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Adolescent FEV₁ decline in cystic fibrosis may be an artefact of the spirometry reference equations used <http://ow.ly/J0com>

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Public awareness on cystic fibrosis: results from a national pragmatic survey



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To the Editor:

Cystic Fibrosis (CF) is a life threatening disorder that primarily affects the lungs and digestive system that represents an important cause of morbidity and mortality. Research has provided effective tools for disease prevention and treatment [1]. Defective gene and its protein product were discovered in 1989 [2–4] and screening systems are available for carrier identification [5]. Moreover, new-borns screening for CF facilitates early diagnosis and genetic counselling. Thanks to more effective drugs availability [6, 7], life expectancy has significantly grown in the last decades [8], with more than 45% of the CF patients aged >18 years and with a satisfying quality of life [9].

Public awareness on genetic disorders is essential for disease prevention, early diagnosis and treatment. Moreover, it is relevant to sensitise institutions to provide the best possible healthcare, social and environmental facilities for patients, families and care managers.

In order to capture information for CF education and advertising campaigns, the Lega Italiana Fibrosi Cistica – Lombardia (Milan, Italy), endorsed a public pragmatic survey, aimed to evaluate Italian adults' knowledge about CF prevention, diagnosis and treatment.

This *ad hoc* survey was conducted by DOXA Marketing Advice (Milan, Italy) who recruited volunteers and attended to the methodology and data analysis. A sample of 1006 individuals that represented the Italian population aged ≥ 15 years (52.1 million people) was used.

The need to accommodate a maximum margin of sampling error was taken into account: considering 1006 respondents out of an overall population of more than 52 million, a sampling error of $\pm 2.88\%$ is reported, which ensures a confidence level of 99% in the worst-case scenario of parameter estimation ($p=50\%$).

The survey was based on interviews in the respondents' homes. A questionnaire, developed by a panel of experts in the treatment of CF patients, which took approximately 15 min to complete, with the support of a laptop computer (computer-assisted personal interviewing system) was adopted.

A total of 130 trained interviewers conducted the survey between May 25, 2014 and June 3, 2014. The community under study was divided into sections or "layers" based on two criteria, the region and the size of the municipality of residence.

The number of interviews conducted in each layer was calculated to ensure that the number of interviews was proportional to the distribution among the various layers of communities under study.

All of the units included in the sample constitute a miniature reproduction of the community being considered (proportional stratified sampling method). The names and addresses of the interviewed were extracted from the electoral roll. People aged 15–17 years, who cannot yet register to vote, were selected using the quotas method.

During the processing phase, the data were weighted and a weighting factor was assigned to each interview to perfectly balance the sample compared to the reference population. The following variables were considered: sex according to age, region according to town amplitude, education (degree/diploma/middle school/elementary school), and employment status (working/not working).

A descriptive analysis of the answers was performed, and the Chi-squared test was used to test for associations between answers and membership in a particular group. The relationship between single items and the general opinion regarding inhaler devices (latent importance) was evaluated using the t-test.

1006 individuals (52% females) were evaluated. To assess their awareness of CF, the respondents were asked whether they had recently heard of any of several lung diseases. 64% of those interviewed claimed that they had previously heard about CF; 47% of them had been informed from watching the television, 13% from

TABLE 1 Subgroup analysis of the 1006 respondents

Awareness of	Answered the question	Respondents								
		Age years			Sex		Has son aged ≤ 14 years		Claimed to know about CF	
		≤ 34	35–64	≥ 65	Male	Female	Yes	No	Yes	No
CF disease features	64	65	73	55	60	68	69	63	40 [¶]	18
CF heritability	20	13 ⁺	28	10 [§]	17	22	30	17		
Age at onset of CF symptoms	21	17	26	17	19	24	25	21	31 [¶]	4
CF life expectancy	9	5	10	9	9	8	10	8	11 [¶]	4
Diagnostic test [#]	16	14	21	8	15	17	26	14	21 [¶]	8
Neonatal screening	47	47 ^f	55	32 ^{##}	45	50	64 ^{¶¶}	44	56	32
Carrier prevalence	8	7	10	5	9	7	9	8	10 ⁺⁺	4
Test for carrier detection	29	22	33	27	28	30	40	265	39 [¶]	11
Oxygen therapy	37	28	41	39	36	38	42	36	48 [¶]	18
Mechanical ventilation	34	24	39	35	33	35	39	33	44 [¶]	15
Lung transplantation	26	185	32	24	27	25	32	25	34 [¶]	12

Data are presented as percentages. #: sweat and genetic tests; ¶: $p=0.0001$ versus claimed not to know about cystic fibrosis (CF); +: $p=0.0001$ versus 35–64 years; §: $p=0.0233$ versus adult; f: $p=0.0398$ versus aged ≥ 65 years; ##: $p=0.0007$ versus 35–64 years; ¶¶: $p<0.0001$ versus without son aged ≤ 14 years; ++: $p=0.003$ versus claimed not to know about CF.

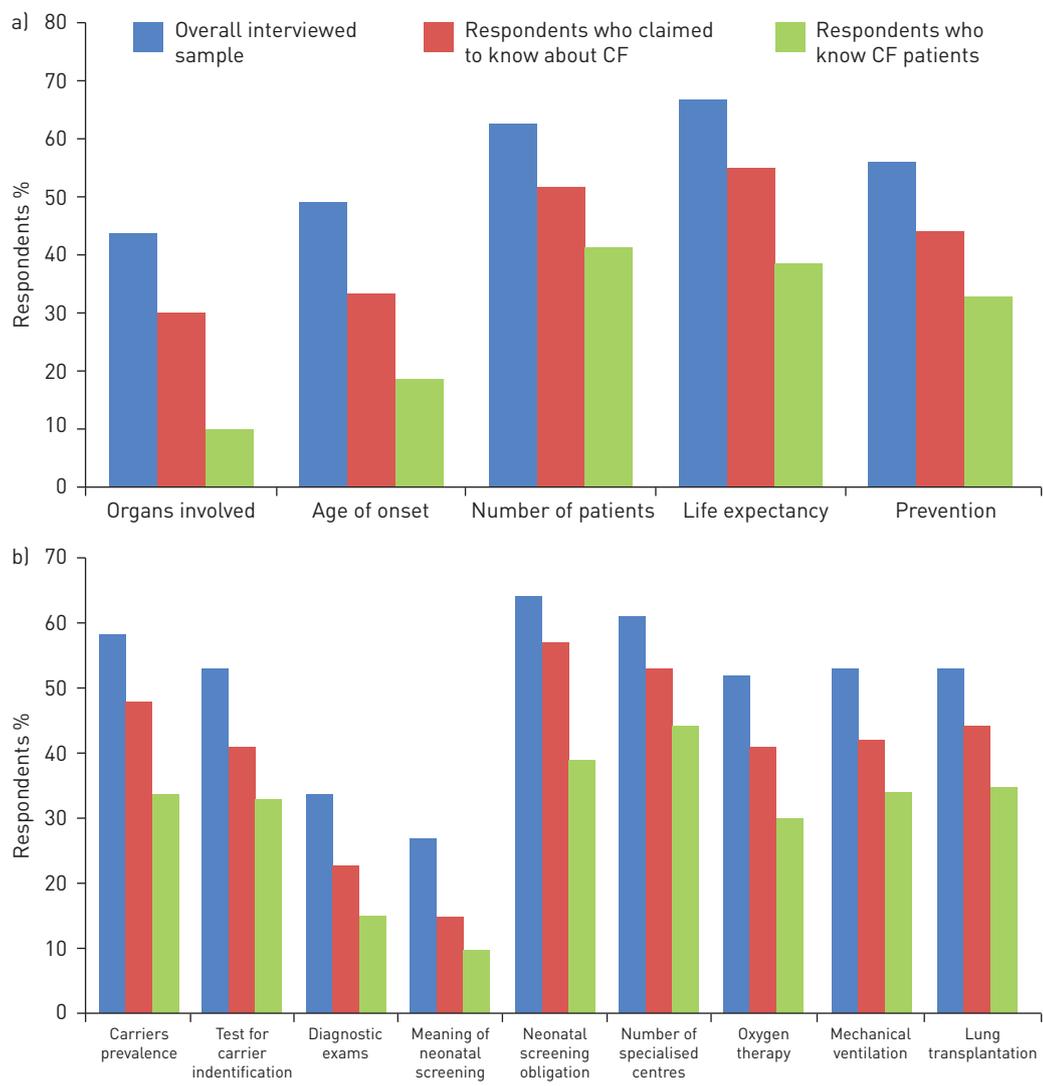


FIGURE 1 Percentage of respondents who provided incorrect answers or were not able to answer the survey questions.

reading newspapers/magazines, 8% know a CF patient, 3% from talking to a physician and 2% from using the web. In addition 20% of the sample was aware about the disease being hereditary. No difference concerning nominal awareness and disease hereditary were shown in accordance with employment, socio-economical status, the size of the town of residence, ownership of a personal computer and internet use. A high degree of education was associated with a higher knowledge of CF transmission. Only 19% of those interviewed and who were aware of CF correctly estimated the the number of Italians suffering from CF (5000 patients [10]), with an odds ratio of 4.9 for correct estimation amongst those interviewed who were aware of CF and an odds ratio of 1.8 among those who know CF patients. 53% of the population aware of CF knew that the lungs and the digestive system were the organs that are most involved in the disease (odds ratio of 4.1 among people who know CF patients). 49% of those interviewed did not know the age of onset for symptoms for CF, while 67% were not aware about life expectancy. 56% did not know if CF is preventable or not, while 20% consider CF preventable. Only 16% of the sample considers sweat test and genetic analysis fundamental in the diagnosis of CF, while 39% and 31% indicate lung CT scan and a blood test, respectively, are essential in diagnosis. Neonatal screening is considered an efficient diagnostic tool by 47% of interviewed and only 10% are aware that, in Italy, this test is mandatory. About two-thirds of the sample is not aware that the carrier status may be discovered using a blood test. Among the persons aware about genetic analysis, 45% suggests that the test should be performed before pregnancy on all parents, while, surprisingly, 19% would make the test available for all couples “during” pregnancy. Frequency of carrier status (estimated as 1 in 30 people [11]) has been correctly identified by 8% of the sample. Respondents were asked if oxygen therapy, mechanical ventilation and lung transplant are used in the end stage of CF; 53% of were not able to answer to the questions while 37%, 34% and 26%, respectively, were aware of the use of these treatments. The results

of the statistical analysis on the above mentioned topics according to sex, having or not having a son aged ≤ 14 years, being knowledgeable or not about CF, and of knowing CF patients are reported in table 1 and in figure 1. The vast majority of respondents (77%) believed that CF patients must be cared for by highly specialised centres although they are not aware about the real existence of these.

The awareness of CF is still poor. In particular, knowledge about the pattern of inheritance seems to be lacking as well as age of onset and life expectancy. Respondents aged 35–64 years were more aware about the heritability of CF and the availability of neonatal screening when compared with either the older or younger population surveyed. No significant difference was found between sex, while respondents with sons aged ≤ 14 years were more aware of neonatal screening than those without sons. Respondents who claimed to know of CF were significantly more aware, than those who do not know of CF, concerning disease features, diagnostic tests, carrier prevalence and detectability, utility of oxygen therapy, mechanical ventilation and lung transplantation. Based on this finding we recommend more educational campaigns targeting all populations with particular attention to those aged ≤ 34 years and males. Informational programmes should educate the public through television broadcasts, life lectures and seminars, emphasising the nature of inheritance, the available preventive measures, the differences between carriers and affected individuals. In Italy, a minority of couples discuss their reproductive plans with a health professional, usually a gynaecologist, before the pregnancy, an attitude which is possibly driven by the fact that this service is reimbursed by the health system in some areas. More resources should be devoted to pre-conceptual counselling, which may explore and illustrate reproductive options and risks and inform about neonatal screening aims and management.



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The results of a national pragmatic survey show that public awareness on cystic fibrosis is poor
<http://ow.ly/JztjM>

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