faulty copper genes (almost always the result of consanguineous marriage) causes Wilson's disease. Heterozygotes are clinically well but may show some of the biochemical abnormalities.

Excess copper damages first the liver and then as copper is released from the dying hepatocytes it affects the brain, eyes, kidneys, bones and joints, and possibly the parathyroid glands.⁸ The definitive biochemical signs are, firstly, a low plasma concentration of caeruloplasmin, usually but not always below 200 mg/l, with low serum concentrations of copper (less than 12.6 $\mu\text{mol/l}$ (80 μg 100 ml)).⁹ Secondly, the urinary copper excretion is high. (A high urinary output of copper is also, however, seen in biliary cirrhosis.¹⁰) Thirdly, the copper content of the liver is raised and histological examination of biopsy specimens shows fine fat droplets, nuclear vacuoles, and a positive stain for copper. Fourthly, there is an overall diminution of copper incorporation into caeruloplasmin and a prolonged turnover of body copper. And, finally, rusty brown Kayser-Fleischer rings near the limbus of the eye are said to occur in all cases with neurological damage.¹¹

Over the past 30 years treatments for Wilson's disease have been largely developed by Walshe.^{12,13} He began with injections of the chelating agent BAL (British—as opposed to Russian—antilewisite; dimercaprol) and then in 1956-7 developed oral treatment with penicillamine—a metabolite of penicillin of no previous importance—and showed that this effectively “decoppered” six patients with Wilson's disease. More important, clinical improvement, though sometimes slow, was spectacular. Renal tubular defects, liver disease, and neurological problems—and the CT scan abnormalities—are all reversed.¹⁴

Treatment with penicillamine needs to be continued for life, and mild or severe sensitivity reactions are not uncommon. Patients who develop a rash, leucopenia, fever, a lupus-like syndrome, or the nephrotic syndrome may need to have the dose reduced, or treatment with cortisone, or the drug may have to be withdrawn. If penicillamine cannot be tolerated other effective treatments are fortunately available, the most satisfactory of which is triethylene tetramine dihydrochloride.¹⁵ Large doses of zinc by mouth also result in a negative copper balance.¹⁶

Our advances in the understanding of Wilson's disease have been spectacular over the past 70 years: over 1000 publications on the topic have appeared.¹ All too often, however, we are missing the diagnosis or making it too late.

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Changes in compensation for occupationally induced bladder cancer

Concern expressed in the *BMJ*¹ and elsewhere has led the Industrial Injuries Advisory Council recently to review neoplasm of the bladder as a prescribed industrial disease (PD 39) under the industrial injuries provisions of the Social Security Act 1975. This advisory body made recommendations in July 1983, which were accepted by the Secretary of State for Social Services and which became law in October 1983.²

The review was mainly concerned with three questions: whether the description of industrial bladder cancer was still satisfactory; what additional substances and occupations, if any, could be added to the terms of prescription; and whether there was evidence that carcinoma in situ could cause disablement before overt evidence of malignancy.

The council—whose members include doctors, lawyers, and representatives of industry, the trade unions, and insurance—recommended that the description of the disease should be amended to include carcinoma in situ and invasive carcinoma: all forms of transitional cell carcinoma of the urothelium are now included. It added benzidine to paragraph (a)ii as many people did not understand the chemical description of benzidine and its related compounds. It clarified the problem regarding auramine and magenta: the handling of these substances is not thought to induce carcinogenesis but their actual manufacture may produce tumours.

Methylene-bis-orthochloroaniline has now been listed as a substance under paragraph (a)i. This recommendation has been derived mainly from studies on animals showing it to be carcinogenic, and in this respect Britain is falling into line with the United States. Research work on this substance will be kept under review. In Britain a few men have probably developed cancer of the bladder from exposure to methylene-bis-orthochloroaniline.

Several other chemical compounds were considered by the council, but since the evidence was not convincing they were not included in the provisions. Nevertheless, research into (and evidence on) the effects of these substances is to be kept under review. Some evidence is accumulating that additional industries may have a raised incidence of neoplasm of the bladder—for example, printing. Concern has been expressed by the trade unions and the council has recommended a continuing review of epidemiological research into the occupational incidence of neoplasm of the bladder in specific occupations.

The last recommendation is that “every effort should be

made to ensure that potential claimants, doctors, and trade unions are aware of the occupational causes of neoplasm of the bladder." At present too few claims are made owing to a lack of awareness by the potential claimants, trade unions, and doctors concerned. The advisory council has made efforts to publicise these changes by sending details to the Royal College of Surgeons, the British Association of Urological Surgeons, the Royal Society of Medicine, the Royal College of General Practitioners, the Trades Union Council, and the Confederation of British Industry.

The changes made in 1983 certainly go a long way to meeting my criticisms expressed in 1982.¹ The disease can now be compensatable at an earlier stage—for example, for carcinoma in situ—and the changes should also allow a better understanding of the condition by people who write reports for either legal purposes or injury benefits. Unfortunately, the recommendations do not include any change in (a)ii, (a)iii, or (a)iv relating to the chemical description of the various chemicals (for the reasons stated in paragraph 9 of the report): the phrases nitro, primary amino, ring substitution by halogeno methyl or methoxy groups mean little to the average medical doctor or to me.

Official policy seems to be based on published reports on occupational or industrial cancer of the bladder, but this seems a very passive outlook. Surely in the future a more positive approach will be needed, perhaps through the Health and Safety Executive or the Trades Union Congress, so that occupations where the risk factors may be present are actively investigated. This might help to allay the suspicions and fears expressed in a recent television programme ("Picture of Health," Channel 4, 20 January 1984).

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Policies on prevention

Setting up committees rarely solves problems, and the proposal that every NHS district health authority should establish an "interdisciplinary heart disease team" seems an expensive way of promoting prevention of the disease. *Coronary Heart Disease Prevention*, the report of a conference held in September 1983, gives this suggestion priority, however, and suggests that around £12 million a year will be needed to finance prevention programmes in all the districts and NHS regions.¹ Surely this is an occasion when one or two regions might act as pilots to explore the possibilities rather than letting them all cut their teeth at the same time?²

Nevertheless, the report does recommend the policies that are essential if Britain is to join the other countries with a declining frequency of coronary heart disease. Firstly—and in our view most important—politicians must be persuaded that a responsible government should have a policy on health and be prepared to take account of that policy in its decisions. The immediate issue is nutrition and its relation with agriculture. As the report explains, "the present operation of the European Economic Community Common Agricultural Policy in relation to dairy products and sugar is directly opposed to the food and health policy the United Kingdom should be aiming for." At a time when farm policies are being reviewed it is essential that the nutritional objectives set out last year by the National Advisory Committee on Nutrition Education should be incorporated into government thinking.³ Food regulations and carcass grading (the amount of fat on cattle and sheep, at present favouring fat animals) should also be revised with the same objectives as targets.

Secondly, a government committed to a health policy would also, we believe, be more aggressive in discouraging smoking and reducing consumption of alcohol. Thirdly, a government initiative is needed for health education to be given priority in all levels of education—but especially in the training of teachers.

The report suggests that primary health care teams "should accept their important responsibility" for prevention. Certainly medical commitment is patchy—perhaps because so many doctors have been taught to be sceptical of the value of screening and health check ups. Disenchantment with multi-channel biochemical screening procedures should not, however, be used by doctors as an excuse for neglecting their obligation to seek out patients with symptomless hypertension or hyperlipidaemia—in whom treatment has been shown to be effective. Whether the proposals in the report for "motivating" the primary health care team will give similar clear cut gains seems less certain: changing peoples' attitudes to exercise and alcohol are not easy, nor are the optimum methods universally agreed.

No one would expect a report of this kind to satisfy all readers: preventive policies tend to provoke strong emotions because they necessarily impinge on individual freedoms and their paternalism upsets many people. The central concept of the report is, however, unchallengeable: as a nation we have delayed too long in formulating a strategy for preventing coronary heart disease. Both the medical profession and consumer bodies such as the College of Health should now maintain pressure on the government to recognise the need for urgent action.

¹ Steering Committee. *Coronary heart disease prevention*. London: Pitman, 1984. (Canterbury report.) (Chairman Geoffrey Rose.)

² Anonymous. Open letter to the new CMO [Editorial]. *Br Med J* 1983; 287:1903-5.

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