WWW.IJRPR.COM

International Journal of Research Publication and Reviews

Journal homepage: www.ijrpr.com ISSN 2582-7421

Managing Cardiovascular/Pulmonary Needs in Duchenne Muscular Dystrophy: A Case Study of GYAN

Sneha Hiren Bhalala (PT)

Assistant Professor
SPB Physiotherapy College, Surat

Introduction:

Duchenne muscular dystrophy (DMD) is a progressive genetic disorder characterized by muscle weakness and degeneration. GYAN, a 14-year-old Indian boy, was diagnosed with DMD at the age of 4. This case study focuses on the cardiovascular/pulmonary needs of GYAN, highlighting the challenges and interventions in managing his condition.

GYAN's medical history reveals a progressive decline in muscle function, with classical physical characteristics such as severe lumbar lordosis and multiple contractures affecting his mobility. Despite his deterioration, GYAN remained ambulatory until recently when he transitioned to using a power wheelchair due to increasing weakness and limited mobility.

Examination of GYAN's cardiovascular/pulmonary status reveals normal vital signs but compromised respiratory muscle strength. His ability to perform essential tasks like running, ascending or descending stairs, and rising from the floor or a chair is severely impaired. Tests and measurements indicate weakened respiratory muscles and limited chest wall expansion, contributing to ineffective coughing and ventilation.

Based on the evaluation, GYAN's respiratory dysfunction is classified as cardiovascular/pulmonary pattern 6E ventilatory pump dysfunction or failure. His prognosis includes 8 to 10 weeks of physical therapy focusing on respiratory muscle strengthening and techniques to improve coughing and ventilation. The intervention plan involves coordination with the muscular dystrophy centre, patient and caregiver education on home exercise programs, and therapeutic interventions such as the active cycle of breathing, inspiratory and expiratory muscle strengthening, and assisted cough techniques.

This case study underscores the importance of comprehensive management of cardiovascular/pulmonary needs in individuals with DMD. Through targeted interventions and collaboration between healthcare professionals and caregivers, GYAN's quality of life can be optimized despite the challenges posed by his condition.

Case Description:

GYAN, a 14-year-old Indian boy with a history of Duchenne muscular dystrophy diagnosed at 4 years of age, was referred for pulmonary physical therapy evaluation. This case study will focus on the cardiovascular/pulmonary needs of this patient.

Examination HISTORY

GYAN was a full-term infant who appeared to be developing normally until approximately 3 years of age when his parents noted that he had difficulty rising easily from the floor and could not easily ascend stairs. Upon stating their concerns to the paediatrician, GYAN was sent for laboratory testing and a muscle biopsy. The testing indicated abnormally high serum creatinine kinase and a muscle biopsy was performed. The resulting diagnosis of Duchenne muscular dystrophy was made. He was referred to a large children's hospital for follow-up and continuing care. As the years progressed, GYAN's weakness became more pronounced, and he developed some of the classical physical characteristics including increasingly severe lumbar lordosis, several contractures including plantar flexion, and hip and knee flexion contractures. He was still ambulatory, although he and his mother reported that the distance was steadily decreasing and the speed was getting slower.

REVIEW OF SYSTEMS

Integument: There were no obvious or stated problems in this area. Musculoskeletal: Numerous contractures in the lower extremities were obvious along with severe lumbar lordosis. Elbow contractures were also noted, although not as severe as those of the lower extremities. There was obvious weakness, particularly in the shoulders and hip musculature. Neuromuscular: Ambulation, balance, and transfers were all significantly abnormal and limited. GYAN currently functions from a power wheelchair. Cardiovascular/Pulmonary: Heart rate, respiratory rate, and blood pressure were all within normal limits. Communication, affect, cognition, language, and learning style appeared unimpaired.

TESTS AND MEASUREMENTS

General Summary of Function: At the time of his referral, an examination of overall physical function revealed that GYAN could ambulate 25 feet in 20 seconds, roll from a prone to a supine position and back to a prone position, and had adequate sitting balance. He was unable to run, ascend or descend stairs, rise from the floor or a chair, sit up from a supine position, or assume a posture on all fours. A modified manual muscle examination indicated strength that was graded from "poor" to "absent" for all isolated muscle groups, except for wrist extensors, which were graded as "fair" to "good." GYAN could function in an electric wheelchair, and he could ambulate slowly using a walker with supervision. Ventilation, Respiration/Gas Exchange: GYAN's breathing pattern was examined and was found to be a diaphragmatic pattern with appropriate intercostal muscle use while at rest and during exertion; his accessory respiratory muscles became active during inspiration and expiration. His respiratory muscle strength was measured using maximum static inspiratory pressure (MSIP), inspiratory capacity (IC), and slow inspiratory vital capacity (IVC). The MSIP was 60% of predicted values; the IC was 45% of predicted values; and the IVC was 45% of predicted values. His maximal static expiratory pressure, a measure of expiratory muscle strength, was 35% of the predicted values. Chest wall expansion was determined with a tape measure and found to be approximately 2.5 cm at maximal inspiratory effort. Passive motion was adequate at the glenohumeral joints. Coughing was evaluated as being weak and questionably functional. As previously noted, his limited IC and reduced expiratory pressures were largely responsible for the impaired cough. Evaluation, Diagnosis, and Prognosis Based on the data gathered specific to his current pulmonary issues, GYAN would receive a classification at cardiovascular/ pulmonary pattern 6E ventilatory pump dysfunction or failure. This would be a pattern specific to his respiratory muscle impairment for which he was referred. His prognosis would be 8 to 10 weeks of care with episodes of physical therapy two times per week for the first several weeks and reduced to weekly thereafter. Interventions Coordination, communication, and documentation would focus on interaction with the muscular dystrophy centre at the children's hospital where GYAN is followed. Patient instruction would focus on instruction of GYAN and his parents and other caregivers regarding a home exercise program to improve his respiratory muscle strength, enhance his coughing, and provide for AC as needed. In addition, they would be trained in proper AC and assisted cough techniques, including various devices to support AC.

PROCEDURAL INTERVENTIONS

Therapeutic Exercise:

Active cycle of breathing to enhance diaphragmatic excursion, maximal inspiration, and FET to aid in improving ventilation to the lower lobes, maintain/improve chest expansion, and aid in secretion removal. Continuing with ongoing strength training and range-of-motion exercise with emphasis on the thorax and shoulder girdle to maintain thoracic compliance. Inspiratory muscle and expiratory muscle strengthening using simple handheld devices to maintain/increase respiratory muscle strength. AC as needed using high-frequency chest wall oscillation with SmartVest or airway oscillation with either Acapella or Flutter device; should secretion removal become problematic, equipment for airway aspiration or insufflation–exsufflation recommended.

References:

1) Eagle, M., Bourke, J., Bullock, R., Gibson, M., Mehta, J., Giddings, D., ... & Bushby, K. (2002). Managing Duchenne muscular dystrophy–the additive effect of spinal surgery and home nocturnal ventilation in improving survival. Neuromuscular Disorders, 12(6), 621-625. https://doi.org/10.1016/S0960-8966(02)00007-9

2) LoMauro, A., Romei, M., Gandossini, S., Pascuzzo, R., Vantini, S., D'Angelo, M. G., ... & Aliverti, A. (2016). Evolution of respiratory function in Duchenne muscular dystrophy from childhood to adulthood. European Respiratory Journal, 48(5), 1508-1517. https://doi.org/10.1183/13993003.00557-2016

3) Finder, J. D., & Birnkrant, D. (2018). Management of Duchenne muscular dystrophy in the United States. Pediatrics, 142(Supplement 2), S86-S92. https://doi.org/10.1542/peds.2018-1095E