high profile, vertical intervention programmes and its stated goal of integrated primary health care.

WHO is entering a period of intense soul searching and internal upheaval. Some insiders fear that it is tearing itself apart. With growing pressure on limited resources, donors are demanding more value for money and greater accountability. Reports from the United Nations, from donors, and from within the WHO itself are demanding radical reform.^{4,6,8} The articles that follow will explore the problems faced by WHO and some possible solutions.

Even WHO's most vociferous critics agree that some form of international health organisation is necessary. The world needs a strong body to take the lead in health matters, to act as an advocate for equity in economic and social development, to set priorities for the use of limited resources, to provide neutral territory for debating sensitive issues, and to give technical advice and support. Such a role can be successfully taken only by an organisation that commands respect and is seen to be above national politics and free from divisive internal wranglings. It

remains to be seen whether the World Health Organisation is capable of reform or whether, as some critics feel, it is doomed, like the rest of the United Nations, to flounder in a morass of petty corruption and ineffective bureaucracy. These articles will, I hope, stimulate a much needed debate on WHO's future.

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Controversies in Management

Screening for carriers of cystic fibrosis

Screening before pregnancy is needed

J A Raeburn

This is the eleventh in a series of articles examining some of the difficult decisions that arise in medicine

I believe that carrier testing for the common cystic fibrosis mutation ($\Delta F508$) should be offered in the United Kingdom before conception, to adults who wish it, after receiving and reading an informative explanatory leaflet. This is simple, unambiguous, and relatively inexpensive. The proposal recognises an individual's right to make an informed choice and separates that choice from life situations such as pregnancy, in which there is pressure to consider only specific options.

Three issues need to be considered: providing and receiving accurate information; whether a person wishes to know if he or she carries a testable mutation of the cystic fibrosis gene; and making a person understand that if the result shows him or her to be a carrier relatives should be told and offered testing.

The carrier screening programme should not aim to achieve a high uptake of screening. If 10% of the population of reproductive age chose to be tested during the first year it would be acceptable and appropriate. Success depends on a slow initial response as the community learns about cystic fibrosis; uptake could increase as the programme proceeds.

In previous approaches to screening a low uptake has been seen as inadequate and they have sought the higher rates achieved in pregnancy screening.¹⁻⁴ Why? An early low response is the bedrock of an ethically directed programme. The views of our communities vary.⁵

Why screen now?

Why should we start screening now, when health care costs and especially new developments are under close scrutiny? The answer lies in the importance of individual informed choice across different groups of the community. Tests for carriers of thalassaemia or

sickle cell disease have been performed since the 1970s among ethnic groups in which they occur. Screening for Tay-Sachs disease has also been offered for five years. Indeed, much of our knowledge of genetic screening has come from the careful studies of Asian, Afro-Caribbean, and Jewish groups.^{6,7} What is so different about cystic fibrosis that should make us delay testing now?

Opponents of screening for carriers of cystic fibrosis suggest that there is a danger of stigmatisation of people found to be carriers.^{8,9} Yet counselling and testing procedures should be confidential. The result would be known to those outside the family only if the person had chosen to tell. Stigma depends on someone being seen to be different and on the community's false assumption that different people are threatening; such labelling shows a lack of knowledge and a need for social education. A slowly developing screening programme would allow people to understand more gradually and should decrease any stigma.

Eugenic policies of 60 years ago are also cited as a reason for withholding cystic fibrosis screening. If this is true, why is it appropriate to offer screening for the commonest recessives in Asians, Afro-Caribbeans, Jews, and southern Europeans? If all people in our community are encouraged to make an informed choice about screening the responses will be diverse. The differing choices exercised will result in the opposite of a directive eugenic policy.

How should we screen?

If instead of focusing on high uptake we aimed at providing a high degree of individual choice, screening before pregnancy would enable more people to choose their preferred package with the same number of tests.

For example, if couple screening were offered to

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6000 pregnant women a year¹—that is, the number of deliveries in a fairly large maternity unit—about 80% might accept.² Among the 4800 couples thus screened about 156 carriers would be expected; after testing partners, five high risk pregnancies would be identified. Because the decision is to report as high risk only those couples who are both carriers, 151 (97%) of the carriers would not know their status (or that their pregnancy had an increased risk of around 1 in 400). It follows that about 600 relatives would remain unaware of their increased carrier risk.

Use the same number of tests a year in a different way—for example, by offering testing to 6000 people of reproductive age—and over 450 carriers would be identified, 156 initially and around 300 by cascade testing. After testing partners about 15 high risk couples could receive counselling before pregnancy. The box shows calculations which suggest that by testing 11.4% of a population we could identify about 30% of carriers.

Advantages of screening before conception

The strength of this approach lies in the opportunity it provides for gradual decision making, as people choose whether to have testing. If those being tested agree to disclose a positive carrier result to their relatives cascade tests can be considered. There is no need for most of a population to be tested, only those who choose. Testing does not need to be focused solely on pregnancy, with its lesser range of options.

Such a policy is easier for health authorities to introduce and is less expensive and less ethically suspect. The family doctor provides information about cystic fibrosis carrier tests,^{4,10} and the tests are then provided by regional genetics centres (with extra funding). The results are disclosed by arrangement between the screened person, the general practitioner, and the genetic centre, almost certainly using the skills of the genetic nurse specialist. By these means

Carrier screening for cystic fibrosis for health authority of population, 1 million

People aged 15-60	605 328
Estimated cystic fibrosis carriers (1 in 22)	27 515
Carriers identifiable (75%)	20 636
Assume 10% uptake of screening	(60 533)
Carriers estimated	2752
Carriers identified	2063
Cascade tests in carriers families (average 4 per family at 50% risk)	8252
Carriers identified	4126
Total carriers identified	2063+4126=6189
% of identifiable carriers	$\frac{6189}{20\ 636}=30\%$
Total screening tests performed	68 785
	(11.4% of population)

screening tests could be introduced in a manner that provides the most options to people who wish them in a way that is relevant for all common autosomal recessive conditions. Above all, those unwilling to be screened are not pressurised and the service develops in response to demand.³

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Psychological consequences are unclear

Theresa Marteau

Advances in genetics enable testing for a growing number of conditions. But simply being able to do a test is not sufficient justification for introducing a screening programme. There are several criteria for deciding whether to start a screening programme, including severity of the condition, accuracy of the test, acceptability of the test and any consequent treatment, and cost.¹ For population based screening programmes in which the intervention is the offer of termination of affected pregnancies, acceptability to patients is critically important. In addition, decisions about implementing a screening programme need to consider its contribution to a population's health needs.

Acceptability of a screening programme can be assessed by both its uptake and its impact on individuals.

How many people want screening?

Assessing demand as opposed to health professionals' enthusiasm for a screening test is difficult, but crucial. Whether people accept the offer of carrier screening for cystic fibrosis depends in part on how it is

offered. While over 70%, and in one study 95%, will have the test if it is offered immediately, only 25% will make a return visit for such a test.² This raises the question of how strongly people want the test.³

Among the few carrier couples who have been detected through population based screening, all the women have had prenatal testing, and all affected pregnancies have been terminated. But how well informed have these decisions been? Over the past 50 years, life expectancy for those with cystic fibrosis has increased from a few months to current estimates of 40 years.⁴ This is likely to improve further with new developments in treatment.

Contrary to the common image of chronic ill health and disability, the results of several surveys of adults with cystic fibrosis show that most are fit, active, and socially integrated.⁵ Women with cystic fibrosis, for example, are more likely to be graduates than women without the disease. Yet the information people having screening are given stresses the chronic ill health picture of cystic fibrosis. Indeed, most leaflets produced by those offering cystic fibrosis screening have been criticised as being too pessimistic about the prognosis for the condition.⁶ The demand for screen-