

MANAGEMENT OF PEDIATRIC PATIENTS IN CYSTIC FIBROSIS WITH TELEMEDICINE

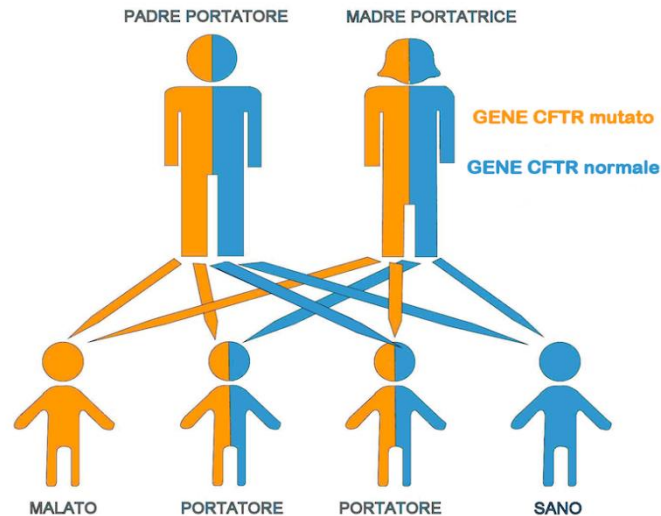
SERGIO BELLA and FABRIZIO MURGIA



Ospedale Pediatrico Bambino Gesù – IRCCS
Dipartimento di Pediatrie Specialistiche
U.O.C. di Fibrosi Cistica
Servizio Speciale A.D.I. nelle Malattie Croniche
Piazza S. Onofrio 4 -00165 Roma

The Cystic Fibrosis

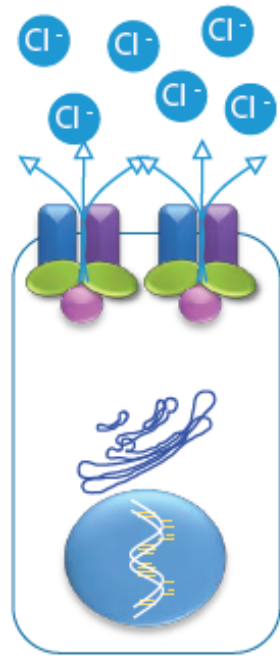
- Genetic, hereditary and chronic disease, widespread among the Caucasian population, resulting in fatal outcome.



Affects about one child every 3,000 births.

In Italy more than 4,000 people currently affected

One Way of Classifying CFTR Mutations



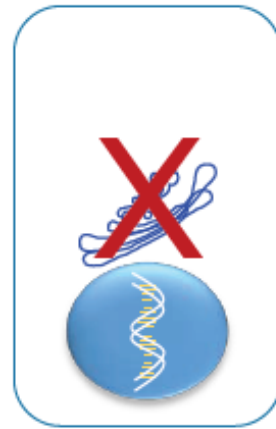
Normal

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



Class I

No functional CFTR created.



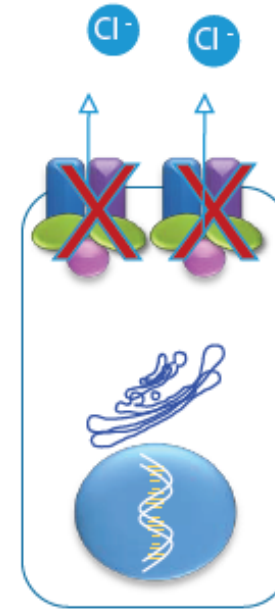
Class II

CFTR protein is created, but misfolded, keeping it from reaching the cell surface.



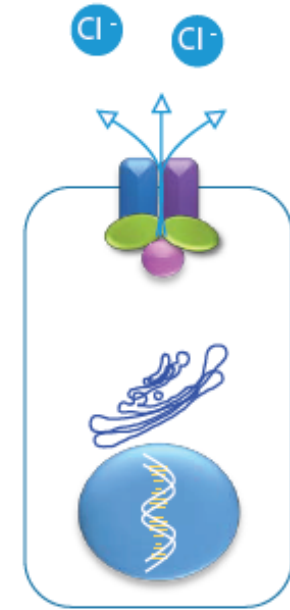
Class III

CFTR protein is created and reaches cell surface, but does not function properly.



Class IV

The opening in the CFTR protein ion channel is faulty.



Class V

CFTR is created in insufficient quantities.

EXAMPLES

G542X
W1282X
R553X

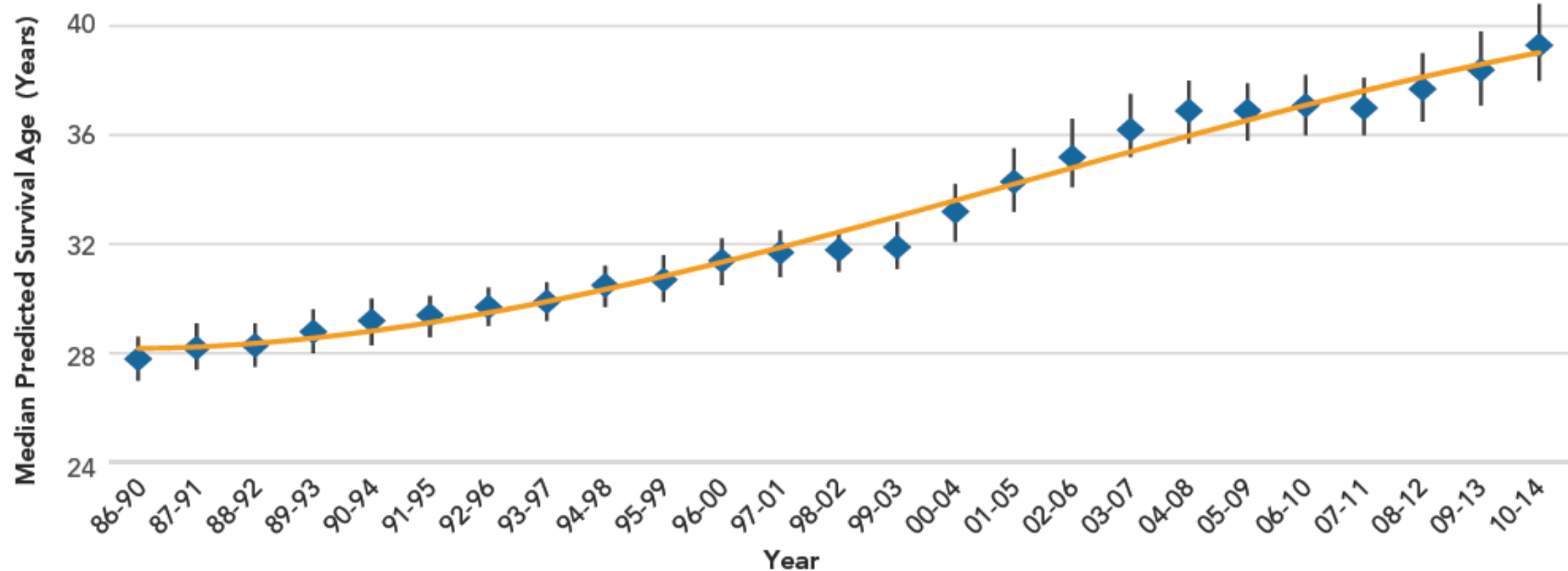
F508del
N1303K
I507del

G551D
S549N
V520F

R117H
D1152H
R347P

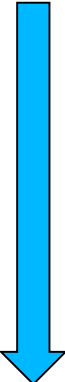
3849+10kbC->T
2789+5G->A
A455E

Median Predicted Survival Age, 1986–2014 In 5-Year Increments



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

Murgia F.^{1§}, Cilli M.², Renzetti E.³, Majo F.¹, Soldi D.⁴, Lucidi V.¹, Bella F.⁵, Bella S.¹

¹Cystic Fibrosis Unit, ²Pediatric Medicine Department, ⁴Epidemiology Unit, Bambino Gesù Pediatric Hospital – IRCCS – Rome; ²II Medical Clinic, “La Sapienza” University of Rome, ⁵Emeritus Professor of Physics, “Roma 3” University of Rome, Italy; [§]External consultant

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 telehome-care.

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 of care and optimization and coordination of clinical

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



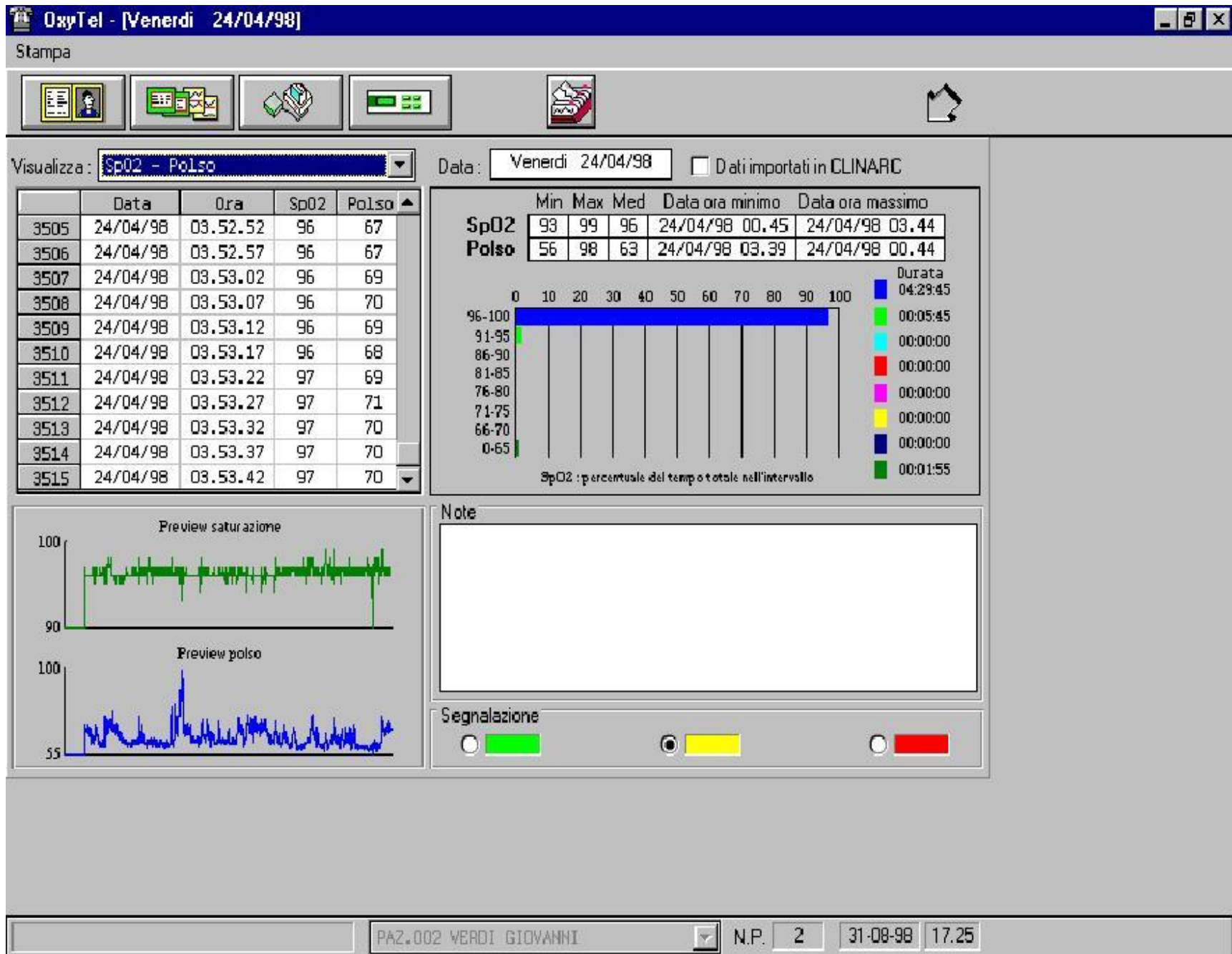
Oxytel[®]


The remote sensing system




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













data processing unit
(PC) in the hospital




OxyTel - [Dati Paziente : A. G.]

File
Utilità

	Data Acquisizione	Ora	Note
<input type="checkbox"/>	Mercoledì	20\03\96 05:09	
<input type="checkbox"/>	Giovedì	21\03\96 18:20	
<input type="checkbox"/>	Venerdì	22\03\96 05:09	
<input type="checkbox"/>	Sabato	23\03\96 05:07	
<input type="checkbox"/>	Domenica	24\03\96 05:07	Lievi desaturazioni dopo le ore 04:00
<input type="checkbox"/>	Lunedì	25\03\96 05:07	
<input type="checkbox"/>	Martedì	26\03\96 05:08	
<input type="checkbox"/>	Giovedì	28\03\96 05:08	
<input type="checkbox"/>	Venerdì	29\03\96 05:08	Dispnea
<input type="checkbox"/>	Sabato	30\03\96 05:08	
<input type="checkbox"/>	Domenica	31\03\96 05:06	
<input type="checkbox"/>	Lunedì	01\04\96 05:06	
<input type="checkbox"/>	Martedì	02\04\96 05:06	
<input type="checkbox"/>	Mercoledì	03\04\96 05:06	
<input type="checkbox"/>	Giovedì	04\04\96 05:07	Monitoraggio notturno con desaturazioni significative...
<input type="checkbox"/>	Venerdì	05\04\96 05:07	Monitoraggio notturno
<input type="checkbox"/>	Sabato	06\04\96 05:06	ATTENZIONE! Sensore posizionato male.
<input type="checkbox"/>	Venerdì	12\04\96 18:34	
<input type="checkbox"/>	Venerdì	12\04\96 18:40	
<input type="checkbox"/>	Venerdì	12\04\96 18:48	Grave desaturazione

Filtro

☒
Visualizza ultimi :
giorni

☐
Visualizza dati del Mese :
Anno:

A. G.

N.P.

1

12-04-96

23.46

Oxytel – OPBG experience (2001-2005)

n. of treated patients	17
Male/Female	6/11
Age at enrolment Years (mean \pm sd)	15.74 \pm 5.8
FEV1 at enrolment % of exp. value (mean \pm sd)	67.48 \pm 21.28
Follow-up duration Months (mean \pm sd)	29.30 \pm 13.32

Five years of Telemedicine in Cystic Fibrosis Disease

S. Bella¹, F. Murgia¹, A.E. Tozzi², C. Cotognini³, V. Lucidi⁴

¹Cystic Fibrosis diagnosis and treatment Unit, ²Epidemiology Unit, ³Information Systems and Organization Department, Children's Hospital Bambino Gesù, Research and Care Institute, Rome, Italy

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 \pm 13.5. The mean age for THC enrolment was 15.74 years \pm 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 \pm 5.22).

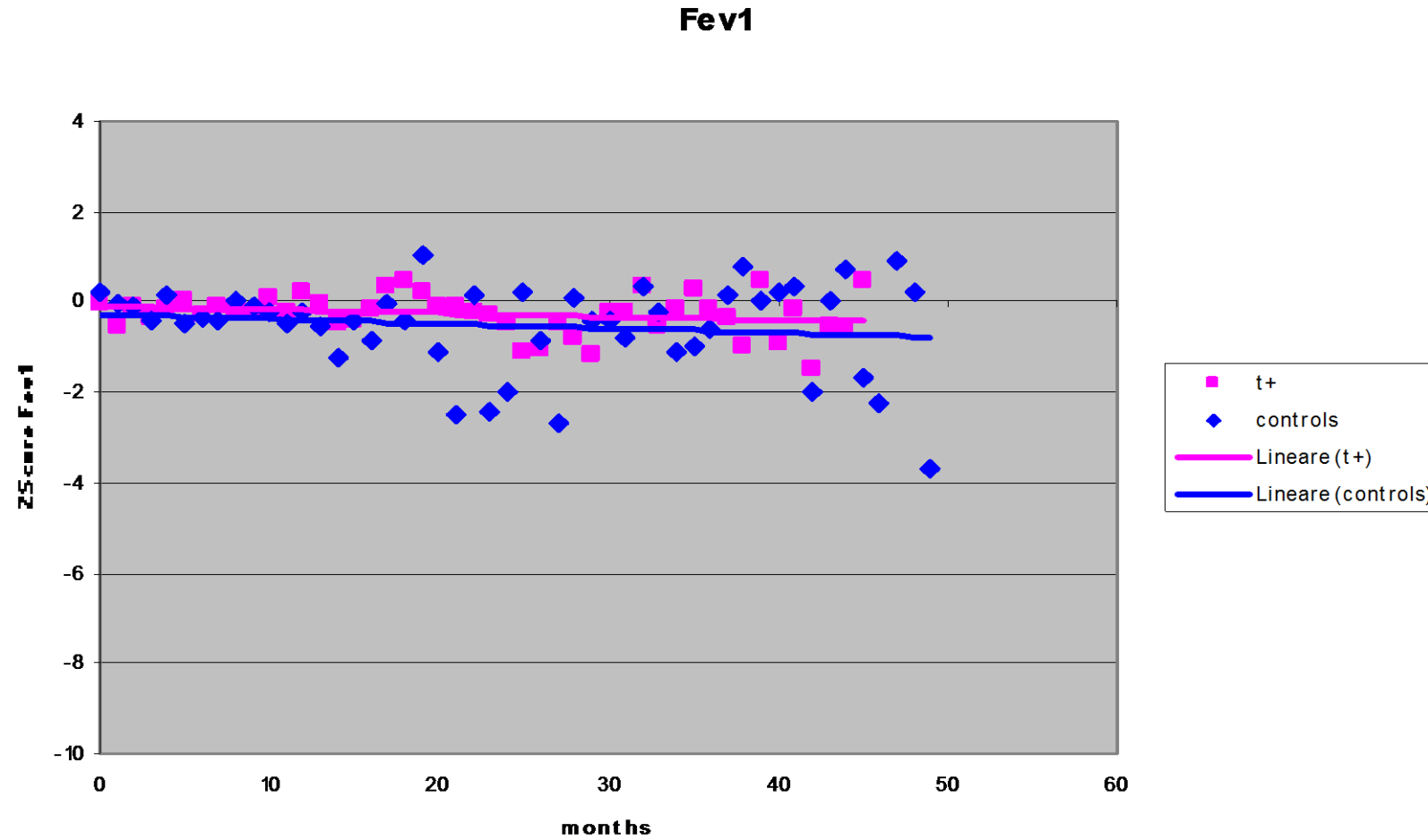
Results. The results show a statistically significant decrease of outpatient accesses and increase of therapy cycles, and a trend of higher stability of the respiratory function, in THC treated subjects

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).

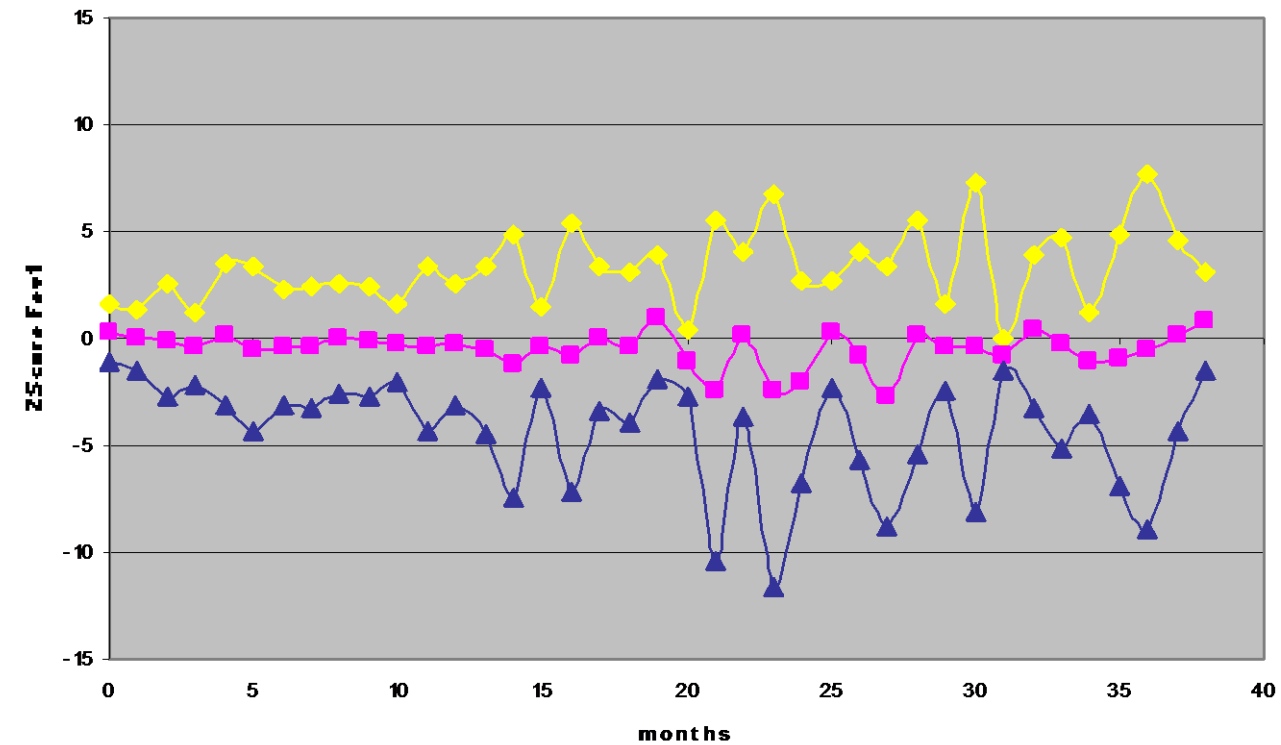
Many researchers demonstrated that, in case of infectious relapse, pulmonary function modifications often precede the clinical symptoms and that monitoring variations in pulmonary function can be useful in children and in adults

FEV1 monthly averages over 5 years



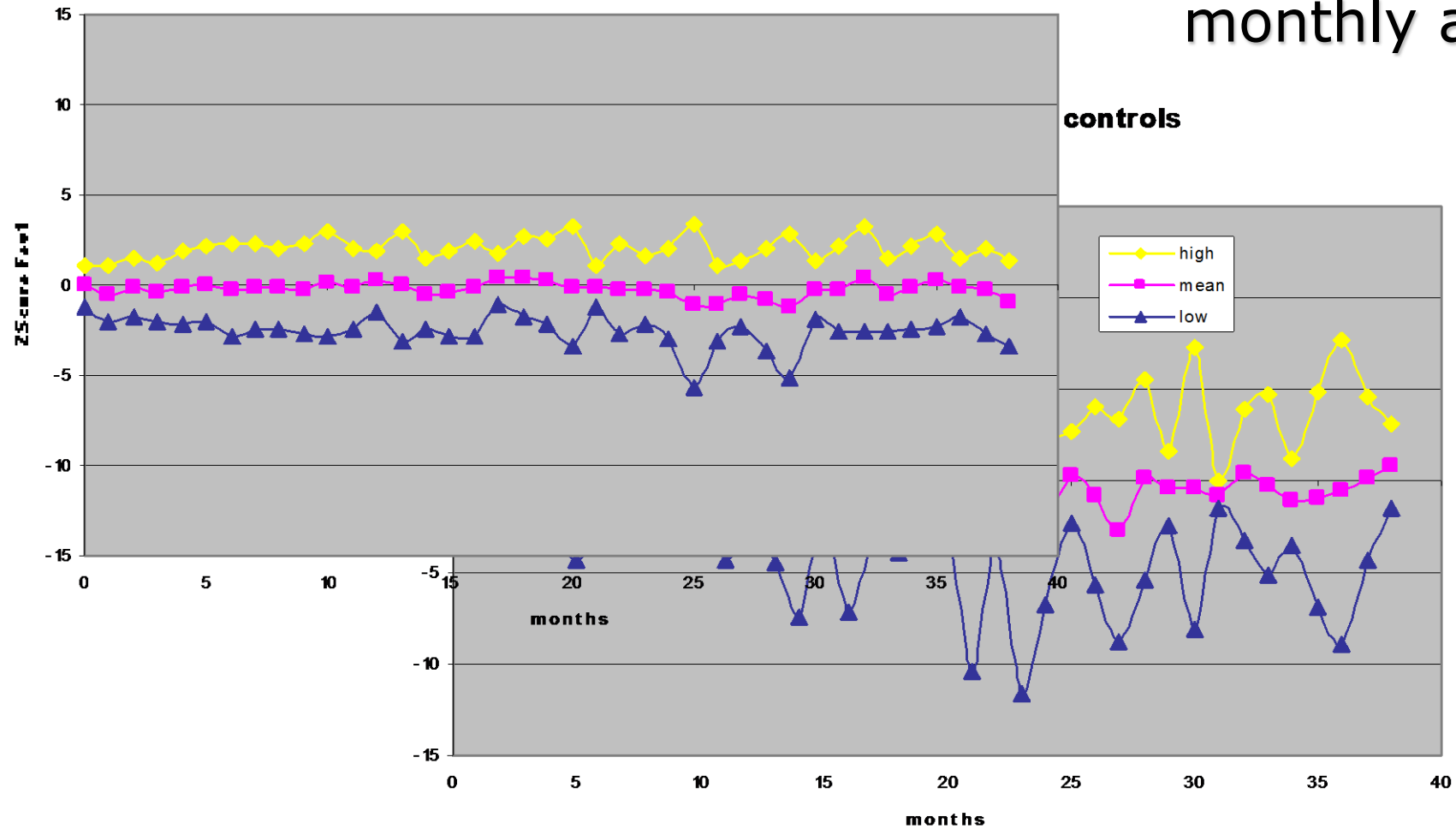
Fev1 monthly averages

Fev1 (mean \pm 2DS) controls



Fev1 (mean \pm 2DS) t+

Fev1
monthly averages



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

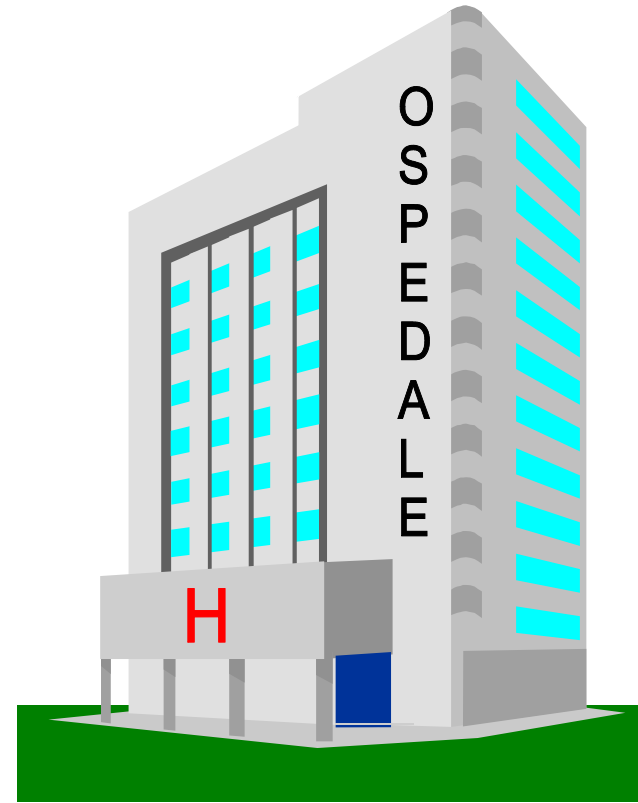


Spirotel - The remote sensing system

Peripheral (in patient's home)



Central Unit
(Web)



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

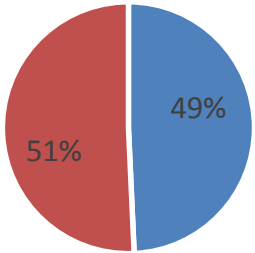
We have included in telemonitoring all CF subjects followed in our Center for which the local INHS Authorities has made available the necessary resources.

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.

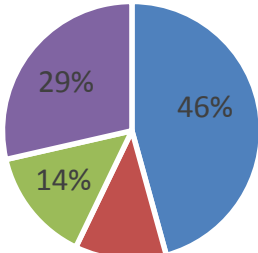
			Balance of Enrolment		15/09/2016					
			Enrolled	69	100,00%					
			Active	34	49,28%					
			Dropout	35	50,72%					
			PoorAdherence	16	45,71%					
			ASL Revocation	4	11,43%					
			Exitus	5	14,29%					
			Other	10	28,57%					

Balance of Enrolment



■ Active ■ Dropout

Causes of dropout

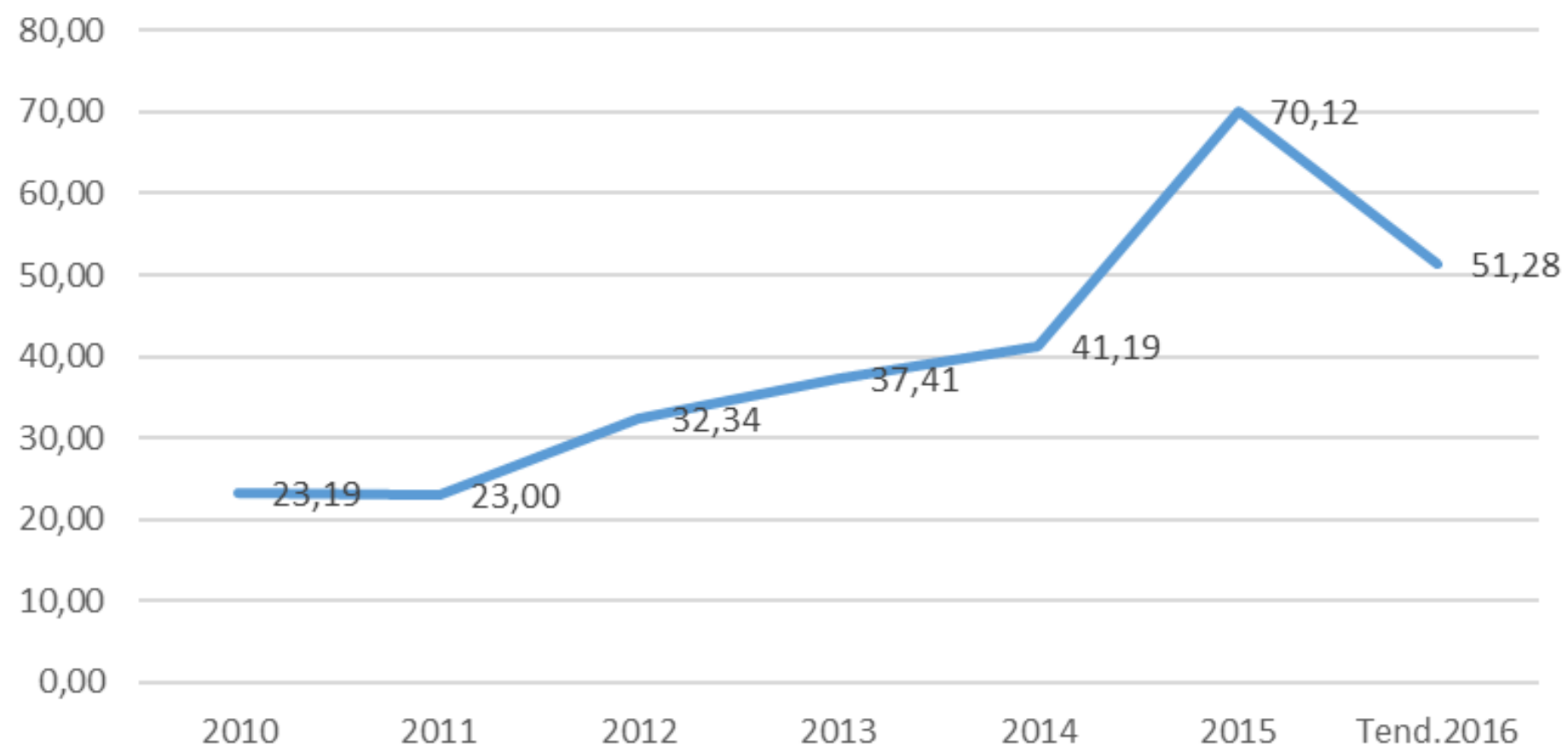


■ PoorAdherence ■ ASL Revocation ■ Exitus ■ Other

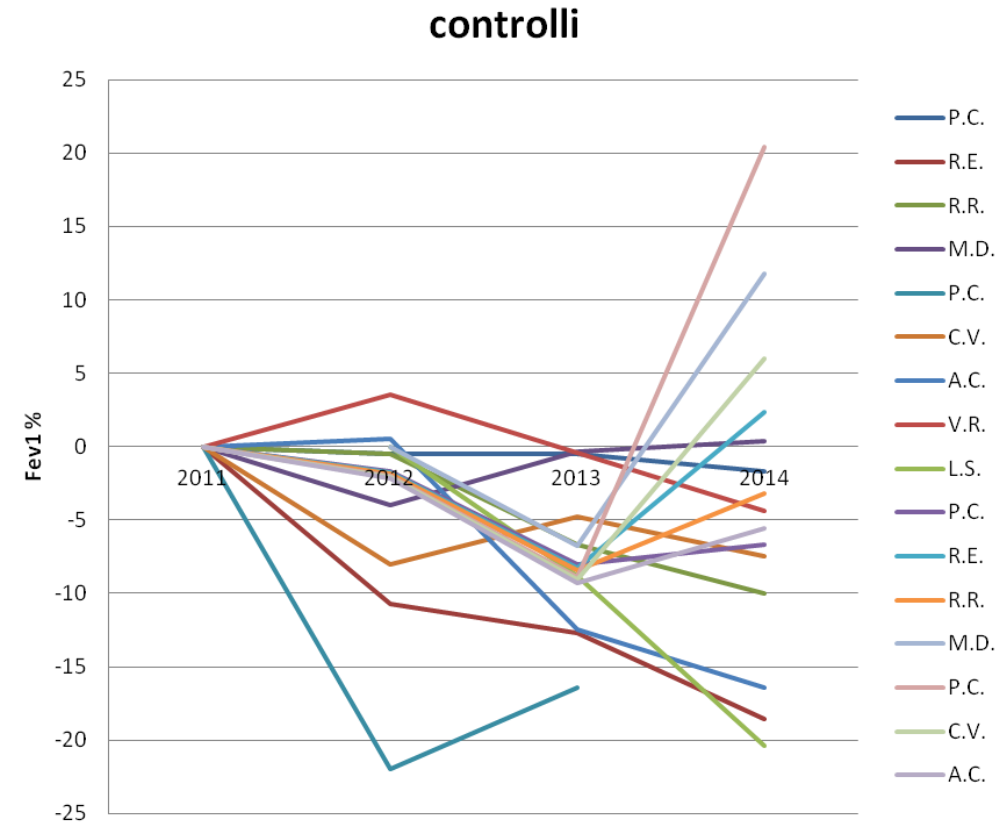
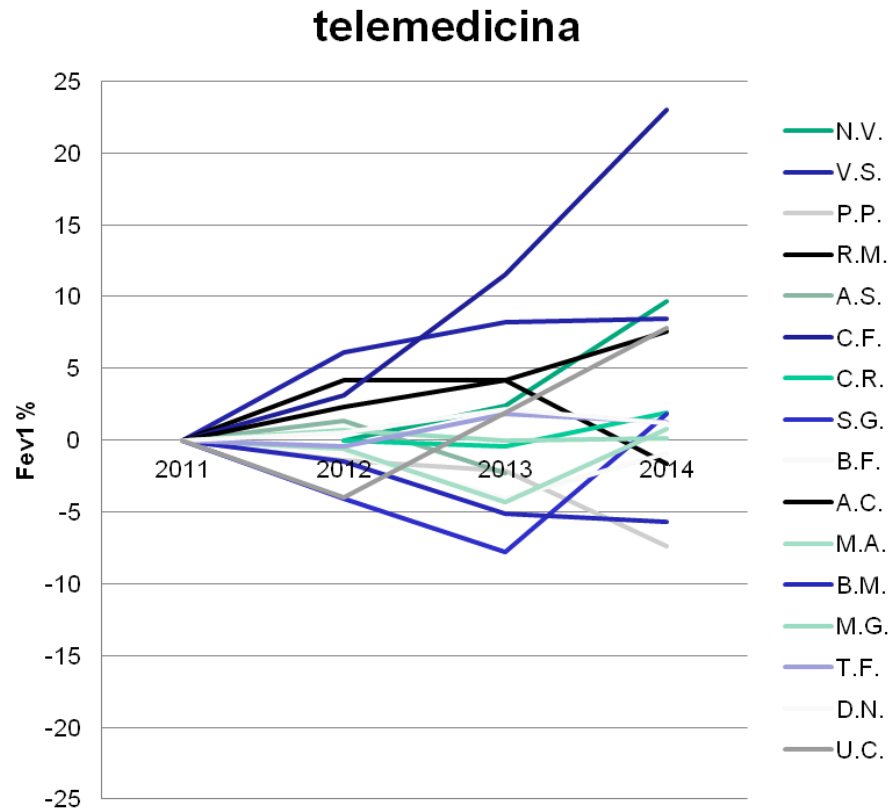
Spirotel: 2010-2016 Activity

Spirotel-Tendency 2016									
period	2010	2011	2012	2013	2014	2015	Tend.2016		Total
patients n.	30	29,7	26,5	24,6	25,1	30	33		28,4 (mean)
days	226	257	243	235	249	254	252		1716
transmissions	466	669	831	868	1029	2127	1706		7696
spirometry	554	985	1060	957	952	952	1495		6955
pulse oximetry	162	211	292	168	62	249	161		1304
symptoms		255	709	755	794	994	18		3525
adherence	23,19	23,00	32,34	37,41	41,19	70,12	51,28		
phone calls	420	592	745	672	669	1194	1012		5304
answers			618	564	573	905	705		
answers/calls			82,95%	83,93%	85,65%	75,80%	69,63%		
inpatients n.	8	15	49	38	35	11	7		163

Adherence



Mean annual FEV1 relative to the 2011value



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 ($\approx 80\%$) 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

xpatient Barcelona Congress - 21 set 2016

Pediatric Hospital Bambino Gesù – IRCCS

Department of Special Pediatrics

Cystic Fibrosis Unit

Special Service of Continuity of Care in Chronic Diseases

Sergio Bella, Fabrizio Murgia, Irene Tagliente, Ambra Carestia,
Vanessa Mercuri, Elena Prosseda, Francesca Cicchetti, Benedetta
Corona - Piazza S. Onofrio 4

00165 Roma

+39 06 6859 2669

telemedicina@opbg.net

<http://www.ospedalebambinogesu.it>