



AGING WITH A DEVELOPMENTAL DISABILITY

A CASE STUDY OF ONE VERY DETERMINED YOUNG LADY WITH SPINA BIFIDA

I first met Nicole and her mother/caretaker, Nancy, in December of 2012 (see picture 1). She was 31 years old and had recently undergone major urological surgery with ongoing medical complications. She had become weaker from the hospitalization, and was also – for the first time since she was 2 years old – developing pressure ulcers. What I did not know at the time was the remarkable course of management and adaptation she and her family had taken since birth in order to get her to this point.

As with many persons with development disabilities, Nicole’s medical management has been varied, complicated and ongoing. Nicole was born in 1981 at a local hospital. She was born with spina bifida, a birth defect in which the vertebrae do not develop properly (see picture 2). In Nicole’s case, her spinal cord was protruding through the vertebral opening at the L4-5 level. Typically, those with spina bifida are also challenged with gastrointestinal issues, urinary issues, paralysis below the spinal injury level, intellectual disability and orthopedic changes.

The pediatrician at her birth suggested not feeding her and “letting her go.” It was her mother’s obstetrician who suggested that Nicole be evaluated at

Boston Children’s Hospital before making a truly life-changing decision. The neurosurgeon at the children’s hospital evaluated her and suggested that medical treatment could afford Nicole the chance to live successfully within the realm of her obvious medical and physical challenges. She underwent surgery within her first 24 hours of life to close the opening in her spine. She started physical therapy twice daily to improve range of motion, as she was born with her toes touching her nose.

She underwent multiple surgeries in her first few years and spent much time in a spica cast. At approximately 2 years old, she started walking using leg bracing and Lofstrand crutches. She was provided with an Enduro brand pediatric wheelchair in kindergarten to use in school for long distances and to protect her from the class of running children. This began a succession of Durable Medical Equipment (DME) provision that has continued throughout her development.

Nicole lived at home and attended public school with the assistance of full-time aides until the ninth grade. Between the ages of 10 and 11, although she continued to walk with braces and crutches, her leg strength was decreasing and she made the decision to use a manual wheelchair full time. Despite encouragement from her physical therapist to continue walking, she felt the wheelchair was more functional and afforded her faster mobility. She was able to better keep up with peers and attend better in school because she was less fatigued.

In her teens, she required an anterior/posterior spinal fusion to stop the progression of her spinal kyphosis and scoliosis. Bracing with a body jacket and complications after surgery required her to remain in bed for three months. Again she relied on DME, this time temporarily, consisting of a Stryker bed frame and Hoyer lift for transfers.

Nicole left the public school system and attended the Massachusetts Hospital School from the ninth grade until she was 22 years old. There she received physical, occupational and speech therapy services, adapted recreation, as well as her education. She began to learn life skills outside of the family home environment, although her mother remained her primary caretaker and advocate. The last few

THE NEUROSURGEON AT THE CHILDREN’S HOSPITAL EVALUATED HER AND SUGGESTED THAT MEDICAL TREATMENT COULD AFFORD NICOLE THE CHANCE TO LIVE SUCCESSFULLY WITHIN THE REALM OF HER OBVIOUS MEDICAL AND PHYSICAL CHALLENGES.

years of her schooling were focused on skills needed to live in the community. Nicole's goal was to live "on her own" and was supported by her family. Her case manager helped find an appropriate assisted living agency (Charles River Association for Retarded Citizens) and a day program with a work component (Opportunities & Visions). She was also at that time connected with a community based primary care medical practice for severely disabled adults called Boston's Community Medical Group. She continues to be followed by this group, which has since expanded and changed its name to Commonwealth Community Care.

Nicole managed to overcome numerous physical and medical challenges, including a life threatening infection when she was 20 years old. She has always been allowed and encouraged to make her own decisions in her care.

DESPITE ENCOURAGEMENT FROM HER PHYSICAL THERAPIST TO CONTINUE WALKING, SHE FELT THE WHEELCHAIR WAS MORE FUNCTIONAL AND AFFORDED HER FASTER MOBILITY. SHE WAS ABLE TO BETTER KEEP UP WITH PEERS AND ATTEND BETTER IN SCHOOL BECAUSE SHE WAS LESS FATIGUED.

She currently lives in a group home setting with caretakers, and spends most weekends with her family at her childhood home. She attends a work-based day program on weekdays that she enjoys. Her mother continues to be very involved with overseeing her care, coordinating medical needs and training caretakers at the group home. She has found it difficult to pull back completely from daily care needs due to inconsistency with multiple agency caretakers.

In the area of Activities of Daily Living, Nicole continues to require moderate assistance with stand pivot or slide board transfers and moderate assistance with showering, dressing and toileting.

Nicole is able to feed herself and perform oral hygiene with set up. She is able to help with food preparation and light house cleaning. She is working on money management and work skills. She is independently propelling her manual wheelchair and using both her cell phone and iPad.

Since 2007, Nicole has been using a Quickie GP manual wheelchair, planar back with lateral trunk supports, Ride seat cushion, left lateral pelvic support, raised footplates and custom tray that she can remove herself on a Mount 'n Mover mounting system.

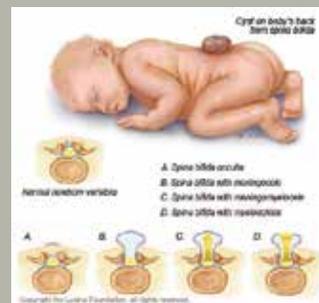
Seating modifications have been required to accommodate her fixed scoliosis and pelvic asymmetry (her spine is fused into her pelvis), fixed hip flexion contractures (from birth), and short leg length. Nicole says that getting the wheelchair tray was "life changing" – allowing her a sense of independence, with the ability to carry books and her lunch in the cafeteria without asking for help. She also uses a tub transfer bench and a hospital bed with Alternating Pressure Pad (APP) mattress overlay.

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PICTURE 1

Nicole and her mother, Nancy



PICTURE 2

Spina bifida



PICTURE 3

Nicole and her power assist wheels

CHANGES NEEDED TO ACCOMMODATE CHANGING CONDITIONS AS NICOLE AGED

At age 31, Nicole had another major surgery with complications including weight loss, decreased leg strength, left hip arthritis and pain, and a pressure ulcer on her left ischial area. She was sitting differently and the Ride cushion no longer fit properly. We trialed numerous cushions and used the Roho X-Sensor pressure mapping system as a tool. She trialed a Roho Quatro Select, a Roho high profile dual valve, and a Comfort Company Vector cushion. After much assessment and trials, a new custom Ride cushion was provided, with good results.

SHE DID NOT WANT TO CONSIDER POWERED MOBILITY, AS THE MANUAL WHEELCHAIR WAS PART OF HER EXERCISE AND WEIGHT CONTROL PROGRAM

Subsequent issues with a pressure ulcer on her right lateral trochanter necessitated another round of assessment and pressure mapping. Although staff from the wound clinic she was being seen at assumed the wheelchair seat cushion was the problem, extensive assessment showed the bed mattress was causing the pressure. Changing from an air overlay to a full Low Air Loss (LAL) mattress solved the problem. We also provided a gel pad on the seat of her tub transfer bench as preventative treatment.

NICOLE PLANS TO STAY ACTIVE WITH FRIENDS. SHE ACKNOWLEDGES THAT SHE NEEDS TO ADJUST EACH TIME EQUIPMENT CHANGES HAPPEN AND “THAT’S OK AS LONG AS IT’S AN IMPROVEMENT.”

In 2013, Nicole began to complain of bilateral wrist pain. This progressed and seemed to be worst when she was propelling her manual wheelchair for long distances. She did not want to consider powered mobility, as the manual wheelchair was part of her exercise and weight control program (Note – the author is not suggesting that a manual wheelchair be used to provide exercise or lose weight).

After discussing options, she agreed to trial some power assist add-on units. We trialed the Frank Mobility E-motion, the Max Mobility Smart Drive, and the Quickie X-tender.

Each brand varied in features and driving. Nicole has short arms and limited arm strength, limiting her capability to control the devices.

Once educated, she was able to utilize the Frank Mobility system fairly well, but had trouble reaching the on/off switch at the axle. She did not find the Smart Drive system as intuitive and she experienced some confusion using it. She also found it too powerful for her to control easily. The Quickie X-tender was the most comfortable for her to control so this was chosen and attached to her current Quickie GP wheelchair. Once received, she underwent training at home and at her day/work program. She has not had any problem using this add-on feature and she has also ceased to have any complaints about wrist pain (see picture 3).

Nicole’s future plans include staying in a community living apartment and participating in a work program. She would like to do more public speaking on disability awareness. She has been involved with training programs for interns at a local medical school. Nicole plans to stay active with friends. She acknowledges that she needs to adjust each time equipment changes happen and “that’s ok as long as it’s an improvement.”

Her message for rehab technology suppliers is:

- 1 – Listen to the client and caretakers regarding their needs and concerns.
- 2 – Give choices, including good explanations and equipment trials.
- 3 – Sometimes you need time to figure things out (don’t rush the decisions).
- 4 – Good people make all the difference (shout out to Guy Joyce from Hudson/NSM).

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NICOLE AND HER FAMILY GRATEFULLY ACKNOWLEDGE THE MEDICAL PERSONNEL AND EQUIPMENT SUPPLIERS OVER THE YEARS WHO HAVE HELPED HER TO REACH HER GOALS AND WHO CONTINUE TO HELP HER FUNCTION AT HER MAXIMUM POTENTIAL.

CONCLUSION:

It is clear to me, after studying Nicole's case, that the success of persons living with developmental disabilities is determined by many factors, including:

- Consistent caretakers who take an advocacy role;
- Proper medical and rehabilitation care;
- Access to enabling equipment that changes as the person's needs change; and
- A person's own determination to make his or her goals happen.

Nicole and her family gratefully acknowledge the medical personnel and equipment suppliers over the years who have helped her to reach her goals and who continue to help her function at her maximum potential. She is truly an amazing young lady.

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