

Background on _____

DOB _____

_____ was diagnosed with Friedreich's Ataxia just before her 11th birthday. The diagnosis took a considerable amount of time to make. Early childhood symptoms were mild, though as later determined, consistent with the neuromuscular disease. Mild scoliosis was noticed during a pediatric check up at age 6, and fatigue was noticeable by her mother with stamina issues progressing through grammar school, in particular ages 8-10. However, _____ was involved in sports, dance, gymnastics, bike riding, and tree climbing and typical age appropriate activities through this time-span. On _____ 10th birthday (August) she broke her right forearm walking with friends, and was casted entering 5th grade. Approximately one month after cast removal, in December, _____ slipped, fell and re-broke the same forearm, requiring surgery and another cast. It was soon after removal of the second cast that an abnormal gait ("toe walking" and wider stance for balance) was noticed by _____ father and balance issues notice by her maternal grandmother and mother. During a pediatric visit in April, a loss of reflex (knee), balance problems, and fine motor/rhythm problems were observed. These symptoms initiated a formal search for the cause of these problems. Friedreich's Ataxia was verified through genetic testing, and both parents were confirmed as recessive gene carriers. Mild HCM of the left ventricle and more pronounced scoliosis was also confirmed. At school, a 504, which was eventually converted to an IEP, was established to deal with the increasing level of fatigue and limit physical activity. PT/OT was eventually prescribed, to help maintain flexibility, range of motion, and loosen heel cords.

_____ continued with most normal activities through 8th grade, with gradual loss of balance, coordination and fine motor skills occurring over this time. It was also during this time that custom orthotic soles were provided for leg and especially foot pronation and that the scoliosis became more pronounced. An aide was provided during freshman year of high school to facilitate walking between classes. Later a service dog "_____" accompanied _____ to high school. The service dog was originally selected as a balance dog. However, _____ walking ability continued to deteriorate and by junior year she could not walk without some assistance or support. During this time a wheel chair was acquired to reduce fatigue and was used by _____ intermittently. During October of _____ junior year she suffered her first heart attack, (irregular and very high heart rate), and was hospitalized for a couple days. Verapamil (a calcium-channel blocker) was prescribed to maintain a steady/slower rate. It was after this attack that more regular use of the wheelchair occurred. However, she was still climbing stairs (with help) to reach her second story bedroom. The scoliosis continued to progress until around age 14, with a maximum upper curve close to 20 degrees and lower curve approaching 40 degrees before it stabilized. Due to the position of the curve, a spinal fusion was avoided. PT/OT continued to be used to maintain capability.

For _____ senior year, she moved to a new high school and house, both newly built for accessibility. The house has step-less entry with a first floor bedroom and customized bathroom with grab bars, roll in shower, undercut sink, etc. for accessibility. "_____" continued to accompany _____ in high school and a wheel chair was routinely used, although she could still walk for short distances with assistance. However, the toe walking and pronation in the legs and feet continued to progress, and balance and fine motor skills continued to deteriorate, despite on-going PT/OT. Within another year or two _____ lost the ability to walk. At this time she was attending and living at Berry College in an accessible dorm room, accompanied by _____, and using a motorized wheel chair to attend classes.

During college, volunteer and eventually paid aides were used, for longer periods of time as the disease progressed, to help _____ get ready for class. This consisted of getting out of bed (supplemented with a super pole), showering, use of the toilet, dressing, grooming, personal care, etc. Parental care in these areas also increased substantially, with eventual multiple visits to the college each week for necessary care. Care for _____ became too difficult, and he eventually (and reluctantly) stayed with _____ parents at home. Until her last year in college _____ graduated with honors after 4 ½ years. She was able to do most of her assignments via computer. During college there were several hospitalizations due to irregular heart problems, and _____ ability to stand (with support) and transfer deteriorated over time. PT was continued throughout college to maintain flexibility and to slow the progression of the disease in this area. Speech therapy was also started during the last year of college and continued after graduation in conjunction with the PT. As part of the PT routine, a stander was used and found to be effective in mitigating problems and improving the ability to transfer. During the last year in college (2009 Spring, Summer and Fall semesters), _____ began to have increasing difficulty with her vision. In addition to nystagmus, she also encountered a loss in color perception and contrast, along a reduction of viewable field of vision. This prevented Laura from reading assignments, and severely reduced her capability to type her assignments. Additional support was acquired for these activities. During this time it was confirmed that the optic nerve had degenerated as a result of the neuromuscular disease.

Upon completing college, and now one year out of school and living at home, transferring has become more problematic. Upon standing (with support) to transfer, _____ heels do not plant on the floor. The heels are 2-3 inches above the floor; within a few minutes they will relax to ¼ to ½ inch above the floor, but standing with support for this length of time for a transfer procedure is not practicable. Also, the right ankle twists during this process, which is often painful, lessens stability and increases the risk of injury. (Given a few minutes to relax the ankle will straighten out better). Active heel cord stretching (by a PT) will bring the angle back to approximately 90 degrees at best. Passive flexing by Laura results in no better than a 130 – 140 degree angle. Severe pronation in the upper and lower legs also prevents _____ from assuming a wider stance for improved balance. The stander has proved to be effective during PT sessions to lessen the problems and pain during transfer. The capability for _____ to have regular use of the standing device would be beneficial to this critical and essential process for her daily living.

In summary, _____ is currently 23 years old and has diagnoses of Friedreich's Ataxia. She cannot ambulate independently, and requires assistance to reposition herself when standing. _____ cannot transfer or stand independently and requires assistance to accomplish these tasks as well. Her bilateral lower-extremity spasticity (adductors, hamstrings and heel cords) and tightness (lack of dorsiflexion), make dressing, using the toilet and bathing difficult for her even with assistance. When standing with assistance, her caregivers often have to force her upper legs apart to facilitate dressing and undressing, and she sometimes experiences pain and discomfort. _____ suffers from chronic constipation which is only alleviated through the use of laxatives several a few times per week to maintain her bowel function. When she has used a supported standing device during physical therapy sessions, for 20-30 minutes, her spasticity is reduced and her hip abduction and ankle dorsiflexion is improved. Her care givers are able to bathe, transfer and dress her more easily, reducing her overall fatigue and discomfort. Although it has been reported that Friedreich's Ataxia patients have confirmed that routine use of a standing device alleviates constipation and bowel difficulties, medical insurance coverage denied further physical therapy sessions – which had included the use of a standing device. Therefore we can only

anticipate that routine use of a standing device would reduce _____ chronic constipation and improve her bowel care program. Her overall activity and participation levels are increased when she has access to a supportive standing device.

The following strategies for supported standing programs have been developed by _____, PT, MS, and DScPT in collaboration with _____, PT, PhD. _____ is an NDT certified pediatric PT from _____. She has worked at NIH, a pediatric rehab hospital (for 8 yrs), and in schools (12 yrs).

Strategies for Supported Standing Programs (for use by _____)

Goal 1: Improve range of motion

ICF category: b7 (Neuromusculoskeletal and movement related functions)

Includes b735 (muscle tone functions) and b7353: tone of muscles of lower half of body:

Functions related to the tension present in the resting muscles and the resistance offered when trying to move the muscles passively.

Studies reported by highest level of evidence

Author	Findings	CEBM Level/sample
Ben et al. (2005)	Tilt table: stretch on ankle dorsiflexors resulted in 4 degrees improvement	1b (RCT) 20 adults with SCI
Baker (2007)	Oswestry Standing Frame 3 times/week, 30 minutes/session, followed by 3-week exercise program: improved hip and ankle ROM (significantly)	2b (RCT) 6 adults with MS
Richardson (1991)	Tilt table, 7-day protocol for 27 days: increased ROM of plantarflexors (dorsiflexion)	3b (Case report) 1 adult with TBI
Salem et al. (2010)	Usual physical therapy treatment, then prolonged standing program 3x/week, in addition to usual physical therapy treatment, then usual physical therapy treatment for 9 weeks: significantly improved stride length (p <.001), gait speed (p <.001), stride time (p <.001), stance phase time (p <.001), double support time (p <.003), muscle tone (p <.02), and peak dorsiflexion angle during midstance (p <.004); improvements not maintained at 3 weeks	4 (Case series) 6 children with CP
Gibson et al. (2009)	Standing frame, 1 hour/day, 6 weeks: significant improvements in hamstring length and possibly increased ease of performance of ADLs; trend for hamstrings to shorten during non-standing phases	4 (Case series) 5 children with CP
Singer et al. (2003)	Tilt table, 30 minutes, with serial casting and Botox injections: improved equinovarus position	4 (Case series) 88 adults with traumatic brain injury (TBI)
Tsai et al. (2001)	Tilt table, one 30-minute session: improved ankle dorsiflexion (statistically significant) but increased tibialis anterior spasticity	4 (Case series) 17 adults with stroke
Bohannon & Larkin (1985)	Tilt table, 5-22 sessions, 2-6x/week, 30 minutes/session: 3-17 -increase in dorsiflexion (average gain of 1 /day)	4 (Case series) 20 adults with stroke
Macias (2005)	Standing program in 55-70 of hip abduction for 45 minutes/day: no loss in ROM of adductor muscles	5 (Presentation) 14 children with CP, 14-17 months old; compared to normal children at age 5

Clinical strategies based on the evidence:

Duration: 30 minutes

Frequency: 5x/week

Goal 2: Decrease spasticity

ICF category: b7 (Neuromusculoskeletal and movement related functions)

Includes b735 (muscle tone functions: Functions related to the tension present in the resting muscles and the resistance offered when trying to move the muscles passively. Inclusions: functions associated with the tension of isolated muscles and muscle groups, muscles of one limb, one side of the body and the lower half of the body, muscles of all limbs, muscles of the trunk, and all muscles of the body; impairments such as hypotonia, hypertonia, and spasticity)

Studies reported by highest level of evidence

Author	Findings	CEBM Level/sample
Tremblay et al. (1990)	Tilt table standing for one 30-minute session: prolonged muscle stretch resulted in reduced spasticity in ankle muscles ($p < .05$); inhibitory effects lasted up to 35 minutes after standing; capacity to voluntarily activate plantar flexors significantly increased post-stretch ($p < .05$)	2b (RCT) 22 children with cerebral palsy (CP): 12 experimental; 10 control group
Taylor (2009)	Standing frame programs 30-40 minutes/day: 16.2% noted reduction in spasticity as a very important benefit	2c (Survey) 386 school-based PTs
Salem et al. (2010)	Regular therapy vs. regular PT plus standing 3x/week, 9 weeks: Improvement in standing group in muscle tone during walking ($p < .02$); improvements not maintained at 3 weeks	3b (Case series) 6 children with CP
Chang et al. (2004)	Tilt table: reduced tone reported by 38 (65.5%)	3b (Survey) 86 PTs in acute care hospitals
Eng et al. (2001)	Supported standing program for 3-4X/week, average of 40 minutes/session: improved reflex activity in 23 adults; 9 reported reduced muscle spasm	3b (Survey) 38 of 126 adult users with SCI
Dunn et al. (1998)	Sit to stand and standing wheelchair 1-6x/week, 30-60 minutes/session: decreased spasticity for 42% of adults, increased spasticity for 4%; correlation between time spent in stander (>30 min) and reported decreased spasticity ($p < .05$)	4 (Survey) 99 adult users with SCI
Tsai et al. (2004)	Tilt table to stretch ankle plantar flexors for 30 minutes resulted in increased passive range of motion for ankle plantar flexors ($p < .05$)	4 (Case series) 17 adults with CVA
Kunkle et al. (1993)	Supported standing program, average of 144 hours over 135 days did not alter range of motion, reflexes, or tone	4 (Case series) 6 adults with SCI
Odeen & Knutsson (1981)	Tilt table standing in at 85° upright in 15° ankle dorsiflexion or plantar flexion during standing for 30 minutes resulted in greatest reduction in resistance to passive movement; average reduction from stretch in supine was 17%	4 (Case series) 9 adults with SCI
Shields & Dudley-Javoroski (2005)	Supported standing wheelchair, 4x/week, 12 min/day, at 61 degrees upright: decreased spasticity	4 (Case report) 1 adult with SCI
Bohannon (1993)	Supported standing at 80 degrees, 30 minutes/session: immediate reduction in lower extremity spasticity lasting until the following morning; no long-term trend after several days	4 (Case report) 1 adult with SCI

Walter et al. (1999)	Re-analysis of data from Dunn et al. (1998): 34% who stood less than once/day and 63% (p<.05) who stood more than 1x/day reported decreased spasticity; 34% who stood less than 30 minutes/day reported decreased spasticity, 51% who stood more than 30 minutes/day	4 (Survey) 99 adult users with SCI
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Clinical strategies based on the evidence:

Duration: 30 minutes can be in short bouts of 15 minutes each

Frequency: 2-5x/week

Goal 3: Improve regularity of bowel function and decrease time performing bowel care.

ICF category: b5 Functions of the digestive, metabolic, and endocrine systems

Includes b525 Defecation functions (Includes b5250-elimination of faeces)

Studies reported by highest level of evidence

Author	Findings	CEBM Level/sample
Eng et al. (2001)	Supported standing program for 3-4x/week, average of 40 minutes/session: improved bowel and bladder function reported in 20 adults	3b (Survey) 38 of 126 adult users with SCI
Hoenig et al. (2001)	Prone standing table for 5x/week, 1 hour/session: improved frequency of defecation (decreased constipation) (p < 0.05); decreased bowel care time (p < 0.05); improvements decreased with reduction in standing from 5 to 3 sessions/week	3 (Case report) 62 year-old with SCI
Shields & Dudley-Javoroski (2005)	Supported standing wheelchair for 4x/week, 12 minutes/day at 61°: improved bowel program and negated improvement when not used	4 (Case report) 1 adult with SCI
Netz et al. (2007)	Standing box to perform exercises, 13-60 sessions: improved sphincter control (p=0.048)	4 (Case series) 13 elderly residents of nursing home (mean of 82 years)
Walter et al. (1999)	Re-analysis data from Dunn et al. (1998): Standing less than once/day: 18% had improved bowel regularity; standing more than once/ day: 33% had improved regularity; standing for more than 30 minutes/day: increased bowel regularity to 39% (p=0.05)	4 (Survey) 99 adult users with SCI
Dunn et al. (1998)	Supported standing program 1-6X/week, 30-60 minutes/session: mixed outcomes for change in bowel care time; positive correlation between frequency and time for program with more regular bowel movements (p=0.001)	4 (Survey) 99 adult users with SCI

Clinical strategies based on the evidence:

Duration: 30-60 minutes/day, divided into 1-2 sessions Frequency: 5x/week