may be a more effective means of changing behaviour in traditional communities than the mass media.

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A little space in Sheffield

Life sciences research in space is still in its infancy despite solving the initial problems of keeping humans alive in such an inhospitable environment. To the earthbound clinician such endeavours have had little professional impact, although there are promises of greater understanding of cardiovascular, respiratory, and vestibular physiology, mineral metabolism, and locomotion. The prospect of such research has encouraged the University of Sheffield to set up the United Kingdom's first institute for space biomedicine, which was inaugurated on 19 October.

The institute's funding is limited at present to a two year grant totalling £50 000 from the university. The announcement about finances produced some surprise at the launch press conference, but the director of the institute, Professor Tim Scratcherd, confirmed that "We're currently thinking in terms of £10³ rather than £10⁹." Scratcherd would welcome government funding and he had been optimistic after initial positive discussions with the British National Space Centre. The rest of the story is history: Roy Gibson, the director of the centre, resigned from his post on 30 September owing to the government's decision not to increase its annual contribution to the European Space Agency's budget. So not surprisingly the timing of the institute's launch provided a forum for considerable discussion on government funding of space research in general.

Sir Geoffrey Pattie, ex-information technology minister, who opened the institute's launch symposium, declared that a "fog of myopia had fallen on the government" on the subject of investment in space. As a world trader and producer Britain could not afford to opt out of this field, yet our spending was "virtually in the noise level." Further evidence of the United Kingdom's lack of commitment to space research, especially in the biosciences, was presented by Dr Heinz Oser, director of life sciences of the European Space Agency. The United Kingdom's contribution to the agency's microgravity programme was about 2% of the total, although smaller countries such as Belgium and Switzerland had each paid over 4%. The bulk of the finance came from Germany (35%), Italy (18%), and France (15%).

Given this impoverished background how can Sheffield possibly succeed in contributing appreciably to space biomedicine? Much will depend on whether the institute will be offered a free flight to put an animal "test bed" into orbit. There were strong hints that the likely benefactor would be the Soviet Union, which would offer a mission in the late 1980s or early 1990s. This will presumably employ the Soviet reusable biosatellites, which were later described at the symposium by Academician Oleg Gazenko, head of the Institute for Biomedical Problems, Moscow. With a weight of about five tons, the craft provides about three cubic metres of useful space for automated studies on the animals restrained within it. In the 1990s a larger capsule is planned, probably three times the present size, which could dock with manned space craft and allow humans to work directly with the animal studies.

If a flight is given to the institute the likely priority for investigation will be the demineralisation of bone that occurs with prolonged space flight. The rapidity with which this occurs in space makes it a useful potential model for predicting the mechanisms concerned in terrestrial osteoporosis. During its investigations the institute would make use of skills and resources from the nine departments at Sheffield with which it has links and, in particular, the department of human metabolism and clinical biochemistry.

Inevitably the institute's autonomy will suffer if its major research expenses are to be met by a third party. I hope that it will have the opportunity to develop into more than just a service laboratory for the Soviets.

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Degenerative ataxic disorders: still perplexing

Degenerative disorders of the nervous system, including dementias, motor system disorders, and the spinocerebellar degenerations, are presenting a bigger problem as the population ages. The disorders result from selective and symmetrical degeneration of one or more groups of neurones. Some are wholly genetically determined, but most are not; often the disease develops because of a combination of genetic and environmental factors. The cerebellar and spinocerebellar degenerations comprise a complex group of over 50 distinct diseases. Their classification is controversial, but in practice they are best subdivided according to clinical and genetic features[1][2] rather than pathological criteria.[3]

Degenerative ataxias with recognisable metabolic defects are rare, but their diagnosis is important for genetic counselling and treatment. Some storage diseases more commonly associated with neurodegenerative disorders of childhood, such as hexosaminidase deficiency and the leucodystrophies,

may present in adults with a multisystem syndrome including cerebellar dysfunction.[4-6] Cholestanolosis, a rare autosomal recessive disorder caused by defective bile salt metabolism, causes ataxia, dementia, spasticity, peripheral neuropathy, cataract, and tendon xanthomas in the second decade of life. Treatment with chenodeoxycholic acid appears to improve neurological function.[7] Longstanding severe vitamin E deficiency causes a syndrome of progressive ataxia, areflexia, and distal loss of vibration and position sense. This may occur secondary to acquired fat malabsorption and is the basis for the neurological syndrome associated with abetalipoproteinaemia.[8 9] Recently patients with isolated vitamin E deficiency, probably inherited as an autosomal recessive trait, have been reported.[10] Adequate doses of vitamin E stop the progression of neurological symptoms and lead to improvement in some cases.[8 9]

Degenerative ataxic disorders of unknown cause that begin in childhood or adolescence are nearly always genetically determined, and most are autosomal recessive. The commonest of these is Friedreich's ataxia, which usually first causes symptoms between 8 and 16 years. Its cardinal features are progressive ataxia of gait and the limbs, dysarthria, and areflexia; signs of pyramidal tract dysfunction and loss of vibration and joint position sense eventually appear in all patients. Scoliosis and electrocardiographic evidence of heart muscle disease occur in about 70% of cases of Friedreich's ataxia. Less common features include pes cavus, optic atrophy, distal wasting, and diabetes. On average patients lose the ability to walk 15 years after symptoms begin and die in their 40s and 50s.[2] The cause of Friedreich's ataxia remains unknown, and early reports suggesting deficiencies of the pyruvate dehydrogenase or mitochondrial malic enzymes have not been confirmed.[11-15] Efforts to localise the gene for the disease to a specific chromosomal region have so far excluded it from about half of the genome (S Chamberlain, personal communication).

When presented with adults with slowly progressive cerebellar dysfunction doctors must exclude hypothyroidism, alcoholism, and malignancy, although paraneoplastic cerebellar degeneration usually causes more acute illness. About one third of patients with degenerative ataxia developing over the age of 20 have similarly affected relatives, and inheritance is usually autosomal dominant. The pathological findings in late onset cerebellar degenerations are variable, but the syndrome of olivopontocerebellar atrophy (loss of cells in the pontine and olivary nuclei and also of the cerebellar Purkinje cells) is seen in at least half of familial and sporadic cases. There are nearly always accompanying degenerative changes in the basal ganglia, spinal cord, and peripheral nerves, so the term olivopontocerebellar atrophy is misleading and not a good diagnostic label.[16]

Most patients with late onset ataxia that is dominantly inherited do not have a pure cerebellar syndrome; commonly they also have features such as supranuclear ophthalmoplegia, optic atrophy pigmentary retinopathy, dementia, and extrapyramidal dysfunction.[2] Sporadic cases are clinically similar; the aetiology of their disorders is not known, but some probably represent fresh dominant mutation.[2] It has been suggested that leucocyte glutamate dehydrogenase activity is specifically reduced in some patients with olivopontocerebellar atrophy,[17 18] but a recent British study has not confirmed these observations (D Aubby et al, personal communication).

Most attempts at treating degenerative ataxias have given disappointing results, reflecting our incomplete knowledge of neurotransmitter function in the cerebellum and its connections.

γ-Aminobutyric acid appears to be the most important inhibitory neurotransmitter, but trials of agonists to γ-aminobutyric acid—such as sodium valproate, baclofen, and isoniazid—have failed to show functional benefit.[2 19] The same applies for cholinergic drugs.[19] Promising results using thyrotropin releasing hormone in treating ataxia have been reported from Japan; these observations need to be confirmed, but they suggest that noradrenergic input to the cerebellum is more important than previously realised.[20]

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Important new treatments for acute ischaemic stroke?

Three recent trials have suggested that glycerol,[1] naftidrofuryl,[2 3] and nimodipine[4] may reduce mortality[1 4] or morbidity[2 3] after acute ischaemic stroke. Treatment with glycerol reduced the odds of death within seven days by 68%,[1] and treatment with nimodipine reduced the odds within four weeks by 70%.[4] Although naftidrofuryl did not have a statistically significant effect on early death, the authors concluded that the observed reductions in morbidity were statistically and clinically significant.[2 3] All three trials were, however, small, including only 173,[1] 164,[4] and 89[2 3] patients. For this reason alone they do not provide evidence reliable enough to justify nationwide use of the treatments.[5]

Combining the results of several small trials that are individually uninformative with all other available methodologically sound trials in a formal statistical overview may