A Successful Structured Inter-Disciplinary Weight Reduction and Rehabilitation Programme for a Morbidly Obese Ventilator-Assisted Adolescent with Prader Willi Syndrome

Poon SWY, Tung YL, Hui CCK, Lau ESK, Ho PY, Chau SK and Lee SL

Department of Pediatrics and Adolescent Medicine, The Duchess of Kent Children's Hospital at Sandy Bay, Hong Kong

Abstract

Complications of obesity account for major causes of mortality and morbidity in patients with Prader Willi Syndrome (PWS). We report a successful weight reduction and rehabilitation program that liberated a morbidly obese teenage boy with PWS from long term ventilator support via tracheostomy. We believe that the concept in designing the program may also be beneficial to other patients with morbid obesity who fail to comply to simple weight reduction measures.

Introduction

Prader Willi Syndrome (PWS) is an uncommon genetic condition that is associated with substantial physical, mental and behavioral problems. Typical clinical features include hyperphagia and rapid weight gain leading to obesity and Type 2 Diabetes Mellitus (DM). Conventional management is mainly targeted at weight control including strict diet supervision, exercise program, counseling for behavioral problem, and management of DM and Obstructive Sleep Apnea Syndrome (OSAS). For OSAS, positive airway machine is often needed. So far, patient with PWS requiring long term ventilator support via tracheostomy has never been reported. We hereby report a case that was successfully liberated from ventilator support and tracheostomy with a well-structured weight reduction and rehabilitation program.

Case Presentation

An 18-year-old Pakistani boy was diagnosed with PWS with abnormal methylation pattern at small Nuclear Ribonucleoprotein-associated Protein N (SNPRN) locus at the age of 2 years. He presented with hypotonia and feeding difficulties in the neonatal period. He started to exhibit food seeking behavior with accelerating weight gain since age 4 to 5. His family often conceded to food demands to avoid tantrums. He was also found to have moderate intellectual disability with features of autism spectrum disorder. He was diagnosed to have OSAS at age 8 and required nocturnal Bi-level Positive Airway Pressure (BiPAP) at age 12. His compliance was unsatisfactory as he often had temper outbursts. As he approached adolescent years, his family found it increasingly difficult to control his food foraging behavior, thus imposed minimal restriction to food. He developed Type 2 DM, hyperlipidemia and hypertension. Attempt for weight reduction in the general pediatric ward at an acute hospital was unsuccessful as he developed aggression towards hospital staff. At age 16, he was admitted to the Pediatric Intensive Care Unit (PICU) for an episode of gastroenteritis with dehydration. He weighed 145 kilograms (kg) with a Body Mass Index (BMI) of 67 kg/m² on admission. He was then intubated for ventilator support as he developed type II respiratory failure despite BiPAP support. Tracheostomy was finally performed. He stayed in the PICU for 16 months and lost 53 kg with aggressive control of diet intake. Yet he remained bed-bound and still required nocturnal BiPAP via tracheostomy. He was also on Ramipril for hypertension; and Metformin and Exenatide for DM. As his family could not take care of him at home, he was subsequently referred to our rehabilitation unit for long term in-patient care.

His Body Weight (BW) was 92 kg (BMI 42 kg/m²) on admission. Baseline evaluation showed that he was only able to walk with one hand supported for 250 m in 6 min due to physical deconditioning.
His lung function was significant impaired. His Forced Vital Capacity (FVC) was 0.55L (19% predicted) and Peak Expiratory Flow (PEF) was 66.6 L/min (18% predicted).

An inter-disciplinary program involving respiratory pediatrician, rehabilitation pediatrician, pediatric endocrinologist, dietician, physiotherapist, Clinical Psychologist (CP), hospital school teacher and nurses was implemented. Treatment strategies included a specially-designed low caloric diet (1200 kcal/day), environmental restriction of food, supervised mandatory exercise with positive reinforcement and visual schedule to maintain compliance to daily routine. Initially, he had frequent temper tantrums, pulling out his own tracheostomy tube, destroying the properties of other patients, hitting himself and harming others when he struggled from being restrained. Functional analysis by CP revealed that the triggers were related to non-scheduled food access, change of routine and lack of sufficient social stimulation. Subsequently, CP designed behavioral modification program coupled with award system and instructed nursing colleagues and personal care assistants to implement distant observation towards aggressive behavior and contingent positive reinforcement towards appropriate behavior. He demonstrated consistent improvement in emotional control, and enjoyed learning activities arranged by hospital school teacher. His body weight steadily decreased to 77 kg (BMI 36 kg/m²) after 6 months. Intensity of physical exercise was titrated up against his lung function. He was able to achieve 398 m in the 6-min walking test at 7 months. His lung function improved with FVC 1.48 L (46% predicted) and PEF 100.2 L/min (34% predicted). At 10 months, he weaned off ventilator support and tracheostomy. Overnight monitoring showed that there was no significant oxygen desaturation and transcutaneous carbon dioxide level was not elevated throughout. His blood sugar profile became more stable. Metformin was weaned to a lower dose at 1 month and Exenatide taken off at 9 months. Caregiver training including setting of meal schedule, designing visual cues for enhancement of communication and behavioral modification skills to parents and school staff was started at 12 months to ensure continuity of the systematic nutritional and behavioral program at the community. He was discharged to a special boarding school with BW of 65 kg (BMI 30.5 kg/m²) at 13 months. His BW was 64.2 kg at 1 month after discharge. He coped well at school with satisfactory glycemic profile.

Results and Discussion

PWS is a multisystem genetic disorder caused by absent expression of paternally inherited imprinted genes on chromosome 15q 11-13 [2]. It is characterized by hypotonia in infancy, followed by early-onset hyperphagia and development of obesity in childhood, as well as short stature, hypogonadism, developmental challenges and behavioral problems. Deaths in PWS adults are most often due to obesity-related cardiorespiratory failure, cor pulmonale exacerbated by obstructive and central apnea, as well as septicemia due to skin infections [3]. Weight control is particularly difficult in PWS due to multiple factors but our subject demonstrated that it is not infeasible. Firstly, PWS patients have neuroanatomical abnormalities in the hypothalamus and elevated level of orexigenic hormone ghrelin that lead to impaired satiety, thus resulting in relentless food seeking behavior. Orlistat, the only FDA approved pharmacological treatment for pediatric obesity, cannot tackle the hyperphagic state. Restrictive bariatric surgeries have not been shown to reduce hyperphagia and achieve long-term weight control [4]. Exenatide, a Glucacon-Like Peptide 1 (GLP-1) receptor agonist is useful in hypothalamic obesity as it acts on centrally located GLP-1 receptors to suppress appetite and increase energy expenditure [5]. It has been shown to result in weight loss in obese adults with and without type 2 DM [6,7]. A recent longitudinal study showed that exenatide led to decreased appetite in youths with PWS, without change in weight or BMI in the short term and its side effects were well tolerated [8]. It was added to metformin in our subject with apparent effect in controlling his appetite. It was later taken off as his body weight was greatly reduced. Secondly, PWS patients have limited coping mechanisms due to cognitive rigidity, variability in adaptive functioning and sensitivity to stress, apart from aggressive outbursts related to food acquisition [9]. Thus, the success of usual weight reduction program is extremely low. Likewise, our case exhibited a lot of tantrums in the referring hospital and the initial days at our rehabilitation unit. After functional analysis, it was noted that changes in daily routine were the most frequent triggers for temper outbursts in our subject. Thus, our behavioral modification program emphasized on predictability, consistency and stability of the environment. We formulated a consistent daily schedule incorporating fixed meal time to reduce unpredictability. Encouragement and descriptive praising were given to reinforce positive behavior in all settings and systemic reward system was implemented to encourage non-aggressive emotional coping. We used social engagement, such as collaborative art work as a form of reward as our subject had unfulfilled social need due to prolonged hospitalization. We also avoided using food as reward to prevent disappointment and frustration for not getting extra food. Anger management skills were also practiced regularly to enhance patient’s emotion regulation skills in face of stress. All these techniques were consistently employed by all the therapists and health care workers. After implementation of this strict behavioral modification program, episodes of emotional outbursts in our subject steadily decreased and he became more compliant to diet restriction and physical training. Thirdly, patients with morbid obesity have impairment of resting lung volumes and gas exchange, which become attenuated during exercise when oxygen cost of breathing and dyspnea increase. Respiratory muscle function is also impaired, inspiratory muscle strength is reduced and respiratory drive is increased. Oxygen uptake at a given workload is increased and maximal workload is reduced [10]. Hence, weight reduction program involving vigorous physical exercises may fail if it is not geared to the cardiorespiratory function of the patient. Our case was further complicated by physical deconditioning and limited mobility due to prolonged PICU stay. Personalized program was designed based on assessment on flexibility, strength, BMI, motor function, physical fitness, pulmonary function and compliance. Intensity of physical activity was gradually titrated up using Frequency, Intensity, Type and Time (FITT) principle [11]. It was tolerated well by our subject and contributed to his weight reduction.

Conclusion

Our subject is the first reported case of PWS that can successfully wean off long term ventilator support. This shows that a well-structured inter-disciplinary program in a rehabilitation setting can address the unique medical and psychiatric needs of morbidly obese PWS patients. We believe that the concept of the program may also be applied to other patients with morbid obesity who fail to comply to weight reduction measures.

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References


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