

**The Association between Cerebral Palsy
and Hearing Loss**

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- Cerebral palsy (CP) is believed to be the most common childhood motor impairment, currently affecting approximately 400,000 school age children (Taft, L. T., 1984).
- Cerebral palsy results from a static insult to the brain during its development, via infection, asphyxia, or trauma occurring during the prenatal, perinatal and postnatal periods.
- A large number of those with CP will have additional impairments, such as visual, auditory, seizures, cognitive and behavioral disabilities (Odding, E., Roebroech, M., & Hendrik, S., 2006).
- Specifically with hearing loss, Robinson, R. O., et. al. (1973) states that 12.5% of those with CP will have some level of hearing loss. This incidence may have changed as a result of more children surviving at smaller birth weights and younger gestational ages, both of which are risk factors for both CP and hearing loss.
- As a result of this association, audiologists and speech, language pathologists (SLP) must understand the causes of CP and hearing loss, as well as diagnosis, treatment and rehabilitation for those children between birth to six years of age that present with both CP and hearing loss.
- Diagnosing hearing loss in children with CP will require modifications to the techniques and equipment used. Once diagnosed, further modifications must be made in the fitting of amplification and subsequent rehabilitation.

Definition of Cerebral Palsy

- Cerebral Palsy is a disorder that involves impaired muscle tone, motor tasks and posture secondary to a static insult to the developing brain (Pearlman, J. M., 2006).
- It is considered the “most prevalent childhood motor disability” today (Johnston, M. V. & Hoon, A. H., 2006) with approximately 8000 infants with CP born in the United States annually (Wu, Y. W., Croen, L. A., Shah, S. J., Newman, T., B., & Najjar, D. V., 2006).
- The control of motor tasks involves complex coordination “between posture and movement through distinct timing and scaling functions of movement variables in order to synchronize the main steps of the task” (Hirschfeld, H., 2007).
- To be considered CP, the damage to the brain must occur either during neonatal development, the birthing process or shortly after birth, and there must be a motor function component.
- Impairment of tone is usually characterized by either hypertonic or hypotonic. Hypertonic is defined as an increase in tone, characterized by a rigidity, or increase tension in the muscles. Alternatively, hypotonia is characterized by a lack of muscle tension, or ‘floppy’ muscle tone (Sanger, T., Delgado, M. R., Gaebler-Spira, D., Hallett, M., & Mink, J.W., 2003).

- The majority of CP is caused by prenatal causes including infection and maternal health.
- Only a small portion can be traced to intrapartum events, such as asphyxia (Johnston, M. V. & Hoon, A. H., 2006).
- The location of the insult to the brain, as well as when during development the insult occurs, can determine the severity and type of CP as well as associated disorders.
- Severity ranges from a mild presentation where very few areas of the body are affected to a more profound presentation that involves most parts of the body.
- In most cases there is not an affect on lifespan of the individual with CP, although, life span can be affected in very severe damage to the brain injury and serious associated disabilities (Koman, L. A., Smith, B. P., & Shilt, J. S., 2004).
- While the insult to the brain is static, or not changing, motor function and tone can change over time, especially during the first few years of life.
- For instance, those with athetoid or dyskinetic cerebral palsy can begin life as “floppy,” or hypotonic, and become more “rigid” or hypertonic by one year of age (Taft, L. T., 1984).
- As a result of this changing pattern of muscle tone, diagnosis of specific types and severities of CP can be difficult and may require multiple modifications to the original diagnosis.

- The incidence in developed countries is considered to be 2 – 2.5 in every 1000 live births.
- It is thought that in underdeveloped, or third world countries, the incidence of CP is higher.
- This difference of incidence rate is thought to be a result of current medical practices, both during the birthing process and medical attention throughout the pregnancy (Johnston, M. V. & Hoon, A. H., 2006).
- While better medical care is being established throughout the United States, the number of term infants born with insults that result in CP is growing.
- According to Wu, Y. W., et al. between 1975 and 1991 the incidence increased from 1.7 to 2.0 per 1000 live births.
 - This increase was from term infants, rather than premature or low birth weight infants. Now this is surprising isn't it?
 - Research is necessary in explaining the increase in incidence of term-births, while there is an increase number of surviving infants and borderline viability (Fawke, J., 2007).
 - While research about etiologies has been mainly restricted to those with CP born premature or at a low birth weight, more than half of those with CP were born at term.
 - Further research in the etiology and risk factors for term births resulting in CP is needed (Wu, Y. W., et. al., 2004).

- While a large majority of those with CP are term infants, it was found that children with CP were found to be eight times more likely to be in the NICU (Wu, Y. W., et. al., 2004).
- Interestingly, there seems to be a gender preference. In a study by Odding, et. al. (2006) it was found that 58% of all those children born with CP were males.
 - This gender preference has not been fully explained and further research in this area is needed.

Etiology and Risk Factors

- Cerebral palsy results from some insult to the developing brain.
- The brain during development is a vulnerable organ, which is sensitive to many infections, trauma and deprivations.
- In the following section, the more prevalent causes of injury to the brain that results in CP will be briefly discussed, including prenatal, perinatal, and postnatal factors.
- These factors include preterm birth, low and very low birth weight, multiple births, maternal infection, fetal infection and fetal anoxic events (Koman, et. al., 2004).
- Prematurity and birth weight of the infant are thought to be the most important risk factors in developing CP.

- Maternal age of over 35 years was also found to be a risk factor for CP (Wu, Y. W., et. al., 2004).
- Multiple births increased the chances of the infant(s) having CP times 1.7 (Wu, Y. W., et. al., 2004). While there are many causes of CP, 20- 30% of those with CP have no clear etiology (Taft, L. T., 1984).
- Lowered socioeconomic status seemed to be negatively correlated with CP. This relationship can be interpreted as a reduced amount of prenatal care, quality of medical staff during birth, as well as nutrition and maternal health.

Prenatal, Perinatal and Postnatal Etiologies and Risk Factors:

- The primary etiology of CP is prenatal factors, or those that occur during the pregnancy and fetal development, which account for 70-80% of CP (Pearlman, J. M., 2006).
- These factors include, maternal health, metabolic disorders, and infection. Preeclampsia, while not considered a cause of CP, it is a risk factor for term infants that can increase the possibility of CP developing.
- Preeclampsia is a condition involving hypertension during the pregnancy, and is considered a risk factor, as a result of the condition inducing spontaneous preterm labor.

- Premature birth, especially if low birth weight as well, has a high incidence of CP developing (Reddihough, D. S. & Collins, K. J. 2003).
- Multiple pregnancies increase the odds of one, or multiple children developing CP. This increased rate is a result of the increased risk of premature labor, low birth weight, poor intrauterine growth and intrapartum complications (Ibid).
- Infections during pregnancy can increase the risk of developing CP, or cause the insult to the brain, which can result in CP. These infections range from rubella to cytomegalovirus (CMV) (Nelson, K.B & Willoughby, R. E, 2000).
 - The CMV has a 50% chance of passing through the placenta from the infected mother and infecting the fetus (Gibson, C. S., MacLennan, A. H., Goldwater, P. H. Haan, E. A., Priest, K., & Dekker, G. A., 2006).
 - According to Stanley, F., et. al. (2000) 5-10% of those with CP the etiology is CMV.
- Although complications during labor are given the majority of press and public scrutiny, it is thought that only 15- 20% of cases that result in CP are caused by perinatal events.
- In other words, less than 1 in 1000 live births at term will result in CP from an intrapartum event for a term delivery (Pearlman, J. M., 2006).
- There is thought to be a difference in incidence rates of intrapartum events in developing countries compared to developed countries; most likely a

result of the state of the medical care in developing countries (Johnston, M. V. & Hoon, A. H., 2006).

- Complications during the birthing process include asphyxia, trauma or infection (Johnston, M. V. & Hoon, A. H., 2006).
- Other risk factors during the perinatal period include fetal distress, pre-eclampsia, sepsis, abruptio placenta, prelabor rupture (Kulak, W., Sobanieck, W., Kubas, B., Walecki, J., Smigielska-Kuzia, J., Bockowski, L., Artemowicz, B., & Sendrowski, K., 2007) and perinatal stroke (Golomb, M. R., Saha, C., Garg, B. P., Azzouz, F., & Williams, L. S., 2007).
 - Perinatal stroke is associated with the development of CP as well as additional impairments.
 - Of those who develop CP, 70% of those with neonatal presentation of the stroke will develop an additional impairment, including speech, cognition and epilepsy (Golomb, et. al., 2007).
 - Perinatal stroke sits under the general umbrella term of Neonatal ischemic stroke; which also includes perinatal arterial ischemic stroke and cerebral sinovenous thrombosis. These types of stroke contribute in the development of CP by causing a focal insult to the brain (Kirton, A., & deVeber, G.,2006).
- Asphyxia as an intrapartum event is defined as an interruption of placental blood flow during the first and second stages of labor, for both term and preterm deliveries, and can result in CP (Pearlman, J. M., 2006).

- Studies have shown that even if the asphyxia was severe and prolonged, the fetus has amazing abilities to adapt in order to preserve cerebral function.
- For instance, the fetus will redistribute blood flow from organs deemed less important to those that are required for survival, such as the brain.
- If asphyxia continues, even this redistribution will not be able to prevent permanent damage to the child (Pearlman, J. M., 2006).
- Persistent asphyxia will limit oxygen supply and cerebral perfusion of blood flow, which in turn leads to a hypoxic-ischemic injury to the brain.
- The clinical diagnosis, in term infants, of hypoxic-ischemic encephalopathy is used if damage to the brain results from the hypoxic-ischemic insult.
- This disorder can include: seizures, muscle tone abnormalities, problems with feeding and respiration as well as a disturbance in alertness level of the child.
- During the first year of life, cerebral palsy may develop in these children (Johnston, M. V. & Hoon, A. H., 2006).
- With regards to the co-morbidity of asphyxia, it was found that hearing loss was 3.5 times more likely to occur in children with known asphyxia (Lima, G. M., Marba, S. T., & Santos, M. F. C., 2006).
- The amount of time the brain is deprived of oxygen and blood can impact the type and diffusion of damage.
- A short but intense asphyxiation, for example that which is seen with total compression of the umbilical cord, can result in selective injuries to the cerebral cortex, basal ganglia, thalamus and/or brainstem.

- Whereas with prolonged periods of reduced oxygen that is not as intense produces a more diffuse or multi-focal array of damage to the cerebral cortex (Johnston, M. V. & Hoon, A. H., 2006).
- Commonly, asphyxia is seen in the vascular distribution in the brain and can include selective neuronal necrosis (SNN), periventricular leukomalacia and ischemic injury, both focal and multi-focal in nature.
- Selective neuronal necrosis is the selective death of cells located in the brain. This cell death can result in permanent, non-progressive damage to the brain, which in turn can lead to CP (Pearlman, J. M., 2006).
- Periventricular leukomalacia (PVL) is damage, or a softening of the white matter near the ventricles in the brain that can lead to cognitive, behavior, motor and sensory abnormalities. This type of damage is seen commonly in premature infants, as a result of the vulnerability of the fetus during cerebral development (Johnston, M. V. & Hoon, A. H., 2006).
- Physiologically, a thinning of the corpus callosum, irregular ventricular enlargement and a loss of white matter can be seen using magnetic resonance imaging (MRI) (Johnston, M. V. & Hoon, A. H., 2006).
- MRIs are useful in detecting the physiological changes that occur with this type of damage to the white matter. Melhem et. al. (2000) has shown a “direct relationship between the degree of white matter injury and clinical severity of motor and cognitive impairment.” Typically, the motor impairment strongly associated with PVL is spastic diplegia cerebral palsy (Johnston, M. V. & Hoon, A. H., 2006).

- The damage caused by PVL differs between preterm infants and term infants, as a result of the changes in vulnerability during the development of the brain, including changes in blood supply; this window of vulnerability is considered to be between 24 to 32 weeks gestation. A second window of vulnerability occurs closer to term, or 40 weeks gestation. In this period areas of the brain that are rich in neurons and neuronal connections are at risk for injury, including the basal ganglia, cerebral cortex, hippocampus and brainstem. PVL occurring at term will damage more subcortical regions of the brain, while preterm infants PVL will result in a periventricular distribution (Johnston, M. V. & Hoon, A. H., 2006).
- Interestingly, studies are showing a promising method in preventing extensive damage to the brain after asphyxia. This method involves cooling the child very soon after birth. This controlled hypothermia is showing promise in reducing the damage, and preventing the possibility of CP (Johnston, M. V. & Hoon, A. H., 2006). Further research is needed in this area, but it shows potential in reducing permanent outcomes when hypoxic-ischemic encephalopathy is diagnosed.
- An important postnatal etiology of CP is hyperbilirubinemia. Hyperbilirubinemia is an abnormally high concentration of bilirubin in the blood, which can lead to jaundice in the infant. If untreated, or if the levels of bilirubin are excessively high, then serious complications can arise from the transmission of bilirubin across the blood-brain barrier to the basal

ganglia and the brainstem nuclei, resulting in kernicterus, or bilirubin encephalopathy. (Sheykholeslami, K., & Kaga, K., 2000).

- This elevation of bilirubin levels has been associated with hearing loss, cerebral palsy and other disorders. Jaundice in the population is considered somewhat common with approximately 60% of all full term infants are subsequently diagnosed with being jaundice during the first week after birth.
- Eighty percent of preterm infants will develop hyperbilirubinemia during this first week.
- Additionally, this preterm population is at a higher risk to develop secondary disorders, as a result of the permeability of the blood brain barrier (Sheykholeslami, K., & Kaga, K., 2000).
- According to Sheykholeslami, K., & Kaga, K. (2000), severe kernicterus usually leads to death of the infant within the first few weeks of life. When the infant survives the kernicterus, athetoid type of CP is often seen, as is hearing loss and cognitive impairments.
 - While it is thought that there are pathological changes in the outer hair cells in the cochlea or the auditory nerve, how specifically hyperbilirubinemia affects the cochlea with regards to hearing loss still remains unclear and further research is needed (Sheykholeslami, K., & Kaga, K., 2000; Sano, M., Kaga, K., Kitazumi, E., & Kodama, K., 2005).

- While the exact cause of the hearing loss may still need to be researched, it is known that hyperbilirubinemia is an important etiology of hearing loss (Lima, G. M., et. al., 2006).

Preterm Births and Birth Weight:

- A child born before 37 weeks gestation is considered premature (Kulak, W., et. al., 2007), and is at a high risk for developing CP as well as other disorders, including hearing loss.
- According to Drougia, et. al. (2007), premature birth is the main risk factor associated with cerebral palsy.
- When preterm birth is in association with periventricular leukomalacia (PVL), the risk for cerebral palsy developing increases by more than 15 times (Drougia, A., et. al., 2007).
- Periventricular leukomalacia is an insult to the white matter near the ventricles in the brain. As stated earlier, this damage, or softening of the white matter, typically will occur as a result of infection and ischemia (Johnston, M. V. & Hoon, A. H., 2006).
- Periventricular leukomalacia is seen in both preterm and term infants, however, with less maturity and decreased birth weight, a more wide spread motor impairment may be seen (Fawke, J., 2007).

- Cerebral palsy following premature birth, especially when associated with PVL, will typically be of the spastic diplegia type.
- In a study performed by Vergani, et al. (2004) PVL and intraventricular hemorrhage (IVH) are the main causes of CP and neurodevelopmental delay, specifically in the preterm population. Intraventricular hemorrhage is a bleeding within or around the ventricles.
- Within the preterm population with CP, both IVH and PVL occur concurrently in approximately 14% (Vergani, P., Locatelli, A., Doria, V., Assi, F., Paterlini, G., Pezzullo, J. C., & Ghidini, A., 2004). Vergani, et al. (2004) suggests that both conditions may have the same causal factors, but more research is needed in determining risk factors and etiologies.
- Research conducted by Fanaroff, J. M., et al. (2006) describes the possible relationship between IVH and hypotension in the neonate, but admits that additional research is needed.
- Outcomes of these two conditions differed slightly, while both could result in CP, those with IVH had a higher rate of neurodevelopmental delay when compared to those with PVL.

- Periventricular leukomalacia is associated with “cognitive, behavioral, and sensory abnormalities,” with more severe presentations at a higher risk for developing CP (Johnston, M. V. & Hoon, A. H., 2006).
- There is an increase in the survival of borderline viable infants, born approximately 23 – 25 weeks gestation.
- This survival is associated with the usage of prenatal steroids and postnatal surfactant therapy (Fawke, J., 2007; Hoekstra, R. E., Ferrara, B., Couser, R. J., Payne, N. R., & Connett, J. E., 2004).
- Surfactants are used in increasing the lubrication between the layers of the lungs, allowing for easier and more efficient respiration by the infant or mechanical ventilation.
- Additionally, the use of steroids in helping to wean infants off of ventilators has been increasing survival of the child, but is associated in an increase in the prevalence of CP (Vohr, B. R., 2007).
- There appears to be an association between gestational age at birth and cognition. Compromised cognitive abilities are correlated with those born below 33 weeks of gestation.
- As the gestational age is reduced there is a higher the incidence of cognitive impairments (Johnson, S., 2007).
- A current survey states that in the United States there are approximately 30,000 extremely low birth weight (<1kg) infants born

each year (Fanaroff, J. M., Wilson-Costello, D. E., Newman, N. S., Montpetite, M. M. & Fanaroff, A. A., 2006).

- Among low birth weight infants, whether born at term or prematurely, the rate of those with CP is higher than those born above 2500 g or approximately 5 lbs.
- The rate of CP in children with low birth weight, less than 1500 g, is 74.2 in 1000 live births, and 11.9 for those children weighing between 1500-2499g (Odding, E., Roebroech, M., & Hendrik, S., 2006).
- According to Winter, S., Autry, A., Boyle, C., & Yeargin-Allsopp, M. (2002) of the children with CP, 25-35% were born “with a birth weight of <1500g.”
- To result in CP, both prenatal and postnatal events can contribute in this population (Johnston, M. V. & Hoon, A. H., 2006).
- Many of these low birth weight infants are deemed failures to thrive and are at a high risk for developing vision, hearing, chronic health problems, behavior and other impairments.
- Vohr, B. R., (2007) warns that the examination of neurodevelopment at term age, or corrected age, is not a strong predictor of future development.

- With regards to hearing loss, pre-maturity is highly correlated with the preterm and low birth weight population.
- Many of those premature infants who have a hearing loss; will have a loss that is sensorineural .
- Additionally, many of these children would have required extensive respiratory intervention, or use of ventilation (Marlow, E. S., Hunt, L. P., & Marlow, N., 2000).
- This association may be a result of additional risk factors, ventilation noise, and administration of antibiotics, which could be damaging to the inner ear (Marlow, et. al., 2000).

Classification of Cerebral Palsy

- Classification and diagnosis of the different types of CP relies on (a) the motor function seen, (b) the areas of the body affected and (c) the severity of the motor impairments (Blair, E., & Watson, L., 2006).
- Four main types of CP are
 - spastic,
 - athetoid or dyskinetic,
 - ataxic,
 - or a mixture of types.

- Each are defined by how motor function is affected in the individual, specifically the tone of the muscles and if there are involuntary movements involved.
- While there are clear differences in the types of CP, there are many incidences where more than one type of CP is seen in one person.
- The type and severity of motor function impairment in those with CP is linked to where in the brain the insult occurred.
- Three of the terms used to define the area of the body affected are: hemiplegia, diplegia, and quadraplegia.
 - Hemiplegia describes the involvement of the ipsilateral arm and leg;
 - diplegia involves significant leg participation with little affect on the arms,
 - while quadraplegia involves all four limbs (Koman, et. al., 2004).
 - Each of these are used in associated with a type of CP to describe the extent to which the person is affected.
 - Spastic diplegia is the most common of all the subtypes of CP, affecting approximately one third off all cases (Rennie, J. M., Hagmann, C. F., & Roberston, N. J., 2007).
 - Additional impairments occur in 25-80% depending on type and severity of CP, for instance
 - 42% of those with hemiplegic CP will have an additional impairment (Odding, et. al., 2006).

- Those with CP, when compared to their able-bodied peers, have less power and muscular endurance in affected limbs, depending on type and severity of CP.
- For those able to walk, the expenditure of energy is considered to be 3 times that of a typically developing peer, causing fatigue.
- Fatigue is seen as worsening of functional skills, reduced quality of life, feeling of pain and emotional drain (Odding, et. al., 2006).
- Spastic CP is defined by its hypertonia, or increase in muscle tone, and is characterized by stiff, rigid and awkward movements that affect a number of limbs in the body. The rigidity is currently being defined as the “resistance passive motion that has a ‘plastic,’ ‘malleable,’ or ‘lead-pipe’ quality” (Sanger, T. D., et. al. 2003).
- As stated above, spastic diplegia is the most common type (Rennie, J. M., et. al., 2007), seen in 80% of those with CP (Odding, et. al., 2006).
 - It is typically associated with preterm birth, PVL and IVH (Fawke, J., 2007).
 - With spasticity, specifically tetraplegic, additional musculoskeletal problems can occur, including: hip

luxations (75%), contractures (73%), and scoliosis (72%) (Odding, et. al., 2006).

- Spasticity can be increased “by anxiety, emotional state, pain, surface contact, or other nonnoxious sensory inputs.”
 - Alertness, activity and posture can affect the rigidity and spasticity of the muscles (Sanger, T. D., et. al. 2003).
 - Unlike spastic CP, which is characterized by difficulty in voluntary movements, athetoid/dyskinetic CP has involuntary, slow, writhing-like movements.
 - The motor impairment can affect all four limbs, hands, feet, face and tongue (Rennie, J. M., et. al., 2007).
 - These involuntary changes in tone can be disconcerting and emotionally detrimental to the child with CP.
 - More severe cases of athetoid/dyskinetic CP can cause a child to be in a constant state of involuntary, unwanted motion (Rennie, J. M., et. al., 2007) that can change daily or even within the same day (Robinson, R. O., 1973).
 - Approximately 10-20% of those with CP will have athetoid/dyskinetic CP, and 80% of these cases will be caused by hypoxic ischemia in term infants (Rennie, J. M., et. al., 2007).
- The mixed type of CP is defined as a number of muscle tones

presented in the same person. In essence those with mixed type of CP, present with symptoms from more than one type. Typically this mixture occurs between spasticity and athetoid/dyskinetic.

- The third type of CP is ataxia.
 - True ataxic CP is rare, seen in approximately 4% of all CP cases.
 - Ataxic CP is usually the result of an insult to the cerebellum.
 - Balance and depth perception are usually affected in the ataxic populations, as is gait, the performance controlled movements, and those movements requiring quick response time.
 - The difficulty in controlled movements can impair writing ability for instance, and reduce quality of life for the person with CP.
 - It is thought that only 5-10% of those with CP have the ataxic type (Rennie, J. M., et. al., 2007).
 - The Gross Motor Functional Classification Scale (GMFCS) is a systematic method of identifying not only type of CP, but also severity of the motor function.
 - Assessments are made from visual observation of the child. The motor function ranges from normal, level 1, to a profound impairment, or level 5 (Vohr, B. R., 2007).
 - The following graph is from Vohr, B. R., (2007) is the five levels of the GMFCS modified to work with 18-22 month old children with CP, where mild CP is equivalent to possible level 1 and level 1, and severe CP is considered level 5.

Modified Gross Motor Classification System used at 12-22 Months	
Score	Definition
Normal	Walks 10 steps independently and fluently
Possible	Walks 10 steps independently but not fluently; may exhibit toe walking (mild dysplasia) or asymmetric walking (possible hemiplegia)
Level 1	Child moves in and out of sitting and floor sits with both hands free to manipulate objects. Child can creep or crawl on hands and knees, pull to stand and take steps holding on to furniture. Children walk between 18 and 24 months of age without holding on.
Level 2	Child maintains floor sitting but may need to use hands for support to maintain balance. Child creeps on stomach or crawls on hands and knees, and may pull to stand and take steps holding on to furniture.
Level 3	Child maintains floor sitting when lower back is supported. Child rolls and creeps forward on stomach.
Level 4	Child has head control but trunk support is required for floor sitting. Child can roll to supine and may roll to prone.
Level 5	Physical impairments limit voluntary control of movement. Child is unable to maintain antigravity head and trunk posture in prone and sitting. Child requires adult assistance to roll.

- The diagnosing of CP can be a difficult and be a very involved process.
- Age and additional disabilities can inhibit readily identifying and qualifying CP.
- The overlap of muscle tone impairment between types of CP, as well as the changes in tone during the first year of life, makes it increasingly difficult to diagnose.
- Working in conjunction with a multidisciplinary team is critical to provide the most effective therapy and diagnosis for a child.

Early Indicators

- There are early indicators that can clue in parents and pediatricians about a possibility of CP.
- The following early indicators are general, and are not necessarily associated with one type of CP, since the motor function impairment during the first year or two of life may alternate between a hypertonic and a hypotonic presentation (Taft, L. T., 1984).
 - An inability to suck or swallow may be the first sign that there is a motor impairment.
 - Sucking and swallowing involves a complex combination of muscular control and timing, and an impairment in this action may be the earliest sign to a parent or pediatrician that further evaluation of the child is needed (Taft, L. T., 1984).
 - Additionally, a child not meeting developmental milestones, such as holding his or her head up independently, sitting, crawling and walking may be indicative of a motor impairment.
 - A child with hemiplegia, where the motor impairment is located on one side of the body (Rennie, J. M., et. al., 2007), may show a hand preference much earlier than a typically developing child. This hand preference is seen

with asymmetrical fisting, or clenching of one hand more often than the other (Taft, L. T., 1984).

- Additionally, a typically developing child will begin to walk using a wide stance to retain balance and posture, using their arms to help with balance.
- With practice, the stance will narrow and the arms will drop; eventually the child will keep his or her arms vertical (Taft, L. T., 1984).
- A child with a motor impairment may begin to walk similar to a typically developing child, but the wide stance will persist (Taft, L. T., 1984).
- Additionally, if a child does not begin to walk, or cannot without assistance, further evaluation may be necessary. A final indicator may be a persistent drooling into early childhood (Taft, L. T., 1984).

Associated Impairments

- The following is a brief description of associated impairments that can occur in conjunction with cerebral palsy.

- Cognitive impairments: Prevalence of cognitive impairments is associated with type and severity of the CP, and increases if epilepsy is present.
 - According to Odding, E., et. al. (2006) 97.7% of those considered to have a severe form of CP have some level of cognitive impairments.
 - In other words, those with tetraplegic CP tend to be severely cognitively impaired.
 - Additionally, 60% of those with hemiplegic CP will have some level of cognitive impairment (Odding, E., et. al. 2006).

- Behavior Problems: Behavior problems are highly correlated with those with CP; it is 5 times more likely to be seen in those with CP when compared to their able-bodied peers (Odding, et. al., 2006).
 - Attention deficit hyperactivity disorder (ADHD), dependency, stubbornness and hyperactivity are commonly seen in those with CP (Odding, et. al., 2006).
 - Parent and self reports are the primary method of retrieving behavioral information (Vohr, B. R., 2007).

- Epilepsy: Epilepsy, or seizure disorder, is seen in 22-40% of those with CP. T

- The variation in prevalence is dependent on type and severity of the CP, as well as the presence of cognitive impairments.
 - In tetraplegic CP with severe cognitive impairment 94% have epilepsy (Odding, et. al., 2006).
- Sensory Impairments: Sensory impairments include hearing, vision, stereognosis, proprioception, and two-point discrimination.
- Stereognosis is the ability to touch or lift an item and determine the weight and shape (Odding, E., et. al., 2006).
 - According to the study done by Odding, E., et. al. (2006), 44-51% of all children with CP will have two-point discrimination problems and stereognosis.
 - Interestingly, unlike epilepsy, the presence and severity of sensory impairments does not follow the severity of the CP (Odding, et. al., 2006).
 - Within the hemiplegic population, sensory impairments seem to be the highest, with 9 out of 10 people having some impairment (Odding, et. al., 2006).
 - Vision impairments are seen in 62% of children with CP. It is thought that the visual problems stem from the original insult to the developing brain (Odding, E., et. al., 2006).

- Hearing impairments are seen, but further research is needed to identify cause and prevalence. Hearing loss and CP is discussed further below.
- Speech Impairments: Speech disorders, such as dysarthria and aphasia, are commonly seen in those with CP.
 - The prevalence of speech disorders is dependent on type and severity of CP, with dyskinetic CP having the highest rate of speech impairments (95%) and diplegic CP the least (20%) (Odding, E., et. al., 2006).
 - Those with CP tend to have different language and communication strategies when compared to typically developing peers.
 - For instance, children with CP will less often initiate conversation, fail to reply, take fewer turns in the conversation and rarely ask questions (Pirila, S., Van Der Meere, J., Pentikainen, T., Ruusu-Niemi, P., Korpela, R., Kilpinen, J., & Nieminen, P., 2007).
 - The language and speech disorders may be a result of the motor impairments in CP.
 - Current research is trying to determine the presence of a specific language impairment (SLI) that occurs within the CP population (Pirila, S., et. al., 2007).

- Endocrine Impairments and Feeding Problems: During the first year of life, children with CP exhibit sucking and swallowing difficulties (57 and 38% respectively), and some difficulties persist into adulthood (Odding, et. al., 2006).

Hearing Loss in the Cerebral Palsy Population

Incidence

- The prevalence of hearing loss within this specific population is a critical component in understanding and treating children with cerebral palsy.
- Cerebral palsy can be accompanied by associated disabilities, including, but not limited to, hearing loss, visual impairments, epilepsy, speech and language disorders, and cognitive impairments (Odding, E., et. al., 2006).
- In order to effectively treat and diagnose a child with CP, the prevalence of associated disorders is important.
- Of those with hearing loss 30.2% will have an additional disability (Daneishi, A., & Hassanzadeh, S, 1984). This webpage will specifically look into the relationship between hearing loss and cerebral palsy.

- According to Robinson, et al. (1973) 12.5% of all those with CP will have a hearing loss as well.
- Considering this research is not current, and that the prevalence of CP has increased, this percentage could be larger.
- Odding, E., et al. (2006) states the incidence of hearing loss is 25% of the CP population of Western Europe.
 - While this incidence is specific to the European population, it can be generalized to the United States because of the similarity of the prenatal care, techniques used during the birthing process and the incidence of CP in the general population.
 - Hypoxic ischemic encephalopathy, hyperbilirubinemia and infection, especially in preterm and very low birth weight infants, can cause both CP and hearing loss (Rennie, J. M., et al., 2007; Sheykholeslami, K., et al., 2000).
 - Unfortunately, much research is needed to find the incidence these exist co-morbidly. In the following section, the method of audiological testing used for typically developing children will be discussed compared to those with CP.

Audiometric Testing

- Before starting any audiological testing, finding out as much information as possible from the parents and family of the child is highly recommended.
- Insights from parents may help you to find the best testing technique for a specific child, as well as increasing the likelihood that you will be able to find an accurate audiometric threshold.
- Following the family-centered approach, taking the parents' insights, goals, and values into consideration will be central to therapy and diagnosis of the child.
- The case study is central in a professional, family-centered, reliable audiometric test.
- In order to fully understand the modifications in the audiometric testing that might be made for a child with a motor function disability, the typical methods of testing need to be explained.
- It is important to remember that all audiometric testing is done with developmental age in mind, rather than the chronological age of the child.

The following modifications are brief, and are the limit of the changes that can be made during the test. For children from birth to six years old with no additional disabilities, and the modifications for CP, the behavioral audiometric testing are briefly broken up as follows:

- **Birth to 6 months of age: Behavioral Audiometric**

Observation (BOA)

- Using this method, the audiologist will present a loud sound and watch for specific movements in the child.
 - For instance, does the child blink, breath in, stop breathing, settle down, or cry.
 - While this is a tool that can be used for the very young child, it does not give us ear specific information or frequency specific information.
 - Additionally, the responses are difficult to see, even for the most seasoned professional.
 - The modifications during this technique of testing would be to make sure the trunk of the body is supported. For very young typically developing infants, this is important as well, but for those with muscle tone disorders it becomes a priority. Being able to see the difference between a startle response and a hypertonic or hypotonic movement is critical in determine if further testing is required.

- **6 months to 2 years: Visual Response**

Audiometry (VRA)

- In this method of testing, the child is conditioned to respond with a physical movement when a sound is presented, using a complex visual stimulus as reinforcement to help condition.
- This method can yield ear and threshold specific information.
- Modifications must be made for a child with CP, even the mildest forms, for this technique of behavioral testing.
- The requirement of a motor function as a way of determining audibility of a sound presents particular problems with a population such as CP, where motor function is impaired.
- A structured seat with straps may be useful and critical in keeping posture of the child throughout testing.
- Understanding the need for multiple visits as a result of the fatigue factor in those with CP.
- If neck is floppy or stiff in the infant using people as the visual reinforcement can benefit the testing.
- Using the motor function that is available to the child, rather than a head turn which may not be, is the basis to whatever modification is chosen.
- The biggest issue of using VRA with children with CP are whether or not the child also has seizures. Flashing lights and moving objects are not recommended for children with a history of epilepsy and other methods of testing the hearing should be found – or different types of reinforcers.

- 2 years to 4 years: Conditioned Play

Audiometry

- Like VRA, reinforcements are used to condition a response in this somewhat older group of children. Rather than looking for a head turn response, we actively participate in a “game” with the child. Toys such as puzzles and blocks can be affective conditioning tools. The child must listen for the tone, when heard place the toy in a bucket indicating that the tone was heard.
- Modifications must be made for those with motor impairments. Again, modifications must be made on a case-by-case situation, and the following are examples of a myriad of options available to the creative clinician.
 - Having the child point to a parent or second clinician rather than placing a toy in the bucket. Reinforcements can be funny faces the parent or clinician makes if the child pointed when the tone was heard. If the child cannot point – the child may be able to lean body in a specific direction or move eyes in the direction of the sound. Any volitional motor movement could be used as a response.
 - If the motor impairment limits the child’s ability to grasp small objects, but the gross motor function is intact, use large toys that are easily held.

- If the child uses an alternative augmentative communication device, perhaps that may be the “toy.” If the child is an effective user of the technology, but may not be able to use the toys, using something they are comfortable with may be helpful. For instance, if there was an animal sound programmed on the AAC, the child may press a button when the tone was heard, making a “moo” as his or her own reinforcement.

- 4 years to 6 years: Audiometric testing

- At this age, the child is usually able to perform the basic audiometric testing that an adult does. Some may need modifications, such as clapping their hands when the tone is heard rather than pressing a button or raising their hand.
- For those with CP, further modifications might have to be made. Like in previous examples, management of posture is critical to reduce frustration, fatigue and pain.
 - For the speech testing involved (SRT and Word Recognition) the level of the child’s speech and language must be considered.
 - Using pictures, and having him or her point rather than say the words. If the motor function of the arms is impaired, then perhaps using the AAC device, or signs that the family knows and understands.

- It is also possible to have only a few options (three toys) and set them a good distance apart in the sound booth. For this general movements towards the three toys could be accepted as heard.
- By separating them inside the sound booth, it can be more obvious as to which toy is being chosen. (Katz, pgs. 469-480).
- All modifications made must be made while looking at the developmental age of the child, for those with CP and without.
 - Since there is a known association with cognitive impairments with cerebral palsy, a flexible plan of testing is critical. A case report from the parents prior to testing will be extremely helpful in determining what testing to do and in what order.
 - Additionally, if the child uses a wheelchair, crutches, or other assistive mobility devices, arranging the sound booth to accommodate these items.
 - The above tests are behavioral, in that they require a behavioral response from the child.
 - Depending on developmental age, and motor abilities, physiological testing may be the only course of action in getting reliable hearing thresholds, since physiological tests do not require a behavioral response.
 - Auditory Brainstem Response (ABR) is a useful tool in finding the hearing threshold of a young child, or one where behavioral measures are unreliable. The ABR measures via electrodes the

activity in the auditory nerve and the brainstem when presented with an auditory stimulus. A major disadvantage of using this test is that muscle movement creates noise, which can make it impossible to record the response. For a child with athetoid/dyskinetic CP, this would be extremely difficult as a result of the sometimes continuous, involuntary movements.

- Sedation can be used, but a medical professional must administer the sedations (Katz).
 - Ototoxic emissions (OAEs) are also a useful physiological test, in which the mobility and function of the outer hair cells within the inner ear.
 - When tones are presented to the ear, the outer hair cells will ‘create’ a sound that is measurable. This measurable output is the OAE. Again, as with the ABR, muscle noise may be problematic, so flexibility is critical (Katz, pgs. 274-298).
- A study by Emmer, et. al. (2003) tells of a possibility of using acoustic reflex thresholds as a possible diagnostic tool for hearing loss.
 - The acoustic reflex threshold is the level of sound required to induce a contraction in the stapedius muscle.
 - Using a broadband noise stimulus, rather than tones, it is possible to find the hearing threshold of a person who is considered difficult to test.

- While this is an intriguing study, more research needs to be done about the efficacy and accuracy of this method of testing.

Rehabilitation and Amplification

- When working with families that have a child with CP and hearing loss a family centered approach can be extremely beneficial.
- A family centered approach is a theory of therapy in which the family is seen as the important decision makers in the child's life rather than the professional.
- In this approach, it is understood that the family knows the child better than anyone else, and therefore should be involved in all decisions regarding the child in question.
- Parents, or guardians are part of the team, rather than observers (Novak, I., Cusick, A., & Lowe, K., 2007).
- This model of therapy applies to all who work with the child and his or her family.
- For the audiologist, listening to the demands of the family and their thoughts on choice of amplification is important in finding the best option for technology.
- A speech language pathologist needs to know the goals of the family in regards to their child's language and articulation, and keep in mind

the differences in language and speech impairments across individuals (Chen, L. & Whittington, D., 2006).

- While a physical therapist and an occupational therapist must know the demands of the household, including what type of house, will there be a large number of stairs that the child is required to use, and goals of the parents in order to provide the most affective therapy and intervention.
- For all professionals involved, working with the parents and the client will be more affective than without.
- However, as the professional, it is important to prioritize and help the parents find effective and goals that are reachable (Novak, I., et. al., 2007).
- Early intervention is important in working with both the hearing loss and the motor impairment (Koman, L. A., 2004). Therapy for children with CP can include speech, language, physical, occupational, optometric, and surgical.
 - Working in conjunction with other therapists and doctors, in addition to the family, can create the best environment for this child (Koman, L. A., 2004).
 - Audiologists must look at all factors related to each individual child.

- Certain factors must be taken into account, such as the physical development, cognitive development, speech and language abilities, as well as goals and needs of the family.
- Each can affect how efficient the audiologic testing is, as well as the efficacy of the treatment.
- For instance, recommendation of cochlear implantation must take into account any additional disabilities that might be contraindicated in pursuing implantation (Daneishi, A., & Hassanzadeh, S., 1984).
- The recommendation of hearing aids, bilateral and unilateral, and listening devices should depend on the hearing loss diagnosed in the child, environmental, cognitive, and familial factors.
- If the hearing loss is bilateral, recommending two hearing aids is optimal, in order to give the child as many opportunities as possible to hear and understand language.
- If the child uses a wheelchair, and has impaired muscle tone in the neck and trunk perhaps unilateral hearing aids, or a body aid is the most effective method of amplification.

- The audiologist must be flexible and creative in working with the child with CP in fitting amplification.
- For instance, this flexibility can be seen when teaching a child to manipulate their hearing aids during insertion.
- In very very unusual cases, children with significant neurological conditions and hearing loss are unable to handle more than one modality to receive language- i.e. visual or auditory.
- In these cases we would evaluate the child's strength - auditory only and then visual only and make a decision about whether the prognosis is better in one modality or the other.
- In such cases, we sometimes do not amplify because multisensory access seems to actually decrease the child's communication and learning progress.

Discussion

- There are no real limitations on the modifications that can be made in both testing and treating this population of children.
- All modifications must be made for the specific child you are testing.

- Each child with cerebral palsy will have a unique presentation of tonal quality as well as severity and additional symptoms.
- A modification for one child may not be effective for the next.
- Remembering the individual variability in this population of children can help the clinician to diagnose and treat affectively.
- Cerebral palsy is a broad and indistinct disorder, ranging in severity, type and affected areas of the body, and being flexible in diagnosing, testing, and therapy is necessary.

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