

An accidental career in a new discipline

Helen P Beange

At seventy-seven it is time to be in earnest

Samuel Johnson — A journey to the Western Isles of Scotland

Nothing about my life has been planned, including medicine. When, to my surprise, I obtained one of the rare scholarships then available to the University of Sydney after the Leaving Certificate, I asked my father what I should do. “Medicine”, he said, “that’s a good career for a woman”. So, despite having no sciences and braving the fierce opposition of my Latin teacher, I obeyed. Nor did I know, even after graduating, what field of medicine to aim for, having spent much of my undergraduate years at Arts lectures and playing bridge.

After doing the odd locum and a bit of general practice, I married a naval aviator and had six children. This removed me from medicine for 13 years. Returning to Sydney from Perth (following the fleet) and being short of money, I went to Royal North Shore Hospital (RNSH) and asked to do a refresher course by attending outpatient clinics. Although feeling like a middle-aged frumpish housewife, I was treated by each of the senior consultants, all men, with the utmost courtesy. I have honoured RNSH ever since.

It was not easy to get a job. I worked first at Grosvenor Diagnostic Centre, assessing children with intellectual disability. The medical officers were at the fringe of a revolution in genetics, constantly learning about recently discovered syndromes and how to identify these as the underlying causes of the conditions in the people we saw. Yet, we were not esteemed by the medical profession, who referred to us as “guardians of the waiting list”, because we assigned priority to those most urgently in need of the few residential care places available. Our chief concern, however, seemed to be for the parents, and how to reconcile them with the pain of accepting that their children were “different”.

From institutions to community care

When my husband was posted to Melbourne, I worked in a large institution in Victoria, which taught me a lot about institutions and the danger of just accepting what occurred in them. I remember admitting a teenage boy, clinging to his tearful mother. She was at last persuaded to leave, too soon to notice that his bed had no pillow. I asked for one, but the busy charge nurse’s reply was, “I won’t allow pillows in my ward. They might fight with them or smother themselves”. Of course, we were understaffed, the few staff we had were overworked, and the residents had little supervision. So little, in fact, that one girl wandered out of her ward one night and fell into an uncovered hole containing a steam pipe — she was scalded to death. When I recollect this tragedy, I

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A trip to the snow



Royal North Shore Hospital Health Promotion Clinic for Adults with Developmental Disability enjoying a weekend in the mountains.

find it hard to accept the pious phrase “the dignity of risk”, which is too often used to justify neglect of people in residential care.

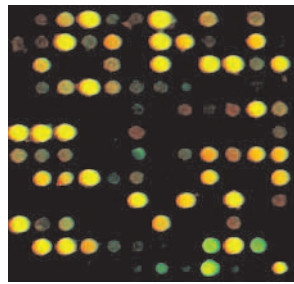
Returning to Sydney, I was lucky to be at the start of the deinstitutionalisation program in New South Wales, but soon realised that this was not to be an easy path. Among the first people to move were a group from a large institution on Peat Island. They moved into a small cottage in Hornsby, and soon the neighbours were complaining, not because of discrimination, but because of the frequent noisy quarrels among the four residents about who should do the dishes! Human nature is no different whatever the IQ level. This same quartet looked marvellous — dressed for the first time in dinner jackets and evening gowns — for their first formal, a ball in Hornsby.

As I grew older, I became interested in exercise and nutrition. I started to jog in the streets near where I lived, embarrassing my children and husband. It felt so good that I wondered how exercise would work for people with a disability. I obtained a Master of Public Health degree by doing a simple research project on this subject. Then RNSH allowed me to start a health promotion clinic for adults with developmental disability. This was soon enhanced by the arrival of a dietitian, Lyn Stewart, and an exercise physiologist, Caroline Bruce, and we became very busy. It was obvious to me that my colleagues, using nutrition and exercise, improved health faster than I did. Caroline also organised the people into teams and they went for trips at weekends, playing indoor cricket and soccer, and returned with amazing stories of their hijinks.

This was a time of fanatical adherence to the philosophy of normalisation — all people with disabilities should live in the community, whatever their special needs, and participate in all community activities. Our staff struck some unrealistic expectations, including carers who refused to be involved in our sports programs, one proclaiming, “We’d rather our residents sat at home all day and never exercised, than take part in segregated activities.” We ignored these thought police, and took one group ourselves for

My mentors and medical detective work

I first met **Professor Gillian Turner** as her locum at Grosvenor. Gillian is now known internationally as a geneticist and has done the fundamental work on identifying the Fragile X syndrome, the most common inherited genetic cause of intellectual disability. This work began when Gillian was a medical officer at Grosvenor and introduced me to the intellectual fascination of dysmorphology and genetic disease. I enjoyed being a medical detective and identifying syndromes. The explosion then occurring in



Microarrays — miniaturised assay systems allowing the structure and expression of thousands of genes to be evaluated

genetics is about to recur, with new technologies such as microarrays (Figure) that will make diagnosis more accessible. Given that a diagnosis can make all the difference to prognosis and treatment of associated physical disease or behavioural phenotype, a group of people much ignored by medicine will be taken more seriously in the future.

I once excitedly asked **Associate Professor Aidan McElduff** (Endocrinologist) in the corridors of Royal North Shore Hospital whether a patient I had referred to him could have Kallman's syndrome. Aidan looked surprised and said he supposed she could. This turned out to be the underlying aetiology. I was in awe of Aidan's many qualifications and his far greater knowledge of medicine, but he has since become a mentor, a coauthor of published articles, a collaborator in studies on osteoporosis and hypogonadism, and an expert on the endocrinology of intellectual disability. Aidan once thanked me for introducing him to a new medical specialty, Intellectual Disability Medicine.

their first sight of snow to luxurious quarters donated by a generous hotelier. At breakfast they sat down to silver and snowy table linen. "Oh look!" said one delighted young man, "we are going first class!"

Latin and the logic of epidemiology

The best teacher I ever had was a Dominican nun, Sister Anselm. She made me love Latin as much as she did. I later took the same delight in epidemiology, the closest thing I have found to thinking in Latin. This prompted my postgraduate studies in public health, completed with a part-time scholarship (awarded to promising public servants in the enlightened practice of the day). I applied epidemiology to the little-studied burden of disease in our patients, in whom I noticed diverse chronic and complex health problems. I obtained a Research and Development Grant from the Commonwealth Department of Community Services and Health for the first population study of health disorders in people with intellectual disability. This showed that, of a random sample of 202 adults with intellectual disability, each had an average of 5.4 medical disorders, half of which had not previously been detected.¹ Colleagues in Victoria and Queensland reported that these findings supported their argument for specialised health clinics in those states. Not so enlightened were authorities in NSW, who closed the RNSH Health Promotion Clinic, in the face of many objections. The Department of Community Services' mysterious decision ended what was, as far as I know, the first health

promotion clinic of its kind in the world. Since then I have tried unsuccessfully to reopen this clinic.

An attempted retirement

At that stage, RNSH enforced retirement at the age of 65 years. I was a bored, cross, retired person, so I leapt at the chance to work for a while at Stockton Centre, a large NSW institution for people with intellectual disability. The enlightened administrator at the time, Lorraine Yudaeff, asked if I could find someone to assess the nutrition of the residents. I recruited Lyn Stewart, who had worked with me at RNSH. Lyn ran a tape-measure over some emaciated residents and recommended an enriched diet. This resulted in another study showing that 69 of the multiply-disabled residents in this 550-bed institution were severely underweight, with low levels of vitamin D, iron and folate. They were given a diet with increased caloric density for 12 months, with significant improvements in weight, in serum calcium and vitamin D levels, and in haematological indicators.² We succeeded in increasing the weight of some very hungry people, but not without protest: one carer said we had ruined the quality of life of an adult resident who now weighed more than his previous 20 kg. He required two people, instead of one, to lift him, and was considered too heavy to take on outings, as they had no hoists. However, the noble staff of Stockton rose to the challenge. Several residents now verge on the overweight, not underweight! As a result of this work, there is now a policy of screening the nutritional and swallowing status of all people with intellectual disability in residential care in NSW.

Battling medical nihilism

It was generally considered inevitable that people with intellectual disability die young, which prompted Seeta Durvasula (Medical Lecturer in Developmental Disabilities, Centre for Developmental Disability Studies, University of Sydney), Wes Baker, Senior Planning Officer, Northern Sydney Health, and myself, to investigate mortality rates in a previously identified cohort of 693 people with intellectual disability in northern Sydney.³ We found that the death rate was five times higher than that of the general population of Sydney's Lower North Shore.⁴ Furthermore, the causes of death were quite different from those in the general population, being predominantly respiratory disease and accidents. Seeta Durvasula recently presented data from this ongoing study at an international conference, showing that up to a quarter of these deaths were preventable. Contributory factors, such as inadequate supervision, delayed presentation and delayed diagnosis of illness, were identified. The challenge is now to reduce premature mortality and to treat remediable disease. The NSW Ombudsman is charged with the responsibility for reviewing all deaths of people with disabilities in care, looking at the causes and patterns of death and recommending ways of improving services to reduce early or preventable deaths. This important initiative has been noted internationally and may be replicated in Scotland.

We also need to beware of medical nihilism. It is one thing to strive officiously to prevent the demise of an elderly person ravaged with disease. It is another thing to deny treatment to a young person with spastic quadriplegia who is enjoying life but swallows unsafely. I am reminded of a senior consultant with whom I discussed the need for supplementary feeding in some young individuals. "Why treat them", he said, "aren't they just vegetables?"

Receiving an award



Member of the Order of Australia (AM) in 2004, receiving the award from Her Excellency Professor Marie Bashir, Governor of New South Wales (left).

An international movement in intellectual disability

I presented papers at meetings of the International Association for Intellectual Disability (IASSID) every four years, but found health professionals at these conferences often cowed by our non-medical colleagues, who considered us relics of the past, detested institutions. Encouraged by the then Australian President of IASSID Professor Trevor Parmenter (now Foundation Chair and Director, Centre for Developmental Disability Studies, University of Sydney), a Dutch physician Professor Heleen Evenhuis and I formed the Health Special Interest Research Group of IASSID. This Research Group now has yearly international meetings, and its achievements include developing health targets for adults with an intellectual disability.^{5,6} These targets address conditions that are highly prevalent, easily identified and amenable to available treatments (eg, regular assessments of hearing, especially if communication problems exist, and of dental and ocular health). The targets have been accepted by IASSID and presented to the World Health Organization for ratification.

The future: developmental disability medicine

A medical specialty will be established in this field. In The Netherlands, such specialists already exist, while, in the United Kingdom, psychiatrists who have specialised in learning disability are often attached to multidisciplinary teams working with people who have learning disabilities.

It is clear, too, that doctors with such specialist knowledge must collaborate with nurses, psychologists, dietitians, speech pathologists, physiotherapists and occupational therapists. If it is possible to have multidisciplinary teams in aged care, such teams should

also be available for people with disabilities. Of Australia's total population, 1.86% have an intellectual disability.⁷ Thus, this group of people constitutes a population about the same size as our Indigenous population. In my opinion, a specialist clinic at each teaching hospital in Australia is required to support general practitioners caring for people with intellectual disability. In Victoria and Queensland, at least, university-affiliated academic centres for developmental disability provide government-funded medical services. In NSW, the Centre for Developmental Disability Studies at the University of Sydney is not so funded, but has helped me to establish a medical clinic with two other doctors, funded by billing Medicare. Our six-month waiting list illustrates the level of unmet need.

Australians with disabilities —predominantly receiving pensions and with no political clout — are caught in a dysfunctional system. Formal responsibility for health and other services has largely passed from the federal to the state governments, with further buck-passing between health and social service departments within the states. Yet, much of their healthcare rests with GPs, funded by the federal government.

I was founding chair of the NSW-based Association of Doctors in Developmental Disability (ADIDD), which lobbied for people with disabilities and their need for specialised health services. As it became obvious that this was a national problem, we have now formed the Australian Association of Developmental Disability Medicine (AADDM), which aims to establish national standards in healthcare delivery, lobby federal politicians, issue policy documents and position statements, and, eventually, train specialists in this field.

I am lucky to have been present at the beginning of a new branch of medicine. I hope that advances in genetics will eventually lead to a therapeutic revolution for people born with intellectual disability. Meanwhile, we need more medical champions for this vulnerable but silent minority.

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