	GMFCS IV (n=6) GMFCS V (n=55)	1619 ±417 1103 ±398

All values are expressed as mean (kcal) ± standard deviation (SD). CP=Cerebral Palsy; GMFCS=Gross Motor Function System; NI= Neurological Impairment.

Similar results were observed when considering the number of limbs involved. Walker et al. [6] and Bell et al. [16] measured TEE depending on involved limbs via the DLW method (see Table 3.3). A trend toward lower ER was observed when the number of limbs involved increased [6].

Table 3.3: **Measured TEE depending on limbs involved**

Author	Subjects	Limbs involved		
		Diplegia	Hemiplegia	Triplegia or Quadriplegia
Walker et al. (2012) [6]	32 children with CP aged 2-9-4.4y	1305.53 ±336	1456.48 ±184	1041.37±250
Bell et al. (2010) [16]	16 children with mild CP aged 5-2-y	1061.19±356	1748.35±221	-

All values are expressed as mean (kcal) ± standard deviation (SD). CP=Cerebral Palsy; y=years.

Motor types were also considered but statistical comparisons could not be made due to the small simple size of children with dyskinesia and ataxia in comparison with spasticity [6] (data not shown). Further research is needed in order to determine the influence of motor type on ER.

3.2 Available equations for estimating energy requirements

Several equations are used to estimate ER (e.g., World Health Organization (WHO) Committee [25] and Schofield et al. [26]). Nevertheless, they are based in healthy children and consequently tend to overestimate calorie needs in children with severe CP [24] even in the absence of the physical activity coefficient or physical activity level. Overestimating energy needs can induce to overfeeding, fat accumulation and overweight [27].

Also, CP-specific equations have been developed [28; 29] but their effectiveness remains questionable [24]. Krick et al. [29] proposed an equation which required IC and estimated energy needs by including factors for muscle tone, movement or level of activity and catch-up growth (see Table 3.4) [29]. This CP-specific equation was later demonstrated to be inaccurate because it overestimated ER in comparison with the reference method (DLW) [24]. The second and third methods shown in Table 4.1. are extracted from a study by Culley and Middleton [30] and Azcue et al. [22], respectively. The European Society for Paediatric Gastroenterology (ESPGHAN) suggests the use of all these equations but it emphasises that these formulas are only a starting point and the effect of dietary intervention must be assessed [17].

Table 3.4: **Equations for estimating ER in neurological impaired patients**

Method	Equation
1. Krick method [29]	Energy intake(kcal/day) = [BMR x muscle tone x activity] + growth Muscle tone: 0.9 if decreased, 1.0 if normal, and 1.1 if increased Activity: if bedridden, 1.2 if dependant, 1.25 if crawling, 1.3 if ambulatory Growth: 5 kcal/g of desired weight gain (normal and catch-up growth)
2. Height-based method [30]	14.7 kcal/cm in children without motor dysfunction 13.9 kcal/cm in ambulatory patients with motor disfunction 11.1 kcal/cm in nonambulatory patients
3. DRI method [22]	1.1 x measured resting energy expenditure (REE), where REE is: Male: 66.5 + (13.75 x weight in kg) + (5.003 x height in cm) - (6.775 x age) Female: 65.1 + (9.56 x weight in kg) + (1.850 x height in cm) - (4.676 x age)

BMR= basal metabolic rate;
REE= resting energy expenditure.
Extracted from [17].

More recently, Rieken et al. [24] created two specific models for estimating energy expenditure in children with severe CP that are explained further on (See Table 3.5).

3.2.1 Factors which influence energy requirements

The initial brain damage and therefore the motor type, distribution and severity of impairment can modify TEE and influence movement patterns and muscle function [6]. Moreover, other factors which influence ER and need to be taken into account are: level of physical activity, altered body composition and malnutrition [27].

3.2.1.1 Body Composition

It has been shown that the best predictor of energy needs in children with CP is fat-free mass (FFM), followed by ambulatory status (ambulant and marginally ambulant or nonambulant) [6; 17]. There is a distinction between FFM and muscle mass. Thus, FFM is also known as lean free mass and it describes all body compartiments except fat, while muscle mass only includes the weight of the muscles. This distinction must be kept and the two must not be confused with one another.

Generally, children with CP have higher percentage of body fat and lower FFM than healthy children [31]. This relatively low FFM may be caused because of wasted muscles and reduced bone mass as a consequence of their immobility [24]. Also, children with severe CP (GMFCS IV-V) may have more intra-abdominal fat than subcutaneous fat [32]. Moreover, although the differences in body composition were more obvious in non-ambulatory children than in ambulatory children, they existed in both groups of children. Furthermore, their reduced muscle mass together with loss of mobility and lower fitness can lead to an increased risk of obesity [28; 33]. Body fat assessment can help to differentiate between underweight patients but nutritionally replete and malnourished patients [15].

Walker et al. [6] found a positive strongly correlation between ER and FFM. Other studies support these findings [20]. In addition, Gurka et al. [28] created CP-specific equations for calculating body composition with the aid of skinfold-thickness measurements. However, they were not cross-validated and Rieken et al. [24] found that the percentage of body fat was overestimated when compared to isotope dilution.

3.2.1.2 Ambulatory status

GMFCS is an important predictor of physical activity in children with CP [34]. Thus, children and adolescents with CP who have walking disabilities have lower levels of physical activity and vice versa. For this reason, ambulatory status can tell us about the amount of daily physical activity and therefore it can influence ER.

Ambulatory children

Ambulatory children with CP have a REE roughly equivalent to that of TDC [1]. Also, different studies found that the TEE was reduced in children with CP compared to TDC mainly because they had lower activity [16; 35]. However, another study based on preschool children with CP [6] found that ER in ambulant children were similar to those TDC. These variations between studies could be partially related to the differences in the structure and purpose of physical activities in younger children [6].

Several studies have demonstrated that ambulatory children with CP expend more energy walking and walk at a lower speed in comparision with TDC [16; 35; 36]. Furthermore, as stated Okur et al. [35] in a study with children with CP and level I and II according to GMFCS, the CP group spent less time and expended less energy in vigorous games. Also, the energy cost (EC) increased in children with diplegia compared to children with hemiplegia [16] and with larger GMFCS [37; 38; 39; 40]. These outcomes indicate that the CP participants cannot reach the same level of physical activity as a result of a poor physical condition [35]. Increased EC was also demonstrated not only during walking but also while standing in a few studies in children with SCP [41; 42]. These findings suggest that modifying the child's position to standing could reduce sedentary behaviour.

Finally, in a cross-sectional study [37] that aimed to compare EC between children and young adults with CP (GMFCS levels I-III) and typically developing peers found that although the EC outcomes were higher for CP participants, TDC and CP peers showed a similar decline in EC when they got older or taller. These findings indicated that it is necessary to evaluate EC and correct this decrease over time. However, further studies should confirm these results.

Marginally ambulatory and nonambulatory children

As explained in the previous section, energy expenditure and physical activity levels in children with CP decline as the severity of the GMFCS increases, as a consequence of movement limitations [37; 38; 39; 40]. Thus, nonambulant and marginally ambulant children are more likely to have lower FFM due to reduced physical activity [23; 24] and have lower ER in comparison with children with major function [24]. Furthermore, ER in children with severe CP who use a wheelchair to get around have been accounted to be 60-70% of those TDC [17; 24].

Rieken et al. [24] established two group-specific equations based in nonambulant children (GMFCS level IV and V) with severe CP for estimating energy expenditure. The first model used a multiple of TBW measurements and performed best according to the authors. However, it might be worse to put into practice because a bioelectrical impedance analysis (BIA) machine is needed and not every professional has it. The second model was based in Schofield equations using weight with corrections for GMFCS level and physical activity. Therefore, if the child has a GMFCS level V plus a low degree of movement, 280 kcal should be subtracted. If the child has a high degree of movement 222 kcal should be added. If the GMFCS level is IV, 431 kcal should be added [19]. Table

3.5 shows the two equations developed by Rieken et al. used in children with severe neurological impairment.

Table 3.5: **Newly developed equations based on Schofield's equation and based on TBW measurements.**

	Model with Schofield equation	Model using TBW from DLW method
Age 3-9y		
M	BMR=(0.095 x weight, in kg) + 2.110	
F	BMR=(0.085 x weight, in kg) + 2.033	
Age 10-18y		
M	BMR=(0.074 x weight, in kg) + 2.754	
F	BMR=(0.056 x weight, in kg) + 2.898	
Equation for TEE (kcal)	1.1 x BMR x 238.8	60.7 x TBW (kg)
Additional corrections		
General correction	-280 kcal	+175 kcal
High degree of movement	+222 kcal	+344 kcal
GMFCS level 4	+431 kcal	+194 kcal

BMR= basal metabolic rate (in mJ); DLW= doubly labeled water; GMFCS= Gross Motor Function Classificaction System; TEE= total energy expenditure.
Extracted from [24]

Nonetheless, it would be useful to cross-validate these equations in a comparable CP-group before they can be recommended in clinical purposes [19]. Walker et al. [6] compared the 2 prediction equations with the data from the nonambulant children of their study and found an underestimation in ER in their population of ~22%, however, it has to be taken into account that their population was younger compared to Rieken et al.'s CP group.

3.2.1.3 Motor type

The motor type affects patterns of movement and function and therefore ER vary with specific CP types [4; 6]. Yet, obtaining evidences is difficult for two reasons [6]:

Firstly, the prevalence of the motor types of dyskinesia and ataxia in comparison with spasticity is low. Secondly, a large enough sample size is needed in order to provide statistically significant results, which requires expensive methodological procedures [6].

However, Rieken et al. [24] found that children with mixed muscle tone had increased ER than children with either hypertonia or hypotonia in a study of children with severe neurological impairment but these outcomes were not added to the amount of varation explained in energy expenditure measured by a DLW method.

Spastic cerebral palsy

Several studies have shown that children with spastic cerebral palsy (SCP) have significantly less ER than TDC [6; 21; 22].Thus, immobile children with SCP often become obese despite their decreased caloric intake normally applicable for weight loss [4].

Azcue et al. [22] found a poor relationship between REE, weight and height and intracellular water in the spastic quadriplegic CP group. Also, they found that REE was significantly lower than in control children and the WHO equations overestimated REE.

More recently, Penagini et al. [21] reached similar conclusions noting that the 5 most commonly used prediction equation formulae (i.e WHO, Harris-Benedict, Schofield, and Oxford formulae) gave imprecise estimates and overestimation was more common than underestimation with the risk of overfeeding most spastic quadriplegic CP children. They also developed a new population-specific formula (see 3.1) but despite being accurate at the population level, it didn't performed better than the other mentioned equations.

$$REE\ (kcal/day) = 24 \cdot weight\ (kg) + 380 \tag{3.1}$$

Another cross-sectional study led by Iñiguez et al. [20] which included 79 participants with SCP from 24 to 16 years nine months found that the mean TEE in their population was 1231 kcal/day, similar to the mean estimated by Walker et al.'s [6] mean which was 1367 kcal/day. The results were really similar to those outcomes obtained by Walker et al. Finally, the study concluded that their estimated energy expenditure (11 kcal/cm/day) would be an applicable and reliable method for estimating ER. However, more accurate approaches are needed in order to estimate ER in this group of children.

Dyskinetic cerebral palsy

As already mentioned, TEE has been shown to be significantly lower in most children with CP. Conversely, children with athetosis (a symptom of dyskinesia) have involuntary movements and therefore it has been hypothesized that they have similar or even increased ER in comparison with TDC [4]. Similarly, Johnson et al. [43] observed that ER were higher in the subjects with dyskinetic cerebral palsy (DCP) than in the subjects without DCP. However, energy expended in physical activity tended to be lower in adults with DCP concluding that the 2 associations removed each other out.

A cross-sectional study which investigated the relationship of dystonia and choreathetosis with activity [44] found lower functional abilities with increasing dystonia in children with DCP. Furthermore, it has been reported that most of the DCP population is confined to wheelchair and therefore is classified at level IV and level V of the GMFCS [44].

Moreover, a population based study from Sweden found that more than half of patients were underweight, probably because of the high energy expenditure owing to involuntary movements combined with poor food intake and gastrointestinal problems [45; 46]. Consequently, ER of ambulatory children with DCP are expected to be comparable to those of healthy children due to an increased activity [4].

In conclusion, few studies have analyzed the functional abilities and activity levels in DCP [44] and its relationship with ER. Furthermore, obtaining evidence is difficult as the prevalence of dyskinesia in comparison with spasticity is low and getting a large enough sample size to provide statistically significant results is challenging [6]. For this reason, further research is needed in order to estimate ER in children with DCP.

3.2.1.4 Distribution of limbs

Walker et al. [6] hypothesized that the distribution of CP had an influence in physical activity and consequently in ER in preschool age children with CP. Therefore, their results showed a trend toward lower ER with an increased number of limbs involved. These outcomes are compatible with other findings as children with triplegia or quadriplegia tend to be shorter and lighter and have lower FFM index and ER than children with hemiplegia [6]. Besides, they are more likely to have higher GMFCS levels and increased movement disabilities.

Chapter 4

Discussion

ER are challenging to estimate in children with CP. The calculation of REE and TEE is the first step towards a personalized nutritional intervention [21].

First of all, a review of the different studies that measured REE and TEE in subjects with CP was made. The methods employed for determining energy expenditure were the gold standard DLW, the reference method IC and a useful equipment for estimating REE and TEE called BIA QuadScan® 4000 equipment. Results showed a variable mean measured REE and TEE and significant differences depending on the level of GMFCS and distribution of limbs (diplegia, hemiplegia and triplegia or quadriplegia). Statistical comparisons between the different motor types could not be made because of the small sample sizes of children with dyskinesia and ataxia in contrast with spasticity. These outcomes demonstrated that ER in this population are different in comparison with healthy children and may vary depending on their specific status. On the other hand, several specific and general equations have been examined. It has been shown that equations based in healthy children such as WHO Comitte and Schofield tend to overestimate energy needs in children with severe CP [24]. However, the ESPGHAN stated that Schofield equation is reasonable in estimating calorie needs [17]. Furthermore, Krick et al. [29] developed an equation that was later proven to be inaccurate in children with severe CP because it also overestimated ER [24]. Interestingly, Rieken et al. [24] developed two group-specific equations for children with severe motor impairment (GMFCS level IV and V) that provided reasonable energy although there is a need to cross-validate them.

With reference to factors which influence ER, body composition, ambulatory status, motor type, and distribution of limbs have been considered. The results have shown that the greatest predictor of ER is FFM, followed by ambulatory status [6]. Ambulatory children with CP expend more energy during walking even when they walk at a lower speed compared to TDC. Conversely, marginally ambulatory and nonambulatory children with CP have lower FFM and physical activity leading to lower ER in comparison with children with higher function. Furthermore, children with spasticity have lower ER than healthy children and often become obese despite reduced caloric intake normally appropiate for weight reduction [4]. On the contrary, it has been hypothesized that children with dyskinesia have similar or even greater ER due to increased involuntary movements at rest. No data was found regarding children with ataxia possibly due to its low prevalence.

Chapter 5

Contributions and suggestions to the topic

In order to provide a starting point to estimate ER in health care centers and special education schools, I have elaborated a summary table of the main specific equations available for children with cerebral palsy and a proposal of a decision tree considering the GMFCS (See Table B.1 and Figure B.1 in Appendix B).

It is noteworthy that these equations should be applied with caution because, as explained above, if they are not applied correctly they can underestimate or overestimate ER. In this sense, if the child presents physical signs of undernutrition or overnutrition, ER must be individualized.

With this work, I have noticed that depending on the ambulatory status of the child, some equations are more effective than others. In this sense, the decision tree can be a useful tool to get an initial idea of the energy needs in children with CP in health centers or schools that do not have reference methods such as IC or DLW. An advantatge of this decision tree is that it considers the influence of the level of the GMFCS, and consequently, the functional status of the child. Paying attention to the ambulatory status of this group of children may help to estimate ER more accurately.

Nevertheless, this decision tree has limitations when calculating requirements at an individual level. For instance, motor type and distribution of limbs also play an important role in estimating ER. However, although it is known that ER in ambulatory children with DCP are expected to be higher in comparison with ambulatory children with SCP, no population-specific equations for children with DCP were found. The same applies to distribution of limbs. For this reason, further studies are required to reach a consensus on which are the more appropriate equations considering factors that could alter ER.

Chapter 6

Conclusions

In this thesis, we have reviewed what is known about ER of children with CP. Thus, a bibliographic research considering the most commonly used prediction formulae and the main factors which alter ER in children with CP has been carried out with the aim of providing a tool to assess ER.

Unfortunately, on the basis of the foregoing, it can be concluded that estimating ER in this children is not straightforward for a variety of reasons. First, it has been observed that children with cerebral palsy have different ER than TDC and therefore methods that estimate energy expenditure in healthy children can overestimate caloric requirements of CP patients. In addition, CP covers a heterogeneity of disorders and concomitant diseases that further complicate the estimation of energy needs. Finally, this high degree of variability in this group of children makes it difficult to reach a consensus and provide general guidelines for ER.

Several equations have been proposed, all of which have limitations. However, bearing in mind that in most health care centers or pediatric services there is no access to calorimeters for calculating energy needs (and even less in schools for children with neurological disabilities), CP prediction formulas can provide a tool to guide health professionals to estimate ER. In this sense, the decision tree created in this thesis could give an initial idea of their ER depending on the functional status of the child. Furthermore, taking into account factors which influence ER in this population may help to provide more precision. Results have shown that FFM and ambulatory status are the best predictors of energy needs.

At present, ongoing assessment and monitoring is the key to ensure that the child is meeting their requirements. Having knowledge about ER in children is crucial as inappropriate energy intake is the main cause of undernutrition, growth failure, and overweight in children with cerebral palsy. Findings of how ambulatory status, FFM, distribution of limbs, and motor type may alter energy needs values can be incorporated into daily clinical practice and early intervention strategies. Further studies are required to develop more population-specific formulas, taking into consideration the factors that may influence ER in children with CP.

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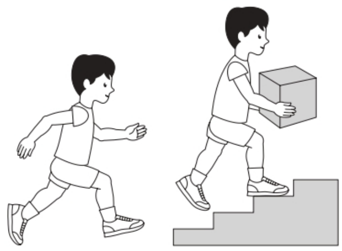
Appendices

Appendix A

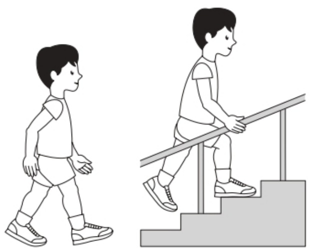
Gross Motor Function Classification System Illustrations

GMFCS E & R between 6th and 12th birthday: Descriptors and illustrations



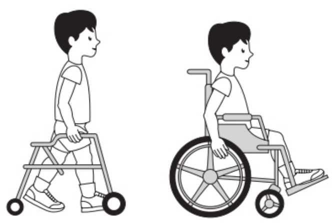
GMFCS Level I

Children walk at home, school, outdoors and in the community. They can climb stairs without the use of a railing. Children perform gross motor skills such as running and jumping, but speed, balance and coordination are limited.



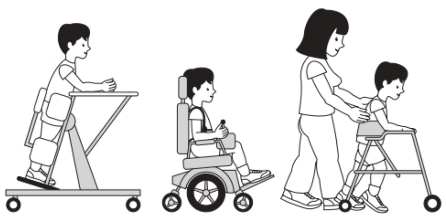
GMFCS Level II

Children walk in most settings and climb stairs holding onto a railing. They may experience difficulty walking long distances and balancing on uneven terrain, inclines, in crowded areas or confined spaces. Children may walk with physical assistance, a hand-held mobility device or used wheeled mobility over long distances. Children have only minimal ability to perform gross motor skills such as running and jumping.



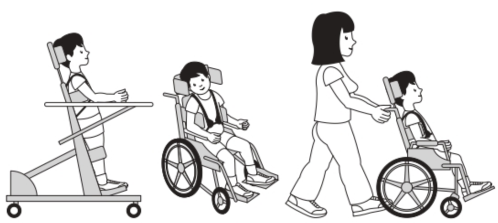
GMFCS Level III

Children walk using a hand-held mobility device in most indoor settings. They may climb stairs holding onto a railing with supervision or assistance. Children use wheeled mobility when traveling long distances and may self-propel for shorter distances.



GMFCS Level IV

Children use methods of mobility that require physical assistance or powered mobility in most settings. They may walk for short distances at home with physical assistance or use powered mobility or a body support walker when positioned. At school, outdoors and in the community children are transported in a manual wheelchair or use powered mobility.



GMFCS Level V

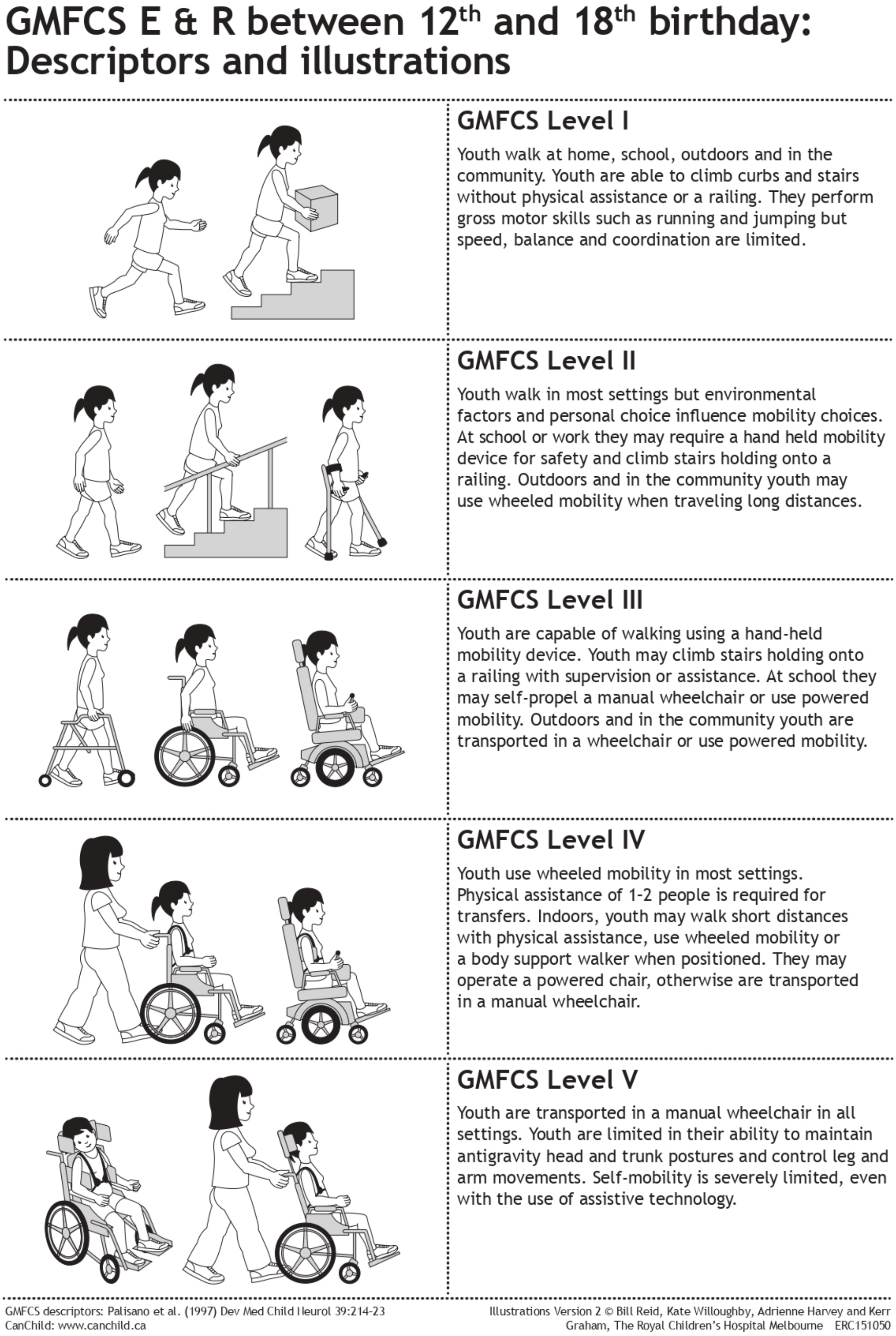
Children are transported in a manual wheelchair in all settings. Children are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements.

GMFCS descriptors: Palisano et al. (1997) Dev Med Child Neurol 39:214-23
CanChild: www.canchild.ca

Illustrations Version 2 © Bill Reid, Kate Willoughby, Adrienne Harvey and Kerr Graham, The Royal Children's Hospital Melbourne ERC151050

Figure A.1: **GMFCS aged 6-12**

Extracted from [10]



GMFCS E & R between 12th and 18th birthday: Descriptors and illustrations

GMFCS Level I

Youth walk at home, school, outdoors and in the community. Youth are able to climb curbs and stairs without physical assistance or a railing. They perform gross motor skills such as running and jumping but speed, balance and coordination are limited.

GMFCS Level II

Youth walk in most settings but environmental factors and personal choice influence mobility choices. At school or work they may require a hand held mobility device for safety and climb stairs holding onto a railing. Outdoors and in the community youth may use wheeled mobility when traveling long distances.

GMFCS Level III

Youth are capable of walking using a hand-held mobility device. Youth may climb stairs holding onto a railing with supervision or assistance. At school they may self-propel a manual wheelchair or use powered mobility. Outdoors and in the community youth are transported in a wheelchair or use powered mobility.

GMFCS Level IV

Youth use wheeled mobility in most settings. Physical assistance of 1-2 people is required for transfers. Indoors, youth may walk short distances with physical assistance, use wheeled mobility or a body support walker when positioned. They may operate a powered chair, otherwise are transported in a manual wheelchair.

GMFCS Level V

Youth are transported in a manual wheelchair in all settings. Youth are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements. Self-mobility is severely limited, even with the use of assistive technology.

GMFCS descriptors: Palisano et al. (1997) Dev Med Child Neurol 39:214-23
CanChild: www.canchild.ca

Illustrations Version 2 © Bill Reid, Kate Willoughby, Adrienne Harvey and Kerr Graham, The Royal Children's Hospital Melbourne ERC151050

Figure A.2: **GMFCS aged 12-18**

Extracted from [10]

Appendix B

Contributions and suggestions to the topic

Table B.1: **Summary table of the main equations available for children with CP**

Authors	Subjects	Equations/methods
Krick et al. [29] (Indirect calorimetry)	Tube-fed children with CP	Energy intake(kcal/day) = [BMR x muscle tone x activity] + growth *Muscle tone*: 0.9 if decreased, 1.0 if normal, and 1.1 if increased *Activity*: if bedridden, 1.2 if dependant, 1.25 if crawling, 1.3 if ambulatory
Culley et al.[30] (Height-based method)	Children without motor dysfunction, ambulatory patients with motor dysfunction and nonambulatory patients	14.7 kcal/cm in children without motor dysfunction 13.9 kcal/cm in ambulatory patients with motor dysfunction 11.1 kcal/cm in nonambulatory patients
Azcue et al. [22] (DRI method)	Children with SQCP	1.1 x measured resting energy expenditure (REE): *Male*: REE = 66.5 + (13.75 x weight in kg) + (5.003 x height in cm) - (6.775 x age) *Female*: REE = 65.1 + (9.56 x weight in kg) + (1.850 x height in cm) - (4.676 x age)
Rieken et al. [24] (Model with Schofield equation)	Nonambulant children with severe CP GMFCS (IV-V)	*Age 3-9y*: *Male*: BMR=(0.095 x weight in kg) + 2.110 *Female*: BMR=(0.085xweight in kg) +2.033 *Age 10-18y*: *Male*: BMR=(0.074x weight in kg) +2.754 *Female*: BMR=(0.056 x weight in kg) +2.898 TEE (kcal)= 1.1 x BMR x 238.8 *Additional corrections*: General correction -280 kcal High degree of movement: +222 kcal GMFCS level IV: +431 kcal
Rieken et al. [24] (Model using TBW from DLW method)	Nonambulant children with severe CP (GMFCS IV-V)	TEE= 60.7 x TBW *Additional corrections*: General correction +175 kcal High degree of movement: +344 kcal GMFCS level IV: +194 kcal
Penagini et al. [21]	Children with SQCP	REE (kcal/day) = 24 x weight (kg) + 380

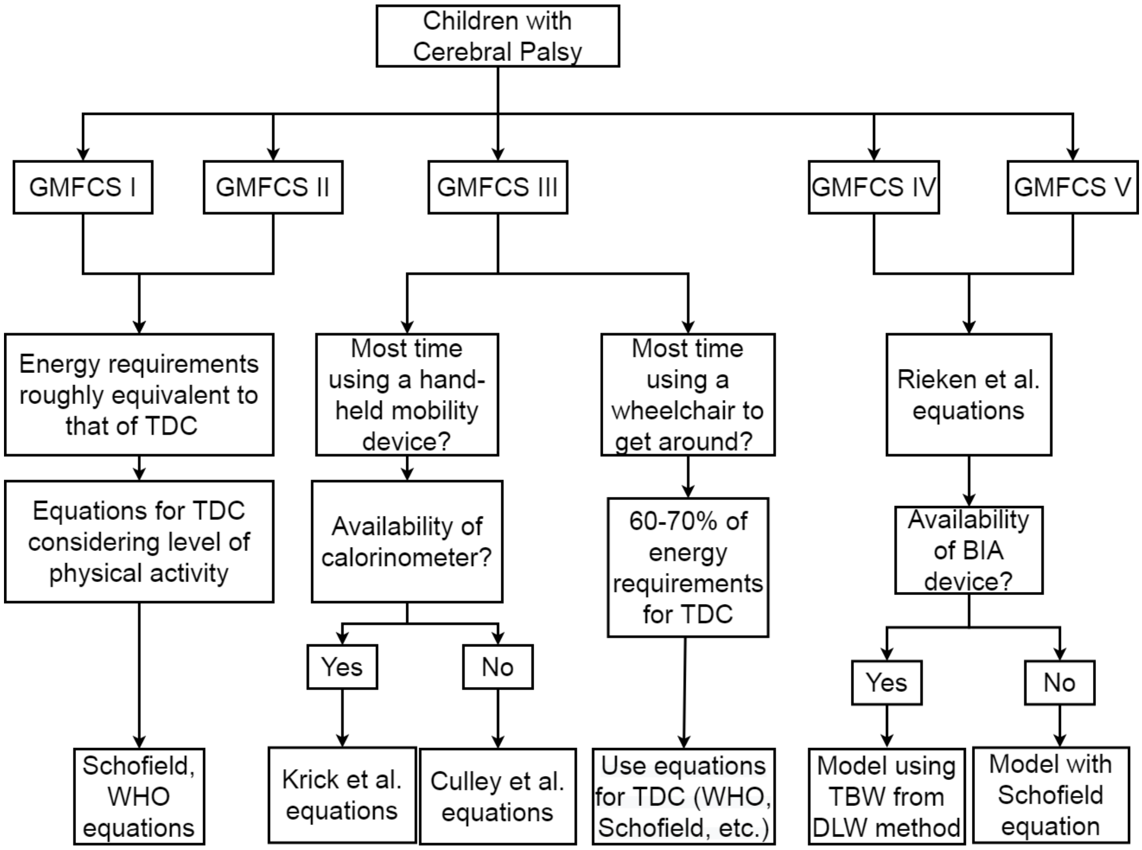


Figure B.1: **Decision tree considering the level of GMFCS**
If a child is growing too fast, then calories can be decreased by 10%. If a child grows slowly, then calories can be increased by 10% with careful monitoring. [47] Abbreviations: BIA= Bioelectrical Impedance Analysis; GMFCS= Gross Motor Function System; TDC=Typically Developing Children