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The application of telemedicine in the follow-up of lung transplantation in a patient with cystic fibrosis

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Abstract

The Cystic Fibrosis (CF) Unit of Children's Hospital Bambino Gesù in Rome (Italy) has more than 25 years of experience in diagnosis and treatment of Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) related diseases. The CF Unit actively collaborates with the Transplantation Division for the follow-up of patients with CF who undergo lung transplantation, performed in our Unit since 1991. We present the case of a 19 year-old girl with CF, with severe respiratory failure for which it was subjected to two lung transplant. During the follow-up the remote monitoring has allowed the identification of an early episode of pulmonary relapse and graft-versus-host reaction even before the onset of symptoms, allowing an effective intervention and a complete recovery of lung function. ***Clin Ter 2014; 165(5):e382-383. doi: 10.7417/CT.2014.1769***

Key words: *cystic fibrosis, lung transplantation, telemedicine, telemonitoring*

Introduction

The Cystic Fibrosis Unit of Children's Hospital Bambino Gesù in Rome has more than 25 years of experience in diagnosis and treatment of Cystic Fibrosis (CF) patients. It represents a support centre in central and southern Italy for critical cases and for Cystic Fibrosis Transmembrane Regulator (CFTR) related diseases.

The CF Unit actively collaborates with the Transplantation Division for the follow-up of CF patients who undergo lung transplantation, performed in our Unit since 1991, and it is a reference centre for lung transplantation for pediatric and young adults patients in Italy.

In the CF Unit at Children's Hospital Bambino Gesù in Rome, THC was introduced in 2001 for the homecare of patients with the aim to early recognize the relapse phases of pulmonary infections.

In the past years, we used, for Telemonitoring, mostly instrumentation able to send information about respiratory parameters (Oxitel®, Spirotel®): spirometry with determination and recording of FVC, FEV1, FEV1%, PEF, FEF 25-75, FET, flow-volume and volume-time curves; pulse oximetry with registration of oxygen saturation (SaO_2) and heart rate also in 24 hours recordings with regard to desaturation events.

Recently, we started using a new equipment that, in addition to the parameters of respiratory function, it also provides information on other organs and systems, useful for monitoring other aspects of chronic multi organ, especially in special situations such as pregnancy and lung transplantation, or monitor other concomitant chronic diseases, such as hypertension.

The CF diagnosis is performed in our FC Centre using the sweat test and the CFTR Gene Mutation Analysis according to current criteria (1).

Parents are in charge of monitoring, managing, and supporting a complex home-based treatment. A decrease of FEV1 >10% is considered a significant sign of infectious pulmonary relapse (2).

The treatment of CF is performed by a multidisciplinary team, having a specific experience in this disease. In our Unit, more than 200 patients are followed and, on the average, 15 new diagnoses are performed every year.

For the follow-up at a distance, we currently use an instrumentation called the Intel® Health Guide, a complete telemedicine device of the latest generation. This is a monitoring solution that integrates a remote device for home care, the Intel® Health Guide PHS6000 with the Intel® Health Care Management Suite, an online interface that allows clinicians to monitor patients and manage their care at a distance. In a recent study we described and discussed the workflow we use in the daily practice, developed and improved over the years (3).

Case report

S.D., 19 year-old, female, Cystic Fibrosis (G542X/N1303K) and severe respiratory and pancreatic insufficiency. Chronic pulmonary colonization with Pseudomonas aeruginosa, Staphylococcus aureus and Stenotrophomonast maltophilia. Clinical course characterized by low average

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FEV1 (33% of expected) and frequent episodes of respiratory exacerbation with Oxygen-dependence with repeated cycles of intravenous antibiotic therapy. One year ago, evidence of atypical mycobacteria in sputum for which was carried out specific therapy. Chest TC: important and widespread lung injury. During the first year of recordings, in the pre-transplant, we found a number of tests similar to the previous period, an increase of DH compared to hospitalizations. The reason for this change was the early recognition of exacerbations, which has prevented hospitalizations by i.v. antibiotic therapy. In May 2013, the patient has undergone dual lung transplantation. We continued the follow-up with the remote monitoring.

D. has had during the post-transplant 2 hospitalizations for pulmonary exacerbations, effectively highlighted by early changes. In the first one she had to do a course of intravenous antibiotic therapy. About 3 months after transplantation is detected at home by telemedicine an acute reduction of FEV1 (-13%) in an apparent well-being state (Fig. 1).

In the phone contact following the transmission are reported agitation and dyspnea. Is given indication for hospitalization. In Intensive Care Unit is treated with oxygen therapy, non-invasive ventilation, sedation and antibiotics. Discharged after about 3 weeks with a diagnosis of acute respiratory failure, acute rejection, noncompliance to immunosuppressive therapy, lung transplantation, D. has continued the follow-up at home by telemonitoring.

Discussion

In the lung transplant the main adverse effects are infection and rejection, which often manifests itself in the form of bronchial obstructive pulmonary disease (4).

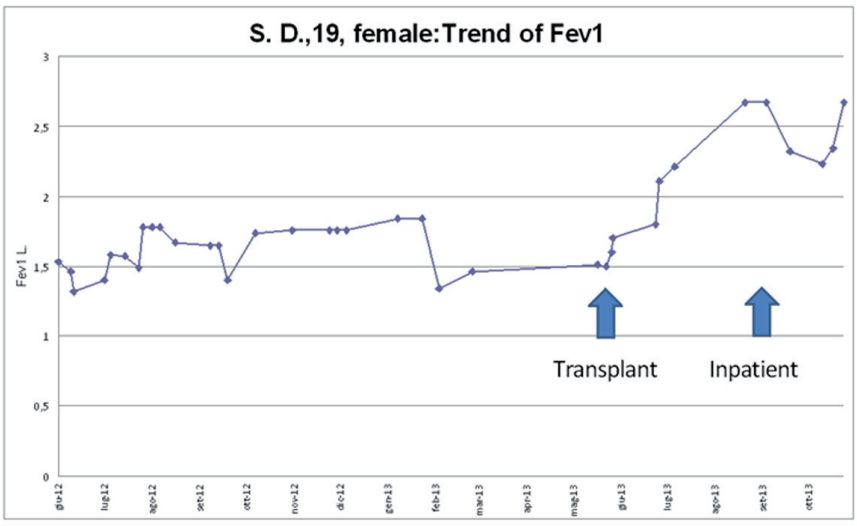

Fig. 1. Trend of Fev1 before and after lung transplantation.

Spirometry, with the determination of FEV1, has long been considered a sensitive method for the systematic search of such complications (5).

Is now universally accepted the recommendation that the follow-up of lung transplant recipients should include the measurement of FEV1 at home by means of a portable spyrometer (6).

In our case the telemonitoring has highlighted early at home a crisis of pulmonary relapse and rejection, allowing medical équipe to make a timely and appropriate therapeutic approach.

Some factors may influence the effectiveness of telemonitoring: first of all understanding by the patient and his collaboration in order to ensure optimal adherence to the recommendations of the treatment team and timeliness to intervene in the presence of significant alterations. Secondly, because the transplant team does not have direct access to the data as a rule, these are not readily available for a daily review (7).

In our case, some data appear significant.

During clinical episode of rejection, the precocity of recognition has avoided the permanent lung damage and therefore the total recovery of lung function until the current value corresponding to approximately 85% of the expected value, as in the pre-transplant period.

Clinical stability during the year preceding the transplant was facilitated by the use of telemedicine, which has also allowed greater sharing of clinical data by the entire treatment team, which is essential for the success of the transplant.

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