Introduction

Fukuyama congenital muscular dystrophy is a hereditary muscular disease and is known to cause restrictive ventilatory defect because of progressive spinal deformity and weakened respiratory muscles, resulting in respiratory failure\(^1\). For patients with such a respiratory disorder who are placed on a ventilator, respiratory care is difficult to provide in the general ward. At our hospital, a respiratory support team (RST) is continuously involved in mechanical ventilation management of patients from the intensive care unit (ICU) to the general ward and provides respiratory care in the ward. In this report, we describe our experience with a patient with Fukuyama muscular dystrophy to whom the RST provided adequate respiratory care.

Case

A 16-year-old man with Fukuyama congenital muscular dystrophy experienced airway obstruction due to sputum, and his face became pale. He stopped breathing, and his radial artery could not be palpated. Hence, an ambulance was called. His mother performed bystander cardiopulmonary resuscitation. Spontaneous heartbeat resumed during transport to a hospital. His consciousness level upon arrival to the hospital was 300 on the Japan Coma Scale, and no spontaneous respiration was observed. The patient was transferred to the nearest critical care center, where intratracheal intubation was performed. Subsequently, he was immediately transferred for further treatment to our hospital where he had been regularly treated.

Vital signs: consciousness level, E1VtM1 on Glasgow Coma Scale and 300 on Japan Coma Scale; respiratory rate, 12/min; SpO\(_2\) 99% (with ventilator); blood pressure, 98/67 mmHg; heart rate, 83/min; and body temperature, 36.2°C

Hematological findings: WBC 20,660/µL, Hb 13.6 g/dL, Ht 43.0%, Plt 242,000/µL, ALT 88 IU/L, AST 46 IU/L, LDH 458 IU/L, T-Bil 0.7 mg/dL, CK 3,838 IU/L, BUN 9 mg/dL, Cre 0.1 mg/dL, Glu 234 mg/dL, Na 140 mEq/L, K 4.1 mEq/L, CRP 0.47 mg/dL, PT-INR 1.39, APTT 89.66 sec, FDP 19.6 µg/mL, Lactate 24.1 mEq/L

Blood gas analysis: Ventilator (Volume Control mode) pH 7.27, PaO\(_2\) 248 mmHg, PaCO\(_2\) 59.9 mmHg, HCO\(_3^-\) 23.2 mEq/L, BE -1.6 mEq/L, SatO\(_2\) 99.2%

Imaging finding: Plain chest radiography (Fig.1) revealed hypolucency in the right lung field. Contrast-enhanced chest computed tomography (Fig.2) revealed infiltration in the right lung field, suggesting pneumonia. The thorax and spine were markedly deformed, and the right main bronchus was compressed.

Clinical course

After the patient was admitted to the intensive care unit (ICU), targeted temperature management was performed for postresuscitation encephalopathy. As his consciousness level gradually improved, he became able to...
communicate on hospital day 14. Because we assumed that weaning from mechanical ventilation was difficult because of thoracic deformity and the weakened respiratory muscles, tracheostomy was performed. On hospital day 19, because his general conditions were stabilized, he was transferred from the ICU to the general ward of the neurology department. Because the hospital to which he would be transferred was not always equipped with any ventilator, Trilogy 100 Plus® (Philips), a home-care ventilator, was connected to him. Because staff members in the general ward were not accustomed to care of patients placed on a ventilator, the RST was actively involved in his care.

Even after he was transferred to the general ward, there were several in-hospital emergency events in which induced sputum caused airway obstruction, followed by hypoxia. However, an emergency department physician who belonged in the RST and continuously involved in his care shared the problems specific to the patient and treatment strategies with the ward staff, such that the patient was properly treated.

In the following section, we present how the RST engaged in the care of the patient in the general ward. (1) Because the intubation tube had been bent leftward, the RST adjusted the place for installing a ventilator and the length and direction of the circuit to prevent the tracheostomy tube from being pulled (Fig.3). (2) After HAMILTON-C2® (Hamilton Medical, Switzerland) was replaced with Trilogy 100 Plus®, the patient complained of respiratory discomfort despite adequate oxygenation and ventilation. As no cause was identified, the RST intervened. When the passive breathing circuit open to atmosphere was switched to the closed active breathing circuit, respiratory discomfort was relieved (Fig.4). Because the active breathing circuit is a closed circuit equipped with an exhalation valve, we assumed that maintenance of consistent positive end-expiratory pressure reduced fatigue of the respiratory muscles. (3) Because the tracheostomy tube was fixed with a string, the string dug into the neck, almost causing a pressure ulcer. It was replaced with a tube holder made of skin-friendly materials (Fig.5). (4) Because skin ulceration was noted around the tracheostomy fistula, the RST recommended consultation with a dermatologist, and the dermatologist prescribed Glymesason and zinc oxide ointments. (5) Because the trachea was compressed by thoracic deformity, even slight displacement of the tracheostomy tube was likely to cause airway stenosis. Thus, the RST presented the observation points for tube management (Fig.6), instructed the ward staff to make evidence of observation visible for record purposes, and examined the patient with the staff during RST rounds. Through the aforementioned respiratory care, the patient was well managed on a ventilator and was transferred to another hospital on hospital day 183.

Discussion

Fukuyama congenital muscular dystrophy is an autosomal recessive disease first reported by Fukuyama et al. in 1960. It is the second most common type of progressive muscular dystrophy after the Duchenne type. The Fukuyama type is caused by mutations of the responsible gene, fukutin (FKTN)\(^2\). Although the annual incidence is 2 to 4 per 100,000 people, this disease is characteristically common in the Japanese. Many patients exhibit moderate to severe mental retardation, and the disease is complicated by convulsion in more than 60% of patients. Joint contracture appears in the early stages. As the disease progresses, plantar flexion of the foot joint and extension of the hip, knee, and elbow joints become limited\(^3\). Prognosis depends on the degrees of respiratory failure due to weakened respiratory muscles and cardiac failure due to dilated cardiomyopathy. The mean life...
span is in the late teens. In Fukuyama congenital muscular dystrophy, factors associated with respiratory dysfunction include spinal deformity and degenerative atrophy of the respiratory muscles, both of which are frequently observed. Miyoshi et al. reported that treatment of chronic respiratory failure is important in the terminal stage of Fukuyama congenital muscular dystrophy. According to them, when chronic respiratory failure is noted, non-invasive ventilatory support should first be implemented, and tracheostomy should also be considered. Because good respiratory management affects the prognosis of Fukuyama congenital muscular dystrophy, administration of adequate respiratory care, including mechanical ventilation management, is important for patients with severe respiratory failure, as is our case.

The RST of our hospital is a multidisciplinary team that strives to improve the safety and quality of respiratory care of patients with respiratory dysfunction who are placed on a ventilator regardless of acute or chronic disease. Its tasks include setting of a ventilator, respiratory care of patients placed on a ventilator, checking and maintaining the operation of the ventilator, supporting patients in weaning from mechanical ventilation, and providing respiratory rehabilitation. The team consists of 10 members, including physicians, certified nurses, clinical engineers, and physiotherapists. While the team makes rounds once a week on patients placed on a ventilator in the general wards, it also examines patients upon request of the ward staff. Physicians in the emergency department and ICU engage in intensive care of patients placed on a ventilator who are expected to be later discharged to the general ward. In addition to these physicians, the RST includes pulmonologists, certified nurses, clinical engineers, and physiotherapists, thereby ensuring a system.
Fig. 4  Switch to an active circuit
Switching from a passive circuit to a closed active circuit relieved respiratory discomfort.

Fig. 5  Skin ulceration around the tracheostomy fistula and a tube holder
Treatment of skin ulceration around the tracheostomy fistula (white arrow) and replacement with a tube holder made of skin-friendly materials (black arrow).

Fig. 6  Observation points for tube management
a: Although the tube appears to be not fitted well, this is in the correct position. b: The space is filled with two pieces of Y gauze. Because of abundant expectoration, gauze should frequently be changed to keep the fistula clean. c: The tube should be kept at an angle where it does not compress the upper edge of the tracheostomy fistula. Because of airway deviation, the tube is bent slightly rightward.
to seamlessly support patients even after transfer to the general ward. Our experience suggests that patients requiring respiratory care, like ours, can receive good respiratory management even in the general ward through supports from the RST.

**Conclusion**

In the general ward, staff members are not accustomed to management of patients with respiratory failure, making it difficult to provide respiratory care, including the installation of a ventilator. Involvement of the RST appears to allow adequate respiratory care, including mechanical ventilation management, to be provided even to patients requiring difficult respiratory management like ours, thereby ultimately ensuring clinical safety.

**Conflict of interest**

All authors declare no conflict of interest to disclose.

**Reference**