Tiffany is a 19 month old child diagnosed at the age of 10 months with SMA Type 1. She receives PT and OT in her home once a week for 60 minutes with services provided by the Child Developmental Services Agency (CDSA), Early Intervention. December of 2012 she was admitted to the hospital for pneumonia and respiratory distress. Prior to admission, she was receiving outpatient PT from another provider to improve gross motor skills, head control, and strength in extremities. Currently she is followed medically by her local pediatrician, Duke Health Care System and ECU Spine Specialist Dr. Reeg. Her specialists include: pulmonologist, neurologist and orthopedic spine specialist. PT referred patient to pediatric orthopedic Dr. Fitch to evaluate spine and lower extremities. Dr. Fitche ordered spine X-Rays and mom reports he saw a 40% scoliosis in sitting. He recommended a sure step TLSO. Previous Hip X-Rays requested by PT and ordered by pediatrician negative for dislocation. Pt has failed 2/3 swallow studies and is scheduled for repeat swallow study next month.

Insurance: BCBS, Mom has applied for Cap C Medicaid

Equipment in the Home: Bi-Pap for used while sleeping, Percussion Vest, Easy S stroller, Snug Seat Stander, Suction Device, Permobil Power Chair on loan from Durable Medical Equipment Provider (DME)

Other Services: Nutrition, Occupational Therapy, Case Management-CDSA

Medications: Depakote, Liquid Albuterol, Benefiber, Miralax, Prevacid, Combivent inhaler, Levocarnitine, Multi-vitamin, Pulmicort

Nutrition: Pt receives all nutrition and liquids through G-tube.

PMH: Normal B Hip X-Rays, GERD, sleep apnea

Social: Pt lives with parents and older sister. She receives 12 hours of nursing 7 days a week.

Family Goals: “Tiffany will sit, move on the floor and learn to use a power chair.”

Objective:

Observations: Tiffany smiles, laughs and talks with PT, nurse and Mom with on word speech. She answers yes and no questions appropriately. In supine her hips are flexed, externally rotated, abducted with knee flexion, B ankle PF or in a windswept position to the right or left. Vision and hearing appear WNL. Pt can manage her secretions with verbal cues during PT but does require breaks in supine to aid with swallowing.

Posture: With support at trunk pt sits with L C-curve of spine (left Convex, R concave). Curvature correctable with manual pressure to left side of trunk.

Trunk/Core Strength: Tiffany can hold her head in midline in supine, supported sitting and while seated in power chair. After 5 minutes she cannot maintain head in midline and laterally flexes head to the right. In prone, she turns her head partially side to side. When passively placed in prone on elbows and assistance to raise head, she can hold head up with eyes and mouth horizontal (90 degrees) for 10 seconds. In suspended prone she cannot lift head or hips into extension. Unable to pull to sit.

AROM/Strength

Tiffany can actively elevated UEs in supine and sitting to 90 degrees, flexes and extends elbows against gravity. Active forearm supination decreased bilaterally. She cannot weight bear on forearms in prone or extend onto hands in prone. She has normal grasp of toys with B hands. In suspended prone she cannot extend hips or head against gravity. Hip flexion in supine WNL. 50% AROM: knee extension, ankle DF, ankle eversion. WNL plantar flexion and ankle inversion.

PROM:

UE and LE PROM WNL except: R knee extension: Lacking 5 degrees from neutral, firm end feel
B Ankle DF: to neutral B firm end feel

Special Tests:

Hamstring Extensibility 90/90: (Popliteal Angle): Right 30 degrees Left 30 degrees

Function/Development

Tiffany rolls to and from prone using trunk rotation, reaching with UEs and LE flexion, however, takes increased time. She is unable to creep on all fours or commando crawl on her stomach. Tiffany can sit with Max A at trunk for no more than 5 minutes with UE support on bench in front of her. She cannot weight bear through LES when held upright. She rolls to prone but cannot lift her head without assistance. Once head is lifted in prone she can maintain for 10 seconds.

Mobility: Non-ambulatory. Power chair: Tiffany can stop/start on demand, navigates around obstacles with verbal and visual cues 50% of the time, navigates backwards but hits people and objects behind her without cues/min A.

Gross Motor Function Measure:

A: Lying and Rolling: 16 B: Sitting: 10 Sections C-E: 0

Spinal Muscular Atrophy Type 1 is a autosomal recessive disease with apoptosis of the cell body of alpha motor neuron in the spinal cord and selective cranial nerve nuclei in the brainstem. [1,2] This destruction causes motor weakness primarily of the proximal muscles of the trunk and extremities. Intercostal muscles are also involved leading to decreased vital capacity and chest expansion. [2] Individuals with this disease are at risk of developing scoliosis, joint contractures, hip dislocation, decreased bone density, fractures and pneumonia from muscle weakness. [1,2] SMA is incurable but pharmacological research has shown some ability to enhance strength and function. [3,4] Tiffany is prescribed two medicines to improve muscle function in SMA-Type 1, liquid albuterol and Depakote. [3,4] Both of these drugs are prescribed in low doses have no effect on her ability to participate in therapy. [4,5] Hepatotoxicity is one adverse side of Depakote if children less than two. [5] While Tiffany is under PT's care if she develops jaundice her pediatrician and neurologist would be contacted.

Additional subjective and objective information necessary includes insurance benefits for durable medical equipment and orthoses, families willingness to use adaptive equipment , deep tendon reflexes, baseline heart rate and respiratory rate at rest, copies of X-Ray, leg length

measure, specialist assessment notes, gentle PROM of trunk lateral flexion and rotation in supine and observations of pelvic obliquity in supine and supported sitting. [6]

Red Flags for this patient might include fever, presence of pain, and change of respiratory rate or pattern. Patient's with SMA are prone to pneumonia and altered breathing patterns due to muscle weakness. [7] Baseline respiratory rate and heart rate should have been measured during the evaluation. If baseline vitals differed in subsequent PT sessions and were associated with symptom increased temperature, fussiness, or decreased oxygen saturation (patient has a pulsoximeter for home use) then she would need to immediately be referred to pediatrician and/or pulmonologist. At this time Tiffany does not appear to be experiencing pain based on observations. However, the Face, Legs, Activity, Cry and Consolability Behavioral Pain Scale (FLACC) should be used to document baseline levels as scoliosis and joint contractures may become painful. [8] Constipation is also associated with SMA and can create pain and may be accompanied by a fever. Symptoms of constipation are hard stools, back pain and general discomfort. [7] Constipation and pneumonia will mostly likely be identified by home health nurses. However, as her regular PT it is important to recognize red flags so that treatment is withheld until she is medically stable.

Hip and spine radiographs were recommended prior to therapeutic trials of a stander and to obtain a baseline measure of the spine. In supported sitting Tiffany was collapsing into right lateral flexion . (See Appendix A for X-Ray) She was referred to an orthopedic to assess her for a TLSO to slow the progression of scoliosis. Development of scoliosis is reported as being as high as 92% and progressing up to 8% a year. [9] Scoliosis is determined by measuring the Cobb angle on a AP radiograph. The Cobb method measures the angle between two perpendicular lines from the superior end plate of the upper most to inferior end plate of the lower most involved vertebra. [10] Prior to trials of a stander, hip AP x-Rays were requested from Tiffany's pediatrician to rule out hip subluxation or dislocation. Hip dislocation and joint contractures are also common secondary impairments in SMA. [1,11,12] Pelvic AP X-rays are used to assess acetabular depth, which is normally less than 30 degrees.[10]

Outcome measures include the GMFM, Power Chair Check List, and the Pediatric Quality of Life Inventory (PedsQL). [13-15] Tiffany's initial evaluation included the GMFM. This measure is found to reliable for patients with SMA although it is was designed for children with cerebral palsy. [14] I chose this outcome measure because it has a significant number of items that assess movement patterns in supine, prone and sitting. Although not initially measured, the Pediatric Quality of Life will be added in a future visit based on research. This tool has the potential to capture quality of life improvements related to the interventions of a power chair, stander and orthoses. These interventions positively impact her function at home and in the community. The power chair checklist assesses her readiness for power mobility and measures progress towards independent use of power chair. [15]

Intervention for Tiffany includes: functional mobility training, sitting balance, prevention of secondary complications including progression of scoliosis, osteoporosis and joint contractures. [1,9,11,12] Specific interventions are stretching of joints prone to contracture progression, use of a stander to provide LE weight bearing and prolonged stretch of LEs, TLSO to improve independent sitting and slow the progression of scoliosis, AFOs and night splints to prevent contracture, power chair training and family education on HEP of stretching and use of orthoses. [1,9,11,12,15-17]

The majority of the evidence to support bracing for scoliosis is in regards to idiopathic scoliosis. [11] Scoliosis in SMA progresses with and without bracing. [2,9] Additionally, some researchers report that TLSO bracing decreases pulmonary function in this population. [2] However, bracing has been found to provide support in sitting. [2,9,11] Tiffany is a young child with early signs of scoliosis secondary to significant proximal weakness. The subjects in most of the research on scoliosis in SMA are children with type II and III. Type II and III SMA are characterized by varying functional abilities. Typically, children with type II can sit and children with type III can walk. [1,9] Although TLSO use may not slow progression of scoliosis, research does support its benefits for sitting. One goal that is important to her family is independent sitting. In only two PT while wearing the TLSO, Tiffany was sat for the first time unassisted for the first time, for 1 minute to play with toys placed in front of her on a bench. Kotwicki et. al.

describes an alternative brace, Suspension Trunk Orthosis (STO) for children with SMA type 1 and 2.[11] This brace differs than the traditional TLSO in that it supports the trunk through a widened posterior base while putting less pressure on the distal anterior portion of the brace. The authors conclude that the STO is tolerated well by children ages 2-7, slows the progression of scoliosis, improves sitting and anecdotally improves respiratory function.[11] This article can be shared with the family, physicians and orthotist for collaborative care. The STO may be an option when she outgrows her current SureStep TLSO.

Tiffany's family was instructed on PROM and stretching into hip extension, knees extension and ankle dorsiflexion. Static stretching has been investigated more thoroughly in children with cerebral palsy and a varied population with neuromuscular disease with mixed results.[1,12,18] Studies have found 10-20 degree found increases in ROM in the lower extremities, but more importantly, maintenance of ROM with stretching.[18] In these studies, discontinued daily PROM, resulted in decreased joint range of motion. On a whole, PROM is recommended at minimum 2x a day for 15-60 second holds.[12] Tiffany's family performs stretches at minimum three times a day with the goal of at least a 30 second hold for three repetitions per motion. Tiffany's family was educated with verbal, written and photographic instructions. (Dad took pictures on his phone of PT stretches for future reference and to educate nurses.)

A Stander is also utilized in Tiffany's plan of care. The purpose of the stander is to provide a prolonged static stretch of hips, knees and ankles with the added benefit of weight bearing through the LES. Standers have been shown to improve or maintain ROM in LEs and there is moderate evidence that they increase bone mineral density in children with CP.[16,17,19] There are no studies were found specific to SMA and standers. A stander is beneficial for Tiffany because she cannot stand or weight bear due to LE and trunk weakness. The stander will be used at least 30 minutes a day with a goal for standing 1 hour a day.

Tiffany's family has private insurance that does not limit her access to PT services or equipment. Her family can support her financially for necessary equipment that insurance does not cover, such as a heated pool for aquatic therapy. This family is fully aware of the realities of

having a child with significant medical needs and they choose to provide her with any advantage they can to promote her well-being and independence. If insurance were an issue her *RECOMMENDED* PT plan would not change, but the actual delivery of care chosen by the family could be altered. With her severe condition, with decreased financial means she would most likely she would be covered under Mediciad for physical therapy. Medicaid would pay for all necessary equipment but their approval process can take up to a year from evaluation to delivery from durable medical equipment provider. If she did not have Medicaid but instead had increased co-pays or deductibles then PT services could be spaced out based on specific goals. Emphasis would be placed on education of home exercise program for PROM and functional activities. Phone consults would be used to advise family on progression of exercises and to alert PT of any area of concerns that might necessitate and face to face treatment. Tiffany could also be seen less frequently after family achieved independence with PROM and equipment use and increased when new learning was necessary. Increased frequency is necessary to educate family and patient for power chair training.[15] Federal and private programs for children with disabilities would be solicited for equipment needs. Equipment could also be loaned from the CDSA until Tiffany is 3.

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Appendix A. Tiffany'