# Management of Spinal Cord Injury in Patient with Down Syndrome

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***Study design:*** Case study

***Objective:*** To evaluate the nursing roll of Spinal Cord Injury in patients with Down Syndrome.

***Introduction:*** One in 733 of newborns has Down Syndrome[1]. Down Syndrome**,** also known as trisomy 21, is a genetic disorder typically associated with physical growth delays and joint laxity resulting in atlantoaxial instability (AAI), which denotes increased mobility of C2 in relation to C1. Management of spinal cord injuries in Down Syndrome is highly complex and requires special care.

Children with Down Syndrome have many problems, and there are many physical differences that are associated with a Down Syndrome diagnosis such as hearing loss, abnormal vision, congenital heart disease, gastrointestinal abnormalities, endocrinological disorder , Speech apraxia, Sleep disorders, Feeding disorders, Developmental disabilities ,Musculoskeletal and movement problems and increased risk of infection[2]. The families of Down Syndrome children also have psychological stress. Nurses in the acute and primary care settings should be having proper knowledge to provide patients with optimal care and be familiar with the types of resources available for families of children with Down Syndrome. After an initial diagnosis of Down Syndrome, families will need extra emotional support throughout their journey of learning how to care for their child.

Nursing care of Spinal Cord Injury with Down syndrome will bring new challenges for nurses. After cervical cord injury, the respiratory, cardiovascular, GI, urinary, and metabolic systems are less of functional[3]. The nurse's role in the case of Spinal Cord Injury with Down Syndrome are special and more complicated. In the initial phase of the injury, it is important to support both patient and family by explaining what is happening in terminology that is easy to understand[4], and help calms some of the parents' fears and worries. Parents must be aware of the effects of the patient being limited to lying in bed; with a very restricted view of what is happening around them there is a risk of medical deterioration, fear, and emotional

exposure greater than the general case of Spinal Cord Injury. During the period of care in the acute phase there is frequent assessment, close observation, and special care skills and knowledge of respiratory, cardiovascular, neurological, bowel, and urinary systems changes are required. Communication is an important to arrange effective physical and psychological care. Additionally, communication with Down Syndrome patients is limit by impaired cognitive skills and inabilities; they may be unable to use their hands or arms when they are talking[4]. Family members are a key to success in solving communication problems during hospitalization. Collaboration is another crucial nursing roll to successful management between multidisciplinary teams to give effective care for the patient. Before discharge, nurses must add a value by educating, helping, encouraging and empowering the patient and family to have rehabilitation and selfcare skills. Finally, after discharge from the hospital, continuous care is required in case of Spinal Cord Injury with Down Syndrome**.**

***Case presentation:*** A five-year-old boy with Down Syndrome was referred from a provincial hospital. He fell from a swing three days previously and developed a progressive neurological deficit. After CT scan he was diagnosed with C1-C2 subluxation causing incomplete spinal cord injury and spastic quadriparesis. During the initial presentation, he was intubated with a ventilator and referred to Chiang Mai University Hospital. During admission, the physiological change toward paralysis was explained to his family by the orthopedic staff with psychological support from the primary nurse. The communication between nurse and patient began in this first period of care. During the hospitalization, the family could stay with him, and the nursing staff gave them information about the daily progress and treatment. A weaning program, conducted by nurses, was started in the second day of admission. Early nutrition support started after three days of admission by nasogastric feeding. At this time there was an assessment about a swallowing problem before admission from the family's history that he could only eat a semi-liquid diet after stopping breast feeding. The record from the provincial hospital did not indicate a history of swallowing problems; only endocrine function and the current trauma problem were on the referral document. Ten days after admission, a tracheostomy was performed because of persistent respiratory muscle paralysis. Nurses gave information about the benefit and risk of the tracheostomy procedure, the progression of eating after tracheostomy, and gave mental support for the family. After the tracheostomy nurses explained about the congenital abnormal tracheoesophageal fistula from Down Syndrome that was found at the time of the tracheostomy operation. Thus, he would be still be fed through a nasogastric tube. Meanwhile, exercise of all extremities was done, encouraging his family to help with passive range of

motion and playing activities. Joint stiffness was also detected in his right foot at the time of admission, and an appropriate night splint was made to release of the joint stiffness at both feet.

The surgical treatment would require a halo vest, which costs around 120,000 Thai baht (3,800 USD) and was not covered by government insurance. The nursing team suggested ways to find payment support, which was obtained after ten days. Twenty-six days after admission to stabilize his condition he underwent an operation to fuse the atlanto-occipital and atlanto-axial joints. External stabilization was done by application of the halo vest intraoperatively to augment the internal stabilization. This made the patient uncomfortable and fearful. Cooperation between the family and nurses could help him relax and accept his condition in time.

After operation the weaning program continued. He was able to breathe by himself one month after injury and attempted ambulation with the halo-vest. The ambulation time made him happy and relaxed, and he cooperated with the nursing care. The orthopedic staff removed the halo-vest one month after surgery and switched to a SOMI brace with a head strap. He had a rehabilitation program for two months in the hospital. Care giver education and training skills for a home program was started after three months in the hospital. It included suction, feeding, and activities of daily living. At discharge he was still breathing with a Jackson tube, continued nasogastric feeding, night splint of his feet, and ambulation with SOMI brace with head strap. His hospitalization in Chiang Mai University Hospital was 104 days.

This case was followed up in Chiang Mai University Hospital every 1-2 months to confirm the stability of fixation by the orthopedic doctor, continuous rehabilitation program by the rehabilitation doctor and management of his fistula and swallowing problem by an ENT doctor. He also had a continuing rehabilitation program in the provincial hospital every week.

A home visit was done every six months by an APN to evaluate the progression of his disability, assess the effectiveness of family care, and help them to solve other problems of suction tube supply, change follow up appointment that they required, and advocate for them to get government support. An APN was available to contact 24 hours a day. At the two months hospital visit, the orthopedic doctor let him ambulate without SOMI brace. Six months after discharge he could sit by himself and had started oral liquid diet; nasogastric feeding and tracheostomy suction continued. One year after discharge he still sitting, could not stand, and

was trying jelly plus nasogastric feeding with less suction. Eighteen months after discharge he started a semi-liquid diet and less feeding in the daytime; he could play on the ground with his brother and friends; and started standing with a bar support. Currently, he has nearly finished with the tracheostomy, and is able to oral feed with semi-liquid diet without aspiration and is undergoing continuous physical therapy for standing and walking. No complications two years after discharge.

***Conclusion:*** In case of Spinal Cord Injury in a child with Down Syndrome, nurses are ideally suited to play an integral role of care in assessment, diagnosis, intervention, and evaluation of the outcome of care. Other roles involve information, education, encouraging, empowerment, helping, supporting, cooperation and communication. The most important and hard role of this case study was communication between the nurse and patient. It took time and good cooperation from the family, but communication role was the key for success in the other roles. A lesson learned from the long way of more than two-year case study is the importance of nursing care in Spinal Cord Injury with Down Syndrome Heightened emphasis should be in place to prevent such injuries in these high-risk populations.

**Keyword:** Spinal Cord Injury, Down Syndrome, Nursing care.

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