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Adolescents' Attitude towards Carrier Testing for Cystic Fibrosis and Its Relative Stability over Time

Myriam Welkenhuysen[a]
Gerry Evers-Kiebooms[a]
Marleen Decruyenaere[a]
Herman Van den Berghe[a]
Jacqueline Bande-Knops[b]
Veerle Van Gerven[c]

[a] Center for Human Genetics, University Hospital,
[b] Department of Youth Health Care, and
[c] Medical School Health Service, Leuven, Belgium

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Abstract
Attitudes towards cystic fibrosis (CF) carrier testing, benefits of and barriers to having such a test were assessed within a randomly selected group of high school students in Flanders, after they had received sufficient basic information about the nature and the mode of inheritance of CF. Attitudes towards carrier testing for CF were not negative, but the majority preferred to wait to have a test. This result changed little after 6 months. A hypothetical testing offer from the Medical School Health Service elicited positive answers from nearly two thirds, suggesting that such an offer may function as a cue to action. Nevertheless, the appropriateness of such an offer may be questioned, considering the disadvantages of testing adolescents. Concern about a negative impact of the carrier status on self-image was reported by 10% of the students. These findings suggest that education about genetics is not only a prerequisite for allowing more informed decisions about CF carrier testing, but also for avoiding negative psychosocial effects of such a test.

Introduction

Cystic fibrosis (CF) is the most common severe, autosomal recessive disease, affecting about 1 in 2,500 live births in the Caucasian population [1]. Approximately 1 in 25 people is an asymptomatic carrier of the CF gene. About 4 years after the localization of CF on chromosome 7 [2], an important breakthrough in CF research was achieved in August 1989: the identification of the CF gene [3]. Meanwhile, more than 300 different CFTR mutations have been documented by the International Cystic Fibrosis Analysis Consortium. Testing for the most common mutations causing CF (ΔF508 and 6–12 other mutations) identifies 85–90% of CF carriers in Caucasian populations. Most infants with CF are born in families with no history of the disease. Therefore, population screening is the only way to reach the majority of couples at risk of having a CF child. However, the implementation of a heterozygote screening programme for CF leads to many complex problems [4–6]. A major problem is the limited test sensitivity: a detection rate of 85–90% implies that 10–15% of all carriers receive a false-negative result, which has to be explained very carefully to all testees. This not only compounds the normal problems of genetic

KARGER
E-Mail karger@karger.ch
Fax + 41 61 306 12 34

1018–4813/96/0041–0052$10.00/0

Prof. Dr. Gerry Evers-Kiebooms
Center for Human Genetics
University Hospital
Herestraat 49
B–3000 Leuven (Belgium)

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counselling, but also requires effective educational programmes to increase public understanding regarding human genetics and genetic disorders as well as common notions of probability [7, 8]. Moreover, education is a prerequisite for ensuring that testing of individuals at risk, or screening of the population for CF carriers, is based on real 'informed' consent, and that people make more informed decisions in a genetic context. The importance of education was a major lesson learnt from the sickle cell and Tay-Sachs screening programmes in the seventies [9, 10].

To study the complex issues involved in offering a population screening programme for CF, pilot projects have been initiated in the US [11], Canada [12, 13] and Europe [14–18]. While most of these studies revealed a poor level of knowledge of CF and its genetic nature, they also showed that the attitude towards population screening for CF carriers was fairly positive. Support for offering the possibility for CF screening ranged from 72 to 100% [19], while the proportion who wished to be tested themselves varied between 54 [14] and 89% [19]. A study within a group of adults in Flanders [20] revealed that nobody considered a mass screening programme for CF unacceptable, while merely 1% thought it was unnecessary. At the same time, only 63% of the group reported that they wanted to know their own carrier status.

The present study is mainly aimed at assessing attitudes towards carrier testing for CF within a randomly selected group of adolescents in Flanders. In the first part of the study, information about the nature and transmission mode of CF was given systematically to all participants. We always checked the understanding of the notion of 'asymptomatic carrier' and corrected it if needed before investigating attitudes towards CF carrier testing. In the second part of the study, we investigated whether understanding of this notion persisted and whether the attitude towards CF carrier testing changed over time.

Method

The study was executed in cooperation with the Medical School Health Service between January and December 1993. In the first part of the study, alternating self-report questionnaires and standardized face-to-face interviews concerning knowledge and opinions about health problems, genetic diseases, perceived susceptibility to genetic disease and expectations regarding genetic testing, were administered within the scope of a 2-yearly medical checkup. Special attention was paid to CF and to carrier identification for CF, in the questionnaire and in the interview. First, initial general knowledge about CF was measured by one open-ended and two multiple-choice questions (appendix A). To make sure that all students would have the same minimal level of basic information about the nature and the mode of inheritance of CF, they were provided with an informative text. This text (one page) concerned the symptoms of the disease, possible treatments, the recessive mode of inheritance, the possibility of CF carrier testing (mouthwash sample), and the possibility of prenatal diagnosis (appendix B). Immediately after reading the text, the adolescents had to answer six multiple-choice questions, evaluating whether they grasped the meaning of CF carriership (appendix C). The number of right responses on these six questions constituted the total score for understanding. Immediately after answering these six questions, wrong answers were always corrected and additional information was given if needed. It is only after this information process that the attitudes towards carrier testing for CF were assessed. This was done by asking the subjects (a) if they would want to know their own carrier status at present, (b) in the future and (c) in the case that the Medical School Health Service would offer a CF carrier test during the next school year. Perceived benefits of and barriers to knowledge of their own carrier status were assessed by four open-ended questions (appendix D).

In the second part of the study, about 6 months later, all participants of the first part were invited to complete a follow-up questionnaire during lunch time at school. This follow-up questionnaire repeated the foregoing questions (appendices A, C and D) but did not contain the informative text about CF. During the second part of the study, we also used a 'control group' for the questions concerning general knowledge about CF. This control group, invited along to the follow-up session, consisted of subjects from the same schools who did not participate in the first part of the study. The control group had only one function in this paper, namely to check whether there had been a change in general knowledge about CF during the 6-month interval due to an uncontrolled factor.

Subjects

In the first part of the study a group of 166 fifth-grade students between 16 and 20 years old was selected at random from name lists we received beforehand from the Medical School Health Service. Four of these subjects did not answer the questions about CF, so that the sample in this paper is confined to 162 subjects. About 6 months later, nearly 64% of them also filled in the follow-up questionnaire. These 103 subjects were considered as the follow-up group. The control group consisted of 117 subjects (17–20 years old), attending the same schools and classes as the initial group of 162 students. Some subjects did not answer all the questions, resulting in a different sample size for some questions. Table 1 gives an overview of some sociodemographic characteristics of the three groups of subjects. About one fifth of the initial sample and the control group, and more than one quarter of the follow-up group were acquainted with a CF patient. These surprisingly high proportions are due to the presence of two CF patients in one school involved in our study. No significant differences were found among the three groups regarding the characteristics described in table 1.

Results

First, a description of the results of the first part of the study involving the group of 162 subjects is given. This group is referred to as the 'initial sample'. Secondly, the

Table 1. Sociodemographic characteristics and proportions of subjects who are acquainted with a CF patient

	Initial sample (n = 162)	Follow-up group (n = 103)	Control group (n = 117)
Sex			
Male	45.1%	41.7%	42.7%
Female	54.9%	58.3%	57.3%
Mean age, years	16.7	17.2	17.4
Education			
General	52.5%	56.3%	56.9%
Technical	47.5%	43.7%	43.1%
Do you know a CF patient?			
Yes	21.0%	27.2%	20.5%
No	79.0%	72.8%	79.5%

results of the follow-up group are presented and compared with the results from the first part of the study. Making this comparison, only the subgroup of 103 subjects who participated in both parts of the study is considered. This group is called 'follow-up group moment 1' when talking about the first part of the study and 'follow-up group moment 2' when reporting on the follow-up session. Inspection of the results tables reveals that the pattern of results within the initial sample as a whole and within its subgroup, i.e. 'the follow-up group moment 1' is largely the same. Therefore, we conclude that no self-selection occurred regarding participation in the follow-up session.

Initial General Knowledge about CF

Only 28.9% of the initial sample had heard of CF; when asked to give a brief description of CF, 88.4% could give at least one feature of the disease, which corresponds to 25.5% of the total initial sample. Respiratory problems are the most salient feature of the disease and were referred to by 20.1% of the initial sample. The genetic cause was spontaneously mentioned by only 2.0%. The multiple-choice question concerning the cause of CF was answered correctly by 4.7% (i.e. genetic cause). When asked about the incidence of the disease in both sexes, 14.8% chose the correct alternative (i.e. CF is equally likely in boys and girls). The proportion of 'I do not know' and missing answers was very high: 85.9% for the first multiple-choice question and 84.6% for the second.

Understanding of Recessive Inheritance and Carriership for CF Immediately after Reading an Informative Text in the First Part of the Study

The answers to the six multiple choice questions (appendix C) are presented in table 2. Wrong answers were always corrected to ensure that the students were correctly informed before answering the next questions concerning attitudes towards CF carrier testing.

A high score was obtained for most of the statements. Except for the third statement (i.e. other people can be infected by a CF carrier), more than four fifths of the initial sample gave a correct answer. This corresponds to a mean total score of 5.2. However, although all subjects had read the text immediately before answering these questions, the proportion of subjects who answered all six questions correctly reached only 48.1%. A t test revealed that the total score for understanding was significantly higher for the subjects who were acquainted with a CF patient ($t = 2.6$; $p < 0.05$).

Initial Attitudes towards Carrier Testing for CF

After correcting the errors in understanding of recessive inheritance and asymptomatic carriership, subjects were asked if they would want to know their own CF carrier status (at the time of the study, in the future, and in the case that the Medical School Health Service would offer a CF carrier test the following year). They could give their answer by choosing one of the following alternatives: 'yes', 'I do not know', or 'no'. The results in table 3 reveal that the proportion of subjects who do not want to know their carrier status at the time of the study was larger than the proportion of those who do. At the same time, only a small minority stated that they did not want to be informed about their own carrier status in the future or by the Medical School Health Service the following year.

To examine if the change in attitude from one question to another was significant, a series of sign tests [21] was conducted. These tests revealed that the attitude towards knowing one's CF carrier status at the time of the study was significantly more negative than the attitude towards knowing it in the future ($z = -9.06$; $p < 0.01$) or if a test were to be offered by the Medical School Health Service the following year ($z = -7.59$; $p < 0.01$). The subjects in the initial sample were more positive towards knowledge about their own carrier status 'in the future' than towards a test offered by the Medical School Health Service the following year ($z = -2.91$; $p < 0.01$). Subsequent analyses showed no significant relation between attitudes towards knowing their own carrier status as measured by the three

Table 2. Percentage of correct answers on the six multiple-choice questions concerning the informative text about CF

	Initial sample (n = 162)	Follow-up group moment 1[a] (n = 103)	Follow-up group moment 2[b] (n = 103)	McNemar change test (χ^2)
A CF carrier is not suffering from CF	90.7%	89.3%	79.6%	2.23
A CF carrier does not get CF later in life	88.3%	88.3%	66.0%	12.90*
Other people cannot be infected by a CF carrier	67.9%	66.0%	44.7%	9.02*
Not all of a CF carrier's children will obligatorily be a CF carrier	83.3%	84.5%	52.4%	20.00*
A CF carrier's children have a chance to be a CF carrier	94.4%	94.2%	76.7%	9.37*
A CF carrier's children have a chance to suffer from CF	96.3%	97.1%	80.6%	10.31*

* $p < 0.01$.

[a] Follow-up group moment 1 = results obtained in the first part of the study from the subjects participating in the follow-up sessions.

[b] Follow-up group moment 2 = results obtained in the second part of the study from the subjects participating in the follow-up sessions.

questions on the one hand, and acquaintanceship with a CF patient (χ^2) on the other.

Subjects who were positive about knowing their own carrier status in the future were also asked at what time in the future they would like to have this information (open-ended question). Most of them (58.6%) stated that they wanted to know their own carrier status before making reproductive decisions. A small proportion (10.5%) thought that knowing one's own carrier status was best only after marriage, while 4.5% would want to have this information before they had a partner.

The following two open-ended questions examined the perceived benefits of having a CF carrier test at the time of the study and in the future. The answers to these questions were coded into broader categories as shown in table 4.

More than one quarter of the initial sample stated that knowing their own carrier status at the time of the study had no benefits at all, but this proportion decreased significantly to less than 5% when considering carrier testing in the future (McNemar change $\chi^2 = 27.84$; $p < 0.01$). On the other hand, more than half the initial sample (53.1%) did perceive some benefit for a carrier test at the time of the study; the most important benefits were 'having certainty, being informed' without mentioning children or reproductive plans (21.0%), and 'knowing the risks for future children' (17.3%). With regard to having a test in the future, 82.1% of the initial sample mentioned some benefit. Here, reproductive plans became more prominent: about half of all the answers dealt with the possibility of making more informed decisions concerning children,

Table 3. Attitudes toward knowledge about their own CF carrier status at the time of the study, in the future, and if offered by the Medical School Health Service (MSHS) during the next school year

	Initial sample (n = 161)	Follow-up group	
		moment 1[a] (n = 102)	moment 2[b] (n = 100)[c]
At the time of the study			
Yes	37.3%	31.4%	31.0%
I do not know	15.5%	21.6%	30.0%
No	47.2%	47.1%	39.0%
In the future			
Yes	82.6%	81.4%	55.0%
I do not know	9.3%	9.8%	34.0%
No	8.1%	8.8%	11.0%
Offered by the MSHS during the next school year			
Yes	68.3%	62.7%	61.2%
I do not know	16.1%	17.6%	22.4%
No	15.5%	19.6%	16.3%

[a] Follow-up group moment 1 = results obtained in the first part of the study from the subjects participating in the follow-up sessions.

[b] Follow-up group moment 2 = results obtained in the second part of the study from the subjects participating in the follow-up sessions.

[c] n = 98 for the MSHS question.

Table 4. Perceived benefits of having a CF carrier test at the time of the study and in the future

	Initial sample (n = 162)	Follow-up group	
		moment 1[a] (n = 103)	moment 2[b] (n = 103)
At the time of the study			
No benefits	27.2%	31.1%	8.7%
Benefits: total	53.1%	49.6%	70.9%
Having certainty, being informed	21.0%	15.6%	37.9%
Knowing risks for future children	17.3%	14.6%	19.4%
If I'm a carrier, urge partner to take the test	8.0%	10.7%	2.9%
Preventing the birth of a CF child	3.7%	3.9%	1.0%
Other	3.1%	4.8%	9.7%
Answer reflecting misconceptions	0.0%	0.0%	5.8%
No answer	19.8%	19.4%	14.6%
In the future			
No benefits	4.9%	5.8%	1.2%
Benefits: total	82.1%	80.5%	77.3%
Informed reproductive decisions	51.2%	48.5%	51.1%
Preventing the birth of a CF child	14.8%	15.5%	12.6%
Informing oneself about CF	6.8%	6.8%	5.8%
Other	9.3%	9.7%	7.8%
Answer reflecting misconceptions	0.0%	0.0%	3.9%
No answer	13.0%	13.6%	17.5%

[a] Follow-up group moment 1 = results obtained in the first part of the study from the subjects participating in the follow-up sessions.
[b] Follow-up group moment 2 = results obtained in the second part of the study from the subjects participating in the follow-up sessions.

and 14.8% mentioned prevention of the birth of a CF child as a benefit.

The last two open-ended questions examined barriers to knowledge about their own CF carrier status at the time of the study and in the future. The answers were coded into broader categories and are presented in table 5.

A minority of 16.7% of the initial sample perceived no barriers at all regarding a CF carrier test at the time of the study. This proportion increased significantly to about one fourth for having such a test in the future (McNemar change $\chi^2 = 5.63$; $p < 0.05$). Complementary to these small proportions, nearly three quarters (74.7%) and more than half (57.4%) of the initial sample mentioned some barrier to knowing their own carrier status at the time of the study and in the future, respectively. As for perceived barriers at the time of the study, nearly one third of the subjects mentioned the expectation of being worried about the increased risk of having a CF child or about transmitting the carrier status to future children in the event that the test result would be positive. Further, a small but substantial proportion of the initial sample (11.7%) was afraid that a positive test result would have a negative impact on self-image. Expressions like 'I would feel abnormal, sick, very depressed ...' or 'I could never be happy again' belong to this category. When barriers to having a CF carrier test in the future were considered, the expectation of worry remained important: it was mentioned by more than one fifth of the subjects. The argument concerning being afraid of becoming pregnant or during pregnancy became a more prominent barrier than for the question concerning having a test at the time of the study; it reached about the same level as the expectation of being worried. Finally, being afraid that a positive test result would have a negative influence on self-image remained a salient barrier for a small proportion of the initial sample.

Are There Differences in General Knowledge about CF 6 Months Later?

The follow-up group, who answered the general knowledge questions twice with an interval in between of 6 months, did better the second time: the proportion of

Table 5. Perceived barriers to having a CF carrier test at the time of the study and in the future

	Initial sample (n = 162)	Follow-up group	
		moment 1[a] (n = 103)	moment 2[b] (n = 103)
At the time of the study			
No barriers	16.7%	15.5%	17.5%
Barriers: total	74.7%	71.9%	62.1%
If I'm a carrier, I will be worried	32.1%	28.2%	35.9%
I'm too young to have children	14.2%	15.5%	3.9%
If I'm a carrier, it will have a negative impact on my self-image	11.7%	12.6%	13.6%
If I'm/my partner is a carrier, having children will be a problem	5.6%	4.9%	3.9%
If I'm a carrier, it will increase anxiety about a/during pregnancy	4.9%	4.9%	1.0%
Other	6.2%	5.8%	3.8%
Answer reflecting misconceptions	0.0%	0.0%	2.9%
No answer	19.8%	19.4%	14.6%
In the future			
No barriers	26.5%	26.2%	19.4%
Barriers: total	57.4%	58.2%	43.6%
If I'm a carrier, I will be worried	21.6%	21.4%	18.4%
If I'm a carrier it will increase anxiety about a/during pregnancy	18.5%	19.4%	1.0%
If I'm/my partner is a carrier, having children will be a problem	8.6%	8.7%	11.6%
If I'm a carrier, it will have a negative impact on my self-image	6.2%	5.8%	8.7%
Other	2.5%	2.9%	3.9%
Answer reflecting misconceptions	0.0%	0.0%	1.0%
No answer	16.1%	15.6%	35.9%

[a] Follow-up group moment 1 = results obtained in the first part of the study from the subjects participating in the follow-up sessions.

[b] Follow-up group moment 2 = results obtained in the second part of the study from the subjects participating in the follow-up sessions.

the follow-up group who spontaneously mentioned at least one CF feature was higher than 6 months before (56.3 vs. 27.2%; McNemar change $\chi^2 = 23.36$; $p < 0.01$); the respiratory problems, again the most salient feature, were also mentioned more frequently 6 months later (48.5 vs. 24.3%) and we observed an increase in the proportion of subjects who spontaneously referred to the genetic cause of the disease (12.6 vs. 2.0%). However, the results on the multiple-choice question concerning the cause of CF were not much better than 6 months before (8.7 vs. 4.7% correct answers). This discrepancy is due to the substantial proportion (10.7%) who incorrectly chose the alternative 'chromosomal abnormality' as the genetic cause of CF. The results on the second multiple-choice question were significantly better: 39.8% (vs. 19.6%; McNemar change $\chi^2 = 14.7$; $p < 0.01$) answered that CF occurs equally likely in boys and girls. On the other hand, the frequency of 'I do not know' and missing answers on these last two multiple-choice questions was still high (67.0 and 50.5%, respectively).

The control group, who answered the same questions as the initial sample to assess existing knowledge about CF, showed no difference at all with the initial sample: 35.9% of the control group had heard of CF before, 30.8% could describe at least one feature of the disease, the respiratory problems were the most frequently mentioned characteristics, only 4.3% could correctly answer the multiple-choice question concerning the cause of CF and 19.6% knew that CF occurs equally likely in boys and girls.

Are There Differences in Understanding of Recessive Inheritance and Carriership for CF 6 Months Later?

As shown in table 2, the results of the follow-up group for five of the six multiple-choice questions concerning asymptomatic carriership were significantly worse

6 months after reading the informative text about CF than immediately after receiving the information. This occurred although additional information was given by the interviewer when needed in the first part of the study. Only for the first question was no significant difference observed. The results on the third statement (i.e. other people can be infected by a CF carrier) were the poorest again. The total score for understanding also reflected this drop in performance after 6 months, with a significant decrease from 5.2 to 4.1 ($t = 5.84$; $p < 0.01$). When the proportion of subjects who answered all six questions correctly is considered, a corresponding decrease from 48.1% immediately after reading the text to 23.3% 6 months later was observed. Similar to the first part of the study, the total score for understanding was significantly higher for the subjects who were acquainted with a CF patient ($t = 2.2$; $p < 0.05$).

Are There Differences in Attitudes towards Carrier Testing for CF 6 Months Later?

The three questions concerning attitudes towards CF carrier testing, which were asked a second time during the follow-up session, elicited largely the same pattern of results as 6 months before (table 3): the proportion of subjects who did not want to know their own carrier status at the time of the study was larger than the proportion that did, while the proportion who did not want to know it in the future or if a test were to be offered by the Medical School Health Service the following year was fairly small. Subsequent sign tests revealed that, similar to the first part of the study, the attitude towards knowing one's CF carrier status at the time of the study was significantly more negative than the attitude towards knowing it in the future ($z = -5.43$; $p < 0.01$) or when a test were to be offered by the Medical School Health Service ($z = -4.85$; $p < 0.01$). However, while in the first part of the study, the follow-up group was more positive towards knowing their own carrier status in the future than towards the offer from the Medical School Health Service ($z = -2.76$; $p < 0.01$), there was no difference in attitude concerning these two options during the follow-up session. In other words, shortly after receiving information about CF and CF carrier testing, the favorite option was knowing one's own carrier status in the future, while after 6 months this latter option seemed equally attractive as learning one's own carrier status when a test is offered by the Medical School Health Service. The difference between the two parts of the study becomes clearer when the results for each of the three questions are compared separately. A series of sign tests revealed only one significant difference: in the second part of the study, the attitude towards knowing one's own CF carrier status in the future was more negative than in the first part ($z = -2.44$; $p < 0.01$). An inspection of table 3 reveals that this was due to a higher proportion of subjects who were indecisive during the follow-up session, rather than to explicit negative answers. Similar to the first part of the study, the relation between tWelkenhuysen towards knowing one's own carrier status and acquaintanceship with a CF patient was not significant. There was no significant rank correlation with the total score for understanding.

When asked at what time in the future they wanted to be informed about their own CF carrier status, the follow-up group answered largely in the same way as in the first part of the study. As for perceived benefits of knowing their own CF carrier status, the results of the two parts of the study were very similar (table 4), except for the following: in the second part of the study, the proportion of subjects who perceived no benefits at all in knowing their own carrier status at the time of the study was much smaller, while the proportion mentioning 'having certainty, being informed' was more than twice as large as in the first part of the study. With respect to knowing their own carrier status in the future, reproductive plans and especially the possibility of making more informed decisions were far the most important benefits, as also observed in the first part of the study.

In the second part of the study, the answers concerning perceived barriers to knowledge about their own CF carrier status were not very different from those in the first part (table 5). The proportion of subjects answering 'no barriers at all' was just as small; with regard to perceived barriers in the future it was even somewhat smaller than in the first part of the study. The expectation of being worried remained the most important barrier, and again a small proportion talked about being afraid that a positive test result would have a negative impact on self-image. With respect to barriers to knowledge about their own carrier status at the time of the study, the argument 'I am too young to have children' was mentioned less frequently than in the first part of the study. Increased anxiety about getting pregnant or during a pregnancy was a less prominent barrier to knowing one's own carrier status in the future.

Discussion

Our study shows that senior high school students' initial general knowledge about CF is poor: only about one third of our subjects had ever heard of CF and less than

5% knew it to be caused by a gene abnormality. This is considerably less than in recent British studies involving high school students, where 59–84% reported they had heard of the disease [15, 16, 19] and 61.6% knew it was inherited [19]. About one quarter of the students in the Flemish study spontaneously mentioned that CF is characterized by respiratory problems. This corresponds more or less to the results of the British studies (between 17 and 34%) and to the results of an earlier Flemish study [20] involving adults (28%).

It is obvious from these data that education about the nature and course of CF will be required before young people can make more informed decisions about CF carrier testing.

Participation in the first part of the study and receiving concise information about CF had an effect on the general level of knowledge, although it was limited. The improvement in the follow-up group cannot be attributed to an uncontrolled factor during the 6-month interval between the interviews and the follow-up session. This can be deduced from the results for the control group: their level of general knowledge was similar to the initial level measured in the first part of the study.

The relationship between knowledge about CF and acquaintanceship with a CF patient remained significant in the second part of the study. This indicates that the long-term effect of the informative text on knowledge was fairly limited. More elaborate and attractive visual information could be helpful in this respect.

In the first part of the study, the answers to the six multiple-choice questions concerning the informative text revealed that, immediately after reading the text, the subjects had enough passive knowledge about recessive inheritance of CF and asymptomatic carriership to allow them to agree with most of the correct statements concerning this topic and disagree with most of the incorrect statements. This is of course much easier than active reproduction of the information. When asked the same questions 6 months later, the students did significantly worse. Moreover, the question concerning benefits of knowing one's own CF carrier status at the time of the study made it clear that at least 6 subjects in the follow-up group transformed the information into some misconception about asymptomatic carriership or about CF being a contagious disease. Together with the rather high proportion of students (about 30%) incorrectly agreeing with the statement that other people can be infected by a CF carrier, this suggests that educational programmes or information about genetics and genetic diseases should pay (more) attention to this 'confusion' about genetic transmission. It is our hypothesis that knowledge about carriership for AIDS might be at the origin of this confusion.

With regard to the attitude towards knowing one's own CF carrier status, most of the students would clearly prefer to receive a carrier test in the future. Only about one third of the group would like to know their own carrier status at the time of the study. This was consistent in both study parts. The attitude towards knowing one's CF carrier status in the future was less positive in the follow-up (55%) than in the first part of the study (80%). The main reason for waiting was the expectation of being worried about risks for children in the case of a positive test result, and this at a stage in life for which plans to have children are usually not yet important. In line with this reasoning, reproductive plans formed the central theme in the indication of benefits of knowing one's own carrier status in the future, and in the answers concerning the best time to receive this information. The majority of positive attitudes towards having a carrier test 'in the future' in the first part of the study corresponds to the results of two British studies [15, 19], both reporting a proportion of 77% of students with a positive attitude.

It is intriguing why so many more students in the second part of the study were indecisive about knowing their own carrier status in the future and why the proportion of positive answers to this question decreased to 55%. A possible explanation for this decrease in positive attitudes is the fact that the informative text, which served as a cue to action [22], was not available during the follow-up session. A similar explanation can be given for the high proportion of positive answers (more than 60%) when subjects were asked about participation in a screening programme offered by the Medical School Health Service: the hypothetical offer by the Medical School Health Service in the next school year can be viewed as an important cue to action. This line of reasoning receives some support from the high uptake rate observed in a study by Kaplan et al. [23]: 70% of the students who attended an information session about CF also participated in the screening session, both sessions being organized in close collaboration with the school board, within curriculum time.

It is important to pay sufficient attention to the increase in indecisiveness about knowing one's own carrier status from the first to the second part of the study. This increase in indecisiveness suggests that the weighing up of the advantages and disadvantages of carrier testing is a dynamic and complex process that needs time. In this respect, temporary indecisiveness should be viewed positively. On the other hand, it also suggests that one should proceed with care when offering carrier testing in a high

school situation. Obviously, screening in high school has some advantages [17, 24]: it is administratively easy, it provides an opportunity to combine the test with a formal education programme and it can usually be considered as 'prepregnancy' screening, so that it leaves open the full range of reproductive options. On the other hand, it has many disadvantages. First, the time delay between obtaining the test result and using the information is long. Secondly, there is no consensus about consent residing with the parents or with the teenager. Particularly in high school, peer pressure towards conformity may occur, so that adolescents cannot always make a really free decision. From a psychological point of view, they may be especially vulnerable to social stigmatization when testing occurs in a group session and loss of self-esteem may also occur [9, 25]. A small proportion of the answers on the questions concerning barriers to receiving a CF carrier test at present and in the future point in that direction, expressing very negative feelings in the situation where one would actually be a carrier. That carriership of a recessive defective gene can have some negative connotations was also found in a study by Marteau et al. [26], which showed that Tay-Sachs carriers perceive their future health with less optimism than do non-carriers. Moreover, a study by Evers-Kiebooms et al. [27] revealed that CF carriers have less positive feelings about themselves than non-carriers and that CF carriership has negative connotations. Furthermore, in a study by Watson et al. [28], nearly one quarter of a group of non-carriers for CF, who attended an information session on CF and carriership for CF before participation in the screening session, stated that it would lower their self-image if they were carriers for CF. Therefore, a well-elaborated educational programme concerning human genetics and genetic disease, here CF, is necessary to enable people to make more informed decisions and to minimize possible negative psychosocial effects of carrier testing for CF and a fortiori of carrier screening programmes.

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Appendix: CF Questionnaires and Information Sheet

A: Existing Knowledge about CF

Have you heard of CF before?
(1) Yes
(2) No

Do you know a child with CF in your surroundings?
(1) Yes
(2) No

Give a brief description of the disease.

_____

_____

_____

(1) CF is caused by environmental factors.
(2) CF is caused partly by hereditary factors and partly by environmental factors.
(3) CF is caused by a chromosomal abnormality.
(4) CF is caused by a gene abnormality.
(5) I do not know.

[Option 4 is the correct answer.]

(1) CF occurs almost exclusively in boys.
(2) CF occurs almost exclusively in girls.
(3) CF is (approximately) equally likely to occur in boys as in girls.
(4) I do not know.

[Option 3 is the correct answer.]

B: Information about Cystic Fibrosis

Cystic fibrosis (CF) is a serious disease occurring in childhood. It is caused by the malfunctioning of certain glands, resulting in the mucus (e.g. mucus in the lungs and in the intestines) being sticky and viscous. Serious problems concerning the lungs, the bronchial tubes and the digestive system can already exist at birth. The sticky mucus clogs the minor bronchial tubes, creating difficulties in breathing. Pneumonia also occurs frequently. The presence of the sticky mucus in the intestines leads to digestive disorders. The physical development and maturation of CF children are often retarded.

CF is an incurable disease. A specially adapted diet and good medical treatment can to some extent reduce and/or postpone the physical symptoms. Despite the progress in treatment already made, many CF patients still die before they reach adulthood.

CF is a hereditary disease. Approximately 1 person out of 25 is an asymptomatic carrier of CF, in other words, carries a defective hereditary characteristic, which can cause CF. *A carrier does not have symptoms of the disease him/herself, but the defective characteristic for CF is present in his/her genetic material.* It is possible that he/she passes on the defective characteristic to his/her children.

A couple can have a CF child **only if both the father and the mother are asymptomatic carriers of the defective gene.**

When two asymptomatic carriers of CF have a child, then the chance is 1 out of 4 (25%) that this child will suffer from CF; 2 out of 4 (50%) that this child will be a carrier of the disease, but does not suffer from it (like the parents); and 1 out of 4 (25%) that this child does not suffer from the disease and is not a carrier, in other words, this child cannot transmit the disease.

Before a pregnancy, the examination of mouthwash samples makes it possible to detect asymptomatic carriers of CF (persons who do not suffer from the disease, but who may pass it on to their children); this procedure is called a **carrier test.**

Prenatal diagnosis makes it possible to detect the disease *during* pregnancy.

C: Knowledge about Recessive Inheritance and Carriership for CF

(1) An asymptomatic carrier of CF is suffering from CF.
(2) An asymptomatic carrier of CF will get CF later in his/her life.
(3) Other people can be infected by an asymptomatic carrier of CF and get CF also.
(4) An asymptomatic carrier of CF will pass the gene with certainty to each of his/her children and they will all be carriers.
(5) There is a chance that an asymptomatic carrier of CF will pass the defective gene to his/her children, so there is a chance that they will be carriers too.
(6) There is a chance that the children of an asymptomatic carrier of CF will suffer from CF.

For each of these six statements the subjects could choose to answer 'correct', 'wrong' or 'I do not know'.

D: Attitudes toward Carrier Screening for CF and Perceived Benefits of and Barriers to Knowledge of Their Own Carrier Status

Your situation being as it is NOW, would you want to know NOW if you are an asymptomatic carrier of CF?
(1) Yes
(2) I do not know
(3) No

What would encourage you to have a carrier test for CF NOW? In other words, according to you, what are the advantages of receiving a carrier test NOW?

_____

_____

_____

What would hold you back from having a carrier test for CF NOW? In other words, according to you, what are the disadvantages of receiving a carrier test NOW?

_____

_____

_____

Would you want to know your carrier status IN THE FUTURE?
(1) Yes
(2) I do not know
(3) No

At what time in the future?

_____

What would encourage you to have a carrier test for CF IN THE FUTURE? In other words, according to you, what are the advantages of receiving a carrier test IN THE FUTURE?

_____

_____

_____

What would hold you back from having a carrier test for CF IN THE FUTURE? In other words, according to you, what are the disadvantages of receiving a carrier test IN THE FUTURE?

_____

_____

_____

Suppose that the School Health Service were to offer all sixth-year students of secondary school the possibility for testing if they are asymptomatic carriers of CF (a test based on a mouthwash sample). What would you do in such a case?
(1) I would let them test if I am a carrier of CF
(2) I do not know
(3) I would not let them test if I am a carrier of CF

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