stated that they were not interested in guided self management plans, describing themselves as "already self managing competently" and "behaving responsibly." This reflects self reliance more than competent self management according to guidelines.

It also indicates a failure to integrate the personal and the medical dimensions of medical care[10]—that is, the integration of the medical agenda with the patient's perspective. Self management schemes have to combine the best of these two elements, but sharing responsibilities implies that patients as well as medical professionals should determine the goals of treatment. Ownership of a management plan is an important precondition to effective treatment for both patients and health professionals. It is not a question of whether guided self management is effective or should be implemented, but rather the challenge is to accept that patients are managing their care one way or another and that we need to create opportunities to clarify how medical input can enhance their personal situation. Cooperation is the key to bridging the gap between the efficacy and effectiveness of asthma care.

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BT has received grants from the Netherlands Organisation for Scientific Research and AstraZeneca for research into self management of asthma. CvW has received financial support for research and education from various pharmaceutical companies that have an interest in the care of asthma.

1 Gibson PG, Coughlan J, Wilson AJ, Abramson M, Bauman A, Hensley MJ et al. Self-management education and regular practitioner review for adults with asthma *Cochrane Database Syst Rev* 2000;3:CD001117.
2 Meijer RJ, Kerstjens HAM, Postma DS. Comparison of guidelines and self management plans in asthma. *Eur Respir J* 1997;10:1163-72.
3 Jones A, Pill R, Adams S. Qualitative study of views of health professionals and patients on guided self management plans for asthma. *BMJ* 2000;321:1507-10.
4 Tirimanna PR, van Schayck CP, den Otter JJ, van Weel C, van Herwaarden CL, Van den Boom G, et al. Prevalence of asthma and COPD in general practice in 1992: has it changed since 1977? *Br J Gen Pract* 1996;46:277-81.
5 Kolnaar B, Beissel E, van den Bosch WJ, Folgering H, van den Hoogen HJ, van Weel C. Asthma in adolescents and young adults: screening outcome versus diagnosis in general practice. *Fam Pract* 1994;11:133-40.
6 Robertson R, Osman LM, Douglas JG. Adult asthma review in general practice: nurses' perception of their role. *Fam Pract* 1997;14:227-32.
7 Thoonen BPA, van Schayck CP, van Weel C, Levy M, Spelman R, Price D, et al. Present and future management of asthma and COPD: proceedings from WONCA 1998. *Fam Pract* 1999;16:312-5.
8 Van Grunsven PM, van Schayck CP, van Kollenburg HJ, van Bosheide K, van den Hoogen HJ, Molema J, et al. The role of "fear of corticosteroids" in nonparticipation in early intervention with inhaled corticosteroids in asthma and COPD in general practice. *Eur Respir J* 1998;11:1178-81.
9 Van den Boom G, van Schayck CP, van Mollen MP, Tirimanna PR, den Otter JJ, van Grunsven PM, et al. Active detection of chronic obstructive pulmonary disease and asthma in the general population. Results and economic consequences of the DIMCA program. *Am J Respir Crit Care Med* 1998;158:1730-8.
10 Sweeney KG, MacAuley D, Gray DP. Personal significance: the third dimension. *Lancet* 1998;351:134-6.

Screening for familial hypercholesterolaemia

Effective, safe treatments and DNA testing make screening attractive

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Familial hypercholesterolaemia is a common disorder of lipid metabolism associated with a high risk of early mortality from coronary artery disease.[1] It is so common that general practitioners often have one or two families with the disorder in their practices, although they are frequently unaware of this.[2] People with familial hypercholesterolaemia often die from atherosclerotic heart disease before the age of 40; this is particularly true for men. These sudden deaths are tragic because they can easily be prevented once the condition has been recognised and treated properly.[3] It has been well established that the clinical sequelae of familial hypercholesterolaemia are the consequence of the extremely high concentrations of low density lipoprotein cholesterol that these patients have been exposed to since early childhood.[1]

The most effective and most widely prescribed class of cholesterol lowering drugs, the statins, was shown to be particularly effective in patients with this condition.[4] While it would seem logical to assume that treatment with statins would reduce mortality if family physicians simply diagnosed and treated this disorder, this line of reasoning is fraught with assumptions. Firstly, we must ask whether familial hypercholesterolaemia is indeed as common as we believe, and secondly, are we as inept in recognising and diagnosing this disorder as most of us believe? But the most important question is: how can we improve our recognition of these patients in general practice and offer them appropriate care?

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Fortunately, recent work from both the United Kingdom and the Netherlands has given clear answers to these questions. A recent survey of four general practices in the Netherlands indicated that familial hypercholesterolaemia occurs in 1 in 400 people; these numbers are likely to be similar in the United Kingdom.[2] A study by Neil et al, which recruited patients from the Simon Broome register for familial hypercholesterolaemia, the Oxford lipid clinic, and general practices throughout Oxfordshire, found that not even one quarter of patients with the condition are recognised in clinical practice, and most are not diagnosed until middle age, when atherosclerotic disease is already rampant.[5]

The weak points, if any, of this careful, prospective endeavour include the low response rate of the general practitioners, the possible errors associated with self reporting of family relationships, and the use of absolute cut-off points for cholesterol for diagnosis instead of DNA testing. The authors' findings are not jeopardised by these weaknesses, but their work may underestimate the true frequency of familial hypercholesterolaemia.

Now that it has been firmly established that familial hypercholesterolaemia is underdiagnosed it should be possible to remedy this situation. Familial hypercholesterolaemia is a genetic disorder that is autosomal dominant and fully penetrant in adolescence; so by definition one patient with the condition will lead you to many more just by examining the patient's family tree.[6]

Durrington and colleagues have used this old wisdom, and in this issue of the *BMJ* (p 1497) show that it is indeed possible to use this approach to find new patients.[7] By testing all first degree relatives of 200 patients with familial hypercholesterolaemia they found another 121 patients. In the general population at least 60 000 tests would have been needed to identify this many people with the condition. With the aid of a nurse specialist, simple cholesterol testing, and the use of small pedigrees Durrington and colleagues convincingly show that adopting an active approach to case finding works for familial hypercholesterolaemia.

Other investigators, including our group in the Netherlands, have come to similar conclusions, with two modest differences in approach. Firstly, testing in the Netherlands was not restricted to first degree relatives but included everyone in the extended family. This obviously reduces the proportion of people identified as having the disorder. On average, over a four year period one index patient led us to 20 additional family members, and eight new patients were identified (unpublished data). The second and most profound difference, however, lies in the use of DNA diagnostics. If the most sensitive test is used—namely age specific and sex specific centiles for total and low density lipoprotein cholesterol—16.6% of cases would have been missed and 12.5% would have been diagnosed as having familial hypercholesterolaemia when they actually had polygenic hypercholesterolaemia. Hence, active screening for a disorder requires a diagnosis that is rock solid, and that can only be provided by using DNA testing to actually find the genetic mutation causing the disorder.

Durrington et al correctly point out that the screening criteria developed by Wilson and Jungner easily apply to familial hypercholesterolaemia,[8] but it is unlikely that DNA testing for the disorder has harmful psychological consequences.[9–11]

We know how to organise the screening, and we have the capacity for testing, be it for cholesterol concentrations or DNA mutations. We also have safe and effective treatment that can save lives and money. Our ministries of health should not hesitate but should support screening and treatment programmes; a few specialised nurses working in close collaboration with lipid clinics could work miracles.

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1 Scientific Steering Committee on behalf of the Simon Broome Register Group. Risk of fatal coronary heart disease in familial hypercholesterolaemia.. *BMJ* 1991;303:893-6.
2 Lansberg PJ, Tuzgöl S, van de Ree MA, Defesche JC, Kastelein JJP. Prevalentie van Familiaire Hypercholesterolemie onder volwassenen in vier huisartsenpraktijken hoger dan werd aangenomen [Prevalence of familial hypercholesterolemia in four general practices higher than supposed]. *Ned Tijdschr Geneeskd* 2000;144:1437-40.
3 Scientific Steering Committee on behalf of the Simon Broome Register Group. Mortality in treated heterozygous familial hypercholesterolaemia: implications for clinical management. *Atherosclerosis* 1999;142:105-12.
4 Mol MJTM, Erkelens DW, Gevers Leuven JA, Schouten JA, Stalenhoef AFH. Effects of synvinolin (MK-733) on plasma lipids in familial hypercholesterolaemia. *Lancet* 1986;ii:936-9.
5 Neil HAW, Hammond T, Huxley R, Matthews DR, Humphries SE. Extent of underdiagnosis of familial hypercholesterolaemia in routine practice: prospective registry study. *BMJ* 2000;321:148-9.
6 Goldstein JL, Hobbs HH, Brown MS. Familial hypercholesterolemia. In: Scriver CR, Beaudet AL, Sly WS, Valle D, eds. *The metabolic and molecular bases of inherited disease*. 7th ed. New York: McGraw-Hill, 1995:1981-2030.
7 Bhatnagar D, Morgan J, Siddiq S, Mackness MI, Miller JP, Durrington PN. Outcome of case finding among relatives of patients with known heterozygous familial hypercholesterolaemia. *BMJ* 2000;321:1497-500.
8 Wilson JMG, Jungner G. *Principles and practice of screening for disease*. Geneva: World Health Organization, 1968. (Public health paper No. 34)
9 Andersen LK, Jensen HK, Juul S, Faergeman O. Patient attitudes to detection of heterozygous familial hypercholesterolaemia. *Arch Intern Med* 1997;157:553-60.
10 Tonstad S. Familial hypercholesterolaemia: a pilot study of parents' and children's concerns. *Acta Paediatr* 1996;85:1307-13.
11 Tonstad S, Novik TS, Vandvik IH. Psychosocial function during treatment for familial hypercholesterolemia. *Pediatrics* 1996;98:249-55.

Emerging arboviral encephalitis

Newsworthy in the West but much more common in the East

The recent outbreaks of West Nile encephalitis in New York and Israel are drawing the western world's attention to the potential threat of arthropod-borne virus (arbovirus) encephalitis.[1] But in many parts of Asia, infection with West Nile virus's sister, Japanese encephalitis virus, is a daily reality.

Epidemics of encephalitis were described in Japan from the 1870s onwards, and Japanese encephalitis virus was first isolated from a fatal case in the 1930s.[2] West Nile virus was isolated from the blood of a febrile woman in Uganda a few years later in 1937.[3] Both viruses are small enveloped RNA viruses, members of the genus *Flavivirus* (family Flaviviridae), named after the prototype yellow fever virus (*flavus* is the Latin for yellow). The flaviviruses are relatively new viruses, derived from a common ancestor 10-20 000 years ago, that are rapidly evolving to fill new ecological niches.[4] Both West Nile and Japanese encephalitis virus are transmitted in an enzootic cycle between small birds by *Culex* mosquitoes, though for Japanese encephalitis pigs are important amplifying hosts. Humans become infected by *Culex* mosquitoes coincidentally, but are not part of the natural cycle.

Although known to be widely distributed across much of Africa, southern Europe, and the Middle East, West Nile virus was, until recently, considered to be relatively benign.[3] It causes a non-specific febrile illness, or a characteristic fever-arthralgia-rash syndrome, which occurred in large epidemics in Israel in the 1950s and South Africa in the 1970s. Direct invasion of the central nervous system to cause encephalitis was thought to be a rarity. In contrast, Japanese encephalitis virus has always been recognised as a killer. Over the past 50 years it has spread relentlessly across Southeast Asia, India, southern China, and the Pacific—reaching Australia in 1998.[5]

Culex mosquitoes are unavoidable in rural Asia, and almost everyone is exposed to the virus. Only about 1 in 300 infections results in disease, and there is a wide range of presentations from a simple febrile illness to a severe meningoencephalitis, as well as a newly recognised polio-like acute flaccid paralysis.[6] There are

BMJ 2000;321:1484–5