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Telemedicine for Facio-Scapulo-Humeral Muscular Dystrophy: A multidisciplinary approach to improve quality of life and reduce hospitalization rate?

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ABSTRACT

Background: Facio-Scapulo-Humeral Muscular Dystrophy (FSHD) is an autosomal dominant inherited disorder characterized by a variable and asymmetric involvement of facial, trunk, upper and lower extremity muscles. Although respiratory weakness is a relatively unknown feature of FSHD, it is not rare. Telemedicine has been used in a variety of health care fields, but only recently, with the advent of sophisticated technology, its interest among health professionals became evident, even in such diseases.
Objective: To demonstrate the telemedicine efficacy in FSHD.
Methods: Four siblings affected by a severe form of FSHD, living in a rural area far away from the referral center for neuromuscular diseases, who used a wheelchair, suffered from chronic respiratory failure and were provided with long-term non-invasive mechanical ventilation, received a 6-month period of telemedicine support. This consisted of video conferencing (respiratory physiotherapy, psychological support, neurological and pneumological assessment, nurse-coach supervision) and telemonitoring of cardiorespiratory variables (oxygen saturation, blood pressure, and heart rate).
Results: We performed 540 video conference sessions per patient, including three daily contacts with short monitoring oximetry measurements, blood pressure, and heart-rate measurements, psychological support, neurological and pneumological assessment, nurse-coach supervision.
Conclusions: Our findings indicate that our telemedicine system was user-friendly, efficient for the home treatment of FSHD, and allowed reducing hospital admissions.

Introduction

Telemedicine is an open and constantly evolving science, using telecommunication and information technology to provide health care at distance, as well as the transmission of medical, imaging and health informatics data from one site to another.[1] The aims of telemedicine are: (i) to provide clinical support; (ii) to overcome geographical barriers; (iii) to use various types of innovative information communication technology; and (iv) to improve health outcomes.[2,3] Telemedicine application consists of two basic types: between health professionals and between health professionals and patients.[1] Telemedicine can be performed in an asynchronous mode, using recorded data at different times, or in a synchronous mode, involving individuals in real time for the immediate exchange of information.[4] In both cases, the transmission of the data can consist of text, audio, video, or images.[5,6]

Telemedicine has been efficiently applied in different neurological fields, such as cerebrovascular or neurodegenerative disorders.[7,8] To date, few data are reported on the application of telemedicine for neuromuscular diseases with chronic respiratory insufficiency, mainly amyotrophic lateral sclerosis. It has been shown that the patients managed with telemedicine received the same quality of care and had similar outcomes to those seen via

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traditional face-to-face encounters, thus guaranteeing high-quality tertiary ALS care.[9–13] No data are available regarding telemedicine application in Facio-Scapulo-Humeral Muscular Dystrophy (FSHD). FSHD is an autosomal dominant inherited disease with an asymmetric involvement of muscles in the facial, trunk, upper and lower extremity regions, with variable severity.[14] FSHD is the third most common form of muscular dystrophy. Patients with FSHD may experience respiratory failure because of a progressive weakness of respiratory muscles and scoliosis. Usually, chronic respiratory failure occurs when the patient complains of other signs of severe functional impairment, such as using a wheelchair.[15]

This study aims at describing a telemedicine system based on video conferencing and telemonitoring of cardiorespiratory variables (oxygen saturation, blood pressure, and heart rate) for the telecare of four siblings affected by FSHD with chronic respiratory failure, living in a rural area far away from the referral center for neuromuscular disorders.

Methods

The present study involved four siblings affected by a severe form of FSHD with chronic respiratory failure treated with long-term non-invasive mechanical ventilation, who used a wheelchair and lived in a rural area far away from the referral center for neuromuscular diseases. They received a 6-month period of telemedicine support based on video-conferencing (psychological support, neurological and pneumological assessment, nurse-coach monitoring for device use) and telemonitoring of cardiorespiratory variables (oxygen saturation, blood pressure, and heart rate), and telerehabilitation (respiratory physiotherapy). The duration of this experimental telemedicine protocol was set at six months, and a follow-up hospital admission was planned to assess the obtained results.

We applied a Telemedicine System that was modified to monitor patients' needs. The patient could interact with the system at any time by accessing a simple menu on the mobile phone. To monitor cardio-respiratory parameters, we used a pulse oximeter (Nonin Onyx® 9500) and an aneroid sphygmomanometer (BOSO). Within the system architecture, the Control Center, the Application Server, and the Database Manager were located at the Hospital Center. The system allowed a Telecare Service Center to perform remote monitoring of biological signals, video conferencing, and to handle emergencies that may arise. An administrator handled database, user privileges, communications, and security from the Hospital. A system firewall protected communications between client and server against unauthorized access. In addition, the system also had a mechanism for user authentication and a password to check the role of the connecting person. The engineers-team visited the patient at home periodically, performing all regular procedures, and checking the equipment by testing all medical and communication devices. Any malfunction was reported to the supervising physician. The local Ethics Committee approved the study and the patients gave their written informed consent to study publication.

Telemedicine protocol

Depending on the severity of the patient's condition, two different telecare protocols were followed. If the patient's condition was stable, cardio-respiratory parameters were carried out three times a day. The patient sent the data to the telemedicine center, where a skilled operator received these; the neurologist or the pulmonologist (who were on call 24/7) were called for consultation in case of alerts. If the patient's condition worsened, the oxygen saturation was monitored overnight, and reported to the physician the next morning. If the measurements were abnormal, new instructions were given to the patient's family by the physician. Other videoconferences were carried out, according to the following schedule: psychological consultation one time a week (about 30-min per session); body mass index (BMI) assessment monthly to monitor the assigned diet (about 15-min per session); cardiorespiratory rehabilitation twice a week (about 40-min per session); neurological and pneumological consultations when required or in emergency.

For the psychological aspect, we also administered at baseline and after the end of the telemedicine protocol the following scales: 36-Item Short Form Survey (SF-36) to investigate changes in quality of life in eight fields (Physical Functioning, Limitations Due To Physical Problems, Limitations due to Emotional Problems, Pain, General Health, Vitality, Social Functioning, Mental Health); and the Hospital Anxiety and Depression Scale (HADS) to evaluate mood and emotional state. The Psychosocial Impact of Assistive Devices Scale (PIADS) and the Caregiver Burden Inventory (CBI) were administered to verify the effect of telemedicine service on some Psychosocial features (i.e., ability, adaptability and self-esteem) and to evaluate the degree of the perceived caregiver burden, respectively.[16–20]

The Cardiorespiratory Rehabilitation was performed through a Virtual Reality Rehabilitation System (VRRS) (Khymeia; Padova, Italy), applying biofeedback exercises for monitoring the respiratory rate and the inhalation and exhalation phases. This module requires a spirometer and the Khymeia dedicated cardiorespiratory monitor, in order to perform the exercises. The advantage of the VRRS consisted of the use of a virtual environment that helped the patients to develop knowledge of the results and the performances. Caregivers required only 4 h of training to learn how to use the system.

Results

Patients were assessed by a neurologist and a pulmonologist before their inclusion in the study. Over a 6-months period, we analysed a series of variables including the use of the system, the satisfaction of the patient, and the clinical impact. Overall, we performed 540 videoconferences per patient, including three daily contacts with short monitoring of oximetry, blood pressure, and heart rate. Additionally, psychological support, neurological and pneumological assessment, respiratory physiotherapist using the VRRS system, and nurse-coach supervision were provided. The system was used on a continual basis. Vital parameters measurements appeared on the TV screen, and were viewed in the Control Center. The hospital alarm was activated through system 45 times, but the events (i.e., low blood/high blood pressure or heart rate or desaturation) were not clinically relevant. The personal information provided by the patients, together with data from oximetry readings, made it possible to identify an acute event managed with the help of the neurologist from the Control Center, which avoided the tracheotomy for patient n.2. Moreover, there were 20 exacerbations associated with infectious disease, requiring pharmacological therapy but not hospital admission. With respect to the clinical impact, after enrolment in the telemedicine program, the total number of hospital admissions for check-up dropped.

As illustrated in Fig. 1, we found a mild improvement in mood and emotional status of all the patients. Only one patient showed significant reduction of depression and anxiety level as per HADS, while three patients reported positive improvement in “mental health” sub-item of SF36 after six months (Fig. 1). BMI remained stable. In addition, PIADDS′ scores revealed higher scores in “ability”, indicating better skills to face problems. On the contrary, telemedicine service did not produce any change in CBI.

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