7° Forum italiano sulla Fibrosi Cistica

forum 2019

PER VIVERE IL PRESENTE COSTRUENDO IL DOMANI

22-24 Novembre 2019

SAPIENZA UNIVERSITÀ DI ROMA

LIFC Lega Italiana Fibrosi Cistica

Semplificazione dei tempi di cura a casa. Minimizzare i tempi di disinfezione delle apparecchiature e protocolli di pulizia

Dr Daniela Savi, *PhD*

Researcher, Policlinico Umberto I Hospital

Department of Public Health and Infectious Diseases

Sapienza University of Rome

La sottoscritta Dott.ssa Daniela Savi

ai sensi dell’art. 76 comma 4 dell’Accordo Stato-Regioni del 2 febbraio 2017

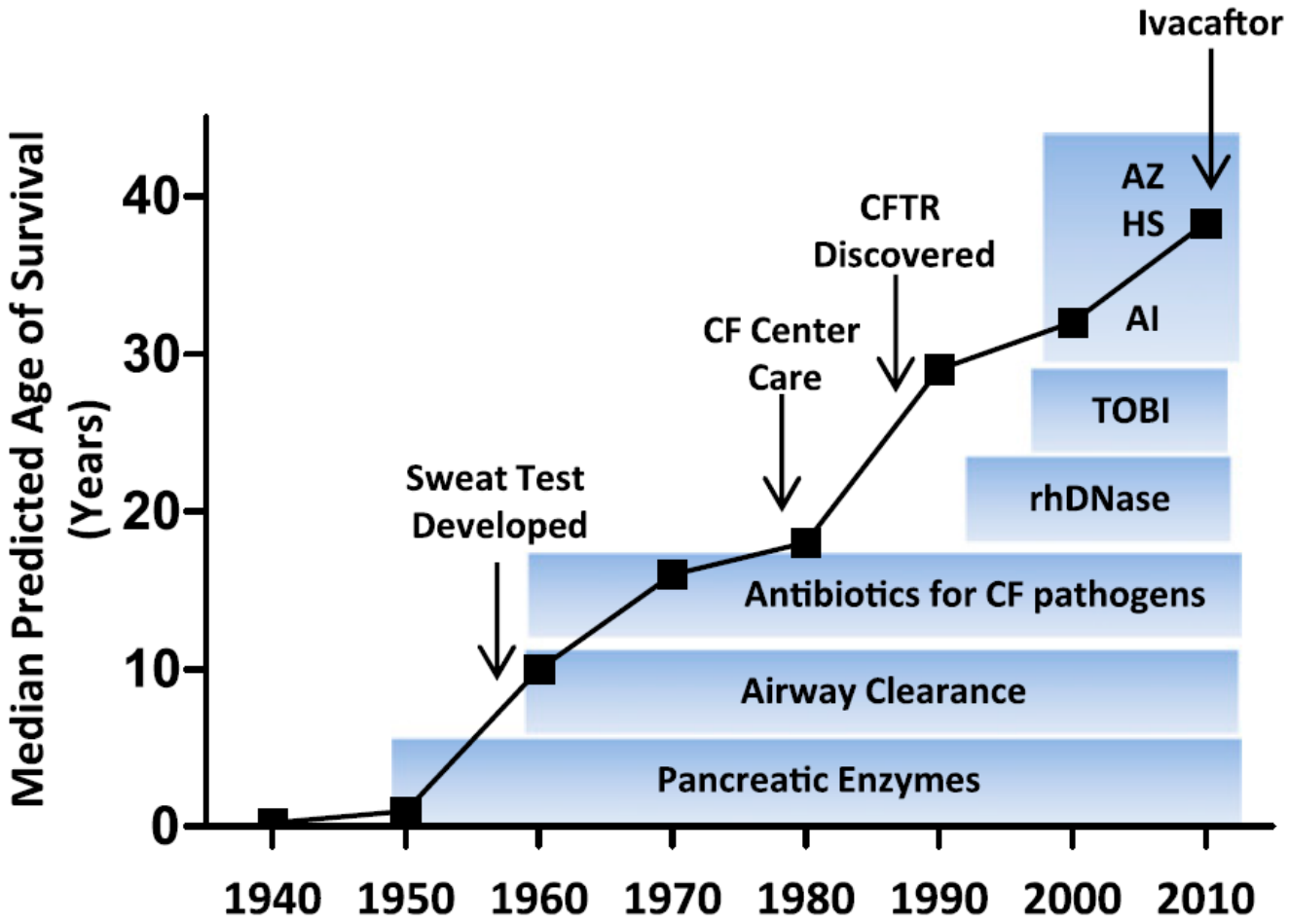
dichiara

per l’evento in oggetto l’**assenza** negli ultimi due anni di rapporti di natura finanziaria e lavorativa con le imprese commerciali operanti in ambito sanitario

- Risultati ottenuti dalle attuali terapie in FC
- Evidenze di scarsa compliance alle terapie in FC
- Interventi per semplificare i tempi di cura e minimizzare i tempi di disinfezione delle apparecchiature

Risultati ottenuti dalle terapie attuali in FC

Survival in CF



Clancy JP AJRCCM 2012

Survival in CF

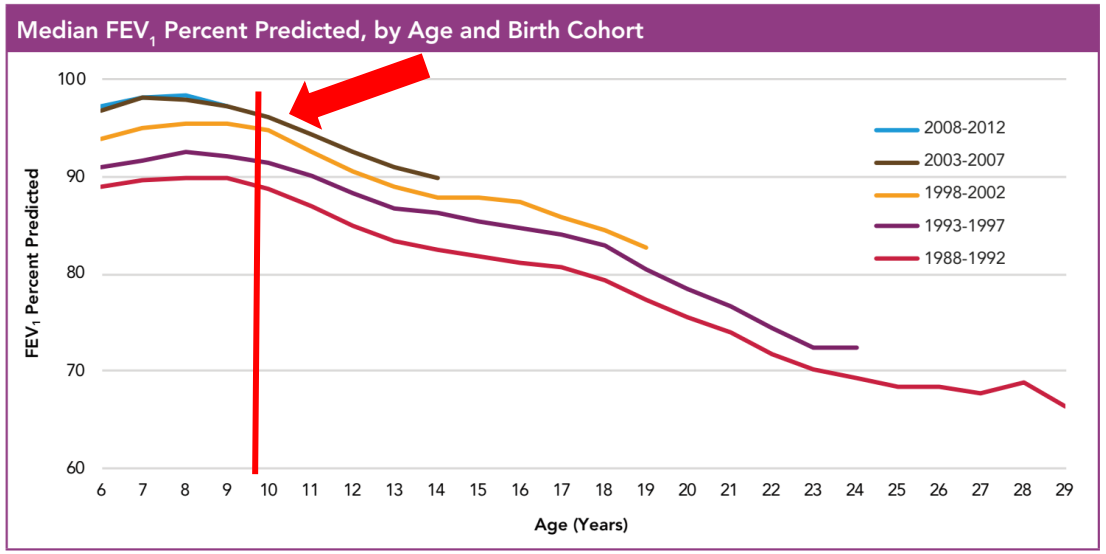
Report of the European Respiratory Society/European Cystic Fibrosis Society task force on the care of adults with cystic fibrosis

JS ELBORN et al. ERS/ECFS report 2016

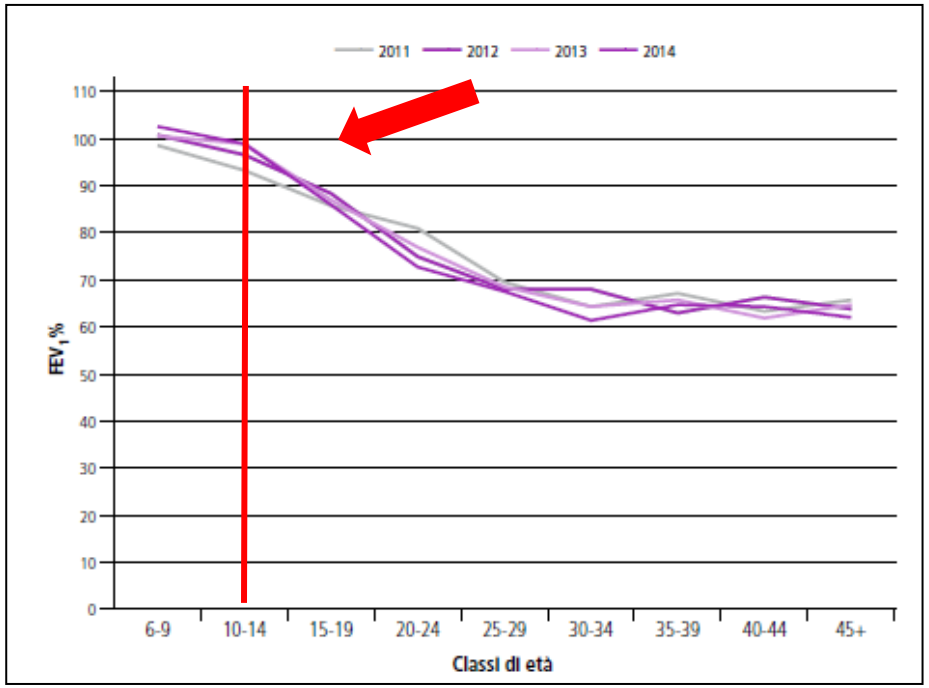
TABLE 1 Reported median survival and numbers of children and adults with cystic fibrosis from selected registries

Country	Year of annual report	Median age at death years	Median predicted age of survival years	Percentage of adult patients (≥18 years)	Total number of patients
Belgium	2011[#]	33	NA	54.7	1171
Denmark	2010 (ECFSPR)[#]	33	NA	56.8	450
France	2012	28	NA	47.2	6196[¶]
Germany	2012	32	NA	52.4	5242
UK	2012	28	43.5	57.6[+]	10 078[§]
Australia	2012	29	NA	49.3	3156
Canada	2012	32	49.7	58.8	3975
USA	2012	NA	41.1	49.1	27 804

[#]: age computed from the 2010 European Cystic Fibrosis Society patient registry (ECFSPR) data; [¶]: patients whose vital status is known, whether they visited the cystic fibrosis centre or not; [+]: ≥16 years; [§]: all patients. NA: not available.

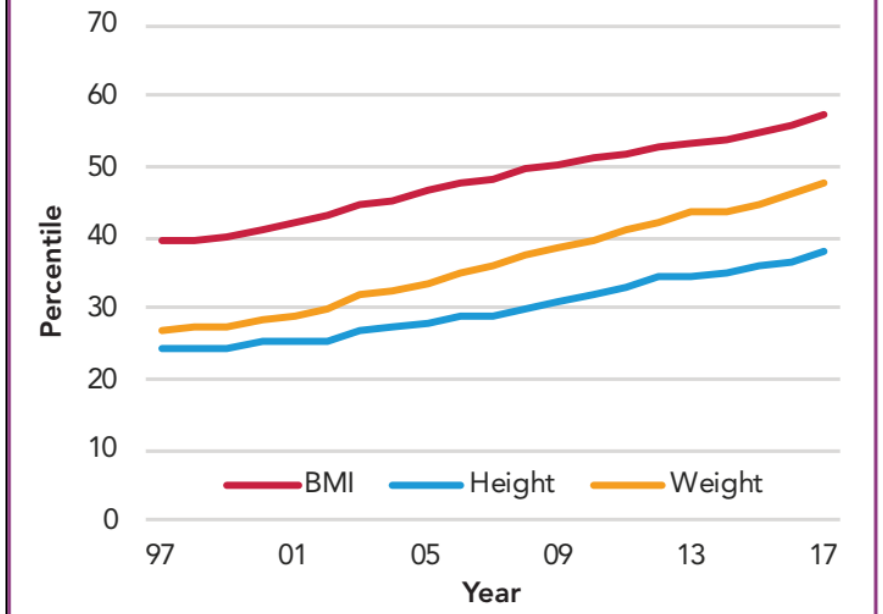


Cystic Fibrosis Foundation 2017

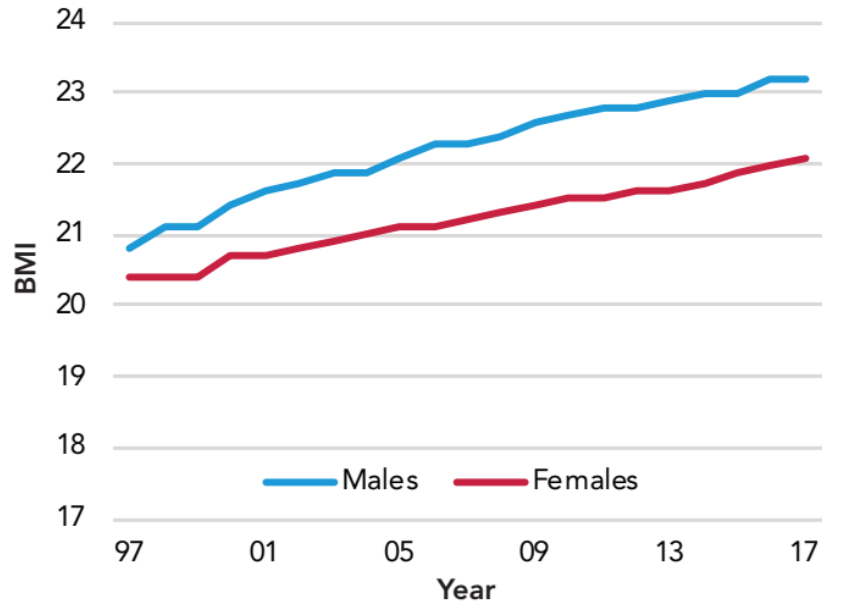


Italian Cystic Fibrosis Report 2018

Median Nutritional Outcomes Percentiles for Individuals 2 to 19 Years, 1997–2017

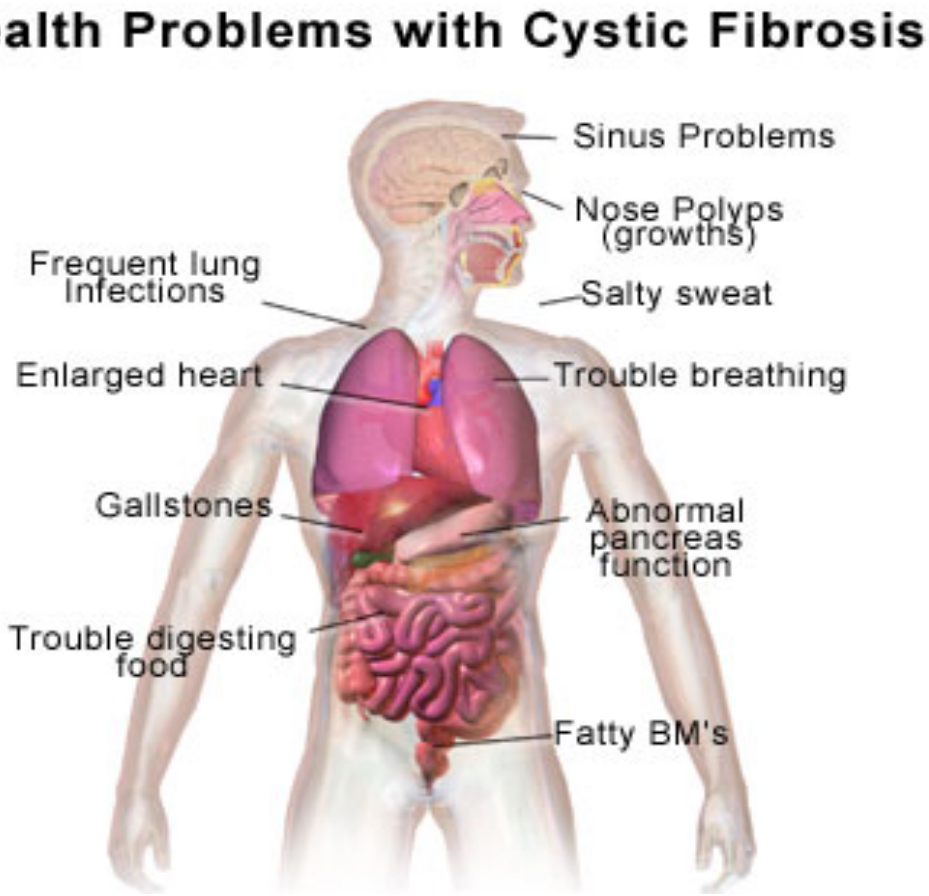


Median BMI Values for Adults 20 Years and Older, 1997–2017



Cystic Fibrosis Foundation 2017

Complessità della terapia



TIME CONSUMING





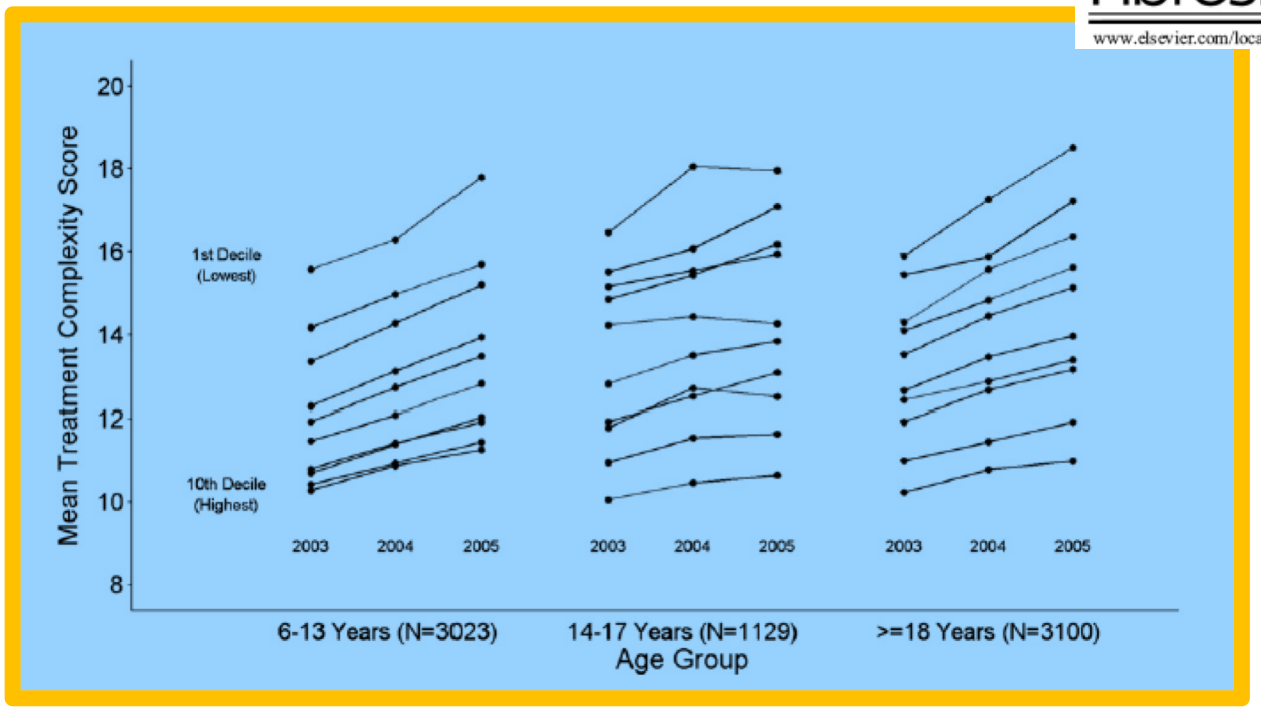

Dalle 7 alle 21 ore settimanali

CAN THE TREATMENT COMPLEXITY AND THE ENORMOUS NUMBER OF DRUGS ADMINISTERED EVERY DAY CAUSE NON-ADHERENCE TO THERAPY?

Treatment complexity in cystic fibrosis: Trends over time and associations with site-specific outcomes

Gregory S. Sawicki [a,*], Clement L. Ren [b], Michael W. Konstan [c], Stefanie J. Millar [d], David J. Pasta [d], Alexandra L. Quittner [e]
for the Investigators and Coordinators of the Epidemiologic Study of Cystic Fibrosis

Journal of Cystic Fibrosis
www.elsevier.com/locate/jcf
2013



Evidenze di scarsa compliance alle terapie in FC

Pulmonary Medication Adherence

The NEW ENGLAND JOURNAL of MEDICINE

REVIEW ARTICLE

DRUG THERAPY

Adherence to Medication

Lars Osterberg, M.D., and Terrence Blaschke, M.D.

Drugs don't work in patients who don't take them.

— C. Everett Koop, M.D.

Determinants of adherence in adults with cystic fibrosis

L J Kettler, S M Sawyer, H R Winefield, H W Greville

Thorax 2002;**57**:459–464

Adherence rates are **27–46%** for pancreatic enzymes and **51%** for frequency of airway clearance

ELSEVIER

Journal of Cystic Fibrosis 10 (2011) 258–264

Journal of Cystic Fibrosis

www.elsevier. www.elsevier.com/locate/jcf

Original Article

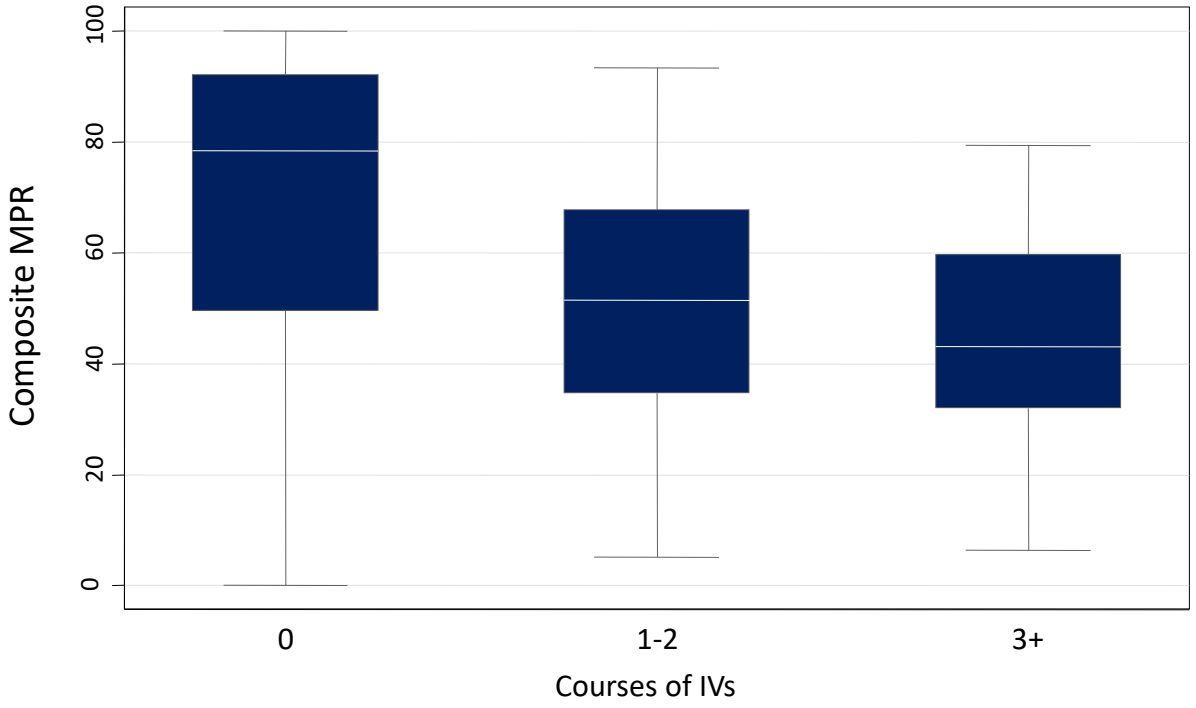
Longitudinal association between medication adherence and lung health in people with cystic fibrosis☆

Michelle N. Eakin [a], Andrew Bilderback [a], Michael P. Boyle [a], Peter J. Mogayzel [b], Kristin A. Riekert [a,*]

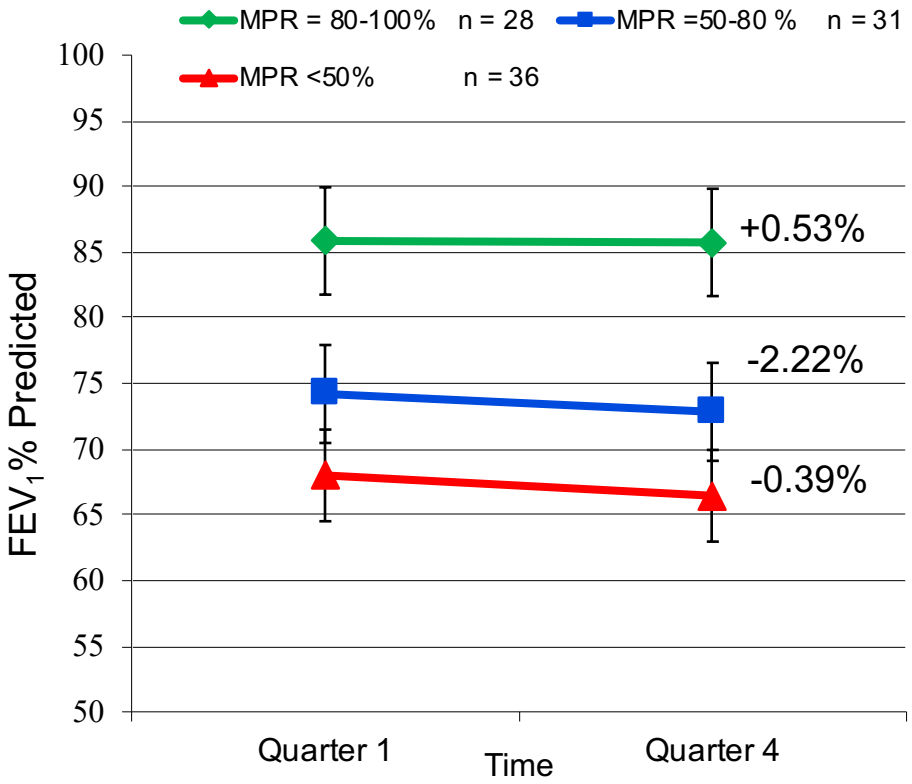
[a] Division of Pulmonary and Critical Care Medicine, Johns Hopkins School of Medicine, USA
[b] Department of Pediatrics, Johns Hopkins School of Medicine, USA

IMPATTO CLINICO DELLA NON ADERENZA

CICLI DI TERAPIA E.V.



FUNZIONALITÀ POLMONARE



ELSEVIER

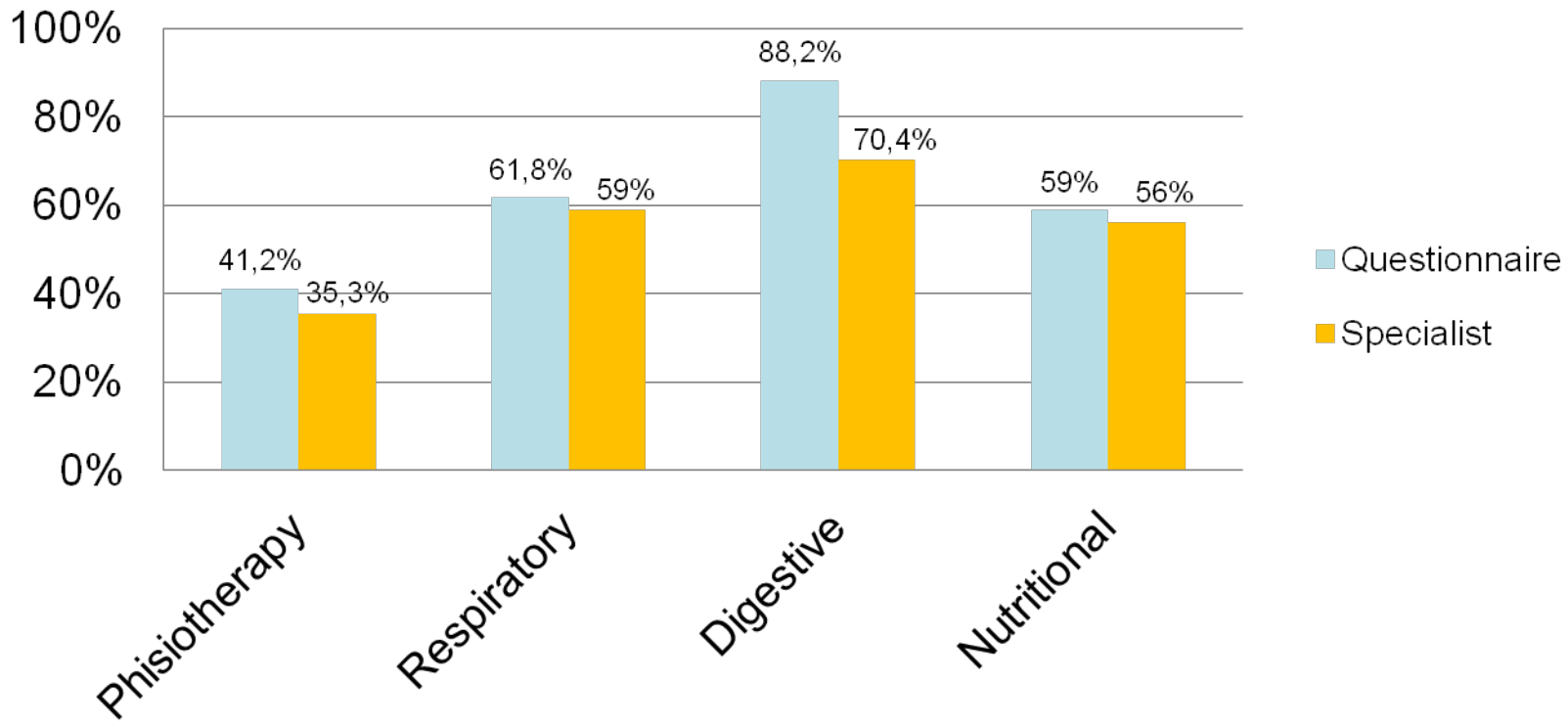
Journal of Cystic Fibrosis 7 (2008) 359–367

Journal of Cystic Fibrosis

www.elsevier.com/locate/jcf

Treatment compliance in children and adults with Cystic Fibrosis

Rosa Patricia Arias Llorente [a,*], Carlos Bousoño García [a], Juan José Díaz Martín [b]



Percentage of therapeutics adherence for each type of treatment based on questionnaire and specialist opinions

Reasons for the poor compliance with different treatments

Predominant reasons for poor compliance	*n* (% patients)
Physiotherapy	
Not enough time	8 (29.4)
I don't think I need it, I feel well without treatment	7 (20.6)
Exercise instead	7 (20.6)
I believe that doing it I don't feel better	5 (14.7)
It interferes with my social life	4 (11.7)
I simply forget it	3 (8.8)
Transplantation	2 (5.9)
Respiratory medication	
I don't think I need it	4 (11.7)
Not enough time	3 (8.8)
Only when I feel worse	3 (8.8)
It interferes with my social life	2 (5.9)
I simply forget it	2 (5.9)
Digestive medication	
I simply forget it	2 (5.9)
Nutritional supplements	
I don't think I need it	5 (14.7)
I don't like the taste or texture	5 (14.7)
I simply forget it	3 (8.8)
I don't believe in it	2 (5.9)

Llorente RPA Journal of Cystic Fibrosis 2008

[Original Research **Genetic and Developmental Disorders**]

CHEST

Pulmonary Medication Adherence and Health-care Use in Cystic Fibrosis

Alexandra L. Quittner, PhD; Jie Zhang, PhD; Maryna Marynchenko, MBA; Pooja A. Chopra, MS; James Signorovitch, PhD; Yana Yushkina, BA; and Kristin A. Riekert, PhD

CHEST 2014; 146(1):142-151

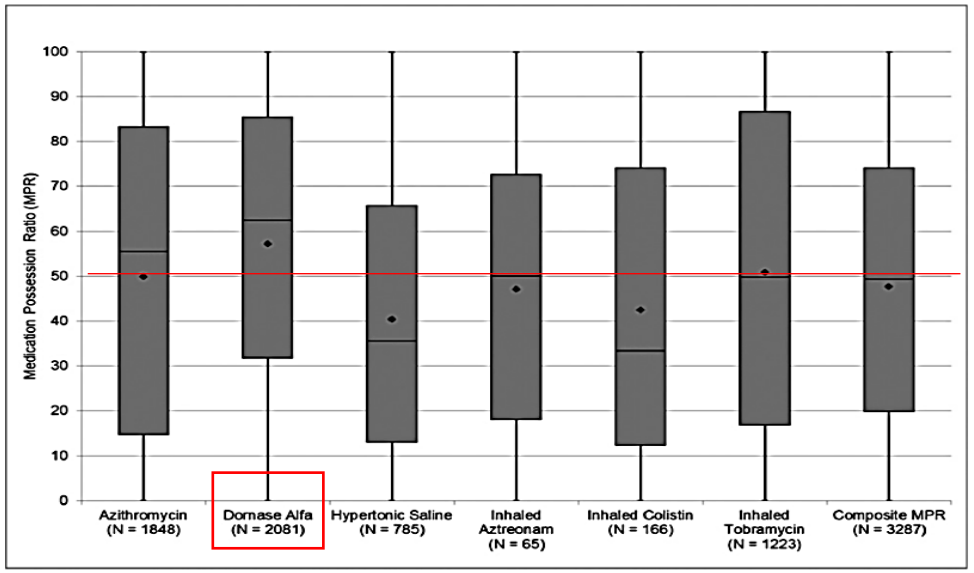


Figure 1 – *Mean MPRs for various long-term pulmonary medications used in cystic fibrosis. The bottom, midline, and top of each box repres lower quartile, median, and upper quartile, respectively. The end points of the vertical lines represent the minimum and maximum values (◆ indicates the mean value). The composite MPR is the average of the individual drug MPRs.*

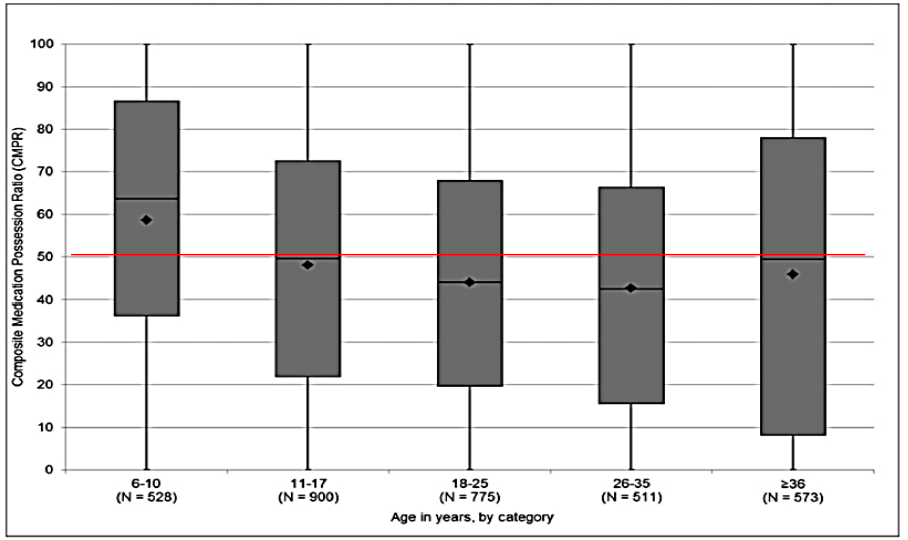


Figure 2 – *CMPRs by age category. The bottom, midline, and top of each box represent the lower quartile, median, and upper quartile, respectively. The end points of the vertical lines represent the minimum and maximum values (◆ indicates the mean value). The CMPR is the average of the individual drug medication possession ratios.*

DIFFERENT MEASURES OF ADHERENCE

- **PARENTAL and SELF-REPORT QUESTIONNAIRE**
  (Treatment Adherence Questionnaire)
- **STRUCTURED INTERVIEW**
- **DAILY PHONE DIARIES**
- **PROXY REPORTS (Caregivers, healthcare professionals)**
- **ELECTRONIC MONITORS (e.g. I-Neb)** Gold standard
- **PRESCRIPTION REFILL HISTORIES (Pharmacy Refill Records)**
- **BIOCHEMICAL ASSAYS**

CHEST Original Research
RESPIRATORY CARE

Accurate Assessment of Adherence

Self-Report and Clinician Report vs Electronic Monitoring of Nebulizers

Tracey Daniels, MSc; Lynne Goodacre, PhD; Chris Sutton, PhD; Kim Pollard, BSc(hons); Steven Conway, MBBS; and Daniel Peckham, MD

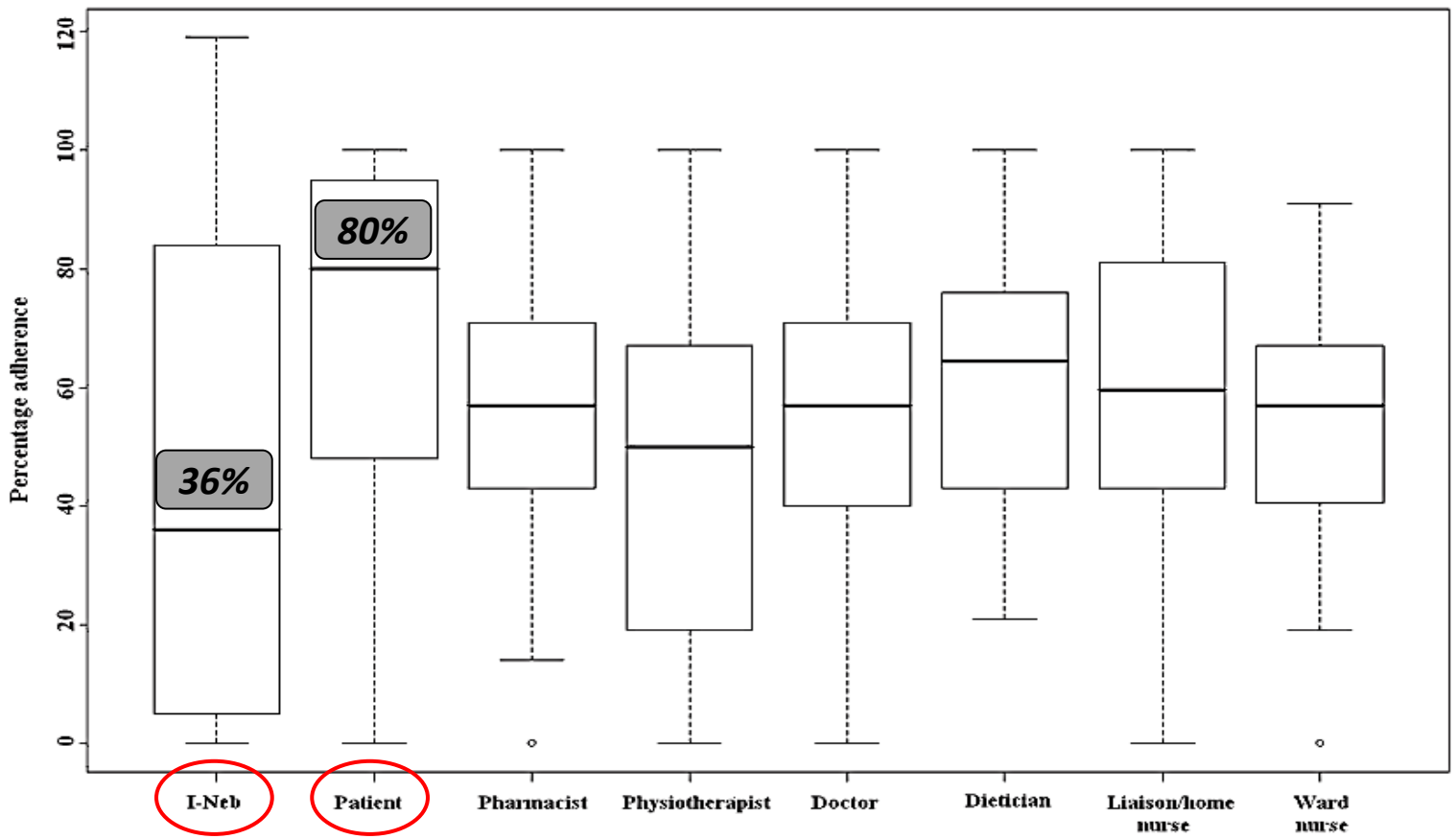


FIGURE 2. Box-and-whisker plot of percentage adherence according to different approaches to assessment.

Paediatric Respiratory Reviews 15S (2014) 16–18

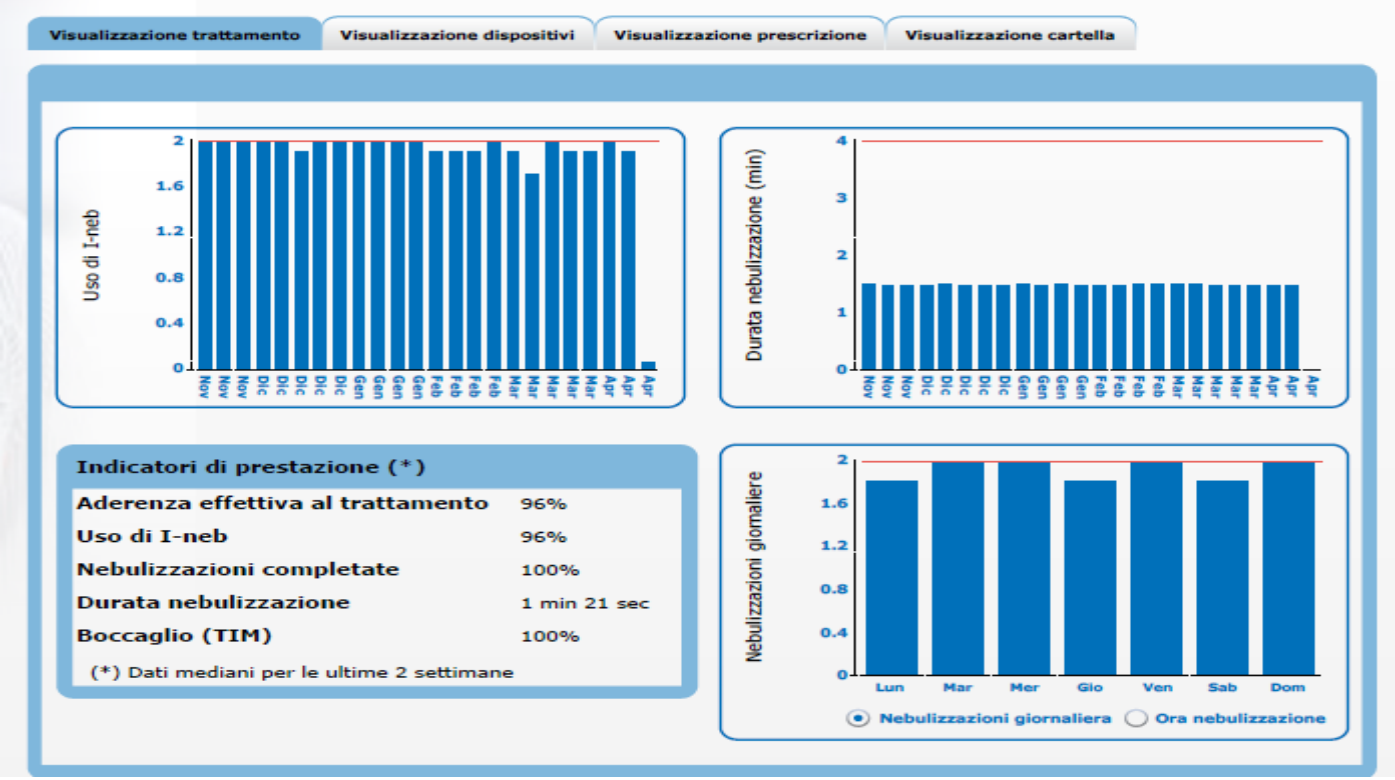
Contents lists available at ScienceDirect

Paediatric Respiratory Reviews

Moving cystic fibrosis care from rescue to prevention by embedding adherence measurement in routine care

Martin J. Wildman [a,b,*], Zhe Hui Hoo [a]

[a] Sheffield Adult Cystic Fibrosis Unit, Sheffield Teaching Hospitals
[b] Health Services Research Unit, School for Health and Related Research (ScHARR), University of Sheffield



Electronic data capture is now considered the ‘gold standard’ for quantifying adherence *

ELSEVIER

Journal of Cystic Fibrosis 14 (2015) 621–626

Journal of Cystic Fibrosis
www.elsevier.com/locate/jcf

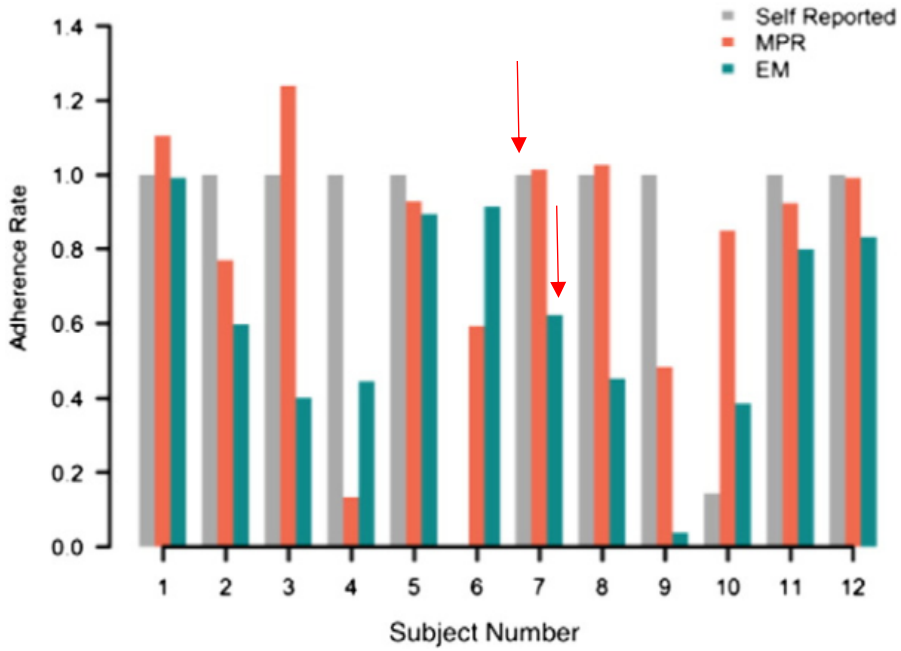
Original Article

Electronic monitoring reveals highly variable adherence patterns in patients prescribed ivacaftor☆,☆☆,★

Christopher M. Siracusa [a,*], Jamie Ryan [b], Lisa Burns [a], Yu Wang [c], Nanhua Zhang [c], John P. Clancy [a], Dennis Drotar [b]

Adherence rates by measurement method (n = 12).

Measurement	Mean (SD)	Range
Monitoring duration, days	117.5 (35.2)	35–182
Self-report	100% [a]	14–100%
Pharmacy refill history (MPR)	84% (31)	13–124%
Electronic monitoring (EM)	61% (28)	4–99%



Interventi per semplificare i tempi di cura e minimizzare i tempi di disinfezione delle apparecchiature

THE CHALLENGES

"multi- disciplinary interventions"

TIME CONSUMING

SATISFACTION OF THERAPIES, SELF-MONITORING

ACCEPTANCE OF THE UTILITY OF THERAPY

NEW NEBULIZER TECHNOLOGY

INVOLVING THE ADOLESCENT IN PROBLEM-SOLVING

SETTING REALISTIC GOALS

SELF MONITORING with new technology

TAILORING THE REGIMEN

SOCIAL SUPPORT

PARENT TRAINING

FAMILY THERAPY

COGNITIVE BEHAVIORAL THERAPY **……**

Haynes et al. Cochrane Database of Systemic Reviews 2008; Kahana et al. J Pediatr Psychol. 2008:590-611

ELSEVIER

Journal of Cystic Fibrosis 8 (2009) 295–315

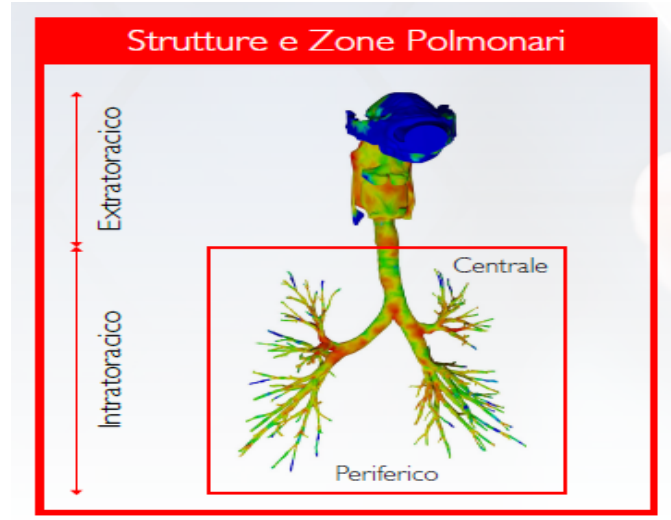
Journal of Cystic Fibrosis
www.elsevier.com/locate/jcf

Inhaled medication and inhalation devices for lung disease in patients with cystic fibrosis: A European consensus ☆

Harry Heijerman *, Elsbeth Westerman, Steven Conway, Daan Touw
Gerd Döring for the consensus working group [1]

Haga Teaching Hospital, Department of Pulmonology, Leyweg 275, 2545 CH The Hague, The Netherlands

Received 31 January 2009; received in revised form 5 April 2009; accepted 8 April 2009
Available online 25 June 2009



DEPOSIZIONE POLMONARE				
NEBULIZZATORI TRADIZIONALI	PARI LC PLUS	eFLOW	DPI	I-NEB AAD
3-8%	9-15%	32%	10-20%	**63-73%**

I-NEB
* TBM: Tidal Breathing Mode – (tempo di nebulizzazione 4,7 minuti)
** **TIM: Target Inhalation Mode - – (tempo di nebulizzazione 3,00 minuti)**

Daniels T, et al., COCHRANE 2013

INFECTION CONTROL AND HOSPITAL EPIDEMIOLOGY AUGUST 2014, VOL. 35, NO. S1

CYSTIC FIBROSIS FOUNDATION GUIDELINE

Infection Prevention and Control Guideline for Cystic Fibrosis: 2013 Update

CAMBRIDGE UNIVERSITY PRESS

SHEA The Society for Healthcare Epidemiology of America

PULIZIA E DISINFEZIONE DEL MATERIALE PER LA FISIOTERAPIA E L'AEROSOLTERAPIA da eseguire il prima possibile dopo ogni utilizzo

PULIZIA	**con detersivo per piatti e** - **acqua di rubinetto** - **acqua di pozzo che rispetta gli standard locali di salute pubblica** - **acqua distillata** - **acqua in bottiglia** **se la pulizia è seguita da disinfezione**

DISINFEZIONE

DISINFEZIONE A FREDDO	- immergere in alcool etilico o isopropilico al 70% - 90% per 5 minuti - immergere in perossido di idrogeno al 3% (10 volumi) per 30 minuti RISCIACQUO del disinfettante: acqua sterile (NO acqua del rubinetto) RISCIACQUO finale: acqua sterile o filtrata (filtro ≤0.2 micron) ASCIUGATURA all'aria

DISINFEZIONE A CALDO - acqua di rubinetto - acqua di pozzo che rispetta gli standard locali di salute pubblica - acqua distillata - acqua in bottiglia	- bollitura per 5 minuti (immergere il materiale nell'acqua bollente) - in sterilizzatore a vapore elettrico - in forno a microonde (2.45 Ghz - il materiale deve essere posto in un contenitore per microonde ed immerso in acqua) per 5 minuti - in lavastoviglie con programma con temperatura ≥70°C per 30 minuti

Source: *Infection Control and Hospital Epidemiology*, Vol. 35, No. S1, Cystic Fibrosis Foundation Guideline (August 2014), pp. S1-S67

Attenzione

- l'acqua di rubinetto e l'acqua distillata possono essere contaminate con germi patogeni per le persone con Fibrosi Cistica

- l'acqua sterile può essere preparata bollendo l'acqua del rubinetto per 5 minuti
- l'acqua sterile può diventare contaminata dopo l'uso e/o la conservazione, ma la frequenza di tale evenienza è sconosciuta: bollire l'acqua immediatamente prima dell'utilizzo minimizza questa possibilità

- sconsigliata la disinfezione con acido acetico (aceto), soluzioni a base di candeggina, benzalconio cloruro

Inhaled versus nebulised antibiotics in Cystic Fibrosis

Respiratory Medicine 138 (2018) 88–94

Contents lists available at ScienceDirect

Respiratory Medicine

journal homepage: www.elsevier.com/locate/rmed

Treatment compliance in cystic fibrosis patients with chronic *Pseudomonas aeruginosa* infection treated with tobramycin inhalation powder: The FREE study

Francesco Blasi[a,*], Vincenzo Carnovale[b], Giuseppe Cimino[c], Vincenzina Lucidi[d], Donatello Salvatore[e], Barbara Messore[f], Marta Bartezaghi[g], Elisa Muscianisi[g], Pasquale Alberto Porpiglia[g], on behalf of the FREE study group

REVIEW
CYSTIC FIBROSIS

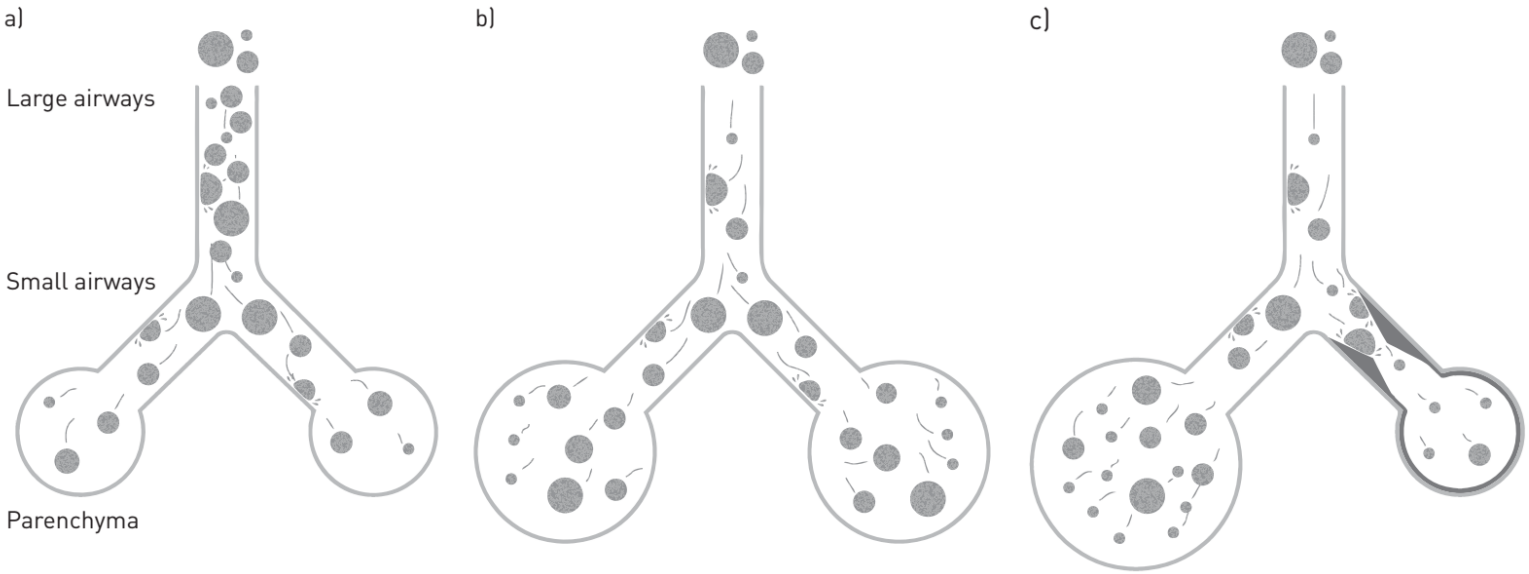
Systematic review of the dry powder inhalers colistimethate sodium and tobramycin in cystic fibrosis

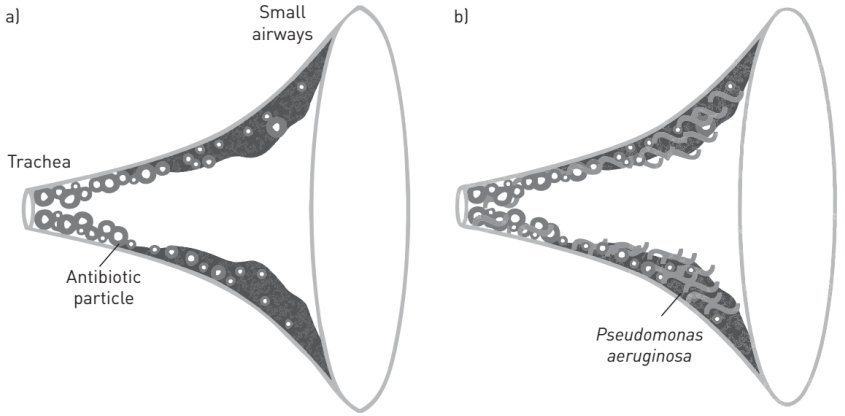
Lesley Uttley[1], Sue Harnan[1], Anna Cantrell[1], Chris Taylor[2], Martin Walshaw[3], Keith Brownlee[4] and Paul Tappenden[1]

REVIEW
INHALED ANTIBIOTICS

Inhaled antibiotics: dry or wet?

Harm A.W.M. Tiddens[1,2], Aukje C. Bos[1,2], Johan W. Mouton[3], Sunalene Devadason[4] and Hettie M. Janssens[1]





Eur Respir Journal 2014

ELSEVIER

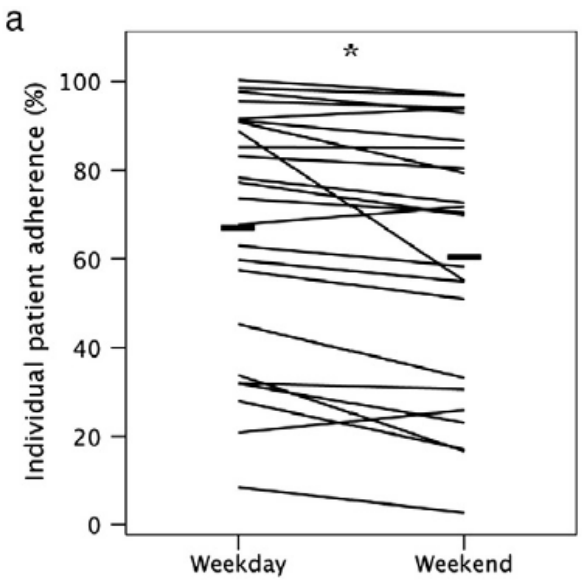
Journal of Cystic Fibrosis 12 (2013) 440–444

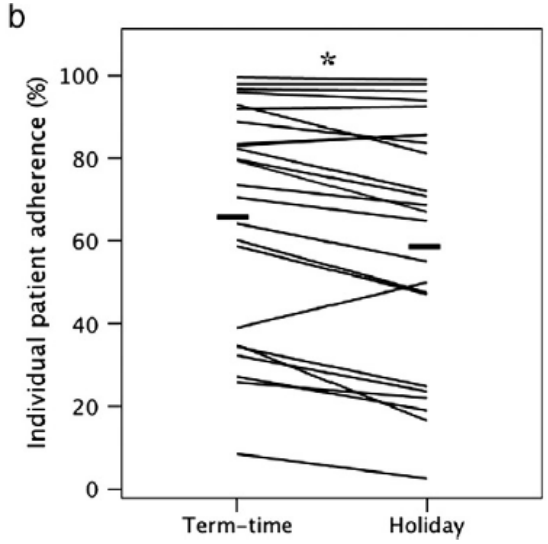
Journal of Cystic Fibrosis
www.elsevier.com/locate/jcf

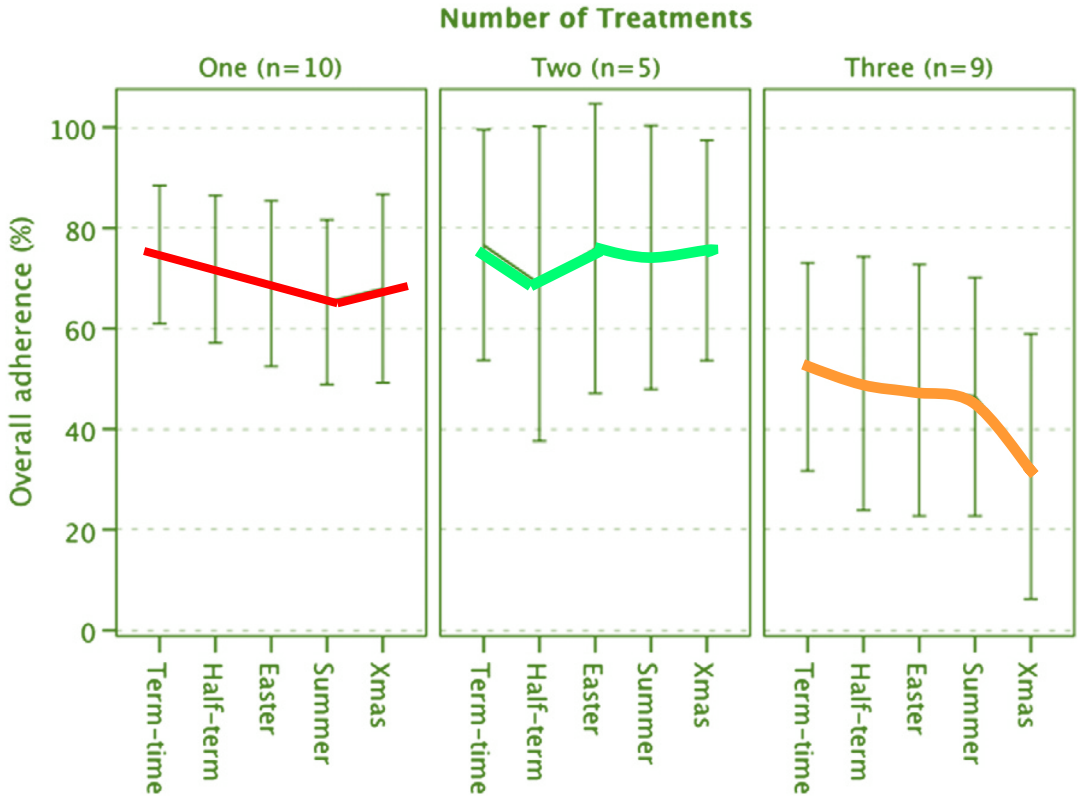
Original Article

Adherence to nebulised therapies in adolescents with cystic fibrosis is best on week-days during school term-time

Rosemary Ball [a, 1], Kevin W. Southern [b, 1], Pamela McCormack [b], Alistair J.A. Duff [a], Keith G. Brownlee [a], Paul S. McNamara [b,*]







Determinants of adherence in adults with cystic fibrosis

L J Kettler, S M Sawyer, H R Winefield, H W Greville

Thorax 2002;**57**:459–464

tion. In addition to the complexity and number of treatments prescribed for adults with CF, adults living with this condition are faced with the challenge of interpreting and understanding the effects and priority of each of these treatments within their own treatment regime. It is not surprising that patients find this difficult, given the lack of consensus among treating physicians and multidisciplinary health care teams about which treatments are most important.[7 30]

People with CF often develop strong relationships with their multidisciplinary health teams. These relationships are usually developed over many years of treatment; CF is one of only a handful of chronic illnesses in which such long term treatment relationships may be developed. This feature of health care in CF will impact (both positively and negatively) on the communication between the health care professional and the patient.[30] Such a relationship may enhance the clarity

- ✓ Consesus among treating physicians and multidisciplinary health care teams
- ✓ Long term treatment relathionships will impact (both positively and negatively) on the communication between health care professional and the patient.

ORIGINAL ARTICLE

Effects of hypertonic saline, alternate day and daily rhDNase on healthcare use, costs and outcomes in children with cystic fibrosis

R Suri, R Grieve, C Normand, C Metcalfe, S Thompson, C Wallis, A Bush

Thorax 2002;**57**:841–846

Effectiveness

Following 12 weeks of treatment there was a mean (SD) increase in FEV_1 from baseline of 16 (25)% in patients receiving daily rhDNase, 14 (23)% for alternate day rhDNase, and 3 (21)% for HS. Comparing mean FEV_1 between the treatments, there was an advantage of 8% (95% CI 2 to 14, p=0.01) for daily rhDNase over HS but none for daily compared with alternate day rhDNase (2%, 95% CI –4 to 9, p=0.55). There was

- ✓ Daily rhDNase vs HS = FEV1 + 8%
- ✓ No difference between daily vs alternate day rhDNase

Table 4 Mean (%) costs (£) and percentages of total costs over each 12 week treatment period

	Daily rhDNase *v* hypertonic saline		Daily rhDNase *v* alternate day rhDNase	
	Daily rhDNase (n=40)	Hypertonic saline (n=40)	Daily rhDNase (n=43)	Alternate day rhDNase (n=43)
Grand total	5694	4285	5711	5198
Mean (95% CI) difference	1409 (440 to 2318)		513 (–546 to 1510)	

STATE OF THE ART

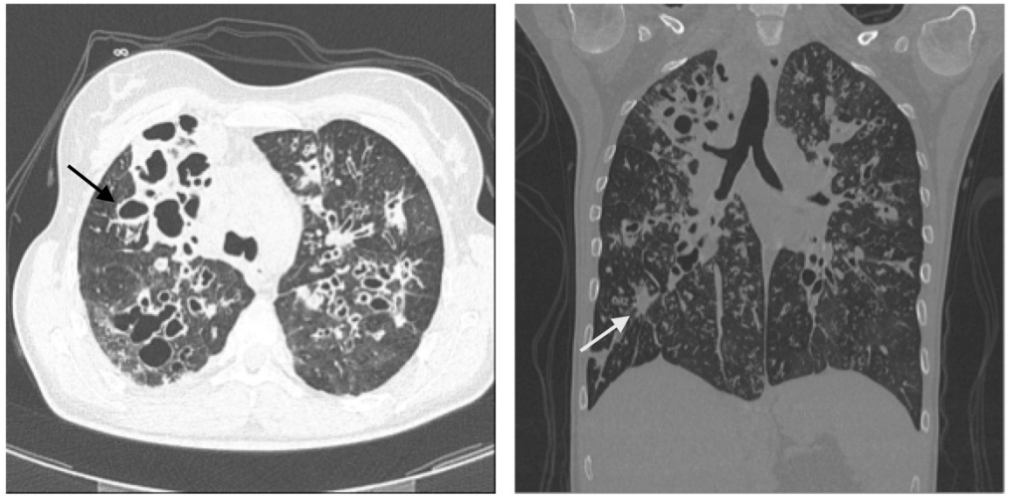
WILEY

Aerosolized agents for airway clearance in cystic fibrosis

Kevin W. Southern MD, PhD[1] | John P. Clancy MD[2] | Sarath Ranganathan MD, PhD[3]

Spiegare il razionale per una terapia aerosolica a lungo termine e promuovere l'aderenza

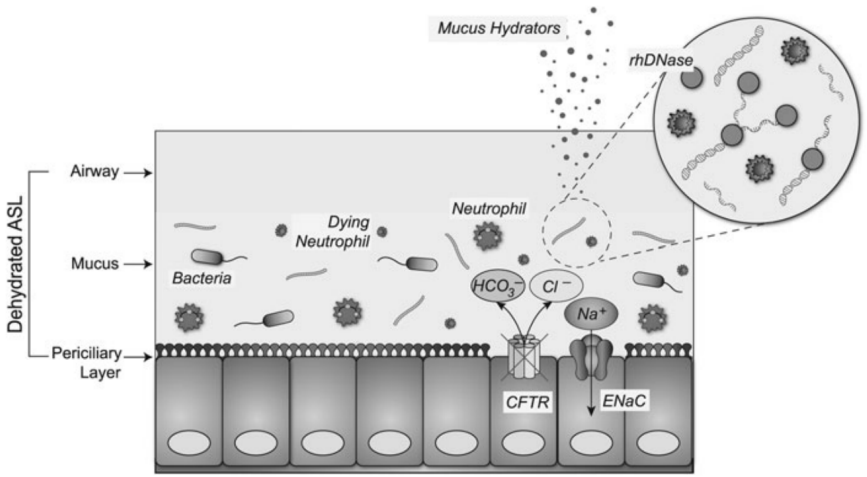
- ✓ Chiara ed aperta discussione con il paziente e la sua famiglia
- ✓ Coinvolgere il paziente nella decisione da prendere
- ✓ Utilizzare immagini visive (RX torace, HRCT)
- ✓ Utilizzare registrazioni broncoscopie



Pediatric Pulmonology 2018

Factors affecting nebulised medicine adherence in adult patients with cystic fibrosis: a qualitative study

Alice Hogan · Mary-Ann Bonney · Jo-anne Brien · Rita Karamy · Parisa Aslani



"Something that makes it go in faster. Whether that means some kind of explosive puffers that you know, blasts it into you, or you know, a nebuliser that goes faster, or just so, some kind of fast delivery system."

"Oh, he [partner] always encourages me. Especially if we, you know, come home late at night, and I say "I'm too tired, I'm too tired." And he'll say "Go in there, do it. I'll come and sit with you or watch TV."

Int J Clin Pharm 2015

Original Paper

Developing Digital Games to Address Airway Clearance Therapy in Children With Cystic Fibrosis: Participatory Design Process

Fabio Balli[1,2], GradD, MAS

Figure 7. The visual of the game Globule: breathing exercise to collect arrows (left), and puzzle (right). Credit: breathinggames.net ,e game Globule. Credit: breathinggames.net.

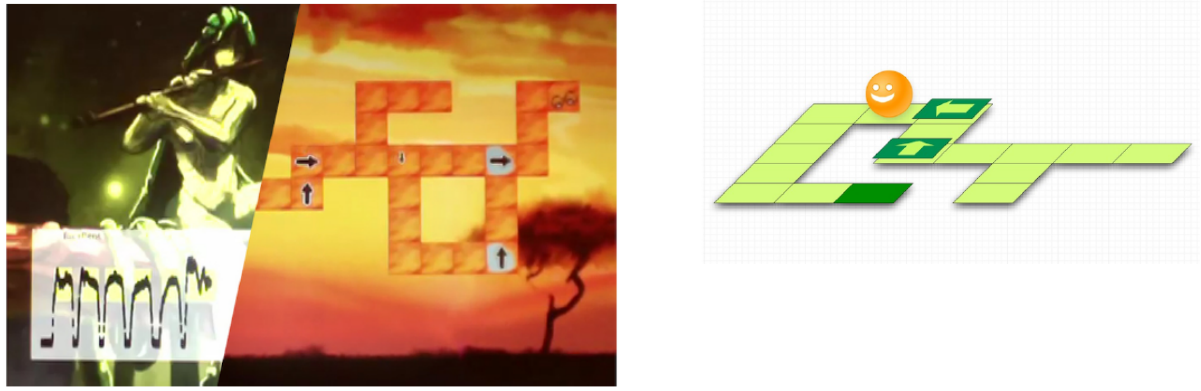


Figure 9. Visuals for the game Ange-Gardien. Credit: breathinggames.net / J Danger.

Figure 13. A visual universe of the game Heritage: breathing exercise. Credit: breathinggames.net / C Skrapits / N Wenk

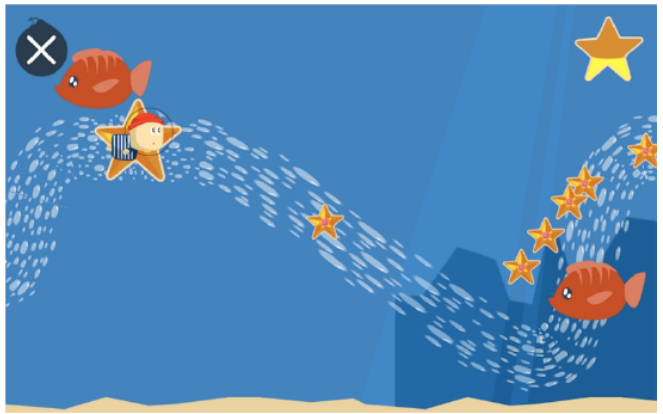
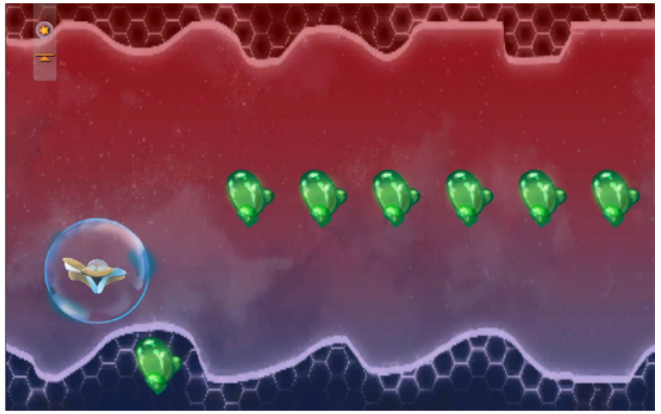


Figure 11. A visual universe of the game PEP Hero. Credit: breathinggames.net / K Berthiaume.

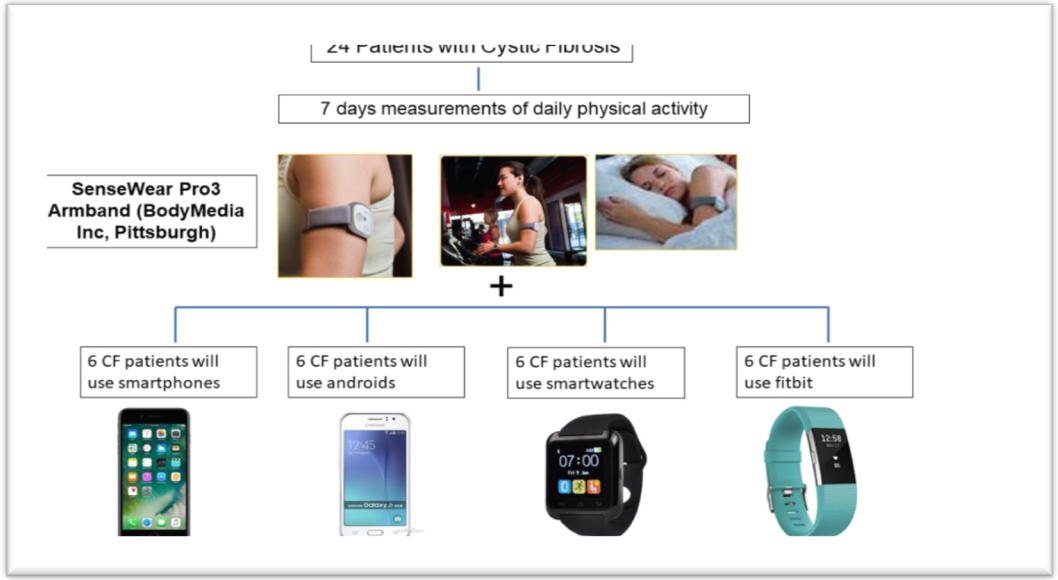


JMIR 2018

Different methods of measuring physical activity in cystic fibrosis: accelerometer versus new electronic devices

Physical activity, Cystic fibrosis, Quality of life

D. Savi[1], L. Graziano[1], S. Schiavetto[1], N. J. Simmonds[2], B. Giordani[3], E. Leggieri[1], M. Rivolta[1], D. Asciutti[1], P. Palange[1], J. S. Elborn[2]



Data presented at the European Cystic Fibrosis Conference, June 2019, Liverpool (UK)

Mobile Phone Apps to Improve Medication Adherence: A Systematic Stepwise Process to Identify High-Quality Apps Monitoring

JMIR Publications
JMIR mHealth and uHealth
ISSN 2291-5222
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To identify high-quality apps to be tested in a future study that will provide evidence on the use of medication reminder apps to improve medication adherence.

GRAZIE !