Early rehabilitation of a child with intensive care unit acquired weakness secondary to membranoproliferative glomerulonephritis: A case report

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Received: 8 September 2014, Revised: 6 January 2015, Accepted: 26 January 2015

SUMMARY: Coşkun-Benlidayı İ, Başaran S, Gül-Mert G, Güzel R. Early rehabilitation of a child with intensive care unit acquired weakness secondary to membranoproliferative glomerulonephritis: A case report. Turk J Pediatr 2015; 57: 422-425.

Intensive care unit acquired weakness presents with flaccid paralysis of the extremities and difficulty of weaning from the ventilator and it has occasionally been reported in children. We report the early rehabilitation of a 12-year-old girl with membranoproliferative glomerulonephritis who developed intensive care unit acquired weakness. She underwent an intensive rehabilitation procedure which included assistive range of motion, bed mobility, airway clearance and breathing techniques, neuromuscular electrical stimulation (NMES) to the quadriceps muscles and resistive therapeutic band exercises. Following the rehabilitation program, muscle strength (Medical Research Council sum score), ambulation (Functional Ambulation Category) and activity (Wee FIM) scores increased significantly.

Key words: intensive care unit acquired weakness, membranoproliferative glomerulonephritis, neuromuscular electrical stimulation, rehabilitation.

Intensive care unit acquired weakness (ICU-AW) is a complication of critical illness such as sepsis and multiorgan failure that presents with flaccid paralysis of the extremities and usually difficulty to wean from the ventilator. The manifestations of this condition are also discussed under the terms such as "critical illness myopathy", "critical illness polyneuropathy" or "critical illness polyneuromyopathy"[1].

Although it was thought to be a rare complication when first described in 1980s, recent data reveals that it affects up to 60% of critically ill adult cases[2-4]. ICU-AW can cause severe prolonged disability and emerging data demonstrates the safety, feasibility and potential benefit of early mobilization in adult patients[1, 5].

The condition has occasionally been reported in children and there are no reliable data on the incidence in pediatric age group[6, 7]. Banwell et al.[8] identified only 1.7% critical illness polyneuromyopathy cases among 830 patients admitted to a pediatric intensive care unit[8]. Although this may relate to the lower incidence of physical comorbidities in critically ill children, it may also be related to the failure of ascertainment. Sepsis, asthma and organ transplantation have been suggested as potential risk factors in children[7].

In this report the safety of early mobilization and neuromuscular electrical stimulation in a pediatric case of ICU-AW is presented.

Case Report

A 12-year-old girl was referred to our hospital due to an ongoing proteinuria for one month. She was diagnosed as dense deposit disease/ membranoproliferative glomerulonephritis by renal biopsy and started on pulse steroids and cyclosporine. She was unresponsive to treatment and on day 77, following plasmapheresis she developed acute respiratory distress syndrome, massive pulmonary hemorrhage and sepsis. Consequently she was put on mechanical ventilation in pediatric intensive care unit (PICU) and sedated. In PICU the first unsuccessful attempt to wean her from the ventilator was

made on day 13, but total weaning was not possible upon many trials until day 42 which she was transitioned to noninvasive ventilation with bi-level positive airway pressure (BIPAP). Meanwhile severe weakness in the limbs was recognized. Electroneuromyography detected no abnormality in sensorial nerve conduction velocities and bilateral motor polyneuropathy was noted. Transcutaneous diaphragmatic electromyography revealed phrenic nerve involvement.

The patient was consulted to the Department of Physical Medicine and Rehabilitation with the problems of difficulty in weaning and weakness. Initial assessment occurred on PICU day 44, she was awake, oriented to person, place and time and receiving BIPAP support. Bedside manual muscle testing Medical Research Council (MRC) sum score was 38 (Table I). In this scoring system the strength of shoulder abduction, elbow flexion, wrist extension, hip flexion, knee extension and ankle dorsiflexion are assessed bilaterally each on a scale of 0 to 5, which when added yield an overall numeric rating between 0-60. The MRC is a valid tool to assess muscle strength. ICU-AW has been inferred based on a sum score of less than 48[1, 9]. Sensation was grossly intact and passive range of motion was full throughout all extremities, bilateral pretibial edema and diffuse striae were present.

On the basis of these findings this critically ill patient was diagnosed with ICU-AW.

Ambulation level was evaluated by using the Functional Ambulation Classification (FAC)[10]. FAC is evaluated in a total of 6 categories from 0 to 5. The FAC category of the patient was 0, which reflects no ambulation.

The functional independence of the patient was assessed according to the functional independence measure for children (WeeFIM)[11, 12]. The scores of the 3 domain subscales (self-care, mobility and cognition) of the patient were 10, 5 and 15, respectively with a total WeeFIM score of 30 (Table I). The WeeFIM instrument contains 18 measurement items, divided into areas of self-care (eating, grooming, bathing, dressing upper body, dressing lower body, toileting, bowel management, bladder management), mobility (bed/chair/wheelchair transfer, toilet transfer, tub/shower transfer, walking/wheelchair, stair climbing) and cognition (comprehension, expression, social interaction, problem solving, memory). A seven-level ordinal scoring system ranging from 7 (complete independence) to 1 (total assistance) is used to rate each item. The minimum possible total score is 18 (total dependence in all skills); the maximum possible score is 126 (complete independence in all skills). WeeFIM is applicable to children older than 7 years with developmental or acquired disabilities[12].

Twice-daily physical therapy sessions of 30 minutes as tolerated were planned and sedation was kept to minimum. Interventions begin as active assisted range of motion and bed mobility including rolling/ supine to sit and air way clearance via percussion/ vibration, active cycling of breathing techniques. The next day, patient tolerated to sit unsupported at the edge of the bed for 10 minutes and progressed to ambulation with the assistance of two staff members, requiring tactile stimulation to prevent knee buckling. By day 48 she was breathing through an oxygen mask with a reservoir and ambulating 6 m with 1 rest break. She started resistive exercises to upper and lower extremity muscles with light resistance elastic bands.

On day 50, she was transferred to pediatric ward. After then physical therapy sessions continued in physical therapy department. Neuromuscular electrical stimulation (NMES) with Russian stimulation to quadriceps muscle was started[13]. This is a 2.5 kHz alternating current applied in 10 milliseconds rectangular bursts at a frequency of 50-Hz, with a duty cycle of 50%, 10 minutes daily treatment sessions were applied (Chattanooga Intelect Advanced 2773MS®). At each NMES session at the physiotherapy department arterial blood pressure, heart rate and respiratory rate were assessed initially and at 5, 10 and 15 minutes. NMES did not cause any hemodynamic instability. The stimulation was safe and free of pain. She was having gradually increasing neuromotor exercises (including strengthening exercises for the abdominal and spinal extensor muscles) which were, in turn, followed by tiptoe and heel stance, march on the spot and gait exercises. After 10 sessions of NMES and slowly progressive rehabilitation program, the MRC sum score improved to 50. When she was discharged on day 62, she could ambulate

Table I. Physical Therapy Outcomes of the Patient

	First assessment (Day 44)	Last assessment (Day 62)
MRC sum score (0-60)	38	50
FAC (0-5)	0	4
Wee FIM		
Self care (8-56)	10	34
Mobility (5-35)	5	24
Cognition (5-35)	15	28
Total (18-126)	30	86

MRC: Medical Research Council, FAC: Functional Ambulation Category, FIM: Functional Independence Measure
Score ranges are given in parenthesis.

independently on level surfaces but required physical assistance to negotiate non-level surfaces and stairs (FAC 4). The total WeeFIM score of the patient was improved to 86 with scores of 34, 24 and 28 for self-care, mobility and cognition subscales, respectively (Table I).

Unfortunately, 15 weeks later she had a subsequent hospital course of 38 days, two days being in the PICU without the need for mechanical ventilation. On follow up 10 months after the disease onset, she was still mobile, moderately independent in activities of daily living and on hemodialysis three days a week.

Discussion

Despite the high incidence in adults, ICU-AW is relatively uncommon in childhood and up until 2007 only 43 children have been reported. Their median age was 11.5 years and 86% of these children have been ventilated for >5 days[7]. Compatible with the previous case reports our case was a 12 year girl who has been ventilated for 42 days.

The incidence, etiology, natural history, and prognosis of the condition in children are unknown[7]. Diagnosis of ICU-AW depends upon the history, physical examination and electrophysiological findings. Current guidelines for ICU physiotherapy suggest that physiotherapy assessments should not be driven by medical diagnoses, but physiological and functional deficiencies[4]. Recently, evidence has been provided for appropriate measurement approaches in ICU-AW in adults. Among those measures for the examination of body function/structure the usage of MRC sum score and for evaluation of activity FIM based scoring have been proposed[1]. In this pediatric case we have evaluated the outcome using these measures.

Physical rehabilitation for individuals with ICU-AW can begin as soon as they have sufficient medical stability to accommodate the increased vascular and oxygen demands that accompany the physical examination and intervention[1]. Managements tended to keep the period of sedation to a minimum including daily cessation of sedation and ambulation of the patients on mechanic ventilators via physical therapy, and rehabilitation can bring about favorable outcomes[14]. Rehabilitation is the fundamental element of ICU-AW management[5]. Early physical therapy not only shortens the hospitalization period but also reduces the readmissions[1, 2, 5]. The intervention techniques include respiratory strategies, range of motion exercises and functional mobility training[1]. Regarding the functional mobility, there are several early mobility procedures in the literature[1, 2, 4, 6, 15, 16]. Sitting on the edge of the bed, standing at bedside and sitting in chair, and walking a short distance are the levels of a three-step process which the patients experience and the rehabilitation of this case has also progressed in that way. It is reasonable to commence with the highest level they can tolerate. As the patients with ICU-AW present with loss of large amounts of muscle mass, electrotherapy can preserve muscle mass in critically ill patients and can be an alternative to active exercises[17, 18].

The present case was managed with a physiotherapy program comprising breathing exercises, active-assistive range of motion and resistive exercises for both upper and

lower extremities. In addition, NMES to quadriceps muscles was performed. The patient experienced a satisfying recovery in terms of the ability of performing daily activities and ambulation.

Accumulating prospective trial evidence supports that early rehabilitation and mobilization in critically ill adults is safe, feasible, cost effective, and improves short-term patient outcomes[4]. In contrast there is paucity of this research in pediatrics and except for case reports, early mobilization has not been evaluated in critically ill children[4, 15]. Prospective series are required to establish the incidence and natural history of ICU-AW in childhood. Standardized clinical, neurophysiologic and pathologic criteria would enable development of a classification system[7]. This case report supports that early mobilization and NMES are safe and effective approaches in childhood ICU-AW. It is of great importance for physicians to be familiar with ICU-AW in pediatric critically ill cases and determine the best rehabilitative approaches.

Acknowledgements

We would like to thank to physiotherapist Yıldız Şahin for her contribution on the rehabilitation process of the patient.

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