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FRAGILITA' E FIBROSI CISTICA DELL'ADULTO: DUE CONDIZIONI UN PARADIGMA?

Dott.ssa Jolanda Somma
Università degli Studi di Napoli "Federico II"
CRR Fibrosi Cistica dell'Adulto
jolanda.somma@unina.it

Fibrosi Cistica: una malattia complessa

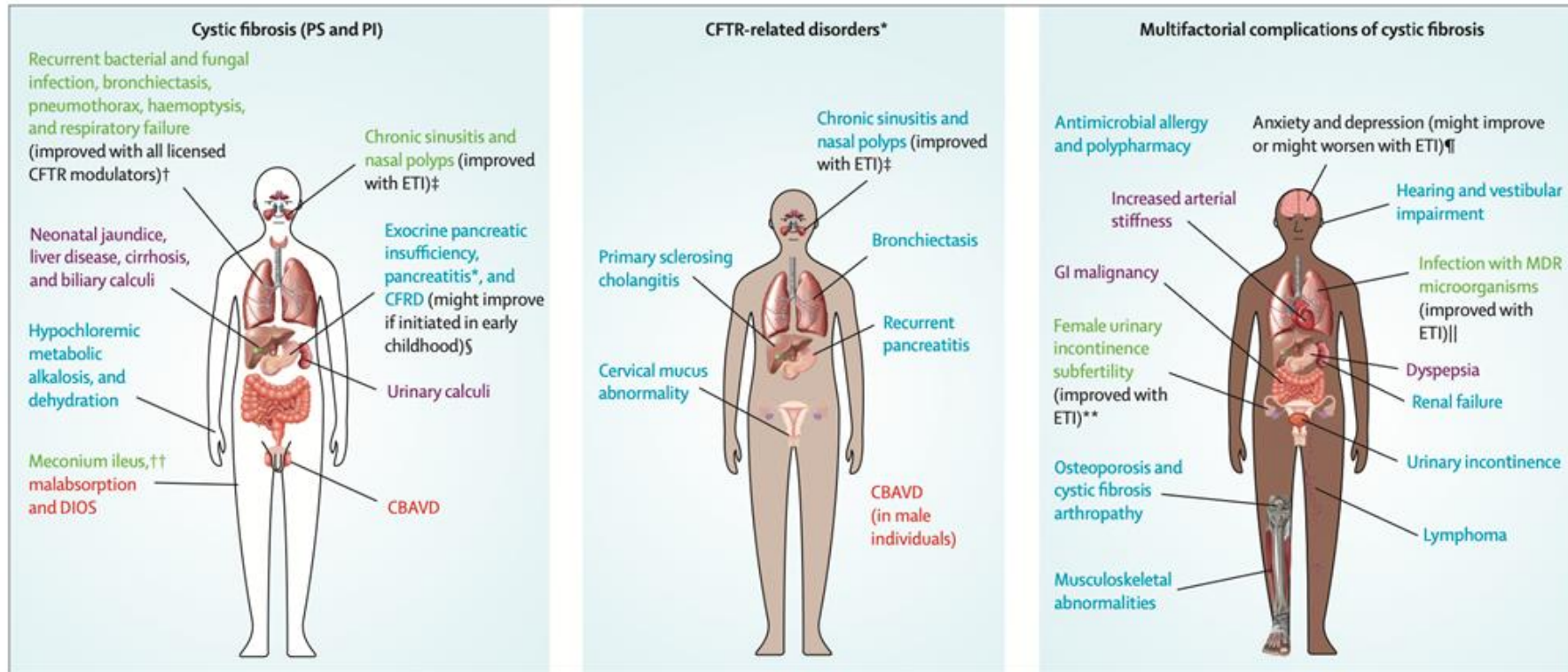
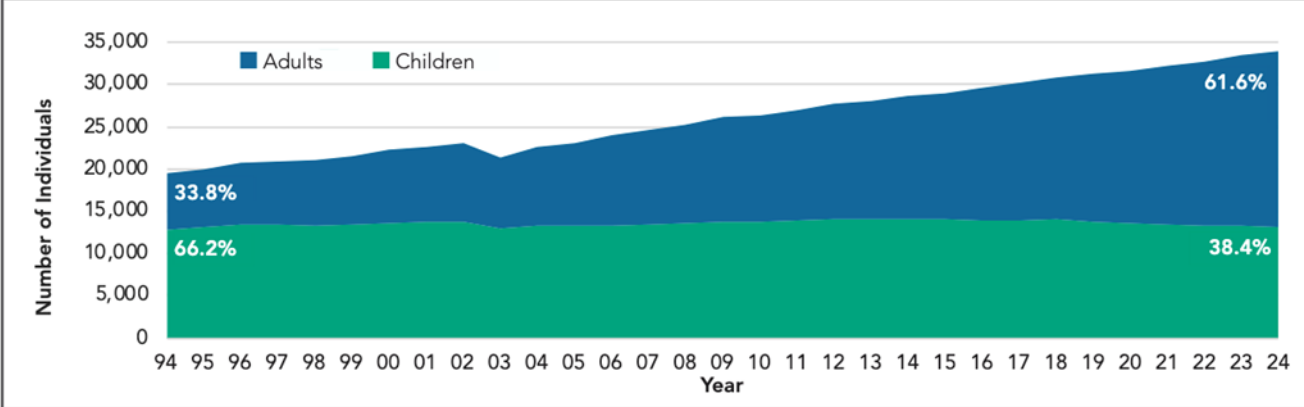


Figure 1: Known and potential effect of CFTR modulation on cystic fibrosis and CFTR-related disease manifestations

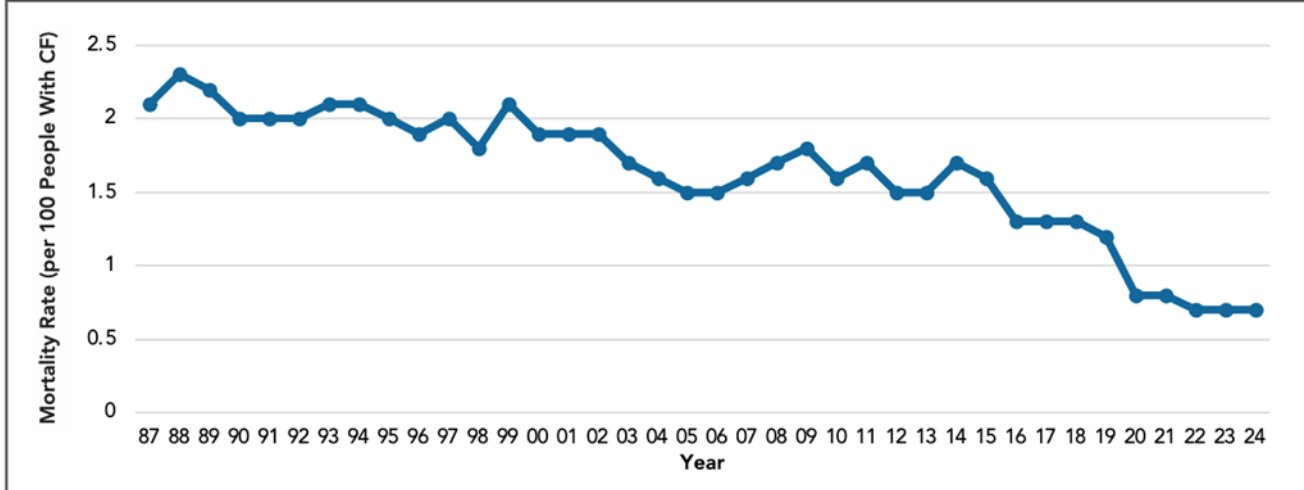
Lancet 2023; 402: 1171–84

Number of Children and Adults With CF, 1994–2024



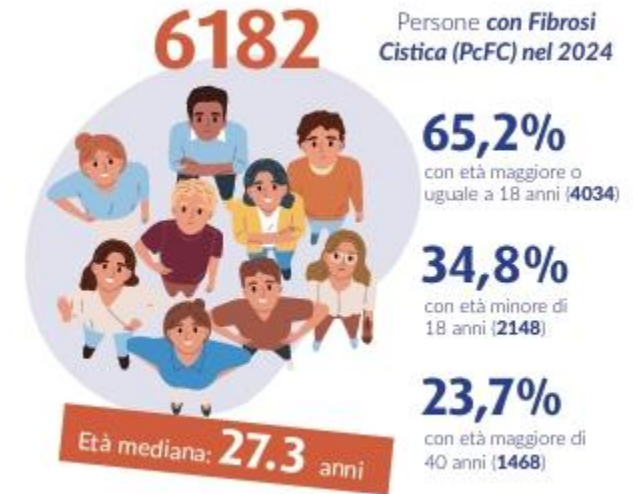
The decrease in the number of individuals reported in 2003 is due to a delay in obtaining informed consent forms before the close of the calendar year at some CF Care Centers.

Annual Mortality Rate (per 100 People With CF), 1987–2024



Annual Data Report 2024 Cystic Fibrosis Foundation Patient Registry

Due to improved medical care and new drugs (CFTR modulators), ageing in Cystic Fibrosis (CF) is becoming more common, and the complexity of the disease and the frailty of awCF increase.



Registro Italiano Fibrosi Cistica
Report at a glance 2024

This study aimed to assess the main clinical and anamnestic characteristics of adult CF patients and to evaluate the association of frailty with the CF genotyping classification.

Methods

An observational cross-sectional study was conducted at the Adult CF Center in Naples. We enrolled 139 CF adult patients (mean age 32.89 years, 46% F). All patients were assessed by spirometry for respiratory function, by ADL and IADL for functional status, and by the Study of Osteoporotic Fractures (SOF) Index for frailty.

Variables	Robust (n = 84)	Pre-Frail/Frail (n = 55)	p
Age, years, mean ± SD	32.38 ± 11.48	33.67 ± 10.12	0.498
Gender, M/W, n (%)	52/32 (61.90/38.10)	23/32 (41.82/58.18)	0.020
BMI, kg/m ² , mean ± SD	24.82 ± 3.76	22.57 ± 4.03	0.001
Smokers, n (%)	6 (7.14)	1 (1.82)	0.160
FEV ₁ , L, mean ± SD	3.28 ± 0.98	1.55 ± 0.75	<0.001
FEV ₁ , %, mean ± SD	84.93 ± 23.95	46.98 ± 22.65	<0.001
FVC, L, mean ± SD	4.26 ± 1.07	2.43 ± 0.86	<0.001
FVC, %, mean ± SD	94.95 ± 16.09	61.71 ± 20.45	<0.001
MMEF, %, mean ± SD	74.37 ± 32.43	29.33 ± 27.65	<0.001
Main Genotypes			0.116
DELTA508/DELTA508, n (%)	10 (12.05)	17 (30.91)	
DELTA508/5T-12TG, n (%)	10 (12.05)	1 (1.82)	
DELTA508/N1303K, n (%)	1 (0.72)	4 (2.90)	
N° pulmonary exacerbations per year, mean ± SD	0.58 ± 1.06	2.56 ± 2.51	<0.001
Cycles of IV antibiotic therapy, mean ± SD	0.08 ± 0.28	0.58 ± 1.32	<0.001
Cycles of oral antibiotic therapy, mean ± SD	0.50 ± 0.91	1.93 ± 1.87	<0.001
N° of hospitalizations per year, mean ± SD	0.11 ± 0.35	0.47 ± 0.88	<0.001
BADL lost, mean ± SD	0.00 ± 0.00	0.24 ± 0.77	0.006
IADL lost, mean ± SD	0.00 ± 0.00	0.65 ± 1.82	0.001
N° of drugs, mean ± SD	4.49 ± 3.80	8.60 ± 3.10	<0.001
N° of CF-related diseases, mean ± SD	2.65 ± 2.57	5.67 ± 2.64	<0.001
N° of non-CF-related diseases, mean ± SD	0.35 ± 0.67	0.42 ± 0.74	0.547
N° of total diseases, mean ± SD	3.00 ± 2.57	6.09 ± 2.80	<0.001

- 60,4% of the enrolled patients were found to be robust.
- The pre-frail/frail group was more frequently females (p = 0.020), had a lower BMI (p=0.001), worse respiratory function, a higher number of pulmonary exacerbations/years, cycles of antibiotic therapy, and hospitalization with respect to robust patients.
- The patients included in the pre-frail/frail group take a polypharmacotherapy and were affected by more CF-related diseases (all p<0.001).

Robust vs. Pre-Frail/Frail	β	95% Conf. Interval Low High	<i>p</i>
Gender			
Women	-0.092	-0.236 0.051	0.205
BMI, kg/m ²	-0.010	-0.027 0.007	0.261
FEV ₁ , L	-0.221	-0.297 -0.145	<0.001
Number of pulmonary exacerbations per year	0.075	0.024 0.125	0.004
Cycles of IV antibiotic therapy	-0.003	-0.149 0.142	0.966
Number of hospitalizations per year	-0.122	-0.326 0.082	0.239
BADL lost	-0.088	-0.293 0.117	0.399
IADL lost	0.073	-0.004 0.150	0.063
Number of drugs	-0.010	-0.040 0.019	0.483
Number of CF-related diseases	0.021	-0.018 0.060	0.287
Number of non-CF-related diseases	0.030	-0.062 0.121	0.525

- The **pre-frail/frail** subjects more frequently showed a **CF-causing/CF-causing** profile compared to the robust patients.
- The **best predictor of the pre-frail/frail status was a low FEV1 level.**

Conclusions

Since CF patients show similarities to pre-frail/frail subjects, CF could be considered an **early manifestation of a geriatric syndrome**. These results could better define the progression of CF but above all, they suggest the usefulness of employing some tools used in the management and therapy of frail subjects to identify subjects with more severe CF.