CASE REPORT

Case Report: Ventilator weaning, tracheostomy decannulation and noninvasive ventilation in an adolescent with autism spectrum disorder and new onset spinal cord injury

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Abstract

Introduction Spinal cord injury (SCI) is a cause of significant psychosocial stress not only to the individual with SCI but also to their family. This is compounded when an individual with a new SCI has premorbid behavioral and medical conditions. For individuals requiring long term positive pressure ventilation, transition to noninvasive ventilation (NIV) can improve the long term outcome and improve quality of life.

Case presentation This case report describes a teenage boy with premorbid autism spectrum disorder who incurred an acute SCI and developed chronic respiratory failure. He was admitted to acute inpatient rehabilitation with tracheostomy and ventilator dependence. Using an interdisciplinary team approach with in vivo desensitization behavioral interventions, he was successfully weaned off mechanical ventilation, his tracheostomy tube was removed, and he was transitioned to NIV.

Discussion This case describes a medically complex adolescent who was successfully transitioned to NIV through behavioral desensitization using a team approach. This is noteworthy given the magnitude of behaviors demonstrated prior to his desensitization protocol. This case demonstrates how serious behavioral barriers to NIV can be overcome using desensitization and strategic behavioral reinforcement techniques.

Introduction

Spinal cord injuries (SCI) in children and adolescents are devastating not only for the individual but also their family.

Caregiver burden increases significantly if the patient has respiratory compromise. For children with SCI necessitating mechanical ventilation or tracheostomy, transition to noninvasive ventilation (NIV) and decannulation in individuals with SCI can be achieved [1]. However, tolerance and compliance with noninvasive positive pressure ventilation (NIPPV) can be challenging for children and adolescents, regardless of cognitive status and premorbid conditions [2].

This case describes an adolescent with acute SCI and significant behavioral disturbances due to premorbid autism spectrum disorder (ASD), intellectual disability (ID), and touch intolerance who successfully weaned off the ventilator, was decannulated and transitioned to NIPPV using behavioral desensitization techniques.

Case presentation

Our patient presented at 16 years of age for posterior spinal fusion (PSF) due to neuromuscular scoliosis and kyphosis. Prior to surgery, he had an 80° right mid-thoracic curve and a 30–40° left lumbar curve. His past medical history was significant for ASD, agenesis of the corpus callosum, ID,

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gastroesophageal reflux disease and von Willebrand factor deficiency. Based on a prior polysomnogram (PSG), he had premonitory obstructive sleep apnea (OSA) but did not require treatment for OSA prior to surgery.

He was nonverbal at baseline but followed simple commands. He had hearing impairment but was inconsistent in his use of required hearing aids due to his behavior. He was independent with ambulation, but required supervision for safety. Prior to spine surgery, he had bilateral upper extremity hypotonia with poor fine motor skills although he was able to self-feed using standard utensils without modifications to food. He had a premonitory history of self-injurious behavior (e.g., head banging, impulsivity, removal of medical equipment from self) that was initially treated as an outpatient with risperidone.

The individual had an uneventful surgical procedure which included PSF from thoracic spine level 2 (T2) to lumbar level 4 (L4), posterior segmental instrumentation from T2 to L4, osteotomies from T2 to L4, and a right-sided bone graft. He had normal motor, sensory and transcranial monitoring during the procedure. On postoperative day 1, he had loss of motor and sensory function to his lower extremities and returned to the operating room to decrease tension on the spinal cord. Posterior spine hardware was removed bilaterally with removal of right-sided thoracic screws and placement of a right-sided titanium rod. No return of function was noted following the procedure. Following removal of hardware, a MRI showed T2 hyperintense signal in the spinal cord at T2 and C7, consistent with spinal cord ischemia. This ischemia resulted in spinal cord damage at the T3–T6 level. The patient remained in the Pediatric Intensive Care Unit (PICU) for a total of 56 days. He required tracheostomy tube placement due to chronic respiratory failure 1-month post procedure. He had one episode of bradycardia and hypoxemia requiring cardiopulmonary resuscitation with chest compressions and epinephrine. He required scheduled catheterizations of bladder due to neurogenic bladder. He had neurogenic bowel with paralytic ileus and resultant significant abdominal distension. Once medically stable, he was transferred to our facility for intensive SCI rehabilitation.

The individual was admitted to the inpatient pediatric rehabilitation unit where he participated in 5 hours of daily physical therapy, occupational therapy, speech therapy, therapeutic recreation, and child life therapy along with support from rehabilitation nursing and respiratory therapy. Physical and occupational therapy focused on activity-based restorative therapy including electrical stimulation.

Given the individual's premonitory ASD diagnosis, pediatric neuropsychology was consulted to evaluate cognitive functioning. Pediatric behavioral psychology was also consulted to assess and develop a developmentally appropriate behavioral plan in efforts to minimize

potentially dangerous behaviors, such as touching or pulling at nasogastric tube or tracheostomy/ventilator tubing.

The individual had a 1:1 behavioral rehabilitation aide with him at all times, including during rehabilitation therapy sessions, to redirect and block dangerous behaviors. These behaviors were so concerning that he required gastrostomy tube placement due to aspiration risk from inadvertent removal of tube during a feeding. The 1:1 aides also blocked intermittent self-harm and aggressive behaviors (i.e., hitting self in head with fist, pulling/pushing staff, hitting legs with open palms, throwing items).

The individual admitted on mechanical ventilation via tracheostomy on synchronized intermittent mechanical ventilation mode with settings of rate of 12, peak inspiratory pressure of 22, pressure control of 10 and pressure support (PS) of 12 with 30% FiO₂ requirement. Pediatric pulmonology was consulted for ventilator management and weaning. Given his level of thoracic SCI, it was thought he had excellent potential to wean off the ventilator. He had significant abdominal distension which impacted his pulmonary status requiring an aggressive bowel regimen and rectal tubes. Once excessive distension resolved, he readily weaned off mechanical ventilation and supplemental oxygen. He required only continuous positive airway pressure (CPAP) with PS without a rate by rehabilitation day 50. He tolerated an open tracheostomy without any ventilator support by rehabilitation day 59.

On rehabilitation day 70, he had an open tracheostomy PSG which showed central apneas with desaturations as low as 77%. There were no obstructive apneas and minimal hypopneas. The apnea–hypopnea index (AHI) was normal at 4.0 events per hour. Following this favorable sleep study result, he began tracheostomy capping and was able to undergo a capped tracheostomy sleep study on rehabilitation day 104.

The capped sleep study demonstrated frequent obstructive apneas. The AHI was 43.9 events per hour, which is significantly abnormal. Despite use of 1 ½ liters per minute of oxygen at the beginning of the study, there were frequent episodes of desaturation and several were severe. Desaturation events were associated with obstructive apneas with an oxygen saturation nadir of 76%. Although the individual had an SCI at a level that would not generally require ventilatory support, his underlying obstructive and central sleep apnea necessitated ongoing treatment.

Given the individual's behavioral profile, the team and family agreed to implement a behavioral protocol to focus on tolerance of nasal bi-level positive airway pressure (BiPAP) for noninvasive ventilatory support. Pediatric behavioral psychology along with rehabilitation nursing, respiratory therapy, and the child's parents worked together to implement a gradual desensitization plan to achieve this goal.

Table 1 Desensitization and behavior therapy procedures used across days.

Day	Tolerance	Parent participation	Interventions	Airflow	Comments
1	25 s nasal mask only	Observation and behavioral intervention	Significant blocking by psychology staff, parent and 1:1 aide	None	
2	10 s mask and headgear straps	Observation and behavioral intervention	Holding hands with aide/parents	None	New mask type obtained which was more difficult to remove by patient.
3	3 min mask only	Observation and behavioral intervention	Arm immobilizer added for safety	None	Minimal distress.
4	60 s mask and headgear	Observation and behavioral intervention	Arm immobilizer	None	
5	50 s mask and headgear	Placed mask for 30 s, four times a day	Minimal blocking	None	Minimal distress. Respiratory illness delayed trials for 2 days.
6	1 min 20 s mask and headgear	Placed mask for 30 s, four times a day	Arm immobilizer	None	
7	2 min mask and headgear	Placed mask for 30 s, four times a day	Arm immobilizer	None	
8	10 min mask and headgear	Placed mask for 10 min, four times a day	1:1 observation	Added low flow air	
9	10 min mask and headgear	Placed mask for 10 min, four times a day	Arm immobilizer	Low flow air	
10	30 min mask and headgear	Placed mask for 10 min, four times a day	1:1 observation	Low flow air	1st night trial: 30 min night time trial with preliminary settings on BiPAP.
11–15	BiPAP with preliminary settings and capped tracheostomy tube tolerated all night	Placed mask for 10 min, four times a day	Significant blocking by 1:1 needed at 23 min mark	Low flow air	2nd night trial: fell asleep within 45 min and slept all night with preliminary settings on BiPAP.
16–24	Home BiPAP setting tolerated all night	Placed mask at bedtime and followed protocol	Arm immobilizer	Low flow air	Continued tolerance of BiPAP settings with repeat PSG for home settings on day 15.
		Continued use of established behavioral protocol	1:1 observation	Home BiPAP settings	Tracheostomy tube removed.
					Discharged to home after tolerating 8 days without tracheostomy tube and on home BiPAP settings.

Prior to starting in vivo desensitization to promote BiPAP tolerance, a clinical interview was conducted with the parents. They reported that the patient had a long-standing history of difficulty tolerating items on his head and face (e.g., hats, hearing aids) which was consistent with his observed behavior of frequently touching and pulling at his tracheostomy and ventilator tubes during rehabilitation.

To address the child's BiPAP intolerance, a gradual systematic in vivo desensitization procedure was used. Desensitization trials consisted of gradual exposure to BiPAP materials (i.e., mask then air pressure) paired with positive reinforcement in efforts to build tolerance. Ten BiPAP desensitization sessions occurred with pediatric behavioral psychology during a 16 day period of time. Desensitization trials were paired with a visual timer and verbal prompt (i.e., "First nose, then [name of reinforcer]/break") to provide antecedent prompts prior to each exposure and predetermined reinforcement (i.e., breaks or item/food). Initial trials included the use of tangible contingent reinforcement, which pairs completion of goal behaviors with a reinforcer or reward. However, it was determined that the individual was able to tolerate desensitization trials better when provided noncontingent access to highly preferred items, which served as distraction during trials.

A preference assessment was conducted by psychology with the individual to determine highly preferred items. These highly preferred items were used as reinforcement and distraction during desensitization trials. A combination of sensory items, music videos, and food was used for initial distraction. Once desensitization trials moved to the child's bed, only music was utilized as a form of distraction as his parents indicated his premorbid sleep hygiene included falling asleep while listening to music.

During in vivo desensitization, all attempts to remove the BiPAP mask prior to the timer indicating the end of a trial were blocked by the family, psychology staff, or 1:1 aide. Once the visual timer indicated the end of a desensitization trial, the child was provided enthusiastic social praise and the BiPAP mask was removed. Initial in vivo desensitization sessions consisted of daytime trials where a psychology staff member incrementally increased the amount of time he wore the BiPAP mask. Later trials included the assistance of respiratory therapists and introduction of nasal air flow. Air flow was initially introduced at the lowest setting and gradually increased based on PSG results.

Once he demonstrated tolerance of BiPAP with psychology staff, the parents were assigned in vivo desensitization homework in order to increase exposure trials, promote tolerance, and generalize learned behavioral management skills across caregivers. Psychology observed the parents conduct BiPAP desensitization trials before assigning parent-led in vivo desensitization homework. During parent-led desensitization sessions, instructions

were to apply the BiPAP mask for a prescribed duration and frequency, always without air pressure, several times per day.

When in vivo desensitization progressed to bedtime, a visual schedule of the bedtime routine was introduced to assist with generalization and incorporation of BiPAP into bedtime. Overnight trials were structured such that the parents began prompting the child through his bedtime routine at 8:45 pm. A respiratory therapist entered the individual's room around 9:00 pm to place the BiPAP mask and turn on air pressure while he was still awake. Parents, nursing staff and 1:1 aides utilized distraction techniques, including music playing in the background and positive verbal encouragement, to assist with self-soothing and to promote sleep onset. He was initially allowed 30 min (ultimately increased to 60 min) to fall asleep with the mask on. If the patient successfully slept throughout the night while wearing the mask, respiratory and nursing staff were instructed to set his visual timer for 1 min prior to removing the BiPAP mask in the morning, in order to continue to pair the association between the timer and mask removal. If the child became agitated during the night, support staff were instructed to block attempts to remove equipment and to prompt him to fall back asleep by providing a soft touch to his hands and using positive verbal encouragement. Initially, if the individual remained awake after 5 min (ultimately increased to 15 min), a timer would be set for 1 min after which time the mask would be removed for the remainder of the night.

The desensitization protocol was implemented by psychology staff, 1:1 rehabilitation aides, rehabilitation nursing staff, respiratory therapist staff, and the parents. See Table 1. He initially did not tolerate the mask, but quickly acclimatized from repeated exposure and differential reinforcement of compliance. Following successful desensitization, the child had a capped tracheostomy polysomnogram with nasal BiPAP with preliminary settings of inspiratory positive airway pressure (IPAP) 15 and expiratory positive airway pressure (EPAP) 8 on day 134. The study showed a normal and significantly improved AHI at 2.7. There were only occasional central apneas noted but no significant obstructive apnea events. Decannulation was recommended and completed following demonstration of continued tolerance to BiPAP with capped tracheostomy. He was discharged to home on BiPAP with settings of IPAP of 12, EPAP of 6 and rate of 10 after 140 days of inpatient rehabilitation.

At the time of discharge, the child was taking a soft diet by mouth with supplemental nutrition via gastrostomy tube. He was assisting with upper extremity tasks such as grooming, dressing and propelling his manual wheelchair but remained nonambulatory. Importantly, he remained compliant with use of the BiPAP following discharge.

Discussion

New onset spinal cord injuries (SCI) in children and adolescents are devastating events that impact not only the individual but also their family. The increased burden of care is significant. For example, the addition of invasive treatments such as bladder catheterization and bowel programs in children with neurogenic bladder and bowel can result in caregiver stress. This is also true if the individual requires ventilatory support or has a tracheostomy in place. Transition to NIV in children with SCI can be achieved. However, it is well documented that tolerance and compliance with NIPPV can be challenging for persons of all ages, regardless of their cognitive status.

The decision to proceed with tracheotomy in a child is difficult for the both the family and medical team providing care. Wilford described the multi-dimensional decision-making process for children with complex medical problems, including mechanical ventilation and tracheostomy [3]. Weaning off mechanical ventilation and decannulation, even for short intervals, can be frightening and uncomfortable for those who are dependent on it. However, weaning can dramatically improve the quality of life for the individual and their family. Additionally, multiple studies have demonstrated increased mortality in children with a tracheostomy [4–6].

In this case, both the parents and medical team were in agreement that the individual's risk for self-harm with a tracheostomy tube was significant due to the severity of behaviors. Treatment of OSA was needed to ensure quality of life for this individual. Although surgical treatment of OSA using mandibular advancement techniques has been documented in children with behavioral issues [7], this patient was able to take advantage of his time in acute rehabilitation to optimize his airway management. Given his distress and avoidance or escape of objects near or on his face, desensitization behavior therapy was essential to achieving tolerance and adherence. The team was able to use both positive reinforcement (distracting stimuli, praise) and negative reinforcement (discontinuation of exposure trials contingent upon a planned period of compliance) to promote increased tolerance of the therapeutic intervention, nasal BiPAP, with the ultimate goal of decannulation achieved.

Use of behavioral protocols to implement use of NIPPV in children and adolescents has been documented in the literature [8, 9]. In a small sample of children requiring noninvasive ventilatory support, Koontz et al. demonstrated that increased compliance with NIPPV was achievable when psychologists used targeted and individualized behavioral strategies, compared with those who did not have psychological support [10]. Inclusion of respiratory therapy in both the outpatient [11] and inpatient setting [12]

has been beneficial in increasing compliance with NIPPV. Successful development and implementation of inpatient behavioral protocols depend on collaboration of family members along with pediatric psychology, respiratory therapy, nursing, and medical staff.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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