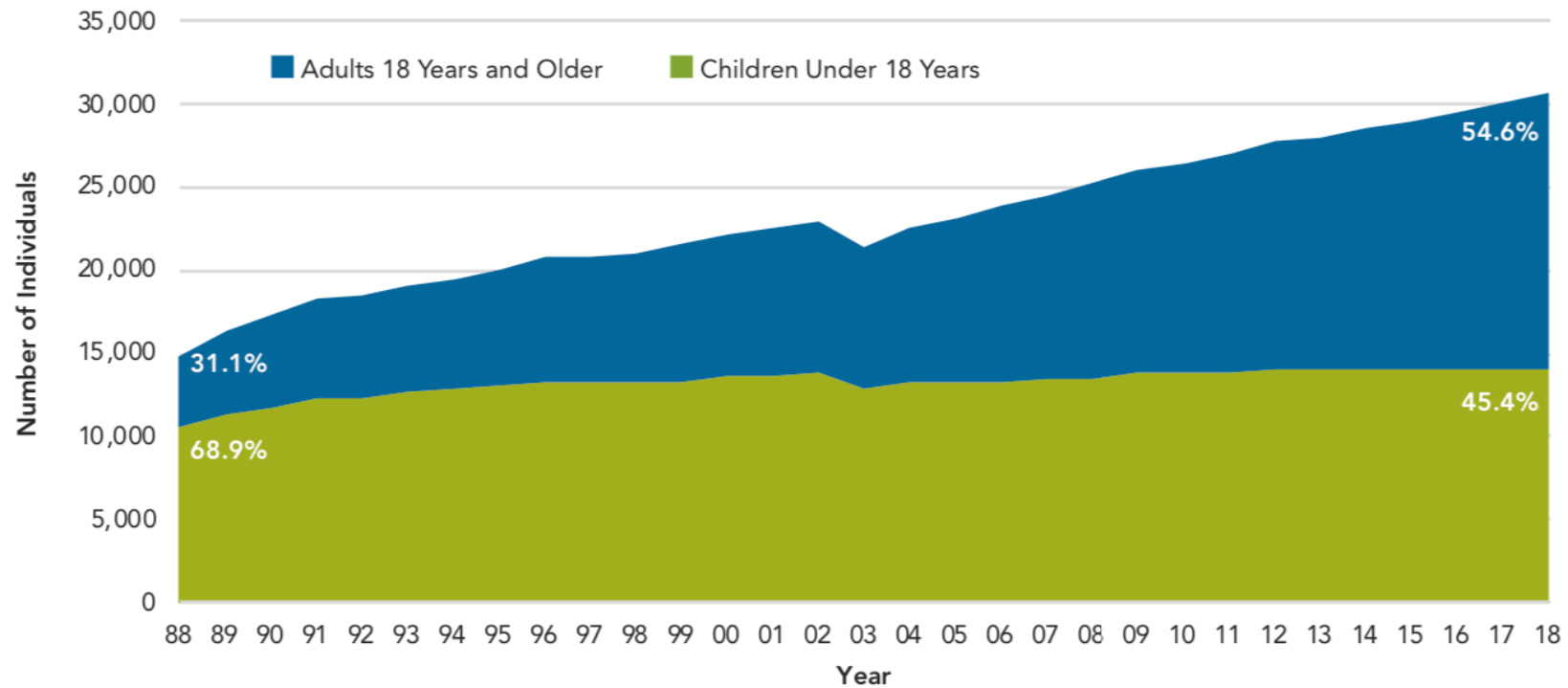




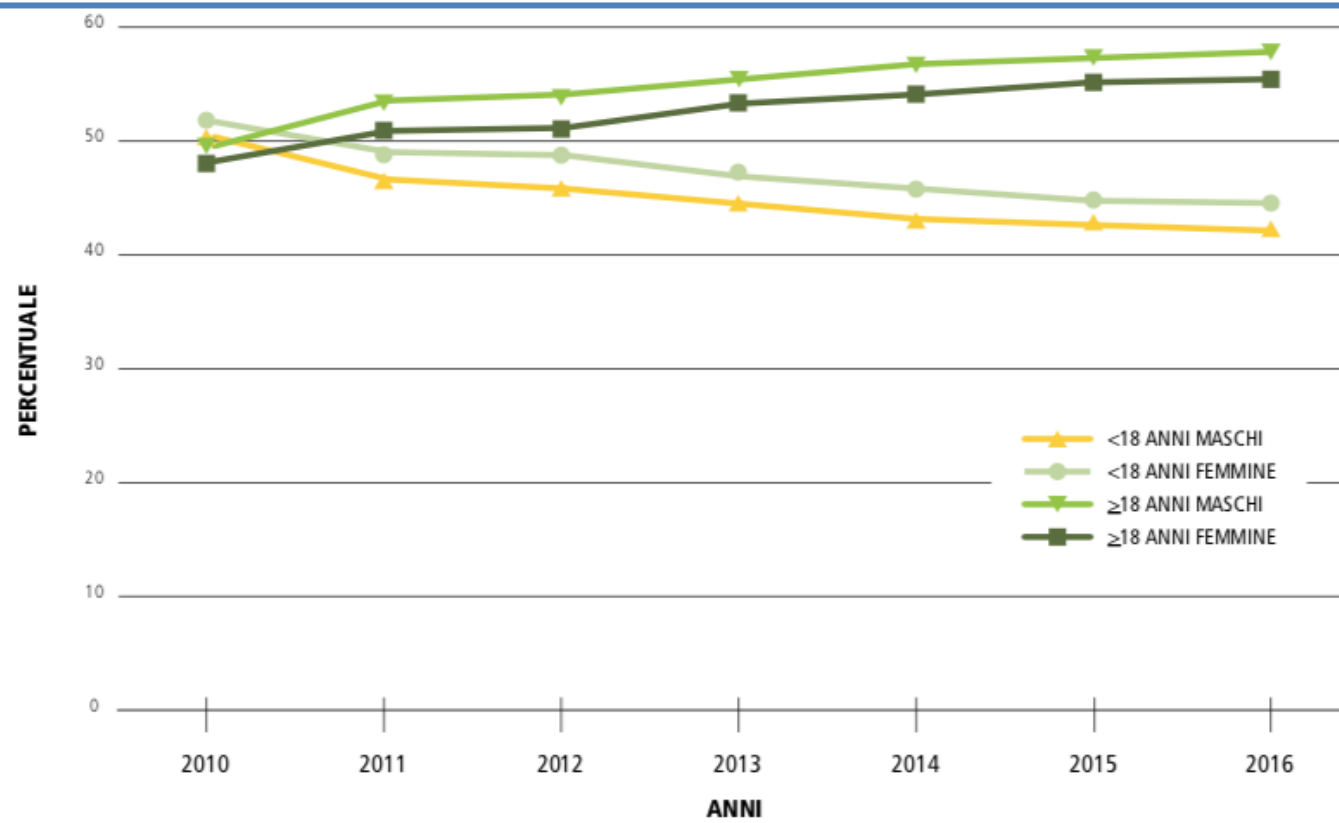
FIBROSI CISTICA DELL'ADULTO

*Dott. Vincenzo Carnovale
Università degli Studi di Napoli "Federico II"
CRR Fibrosi Cistica dell'Adulto
vincenzo.carnovale@unina.it*

Number of Children and Adults with CF, 1988–2018



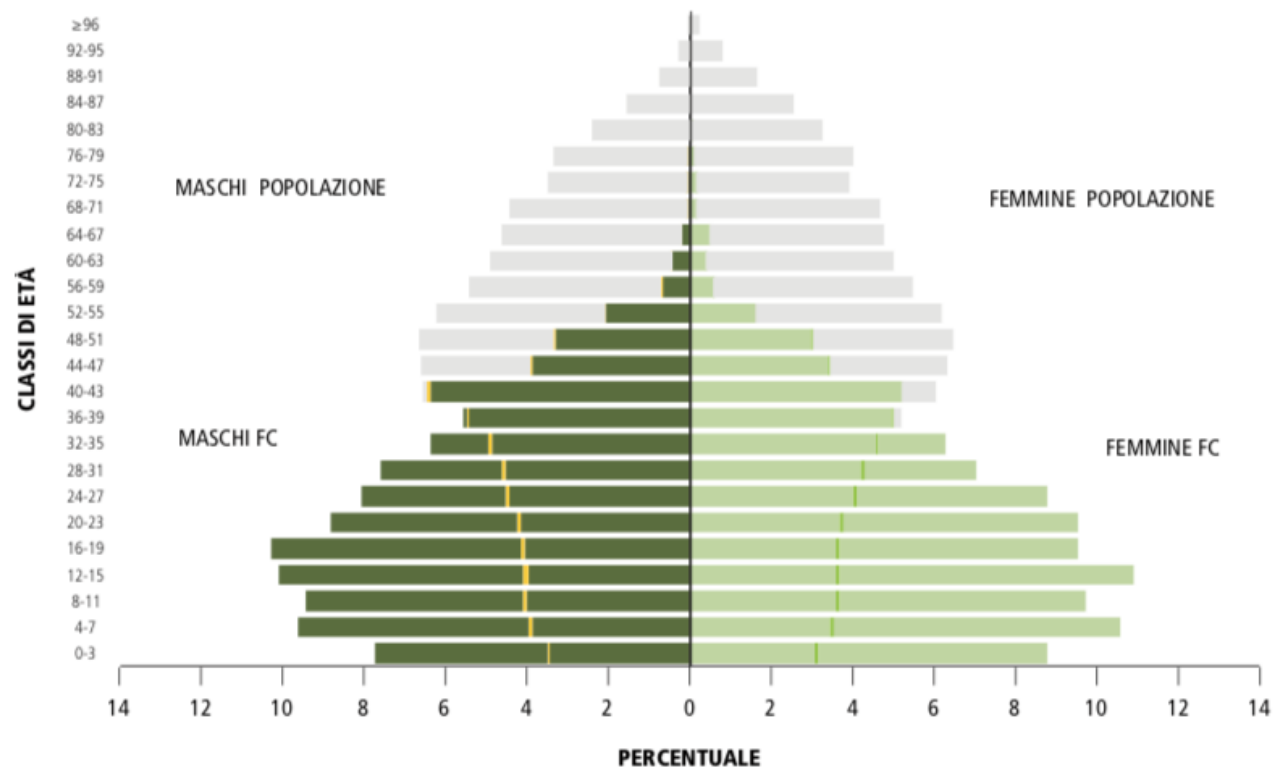
Distribuzione dei pazienti secondo gruppi di età, sesso e anno. Anni 2010-2016.



Registro Italiano Fibrosi Cistica, 2016

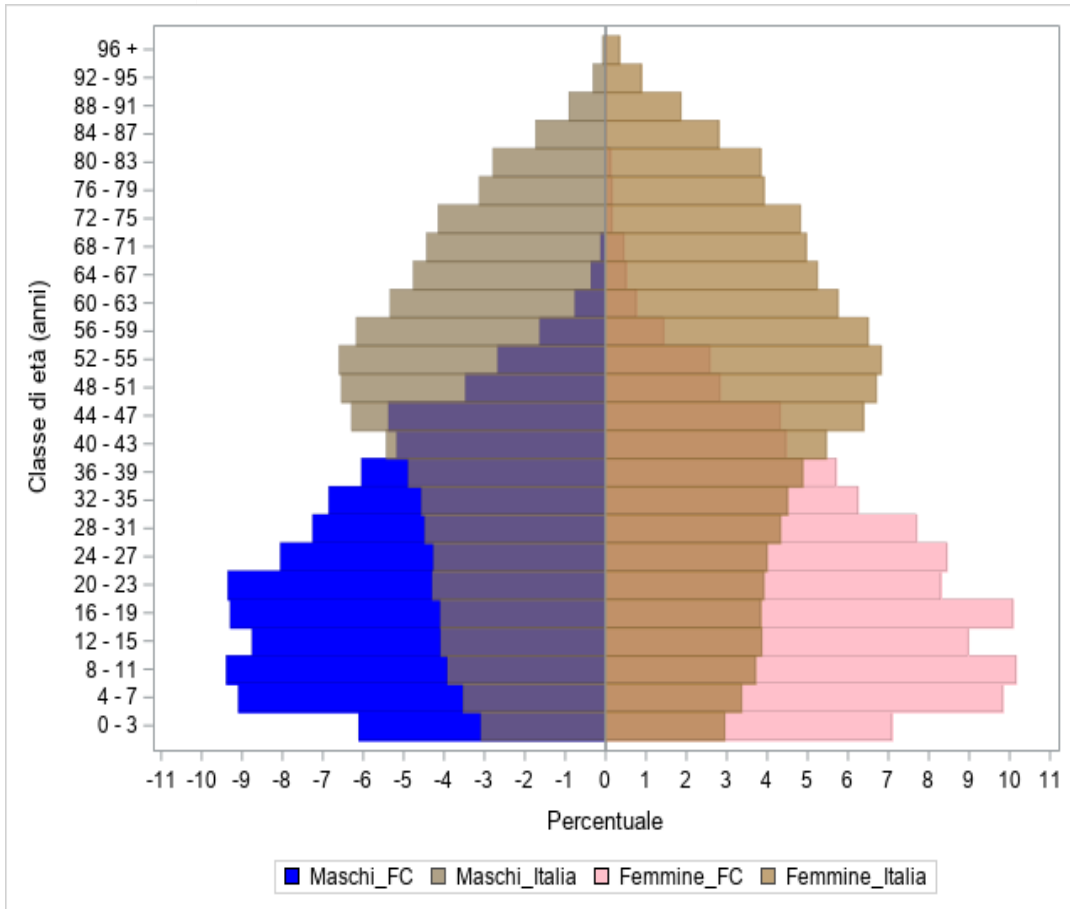


Piramide dell'età dei pazienti FC vs popolazione italiana. Anno 2016.



Registro Italiano Fibrosi Cistica, 2016

Piramide dell'età dei pazienti FC vs. popolazione italiana - Anno 2020



- Per le classi di età più giovani la quota dei pazienti FC è maggiore; l'invecchiamento della popolazione italiana risulta invece evidente osservando l'elevata quota delle classi di età più anziane.
- Come negli anni precedenti, si conferma la presenza della popolazione FC nella sesta e settima decade di vita.

Fonte: Istat 2020: Demo Istat.

Future-Trend in Fibrosi Cistica dal 2010–2025

Country	Children age ≤17 years				Adults age ≥18 years				Total			
	2010	2015	2020	2025	2010	2015	2020	2025	2010	2015	2020	2025
Austria	361	393	419	433	344	440	530	612	705	833	949	1045
Switzerland	294	320	341	353	281	360	433	500	575	680	774	853
Cyprus	19	21	22	23	18	23	28	32	37	44	50	55
Spain	1311	1429	1521	1573	1250	1600	1925	2225	2561	3029	3446	3798
Finland	22	24	28	40	22	41	69	57	65	77	87	97
Greece									589	697	793	874
Italy →									5258	6219	7075	7798
Luxembourg									22	26	30	33
Malta									23	27	31	34
Portugal	147	160	171	176	140	179	216	249	287	339	387	425
All in group	5180	5645	6010	6215	4942	6326	7612	8797	10 122	11 971	13 622	15 012

Incremento pazienti
pediatrici del 20 %

Incremento pazienti
adulti del 77 %

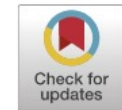
Assumptions on population dynamics based on observed dynamics of countries within group A: increase by 9% in children and by 28% in adults between 2010 and 2015; increase by 16% in children and by 54% in adults between 2010 and 2020; increase by 20% in children and by 78% in adults between 2010 and 2025.



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Respiratory Medicine

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Effectiveness and safety of elexacaftor/tezacaftor/ivacaftor in patients with cystic fibrosis and advanced lung disease with the *Phe508del*/minimal function genotype

Vincenzo Carnovale^{a,1,*}, Paola Iacotucci^{a,1}, Vito Terlizzi^{b,1}, Carmela Colangelo^c,
Pietro Medio^a, Lorenza Ferrillo^a, Fabiola De Gregorio^c, Michela Francalanci^b,
Giovanni Taccetti^b, Serena Buonauro^a, Marcella d'Ippolito^a, Giovanni Marsicovetere^c,
Michele D'Andria^c, Nicola Ferrara^a, Donatello Salvatore^c

^a Department of Translational Medical Science, Cystic Fibrosis Center, Adult Unit, University of Naples "Federico II", Italy

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^c Cystic Fibrosis Center, Hospital San Carlo, Potenza, Italy

Elexacaftor/Tezacaftor/Ivacaftor in Patients with Cystic Fibrosis Homozygous for the *F508del* Mutation and Advanced Lung Disease: A 48-Week Observational Study

Vincenzo Carnovale ^{1,*†}, Paola Iacotucci ^{1,†}, Vito Terlizzi ^{2,†}, Carmela Colangelo ³, Lorenza Ferrillo ¹, Angela Pepe ^{3,4}, Michela Francalanci ², Giovanni Taccetti ², Serena Buonauro ¹, Assunta Celardo ¹, Laura Salvadori ¹, Giovanni Marsicovetere ³, Michele D'Andria ³, Nicola Ferrara ¹ and Donatello Salvatore ^{3,†}



Journal of
Clinical Medicine

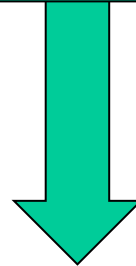
2022, 11, 1021.

<https://doi.org/10.3390/jcm11041021>

Sostenere una transizione efficace

La transizione delle cure ha un'accezione ampia che vale per gli adolescenti e i giovani adulti

“Il passaggio mirato, pianificato, di adolescenti e giovani adulti affetti da condizioni fisiche e mediche croniche dai sistemi sanitari per bambini a quelli per adulti”



Un ***processo graduale e in evoluzione*** al fine di garantire la prestazione di cure ininterrotte, coordinate, adeguate in termini di sviluppo e complete ***prima, durante e dopo il periodo di transizione***

Blum RWM, et al. *J Adolesc Health*. 1993;14(7):570–6.

Gli obiettivi del processo di transizione mirano a sostenere lo sviluppo di ciascun paziente

1

Per **offrire delle cure mediche di qualità e adeguate in termini di età e di sviluppo** che siano flessibili e responsive^{1,2}

2

Per **favorire lo sviluppo di abilità comunicative e decisionali** al fine di mantenere l'assertività e la difesa dei propri interessi^{1,2}

3

Per **garantire un senso di controllo e indipendenza** nella vita sanitaria e personale di un paziente¹

4

Per **massimizzare il funzionamento permanente** e il potenziale¹

Per una transizione efficace è necessario superare potenziali sfide poste da diverse parti interessate¹⁻⁵

Paziente

- Grado di preparazione alla transizione
- Ansia relativa all'abbandono del team familiare pediatrico
- Preoccupazioni in merito alla mancanza di sostegno da parte dei servizi per adulti
- Le consuete sfide fisiche e psicosociali dell'adolescenza

Genitore/ assistente sanitario

- Rinunciare al controllo, passare da un ruolo "di controllo" a un ruolo "consultivo"
- Preoccupazioni relative all'assicurazione sanitaria
- Mancanza di supporto

Centro pediatrico

- Riluttanza a effettuare la transizione per paura di perdere il "legame speciale" instauratosi con i pazienti
- Preoccupazione che i servizi per adulti non siano in grado di prestare cure simili
- Mancanza di dati relativi agli esiti sanitari post-transizione

Centro per adulti

- Mancanza di opportuni servizi per adulti dotati di risorse adeguate
- Mancanza di una comunicazione regolare con il team di cure pediatriche
- Limitata documentazione relativa all'anamnesi sanitaria e alle complicazioni correlate alla FC
- Differenze logistiche tra la prassi pediatrica e quella per adulti



Il processo di transizione può essere composto da quattro componenti fondamentali

Coordinato ¹⁻³	Collaborativo ^{3,4}	Responsivo ^{3,4}	Valutato ⁴
• Utilizza un approccio graduale, coordinato, pluridimensionale, che si focalizza sulla famiglia • Affronta le comuni preoccupazioni di pazienti e genitori • Promuove l'autonomia e fornisce la flessibilità per soddisfare le esigenze individuali e familiari	• Comporta il legame tra il team di cure pediatriche e quello per adulti per la FC • Incoraggia la responsabilità congiunta dei team	• Si verifica in risposta al raggiungimento da parte del paziente di tappe di sviluppo prestabilite • Garantisce che i pazienti abbiano risorse adeguate per preservare lo stato di salute e accedere alle terapie in un contesto adulto	• Utilizza un programma valutato regolarmente • Assicura esiti del paziente sostenibili



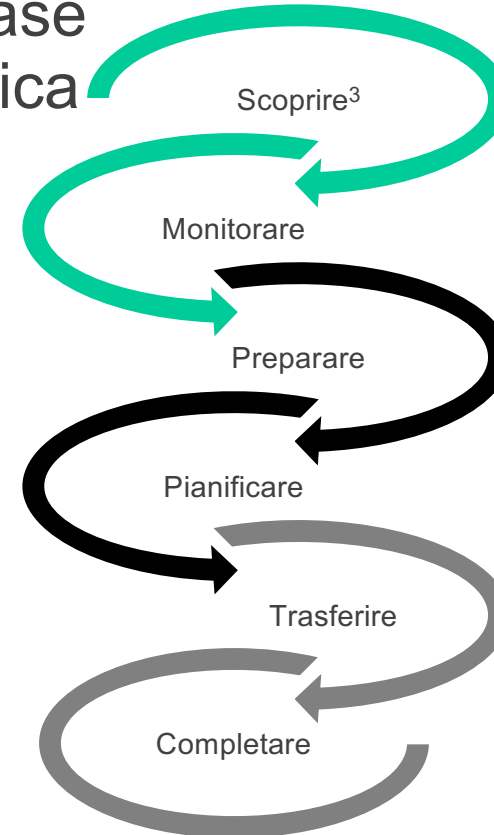
L'età ottimale per le diverse fasi del processo di transizione deve essere personalizzata

Il “grado di preparazione” alla transizione è flessibile e varia in base all'età cronologica e la maturità fisica e cognitiva dell'adolescente^{1,2}

Adolescenza →

Tarda adolescenza →

Prima età adulta →



Un'analisi retrospettiva statunitense dimostra che la transizione comporta un declino meno rapido della funzione polmonare

I pazienti che hanno portato a termine la transizione alle cure per adulti sono stati messi a confronto con quelli rimasti nelle cure pediatriche

Analisi retrospettiva del Database del Registro FC (1997–2007)

661 pazienti assegnati a ciascun gruppo mediante un'analisi di matching che sfrutta il propensity score (1:1)

Caratteristiche generali ed età simili

Misure di esito	Modifica iniziale (1 anno dopo la transizione)		Tasso di variazione (2 anni dopo la transizione)	
	Stima (Intervallo di confidenza [CI] al 95%)	Valore p	Stima (Intervallo di confidenza [CI] al 95%)	Valore p
% predetta di FEV ₁	0,76 (−0,30–1,83)	NS	0,78 (0,06–1,51)	0,035
% predetta di CVF	1,40 (0,40–2,40)	0,006	0,45 (−0,18–1,08)	NS
Indice di massa corporea (IMC)	0,22 (0,04–0,39)	0,015	0,05 (−0,06–0,16)	NS
Numero di ricoveri	−0,04 (−0,15–0,08)	NS	−0,02 (−0,10–0,06)	NS
Numero di eventi con antibiotici EV a casa	0,08 (−0,03–0,19)	NS	0,04 (−0,03– 0,12)	NS

La transizione ha portato a un declino meno rapido della funzione polmonare e non influisce sugli esiti sanitari a breve termine per i pazienti con FC

Telemedicina

ePS1.07

Patient experience of virtual consultations: survey results

H. Parrott^{1,2}, L. Funnell², L. O'Connor², L. Bosaeus¹, U. Bertello¹, S. Madge².
¹NuvoAir AB, Stockholm, Sweden; ²Royal Brompton and Harefield NHS Trust, Department of Adult Cystic Fibrosis, London, United Kingdom

Introduction: Digital health tools have the potential to support the care of a growing adult CF population. In addition, virtual consultations paired with home spirometry assessment could reduce the burden and cross-infection risk associated with clinic visits.

Aim: To evaluate patient-reported perceptions of healthcare utilisation and acceptability of NuvoAir technology in patients who have taken part in a virtual consultation service for more than 6 months (the service commenced in 2017). The NuvoAir platform consists of a smartphone application and Bluetooth spirometer, with a healthcare practitioner portal allowing patients to share data with their healthcare team.

Methods: A survey was emailed to eligible patients, including questions on patient-reported healthcare utilisation, use of the technology and understanding of their CF. All patients provided consent.

Results: Response rate: 41/67 (61%); males 46.3%, mean age 37.3 years (SD 10.2), mean FEV₁ 57.3% predicted (range 24.2–103.0). Length of time using the virtual consultation service: mean 13.0 months (range 6.9–25.8). The virtual service reduced the number of booked face-to-face consultations by 30.9% (from mean 5.4 to 3.7 visits/year; $p < 0.001$) and the number of urgent face-to-face consultations by 39.8% (from 2.2 to 1.3 visits/year; $p < 0.01$). A total of 18/41 (43.9%) patients felt that they understood their CF better since starting the virtual service (23 [56.1%] unchanged; 0 worse) and 7/41 (17.1%) said their medication adherence improved (34 [82.9%] unchanged; 0 worse). Overall acceptance of virtual consultations was high (mean 9.4/10 [range 7–10]). The numbers of antibiotic courses and hospitalisations were not significantly changed.

Conclusions: These self-reported data show the acceptability of virtual consultations and the potential to substantially reduce overall healthcare utilisation. These results will help plan the service's further development in a larger population and with a longer follow-up period.

E-Posters / Journal of Cystic Fibrosis 19S2 (2020)



OUR SOLUTION

Transforming Remote Care for IPF and CF patients

- NuvoAir has CE certified a best-in-class home spirometer and a mobile application to help patients with respiratory problems to measure their lung function, monitor their disease progression and support physicians to intervene in a timely manner
- In patients with IPF Daily measurements of FVC provide a more sensitive and earlier prediction of disease behavior and outcome than do traditional periodic hospital-based readings¹
- In patients with CF, home spirometry could be used to monitor benefits of IV antibiotics², improve medication adherence³ and detect exacerbations⁴, among other things

¹ Daily Home Spirometry: An Effective Tool for Detecting Progression in Idiopathic Pulmonary Fibrosis. American Thoracic Society. 2019; 100(1):1-10. doi:10.1183/13659033.01001119

² Daily spirometry in an acute exacerbation of adult cystic fibrosis patients. Medical Research - Free Lung - Open Access. 2019; 10(1):1-10. doi:10.1183/13659033.01001119

³ Impact of home spirometry on medication adherence among adolescents with cystic fibrosis. Thorax. 2019; 74(1):1-10. doi:10.1183/13659033.01001119

⁴ Home Monitoring of Patients with Cystic Fibrosis to Identify and Treat Acute Pulmonary Exacerbations. eCF Study Results. American Thoracic Society. 2019; 100(1):1-10. doi:10.1183/13659033.01001119



NuvoAir and the Royal Brompton Hospital transforming Cystic Fibrosis Remote Care

ARIA FOR HOME-USE

Having lung health as cornerstone of the solution

The Core

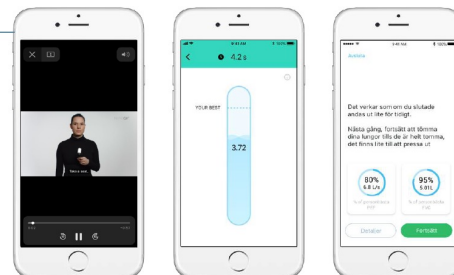
Best-in-class user experience for spirometry with clear instructions and world-leading intelligent feedback.
Data collected: FVC, FEV1, PEF, flow-volume-curve and more

Quality assurance

Intelligent error handling and session grading

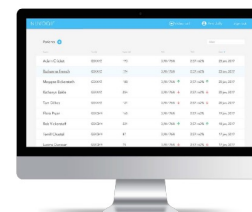
Simplified spirometry

through cloud based intelligence on historic values



NUVOAIR
by FIBROSIS

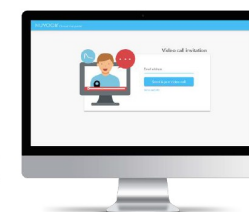
DOCTOR'S TELEMEDICINE PORTAL - Available for critical care units of hospitals



Doctors can access an **overview of the results of their patients** - if these decide to share the data with them, remotely, via the cloud

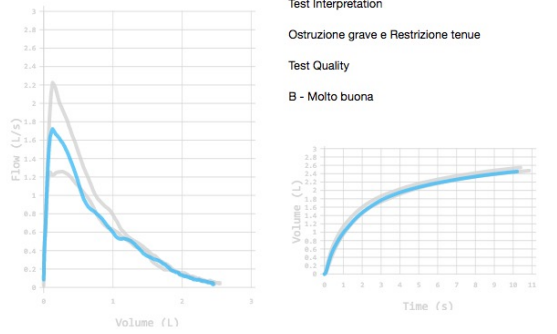


The portal can be accessed via a log-in secured website and provides the **caregiver access to all individual tests**

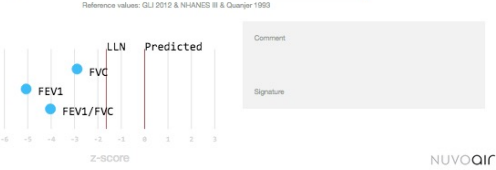


The portal allows the patients to set up a video call with the patient to initiate a **remote consultation**

NUVOAIR
by FIBROSIS

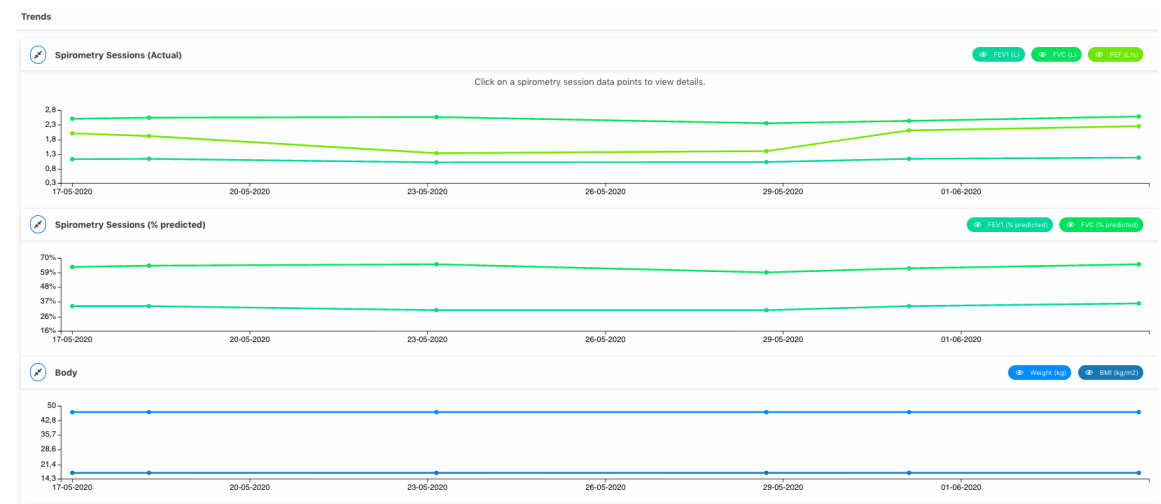


	Act	Pred	%Pred	LLN	z-score	1st best	2nd best	3rd best
FVC (L)	2.55	3.00	85	3.12	-2.89	2.46	2.55	2.48
FEV1 (L)	1.16	3.25	36	2.80	-5.07	1.02	1.16	0.99
FEV1/FVC	0.45	0.84	54	0.72	-4.03	0.41	0.45	0.40
PEF (L/s)	2.21	6.94	32.0	5.46	-5.25	1.71	2.21	1.26
FEV6 (L)	2.25	3.82	59	3.12	-2.17	2.17	2.25	2.17
MEF25 (L/s)	0.17	1.99	9.0	0.86	-2.83	0.19	0.17	0.18
MEF50 (L/s)	0.52	4.35	12.0	2.55	-3.48	0.52	0.52	0.50
MEF75 (L/s)	1.22	6.06	20.0	3.85	-3.58	0.91	1.22	1.00
FEF2575 (L/s)	0.46	3.83	12.0	2.44	-3.97	0.43	0.46	0.40
Duration Exhale (s)	10.40					10.22	10.40	10.86



Primi mesi di follow up:

- 50 pazienti arruolati;
- 100% aderenza;
- In media sessioni spirometriche eseguite mensilmente.



WS14.3 VIRTUAL CONSULTATION IN CYSTIC FIBROSIS: A FIRST ITALIAN EXPERIENCE

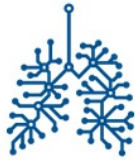
Vincenzo Carnovale¹, Paola Iacotucci¹, Pietro Medio¹, Lorenza Ferrillo¹, Serena Buonauro¹, Marcella d'Ippolito¹, Assunta Celardo¹, Steven Coughlin², Daniela Savi³

¹Department of Translational Medical Science, "Federico II" University of Naples, Italy; ²NuvoAir AB Stockholm, Sweden; ³Department of Public Health and Infectious Diseases, "Sapienza" University of Rome, 00185 Rome, Italy



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NUVOair



SAPIENZA
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ASSESSMENT OF NUVOAIR PLATFORM USE ON CLINICAL OUTCOMES IN ADULTS WITH CYSTIC FIBROSIS: A FIRST ITALIAN EXPERIENCE

Vincenzo Carnovale¹, Paola Iacotucci¹, Pietro Medio¹, Lorenza Ferrillo¹, Serena Buonauro¹, Marcella d'Ippolito¹, Assunta Celardo¹, Steven Coughlin², Daniela Savi³

¹Department of Translational Medical Science, "Federico II" University of Naples, Italy; ²NuvoAir AB Stockholm, Sweden; ³Department of Public Health and Infectious Diseases, Sapienza University of Rome, 00185 Rome, Italy



Long-term clinical impact using a virtual model of care in Cystic Fibrosis

Vincenzo Carnovale^a, Paola Iacotucci^a, Dandi Qiao^b, Lorenza Ferrillo^a, Jolanda Somma^a, Serena Buonauro^a, Marcella d'Ippolito^a, Assunta Celardo^a, Daniela Savi^c

^a Department of Translational Medical Science, "Federico II" University of Naples, Italy; ^b NuvoAir U.S. Inc, Boston, MA, USA; ^c Department of Public Health and Infectious Diseases, "Sapienza" University of Rome, Italy

BACKGROUND

The opportunity provided by digital technologies for managing chronic respiratory diseases remotely has been explored and tested by health professionals during the coronavirus disease 2019 (COVID-19) pandemic. Further investigations into whether these new approaches to care delivery provide an opportunity to improve Cystic Fibrosis (CF) management are required. The aim of this study was to assess the long-term clinical impact of the application of e-health as part of a virtual model of care in CF.

METHODS

NuvoAir Home platform consists of a smartphone application, Bluetooth spirometer and a clinician portal. Patients were trained to use the platform and asked to do home spirometry monthly. Data on pulmonary function and pulmonary exacerbations were collected at baseline and after twelve months. A survey was emailed to evaluate patients' experience using the technology.

RESULTS

A cohort of 43 CF patients were recruited (26 females; mean age 31.6±6.8; 16 homozygous for delta F508; FEV₁ 48.4±16.3 % predicted). Sustained improvement in FEV1 both expressed as absolute and percentage of predicted value was seen through 12 months (mean change 100 ml, p <0.02 and 3.8% p <0.005, respectively). We found significant improvement in FVC both expressed as absolute value with mean change of 230 ml (p <0.006) and expressed as percentage of predicted with mean change 6.2 % p < 0.002. The average of number of exacerbations per person was 0.84 1-year before vs 0.09 1-year after the use of digital technology, p <0.0001. 90% of patients reported they understood their CF better since starting the virtual care service. No changes in medical treatment were reported during that time.



CONCLUSIONS

The application of digital technologies in the management of adults with CF showed an improvement in lung function and pulmonary exacerbations. Using a virtual model of care was well accepted by CF patients and improved their understanding of their medical condition.



NACF 2022





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Respiratory Medicine

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Original Research

Clinical outcomes of digital health in adults with cystic fibrosis

Vincenzo Carnovale^{a,1}, Paola Iacotucci^{a,1}, Dandi Qiao^b, Lorenza Ferrillo^a, Jolanda Somma^a,
Serena Buonauro^a, Marcella d'Ippolito^a, Assunta Celardo^a, Daniela Savi^{c,*}

^a Department of Translational Medical Science, "Federico II" University of Naples, Italy

^b NuvoAir U.S. Inc, Boston, MA, USA

^c Department of Public Health and Infectious Diseases, "Sapienza" University of Rome, Italy



J16 Modeled Survival Gains and Cost-Effectiveness of Trikafta (Elexacaftor/Tezacaftor/Ivacaftor) in the Treatment of Patients with Cystic Fibrosis in the US

*Lopez A¹, Booth J¹, Bailey E¹, Ektare V², Rubin-Cahill J¹;
Andrea_Lopez@vrtx.com*

RESULTS: ELX/TEZ/IVA was projected to increase median survival of F/MF patients aged ≥ 12 yrs by 21.8 yrs (61.2 vs 39.4 yrs for BSC



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Journal of Cystic Fibrosis xxx (xxxx) xxx



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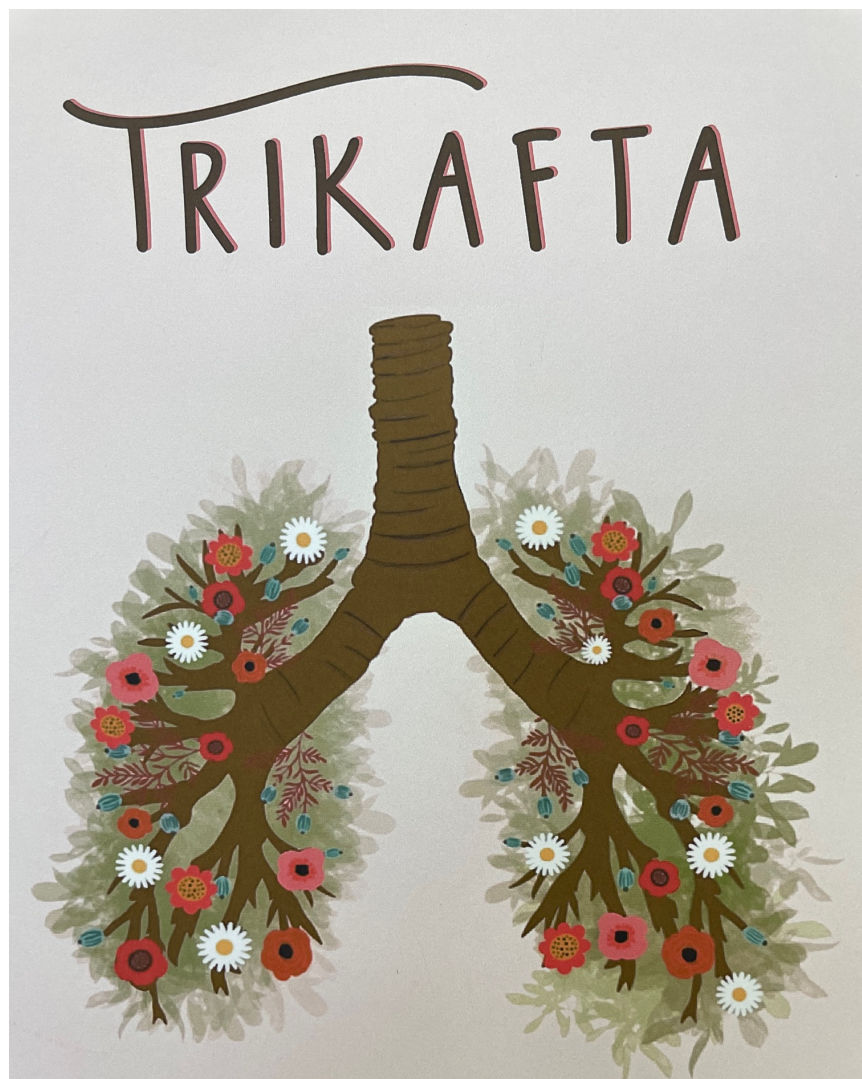


Case Report

A case report of CFTR modulator administration via carrier mother to treat meconium ileus in a F508del homozygous fetus

Sylvia Szentpetery*, Kimberly Foil, Sara Hendrix, Sue Gray, Christina Mingora, Barbara Head, Donna Johnson, Patrick A. Flume

Medical University of South Carolina, Charleston, SC 29424, USA





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Nature 428: 116-8

Growing old gracefully



FIBROSI CISTICA DELL'ADULTO

GRAZIE PER L'ATTENZIONE

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