Should we be testing and training muscle strength in cerebral palsy?

Annotation

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Cerebral palsy (CP) is a collection of disorders characterized by an insult to the developing brain that produces a physical disability as the primary or distinguishing feature. The spastic form of CP is most common and in those patients, additional clinical signs may include muscle shortening, diminished selective control, and weakness. The recognition of weakness as a component of CP has been longstanding as evidenced by the names given to this disorder and its subtypes. 'Cerebral palsy' means weakness originating from the brain, and the use of the suffixes 'plegia' or 'paresis' also indicate that weakness is a prominent feature. More than 50 years ago, Phelps contended that resisted exercise 'to develop strength or skill in a weakened muscle or an impaired muscle group' was an integral part of treatment in CP. (p 59)[1] Since that time, physical educators have also advocated strengthening.[2–8] Yet for years, conventional clinical wisdom in physical therapy argued against the use of strength testing and training in children with CP and, indeed, in all persons with CNS disorders. The rationale for this exclusion appears to be multifaceted. First, therapists were discouraged by the relatively meager functional responses to strength training in patients with spasticity compared with those with polio.[9] Clinicians also feared that strong near maximal effort would exacerbate spasticity and muscle tightness in those who were already 'stiffer' than normal.[10] Many also attested that impaired selective control in CP essentially prohibited performance of strengthening activities. Consequently, this approach was discarded in favor of a more direct focus on the brain. Only recently has strength testing and training experienced a resurgence in habilitation and rehabilitation programs for this population and for other spastic motor disorders. However, hesitation and even resistance to their incorporation are still encountered despite the lack of evidence to suggest that strengthening is detrimental in the presence of spasticity and accumulating evidence to support this type of exercise. The purpose of this annotation is to summarize existing research on strength testing and training, primarily focusing on CP and address the following clinical questions: (1) Can strength be measured reliably and in a valid way in cerebral palsy? (2) Is weakness a significant impairment in CP? (3) Is strength training effective in increasing force production and improving motor function and disability in CP? (4) Is strength training safe in the presence of spasticity and for children and adolescents, regardless of health status, who have an immature musculoskeletal system?

Can strength be measured reliably and in a valid way in CP?

Manual muscle testing in the grades above 'fair' involves isometric resistance of an examiner-imposed force.[11] An isometric contraction measures the ability of a muscle group to produce force without a change in overall muscle-tendon length; therefore, stretch responses should not be evoked. A maximal isometric contraction is only indicative of the capacity to produce force in that condition and at that particular muscle length, and cannot necessarily be extrapolated to conditions where the muscle length is different or changing throughout the task. Other factors besides muscle weakness, such as excessive cocontraction and impaired selective motor control, may inhibit the ability to produce agonist force. However, there is evidence that strength training, and not repetitive practice, can markedly increase the measured force in many individuals with CP. This points to weakness as a major factor in CP and argues for the validity of strength testing in CP and other spastic disorders.[6,12]

Isometric and isokinetic dynamometry testing, particularly at slower speeds, have been shown to be reliable in this population (even in children as young as 4 to 5 years of age) as well as in other spastic disorders for selected muscle groups.[13–16] Some normative data are also available in the literature for some age ranges, muscle groups, and testing protocols.[17–20]

There are practical difficulties in the measurement of strength in children with CP. The person being examined must be able to comprehend and repeatedly comply with producing maximal effort. Test positions may require some modifications in this population because of muscle shortening and the examiner must be careful not to exert a counter force at the point of joint contracture. Testing positions that promote or

inhibit the use of flexor or extensor synergies may also affect the strength values differentially in CP. Poor selective control in some muscle groups may prevent an individual from being able to perform the task,[20] although for less severely involved children with CP, motor control limitations are most likely not a substantial factor in the ability to generate force. As an example, in a lower-extremity strength investigation of children with mild to moderate spastic diplegia and hemiplegia who were tested in multiple muscle groups, the only muscle group and test position where selective control interfered with task performance was the ankle dorsiflexors which were tested with the knee in extension for only 2 of 30 participants.[20] However, motor control deficits may disrupt strength testing and training to a greater extent in those with greater neurological involvement.[21]

Some procedural factors that may enhance reliability of measurement in this and other populations include the use of the peak value achieved in a series of trials rather than a mean value across trials,[22] and use of a 'make' test where the person is asked to exert force against a rigid surface versus a 'break' test where the examiner attempts to exceed the resistance.[23] The point on the extremity where resistance is applied will also affect the measurement due to differences in the lever arm; therefore, data that will be compared across individuals or time periods may need to be reported as a torque measurement (force × lever arm) or the test position standardized with respect to the location of dynamometer placement.

Is weakness a significant impairment in CP?

While spasticity was once thought to be the primary contributor to the motor dysfunction noted in CP, many have challenged this perspective and now consider 'negative' signs such as muscle weakness to be more harmful to function.[24] Leg strength has been shown to be related to freely selected walking velocity and to the Gross Motor Function Measure in children and adolescents with CP.[25–27] For example, the amount of variance in walking speed that can be explained by muscle weakness was found to be 50% for a group of children with spastic diplegia and hemiplegia.[26]

Even children with CP who have mild disabilities demonstrate substantial weakness compared with age-related peers. [18,20,28,29] The lower level of physical activity observed in this population is one potential contributor to weakness,[30] but is hardly the sole explanation. Other possible factors include decreased central input to the muscle due to a pyramidal tract insult,[31] changes in the elastic properties of the muscles themselves,[32] aberrations in the reciprocal inhibition pathways in agonist–antagonist muscle pairs,[33] and heightened stretch responses or spasticity.[21] It is possible that some of the above factors may be secondary, rather than primary impairments, and may be preventable, at least in part, if sufficiently intense intervention is provided before these secondary factors ensue.

Weakness in CP may be exacerbated by procedures that address other impairments in these patients. In fact, none of the major neurosurgical or orthopaedic interventions that are prescribed in CP has a direct positive effect on muscle strength. Selective dorsal rhizotomy unmasks weakness by reducing antigravity support that may have been provided by spasticity.[34] Orthopaedic surgery that lengthens or transfers tendons may have a negative effect on the force production of the muscle addressed, at least in the short term.[35–37] Botulinum toxin directly and temporarily weakens the injected muscle to reduce its spasticity or over activity.[38] Intrathecal baclofen acts on contracted muscles to reduce spasticity and muscle spasms, and may have a direct negative effect on strength that warrants further exploration.[21,39] Each of these interventions may have an indirect positive effect on strength in the muscles opposite those that were spastic or short,[40] which could be enhanced even further by strength training. Other common treatments such as the use of orthoses or serial casting can also exacerbate weakness due to immobilization. Directly loading the muscle through specific exercises, activities, or sufficiently intense electrical stimulation is the only direct way to increase muscle strength in CP, and may be particularly useful in augmenting or maximizing the functional outcomes of other interventions that address different components of the motor disorder.

Is strength training effective in increasing force production and improving motor function and disability in CP?

In view of current models of disablement which have been popularized in recent years,[41,42] health professionals are encouraged to consider and hopefully measure the effects of their interventions at multiple levels to ensure that a change in an impairment has an appreciable effect on an individual's function or health-related quality of life. Reports to date consistently show that strengthening programs predictably increase the ability to produce force, and that training programs of short duration can improve gait, wheelchair propulsion, and other aspects of motor performance.[5,25,26,43–48] MacPhail and Kramer[46] reported positive functional effects, as measured by the Gross Motor Function Measure, from isokinetic training of knee flexors and extensors in adolescents with CP. Damiano and colleagues[25–26] have reported on two different isotonic training programs, one for the knee extensors alone and one for multiple lower extremity muscles, depending on individual areas of weakness, including hip flexors, extensors, and adductors, knee extensors, and ankle dorsiflexors and plantarflexors, each documenting improvement in strength and gait parameters. Darrah and colleagues[49] found that a community-based training program not only improved strength but also significantly enhanced the perceived physical appearance of a group of adolescents with CP.

The basis for producing strength gains in children with CP appears to be the same as those for people without chronic motor disorders. McCubbin and Shasby[6] in a randomized design showed that repetition alone without the use of resistance did not significantly improve torque production, suggesting that the physiological response to muscle loading, and not merely motor learning, is responsible for the increase in torque. Children with CP also appear to gain strength at the same rate as persons with weakness who have no CNS pathology in programs of similar intensity and duration.[18] The principles of strength training with respect to overload, progression, and specificity can be gleaned from the sports medicine literature[50–51] and require few if any modifications when designing programs for most persons with spastic CP.[52]

Many issues still remain unknown about strengthening in CP. More controlled studies need to be conducted to establish the efficacy of different types of strength training programs. Also, published reports on CP have focused primarily on persons with spasticity, with no available evidence to

date on the effectiveness of strength testing and training in those with extrapyramidal disorders.

In summary, strength training can have positive effects at the impairment level by increasing the muscle's capacity to exert force; at the functional level through improvements in gait and other motor skills; and at the personal, disability or societal level by enhancing fitness, participation,[30] and self-perception.

Is strength training safe in the presence of spasticity and in children and adolescents who have an immature musculoskeletal system?

While clinicians feared that exerting maximal effort could exacerbate spasticity, this has not been verified empirically. In a single case study, Horvat[4] found increased range of motion in a spastic muscle after strengthening its antagonist, which countered the suspicion of increasing muscle tightness resulting from strengthening. In another study, hamstring strength was measured before and after a quadriceps strengthening program in children with CP to determine whether the program caused an inadvertent increase in strength in the spastic muscle due to abnormal cocontraction or stretch responses elicited in the antagonist during agonist strengthening. The quadriceps showed a mean strength increase of more than 50% with no significant change in the hamstring values.[25]

Although it remains unknown whether spasticity causes reduced force production in the short or long term, some indirect evidence of a potential correlation has been noted. Eccentric weakness was shown to be less marked than concentric weakness, a difference that may be explained by heightened stretch responses in the muscle being lengthened which is the antagonist during concentric activation, but the agonist during eccentric activation.[53] Also, concentric torque is relatively more impaired with increasing movement speed which could also be explained by heightened stretch responses with increasing velocity.[53] Investigations have attempted to correlate the amount of resistance torque with the degree of weakness in both spastic antagonist and agonist muscles. Ross and Engsberg found no relation between strength and antagonist spasticity in a group of mildly involved children with CP.[54] However, in a sample of children with a broader range of involvement, those with greater muscle spasticity in the antagonist tended to have greater agonist weakness.[53] Since correlation procedures are particularly sensitive to truncated ranges in either or both variables, the marked differences across studies in the range of involvement and strength values may account for the seemingly disparate results.

The risks and benefits of strength training have been a controversial topic in normally developing children and adolescents, particularly during the period of maximal physical growth in puberty since the epiphyses may be particularly susceptible to injury at that time,[55–57] although the literature is neither extensive nor conclusive. The American Academy of Pediatrics has synthesized and summarized the available evidence, and has issued a policy statement to help guide clinicians.

Their general recommendation is as follows: 'Strength training programs for preadolescents and adolescents can be safe and effective if proper resistance training techniques and safety precautions are followed'. (p 1470) [56]

Until more definitive safety data are available, caution is urged when using maximal resistance or overloading the muscle in normally developing children and perhaps more particularly in those with musculoskeletal pathology such as CP. Further research on the optimal methods of strength training is also warranted particularly with respect to long-term implications for joints that may already be stressed by persistent abnormal loading patterns. For example, resistance programs that minimize the weight bearing stress on joints (e.g. water-based training programs) may be preferable for many of these individuals.

Conclusions

While therapists have been resistant to strength testing and training for several decades, others in the physical education and medical communities have not concurred with this viewpoint[2,4,6,58] and have continued to support and even promote strength and endurance testing and training in CP and other neuromuscular diseases. In the past decade, two literature reviews have been published on strength training in the physical therapy literature and concur that this is now an accepted therapeutic approach in these patients.[43,44] Much of what has been learned in CP in this area has also been mirrored by research on adults with spasticity and other disorders. As an example, in chronic stroke direct muscle strengthening improved functional performance in persons whose recovery had plateaued before this intervention, and was not shown to increase spasticity.[59]

Data on the specific treatment regimes to train differentially for strength, endurance, or power in this population, or on which muscles can and should be strengthened to impart the greatest functional benefit are not yet available specifically for CP. However, many useful guidelines may be found in the orthopaedic and athletic training literature.[50,51,60] Both absolute and relative strength across a joint should be considerations when designing protocols so as not to exacerbate muscle imbalance and contractures that may result from this.

Finally, medicine is focusing more on prevention of secondary impairments and on promotion of health and fitness in all children and adults, with increasing emphasis on populations such as those with disabilities who demonstrate health disparities. Strength and endurance training is an important component of fitness and these programs may promote more optimal health across the lifespan and increase participation in recreational, social, and occupational activities in children and adults with CP.

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Erratum

'Idiopathic Central Pontine Myelinolysis in Childhood'
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The published version of the Figure 3 caption in the October 2001 issue of DMCN was incorrectly labelled. The correct caption should have read:

Figure 3: *Axial Flair image through basis pontis showing bilateral symmetrical hypointense areas centrally in pons characteristically sparing ventral lateral tracks.*