patients" completely prevents the development of clinical disease. Furthermore, the results of treatment in patients with overt disease are also impressive: in most cases symptoms and signs regress and the prospects of survival are good, whereas formerly virtually all patients died. Strickland et al examined the prognosis in 142 patients with Wilson's disease, including 88 who had been treated with penicillamine for up to 16 years. Of 36 symptomatic patients who had not been treated, all but one were dead; while 31 of the 35 symptomatic patients being treated with penicillamine remained alive, including 22 who had had penicillamine for two years or more; only six of these had residual symptoms. The overall experience suggests that the disease is completely and permanently controlled in patients who are lucky enough to be diagnosed and treated at an early stage.

Other patients do not fare as well—those with established cirrhosis and portal hypertension and renal tubular damage. Some patients present with fulminant hepatitis, often associated with acute intravascular haemolysis, when the only hope may be peritoneal dialysis. The rare patients who cannot tolerate penicillamine may be treated with the chelation agent triethylene tetramine hydrochloride. Hepatic transplantation is one radical alternative in patients with advanced disease; it may even correct the underlying metabolic abnormality.

The hepatic manifestations of Wilson's disease vary from asymptomatic cirrhosis to fulminant hepatic failure. Doctors must maintain a high index of suspicion, especially in young patients, though the middle aged may also present with hepatic disease. This need for awareness is especially true of chronic active hepatitis, which is thought to be due to copper overload in as many as 5% of cases. Scott et al. from the Royal Free Hospital recently published a series of such patients in whom the outcome was disappointingly poor even with penicillamine treatment: over half of them died within two years of presentation. They suggested that diagnosis may frequently be delayed beyond the point of optimal response to treatment in this syndrome, which is clinically and biochemically indistinguishable from other types of chronic active hepatitis unless copper metabolism is studied. Once suspected, the diagnosis may be established without undue difficulty on the basis of raised copper concentrations in the liver and in the urine. Scott et al. found that these were raised in all cases, though the serum caeruloplasmin concentration was normal in three patients; the results of ruban acid staining of liver tissue for copper proved unreliable.

Clearly the best use of the lifesaving potential of penicillamine treatment in Wilson's disease comes from early diagnosis. Suitable diagnostic procedures have been reviewed recently by Sternlieb, who argued that—despite the many reported pitfalls—most patients are diagnosed correctly with standard investigations and that few need such refinements as radiocopper loading tests. Nevertheless, the clinician's most potent ally remains an awareness of the possibility of Wilson's disease in the differential diagnosis.

Storage and cooking of poultry

As Christmas approaches, it is safe to predict that there will follow the usual reports of food poisoning in people who have eaten turkey, chicken, duck, or goose. The amount of distress and trouble caused by such food poisoning is well known. Not infrequently cold chicken is the vehicle of infection. The bird is often one that has come from a deep freeze and has been thawed for only an hour or two before being roasted for too short a time at too low a temperature to guarantee that even vegetative bacteria will be killed. Inadequate heating is often followed by too slow cooling, and, worst of all, the chicken may simply be left overnight at room temperature. In this way a temperature favourable for bacterial growth is maintained for enough hours to ensure that the carcass is heavily colonised by the rapid growth of any salmonellas not killed by the cooking.

As a result a large proportion of those who have eaten the chicken are shortly stricken with severe abdominal pains, diarrhoea, vomiting, and possibly a generalised infection with fever. Food-handlers may be laid off work for many weeks if—as often—they carry the salmonella that has infected them from the chicken. The inference is not hard to draw and the lesson to be learned should be: let your hot be hot and your cold be cold. Such incidents, causing much distress and an incredible amount of work to all concerned in their investigation and management, are not properly described as “accidents” because they are readily preventable by applying the elementary rules of kitchen hygiene.

In present conditions, it is well to assume that any chicken, goose, duck, or turkey carcass may be contaminated with food-poisoning salmonellas or spoilage bacteria. The probability of such contamination is much higher in deep-freeze poultry than in unrefrigerated birds. One day, perhaps, salmonella infection of poultry may be cut off or much reduced by measures taken in breeding establishments and processing plants and in animal-feed production; but this remains only a future hope. So it is essential to make sure that frozen poultry is well thawed, properly cooked, and stored in the cold if not to be eaten at once after cooking. This advice can be specified in some detail. Thawing a deep-freeze carcass should mean leaving it for a full 24 hours at room temperature; because, if a bird is only partially thawed, the penetration of heat will not be enough to destroy even vegetative bacteria in the deeper parts of the bird. Cooking must be done in a pre-heated oven and the time and temperature requirement is about 180-200°C for 20-30 minutes per pound plus 30 minutes extra. The higher times and temperatures are for larger birds—say, over 10 lb—and the weight of any stuffing must be included in the calculation.
If the roasted bird is not to be eaten until later it must be stored in a properly managed refrigerator, preferably at 4°C or below. The importance of this requirement is brought out in the report of a well-designed and conducted study by Toule and Murphy. They identified the bacteria contaminating refrigerated cooked chicken and examined their spoilage potential and possible origin. Briefly, they found that the variable temperature range common in a normal kitchen refrigerator (2-13°C) resulted in a greater number of bacterial species and in total counts tenfold greater on the cooked chickens thus stored than in those kept at 4°C or below. The lesson is that refrigerators should be kept away from the humid atmosphere of cooking ovens, preferably in a different room, and that doors should be opened as seldom as possible and closed as quickly as possible. They found that cooked chicken was much more vulnerable to bacterial spoilage than was raw chicken and that a normal kitchen provided numerous sources of spoilage bacteria—including, for example, air of the kitchen and refrigerator, working surfaces, cutting equipment, socially clean plates, and water taps. Since many of these spoilage bacteria grow well and rapidly at refrigerator temperatures, they conclude that the shelf-life of cooked chicken is often no more than two days at 4°C.

Hence, the precautions needed to keep our Christmas fare both safe and wholesome mean a reasonable amount of thought and care but should cause no insuperable problems.

1 Toule, G, and Murphy, O, *Journal of Hygiene* (Cambridge), 1978, 81, 161.

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**Who cares for the mentally handicapped?**

Established policies for the care of both the mentally ill and the mentally handicapped have been challenged and almost reversed in recent years. In the nineteenth century the asylum was a place of refuge for idiots; and Colonel W W Ireland, among others, made a plea for more adequate medical care in such places. The notion that institutions protected such people from being exploited by the public persisted until the beginning of the National Health Service—the question “Whether ever in moral danger?” being printed in local authority case notes alongside “Whether convicted or been in the hands of the police?” There was also a strong lobby in favour of stopping the “propagation of the unfit” by locking them up.

Not surprisingly, with this legacy of different and sometimes contradictory goals, hospitals for the mentally retarded are not ideally equipped as a base for rehabilitation for life in the community or even as a substitute home for the inmates. These hospitals became an integral part of the Health Service when it was founded; but, as Professor P Mittler and his colleagues of the National Development Group for the Mentally Handicapped say in their report to the Secretary of State for Social Services, these institutions often find it hard to get expert help for the many complications to which the mentally retarded are prone. The report sets an ironic goal—to ensure at least as good a quality of specialist help for those who happen to be in hospital as for those who live in the community.

In his introduction to the report Mr David Ennals admits that mental handicap hospitals have long been one of the most neglected and deprived areas of the NHS: though many improvements have been made recently, any visitor cannot fail to be impressed, he points out, by the contrast between the life lived by many of the residents and that which most of us enjoy. The Secretary of State commends improvements that can be made within existing resources but makes no promises about additional financial help. But, as this report shows, more funds are essential from both the NHS and local authorities if we are to achieve any real improvement.

Some 50,000 citizens of Britain are living in hospitals for the mentally handicapped, and 20,000 have lived in hospitals for 20 years or more. The report repeats that many thousands do not need to be in hospital at all and wants the Government to do much more to provide the resources required by their policy of community care.

Meanwhile we are left with hospitals as they are and think the report spells out the shortcomings. One of the cardinal problems at present is lowered morale and uncertainty about the future. Money for more nurses is hard to come by, reducing beds improves the ratio, but even when there is money for staff those of the right calibre may not be forthcoming. And the discontent of nurses extends to other professions.

The report again commends a district attachment for the psychiatrist concerned with the mentally handicapped; and it again emphasises that we should avoid consigning mentally handicapped children to hospital, and instead put them under the care of paediatricians and community physicians. Doctors and other specialists are, however, reluctant to opt for this work, while deplorably few psychiatrists in training are prepared to devote themselves to the care of the mentally handicapped.

We need more work like that of pioneers such as Lionel Penrose to raise the academic status of the subject; the Medical Research Council and the universities should give a clearer lead. Uncertainty should be ended, and if these hospitals are to stay for 20 to 40 years or more this must be said boldly. Joint funding by the NHS and local authorities may be a partial answer. At all events, professional staff must be guaranteed a future within an integrated service for the mentally handicapped.