**Spastic Diplegia**

**Epidemiology:**

According to the Center for Disease Control, spastic cerebral palsy accounts for about 80% of all cerebral palsy diagnoses1. Spastic diplegia accounts for 22.4% of cases in 8-year olds, as reported by a 2002 survey of 3 large geographic areas (Atlanta, Northern Alabama, and Southeastern Wisconsin)2. Approximately 8,000 to 10,000 new cases of cerebral palsy are diagnosed each year3. Prevalence of CP in the world has been reported from 1.9 to 4.1 per 1,000 births4. In the United States prevalence appears to be more between 2.3 and 3.6 per 1,000 births3.

**Pathology and pathophysiology of disorder/condition:**

Spastic diplegia is a non-progressive neurological condition characterized by an insult that occurs in the prenatal, perinatal, or postnatal brain9. In the prenatal environment the fetus may be exposed to maternal fever, environmental toxins, or abdominal trauma leading to a brain lesion9. Other factors that affect the fetus in the prenatal period include: multiple gestations by the mother, genetic disorders, hypoxia, metabolic disorders, and intrauterine infections9. Perinatal risk factors include: asphyxia, pre-mature birth or low birth weight, instrument delivery, or blood incompatibility9. Postnatal complications such as sepsis, meningitis, respiratory distress, intraventricular hemorrhage, head injury, and periventricular leukomalacia may lead to the development of CP9. Due to the wide variety of risk factors and complications, CP presents differently in each individual.

As previously stated, spastic diplegia is a non-progressive condition, however, the impairments of an individual may change over time. Spastic diplegia involves a disruption of motor and postural responses, with the amount of involvement based on the size and area of the lesion2. Lesions created by ischemia or infection can lead to periventricular leukomalacia (PVL)13. PVL most commonly occurs in premature infants and in very low birth weight (<1500g) children13. The brain and its vasculature are not fully developed or supported prior to 32 weeks gestation, leaving the periventricular white matter susceptible to injury13. Hemmorrhage near the periventricular white matter is possible and can disrupt the function of the internal capsule and motor cortex14. Damage to these regions leaves the corticospinal tract unable to relay messages for voluntary movement to the skeletal muscles14. In spastic diplegia the vestibular and reticular nuclei can also be damaged, leading to incorrect signaling, spasticity, and altered posture14.

**Disease progression**: **– effects on systems**

*CNS:* Dependent on the location and trigger of the insult. As discussed in the case of PVL, the internal capsule, corticospinal tract, and vestibular and reticular nuclei can all have their functions disrupted14. Damage in the CNS presents differently in each individual and sometimes the impairments are not seen until the child begins to miss motor milestones.

*Cardiopulmonary:* Children with spastic diplegia have a lower level of physical fitness7, ranging from 13-53% less than typically developing age-matched peers8. Abnormal tone can alter posture, making respiration more labor-some and less efficient, leading to decreased endurance abilities9. Spasticity leads to abnormal position of the joints in the LEs and causes gait to be more energy consuming12.

*Integumentary:* The integumentary system can be threatened by several complications with treatment and care of CP. Many times splints or orthotics are used to help correct alignment, improve/maintain muscle length, or to increase functional abilities, however, they require proper fit, skin evaluation, and maintenance of the equipment. The abnormal alignment of an individuals LEs causes unwanted torque and forces on the equipment, leading to rubbing and increased friction on the patient’s skin. Skin breakdown from equipment is very common and the PT and/or caregivers should regularly perform a skin assessment. Increased inactivity can also lead to prolonged sitting/lying causing ischemia to tissues around boney prominences.

*Neuromuscular:* Increase spasticity and tone especially in the LEs. Positive Babinski’s and clonus, as this is an UMN lesion. Decreased voluntary control of muscle action due to damage in the motor cortex tracts14. Sensory changes such as two-point discrimination have been found in children with spastic diplegia15.

*Musculoskeletal:* Individuals have decreased anaerobic power and muscular endurance7. A crouched gait with increased hip adduction/IR, knee flexion/valgus, ankle PF, lack of hip/knee extension, no heel strike, decreased stride length/stance time bilaterally, and decreased pace. Abnormal joint position and muscle lengths put increased wear and tear on the hip, knee, foot, and ankle. Decreased PROM and AROM can vary from patient to patient. Generation of muscle force7 is slower and not uniform compared to typically developing children.

*Other:* Pain is common in general with the population of patients with CP, with up to 75% reporting pain10. Other possible impairments seen in CP: vision loss or strabismus, hearing loss, loss of bladder control, behavioral problems, and sleep disturbances10.

**How does the disorder affect activity, participation and quality of life?**

*Activity:* Calley et al conducted a study comparing typically developing children and children with CP16. Evaluation of activity was based off the ICF model definition of activity, “a specific task or action undertaken by an individual”16. Children with CP were found to have no difference in the realm of school/productivity, sports, or hobbies/social activities, when compared to typically developing peers16. The major difference in activity participation came from lack of endurance, as measured by the 6MWT16. Typically developing peers had significantly better scores on both the 6MWT and the TUG compared to the children with CP16. These results show that children with CP will have a harder time maintaining pace with peers throughout an activity or simply walking side-by-side in the hallways with them16. The lower TUG scores indicate a challenge with safely and timely maneuvering from a desk to get an assignment and sitting back down again. The same could be said for getting a lunch tray and returning to the lunch table. From my clinical experience I also noted a challenge with stair climbing in children with CP. The increased tone in the LEs made ascending the stairs with a reciprocal pattern extremely difficult. The children would take more than 30 seconds to ascend a flight of stairs, making them at risk for being trampled during passing times. On several occasions we had to make protocols to allow for these children to be released early from class in order to make it safely to their next destination.

*Participation:* Participation has been defined as “an involvement in a life situation” by the WHO16. Larger disparities between typically developing children and children with CP have been found in regards to participation16. Calley et al found that the most significant differences between typically developing children and children with CP were in the domains of community life and recreation16. Community life might include items such as riding public transportation, participation in fundraisers/parades, or volunteering. Given the lack of endurance of many of the children with CP, conserving energy for ADLs and normal ambulation is critical. Getting on public transportation can be difficult if the steps are too large to conquer, not to mention the ability to safely walk to a seat before the bus starts moving. Recreation also has its limitations due to endurance and accessibility. I like to think of the example of the Washington Mall for recreation. The Mall is filled with many recreation activities but even for typically developed peers it can be an exhausting journey to complete. A child with CP may not be able to complete the museums, have a picnic, or cross the street without becoming overly exhausted. The variance in terrain, the energy expenditure, and the speed at which these activities must be completed is not feasible for many individuals with CP. Participation can go a step further and interfere with an individual’s ability to drive or maintain employment. Depending on cognitive abilities and motor control driving may not be an option for an individual with CP. Working choices may also be limited due to the demands of the job. Jobs that require excessive standing or lifting are going to be difficult for individuals with CP, due to the lack of endurance, abnormal posture, and decreased peak muscle force generation7.

*Quality of Life (QoL):* QoL has been found to remain at levels similar to typically developing peers in the domains of emotional and social health16. Several studies have expressed that a “disability paradox” occurs where those with CP learn to adjust to their condition and accept it because it is all they know16. In the minds of these individuals life is still good and they still manage to have good relationships16. The difference is QoL comes from the perception of function abilities16. Children with CP rate QoL lower in the functional ability domain than their typically developing peers16. I believe this is a positive considering the amount of diseases/conditions where individuals have a low QoL score in the emotional or social domains. This means that individuals are more emotionally stable and less likely to become depressed or suicidal.

**Intervention:**

Selective dorsal rhizotomy (SDR) is an invasive surgical technique where specific nerve rootlets are severed in order to reduce the amount of spasticity in muscles they innervate. St. Louis Children’s Hospital is the leader in surgical intervention for selective dorsal rhizotomy and doctors there have reduced the invasiveness of the procedure from 5-7 vertebral laminectomies to merely 1-2 levels5. Surgeons typically test nerve rootlets in bundles of 3-5 in order to determine (rate) the severity of spasticity they produce5. Levels L2-S2 are the main areas affected with spastic diplegia and are the target of SDR5. SDR is not a fit for all individuals with spastic diplegia and may produce several complications.

The most appropriate candidates for SDR are children age 2-4 years old, yet SDR has shown positive improvements in individuals up to the age of 40 years old5. Individuals with spastic diplegia, hemiplegia, tetraplegia, and triplegia may all benefit from surgical intervention5. Individuals who are independent in mobility, exhibit potential for functional improvement, and have no basal ganglia involvement also have better outcomes5. SDR also requires a significant amount of commitment by the patient, the patient’s family, and the physical therapist. If the patient is not motivated or making progress towards motor milestones, they may not be appropriate for SDR5. Inappropriate candidates would include individuals who are unmotivated, have a mixed case of CP, history of meningitis, have severe scoliosis, have congenital hydrocephalus, or have a diagnosis not related to premature birth5.

According to St. Louis Children’s Hospital, they have only had 4 patients out of 2,300 who required a secondary surgery due to complications (spinal fusion, 3 spinal fluid leaks)5. Other possible complications include: lower extremity paralysis, impotence, meningitis, incontinence, and loss of lower extremity sensation5. Outcomes also appear related to age, severity of condition, muscle strength, and level of independent mobility. Grunt et al found that spinal deformities following SDR were more frequent than reported at St. Louis Children’s Hospital6. Scoliosis was the most prevalent complication, ranging from 41-56% of patients, while lumbar lordosis, thoracic kyphosis, spondylolysis, spondylolisthesis, and LBP were also frequently reported6.

Grunt et al performed a systematic review of studies on SDR and outcomes that included a total of 966 patients6. Patients were between 2-27 years old, included multi-level SDR and single-level SDR, with follow-ups greater than 5 years6. Follow-up of patients at 2 years post-operation were found to have decreased spasticity, increased PROM, improved gait pattern, and better gross motor function6. More specific improvements were found in step length and gait speed that persisted up to 3 years post-operation6. Grunt el al included one study that found positive results in gait and kinematics, close to healthy norms, after 20-year follow-up6.

MacWilliams et al compared the results of 26 patients who received SDR, orthopedic surgery, or nothing at all after the age of 1011. Appropriate candidates and successful outcomes have been associated with patients’ age 2-45 or 5-1011. Patients electing to undergo SDR after age 10 were found to have a significant decline in function as measured by the GMFM and GMFCS11. Meanwhile, the orthopedic surgery and no surgery groups both improved in functional outcomes11. Deviations in gait did improve in the SDR group but not significantly like they did in the orthopedic surgery group11. Findings in this study indicate that earlier detection and selection of appropriate candidates is necessary for optimal results following SDR.

Thomas et al also compared SDR versus orthopedic surgery, but the average age of subjects were 6 years and 6 years, 5months respectively12. Patients undergoing SDR had a 1 month hospital stay with PT 2x/day and OT 1x/day, followed by OP PT (3-4x/week for 6 months, then 1-2x/weeks for 6-12months) and OT (1-2x/week for 6 months)12. The orthopedic group had a variable hospital stay and only PT for 2-4 months12. A 2-year follow-up showed significant reduction in tone in all LE for the SDR group, and only 3 LE muscles in the orthopedic group12. ROM improved in the dorsiflexors and knee extensors of the SDR group, while the orthopedic group had significant improvements in hip/knee extension12. The orthopedic group had significant improvements in stride length, but no such improvement was seen in the SDR group12. Velocity of ambulation and DF ROM during gait both saw improvements in the SDR group12. Both groups showed improvement in oxygen cost of gait, with the orthopedic group’s results significant12.

This information on SDR has helped me understand that surgery is not always better, however, it may be appropriate for some children with spastic diplegia. Prior to this paper I had assumed that SDR would be beneficial for patients with CP no matter what age. The research and leaders in the field have clearly found that SDR is more appropriate for younger individuals (2-10 years old). Although the surgery has shown positive results in this population it is also important to critically assess the patient and their support before intervention is initiated. SDR takes a significant amount of resources, both financial and emotional. I believe this puts it in perspective that as PTs we should not rely on surgical interventions to solve our patients’ problems and that sometimes we have to work with what we have. A whole patient perspective can help to determine if an individual is an appropriate candidate for SDR or not.

References:

1. Centers for Disease Control and Prevention. Cerebral Palsy. 2012. Retrieved from: <http://www.cdc.gov/ncbddd/cp/facts.html>
2. [Yeargin-Allsopp M](http://www.ncbi.nlm.nih.gov/pubmed?term=Yeargin-Allsopp%20M%5BAuthor%5D&cauthor=true&cauthor_uid=18310204), [Van Naarden Braun K](http://www.ncbi.nlm.nih.gov/pubmed?term=Van%20Naarden%20Braun%20K%5BAuthor%5D&cauthor=true&cauthor_uid=18310204), [Doernberg NS](http://www.ncbi.nlm.nih.gov/pubmed?term=Doernberg%20NS%5BAuthor%5D&cauthor=true&cauthor_uid=18310204), [Benedict RE](http://www.ncbi.nlm.nih.gov/pubmed?term=Benedict%20RE%5BAuthor%5D&cauthor=true&cauthor_uid=18310204), [Kirby RS](http://www.ncbi.nlm.nih.gov/pubmed?term=Kirby%20RS%5BAuthor%5D&cauthor=true&cauthor_uid=18310204), [Durkin MS](http://www.ncbi.nlm.nih.gov/pubmed?term=Durkin%20MS%5BAuthor%5D&cauthor=true&cauthor_uid=18310204). Prevalence of cerebral palsy in 8-year-old children in three areas of the United States in 2002: a multisite collaboration. Pediatrics. 2008 Mar;121(3):547-54.
3. Stern Law Group. Prevalence and Incidence of Cerebral Palsy. Retrieved from: http://cerebralpalsy.org/about-cerebral-palsy/prevalence-of-cerebral-palsy/
4. [Pakula AT](http://www.ncbi.nlm.nih.gov/pubmed?term=Pakula%20AT%5BAuthor%5D&cauthor=true&cauthor_uid=19643346), [Van Naarden Braun K](http://www.ncbi.nlm.nih.gov/pubmed?term=Van%20Naarden%20Braun%20K%5BAuthor%5D&cauthor=true&cauthor_uid=19643346), [Yeargin-Allsopp M](http://www.ncbi.nlm.nih.gov/pubmed?term=Yeargin-Allsopp%20M%5BAuthor%5D&cauthor=true&cauthor_uid=19643346). Cerebral palsy: classification and epidemiology. Phys Med Rehabil Clin N Am. 2009 Aug;20(3):425-52.
5. Children’s Hospital St. Louis. About Selective Dorsal Rhizotomy. Retrieved from: http://www.stlouischildrens.org/our-services/center-cerebral-palsy-spasticity/about-selective-dorsal-rhizotomy-sdr
6. Grunt, S., Becher, J., & Vermeulen, R. Long-term outcome and adverse effects of selective dorsal rhizotomy in children with cerebral palsy: a systematic review. *Developmental Medicine and Child Neurolog. 2011;* *56*(6): 490-498.
7. Berg-Emons HJG van den, van Baak MA, Speth L, Saris WH. Physical training of school children with spastic cerebral palsy: Effects on daily activity, fat mass and fitness. Int J Rehabil Res 1998;21:179 – 194.
8. [Carlon SL](http://www.ncbi.nlm.nih.gov/pubmed?term=Carlon%20SL%5BAuthor%5D&cauthor=true&cauthor_uid=23072296), [Taylor NF](http://www.ncbi.nlm.nih.gov/pubmed?term=Taylor%20NF%5BAuthor%5D&cauthor=true&cauthor_uid=23072296), [Dodd KJ](http://www.ncbi.nlm.nih.gov/pubmed?term=Dodd%20KJ%5BAuthor%5D&cauthor=true&cauthor_uid=23072296), [Shields N](http://www.ncbi.nlm.nih.gov/pubmed?term=Shields%20N%5BAuthor%5D&cauthor=true&cauthor_uid=23072296). Differences in habitual physical activity levels of young people with cerebral palsy and their typically developing peers: a systematic review. Disabil Rehabil. 2013 Apr;35(8):647-55.
9. Ollendick, K. Cerebral Palsy (Powerpoint). Neuromuscular Course, University of North Carolina-Chapel Hill. 2011.
10. [Ashwal S](http://www.ncbi.nlm.nih.gov/pubmed?term=Ashwal%20S%5BAuthor%5D&cauthor=true&cauthor_uid=15037681), [Russman BS](http://www.ncbi.nlm.nih.gov/pubmed?term=Russman%20BS%5BAuthor%5D&cauthor=true&cauthor_uid=15037681), [Blasco PA](http://www.ncbi.nlm.nih.gov/pubmed?term=Blasco%20PA%5BAuthor%5D&cauthor=true&cauthor_uid=15037681), [Miller G](http://www.ncbi.nlm.nih.gov/pubmed?term=Miller%20G%5BAuthor%5D&cauthor=true&cauthor_uid=15037681), [Sandler A](http://www.ncbi.nlm.nih.gov/pubmed?term=Sandler%20A%5BAuthor%5D&cauthor=true&cauthor_uid=15037681), [Shevell M](http://www.ncbi.nlm.nih.gov/pubmed?term=Shevell%20M%5BAuthor%5D&cauthor=true&cauthor_uid=15037681), [Stevenson R](http://www.ncbi.nlm.nih.gov/pubmed?term=Stevenson%20R%5BAuthor%5D&cauthor=true&cauthor_uid=15037681); [Quality Standards Subcommittee of the American Academy of Neurology](http://www.ncbi.nlm.nih.gov/pubmed?term=Quality%20Standards%20Subcommittee%20of%20the%20American%20Academy%20of%20Neurology%5BCorporate%20Author%5D); [Practice Committee of the Child Neurology Society](http://www.ncbi.nlm.nih.gov/pubmed?term=Practice%20Committee%20of%20the%20Child%20Neurology%20Society%5BCorporate%20Author%5D). Practice parameter: diagnostic assessment of the child with cerebral palsy: report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. Neurology. 2004 Mar 23;62(6):851-63.
11. MacWilliams, B., Johnson, B., Shuckra, A., & D'Astous, J. Functional decline in children undergoing selective dorsal rhizotomy after age 10. *Developmental Medicine and Child Neurology. 2011;53*(8): 717-723.
12. Thomas, S., Buckon, C., Piatt, J., Aiona, M., & Sussman, M. A 2-year follow-up of outcomes following orthopedic surgery or selective dorsal rhizotomy in children with spastic diplegia. *Journal of Pediatric Orthopaedics B*. 2004; *13*(6):358-366.
13. Volpe, JJ. Neurology of the Newborn, 4th ed, WB Saunders, Philadelphia 2001.
14. British Medical Journal Best Practice. Cerebral Palsy: Pathophysiology. British Medical Association. 2012; Accessed from: <http://bestpractice.bmj.com/best-practice/monograph/674/basics/pathophysiology.html>
15. [Lesný I](http://www.ncbi.nlm.nih.gov/pubmed?term=Lesn%C3%BD%20I%5BAuthor%5D&cauthor=true&cauthor_uid=8495821), [Stehlík A](http://www.ncbi.nlm.nih.gov/pubmed?term=Stehl%C3%ADk%20A%5BAuthor%5D&cauthor=true&cauthor_uid=8495821), [Tomásek J](http://www.ncbi.nlm.nih.gov/pubmed?term=Tom%C3%A1sek%20J%5BAuthor%5D&cauthor=true&cauthor_uid=8495821), [Tománková A](http://www.ncbi.nlm.nih.gov/pubmed?term=Tom%C3%A1nkov%C3%A1%20A%5BAuthor%5D&cauthor=true&cauthor_uid=8495821), [Havlícek I](http://www.ncbi.nlm.nih.gov/pubmed?term=Havl%C3%ADcek%20I%5BAuthor%5D&cauthor=true&cauthor_uid=8495821). Sensory disorders in cerebral palsy: two-point discrimination. Dev Med Child Neurol. 1993 May;35(5):402-5.
16. [Calley A](http://www.ncbi.nlm.nih.gov/pubmed?term=Calley%20A%5BAuthor%5D&cauthor=true&cauthor_uid=22200241), [Williams S](http://www.ncbi.nlm.nih.gov/pubmed?term=Williams%20S%5BAuthor%5D&cauthor=true&cauthor_uid=22200241), [Reid S](http://www.ncbi.nlm.nih.gov/pubmed?term=Reid%20S%5BAuthor%5D&cauthor=true&cauthor_uid=22200241), [Blair E](http://www.ncbi.nlm.nih.gov/pubmed?term=Blair%20E%5BAuthor%5D&cauthor=true&cauthor_uid=22200241), [Valentine J](http://www.ncbi.nlm.nih.gov/pubmed?term=Valentine%20J%5BAuthor%5D&cauthor=true&cauthor_uid=22200241), [Girdler S](http://www.ncbi.nlm.nih.gov/pubmed?term=Girdler%20S%5BAuthor%5D&cauthor=true&cauthor_uid=22200241), [Elliott C](http://www.ncbi.nlm.nih.gov/pubmed?term=Elliott%20C%5BAuthor%5D&cauthor=true&cauthor_uid=22200241). A comparison of activity, participation and quality of life in children with and without spastic diplegia cerebral palsy. Disabil Rehabil. 2012;34(15):1306-10.