the present stage—which entailed the design, synthesis, and screening of over 70 chelators in vitro and in vivo and the identification of several other promising 1-substituted-2-alkyl-3-hydroxypyrid-4-one derivatives similar to L1—took nine years.⁸ The absence of side effects in over 20 patients and volunteers who received L1 18 months ago, some for up to three months and at doses of up to 8 g (4 × 2 g) in a day, suggests that the drug has a high degree of safety when therapeutic doses are given long term.

Despite the above results and the recent discovery of a new method of synthesis of L1 and related derivatives, which will make them about 50 times cheaper than desferrioxamine,³ pharmaceutical companies may become fully interested in orphan drugs such as L1 only after scientists carry out most of the work required for licensing, including both toxicological and clinical trials. The prospect of fully developing L1 and related chelators will also improve if other uses, such as the treatment of aluminium overload, are taken into account. It is highly likely with further positive results from our expanding and continuous clinical trials that L1, and possibly other related oral drugs, will be available for the treatment of patients with thalassaemia within the next few years.

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Outpatients

SIR,—The article by Mr M Duncan and others (30 April, p 1247) makes sorry reading. I have carried out a similar study of the efficiency of the outpatient service in the department of internal medicine in this hospital.

We use three or four consulting rooms, where the patients are seen initially by a junior member of staff. The professor moves from room to room, where he is given a summary of the problem and a plan of management. Thus he sees every patient. This system also provides a useful educational forum and avoids interval time as defined by Mr Duncan and others. In their study this accounted for 25% of the total clinic time.

Since January, basing my calculations on 13 sessions, an average of 25.1 (SD 3.46) patients were seen at each session, of whom 25% were new referrals. Each session lasted 286 (20) minutes, and thus the patient-doctor contact was 11.4 minutes—some 50% longer than in the authors' study. Altogether 327 patients were seen. In not one instance were notes or relevant clinical data missing.

This department, with responsibility for 60 beds, has three full time medical secretaries and

one records clerk. The secretaries are paid Frs 5000-6500 monthly and the clerk about Frs 4500-6000 (before tax). In the private sector they might command up to 50% more. I support Mr Duncan and his colleagues in their conclusion that these staff play a vital role in the efficient working of outpatient departments but suggest that useful alterations might be made in the manner in which these sessions are run.

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Care of physically handicapped young adults

SIR,—Dr Martin C O Bax and others (23 April, p 1153) are to be congratulated on their study of the needs of handicapped young adults. Support does not necessarily require specific teams as is shown by the following case report.

A 44 year old divorced woman is quadriplegic as a result of multiple sclerosis and depends on others for turning in bed, transfers, washing, dressing, feeding, and all household tasks. Specific help is needed for bladder and bowel functions. Her mental state is normal, and there is no evidence of intellectual or cranial nerve involvement in the disease. She lives with her daughter in an extensively modified council house.

In 1980 she developed a large ischial pressure sore. By 1981 she was needing about 80 hours' care each week by the community nurses (including two calls nightly) and social services. Stress among these professionals was increasingly apparent, and interprofessional problems occurred almost daily.

Case conferences with senior representatives of all the professions concerned confirmed a commitment to maintain the mother and daughter in the community and avoid residential care for the mother and foster care for the daughter. Close liaison between the care attendants and community nurses ensured that the patient was not left alone for more than three hours. This overcame the isolation which had resulted in her making excessive demands on nursing time. A low air loss bed was provided, avoiding the need for readmission to hospital for pressure care over the six years of follow up.

Who does what?

Care attendant

Meals, transfers from chair to bed and back, hair washing, emptying urine bag, help with washing, sewing, mending, writing letters, dressing

Home care worker

Cleaning house, laundry, shopping, and paying bills

Community nursing

Specific nursing care for pressure sores, catheter and bowel care, and bathing

Social worker

Counselling

Disabled living adviser (occupational therapist)

Advice on practical changes in the environment—for example, Possum

Adaptation technician

Minor alterations to home

Domiciliary physiotherapist

Maintenance of level of function, management of spasticity

Hospital

Support for community services when needed

General practitioner

Continuing personal care

It has been estimated that between 1968 and 1986 the health and social services capital costs exceeded £30 000, and the costs for 1986-7 were at least £330 each week.¹

This woman obtains her community support from the pool of domiciliary workers (see box) and not from a specific team. The willingness of the medical staff to work with the social services management,² the production and monitoring of a plan by multiprofessional conferences,^{3,5} the presence of a health service equipment budget to support people in the community and the devolution of many care tasks from the community nurses to social services workers⁶ were important features of her care. This patient has provided a model for the development of community care for severely physically handicapped people in Harrow.

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Diagnosing Marfan's syndrome

SIR,—I agree with Dr Maurice Super (14 May, p 1347) that the main reason for diagnosing Marfan's syndrome is to offer screening by echocardiography for the one fatal complication—aortic aneurysm, which can rupture or dissect with an 80% mortality rate. Elective replacement of the aneurysm with a dacron graft has a 5% mortality rate with an 87% survival rate after five years of follow up.¹ This operation is offered when the aortic root diameter reaches 5 or 6 cm, but the family history must be taken into account. We have advised surgery at an aortic root diameter of 4.4 cm in an affected daughter with severe unexplained chest pain whose father had died suddenly of aortic dissection at that precise aortic diameter.

When can you make a diagnosis of Marfan's syndrome? The consensus of opinion seems to be when two of three systems (eye, heart, and skeleton) are classically affected. Preferably there should also be a positive family history. The most common diagnostic errors such as dominantly inherited mitral valve prolapse with marfanoid habitus (in the absence of aortic root dilatation)² and joint hypermobility syndrome with marfanoid habitus³ are thus avoided. Dr Super's suggested scoring of major and minor criteria would make an erroneous diagnosis of Marfan's syndrome in both these instances and in mild, dominantly inherited Ehlers-Danlos syndrome type IV.⁴

If a patient over 20 years of age has a normal echocardiogram the longterm prognosis is excellent, and a letter stating this should help to obtain reasonably priced insurance and mortgages. When the diagnosis is in doubt records should never contain reference to Marfan's syndrome but rather to a marfanoid condition, even though regular surveillance may be continued until the diagnosis is certain. Ocular, skeletal, and major cardiovascular complications should have developed by age 18, and in their absence the family can be counselled that this is not Marfan's syndrome and that the prognosis is good.

The Marfan Association has 450 member families