



# SEATED STABILITY FOR SWITCHED POWER WHEELCHAIR USERS

Written by: **JAN FURUMASU**, PT, ATP

Assistive technology is the key to unlocking a person's potential and compensating for disability. Seating and wheelchair mobility can empower children and adults with disabilities to be recognized as individuals of value and to build the self-confidence necessary to interact positively with the world at large. Powered mobility using switches can open up an entire world and potential to actively participate and interact.

Jose is a 19-year-old young man with significant physical disabilities because of Lesch-Nyhan syndrome. Lesch-Nyhan syndrome (LNS), also known as Nyhan syndrome and juvenile gout, is a rare inherited disorder caused by a deficiency of an enzyme leading to over-production of uric acid. LNS affects about one in 380,000 live births.

Uric acid increases in all body fluids, resulting in severe gout and kidney problems. Neurological signs include poor muscle control and moderate intellectual disability. These complications usually appear in the first year of life. Beginning in the second year of life, a particularly striking feature of LNS is self-mutilating behaviors, characterized by lip and finger biting. Neurological symptoms include facial grimacing, involuntary writhing, and repetitive movements of the arms and legs like those seen in clients with Huntington's disease. Infants who had previously been able to sit upright typically lose this ability. Initially, muscles have low tone, (hypotonia) which makes holding the head in an upright position difficult. Affected infants may fail to reach developmental milestones such as crawling, sitting, or walking (developmental delay). Eventually, most children with Lesch-Nyhan syndrome experience abnormally increased muscle tone (hypertonia) and muscle rigidity (spasticity). Deep tendon reflexes are also increased (hyperreflexia). Cognitive impairment may occur and is typically moderate, though some clients have normal cognitive function. Accurate evaluation of intelligence may be difficult because of poorly articulated speech (dysarthria).

Jose's sister reports there are different levels of involvement, and that Jose never demonstrated self-harming or self-mutilating behavior, even as a child. Whereas her nephew, who also has Lesch-Nyhan, is more cognitively involved and does demonstrate self-mutilating behaviors. Jose is not as involved, is cognitively age appropriate, and has never demonstrated those self-harming behaviors, allowing him to use a power wheelchair safely.

Jose is unable to sit upright secondary to extensor posturing and dystonia. His speech is dysarthric, but understandable. He has completed high school with the

assistance of a one-on-one aide. He will be followed medically through California Children's Services (CCS) until age 21. Last year, Jose was hospitalized several times as he has chronic problems with kidney stones due to his high uric acid levels. Currently, Jose is doing well. He likes to play video games.

Jose postures in extension demonstrates dystonic type movement in his extremities. The extensor posturing is seen in conjunction with hip adduction and extension of his hips, trunk and neck. He wears splints on his arms to provide stability and to reduce the dystonic type movement. Per his mother and sister, when not in his wheelchair, Jose sits comfortably in bed or on the sofa with his trunk reclined and wearing his arm splints for stability. He can play Xbox in bed using a standard controller with his right thumb if his hands and the controller are underneath a pillow to control the extraneous movement of his arms.

Jose was seen initially at the Center for Applied Rehabilitation Technology (CART) at Rancho Los Amigos National Rehabilitation Center by an occupational therapist, speech language pathologist and physical therapist. His primary goal was to try a power wheelchair. Stability in the seated posture is critical to ensure consistent, accurate access. Jose needed to sit as upright as

possible with adequate stability to control a power wheelchair. In his current Kids Rock manual wheelchair, Jose sat in thoracic kyphosis and a posterior pelvic tilt with his hips sliding out of the seat. The flexibility of his legs, especially of his hip flexion and abduction, was assessed in supine. During the mat evaluation, we found Jose had significant extensor tone in his hip muscles. When his hips were flexed to 100 degrees, the persistent extensor tone and spasticity in his muscles around his pelvis relaxed. Sitting upright in his manual wheelchair, his hips are currently flexed less than 90 degrees. This leads to Jose sliding out into a posterior pelvic tilt and a windswept posture from his extensor spasticity. As a result, his seat-to-back angle was closed (the seat was wedged up past 90 degrees) to prevent his posterior pelvic tilt and extensor thrusting.

When Jose was in a stable position, his thumb control was accurate for pushing down and coming off a switch, however he did not have the fine motor control needed to operate a small mini proportional joystick. Prior to his next appointment, we contacted the manufacturer's representative and the equipment supplier who worked with this young man at the CCS Medical Treatment Unit (MTU). We requested a non-proportional switch system to allow directional control using low force mechanical switches to drive the power wheelchair. At the time of the next evaluation, we had an Invacare Spree power wheelchair with Ultralight switches (Adaptive Switch Labs) ready and mocked-up with the required seating. A Comfort Company cushion was modified by adding a foam wedge in the front to increase the hip flexion angle to approximately 100 degrees. A foam abductor, which was positioned between his thighs, limited adduction and, subsequently hip extension. Foam wedges were also used as lateral thigh supports to keep his pelvis midline and stable. To assist in limiting extension, his knees were flexed to 90 degrees with his feet stabilized in shoe holders. Jose has poor head control and, if his neck can extend, his total body extension increases. Supporting his head in a forward or neutral position reduced extensor thrusting. A wide chest strap (Bodypoint universal strap) provided input to his chest to prevent arching or extension of his trunk (see Picture 1). Jose uses splints on his arms to maintain elbow extension, providing stability. Both arms were positioned on top of the laptray and further stabilized with a belt (see Picture 2). The laptray was short enough that he could wrap his fingers along the edge to stabilize his hand to use his right thumb optimally. He was able to use his right thumb to activate three switches and could have easily used a fourth, if needed. The three switches were placed around his thumb to allow easy, consistent, and accurate access (see Picture 3).

Jose immediately understood the directional representation of the switches and could move the power wheelchair forward, right and left. Outside, he was able to use the power wheelchair to follow his sister, maneuver the power wheelchair along a wall, turn a corner, negotiate obstacles, and drive along a curving sidewalk (see Picture 4). He demonstrated good cognitive skills and physical endurance, driving the power wheelchair for 45 minutes with verbal cues and close standby assist. No hands-on assistance was needed, and he demonstrated good judgment with just verbal cues. He needed close standby assistance to go through a doorway, but demonstrated good judgment by requesting to change to indoor speed for this task.

Jose and his family live a distance from our program - more than one and one-half hours. Further practice at the local MTU was recommended as a result. The manufacturer's representative brought the same demonstration power wheelchair to the MTU for further trial and practice using the present Ultralight switches on the lap tray. Recommendations were also made for seating to optimize the stability of his body to access the switches for control of driving, as well as a power tilt-in-space for pressure relief and repositioning his body in space. The equipment supplier will work with the MTU therapists to recommend a definitive system once he received additional training. His mother and sister were pleased and pleasantly surprised at how well he did. They took pictures and video to show the therapists and supplier.

CONTINUED ON PAGE 42

**SEATED STABILITY FOR SWITCHED POWER...**  
 (CONTINUED FROM PAGE 41)

In populations with extraneous movement disorders, providing seated stability is critical to ensure energy efficient, accurate, and consistent access to switch controls. By providing consistent access, Jose was able to become independently mobile for the first time in his life, and the power wheelchair will hopefully open doors and opportunities for more life experiences.



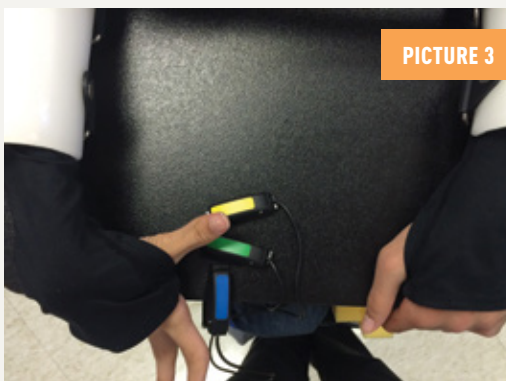
PICTURE 1



PICTURE 2

**CONTACT THE AUTHOR**

Jan may be reached at [JFURUMASU@DHS.LACOUNTY.GOV](mailto:JFURUMASU@DHS.LACOUNTY.GOV)



PICTURE 3

PICTURE 1: Lateral view sitting with his hip, trunk and head in flexion to control his extensor thrusting

PICTURE 2: Arms are in splints and strapped down to the laptray with his fingers wrapped around the edge for stability to prevent extraneous movement

PICTURE 3: Switch access using his right thumb

PICTURE 4: Demonstrating good control and safety judgment driving the wheelchair using switches with his right thumb



PICTURE 4

Jan Furumasu, BS PT, ATP is a physical therapist who has worked at Rancho Los Amigos National Rehabilitation Center since 1979. She is currently an instructor working in the Seating Center and the Center for Applied Rehabilitation Technology (CART), evaluating seating, mobility and assistive technology needs. She is also a consultant for the pediatric inpatient pressure ulcer management service. Since 1990, she has worked on three nationally funded research projects investigating cognitive readiness for powered mobility in young children—most recently a grant on custom wheelchair configuration, funded by the Department of Defense. She has published and presented extensively in areas including pediatric powered mobility, seating, mobility, gait and patients with muscle diseases. Furumasu was the recipient of the Excellence in Clinical Teaching Award in 1994 and the Amistad, Lifetime Achievement award at Rancho Los Amigos National Rehabilitation Center in 2008.

