MANAGEMENT OF PEDIATRIC PATIENTS IN CYSTIC FIBROSIS WITH TELEMEDICINE

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The Cystic Fibrosis

- Genetic, hereditary and chronic disease, widespread among the Caucasian population, resulting in fatal outcome.

Affects about one child every 3,000 births.

In Italy more than 4,000 people currently affected
# One Way of Classifying CFTR Mutations

**Normal**
- CFTR is created, reaches cell surface and functions properly, allowing transfer of chloride and water.

**Class I**
- CFTR protein is created, but misfolded, keeping it from reaching the cell surface.

**Class II**
- CFTR protein is created and reaches cell surface, but does not function properly.

**Class III**
- The opening in the CFTR protein ion channel is faulty.

**Class IV**
- CFTR is created in insufficient quantities.

### Examples
- **Normal**: G542X, W1282X, R553X
- **Class I**: F508del, N1303K, I507del
- **Class II**: G551D, S549N, V520F
- **Class III**: R117H, D1152H, R347P
- **Class IV**: 3849+10kbC->T, 2789+5G->A, A455E
The Cystic Fibrosis

Natural history:
- recurrent episodes of respiratory infection
- progressive pulmonary damage
- decay of long-term lung function leading to death

Goal of home follow-up
- short term: prevention and control of lung infections
- long-term: slow down the decline in pulmonary function
The Cystic Fibrosis: clinical manifestations

Trend of Spirometry
- 2% /year decrease of FEV1 (forced expiratory volume)
- long term decrease of FVC (current Volume)

In case of infectious relapse
- changes in pulmonary function often precede the clinical symptoms
- monitoring variations in Spirometry is useful both in FC children and in adults

An early antibiotic treatment
- prevents more serious complications
- limits the pulmonary damage in the long term
- allows less invasive antibiotic therapies (orally)
Remote telematic control in cystic fibrosis

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Abstract

Introduction. In this study we describe and discuss the way we daily act in remote telematic tracking of CF outpatients, a procedure which has been improved through our daily experience in telehomecare.

Materials and methods. Currently, there are almost 30 patients

Introduction

Recent advances in technology gave us the possibility to receive, even at big distances, clinical information directly from patients such as measurement of biological parameters. This possibility shed new light in terms of continuity
At Home

Patient:
- Pulse oximetry by night
- spirometry in the morning, after chest FKT and after answering a questionnaire about pulmonary subjective symptoms
- data transmission

- recommended frequency of transmissions: twice a week.
- patient may autonomously decide to send data
In Hospital

Healthcare professionals daily:

• download the data in a protected way

• interpret the data through the application software

• print and store every transmission in a paper archive.
Application Software provides:

- spirometric curves and main parameters (FEV1, FVC, PEF, FEF25-75).
- comparison with previous parameters
- variations expressed as % difference.
- trend of FEV1 short, medium and long term

- graph of the night SaO2 and hearth rate (mean, minimum and maximum SaO2, T90 and T89)
Intervention Parameters

we consider significant:

• Acute reduction of FEV1 (>10% compared to previous values recorded in stable clinical conditions)

• Reductions below 90% of the maximum value of oxygen hemoglobin saturation and of mean SaO2, increase of T90.
Work Flow

• Every patient is called back to retrieve anamnesis data and to share the results.

• Anamnesis data and graphs are discussed in a mid-day briefing between clinicians for
  • overall evaluation
  • any therapeutic action
Decisional Flow

- Patients with significant decrease of SaO2 and/or FEV1 are invited to transmit soon further test.

- In some cases antibiotic home therapy is prescribed on the basis of the last sputum culture collected in hospital.

- In other cases patients are invited for a clinical evaluation, to perform further testing, or to be admitted.

In any case the next data transmission is scheduled
Technological Evolution in Respiratory Telemedicine

2001: Oxytel
- Pulse oximeter, external spirometer

2005: Spirotel
- Pulse oximeter, integrated spirometer

2014: Spirotel2
- Pulse oximeter, integrated spirometer
Oxytel – at patient’s home
Oxytel®
The remote sensing system

data collection and transmission device unit (In patient's home)

data processing unit (PC) in the hospital
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## Oxytel – OPBG experience (2001-2005)

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<tr>
<td>Age at enrolment (Years (mean ± sd))</td>
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<tr>
<td>FEV1 at enrolment (% of exp. value (mean±sd))</td>
<td>67.48 ± 21.28</td>
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<td>Follow-up duration (Months (mean±sd))</td>
<td>29.30 ± 13.32</td>
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Five years of Telemedicine in Cystic Fibrosis Disease

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Abstract

Aims. We studied the effect of Telehomecare (THC) in a group of Cystic Fibrosis (CF) patients, with the aim to early recognize the relapses of pulmonary infections. Data obtained with Vivisol (OXYTEL) instrumentation were collected from 2001 to 2005.

Materials and Methods. The study has involved 17 patients (11 f, 6 m) affected by CF, treated with THC, in addition to the usual therapeutic protocol, for an average period of 29.6 months ± 13.5. The mean age for THC enrolment was 15.74 years ± 5.8. As controls, the study has involved the same patients during the 12 months prior to THC start-up and 28 patients affected by CF treated at our Unit (13 f, 15 m; average age 14.77 ± 5.22).

Results. The results show a statistically significant decrease of outpatient accesses and increase of therapy cycles, and a trend of higher stability of the respiratory function in THC treated subjects.

The natural history of Cystic Fibrosis (CF) is characterized by a progressive lung destruction, caused by obstruction of the airways due to dehydrated thickened secretions, resultant endobronchial infection and an exaggerated inflammatory response leading to development of bronchiectasis and progressive obstructive airways disease (4). Prevention and control of lung infections is one of the main objectives of therapy in CF patients with the aim to slow down the progressive decline in pulmonary function with time (5).

An acute decrease of FEV₁ >10% is considered a significant sign of infectious pulmonary relapse (6).

Many researchers demonstrated that, in case of infectious relapse, pulmonary function modifications often precede the clinical symptoms and that monitoring variations in pulmonary function can be useful in children and in adults.
FEV1 monthly averages over 5 years
Fev1 monthly averages

Fev1 (mean ±2DS) controls

![Graph showing monthly averages of Fev1](image-url)
Oxytel – OPBG experience (2001-2005)

Results:

in THC treated subjects compared to controls

• statistically significant decrease of outpatient accesses and increase of therapy cycles
• trend of higher stability of the respiratory function.

Conclusions:

• Telehomecare seems to increase in general the rate of access to health care without any clear effect of pulmonary function.
Spirotel® - at patient’s home
Spirotel – transmission of data
Spirotel - The remote sensing system

Peripheral (in patient's home)
Design of the Study

Consent to participate in research has been granted in writing by all participants or, in the case of minors, by their parents.

For this study, we didn’t receive dedicated funding, except INHS (Italian National Health Service) fees for provided service.

Since telemonitoring actually in Italy is not yet included in the essential basic assistance levels (LEA), its viability depends in the individual cases from voluntary resources provided by INHS local Authorities.
Admission Criteria

Regarding the admission criteria in telemonitoring, at today, in CF subjects the definition of general criteria for inclusion is difficult as there is no experimental data that ensure in the various expressions and clinical types significant differences about a better efficacy of the THC procedure.

We have included in telemonitoring all CF subjects followed in our Center for which the local INHS Authorities has made available the necessary resources.

Exclusion criteria from the study were essentially the interruption or rarefaction of transmissions or death.

All co-morbidities were disease-related major and did not constitute grounds for exclusion.
Balance of Enrolment 15/09/2016

- Enrolled: 69 (100.00%)
- Active: 34 (49.28%)
- Dropout: 35 (50.72%)
- Poor Adherence: 16 (45.71%)
- ASL Revocation: 4 (11.43%)
- Exitus: 5 (14.29%)
- Other: 10 (28.57%)
Spirotel: 2010-2016 Activity

### Spirotel-Tendency 2016

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<td>35</td>
<td>11</td>
<td>7</td>
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Mean annual FEV1 relative to the 2011 value

telemecina

controlli
Discussion

Patients followed in telemedicine showed in most cases a lower decay of respiratory function compared to those in follow-up with the traditional method.

The data show an increase over time of sent exams despite the number of patients in the telemonitoring has remained virtually constant.
Discussion

One of the critical aspects in the follow-up of chronic patients is poor adherence to therapy. We have shown over time a considerable increase of % adherence to telemonitoring.

The home telemonitoring has been accepted in most cases positively by patients. The percentage of telephone answers (≈ 80%) is to be considered high (as if the patient expected the contact with the center) but insufficient to consider the phone a completely reliable means of communication.
Conclusions

In our experience, gained over a relatively long period, telemedicine is a method useful in the follow-up of chronic diseases because it allows:

• Less deterioration of the pulmonary function, with consequent less need, in the long term, to employ invasive therapies

• A radical change of the motivations of the hospital accesses, which become more rational and less demanding route for both the patient and for the treating staff

• Overall a better quality of life
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