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Spinal Muscular Atrophy Type II, Implications for the Life Care Plan: A Case Study Involving Opposite-sex Siblings with Spinal Muscular Atrophy Type II

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Abstract

This paper presents a case study involving a family in which male and female siblings have spinal muscular atrophy (SMA) type II. The paper explores the unique needs of a male and female client with spinal muscular atrophy type II and the implications for the life care plan, as well as, considers the dynamics of care planning for a family with multiple individuals with a disability.

Background

According to Hirtz, Iannaccone, Heemskerk, Gwinn-Hardy, Moxley, and Rowland (2005), "Spinal muscular atrophy (SMA) is the most common fatal neuromuscular disease of infancy and the third most common diagnosis of neuromuscular diseases seen in clinics for children < 18 years" (p. 1352 as cited Iannaccone & Khatri, 2005). Spinal muscular atrophy is a non-sex-linked inherited disease causing the degeneration and death of motor neurons in the anterior horn of the spinal cord (Rubin, 2008; Wang, Kleyn, Vitale, Ross, Lien, Xu, et al., 1995). As such, SMA results in progressive muscle weakness and wasting (Rubin, 2008). The National Institute of Neurological Disorders suggests, "It is caused by an abnormal or missing survival motor gene (SMN1), which is responsible for the production of a protein essential to motor neurons. Without this protein, lower motor neurons in the spinal cord degenerate and die" (National Institute of Neurological Disorders and Stroke/NINDS, 2008). Further, a collaborative study on the natural history of children and juvenile onset SMA reveals that "over 90% of cases of SMA are recessively inherited with the responsible gene located on chromosome 5q" (Zerres, Rudnik-Schöneborn, Forrest, Lusakowska, Borkowska, & Hausmanowa-Petrusewicz, 1997, p. 67). Also reported, "one in 50 people carry this autosomal recessive gene" (Hirtz, et al., 2005, p. 1352, as cited in Emery, 1991; Merlini, Stagni, Marri, & Granata 1992; & Pearn, 1978), "with a frequency of 8 to 11 per 100,000 live births."

In the United States, SMAs are reported to be the second most common autosomal-

recessive inherited disorders after cystic fibrosis (Tsao & Armon, 2009). The Spinal Muscular Atrophy Foundation reports, "one in 35-40 Americans are carriers, or approximately 7 million people. Over 25,000 Americans are believed to suffer from SMA" (SMA Foundation, 2009). Per Wang, Finkel, Bertini, Schroth, Simonds, Wong, et al. (2007), there are other forms of severe SMA not linked to the SMN gene that have varying inheritance, linkage, and genes; however, such a discussion is beyond the scope of this paper.

Classification and Disease Process

Per Wang, et al. (1995):

> "Childhood-onset SMAs are often divided into three categories based on disability and age at onset: SMA type I, or Werdnig-Hoffman disease, is the most severe, leading to death in the first years of life; SMA type II, or intermediate SMA; and SMA type III (Kugelberg-Welander disease) are milder, with later age at onset" (p. 202).

SMA type II is also known as intermediate, chronic or juvenile SMA (Hirtz, et al., 2005; NINDS, 2008). Some classification systems such as the International Spinal Muscular Atrophy Consortium (ISMAC) divide the SMA groups into 4 categories (Wang, et al., 2007, as cited by Dubowitz, 1995; Munsat & Davies, 1992); however, the majority of the literature researched for this paper categorized SMA as types I, II, and III, so all references to SMA II will be in reference to the use of 3 categories unless otherwise noted.

According to Hirtz, et al., (2005):

> "Milestones are usually normal until the onset of weakness between 6 and 18 months of age. The legs tend to be weaker than the arms, so children often come to medical attention because of failure to walk. The pattern of deep tendon reflexes may be variable. Children can sit without support when placed" (p. 1352-1353).

Tsao and Armon (2009) also state that "patients with SMA often have above-average intelligence quotients (IQs) and demonstrate high degrees of intelligence." Further, Zerres, et al. (1997) report "motor functions may vary; some children have early difficulty sitting or rolling over, whereas others learn to crawl or stand with support" (p. 69). The National Institute of Neurological Disorders (2008) notes that other children with SMA may be able to walk with help, while Russman (2002) notes that although "sensation is intact in all patients with SMA II, approximately 70% lack tendon reflexes" (p. 1005). Nordgren (2005) further notes that "some muscle groups are preserved and others severely involved. Often, reflexes are preserved when a muscle group is preserved. Many children with SMA have minipolymyoclonus, a regular, small amplitude of movement that is most prominent in the distal extremities" (p. 1193).

In a study by de Groot and de Witte (2005), "the rate of progression of SMA is related to the severity of impairments such as scoliosis or lung function" (p. 37, as cited by Robinson, Gakasji, Delaney, Williamson, & Barrie, 1995; Carter, Abresch, Fowler, Johnson, Kilnmer, & McDonald, 1995). Zerres and Rudnik-Schöneborn also cite that functional abilities are a factor (as cited in Zerres & Rudnik-Schöneborn, 1995; Rudnik-Schöneborn, Hausmanowa-Petrosewicz, Borkowska, & Zerres, 2001, as cited in de Groot, IJM., de Witte, LP. (2005)). The clinical course may also have periods of arrest (Zerres, et al., 1997).

As the child grows, there is an increased demand placed upon the remaining motor neurons resulting in a progressive loss of muscle control and movement, if ever acquired (SMA

Foundation, 2009). Iannaccone, Hynan, and the American Spinal Muscular Atrophy Randomized Trials (AmSMART) Group (2003, as cited in Iannaccone, Russman, Browne, Buncher, White, Samaha, et al., 2000) showed "in a long-term prospective study that patients with SMA are very stable with regard to strength" (p. 1131), and that "motor neuron unit counts seem to remain stable up to age 18 months" (Iannaccone, et al., 2003, p. 1135). Persons with SMA II retain their general level of strength, but as they grow physically, that level of strength is not sufficient to operate larger, heavier muscles at the previous functional level.

In their research, the SMA Foundation (2009) reported that, "children suffer increasing orthopedic deformities, such as the spine, hips, contractures and ultimately fatal respiratory complications. Repeated surgery is often required to prolong life, which may include procedures such as tracheostomies to assist in breathing; tube feedings for nutrition; and spinal fusion to mitigate spinal damage" (Frequently Asked Questions, http://www.smafoundation.org/index.php?option=com faq&Itemid=32). Additionally, the National Institute of Neurological Disorders and Stroke (NINDS, 2008) reported that "There is no cure for SMA. Treatment consists of managing symptoms and preventing complications" (Fact Sheet, http://www.ninds.nih.gov/disorders/sma/detail_sma.htm?css). As previously noted, "the rate of progression of SMA is related to the severity of impairments" (de Groot & de Witte, 2005, p. 37).

Complications of SMA

Respiratory: "Pulmonary disease is the major cause of morbidity and mortality in SMA types 1 and 2. Respiratory care of patients with SMA is essential to their survival and quality of life" (Wang, et al., 2007, p. 1037). Factors affecting respiratory function are swallowing dysfunction, presence of reflux, and scoliosis (Wang, et al., 2007) and individuals have a progressive decline starting with recurrent chest infections, oxygen desaturation and hypoventilation at night until displaying daytime hypercarbia and respiratory failure (Wang, et al., 2007, as cited by Mellies, Dohna-Schwake, Stehling, & Voit, 2004; Mellies, Raguette, Dohna-Schwake, et al., 2003; Raguette, Mellies, Schwake, et al., 2002). "Acute and chronic management includes mechanical insufflation/exsufflation or manual cough assist and non-invasive ventilatory support" (Wang, et al., 2007, p. 1038).

Gastrointestinal (GI): GI concerns with SMA type II are feeding and swallowing problems related to bulbar dysfunction and GI dysfunction related to decreased motility and reflux (Wang, et al., 2007). According to Wang, et al. (2007), feeding problems should be assessed by speech or occupational therapy. When reflux is combined with respiratory compromise in an individual, the risk for aspiration and pneumonia increases.

Nutritional: Excessive weight gain is more common in SMA type II patients who are able to sit (Wang, et al., 2007). "Children with SMA may have acceptable fat mass, but may plot as underweight based on height/weight criteria due to the decrease in lean body mass. Decreased activity and lean body mass will lead to reduced resting energy expenditure and increased risk of obesity" (Wang, et al., 2007, p. 1040). To monitor weight and nutritional status, Wang, et al. (2007) recommend a nutritional assessment by a dietician or other healthcare provider proficient at nutrition at each evaluation for growth. They also suggest that SMA patients are particularly vulnerable to catabolic states which may progress to metabolic decompensation and fasting states in which they are more likely to develop hypoglycemia (Wang, et al., 2007).

Orthopedic: "Muscle weakness of varying severity limits motor function of the trunk and upper and lower extremities, resulting in contracture formation, spinal deformity, limited

mobility and activities of daily living, and increased risk of pain, osteopenia, and fractures" (Wang, et al., 2007, p. 1042). "In sitters, wheelchair mobility, contracture management, physical therapy, and occupational therapy are of the highest value, with strong considerations for spine and limb orthotics and spine surgery" (Wang, et al., 2007, p. 1042). Further, scoliosis occurs in 50% of the general SMA population (Wang, et al., 2007). "Hip subluxation is common in SMA, however is rarely painful and there is a high risk of recurrence despite surgical correction" (Wang, et al., 2007, p. 1042, 1045). Zenios, Sampath, Cole, Khan, and Galasko, (2005) reported that they found no justification to operate on hips which are at risk of dislocation due to the complexity of hip surgery and anesthesia in patients with SMA, their frequent hospitalization for other medical reasons, and the high rate of recurrent subluxation. It is noteworthy that in a study by Abresch, Carter, Jensen, and Kilmer (2002), the adults with SMA did not show significant frequency and severity of pain associated with other slowly 54 progressive neuromuscular disorders and that the SMA sample reported no more pain sensitivity than the general population.

Diagnostics

Several papers report that "blood testing of the DNA for the survivor motor neuron deletion is the first diagnostic test that should be done" (Nordgren, 2005, p. 1193; Rubin, 2008; NINDS, 2008), which "detects the causative mutation in about 95% of patients" (Rubin, 2008). Muscle biopsy is now only done occasionally (Rubin, 2008) although "on muscle biopsy, patients with SMA show a pattern called "grouped atrophy" (Nordgren, 2005, p. 1193), with fiber size variation (Wang, et al., 1995). Serum enzymes (e.g., creatine kinase/CK, aldolase) may be slightly increased, and amniocentesis, done if family history is positive, is often diagnostic (Rubin, 2008). Tsao and Armon (2009) report that "the accuracy of prenatal prediction by chorionic villi sampling and amniocentesis was 88-99%; however caution is recommended in the presence of atypical features because these clinical variations may represent other pathogenic processes" (http://emedicine.medscape.com/article/1181436). Other diagnostics may include electrodiagnosis with nerve conduction velocities/EMG (NINDS, 2008).

Prognosis

Tsao and Armon (2009) report that "SMA II survival probabilities at ages 2, 4, 10, and 20 years were 100%, 100%, 98%, and 77% respectively" (p. 10). Further, Nordgren (2005) states that "the prognosis is variable, with some patients surviving until the second or third decade" (p. 1193); however, "the SMA II prognosis is improved primarily due to supportive care" (Hirtz, et al., 2005, p. 1352).

Per Zerres, et al. (1997), "In an individual patient, the predication of life expectancy is not possible on the basis of age at onset alone" (p.70). "Among siblings, the age of death varies among deceased sibling pairs with chronic childhood SMA despite a similar 'age of onset' and comparable motor functions" (p. 70, as cited from Rudnik-Schöneborn, et al., 1994). "Patients with SMA II who can stand, however, have a better prognosis" (p. 71). Per the study performed by Zerres, et al. (1997) on the natural history of childhood and juvenile onset proximal SMA II and III, "the data show that a rigid classification system with defined age at onset and age of death is still limited, because life span describes only one aspect of natural history. Others, like periods with preserved motor functions (e.g. intervals between onset and wheelchair), are other important characteristics in the clinical course. Strictly defined ages of onset and death, as used in many classifications should be replaced by the definition of achieved motor

functions (ability to sit or walk) in addition to data about probability for survival and preserved motor functions" (p. 71).

It is noteworthy that many patients with SMA do not fit neatly into their diagnostic classification at onset and may functionally seem to be closer to a higher or lower diagnostic classification (de Groot & de Witte, 2005; Wang, et al., 2007); thereby further disproving rigid life expectancies predicated by disease classification alone. For the SMA II group, quality of care, level of function and nature of complications will greatly affect life expectancy (de Groot & de Witte, 2005; Russman, 2002; Zerres, et al., 2007). The life care plan will be a tremendous tool in assisting to plan to prevent complications, enhance quality of life, possibly prolong life, and provide a tailored cost projection to support these measures. For purposes of the life care plan, life expectancy should be made by a physiatrist who has thoroughly reviewed the patient's medical records (Bonfiglio, 2009, as cited Weed & Berens, 2009).

The Future

Genetics: "Several studies are looking at ways to stimulate the production of the SMN protein from the SMN2 gene. Most children with SMA have functional SMN2 genes. This research is being tested in animal models" (NINDS, 2008; Iannaccone, et al., 2003, as cited in Lorsen, et al., 1998; Monani, et al., 2000). Look for researchers to determine whether the Gross Motor Function Measure (GMFM) changes over time in patients whose mutation analysis and SMN2 copy number were known to provide some phenotype-genotype correlation with motor function (Iannaccone, et al., 2003, p. 1135)." The American Society of Human Genetics Board of Directors and The American College of Medical Genetics Board of Directors (ASHG/ACMG) report in 1995 that "As genetic testing for children and adolescents becomes increasingly feasible, research should focus on the effectiveness of proposed preventive and therapeutic interventions and on the psychosocial impact of the tests" (p. 1234).

Stem Cell Research: The Muscular Dystrophy Association (MDA) is supporting the Rothstein group to refine stem cell treatment by determining how cells promote spinal cord repair in rodents. The MDA supports other research groups for stem cells research (SMA Foundation, 2009).

Clinical trials: According to Hirtz, et al., (2005), "families should be referred to enroll in clinical trials as soon as they are diagnosed" (p. 1355). The American Spinal Muscular Atrophy Randomized Trials (AmSMART) Group is an organization of five pediatric medical centers performing clinical trials in children with SMA to develop valid and reliable outcome measures (Iannaccone, et al., 2002). Additionally, the "National Institute for Neurological Disorders and Stroke (NINDS) conducts research on SMA in laboratories at the National Institutes for Health (NIH) and also supports research through grants to major medical institutions across the country" (NINDS, 2008). "Clinical trials are testing the benefits in children of hydroxyurea, riluzole, valproic acid, and phenylbutyrate-drugs that have a promise in treating the symptoms of SMA" (NINDS, 2008). According to published research, "a new requirement for clinical trials is measurement of the patient's perception of disease state, the quality of life (QOL) and may include family members" (Iannaccone, et al., 2002, p. 1449, as cited in Hanestad, 2001 and Farquhar, 1995).

Drugs and nutrition: "The MDA supports the investigation of other possible treatments for motor neuron disease, including nerve growth factors and dietary supplements" (SMA Foundation, 2009, http://www.smafoundation.org/index.php?option=com faq&Itemid=32). The Spinal Muscular Atrophy (SMA) Project, funded by The National Institute for Neurological Disorders and Stroke (NINDS), is working to identify drugs already in use that

increase the level of SMN protein for potential leads for further drug discovery and clinical testing" (NINDS, 2008). "Five drugs are known to increase SMN2 expression in cultured cells as well as have been used previously in humans: aclarubicin, hydroxyurea, indoprofen, phenylbutyrate, and valproate. Because a correlation between histone acetylation and gene expression has been discovered (Zupkovitz, Tischler, Posch, Sadzak, Ramsauer, Egger, et al., 2006, as cited in Allfrey, Faulkner, & Mirsky, 1964), histone deacetylase (HDAC) inhibitors are also under consideration for further evaluation" (Hirtz, et al, 2005, p. 1354).

Future research: Recommendations have been made to establish an international SMA study group, expedite drug development for clinical trials in SMA, form a working group on outcome measures for clinical trials, expand the existing SMA patient registry to an international registry, and develop guidelines for clinical care (Hirtz, et al., 2005, p. 1356). Trends in research are predicted to "assess the impact and economic burden of SMA, quality of life, natural history, epidemiology, and family history" (Hirtz, et al., 2005, p. 1355).

Methodology

Verbal and written consent and assent was obtained by the individuals and their mother to construct a case study based on the children's personal disease process, family dynamics and implications for the life care plan. A questionnaire was developed and modified utilizing recommended data collection by the Spinal Cord Injury Information Network (2006) and information applicable to the life care plan from the *Life Care Planning and Case Management Handbook, 3rd ed.* (Weed & Berens, 2009). The first questionnaire was given to the siblings' mother and was returned with written responses. After the literature search and review was completed, a second questionnaire was developed unique to the SMA II disease process. The second questionnaire was answered during a telephone interview with each individual: the affected male and female siblings and the individuals' mother for consistency and accuracy. Further questions incurred during the writing of the paper were answered by the siblings' mother via email correspondence and/ or in person. After completion of the questionnaires, answers were reviewed by the life care plan student who is a registered nurse. An in-person home interview was not conducted; however the individuals discussed in this paper are personally known to the author and have been randomly observed several times a month throughout a period of five years. The author also has been a guest in their home on one occasion. There was no formal medical record review conducted. All information was obtained from interviews with the individuals, their mother who has a healthcare background, and pertinent medical literature pertaining to the SMA II disease process. Four principle researchers among three universities were consulted to verify the author complied with ethical guidelines in case reporting according to Institutional Review Board standard recommendations (University of Florida, 2007). The case study itself and beneficial information related to life care plans for individuals wit spinal muscular atrophy type II were shared with the family.

The SMA Siblings

SMA II male: The SMA II male sibling, who will be referred to as "Victor," was born via uncomplicated vaginal delivery. There was no genetic testing performed during the pregnancy and no family history of SMA. The parents initially noticed that when they changed his diapers his legs were somewhat floppy, as opposed to the older siblings. He would never take a step, but would stand and fall forward into their arms, thinking it was a game. During frequent medical testing to diagnose Victor, his family was informed that he would only live

for six months. His SMA II was diagnosed by muscle biopsy when he was just over two years old. His parents were told he would not live past age five. Healthcare providers recommended the parents offer the child comfort care; however Victor was kept as active as possible. He received physical therapy from age 3 to 5 about once a month and occupational therapy approximately 4 times a year. He used a walker until he was between the age of three and five and used a standing wheelchair to play tee ball. His mother reported that he used a walking wheelchair intermittently with the power wheelchair. Victor was able to crawl until age thirteen. He developed scoliosis that progressed to 90 degrees. During an attempted anterior spinal fusion, Victor had significant intra-operative bleeding and was reported to have had an infarct/ischemia in one of his thoracic vertebrae. The father reported he was told the instrument measuring nerve transmissions to Victor's legs indicated there were no transmissions. Post-operatively, he was found to be paraplegic. Several weeks later his scoliosis was corrected with a posterior spinal fusion.

At the time this paper was drafted, Victor was 22 years-old, five feet four inches tall and weighed ninety-five pounds. He denied any need for counseling related to coping with the SMA. He does not require any hearing or visual aids. He has not had any respiratory complications related to the SMA. He does not have any known cardiac disease. On several occasions after some episodes of vomiting, Victor experienced hypokalemia requiring hospitalization. His healthcare providers have been assessing his condition to rule out diabetes. He required insulin for a brief period, but Victor reported he no longer required insulin injections and did not take oral hypoglycemic medication. Victor monitors his blood sugar independently at home. He is not on a special diet, but per his mother, he watches his carbohydrate intake and will take short-acting insulin if he ate a large meal. Given the possibility of diabetes, he has his foot care provided by a podiatrist. Victor self-catheterizes himself several times per day and does not require any special bowel program. He has mild contractures of his elbows. He is able to write and type despite slight tremors in his fingers. Victor has bilateral contractures of his legs with no movement except a couple of his left toes. He does not have any unilateral weakness post-infarct. He retained most sensation in his legs post-paralysis. He is able to shift his own weight in his wheelchair, but is dependent for any transfers in and out of bed, and transfers for toileting. He does not use any special seating beyond the standard wheelchair cushion. He has not had any problems with skin breakdown. Victor uses a "picker-upper" to assist with reaching. He and his sister are evaluated yearly in a SMA clinic. His immunizations were current. On multiple occasions, Victor became ill requiring hospitalization after receiving the influenza immunization. He no longer receives an annual flu shot. His additional past surgeries include wisdom teeth extraction. He does not wear a medical alert bracelet and there is no medical alert system in the home. He has not discussed end-of-life issues with his family. He has not made an advanced directive or medical living will to date. He reported that he had not given much consideration to whether he would be willing to use a ventilator or have tube feedings should his condition deteriorate.

Victor received home schooling and several years of education in a local private school. He received state-sponsored vocational testing after high school and was assessed to need only driving services. He completed six months worth of driving in a modified van and obtained his driver's license. He plans to buy a modified vehicle after college. He currently uses the local mobility bus to go to college or is driven by family. Victor is to receive state-sponsored vocational counseling after he graduates college. He is due to graduate with degrees in math and computer science. He is employed part-time through a work-study program editing college sports videos and is training to assist with data entry for a local business. Upon

graduation, Victor would like to obtain employment with the government using his math degree. He is undecided whether he would like to pursue marriage and a family, stating that for now, it was "not a priority." He was unsure whether he would consider genetic testing if he pursued a family of his own. He reported he was content to live with his family members who remain in the home which consists of his parents, his sister with SMA, and a younger sibling.

Victor reported his friends and family are supportive. In the past he was active in organizations related to SMA. He played power hockey, power soccer, and power football at a facility designed for athletes with special challenges. He has competed in national and one international tournament. His practice area is in his home living room/dining area.

A typical day for Victor consists of being transferred out of bed by one of his parents, sometimes requiring the use of a Hoyer lift, to a powered wheelchair. He reported he is able to dress his upper body, but needs assistance for his lower body. He is able to participate in homemaking activities such as folding clothes, general picking up, and some cleaning providing it is at a level that he can reach. He attends college four to five days per week and uses transportation as stated above. He reported he generally brings lunch to college, but occasionally purchases his food. In the evening, he is assisted in transferring to a chair in the shower. He is unable to lie prone at night. He is unable to turn himself during the night and will call one of his parents using his cell phone when he wants to be repositioned. He does not use any night splinting.

SMA II female: The SMA II female sibling, who will be referred to as "Faith," was born via uncomplicated vaginal delivery. There was no genetic testing performed during the pregnancy despite having a sibling with SMA II. An early assessment by Faith's doctor revealed that the child was believed not to have SMA. As an infant, Faith was able to sit up, crawl and walk holding onto items such as chairs. Her mother began noticing that the child had similar symptoms to the older SMA sibling such as manually moving her legs with her arms for purposeful movement. Faith was diagnosed with spinal muscular atrophy type II at the approximate age of 11 months.

By the age of three, Faith's parents involved her in swimming lessons at a local medical center specializing in spinal injuries and diseases. She was able to ambulate with the use of a walker and leg braces until approximately age four, at which time she began using a powered wheelchair. She received courses of occupational therapy from age three to five, when she "aged-out" of the local district program. Faith was able to crawl until approximately age six, when she broke her leg after falling out of her wheelchair. After the fall and fracture, she was largely immobilized and developed bilateral leg contractures. Post-injury, she developed pneumonia, but did not require any respiratory therapy or mechanical equipment to assist with breathing. At age 11, Faith dislocated her right hip. She reported it was painful at first, but that the pain has subsided to intermittent pain, and that she does not require any pain medication. Faith's family owns a blow-up pool and Faith is able to walk on her feet in the water. The child reported the hip pain is worse when walking in the pool. She reported that she would like to inquire about having the dislocation surgically reduced about the time she has her spinal fusion to correct her scoliosis.

At the time this article was drafted, Faith was 12 years-old, five feet four inches tall and weighed eighty-five pounds. She is an alert and intelligent child who knew a great deal about her disease process. She and her mother reported that she has anxiety unrelated to the SMA, which Faith described as being related to being "a perfectionist" and for which she takes medication daily. Her only surgery to date has been a tonsillectomy and adenoidectomy. Faith

wears glasses for watching TV. She denied any problems with swallowing. Other than one episode of pneumonia, she has not had any respiratory complications. She is not on a special diet. Faith has scoliosis which has exceeded 80 degrees and is scheduled for a posterior spinal fusion. She wears a thoracolumbosacral orthosis (TLSO) during the day and is able to sleep without the brace at night, although she reported she can feel her back "rebounding" to the pathological degree of curvature. She is able to roll from her back to her stomach, but is not able to roll from her stomach to her back. She uses a powered-wheelchair for mobility. She reported her strength is that of a 2-year-old. She has the classic tremors of the hands that are common with SMA. She is able to move her legs and likes to sit on the edge of her wheelchair and swing them. She is partially dependent on her mother for dressing and totally dependent for transfers in and out of bed and to the toilet. She has not had any problems with skin breakdown. She uses a "picker-upper" to assist her with reaching items. Faith is able to control her bowel and bladder. She reported she has had several urinary tract infections which she believed was because she waited too long to relieve herself when she was preoccupied. Her immunizations are reported to be current, including an annual flu shot. She does not wear a medic alert bracelet and as cited above, there is not a medical alert system in the home. Faith has not had any end-of-life discussions related to her medical care with her family.

Faith is homeschooled as were her other 5 other siblings before her. She is schooled in a seventh-grade curriculum and participates in a local private school co-op. She enjoys swimming, music, and watching movies. She likes to play board games with her brother and mother. Faith reported she likes math and novels that are part of a series. She is able to help with household chores such as folding laundry and food preparation provided the working surface is low enough. She has considered driving when she gets older, but she reported that she is nervous about the written driver's test and so is "not in a hurry to drive." She was unsure what career if any she wanted to pursue after high school. She stated that she would like to be married some day and be a mother. She stated that she would want to pursue genetic counseling with her future spouse to determine the probability of conceiving a child with SMA.

Faith described her family and friends as "awesome." She has not required counseling to date to deal with any loss of function. She is active within her local church and with associations pertaining to her disease process and has participated in a drug study. She attends an over-night camp one week per year that provides a 1:1 counselor per camper and is tailored to needs related to her disease process.

Faith has reported her typical day is as follows: Upon awakening, her mother lifts her out of bed and transfers her into the bathroom. Her mother applies her TLSO brace and assists her with dressing. She then has a "quiet time" and eats breakfast. After breakfast she is schooled and takes part in home recreational activities such as using the computer, swimming, and playing with one of her siblings. She then generally has a bathroom break followed by lunch. Her afternoon consists of playing and dinner. After toileting at night, Faith's father transfers her into bed and performs some range of motion exercise to her legs, talks with her, and prays. When asked by the author if she had anything to communicate about her disease or life, Faith stated, "I used to think, why am I in a wheelchair? Then I realized that I wouldn't have met some of the friends I have if it I wasn't."

Family dynamics: The SMA siblings' family consists of 2 parents and 6 children. The father is a teacher and a small business owner. The mother has a degree in the healthcare industry, but does not currently practice in her specialty. She has homeschooled all of her children and runs a local homeschool co-op. Three siblings are grown and no longer live in

the home. One unaffected sibling lives at home with the SMA siblings. There has been no further genetic testing for SMA to the knowledge of the family members questioned for this paper. There is a known family history of diabetes, cancer, heart disease, myocardial infarction, high cholesterol and irritable bowel syndrome. Various family members have volunteered in a SMA support organization for approximately 20 years.

The mother reported the effects of caring for two disabled children included increased stress and financial burdens, challenges in time management, and marital stress. She reflected on how during the early years, she had to fight with providers to obtain a walker for Victor. The family provides all routine care related to activities of daily living, housework, and household repairs. The family had once used personal care attendants to assist with the care of the SMA siblings, but discontinued the home health aid service to care for their own children. Since the older siblings have now moved out of the home, caring for the SMA siblings has become more difficult. This includes being mindful of changes such as rethinking and planning an emergency escape route in case of fire or disaster. Faith had a nurse case manager for her SMA until she became ineligible due to a change in the family health insurance. Victor carries his own health insurance and has a nurse case manager to help navigate the medical and insurance systems. The mother keeps a computerized journal and a folder of the siblings' health history in case of emergencies. There is no family will or advanced directives for the children or parents. The mother of the SMA siblings and Faith reported that they believed the family had a good social support system. Victor reported he had support and that he did not know another way of life.

The family has working fire alarms and an escape plan; however the mother reported the escape plan will need to be revised since one of the older siblings no longer lives in the home. Other details related to the current and projected care of the SMA siblings are listed below in the next section.

Implications for the Life Care Plan

Per Evans, Drennan, and Russman (1981) (as cited in Dubowitz, 1974),

> "The majority of patients with chronic infantile and juvenile forms of spinal muscular atrophy survive to adult life and require careful planning for their long-term care and support. Maximal physical function developed by the individual, correlates well with the onset and severity of secondary deformity...This information allows anticipation of the problems and plans for the treatment to be made from early childhood" (p. 520).

Evans, et al. (1981), also suggest that "a specific programme of care is recommended for each of the function groups" (p. 516). Further support for detailed planning of care for those with SMA is cited by Hirtz, et al. (2005) who stated the prognosis of SMA II "is improved primarily due to supportive care" (p. 1352), and by Russman (2002) who reported "the prognosis for life expectancy is more closely related to maximum function achieved than to age of onset" (p. 1005). Tsao and Armon (2009) call for maximizing the patient's independence and quality of life at each stage of the disease by a multi-disciplinary approach.

As cited in Weed and Berens (2009):

> "A Life Care Plan is a dynamic document based upon published standards of practice, comprehensive assessment, data analysis and research, which provides an organized concise plan for current and future needs with associated costs, for individuals who have experienced catastrophic injury or have chronic health care needs" (p. 3).

Maybe the best support for the life care plan as applied to those with SMA is *The International Standard of Care Committee for Spinal Muscular Atrophy*, who stated "the most appropriate care for patients with SMA should be tailored according to their current functional status rather than the original classification of disease types because these represent the best level of function rather than the present status" (Wang, et al., 2007, p. 1030).

In 2005, *The International Standard of Care Committee for Spinal Muscular Atrophy* was formed, and in 2007, a *Consensus Statement for Standard of Care in Spinal Muscular Atrophy*, was published in the *Journal of Child Neurology*. Per the authors, "the goal of the consensus statement was to establish an initial practice guideline for clinical care, not to serve as a practice standard. The authors called for a practice standard to help with the multidisciplinary care of SMA patients" (Wang, et al., 2007; Tsao & Armon, 2009). There is no known published literature to date that has applied these guidelines to the life care plan.

Categories of care, also known as "Elements of the Life Care Plan" covered in the application of the life care plan to the SMA siblings was described in the *Life Care Planning and Case Management Handbook, 3rd ed.* (Weed & Berens, 2009). For purposes of this paper, general recommendations will be made followed by gender-specific recommendations. A sample life care plan on the male sibling can be found in Table 1. This section is not meant to discuss how to compose a life care plan, but to apply the principles of life care planning to the SMA siblings using information obtained from the sibling interview assessments and relevant published literature.

<u>*Routine medical care:*</u>

Ophthalmology: Each sibling will require a yearly ophthalmology exam. Faith's glasses, including a spare pair, would be written into the life care plan, but not listed as a cost as it is not related to her disability. Replacements could be expected as needed for changes in prescriptions as the child only wears them to watch television. Costs would be covered by the family or insurance.

Pulmonology: "Pulmonary disease is the major cause of morbidity and mortality in SMA types 1 and 2. Respiratory care of patients with SMA is essential to their survival and quality of life" (Wang, et al., 2007, p. 1037). Faith has attended yearly pulmonary visits for pulmonary function testing. Victor is evaluated approximately every 2 years. Both children have had multiple sleep studies to rule out sleep-disordered breathing/ventilatory insufficiency due to their neuromuscular disorder (Mellies, et al., 2003), which have reportedly been stable. "Respiratory assessment should include evaluation of cough effectiveness, observation of breathing, and monitoring gas exchange" (Wang, et al., 2007, p. 1038). "Parents should be instructed in chest physiotherapy to clear the lungs and airway" (NINDS, 2008), and chest physiotherapy can be taught to the parents by the pediatrician, primary care physician (PCP), or pulmonologist during a routine evaluation at no cost.

Neurology: Victor had been evaluated by a neurologist on a yearly basis and Faith has attended 1-2 times per year.

Orthopedic evaluations: Victor has had spinal x-rays every 1-2 years post-spinal fusion. His sister has required orthopedic evaluations every 3-6 months with x-rays for her scoliosis. She can be expected to attend orthopedic evaluations every 1-2 years after her spinal fusion and recovery.

Endocrinology: Victor is evaluated by an endocrinologist every 3 months to rule out diabetes or a metabolic disorder. This would be projected to change to 1 time per year when stabilized unless changes in condition occurred.

Podiatry: Victor has visited the podiatrist every 9 weeks for foot care related to his possible diabetic condition.

Primary Care Physician: Victor is evaluated by his PCP yearly for routine health evaluations.

Pediatrics: Faith is evaluated by her pediatrician every 6 months for general health monitoring. Her care under a pediatrician can be expected to continue until ages 16-18 when her care should be transferred to an adult PCP practitioner.

Gynecology: Yearly gynecologic exams are anticipated for Faith to begin after the onset of menses and/or as needed to screen for cervical and breast cancer.

Routine lab tests: Routine lab tests anticipated for both siblings are yearly serum enzymes, chemistry panels, urinalysis and blood counts. A prealbumin will be necessary to evaluate protein status. Victor can expect HgbA1c tests for evaluation of his blood sugar status. He may also receive more frequent blood work due to his history of metabolic complications and possible diagnosis of diabetes, especially if the diabetes is confirmed and/or he has recurrent metabolic illness.

Projected evaluations:

Physical therapy evaluation: A physical therapy evaluation is recommended yearly for life for each sibling to evaluate and preserve function. The evaluation has been conducted at the SMA clinic annually. See also *Therapeutic Evaluations*. The siblings' both have contractures. According to Wang, et al. (2007), "Contracture management and exercise are a major focus of treatment, with implementation of a regular stretching and bracing program to preserve flexibility" (p. 1043).

Psychological evaluation: A psychological evaluation can be expected as needed for each sibling for coping with multi-dimensional loss and altered relational dynamics. Ongoing counseling may or may not be required.

Family counseling: Family counseling should be provided for in the life care plan as the family relationships and related stress will impact the quality of care of the siblings emotionally, physically, mentally and spiritually.

Recreational therapy: A recreational therapy evaluation is recommended every 5 years for each sibling or to coincide with each stage of functional loss throughout life. "Regular exercise should be encouraged to maintain fitness and endurance and might include swimming and adaptive sports" (Wang ,et al. ,2007, p. 1043). Given the advances and creativity of physical therapy (PT) facilities, it is possible that the appropriate PT facility could also cover some of the recreational therapy recommendations.

Nutritional consultation: "Ensuring optimal caloric intake enables patients to use weak muscles to their maximum capacity without incurring obesity as a comorbid condition" (Tsao & Armon, 2009, p. 9). Further, Wang, et al. (2007) recommended "assessment of nutritional intake by a dietician or other healthcare provider proficient at nutrition at each visit" to a physician for growth evaluation (p. 1041). Nutritional assessments should be routine throughout the lifespan.

Physiatry consultation: Both siblings would benefit from an initial physiatry consultation as the physiatrist specializes in rehabilitative medicine.

Home evaluation: The home evaluation is a one-time allowance for architectural renovation recommendations.

Vocational evaluation: Faith will need a one-time vocational evaluation in senior year high school for recommendations for career options. This service is provided free of charge

through her state of residence. Vocational evaluation by a private agency would be approximately $65-85 dollars per hour.

Vocational counseling: For both siblings upon completion of school, specialized training, or college to assist with job placement/options. This service is provided to the siblings at no charge by their state of residence. Vocational counseling by a private agency would be approximately $65-85 dollars per hour.

Legal consultation: "A decision should be made about how patients wish to be treated when they develop ventilatory failure, preferably well before it occurs. Advance directives are in order for adults" (Tsao & Armon, 2009, p. 11). It is recommended Victor consult with an attorney to design an advanced directive, general will, and medical living will. Attorney consultation can cost $150 to 250 per hour; however, there are organizations that offer some pro bono services such as the Christian Legal Society.

Driving evaluation: Faith should have a one-time driving evaluation/assessment upon coming of age in her state of residence to determine if driving will be feasible for her followed by training if plausible. "Driver's education alternatives and consideration of customized driving controls should be part of the overall rehabilitation management of the adult with SMA" (Wang, et al., 2007, p. 1044). "As weakness progresses, patients with spinal muscular atrophy may reach a point when it is unsafe for them to operate a motor vehicle or other machinery that requires dexterity. The physician should instruct them to comply with state regulations and to apply common sense" (Tsao & Armon, 2009, p. 10-11).

Genetic counseling: Each sibling can be expected to have genetic counseling/testing once in their adult life if consideration is given to having children of their own. If there are no plans to produce children, testing of the mate is not necessary.

Fertility evaluation: Individuals with SMA are still capable of producing children. In light of Victor's paraplegia, a fertility evaluation is recommended should he desire to father a child. A fertility evaluation will include a comprehensive physical and psychological assessment including "routine blood chemistry tests, hormonal assays, determination of the sensory threshold of the genital area, vascular assessment, neurologic examination of male sexual dysfunction, and nocturnal penile tumescence monitoring with sleep EEG" (Ducharme, http://www.stanleyducharme.com/pdf/sexual_aspects_pd.pdf). Faith would consult with her gynecologist and would be referred to a fertility specialist for any fertility dysfunction; however, costs related to a fertility specialist would be cited in the life care plan, but not covered in the life care plan since SMA does not affect her fertility. High risk prenatal care would be included in the life care plan should she become pregnant.

Hospice evaluation: Upon appropriate deterioration of condition, "hospice referral or other provision for the specific issues regarding terminal care, grief, and bereavement support is important" (Wang, et al., 2007, p. 1045). Hospice care is generally recommended by the treating physician.

Financial Planning: Financial planning along with care planning is recommended to adequately provide for the costs incurred by caring for multiple individuals with a disability. Economists calculate the rate of inflation on services and use the life expectancy provided by the physician or physiatrist to provide final estimates (Weed & Berens, 2009).

Therapeutic modalities:

Physical therapy: Each sibling has attended physical therapy 1-2 times per year for positioning and posture related to the wheelchair (costs allowed once in the LCP).

Nurse case manager: "Successful teams have a point person who is mindful of the many

needs and can obtain appropriate medical, social, and spiritual assistance as appropriate" (Wang, et al., 2007, p. 1045). Faith would benefit from a nurse case manager to assist the family in obtaining resources and confirm the child is receiving optimal care. Sources of case management are the family's health insurance if available, or by private hire/agency at $65-85 per hour for an estimated 4 hours per month. Victor has had a nurse case manager through his own insurance. Should his insurance circumstances change for the worse, he could be expected to require similar case management. Case management can be expected to increase to up to 8 hours per month should either of the siblings' condition significantly deteriorate.

Diagnostic testing/educational needs:

The SMA male has had no cognitive challenges and is in college with no special services. The SMA female is homeschooled, also without cognitive deficits. She functions academically at or above her grade level. She does not receive any special educational services. She will however, require driving evaluation and a vocational evaluation and testing with vocational counseling for job placement should she pursue a career. This was previously cited under Projected Evaluations. Normal schooling in patients with SMA, especially types II and III, is highly recommended because their intelligence is normal or even superior to that of other individuals (Tsao & Armon, 2009, p. 10). Vocational testing and counseling can be provided at no cost through the siblings' state of residence. The cost of homeschooling, private school or college would be a normal cost incurred by an individual or family with children; however, since it is obvious that the individuals would be unable to work a manual job due to weakness, financial information pertaining to college has been included in the life care plan.

Wheelchair needs:

The SMA male uses a power wheelchair for mobility in and out of the home. His current model has been discontinued and he uses parts from an older wheelchair to service the discontinued model. He has a replacement power wheelchair which he does not like the fit and so it has not been used. Based on his consistent involvement in sports, an athletic wheelchair is recommended until his condition deteriorates and/or he no longer desires to pursue sports. A physician's prescription is necessary for a wheelchair; however, for purposes of the sample care plan, a price was researched and listed in Table 1. As noted in a previous category, both siblings are evaluated by a physical therapist for seating and posture. Sales representatives come to the siblings' home to measure and fit the siblings for new chairs when replacement is required. The SMA female uses a power wheelchair, but a different model. There is one manual wheelchair that the siblings share when the power chair is not working or if a location is inaccessible to their power chair. According to the National Institute of Neurological Disorders and Stroke (2008) "Children with SMA II can be taught to operate a power wheelchair at two to three years of age." As previously noted, Faith is scheduled for a spinal fusion to correct her scoliosis. Per Wang, et al. (2007), after scoliosis surgery, Faith may have an increased sitting height, and require van modifications. In addition, "new wheelchairs or wheelchair modifications of the seat, back, arm, leg, or headrests are likely to be required" (p. 1044).

Wheelchair accessories and maintenance:

A local vendor services the children's wheelchairs as needed to evaluate and repair the chairs and replace the batteries. A yearly maintenance visit is recommended for the purposes of assigning a schedule and cost. There is one charger for the wheelchairs. A second charger

and a generator are recommended in cases of simultaneous low batteries or an extended power outage. Maintenance costs begin after the warranty has expired, generally 1 year. Both siblings use the standard wheelchair cushion. Their cushions are vinyl and are not removable. The siblings are reported to prefer the vinyl seat so that they can shift themselves by moving their wheelchairs. The cushions are replaced when the chair is replaced. A spare removable cushion and cover is recommended for each wheelchair in cases of premature wear and tear and could be used for alternate positioning. An alternative option would be to have the vendor replace the cushion attached to the power chair, but most likely at a significant increase in price over a spare individual cushion. The mother has made side pouches to fit the wheelchairs and accommodate their needs. Additional recommended accessories for both siblings include cup holder; replacement backpack for the future, as the mother may not always be able to make replacements; and canopies to protect siblings from prolonged sun or rain and to leave their hands free. As each sibling ages, sheepskin padding for each section of the wheelchair will be needed to protect skin integrity at two sets each. Two sets of padding will allow for a spare in cases of soiling and will extend the life on both sets of padding by reduced wear and tear.

Orthopedic equipment:

None. Future orthopedic equipment would be based on physical or occupational therapy recommendations.

Orthotics and prosthetics:

The SMA male did not require any orthopedic equipment at the time of this paper. His sister requires a thoracolumbosacral orthosis (TLSO) body jacket for her scoliosis. "Scoliosis occurs at some point in the majority of children with SMA II. Custom seating systems, seating aids, and a body jacket can be used to prevent severe scoliosis. Spinal fusion surgery may be necessary for some children" (NINDS, 2008). Faith is scheduled to undergo a posterior spinal fusion for scoliosis exceeding 80 degrees. If the surgery is successful, she will not require a replacement TLSO brace. Without a spinal fusion, the care plan would reflect a replacement jacket every 4 years (Evans, et al., 1981). Per Evans, et al. (1981), an alternative prescription to spinal orthotics in those unfit for surgery with deformity and loss of balance when sitting is a custom-built supporting seat.

Architectural needs:

The SMA siblings' home has many accessibility challenges and an architectural consultation for home accessibility was recommended in the *Projected Evaluations*. The home is a Cape Cod design built in 1947. It has 13 rooms between the 3 levels with hardwood, vinyl and tile floors. The only access to the home for the siblings is on a wooden ramp at the back of the house. The ramp is 20 years-old and is reported to have begun falling apart. Repairs to the existing ramp and a second ramp is recommended for safety and accessibility. There is no handicapped access to the upstairs or basement which makes two-thirds of the home inaccessible to the siblings and limits their routes for emergency evacuation. A second escape route should be considered for addition to the main level of the home. The doorways are reported to be tight throughout the house with pieces of the frames knocked off and chipped away by the wheelchair traffic. Some of the door frames are entirely removed from being knocked off on multiple occasions. Handicap-accessible doorways are recommended for mobility, safety and preservation of the house. The bathroom is reportedly too small to adequately maneuver the power wheelchairs and the toilet often gets knocked over. The

shower floods the bathroom every time the siblings are bathed. The vanity was removed from the sink to improve accessibility, but the sink no longer has the counter around it so the siblings often drop whatever hygiene items they are working with. Recommendations for architectural consideration in the bathroom would be a wheelchair accessible shower and sink and enlarging the bathroom in general.

In the kitchen, the siblings are unable to reach dishes, drinks, food, the microwave, sink, and toaster. Because they have no independence in the kitchen, they are totally dependent on others to reach and prepare most of their snacks and all of their meals. The siblings have a picker-upper for extended reaching; however, because strength is an issue as many items should not be lifted or reached with a reaching aid due to the height and safety concerns. Part of the architectural evaluation should include the design of a handicap-accessible kitchen.

The siblings' bedrooms are very small which makes maneuvering their power wheelchairs and transfers very difficult. The siblings are unable to reach their clothes, open their bureau drawers, and unable to reach some of their sports equipment and/or toys. Enlarging the room and placing low cabinets with doors versus drawers would improve accessibility to personal items as well as allow the siblings to perform some independent homemaking chores such as putting clothes away and dusting and cleaning lower surfaces provided the surface is not too low.

The family has working fire alarms and an escape plan; however, the escape plan will need to be revised since one of the older siblings no longer lives in the home. There is no medical alert system in the home and addition of a medical alert system would improve the likelihood of timely intervention in cases of choking, falls or respiratory distress.

Home furnishings and accessories:

There is no handicapped access to the upstairs or basement which makes two-thirds of the home inaccessible to the siblings. A platform or inclined wheelchair lift(s) and a stair lift would improve accessibility to the rest of the home. A stair lift however would require the siblings to be transferred from the wheelchair to the stair lift and back into another wheelchair when transporting to another level of the home.

Accessories for the bathroom include shower and sink caddies and a hand-held showerhead. Additional recommendations include a table-type sink that would allow the siblings to roll their wheelchair under the sink, while improving their reach and providing a surface on which to set hygiene items.

The kitchen has poor accessibility. In addition to the renovations listed above, liberal use of multi-leveled Lazy Susans would give the siblings maximal access to food, medications, and utensils while preserving space. A refrigerator with an ice and water dispenser on the door would give the siblings ready access to fluids.

Bedroom accessories should include low cabinets, if not inserted during any architectural renovation, with doors versus drawers. Multi-leveled Lazy Susans could also be used in the bedroom.

A Hoyer lift is used for some transfers.

Aids for independent function:

Both the siblings have pouches attached to their wheelchair for personal items. The male has a TV tray table in his bedroom to hold various items. School supplies, toys and mirrors have been lowered to accommodate the siblings. The siblings both have a picker-upper for extended reaching. Additional items for independent living are electric can openers; and drink

dispensers with a flip spout.

Medications:

The male takes a bladder relaxant at bedtime, daily. This medication does not require any special diagnostic testing. Side effects of this anticholinergic include delayed gastrointestinal function and dry mouth. He also has been prescribed short-acting insulin; however, his healthcare providers are still in the process of confirming the diagnosis of diabetes. His sister is reported to have no chronic medication related to the SMA.

Supply needs:

The male has a glucometer, including testing strips and insulin syringes. He uses new sterile 14 French catheters 4 times daily with surgi-lube.

A plastic lawn chair serves as a shower chair due to the size of the bathroom. A shower chair, transfer board and bench are recommended for safety.

Home care/attendant care/facility care:

Both siblings are cared for by their parents. A home health aide was used at one point to assist with bathing and routine care, but the parents opted to care for the children independently. There are several options for present and future care of the siblings; four (4) options are listed below:

Option 1: Continue to care for the siblings independently.

Option 2: Care for the siblings with the assistance of a home health aid. This could be arranged on any number of schedules to best accommodate the family. As the parents of the siblings age, this may become a more feasible option. Victor's insurance was reported to cover a portion of home health assistance; however, the mother was unsure if the family's insurance would cover a home health aide for Faith. Another alternative in the nature of the home health aide option, would be to let Faith attend public school and utilize the resources provided under the federal Individuals with Disabilities Education Act (IDEA).

Option 3: Complete skilled nursing in the case of major pulmonary or swallowing deficits either through an agency or private pay. In the event one or both siblings' condition deteriorates to include tube feedings and/or ventilatory support, an appropriately skilled nurse would be required. Since the siblings' mother has the appropriate healthcare background, and if her own health permitted, she may opt to manage either or both siblings in a deteriorated condition with routine assessments and assistance as needed from a skilled nurse.

Per (Tsao and Armon, 2009),

> "A decision should be made about how patients wish to be treated when they develop ventilatory failure, preferably well before it occurs. Advance directives are in order for adults. Early discussions regarding end-of-life issues with parents of affected children and adolescents are highly recommended" (p. 11).

Agency versus private duty nursing is listed in the sample care plan for the SMA male (Table 1). The family must recognize and consider that to use a nurse under private hire makes the family or individual an employer who is responsible for withholding state and local taxes as well as any other regulations put forth by the state of residence.

Option 4: Residential assisted living. In the event the parents are no longer able to care for the siblings or the siblings survive the parents, residential assisted living is available. The

siblings are cognitively bright, and although limited by the use of their lower extremities, are functional for everyday life and have the potential to pursue rewarding non-manual careers.

Since the siblings live in the home with their parents, housekeeping, home maintenance, and yard work are provided. As the parents age, these services may need to be arranged.

Aggressive future medical care or surgical intervention:

Any aggressive future medical care or surgical intervention for the SMA male would be dictated by a physician. The SMA female is scheduled for a posterior spinal fusion to correct her scoliosis. Per Evans et al. (1981), "There is a risk of functional loss after spinal fusion (as cited by Schwentker & Gibson, 1976) such as the loss of the ability to walk or transfer independently. This complication has been prevented by intensive physiotherapy and an early return to the activities performed before the operation" as cited by Hensinger and MacEwan (1976).

Preoperatively, Faith can anticipate routine blood work, pulmonary function tests, and a chest x-ray, along with extensive patient/family education regarding the preparation for surgery, surgery itself and postoperative teaching regarding rehabilitation.

With regard to Faith, she has some ability to walk when in water and has some movement in her legs. Her legs have contractures. The physician will determine whether she would benefit from formal physiotherapy or routine range of motion exercises post-operatively to preserve function. Most of the cost of the surgery will be covered by insurance except for the family deductible and any doctors who provided care in the hospital who are not in the family's insurance network. There is a known up-front cost to the parents of $500.

Faith and her parents reported they planned to inquire about whether surgical repair of the dislocated hip was feasible. According to Zenios, et al., (2005), surgical repair of subluxed hips was not associated with a better functional outcome and that the subluxation often recurred.

Preoperative teaching and preparation is imperative for the SMA patient who is at risk for pulmonary complications. "Perioperative care includes a thorough preoperative evaluation of respiratory status and anticipatory guidance of the surgical team and postoperative management team regarding optimal care" (Wang, et al., 2007, p. 1038). Perioperative management is customized to the individual, family and treatment team and includes modification of the patient's environment and orthotic plan (Wang, et al., 2007). Wheelchair modifications may need to be required (Wang, et al. 2007). Postoperative care will focus on range of motion, incentive spirometry, and mobility in addition to the standard medical surgical care (Wang, et al., 2007).

Transportation needs:

The family had a power lift installed in the family van to allow the siblings access. They have tie downs to secure the wheelchairs in the van and seat pouches to hold items needed during travel. The SMA male plans to purchase a van after college. A state agency will assist in modifying the van to allow him to drive independently. As cited earlier, he has already obtained his driver's license. A state agency will pay for the entire cost of the van modification once he purchases a new van. The estimated cost of the van modifications provided to the parents was $90,000. There would be an offset to the cost of the modified van as the individual would have likely owned his own vehicle had he not had the disability (Dillman, 2009, as cited in Weed & Berens, 2009).

Maintaining safety should be a priority for SMA individuals choosing to drive. "As

weakness progresses, individuals with SMA may reach a point when it is unsafe for them to operate a motor vehicle or other machinery that requires dexterity. They should comply with state regulations and apply common sense" (Tsao & Armon, 2009, p. 10-11).

After scoliosis surgery, Faith may have an increased sitting height and require van modifications (Wang, et al., 2007).

Leisure time/recreational needs:

"The goal of active but nonfatiguing exercises is to maintain range of motion, increase muscle flexibility, and prevent contractures. These exercises should not produce pain or exhaustion" (Tsao & Armon, 2009, p. 9). The SMA male's hobbies include wheelchair hockey and he participates in various wheelchair sports twice a year for eight to ten week sessions. He loves sports and owns seasonal tickets to the local state football team's games. He enjoys math, computers and is skilled in editing video. His life care plan should include a membership to an athletic club for challenged athletes. The current sports facility that he uses costs $80-120 per eight to ten-week session depending on the sports class.

The SMA female enjoys swimming, singing, animals, babies, dolls and crafts. She frequently volunteers for local organizations supporting SMA and regularly attends church. The family home has a computer and DVD player accessible to the siblings. The family also owns a 3-foot blow up pool in which the SMA female walks and swims. Recommendations would include maintenance and repair of the pool and a pool membership to an indoor facility for the winter months.

Vocational Interests and Pursuits:

Victor is to receive state-sponsored vocational counseling after he graduates college. He is due to graduate with degrees in math and computer science. He is employed part-time through a work-study program editing college sports videos and is training to assist with data entry for a local business. Upon graduation, Victor would like obtain employment with the government using his math degree. See the life care plan sample in Table 1 for associated costs. College costs have been $7,200 per year times 4 years. Computer maintenance and supplies would normally be owned by an individual in this culture; however, since it is obvious that Victor will not be able to work a manual job and has pursued a computer-related degree, the computer and maintenance will be included in the life care plan as a necessity. According to Wang, et al., (2007), "Assistive technology and other adaptive equipment to enhance work and play should be considered" (p. 1043).

Faith was unsure what career, if any, she wanted to pursue after high school. She stated that she would like to be married some day and be a mother.

Discussion

Spinal muscular atrophy type II leaves cognitively normal individuals in progressively deteriorating bodies. Over 115 years of care of persons with SMA has revealed that their functional level directly impacts their life expectancy and quality of life (Iannaccone, 2002). As reported in this paper, researchers and physicians alike agree that prudent planning incorporating a team approach for medical care and activities of daily living is vital to survival and quality of life (Tsao & Armon, 2009; Wang, et al., 2007). The life care plan takes into consideration the unique aspects of the individuals' condition, family life, geography, and available treating providers. While both siblings share the same diagnosis of SMA type II, they do not share the same pathological progression, treatments, and do not have the same goals for

their future. Even within a family, the life care plan must be tailored to the individual for optimal outcomes.

Further differences in recommendations for the siblings were gender-specific to care for and evaluate the reproductive systems. The siblings' family dynamics were affected by their age, changes in disease progression, complications and lack of complications, finances, insurance, the aging of parents and the aging of the unaffected siblings who reached adulthood and left the home.

The life care plan facilitates communication between family members, healthcare providers, legal and insurance agents, and financial planners regarding care recommended by the treatment team and supported by valid medical research on the part of the life care planner. A well-written life care plan, executed by all parties, provides for the optimal well-being of the SMA individual and can promote harmony and understanding to families struggling with the many aspects of loving and caring for their family member(s) with a disability.

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References

Abresch, R.T., Carter, G.T., Jensen, M.P., & Kilmer, D.D. (2002). Assessment of pain and health-related quality of life in slowly progressive neuromuscular disease. *Journal of Hospice and Palliative Care, 19*(1), 39-48.

Allfrey, V.G., Faulkner, R. & Mirsky, A.E. (1964). Acetylation and Methylation of histones and their possible role in the regulation of RNA synthesis. Proceedings of the National Academy of Sciences USA, 51, 786-94.

American Society of Human Genetics (ASHG) Board of Directors and The American College of Medical Genetics (ACMG) Board of Directors. (1995). ASHG/ACMG Report. Points to consider: Ethical, legal, and psychosocial implications of genetic testing in children and adolescents. *American Journal of Human Genetics, 57,* 1233-1241.

Bonfiglio, R.P. (2009). The role of the physiatrist in life care planning. In R.O. Weed & D.E. Berens (Eds.), *Life Care Planning and Case Management Handbook (3rd ed.)*. Boca Raton, FL: CRC Press, 17-25.

Cheung, L., Fitch, K., & Pyenson, B. (2001). The costs of hospice care: An actuarial evaluation of the Medicare hospice benefit. 2001. New York: Milliman USA, Inc. Retrieved July 12, 2009 from: http://www.nhpco.org/Files/members/TheCostsofHospiceCare-Millman.pdf

De Groot, I.J.M., & de Witte, L.P. (2005). Physical complaints in ageing persons with spinal muscular atrophy. *Journal of Rehabilitation Medicine, 37*, 258-262.

Dillman, E. (2009). The role of the economist in life care planning. In R.O. Weed & D.E. Berens (Eds.), *Life Care Planning and Case Management Handbook, (3rd ed.)*, Boca Raton, FL: CRC Press, 303-317.

Dubowitz, V. (1995). Disorders of the lower motor neuron: The spinal muscular atrophies. In Dubowitz V. (Ed.), *Muscle Disorders in Childhood. London:* WB Saunders, 325-367.

Dubowitz V. (1974). Benign infantile spinal muscular atrophy. *Developmental Medicine & Child Neurology, 16,* 672-75.

Ducharme, S.H. Sexual Aspects of Physical Disability, Chapter 55, p. 966-980. Retrieved July 10, 2009 from: http://www.stanleyducharme.com/pdf/sexual_aspects_pd.pdf

Emery, A.E.H. (1991). Population frequencies of inherited neuromuscular diseases - a world survey. N*euromuscular Disorders, 1,* 19-29.

Evans, G.A., Drennan, J.C., & Russman, B.S. (1981). Functional classification and orthopaedic management of spinal muscular atrophy. *The Journal of Bone and Joint Surgery; 63-B*(4), 516-522.

Hirtz, D., Iannaccone, S., Heemskerk, J., Gwinn-Hardy, K., Moxely III, R.,& Rowland, L.P. (2005). Challenges and opportunities in clinical trials for spinal muscular atrophy. *Neurology, 65*(1), 1352-1357.

Iannaccone, S.T., & the American Spinal Muscular Atrophy Randomized Trials (AmSMART) Group. (2002). Outcome measures for pediatric spinal muscular atrophy.

Iannaccone, S.T., Khatri, I.A. (2005). Childhood spinal muscular atrophy. In D.S. Younger (Ed.), *Motor Disorders*. Philadelphia: Lippincott Williams & Wilkins.

Iannaccone, S.T., Hynan, L.S., & the American Spinal Muscular Atrophy Randomized Trials (AmSMART) Group. (2003). Reliability of 4 outcome measures in pediatric spinal muscular atrophy. *Archives of Neurology; 60,* 1130-1136.

Iannaccone, S.T, Russman, B.S, Browne, R.H, Buncher, C.R, White, M, & Samaha, F.J., (2000). DCN/SMA Group. Prospective analysis of strength in spinal muscular atrophy. *Journal of Child Neurology, 15,* 97-101.

Mellies, U., Ragette, R., Schwake, C.D., Boehm, H., Voit, T., & Teschler, H. (2003). Long-term noninvasive ventilation in children and adolescents with neuromuscular disorders. *European Respiratory Journal, 22,* 631-636.

Merlini, L., Stagni, S.B., Marri, E., & Granata, C. (1992). Epidemiology of neuromuscular disorders in the under-20 population in Bologna Providence, Italy. *Neuromuscular Disorders, 2,* 197-200.

Munsat, T.L., & Davies, K.E. (1992). International SMA consortium meeting (26-28 June 1992, Bonn, Germany). *Neuromuscular Disorders, 2,* 423-428.

Muscular Dystrophy Association. (2000). MDA News: New research opens door to stem cell therapy for ALS and SMA. Retrieved May 20, 2009 from: http://www.mda.org/news/001108newresearchalssma.html

National Institute of Neurological Disorders and Stroke. (2008). Spinal muscular atrophy fact sheet. Retrieved May 18, 2009 from: http://www.ninds.nih.gov/disorders/sma/detail_sma.htm?css

National Spinal Cord Injury Database (2006). Model SCI Care Systems' Data Collection Forms 2006-11. Retrieved May 18, 2009 from: http://www.spinalcord.uab.edu/show.asp?durki=24480

Nordgren, R.E. (2005). Myopathies and Neuropathies. In L.M. Osborn, T.G. DeWitt, L.R. First & J.A. Zenel (Eds.). *Pediatrics*. Philadelphia. Elesvier Mosby, 1191-1193.

Pearn, J. (1978). Incidence, prevalence and gene frequency studies of chronic childhood spinal muscular atrophy. Journal of Medical Genetics, 15, 409-413.

Rubin, M. (2008). Spinal Muscular Atrophies. In Spinal Muscular Atrophies: Peripheral Nervous System and Motor Unit Disorders, Merck Manual Home Edition. Retrieved March 1, 2009 from: http://www.merck.com/mmpe/print/sec16/ch223j.html

Rubin, M. (2008). Spinal Muscular Atrophies. In Spinal Muscular Atrophies: Peripheral Nerve Disorders, Merck Manual Home Edition. Retrieved March 1, 2009 from: http://www.merck.com/mmhe/print/sec06/ch095/ch095j.html

Rudnik-Schöneborn, S., Hausmanowa-Petrosewicz, I., Borkowska, J., Zerres, K. (2001). Predictive value of achieved motor milestones assessed in 441 patients with infantile spinal muscular atrophy type II and III. *European Neurology, 45,* 174–181.

Russman, B. (2002). Spinal Muscular Atrophy. In L. Finberg, L. & R.E. Kleinman (Eds.). *Saunders Manual of Pediatric Practice (2nd ed.)*, Philadelphia. W.B. Saunders Company, 1004-1006.

SMA Foundation. (2009). Frequently Asked Questions. Retrieved May 20, 2009 from: http://www.smafoundation.org/index.php?option=com faq&Itemid=32

Spinal Cord Injury Information Network. (2006). National Spinal Cord Injury Database: Data Collection Forms. Model SCI Care Systems' Data Collection Forms 2006-11. Retrieved March 1, 2009 from: http://www.spinalcord.uab.edu/show.asp?durki=24480

Tsao, B., & Armon, C. (2009). Spinal Muscular Atrophy. Retrieved May 20, 2009 from: http://emedicine.medscape.com/article/1181436

University of Florida Health Center Institutional Review Board. (2007). Opinion paper: Case reporting. (IRB-01). Gainesville, FL.

Wang, C.H., Kleyn, P.W., Vitale, E., Ross, B.M., Lien, L., Xu, J., et al. (1995). Refinement of the spinal muscular atrophy locus by genetic and physical mapping. *American Journal of Human Genetics, 56*, 202-209.

Wang, C.H., Funkel, R.S., Bertini, E.S., Schroth, M., Simonds, A., Wong, B., et al. (2007). Consensus Statement for Standard of Care in Spinal Muscular Atrophy. *Journal of Child Neurology, 22(8)*, 1027-1049. Retrieved May 18, 2009 from: http://jcn.sagepub.com

Weed, R.O. & Berens, D.E. (Eds.) (2009). *Life Care Planning and Case Management Handbook (3rd ed.)*. Boca Raton, FL: CRC Press, Inc.

Zenios, M., Sampath, J., Cole, C., Khan, T., & Galasko, C.S.B. (2005). Operative treatment for hip subluxation in spinal muscular atrophy. *The Journal of Bone & Joint Surgery (Br), 87-B*(11), 1541-1544.

Zerres, K. & Rudnik-Schöneborn, S. (1995). Natural history in proximal spinal muscular atrophy. *Archives of Neurology, 52,* 518–523.

Zerres, K., Rudnick-Schöneborn, S., Forrest, E., Lusakowska, A., Borkowska, J., & Hausmanowa-Petrusewicz, I. (1997). A Collaborative Study on the Natural History of Children and Juvenile Onset Proximal Spinal Muscular Atrophy (type II and III SMA): 569 patients. *Journal of the Neurological Sciences, 146,* 67-72.

Zupkovitz, G., Tischler, J., Posch, M., Sadzak, I., Ramsauer, K., Egger, G., et al. (2006). Negative and positive regulation of gene expression by mouse histone deacetylase 1. *Molecular and Cellular Biology, 26(21)*, 7913–28.

Table 1: The SMA male

Note: **This is a sample life care plan**. A formal life care plan would include a thorough medical record review and healthcare providers would be consulted for recommendations for the plan of care. An economist would calculate the rate of inflation on services and use the life expectancy provided by the physician or physiatrist to provide final estimates (Weed & Berens, 2009). Costs reflect the cost cited by the parent and current local or national market. The costs remain estimates as the patient's providers were not consulted. Frequencies were per the parents, literature, vendors or estimates provided by the author for demonstration.

Routine Medical Care			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Dentist	2 times per year to life expectancy	Hygiene; Evaluate for oral complications	Exam $50-$135 Deep cleaning $100-$400 per quadrant X-rays $15-$130
Pulmonologist	Every 1-2 years until signs of decompensation begin (recurrent lung infections, etc. (Wang, et al., 2007)) then 3-4 times per year/as needed.	Pulmonary function testing and sleep studies. Teach parents chest physiotherapy to clear airway secretions	Pulmonology evaluation $150-$300 Sleep Laboratory (overnight evaluation) $1500 PFTs $70-$325 depending on tests Costs covered by insurance
Neurologist	1 time per year to life expectancy	Monitor neurological status	$300-$500 per exam Costs covered by insurance
Orthopedist	Every 1-2 years to life expectancy	Spine x-rays post fusion; Monitor for orthopedic changes that may impact lung function	$150-$250 per exam Costs covered by insurance
Endocrinologist	Every 3 months. This would be projected to change based on a formal endocrinology	Rule out diabetes/ metabolic disorder	$125 per exam Costs covered by insurance

	diagnosis		
Podiatrist	Every 9 weeks to life expectancy unless diabetes is ruled out.	Foot care related to his possible diabetic condition	$290 per year Costs covered by insurance
Primary Care Physician	Annually to life expectancy	Routine health evaluations	$80 per visit Costs covered by insurance
Routine lab tests*:* serum enzymes, chemistry panels, urinalysis and blood counts, prealbumin levels, HgbA1c	4 times per year until a metabolic condition is ruled out and then 1-4 times per year until life expectancy.	Monitor general condition; screen for infection; early detection of urinary tract infections; assess nutritional status	$93 for routine blood panels; $50 for additional lab testing Costs covered by insurance
Projected Evaluations			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Physical therapy	1-2 times per year for life. The evaluation has been conducted annually at a SMA clinic.	Evaluate and preserve function; contracture management. According to Wang, et al., (2007, p. 1043) "Contracture management and exercise are a major focus of treatment, with implementation of a regular stretching and bracing program to preserve flexibility."	See *Therapeutic modalities*
Psychological evaluation	1 time or as needed to life expectancy. Ongoing counseling may or may not be required.	Assist with coping with multi-dimensional loss and altered relational dynamics.	$80-$140 per session
Family counseling	Every 1-3 years if needed while individual lives	Assist with coping and improve the	$75-$200 per hour
	with his parents/siblings or significant other	quality of life	

Recreational therapy	2009, then every 5 years or to coincide with each stage of functional loss throughout life if desired.	"Regular exercise should be encouraged to maintain fitness and endurance and might include swimming and adaptive sports" (Wang, et al., 2007, p. 1043). It is possible that the appropriate PT facility could also cover some of the recreational therapy recommendations	$175-$375 per evaluation
Nutritional consultation	1 time per year until condition deteriorates, then 4 times per year to life expectancy. May be monitored by PCP.	Ensure optimal caloric intake without incurring obesity or a catabolic state (Tsao & Armon, 2009; Wang, et al., (2007)).	$65-$75 per evaluation
Physiatry consultation	Yearly to life expectancy	The physiatrist specializes in rehabilitative medicine and physical function.	$125-250 per visit
Home accessibility evaluation (may be conducted by OT)	1 time	Architectural renovation recommendations for handicapped-access	$125-$150 per evaluation
Vocational evaluation	1 time (completed)	Vocational assessment services	This service was provided free of charge through his state of residence. Vocational evaluation by a private agency would be $72-$85 per hour.

Vocational counseling	Upon completion college	Assist with job placement/options	This service is provided at no charge by the state of residence. Vocational counseling by a private agency would be $72-$85 per hour.
Legal consultation	1 time	Create an advanced directive, general will, and medical living will	$500-$750; however there are organizations that offer some pro bono services and there are 'do-it-yourself" online legal alternatives
Driving evaluation	(Completed)	"Driver's education alternatives and consideration of customized driving controls should be part of the overall rehabilitation management of the adult with SMA" (Wang, 2007, p. 1044).	State-funded
Genetic counseling:	1 time for mate unless there are no plans to produce children	Predict chances of having a child with SMA	$100-$2,000 depending on tests Some insurances will cover
Fertility evaluation: includes comprehensive physical and psychological	1 time	Fertility evaluation as related to paraplegia	$400 for initial consultation, then $1200 for initial exam/testing. Other visit costs
			unknown and would depend on options pursued.

Hospice evaluation/care	36-48 days (Cheung, Fitch, & Pyenson, 2001)	Provision for terminal care (listed as a resource for individual/family information only)	$117.10 per day (routine home hospice)
Financial Planning	Every 1-3 years	Provide for costs incurred by disability	$100 per hour
Therapeutic Modalities			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Physical therapy	1-2 times per year	Positioning and posture related to the wheelchair	$150-$200 per visit. See also *Projected evaluations*
Nurse case manager	4 hours per month to increase to 8 hours per month should he significantly deteriorate	"Successful teams have a point person who is mindful of the many needs and can obtain appropriate medical, social, and spiritual assistance as appropriate" (Wang, 2007, p. 1045).	Covered by Victor's health insurance. Private hire/agency at $65-85 per hour
Diagnostic Testing/Educational Needs			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
See *Projected evaluations* regarding vocational assessment			
College education: tuition, lab fees, books, and supplies	1 time per year for 4 years (due to graduate 2009)	Obtain the most appropriate non-manual job to provide for self	$7,200 per year
Wheelchair Needs			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Power wheelchair	Every 5 years beginning in 2015 His current model has been	Mobility in and out of the home	$2,890 Costs covered by insurance

	discontinued and he uses parts from an older wheelchair to service the discontinued model. He has a replacement power wheelchair that has not been used.		
Athletic wheelchair	1-2 times beginning in 2009 until/unless his condition deteriorates and/or he no longer desires to pursue sports	Recreation and exercise	$1535
Manual wheelchair	Beginning 2009 then every 10 years	Back-up to power chair; For use where power chair cannot be accommodated Family has shared 1 manual wheelchair between 2 disabled siblings	$145
Wheelchair Accessories and Maintenance			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Routine maintenance (each wheelchair purchased)	1 time per year to life expectancy	Evaluate and repair the chair	Maintenance costs begin after the 1-year warranty at 10% cost of chair.
Batteries	2011 then every 2-3 years to life expectancy	Power the wheelchair	$280 each
Battery charger	Every 5 years beginning 2011 until life expectancy	The disabled siblings share a battery charger. Each individual should have their own in case of	$130
		emergency.	

Back-up generator	1 time	In cases of power outage.	$380
Removable wheelchair cushion	Every 5 years to life expectancy. Victor uses the standard wheelchair cushion that is fixed to the power chair.	Recommended in cases of premature wear and tear on power chair seat; prevention of pressure sores	$320
Wheelchair cushion cover (2)	Every 2 years (or when new removable cushion is purchased) to life expectancy	Spare cover in case of soiling; reduces wear and tear	$55
Cup holder	1 per year to life expectancy or deteriorated condition prohibiting oral fluids	Promote hydration and independence	$22
Side pouches	Every 1-2 years to life expectancy	Promote independence; store items needed	$17
Backpack	Every 1-2 years to life expectancy	Promote independence; store items needed	$26-$45
Canopy	1 time	Protect from prolonged sun or rain while leaving the hands free	$150
Sheepskin padding for each section of the wheelchair. Two sets of padding	1-2 times until life expectancy	Protect skin integrity at two sets each; spare for cases of soiling and extend life on both sets of padding	$59 each set
Orthopedic Equipment			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
None.			

Orthotics and Prosthetics			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
None.			
Architectural Needs			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Repairs to the existing wheelchair ramp Or Replace ramp	To begin 2009 then every 1 years to life expectancy To begin 2009 then every 15 years to life expectancy	Allow access to home	Per architect's estimate
Second wheelchair ramp	To begin 2009 then every 15 years to life expectancy	Safety as an alternate escape route; accessibility	Per architect's estimate
Handicap-accessible doorways	1 time	Mobility, safety and preservation of the house	Per architect's estimate
Handicap-accessible bathroom	1 time	Mobility; safety; hygiene	Per architect's estimate
Wheelchair-accessible shower	1 time	Mobility; safety; hygiene	Per architect's estimate
Wheelchair-accessible sink/vanity	1 time	Access; hygiene; independence	Per custom production estimate
Handicap-accessible kitchen	1 time	Nutrition; independence; safety	Per architect's estimate
Handicap-friendly bedroom	1 time	Independence; safety; access	Per architect's estimate
Medical alert system Monitoring fee	1 time Monthly beginning 2009 to life expectancy	Safety; prompt medical care	Some systems require a portion of the system to be purchased at $200, then charge a monthly monitoring fee. $25 to $45 per month monitoring fee.

Extended warranty	1-year warranty included. Begin 2010, ends 1-3 years		Warranty $49-$69
Pendant	1 time beginning 2009		Pendant $30-$60
Spare pack pendants (5)	1 time beginning 2009		Spares $99-$139
Replacement batteries (5-pack)	Beginning 2009 then 2 times per year to life expectancy		Batteries $15
Home Furnishings and Accessories			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Inclined wheelchair lift (2).	One-time purchase to begin 2009	Accessibility on each level of home; safety in case of flooding or tornados	$2,500-$4,000 each
Warranty	20-year warranty		None.
Maintenance			Maintenance fees to depend on warranty
Accessories for the bathroom include: Shower caddy	Patient owns. Replace every 5 years beginning 2010 until life expectancy	Access to hygiene items; independence	$15-21
Sink caddy	Patient owns. Replace every 5 years beginning 2010 until life expectancy		$6-$10
Hand-held showerhead	Patient owns. Replace every 5 years beginning 2010 until life expectancy	Safety	$30
Shower bars	1 time beginning 2009		$30-45 each

Table-type sink	1 time beginning 2009	Allow legs to be wheeled under sink, access to hygiene	See *Architectural needs*
The kitchen: Multi-leveled Lazy Susans for cabinets (5)	Beginning 2009 then replace every 5 years until life expectancy	Access to food, medications, and utensils while preserving space	$49 to $169
Multi-door refrigerator with ice and water dispenser and drawer(s)	Beginning 2009 to life expectancy	Access to fluids for hydration; Easier to open	$2800 to$3300
Extended warranty	Beginning 2010-2012 (depends on warranty)		(Depends on warranty)
Maintenance	To begin when warranty expires to life expectancy		$75 per evaluation
Bedroom: Low-set cabinets, with doors versus drawers.	Beginning 2009 then replace every 5 years until life expectancy	Access to personal items; independence	See *Architectural needs*
Multi-leveled Lazy Susans	Beginning 2009 then replace every 5 years until life expectancy		$49 to $169
Hoyer lift	Patient owns. 1 time expense		NA
Maintenance			Depends on warranty
Aids for Independent Function			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Picker-upper (2)	(Has). Replace every 3-5 years	Extended reaching; spare	$20 each
Electric can opener	One-time	For independent living; nutrition	$25
Drink dispenser with a flip spout (2)		For independent living; hydration; Recommend	$7 each
		plastic for safety	

Cellular phone	(Has). Replace every 5 years depending on wear and tear	Safety; communication; independence	No additional cost above general population
Medications			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Anticholinergic/antispasmodic	Monthly until life expectancy or changed by PCP	Bladder relaxant	$62.10 (30-day supply)
Short-acting insulin	As needed; 2009 to unknown	Blood sugar control/diabetes	$47.23
Supply Needs			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Glucometer	(Has) replace every 5 years if diabetes confirmed until life expectancy	Blood sugar control	$150-$200
Testing strips; control solution;	If diabetes confirmed 1-4 strips daily		$35 (200 count)
Lancets	If diabetes confirmed 1-4 daily		$59.39 (100 count)
Insulin syringes	If diabetes confirmed 1-4 daily		$21 (90 count)
14 French catheters	4 times daily to life expectancy	Self-catheterization	$52.80 (30 count)
Surgi-lube	4 times daily to life expectancy	Lubricant	$0.14 (144 packets)
Vinyl gloves	4 pairs daily to life expectancy	Prevent UTI; reduce latex allergen exposure	$6 (box of 50 pairs)
Aluminum shower chair commode with casters	One-time	Hygiene; convenience portable commode; smaller size to	$194
		accommodate bathroom/house	

MRI safe transfer board	One-time	Transferring	$86-$146
Transfer Bench	One-time	Transferring	$164
Home Care Attendant Needs			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Option 1: Continue to care for Victor independently	Daily until life expectancy	Care for Victor independently	None
Option 2: Care for Victor with the assistance of a home health aid	Daily until life expectancy or upon individual/parental decision. Number of hours will depend on level/amount of care provided.	Care for Victor with assistance to prevent care giver burn-out and injury	Victor's insurance covers a portion of home health assistance. In-home care cost approximately $19 per hour.
Option 3: Complete skilled nursing (RN) either through an agency or private pay. This would be per physician's recommendations.	In the case of major pulmonary or swallowing deficits until life expectancy. Number of hours would depend on level of care needed.	Provide skilled nursing care and assessment for prevention of complications and routine care. (Since Victor's mother has the appropriate healthcare background, and if her own health permitted, she may opt to manage his care in a deteriorated condition with routine assessments and assistance as needed from a skilled nurse).	$65 per hour via agency If through private hire, individual/family is considered an employer and must withhold taxes and abide by stated regulations.
Option 4: Residential assisted living	Upon decision of the individual/family until life expectancy	In the event the parents are no longer able to care for Victor, or he survives his parents, or	$500-$4,000 per month
		decides to live outside of the home	

Housekeeping	2 hours every 1-2 weeks 2009 to life expectancy	Provide clean sanitary environment; Prevent care giver burn-out	Provided by parents. $25-$30 per hour for independent housekeepers. $70-$96 per hour through agency. Some agencies send a team of 2 housekeepers.
Interior/exterior home maintenance	2 hours every week 2009 to life expectancy		Provided by parents. $15-$75 per hour depending on nature of services
Aggressive Future Medical Care or Surgical Intervention			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
None to date. See *Complications* in body of paper.			Unknown
Transportation Needs			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Van	Beginning 2009, replace every 10 years until life expectancy	Independent transportation	$20,000
Maintenance	Once per year excluding oil changes		$75-$125 per year
Modifications	One-time per vehicle	Handicap-accessible	$90,000: covered through state-sponsored program when individual purchases a new van. Cost to be offset by price of average vehicle.

Leisure Time/Recreational Needs			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Membership to an athletic club for challenged athletes	2 sessions per year 2009 until life expectancy or condition significantly deteriorates	Maintain overall physical and mental well-being; maintenance of musculoskeletal function; community socialization; dignity	$80-$120 per eight to ten-week session
Vocational Interests and Pursuits			
Recommendation	***Frequency***	***Purpose***	***Anticipated Cost***
Computer system Maintenance Technology upgrades See *Diagnostic Testing/Educational Needs*	(Has) Replace every 5 years beginning 2010 to life expectancy or significant deterioration in condition Every 2 years	A computer would normally be owned by an individual in his culture; however since it is obvious that Victor will not be able to work a manual job and is obtaining a degree in the computer industry, the computer is necessary due to his disability and will be included in the life care plan. According to Wang, et al., (2007, p. 1043), "Assistive technology and other adaptive equipment to enhance work and play should be considered.	$2,500-$3000 $129 plus tax per visit $250-$500