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ISSN 0041-6193

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THE ULSTER MEDICAL JOURNAL

Published by
THE
ULSTER MEDICAL SOCIETY

The Ulster Medical Journal, Volume 70, No. 1
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The Ulster Medical Journal

The Journal of the Ulster Medical Society. First published in 1932.
Successor to the Transactions of the Ulster Medical Society (1884-1929), and the Transactions of the Belfast Clinical and Pathological Society (1854-1862)

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The Ulster Medical Journal, Volume 70, No. 1, pp. 1-2, May 2001.

Editorial

Future direction of medical genetics

The genetic revolution began in 1953 when Crick and Watson proposed the double-helix structure of DNA. On 26th June 2000, the first draft of the 3.2 billion bases of DNA (the human genome) was made public. The completion of the sequencing of human DNA, undoubtedly the greatest biological achievement will have major implications for the future direction of medical genetics and medicine. The practical value of the completed DNA sequence will be to provide more rapid approaches in the search for disease-causing genes which will lead to the unravelling the molecular basis of disease. It will also provide techniques for accurate and improved diagnosis and for the presymptomatic screening of 'at risk' individuals within families and populations. An understanding of the genetic factors involved in multifactorial conditions, such as ischaemic heart disease and diabetes, will result in the development of personalized prevention, tailored drug treatment and eventually 'cure.'

The clinical geneticist's primary function will continue to be the provision of diagnosis and counselling of families with genetic and partly genetic disorders. They will have the facilities to make accurate diagnoses and to screen family relatives more efficiently for the disease causing gene mutations. It will be possible to make accurate assessment of risk and to provide more detailed information to enable patients and their relatives to make informed decisions. As many clinical disciplines embrace molecular medicine, the clinical geneticist will be a member in multi-discipline teams managing patients with single gene defects and complex disorders Clinicians will have direct access to analysis of gene expression using DNA micro-array technology. This will enable the simultaneous analysis of a thousands of unique DNA fragments, each fragment able to detect mRNA expressed from its corresponding gene. Micro-arrays have huge potential for many different fields in medicine, particularly in gene expression pattern recognition which characterizes disease states.[1] Gene expression profiling will not only be useful for haematological diseases and cancers, but also in unravelling the molecular aetiology of learning disability, skeletal dysplasias and 'dysmorphic' syndromes. Chromosomal analysis (cytogenetics) remains the main diagnostic test for learning disability, genetic syndromes and for reproductive loss and infertility. Currently, chromosomal abnormalities are diagnosed by conventional G- or R-banding methods but new techniques based on DNA hybridisation (FISH), spectral karyotyping (SKY) and spectral colour banding (SCAN) will enhance the ability of clinical cytogenetic laboratories to detect subtle genomic changes.[2] The development of whole genome matrix-comparative genomic hybridisation is likely to achieve a resolution equivalent to that of standard of G-banding. A whole genome micro-array will be ideal for screening for microdeletions and microduplications and will greatly assist in the identification of cryptic telomeric aberrations which have a role in unexplained learning disability and dysmorphic syndromes. It will also be a valuable asset in the study of chromosomal aberrations in cancer and will have a major impact on the understanding of disease, disease progression and outcome and also on responses to drug and radiotherapy treatments.

Clinical geneticists and obstetricians are involved in assessing the health of the human fetus. A major challenge is to develop methods which do not compromise the fetus. Currently, prenatal diagnosis of chromosomal abnormalities such as Down syndrome and gene disorders such as Duchenne muscular dystrophy can only be accurately achieved by chorion villus biopsy and amniocentesis which are associated with a 1-5% fetal loss. In the future, clinicians will be able to screen the fetus for chromosomal abnormalities and gene disorders on maternal blood at 10 weeks gestation. The recent discovery of acellular fetal DNA in plasma from pregnant women offers an alternative to invasive procedures.[3] Many pregnant women who are 'at risk' of having an infant with a single gene disorder or a congenital abnormality are reluctant to undergo prenatal diagnosis because abortion is unacceptable. Another approach for such women will be

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preimplantation genetic diagnosis(PGD) which will avoid the birth of affected children.[4] Several embryos are produced by IVF and cultured for 3 days to reach a 6-10 cell stage. One or two cells are removed from each embryo and analysed for the specific genetic disorder. Unaffected embryos are transferred to the mother's uterus and thus the resulting baby must be free of the genetic disorder. Currently, PGD is available only for chromosomal trisomies and a few gene disorders such as cystic fibrosis and Duchenne muscular dystrophy. The goal in medicine is to 'cure' disease. Understanding the molecular basis of genetic disorders offers the possibility of correction of the gene defect. Gene therapy is only 10 years old but despite set-backs and obstacles, evidence now is emerging of beneficial outcomes. Trials of gene therapy for severe combined immune deficiency(SCID) and haemophilia have demonstrated that it is possible to restore and maintain normal gene function. Currently gene therapy involves the delivery of the wild type (normal) gene into the affected cells but within the next decade more efficient approaches will be available such as gene repair, engineered exon skipping and 'in utero' gene therapy.[5]

The advances in human genetics raise many legal, ethical and social issues. Clinical geneticists will become increasingly involved in public debate about the application of genetic knowledge not only to families but also to society. Despite the potential for misuse of genetic knowledge, there are enormous benefits of the 'new genetics' to tomorrow's clinical geneticists and physicians. When questioned about the implications of the sequencing of the human genome, J D Watson replied, "Use it . . . to understand more about disease, prevent genetic diseases coming into existence and possibly finding a way to cure them." Clinical genetics has an promising future.

N. C. Nevin

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The Ulster Medical Journal, Volume 70, No. 1, pp. 3-4, May 2001.

Dr Robert William Magill Strain (1907-2000)

Dr Robert William Magill Strain was born on 12 April 1907 at 3 University Square, Belfast. His father was David Strain, manager of the Ulster Bank in Donegall Place, and his mother was Helen Louise Magill in 1906. His mother's sister Florence had married Dr Robert James Johnstone, later Sir R J Johnstone, Professor of Gynaecology at Queen's University and University MP in the Stormont Parliament. R J Johnstone had a considerable influence on his nephew and there was certainly some similarity in their early careers.

Bill Strain, as he was always known, went to school at the RBAI (Inst) and then on to study medicine at Queen's University. He took time out in the middle of the course to work for a BSc degree in physiology and biochemistry (2nd class honours) in 1927, obtaining the Musgrave Prize in Pathology in 1928 and graduated MB, BCh, BAO, with 2nd Class honours in 1930. As a student he also found time to be a committed member of the Student Union Society, Literific Society, BMSA (also Hon. Secretary and Vice-President) and Students' Representative Council.

For his first year after qualifying he was house physician and house surgeon at the Royal Victoria Hospital and during 1931-3 returned to the department of physiology at Queen's to work with Professor T H Milroy. He has recorded how the group of young doctors in between supervising the practical classes for the medical students, were all engaged in research on the nucleotides of muscle, work which led elsewhere to the discovery of DNA. He obtained the MD (with Gold Medal) for his thesis on the "Acid-Soluble Phosphorus Compounds of Mammalian Cardiac Muscle" in 1933. He then spent a period as postgraduate student at the Rotunda Hospital, Dublin, house officer at the Princess Louise Kensington Hospital for Children and medical registrar at the Royal Victoria Hospital. This pre-war period also saw the start of an interest in the Order of St John and the St John Ambulance Brigade, which he maintained throughout his working life. At the end of this broad training, he was appointed clinical assistant in medicine at the Ulster Hospital for Children and Women 1935-38 and honorary assistant physician 1938-44.

After the outbreak of World War II he joined the RAMC (October 1939) and served as medical specialist or acting officer in charge of the medical divisions of various General Hospitals, with the rank of lieutenant-colonel. This took him eventually to France, Holland and Germany, before he was demobilised in November 1945. He then returned to the Ulster Hospital as honorary physician.

About the beginning of the war he met Eileen Clapham and they were married in 1947 after he returned home. It was a long and happy marriage, and she died only in 1997. They had no children.

He obtained the MRCP Ireland in 1949 while already a consultant physician (as was possible at that time) and was elected FRCP in 1953. During the post-war years in the Ulster Hospital he worked as a general physician and was a fellow or member of such organisations as the Association of Physicians of Great Britain and Ireland, the British Medical Association, the British Geriatric Society, the Royal Society of Medicine, the Ulster Medical Society (President in 1968-9), the Ulster Paediatric Society and the Corrigan Club, serving

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on a variety of subcommittees of many of them. He was also appointed visiting physician to the Belfast Charitable Society in 1946, a post which he held until he retired, and took an active interest in the health of residents. This led to his historical work on the Charitable Society and after retirement he was president of the Society 1971 - 82. Yet another strand of his work was with Queen's University, as lecturer in medicine for dental students from 1949-71.

Bill Strain's historical studies began with an article on the Belfast Charitable Society in the Ulster Medical Journal as early as 1953. This led to the degree of Doctor of Philosophy of Queen's University in 1955 and the much larger volume "Belfast and its Charitable Society" published in 1961. This used the extensive archives of the Society to re-create the social history of Belfast over two centuries. It is in fact the wide range of the Society's activities covering care of the poor, literally from cradle to the grave, training for work management of beggars, provision of a water supply for the city, provision of medical care, and finally creation of the city's most interesting graveyard, which makes the book essential reading for all concerned with the history of Belfast.

His main involvement with the Ulster Medical Society was as member of the Council, honorary archivist and President for the year 1968-9. He wrote first a history of the Society (1967) which is an update of Richard Hunter's earlier paper and is notable for its detailed and rather moving account of the last days of the Whitla Medical Institute. His presidential address was entitled "University Square - A Sentimental Retrospect" and in 1970 he gave another paper on "The Foundations of Belfast Medicine".

On retirement which was at the height of the "troubles", Bill and his wife decided to leave Northern Ireland and they settled in Cornwall for 14 happy years. As his brother-in-law, Angus Wood, commented "There he was able to absorb the Celtic atmosphere, so reminiscent of Ireland, became an active member of the painting fraternity producing numerous paintings, many of which adorn the wall of his circle of friends. He landscaped his beautiful garden and, as he had done in Belfast, acquired an encyclopaedic knowledge of the county and still found time to do some part-time work for the Department of Social Security. When his sister-in-law and husband moved to Cheltenham in 1988 he and his wife decided to follow. Cornwall was beginning to feel rather remote and the garden was getting too much for him. On the death of his wife in 1997 he moved into a care home, although still retaining his apartment in Cheltenham. There he was well looked after and continued to paint to the end, and even last year, his ninety-second, produced his usual quota of over 50 painted Christmas cards which he sent to his friends. He even had published another book "The Silver Bracelet". As time passed he became frustrated by physical handicap, but his mind and memory still remained as sharp as ever. He still recounted amusing anecdotes from seventy odd years of student, army and working life and those stories were still exceedingly well told. His health deteriorated rapidly and he died on 27 December 1999.

Before his death Bill Strain took care to disperse his papers and memorabilia safely between the Ulster Medical Society and the Archive Office of the Royal Victoria Hospital. In his will also, he remembered his home area, particularly charities within the province and bodies involved in the welfare of animals, another love of his life. Altogether, Bill Strain is a man who should be remembered in Ulster as physician, historian and benefactor as well as for his great warmth of character.

R S J Clarke, Honory Archivist,
Royal Victoria Hospital, Belfast.

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The Ulster Medical Journal, Volume 70, No. 1, pp. 5-14, May 2001.

The History of the teaching of the specialty of General Practice in Northern Ireland

Presidential Address to the Ulster Medical Society

Dr Robin Harland MB, BCh, BAO, FRCGP, FISM

The weight of history is upon me, both physically and metaphysically. I am both very grateful and extremely humble on receipt of this magnificent chain of office, and of the responsibilities it implies. I shall make every effort to maintain the high standards set by all my predecessors. In that regard I would particularly want to congratulate Bob Stout on a very successful year. I have a second title to my paper and it is this: The Ugly Duckling – and What Became of It!

MEDICAL INDEX?

Dissection is the art of cutting up to display structure and relation to internal parts. As we come to dismember this body it will soon be only too obvious that it is a grossly obese subject. In the PowerPoint presentation[1] we worked our way through ten cross-sections. But, in this published version I have had to use even more liposuction; and I can only proffer one big Excuse Me for flying past or omitting your own favourite piece of the action in this published version.

Analysis of the Title

History.

History may be regarded as being simply a record or account of past events, much as the scientist may report on what is going on in his test-tube. But, when one is actually inside that experiment, within the bubbling reaction all around, rather than at arm's length, then the chronicler and the chronicle are going to keep changing - ever changing - as time passes. I turn to T.S. Eliot's Four Quartets – The Dry Salvages.

When the train starts, and the passengers are settled
To fruit periodicals and business letters
(And those who saw them off have left the station)
Their faces relax from grief into relief
To the sleepy rhythm of a hundred hours.

Fair forward travellers! Not escaping from the past
Into different lives, or into any future.
You are not the same people who left that station
Or who will arrive at any terminus
While the narrowing lines slide together behind you.

Certainly, I have changed into a very different person to the one who started down his medical journey in 1943; and I am being changed again by this experience. But so are we all! History personified!

Teaching

A quarter of a century ago Prof. D.A.D. Montgomery in his Presidential Address[2] to us said "Do not let us be fooled by the technologist; the true Art of Medicine is our most prized possession". We are all agreed; but how do we teach it? The Annual Will Pickles Lecture is given to The Royal College of General Practitioners. In his fine 1971 lecture entitled "The Art So Long to Learn", our own John McKnight,[3], chose Saint Patrick's famous symbol of the Holy Trinity and gave it a secular form. He demonstrated how Continuing Education, Postgraduate Education, and Undergraduate Education were as closely linked as the leaflets of a shamrock.

What is a Specialty?

In the beginning every doctor was a generalist. The words 'specialist' and 'specialism' were not used in their modern sense until 1856, but the explosion of knowledge from chloroform to vaccines made this process inevitable. It seems *de rigeur* to speak of William Osler, during each and every Presidential Address of the UMS, so I have put him in early on, because a report on a singular medical man may explain the issue more easily than general observation.

Professor William Osler, that doyen of physicians, was a Canadian who, at the age of only 25, was already the professor in charge of the medical institutes in his Alma Mater, Magill University, Montreal. He spent 20 years working in the USA - from 1884 to 1889 in Philadelphia, and then from 1889 to 1904 in Johns Hopkins in Baltimore. He only took up the Regius Chair of Medicine at Oxford in his 54th year. He was a true generalist,

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as a study of his publications reveals. His articles covered such varying topics as Blood Platelets - which he was the first to describe - (Research Scientist and Haematologist); Cerebral Palsies in Children, and Chorea and Choreiform Affections (Paediatrician and Neurologist); Abdominal Tumours (Gastro-enterologist and Oncologist); Angina Pectoris (Cardiologist); and Aequanimitas (Philosopher)[4]. . He was a fine example of the generalist specialist. Nowadays we all accept the concept of knowing more and more about less and less.

What is General Practice?

Although most patients seemed to have a fairly clear image of the service provided for them, the doctors in this group have had some difficulty in choosing a suitable label or marker to cover the essential qualities which distinguished them from their hospital-based colleagues. 'General Practice' and 'General Practitioner' seem to have been in common usage, while many Americans favoured 'Family Physician'. The term 'Primary Care Team' was a much later invention.

John Horder also wrote elegantly about the family doctor.

"The personal doctor can offer care which is accessible, of broad range, relatively continuous, and above all, integrative. No one of these characteristics is easily provided by a specialist; in combination they can only be provided by a generalist. This forms a distinctive role which people value as a basis to which specialist care can be added when necessary. In this way specialists and generalists can relate to each other, not as two people doing the same job, one better than the other, but as two people doing complementary jobs of equal status within the profession and society more widely"[5].

Foundations

In the early part of the nineteenth century most doctors were trained in apprenticeships, and their work in Ulster, especially amongst the poor, was governed by the Dispensary System[6]. There were four supporting legs in this education process: the Learned Societies, the Royal Colleges, and the Universities,(often with an overlapping personnel) accepted the planning and burden of undergraduate and postgraduate medical education - both in the whole country and locally too in our Province. The fourth important leg was supplied by the Parliament and Government at Westminster.

Evolution

At the very start of my time in General Practice in February 1950 standards were at a very low ebb. An Australian visitor, Dr. J.S.Collings, wrote a definitive study that year and his report was scathing. It hit home. He particularly criticised the two-room surgery premises, *"so often ill-furnished, and under-equipped"*. He found these all over the kingdom. *"There did not seem to be any place for adequate record keeping, nor for any ancillary staff."* His portrait was terribly bleak, but an accurate contemporary record, and it had a profound effect on a whole generation of doctors. He continued, *"General Practice is unique in other ways too. For example, it is being accepted as being something specific, without anyone knowing what it really is. Neither the teacher responsible for teaching future general practitioners, nor the specialist who supposedly works in continuous association with the GP, nor for that matter the GP himself, can give an adequate definition of general practice."*[7] *"My observations have led me to a condemnation of general practice in its present form; but they have also led me to recognise the importance of general practice, and the dangers of continuing to pretend that it is something which it is not"*[8]

This was a time of deep unrest in the profession. A contemporary writer described it like this: *"The position in which some doctors find themselves is more akin to slavery than to service"*[9]. The Government had been very slow to respond to the Pilkington Report[10] which had recommended £11,000,000 extra for general practitioners and £8,000,000 for hospital staff. But it wasn't until 1962 that the Review Body which the Commission had recommended in 1960 was appointed,[11] and their deliberations took a further year before the profession was offered a 14% increase in pay.[12] But problems were not just related to money. Whatever the reasons some 25% of British graduates were emigrating. Junior hospital staff was in very short supply, and many posts were being filled by graduates from abroad, most noticeably from India and Pakistan. Lord Taylor had been particularly critical when he opened the debate in the House of Lords in December 1961. Unhappy comments featured weekly in the medical press under the very descriptive title he had used "This ghastly awful mess."

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"This sense of lessened status and decline in prestige has been too general for too long, to be dismissed as of little account, or as the complaints of disgruntled minority"[13]

Unlike their hospital-based colleagues the general practitioners became very confrontational against the Government mainly through the leadership of the Local Medical Committees, and the General Medical Services Committee of the British Medical Association. In 1965 this almost led to a mass resignation from the NHS.

Curwen wrote brilliantly as he illustrated the dichotomies. *"On the 17th January 1958, Lord Moran of Manton, who was at that time the Chairman of the Awards Committee administering awards for consultants in the National Health Service, was giving evidence before the Royal Commission on Doctors' and Dentists' Remuneration". "He was defending the principle of Merit Awards against a certain amount of criticism by members of the Commission, and he made the point that those selected for the awards were chosen from a group of doctors, the consultants, who had already distinguished themselves from the rest of the profession by achieving that status". He described the process by which they did so, and mentioned 'a ladder which people are constantly falling off'. The Chairman asked him the following question - "It has been put to us by a good many people that the two branches of the profession, general practice and consultancy, are not senior or junior to each other, but level. Do you agree with that?" To this he replied as follows "I say emphatically 'No'. Could anything be more absurd? I was Dean of St. Mary's Medical School for 25 years..., and all the people with outstanding merit, with few exceptions, aimed to get on the Staff. There was no other aim, and it was a ladder off which some of them fell. How can you say that the people who get to the top of the ladder are the same as the people who fell off it? It seems to me so ludicrous!" In reply to further questions the noble lord made evident his distaste at having to discuss such contentious matters in public, but he stuck to his guns, and maintained that 'his ladder' was real enough, although that this did not imply that general practitioners did not include amongst their number men of ability doing splendid work in their own fields"*[14]

There was one new line-up of doctors, from 1952 onwards, which had a profound effect for the better in the education and training of general practitioners. This was the College of General Practitioners founded by a Steering Committee on 19th November 1952.[15][16] Its progress was rapid, and in 1967 the royal charter was conferred by Her Majesty the Queen, allowing the name to change to the Royal College of General Practitioners. Like its precursor of over 100 years earlier this organisation too met with a great deal of initial opposition. But on this occasion the planners overcame internal dissentience, external resistance, and most of all the natural opposition of the multitude of independent minds of the many potential members. It can be no surprise that even today the policies of the Royal College of General Practitioners are not universally popular.

One of its strengths was the early decentralisation of the organisation, with many regional faculties throughout the United Kingdom and Ireland. The Northern Ireland Faculty was founded on 30th. April 1953, in the Whitla Institute, Belfast, under the chairmanship of Dr. J. Campbell Young.[17] Dr. Young had been a member of the original steering committee and a founder member of the College.

The College moved very quickly on the educational issues. Curwen's prophetic words in 1964 spell out this progress, although the precepts must have seemed outlandish to many people at that time. He wrote about general practitioner training in these words:

"His apprenticeship must be as long and as demanding as the present-day consultant, and at the end of it he must pass an examination as least as difficult as that now required for entry into a 'minor' specialty."[18]

Beginnings

As early as 1931 there are records of organised postgraduate education courses for general practitioners in Northern Ireland. Originally this series was offered by the consultant staff of the Royal Victoria Hospital, Belfast. The proposal for such a course came from Professor Andrew Fullerton, the Professor of Surgery at Queen's University, and the Chairman of the Medical Staff at the Royal Victoria Hospital Belfast. Dr. Sydney Allison had been appointed to the visiting staff the previous year, and he was put in charge of the arrangements. In 1970 Dr. Allison was to write of his experiences to Dr. John McKnight.

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"Although a novelty in Belfast, and in many other centres in 1931, the only established post-graduate teaching centre in the United Kingdom was at the West London Hospital, Hammersmith, where a school had been in existence for many years, with a library, reading rooms, lecture theatre, and other amenities for the twenty to thirty doctors usually in attendance, and most of them drawn from the army, navy, and colonial medical service".[19]

I just love the style of writing from a by-gone age. The proposal for such a course was not universally popular. Allison went on to say: *"However, it is interesting to recall that there were one or two dissentients who believed that the dissemination of professionalised knowledge and techniques could harm the position of the specialists, in giving implicit encouragement to practitioners to undertake work themselves which was primarily the province of the specialist. These criticisms were voiced, but not taken seriously, most of the staff recognising that nothing but good could come from the new move".*[20] What Dr. Allison reported was that one ENT surgeon had exclaimed *"If we teach the general practitioners too much, they will take out tonsils and adenoids, and we will soon all be in the Work House!"*

This first course[21] was attended by practitioners from as far away as Bushmills and Cookstown and Keady, as well as those from nearer at hand[22]. It was held each Wednesday afternoon at 4 p.m. from January until June, and again in October and November of that year. 27 different members of the hospital staff gave one talk each. The fee of 2 guineas was payable in advance. 29 doctors attended the first session, including two women doctors. It is recorded that this course was given a very enthusiastic reception, and that practitioners were invited to enrol again for the following year.

By 1938 these courses were on a much more formal basis. Government officials from the Ministry of Labour were pressing the Faculty of Medicine and the staff of the Royal Victoria Hospital to provide two courses annually. There was an obvious hiatus through the Second World War 1939 - 1945 when many of the hospital staff and general practitioners had volunteered to serve in the RAMC. As soon as the war ended the entire energies of the medical profession were dedicated to the creation of the National Health Service on 1st. July 1948.

Turbulent Times

The National Health Service was ten years old when Lord Moran talked about his ladder After this decade of NHS operation the medical profession had appointed its own Medical Services Review Committee; and this became known as the Porritt Committee - and it eventually reported in 1962.[23]. Paragraph 211 of this report is significant: *"The Royal Colleges and the Universities have an important role to play in postgraduate preparation, but the Government has an equally important part, as virtually the monopoly employer of doctors. We recommend that the Royal Colleges and the Universities, and the Ministry of Health should combine to organise postgraduate training throughout country".*

As befits such an ancient and august institution, the Faculty of Medicine of the Queen's University of Belfast counts its meetings by numbers as well as by dates. At its 420th meeting on 20 November 1962, the Faculty responded to a request from Dr. Frank Main, then Chief Medical Officer of the Ministry of Health and Social Services in Northern Ireland. It was his wish to consider the immediate problem of continuing education for general practitioners. Over the next two years the Civil Servants at Stormont pressed for an organised continual education programme for GPs. Professor John Henry Biggart, the Dean of the Faculty of Medicine[24] at Queen's, saw a different end-point, for he wished to include the needs of the specialist registrars as well.

By the 425th meeting of Faculty on 25 June 1963 Professor Biggart was able to report that the Ministry had found adequate funds to allow the appointment by the University of clinical tutors, at an honorarium not exceeding £150 per annum each, and also of paying clinical lecturers at a rate of four guineas per half-day session. There was now a commitment by the Ministry of Health to provide a sum of £55,000 (£11,000 for five years) for the development of postgraduate medical education in Northern Ireland.[25] This was money which could or should have been distributed to all the GPs in Northern Ireland, as the Pool Balancing Cheque. But the GMSC(NI), under the astute chairmanship of Dr. Dan Chapman, agreed to use it instead as a fund for continuing education.

The End of the Beginning

The staff at the Ministry of Health and Local Government had prepared a draft paper on

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'Training for General Medical Practice'[26]. There was much deliberation on schemes already operating in Inverness and Wessex. An important decision was taken in principle, when it was agreed, ***"That there should be a compulsory period of training prior to entry to general practice, and the Board's list".***

The Civil Servants were quite clear of the implications. There would have to be extra funding found by the Exchequer, and this would require further inter-departmental discussion. But it is quite remarkable that this Government statement was written in November 1963, and that it took nearly 17 years to implement!

By 1964 Continuing Education was well established. Vocational Training was accepted as inevitable and Undergraduate Education was already at the embryonic stage. The die was cast. Professor Biggart was able to report that a **Board of Postgraduate Education** had now been accepted by the Academic Council and Senate of Queen's.[27] It was agreed that the Board would have both planning and advisory functions, and that it would cover the needs of other medical specialties, as well as general practice. On 21 April 1964 its membership was reported[28]

The Postgraduate Medical Education Board.

The first meeting of the Postgraduate Medical Education Board was on 1 June 1964.[29] A decision had been made to appoint a Postgraduate Dean. It has been suggested that John Henry Biggart was reluctant to share his own title of Dean with anyone else, even though this was the accepted mode of address in Britain. He favoured the more dictatorial 'Director'. But there continued to be great uncertainty as to the exact type of person who would fill this new post. Professor Biggart again stressed 'the importance of finding someone with fire in his belly'.

It was after another longish period until the Dean reported to Faculty at its 441st meeting on 29 June 1965 *"that Dr. John E. McKnight was to be appointed the Director of the Postgraduate Medical School, and would take up duties at the beginning of October".*[30]

So with the Director in post at last, things started to move again; and Dr. John McKnight's task must have been daunting. Expansion and development were the watchwords from 1965 to 1970. Demands seemed to crowd in from all sides.

For the general practitioners he was to double the continuing education courses for established practitioners, which had to be Province wide, and not just limited to Belfast. At the same time he had to start up a 'Training for General Medical Practice Scheme' for a new breed of trainees[31], and trainers. Again, all the other specialties were demanding his best effort on their behalf.. Yet again, this was a period when great efforts were being made to attract married women doctors back into practice; and Dr. McKnight had to cater for their special needs too.

The Training for General Medical Practice Scheme.

The staff in the Ministry of Health and Social Services at Stormont had been pressed further by the thinkers in the College of General Practitioners to produce a scheme for postgraduate training during the early sixties. Long before the Todd Report[32] on Medical Education had been published in 1968, these problems had been tackled in very many different ways in various places in England, Wales, and Scotland.

In August 1959 the first trainee of the Wessex Scheme was in post. It was based in the hospitals in Southampton, Portsmouth and Winchester. In 1962 combined hospital and GP schemes were opened in Canterbury and Durham. I was the first Trainer in that scheme in Durham.[33] By 1964 there were further similar developments in Lancaster and Birmingham.

One of John McKnight's early tasks was to organise such a training scheme in Northern Ireland, and his Board appears to have modelled its plans on the one in Wessex. He acted as quickly as he could. At the 445th meeting of Faculty on 25 January 1966 the Dean reported the appointment of 12 trainers under the General Medical Practice Trainers Scheme. They had been carefully chosen from some 80 applicants.[34] The initial criteria for appointment look very simplistic by today's sophisticated standards.[35] Even this task had been far from easy, and there were several disgruntled practitioners who were very aggrieved at non-selection, and did not hesitate to say so.

As it turned out, this unhappiness was misplaced, because there was a complete absence of trainees. At the outset there had been two doctors accepted; but both withdrew; one was given 'an Assistantship with a view to partnership' and the

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other decided to stay in hospital medicine. The first trainee started the course in August 1966. He was Dr. Myles Shortall, who had graduated from Queens in 1964, and who is now the senior partner in a group practice in Newry, Co. Down. The first trainer was the late Dr. G.W.C. ('Garry') McCartney of Lisburn. Dr. Shortall was the only trainee to complete the course in 1968, and there was not a single completion in 1969. There was a simple explanation for the acute shortage of applicants. A memorandum from the Ministry of Health and Social Services[36] explained the problem in this way: *"Clearly, however, the main weakness of the present scheme is* its *lack of financial incentives in a period of acute shortage of doctors"*.

As well as offering the possibility of financial incentives, this Memorandum reaffirmed their previous decision, and recommended that consideration be given to making the vocational scheme compulsory instead of it being voluntary.

A second group of twelve practitioners[37], was selected for trainer posts in June 1969. It is clear that there had been an upgrading of standards when one studies the new criteria[38], which show clear evidence of the influence of the Royal College of General Practitioners, and its Education Committee, on which John McKnight had been serving with such distinction. Even so, it is worth pointing out that those originally chosen were all interviewed again, and all were re-selected. These extended duties had to be delegated to a special General Practice Sub-Committee whose membership was first minuted by the Board on 4 February 1969.[39] Four more 'teaching practices with a designated 'teacher-in-charge' were appointed by this new committee in May 1970.[40]

On 29 April 1968 the sub-committee met and selected six of the applicants[41] as trainees; and in April 1969 they appointed 18 more trainees, some of whom are now very well known in Ulster's medical services.[42] These were boosted by six more in November 1969[43]. The Vocational Training Scheme had become accepted at last. Between 1 February 1966 and 1 February 1970, 43 doctors were accepted on the Scheme. Of these, 3 had completed their training, 5 did not enter the scheme, 9 withdrew for various reasons between 4 weeks and 18 months; and 26 were still in training, when this data was presented to a meeting of The Board of Postgraduate Medical Education on 5 May 1970.

The Postgraduate Medical Education Board of Queen's University had fulfilled its purpose, and it was time to think afresh. Some of the recommendations of the Todd Report were to be implemented, after modification. Plans were put forward by Dr. F.D. Beddard, the new Chief Medical Officer at the Ministry of Health at Stormont, to create a new Council in Ulster. On 4 December 1970 the Northern Ireland Council for Postgraduate Medical Education was born. The following important piece of the puzzle will have to be told on another occasion.

Undergraduate GP Teaching

It was within the heady atmosphere of university expansion during the Sixties that the Department of General Practice was born. At the end of 1968 Queen's University accepted a recommendation that a Chair of General Practice be created. This was made possible by the remarkably generous decision of the general practitioners of Northern Ireland to recommend that the Practice Improvement Fund be diverted from their own pockets to this worthy purpose. This was a remarkable gift of £59,653. A decision was taken to advertise this Chair late in 1969. Dr. William George Irwin was appointed on 1st October 1971.[44] He was a Principal in the National Health Service structure in south Belfast, and became the Head of an autonomous Department in the Faculty of Medicine. The department was to be based in a newly built teaching health centre on the corner of Dunluce Avenue and Lisburn Road. Four other British Universities had already entered this new field of medical education[45].

It took a full nine years of effort before the Dunluce Teaching Health Centre was opened close to the Belfast City Hospital.[46] On the ground floor there was a pharmacy and a family planning unit. The second and third floors were home to four different partnerships and one single-handed general practitioner; giving a total of over 24,000 patients catered for in twelve consulting suites. Each consulting room had a one-way mirror, so that real consultations could be observed from the viewing room next door by medical students, some two or three at a time. Informed Consent had to be obtained before any such viewing took place. In addition each consulting room was equipped with a video-camera so that everything which took place could be recorded for use in small-group discussion afterwards. In addition each consulting room was linked to the seminar

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rooms on the fourth floor, allowing consultations to be discussed with larger groups. The top, fourth floor, housed the University Department with a library, teaching space, and offices for clinical academic staff, non-clinical academics, research fellows, and secretarial staff.

There were two possible alternative organisational arrangements, well described in the MacKenzie report[47]. Although on the face of it the Practice Based Department might have theoretical advantages in providing a solid research base, time showed some serious disadvantages. The full-time University lecturers were swamped by the ever-increasing demands of patient care, and academic productivity was low. Professor Irwin chose the Practice Linked Model, and was the very first in the UK to do so. With hindsight it can be seen that this was the correct decision because academic productivity in both teaching and research were of a much higher standard.[48]

Every academic has a trilogy of duties – Teaching, and Research and Administration. Sadly, only the first of these duties can be discussed in this paper. The needs of academic general practice were spelled out by George Irwin in his Inaugural Lecture.[49] In this he pointed out that there is a considerable difference between, on the one hand, Disease with all its pathology, and on the other Illness with all "its behavioural aspects and social consequences".[50] Professor Irwin saw academic general practice as largely an applied clinical discipline of primary care, with health promotion and preventive medicine as part of the mix. From its earliest days the department had to develop, by process, a core content of knowledge, skills, and attitudes.

Teaching and Assessment

It was clear that small-group teaching should be used whenever possible, and this meant the recruitment of many new general practitioner teachers from a variety of practices. These were loosely divided into 12 core practices, and around 100 'outer ring teachers'. Teaching took place in the seminar rooms in the department, in the consulting rooms, and during home visits. Each student entering the Department in the clinical years was given a Handbook which listed the learning objectives of general practice, and the methods of assessment in class examinations and the Final MB Examination.

Long before Dunluce Health Centre was ready for occupation Professor Irwin had to formulate a philosophy of education, defining learning objectives and methods of assessment. These objectives cannot be fully achieved in the teaching hospital where specialisms have fragmented care and where illness\disease is usually seen at quite a late stage. As a result less weight is given to social and psychological factors. From the early 1970's the Department made a substantial weekly contribution to small group teaching in the Pre-Clinical Behavioural Science Course; but the main impact of the teaching was in the clinical phase of the curriculum.

Just at the time of Professor Irwin's appointment in 1971 the Medical Faculty had completely remodelled its teaching into an integrated teaching course. At first the departments of General Practice, Geriatrics, and Mental Health joined together, and were later joined by Community Medicine. This avoided the problems of vain repetition while combining the very different skills of the specialists involved. The topics included communication, terminal care, bereavement, alcohol problems, maternal and child care, the confused elderly patient, disability, human sexuality, coronary care and screening.[51] The department's teaching skills have been developed over the years, and members of staff are involved in all 5 years of the undergraduate course. One interesting innovation is the introduction of first year students to individual families so that they may learn how families and individuals perceive, understand, and manage their own health[52].

Teaching Communication Skills

With the advent of the new Teaching Health Centre came the dawning of a whole new teaching programme exploiting the relatively new science of Closed Circuit Television (CCTV). The rising generations have come to accept the intrusion of a television camera recording every clue – verbal and non-verbal – of the consultation. Indeed the practice has become so well established that the normal way of assessment of the consulting skills module of the MRCGP examination is the submission of 15 video recordings of a variety of different types of consultation by the candidate. The students at Queen's acquired first hand knowledge of the illnesses seen in primary care and of the diagnostic and management skills needed by GPs to differentiate trivial illnesses from more serious pathology either physical or mental or both.[53]

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Involvement in Undergraduate Examinations.

Having created new aims and new general practice courses for undergraduates in their penultimate and final years Professor Irwin and his team turned their attention to assessment of learning. Communication skills were evaluated by direct observation of a student consulting with a real patient in the surgery using the one-way mirror, and scoring for each attribute. Each student was given 20 minutes to take a history and perform a relevant physical examination. This method became so successful and widely acclaimed in the Faculty of Medicine that it became part of the integrated Final MB Examination in 1981. Some 16 students were randomly allocated on the day of the clinical examination to Dunluce Health Centre. There they did their major cases in General Practice, interviewing, examining and diagnosing patients with a mixture of physical and emotional disorders, and watched for 20 minutes by the examiners, who then interviewed each candidate about the diagnosis and management of their case. One External Examiner, a London Professor of Surgery, was so impressed by this system that he arranged for an ITV crew to come over from London and record similar proceedings; this was shown on National Television. It is clear that the allocation of major cases in the Final MB Examination was a real achievement for the Department of General Practice.

In 1973 the final MB examination had been integrated (always with the exception of midwifery and gynaecology). The clinical and oral examination remained, although the examiners came from much more diverse areas, but it was in the written papers that much bigger changes occurred. Instead of separate essay papers in Medicine and Surgery the new examination had a Multiple Choice Question Paper (MCQ), an Essay Paper, and a Modified Essay Question Paper (MEQ). The Department of General Practice was chosen to assess the written work of students using the Modified Essay Question (MEQ) which had been developed by the Royal College of General Practitioners for its own entrance examination. It is reliable and valid examination tool[54] Each short paragraph of narrative is followed by a couple of questions, and the picture of a family or families in crisis gradually builds up. The MEQ aims to evaluate three areas of cognitive ability – recall and recognition of specific information; interpretation of data; and problem solving. It was natural that this paper should be set, invigilated, and marked by the Department of General Practice. Within a very short period of time the setting and marking of the MEQ was formalised. Some fourteen doctors would gather around a table, and each examiner would mark the same section over and over until all 150 papers had been circulated. Ideal answers were provided[55]. This guaranteed fairness and objectivity, with an annual pass rate of between 92 – 95%. The whole process was reviewed with care.[56]

MILESTONES

Steps along the way for Postgraduate Education

We have to bypass the important story of the QUANGOs of the General Medical Council and the Northern Ireland Postgtraduate Medical and Dental Education Council. But much of this part of the history of the teaching of General Practice in Northern Ireland has already been written and spoken about in the Presidential Address to the UMS by Dr. A.G. ("Lexie") McKnight 1988[57], and by Dr. Ben Moran.[58]

So the dream was realised after some 30 years of toil. All trainees, now known as General Practice Registrars, have to face a three year training programme and an entry examination into the specialty. This is called the Summative Assessment[59], and may only be entered after completion of training. Since 1997 this goal has been made compulsory. Without reaching this minimum standard one cannot become a Principal in General Practice. The Membership examination of the Royal College of General Practitioners has different goals[60], but in Northern Ireland all the GP Registrars are encouraged to sit both examinations.

Much of the training is given individually and can be said to resemble the old apprenticeship system. But added to this is an intensive planned scheme provided by the Course Organiser[61] The very fact that a book of over three hundred pages has been written on this single topic of organised teaching for GP Registrars underlines just how highly developed the single task has become. That there is already a second edition after only five years confirms the ever-changing scene. That the author, although writing for a national and international readership, is an Ulsterman is also highly significant. Patrick McEvoy is a highly respected GP in the City of Londonderry.

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The Art so Long to Learn was where we started. The Ugly Duckling has become a very fine swan indeed. But is it paddling in the right direction? For example, we all know that the modern medical students are very high-powered in the intelligence field and very well taught; but some fear that they are under-powered in the empathy stakes. Don't let this happen! Please! Let us answer that question – from now on – by focusing on the Patient!

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19. McKnight J. Private papers.
20. McKnight J. ibid.
21. The original notice for these meetings reads like a Who Was Who of Ulster medicine. It reads - "The lectures will begin in January 1931. They will be given on Wednesdays at 4 pm at the hospital, by members of the Honorary Visiting Staff. They will continue each week until June. After the summer vacation, a second series will begin in the autumn. Medical men desirous of taking the course should apply to the Honorary Secretary of the Medical Staff at the hospital. The fee for attendance from January to June 1931, is £2 2s 0d., payable in advance. The programme which has been arranged is subject to alteration, but so far as possible the enclosed programme will be followed".

    Sir Thomas Houston 'Vaccine Therapy and Bacteriological Diagnosis': Mr. T.S. Kirk. 'The Treatment of Fractures': Dr. J. C. Rankin. 'Modern Methods of Treatment in Venereal Diseases': Prof. W.W.D. Thomson 'Modern Conceptions of Renal Disease': Prof. Andrew Fullerton. 'The Surgery of the Hand - illustrated by cineomatograph demonstration': Prof. R.J. Johnstone 'The results of Treatment of Cancer of the Cervix Uteri': Dr. Foster Coates. 'The Medical Treatment of Gastric and Duodenal Ulcer': Mr. Howard Stevenson. 'Surgical Affections of the Gall Bladder and their Treatment': Mr. James Craig. 'The Diagnosis of some common Eye Diseases, with demonstrations': Dr. Boyd Campbell. 'Coronary Thrombosis': Mr. S.T.Irwin. 'The Diagnosis and Treatment of Hip Joint Disease': Prof. C.G. Lowry. 'The Importance of Uterine Haemorrhage as a Symptom': Dr. V.G.L. Fielden. Recent Advances in Methods of inducing Anaesthesia': Dr. Robert Marshall. 'The Diagnosis and Treatment of Cerebral Tumour': Mr.P.T. Crymble. 'The Surgical Aspect of Gastric Ulcer': Mr. Henry Hanna. 'The Diagnosis and Treatment of Squint': Dr. S.I. Turkington. 'Pulmonary Tuberculosis': Mr. R.J. McConnell. 'The Acute Abdomen': Dr. R. Maitland Beath. 'X-rays in the Diagnosis of Alimentary Disease, Demonstration of films': Mr. H.L. Hardy Greer 'Birth Injuries and their Sequelae': Dr. J. A. Smyth. 'Diabetes': Mr. H.P. Malcolm. 'The Treatment of Pott's Fracture': Mr. Fred Jefferson. Superficial Diseases and Injuries to the Eye': Dr. J.T. Lewis. 'The Diagnosis and Treatment of Pernicious Anaemia': Mr. J.R.B. Purce. 'Oesophageal Obstruction': Dr. F.P. Montgomery. 'Radium in the Treatment of Cancer': Dr. R.S. Allison. 'Clinical Examination of the Nervous System. Demonstration of cases'.
22. The GPs who enrolled are listed as follows:-
    Dr. S.M. Magowan, Glenarm, Co. Antrim
    Dr. Wm. Hunter, Camcairn, Crumlin, Co. Antrim.
    Dr. D. Huey, Ballaghmore, Bushmills, Co. Antrim.
    Dr. C.L. Gaussen, Hollydene, Holywood, Co. Down.
    Dr. W.R,Hayden, 100 Cliftonville Rd. Belfast.
    Dr. J. Armstrong, Audley Lodge, Ballymena, Co. Antrm.
    Dr. Wallace, Lavin House, Knockahollet, Co. Antrim.
    Dr.J.C.Gilbert, 36, Hamilton Road, Bangor, Co. Down.
    Dr. A. McComiskey, Downpatrick, Co.Down.
    Dr. Annie Watson, 126, Bloomfield Road, Belfast.
    Dr. J. Matson, Mansefield, Limavady, Co. Derry.
    Dr. J. Rodgers, Ashley House, 174, Albertbridge Rd., Belfast.
    Dr. W.R.Davison, Broughshane Rd., Ballymena, Co. Antrim.
    Dr. J. C. Robb, County Infirmary, Downpatrick, Co. Down.
    Dr. S.J. Killen, High Street, Carrickfergus, Co. Antrim.
    Dr. J. McMaster, Broughshane, Co. Antrim.
    Dr. W.S. Boyd, J.P., Roden House, Hillsborough, Co. Down.
    Dr. T. Kennedy, 10, University Square, Belfast.
    Dr. Isabel L. Glasgow, Corbally, Bishopscourt, Downpatrick.
    Dr. J. S. Erwin, 68, Woodstock Road, Belfast.
    Dr. W. Dickey, 86, Antrim Road, Belfast.
    Dr. E.O. Blake, 28, University Road, Belfast.
    Dr. S. Acheson, 207 Lorne Terrace, Albertbridge Rd. Belfast.
    Dr. S.E.A. Acheson - same address.
    Dr. J. Cowan Adams, 186, Ravenhill Road, Belfast.
    Dr. H.J. Ritchie, 219 Springfield Road, Belfast.
    Dr. J. Cavanagh, Hy Niall, 75 Falls Road, Belfast.
    Dr. S.R. Hunter, Dunmurry, Belfast.
    Dr. Kirk Forsythe, 32, Ranfurly Avenue, Bangor, Co. Down.
    Dr. C.O.S. Blyth Brooke, Danesmere, Rosetta Avenue, Belfast.
    Dr. G. Hamilton, Solarium, Balmoral Avenue, Belfast.
    Dr. T.H. Houston, Cullybackey, Co. Antrim.
    Drs H.H.G. Dornan of Keady, Co, Armagh; and Dr. A.G.P. Alexander also appear to have attended.
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24. Weaver J. "John Henry Biggart 1905 – 1979; a Portrait in Respect and Affection". UMJ. April 1985. He was a colossus in medicine in Northern Ireland and far beyond.
25. Confirmed in a letter from the Ministry of Health and Local Government, reported to 426th Faculty of Medicine meeting on 1 October 1963.
26. Those present were Mr. Norman Dugdale (Chairman), Dr. Kidd, Mr. Duncan, Mr. Kells and Miss Kerr of the Ministry of Health and Local Government; Mr. Stewart, Dr. Hunter & Dr. Maybin of the General Health Services Board; Dr. Armstrong representing the Northern Ireland Hospitals Authority; and Professor John Pemberton of Queen's University.
27. In fact, this was on 11 October 1963 when the Academic Council minutes record that the Faculty be empowered to establish a Board of Graduate Medical Education, to develop a programme for the education of general practitioners and medical and surgical specialists. This Board should have representatives from other bodies interested in graduate training''.
28. "Heads of all Medical Departments; Dr. N.D. Wright and Dr. J. Hunter (Northern Ireland General Health Services Board); Dr. N. S. Dickson (British Medical Association); Dr. J. A. Smiley (Industrial Medical Officers); Dr. J. B. McKinney (Medical Officer of Health); Dr.J.N.W. Ritchie (College of General Practitioners); Dr. Cecil Kidd (Ministry of Health and Local Government); and Drs. J. S. McKelvey and K.P.D. Porter (Northern Ireland Hospitals Authority)".
29. Reported to the 433rd meeting of the Medical Faculty on 16 June 1964.
30. Confirmed at Academic Council on 2nd July 1965.
31. There was considerable argument as to their title. Should it be Trainee\Assistant, or General Practice Registrar, or just Trainee?
32. Royal Commission on Medical Education 1965 - 1968 Report. London HMSO 3569.
33. Dr. Alistair McCall, later a Principal in Dingwall, Ross-Shire was that trainee.
34. Dr. Kathleen Herron 374 Beersbridge Road, Belfast 5
    Dr. Noel Wright 137 Ormeau Road, Belfast 7
    Dr. Michael McSorley 442 Antrim Road, Belfast 15.
    Dr. W.G. Irwin 13, Finaghy Road South, Belfast 10
    Dr. Garfield W.C. McCartney 68 Bow Street, Lisburn, Co. Antrim.
    Dr Geoffrey A.C. Miller Linenhall Street, Banbridge, Co. Down.
    Dr. John McDowell Newtown Street, Strabane

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Dr. Robert E. Hadden 'Magheree', Thomas Street, Portadown.
Dr. Charles A.K Tully 2 Crevenagh Road, Omagh.
Dr. John J. Doherty 26 Church Street, Enniskillen.
Dr. Robert J. Miller, Ballynure, Co. Antrim.
Dr. Sheelagh K.M. Woods, The Mall, Armagh.

35. 1. Ten years a principal in general practice.
2. Less than 50 years old.
3. In a partnership.
4. List size with less than 2,500 in an urban, and 2,000 in a rural practice.
5. Suitable practice premises and organisation.
6. Suitable geographical spread
7. They had to practise midwifery.

36. Memorandum by Ministry of Health and Social Services on Vocational Training for General Practice in Northern Ireland - written evidence to the Royal Commission on Medical Education September 1966.

37. Dr. Cecil Burns 10, Queens Street, Ballymoney.
Dr. J.J. Cosgrove 28 Pump street, Londonderry.
Dr. P.T. Fallon 3 Aberfoyle Terrace, Londonderry.
Dr. R. Graham 463, Springfield Road, Belfast 12.
Dr. G.T.C. Hamilton 30 Great Victoria Street, Belfast 2.
Dr. T.P Herriot 202 Connsbrook avenue, Belfast 4.
Dr. L.J. Higgins Fair Hill, Magherafelt.
Dr. Tom Horner 33 Gilnahirk Road, Belfast 5.
Dr. J.N. Lewis 17, Ballygomartin Road, Belfast 13.
Dr. W.M. Loughridge 10 Governor's Place, Carrickfergus.
Dr. I.B. Moran Linenhall Street, Banbridge.
Dr. A.G. McKnight 209, Shore Road Belfast 15.

38. The emphasis for selection as a trainer now lay with teaching abilities. 1. A desire to teach by the applicant, evidenced by past and present activities in the teaching of undergraduate or postgraduate students and paramedical staff, by interest in teaching methods, and attendance at teaching courses. 2 The provision of time to teach or readiness to make time - at least one session per week.3. Attitudes towards patients, partners, colleagues, previous trainees (if any) and general practice itself. 4. The trainer's and partners' special interests should be noted, particularly research. 5. The ability to make available the necessary time for teaching by means of good organisation - in particular the use of appointment systems, employment of appropriate ancillary staff, and an efficient record system

39. 14 names are listed; only a very few were not already members of the Board. COM of the MOHSS or his representative, Profs O.L. Wade, J. Vallance-Owen, J.H. Pinkerton, J. Pemberton, and I.J. Carre. Drs. C. Burns, J. F. Harrington, W.G. Irwin, R.P. Maybin, R.J. Millar, D.T. Patton, N.D. Wright & J.E. McKnight.

40. Dr. F.C. Calvert 36 Main Street, Limavady.
Dr. J.C. Loughridge 430, Antrim Road, Belfast 15.
Dr. J.M. McKelvey Ballynahinch Road, Saintfield.
Dr. S. Moore 18 Upper Library Street, Belfast 1.

41. Dr. F.R.Elder; Dr. D.H.M. Groves; Dr. J.A. Jefferson; Dr. B. Leitch; Dr. M.R. Phillips; and Dr. J.W. Richardson.

42. Dr. P.M. Reilly; Dr. M.L.W. Crooks; Dr. B Khosravinezhad; Dr. A. Glennie; Dr. L.A. Watterson; Dr. G. Fitzpatrick; Dr. A. (D.J.) Houston; Dr. W.F. Foster; Dr. D.S. White; Dr. J.R.B. Kane; Dr. B.R. Watterson; Dr. C.I. Andrew; Dr. D.E. Dorman; Dr. M.J. Boyle; Dr. J.H.S. Empey; Dr. M.G.B. McElhinney; Dr. W.D. McGimpsey; Dr. P.G.M. Kelly.

43. Dr. J.D. Boyd; Dr. R.D. Finch; Dr. D.P.W. Hoey; Dr. J.I. McCully; Dr. N.A. McKenny; and Dr. T.J. Smyth.

44. He had been an occasional lecturer for Prof. Pemberton since 1966, and named part-time Lecturere in GP from 1967

45. University of Edinburgh 1948; University of Manchester 1954; University of Newcastle-upon-Tyne 1964; University of Aberdeen 1967.

46. January 31st, 1980

47. Howie JGR, Hannay DR & Stevenson JSK. The Mackenzie Report. General Practice in Medical Schools of the UK 1986. p 7 Pub. Macdonald Printers (Edinburgh) 1986 1. The term PRACTICE BASED DEPARTMENT is used to describe a unit which has sole responsibility for running its own NHS Practice. In this model, the Principals, with their own NHS lists, are all Clinical Lecturers. The first four academic departments is general practice had all followed this model. They served either over-populated inner city areas of new housing developments.

2. The second model is the PRACTICE LINKED DEPARTMENT, where the academic clinical staff undertake part-time clinical work (of about 21 hours per week) in practices which are mainly staffed by full-time general practitioner Principals. This model allows the academic staff to organise their working lives much more efficiently. This is particularly true when all the work is housed in a single building close to a teaching hospital.

48. This was used later by many other centres in Great Britain (e.g. Leicester, Nottingham, Liverpool, and Glasgow).

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The Ulster Medical Journal, Volume 70, No. 1, pp. 15-18, May 2001.

Double contrast barium enema and colorectal carcinoma: sensitivity and potential role in screening

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Accepted 20 November 2000

SUMMARY

To establish the sensitivity of double contrast barium enema (DCBE) for detection of colorectal carcinoma in a tertiary referral centre and consider its possible role as a suitable imaging method in screening for this disease.

A total of 160 patients with a histopathologically proven diagnosis of colorectal carcinoma over a two year period were reviewed. Subsequently 112 of the 160 patients were identified as having undergone DCBE, the results of which were analysed to determine its sensitivity for detecting colorectal carcinoma.

Colorectal carcinoma was missed in 4 of the 112 barium enemas performed. This corresponds to a sensitivity of 96.5% with a false negative rate of 3.5%. The Dukes Classification in these 4 cases showed that Dukes stage B, C and D were missed, with tumours located in the right and the sigmoid colon. The mean delay to operation in these four cases was 6 weeks.

Our study correlates with previous studies showing a false negative rate for DCBE of 3.5%. Colonoscopy also fails to detect small numbers of tumours with false negative rates reported as high as 10%. We suggest that double contrast barium enema should be effective as a screening method in any future colorectal cancer screening program.

INTRODUCTION

Colorectal cancer is the second most common malignancy in the United Kingdom with 28,000 new cases each year. There are 600 new cases and 480 deaths per annum in Northern Ireland. We wished to examine the sensitivity of double contrast barium enema (DCBE) for the detection of colorectal carcinoma in a tertiary referral centre and consider its possible role as an imaging method in screening for this disease.

MATERIALS AND METHODS

All histopathologically proven cases of colorectal carcinoma in the Royal Victoria Hospital within a two-year period were reviewed. A total of 160 patients were found to have colorectal carcinoma over this 2 year period. Subsequently 112 of the 160 patients were identified as having undergone barium enema examinations. The remaining 48 patients with colorectal carcinoma were excluded from the study as they had the diagnosis established by means other than barium enema.

The examinations were performed using a standard double contrast technique, following colonic preparation with 10 mg sodium picosulphate (Picolax, Nordic) at 8 am and 6 pm on the day before the examination, and a clear fluid diet. Hyoscine bromide or glucagon was not used routinely.

RESULTS

Colorectal carcinoma was not identified in 4 of the 112 barium enemas performed (Table I). This corresponds to a sensitivity of 96.5% with a false negative rate of 3.5%. The age range of patients with unidentified tumours was 59 to 78 years, with an average age of 68 years.

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TABLE I

Undidentified colorectal carcinomas following DCBE

	Age	*Site*	*Duke's Stage*	*Delay to Operation*	*Diverticulosis Present?*	*Faecal Residue Present?*
Missed Tumour 1	73 yrs	Caecum	D	10 Days	No	Yes
Missed Tumour 2	69 yrs	Sigmoid	D	9 Weeks	Yes	Yes
Missed Tumour 3	59 yrs	Recto-sigmoid	C	9 Weeks	Yes	No
Missed Tumour 4	63 yrs	Recto-sigmoid	B	8 weeks	No	No

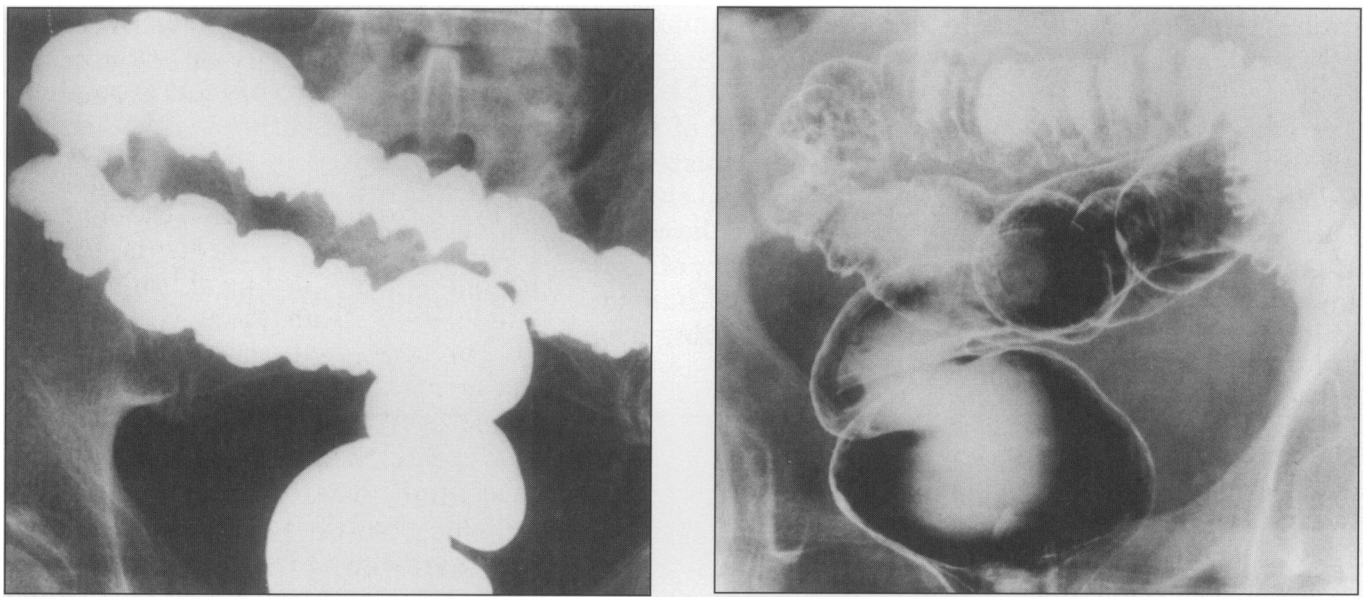

Fig 1 (a & b). Single and double contrast barium enema views of a colorectal carcinoma that was not identified in the mid-sigmoid colon.

One malignancy was missed in the caecum. In this case, marked right-sided faecal residue had been noted in the caecum and although no gross abnormality was noted on the report, a CT scan of abdomen was recommended to evaluate the region further. This malignancy was subsequently staged as a Dukes stage D carcinoma and the delay to operation following the barium enema was 10 days.

The three remaining unidentified malignancies were in the sigmoid colon or at the rectosigmoid junction. The unidentified sigmoid lesion measured 3 cm and involved the full circumference of the sigmoid colon. This patient was staged, subsequently, as a Dukes D colorectal cancer. It was visible on retrospective reanalysis of the barium enema films (Figs.1). The delay to operation was 9 weeks.

The 2 remaining unidentified lesions were polyps at the recto-sigmoid junction, one measuring 1.3 cm and the other 2.5 cm. The delay to operation was again between 8 and 9 weeks. Neither of these 2 polyps could be identified on retrospective viewing of the films.

DISCUSSION

Screening for colorectal carcinoma has not yet received widespread acceptance; however, there

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is indirect evidence that screening for this disease between the ages of 50 and 75 years could reduce the chance of developing or dying from colorectal carcinoma by 10 to 75%, depending on which screening tests are used and how often they are done.[1] The increased reduction in mortality is more likely to occur if the entire colon is visualised, with less reliance on faecal occult blood tests and flexible sigmoidoscopy. We would propose that if the entire colon is to be visualised, then DCBE should be considered as an appropriate imaging method for screening.

Colonoscopy, although having the advantage of biopsy and resection of lesions, also fails to detect a small number of colonic carcinomas, with a false negative rate reported as high as 10%.[2] Furthermore, it is not always possible to negotiate a tortuous colon and reach the caecum. There is also a small risk of perforation during colonoscopy. This has been estimated at 1 in every 1000 diagnostic procedures, with a higher rate if polypectomy is performed. The estimated perforation rate during a DCBEis 1 in 25,000 examinations.[3] Expense, a critical consideration with a large screening programme, is another factor favouring barium enema, with projected costs showing DCBE to be less than half that of colonoscopy. A disadvantage of DCBE is radiation dose, which is estimated at 7mSv.[4]

We have demonstrated that the DCBE, as performed at our institution, has a sensitivity of 96.5% and a false negative rate of 3.5% for the detection of colorectal carcinoma in a symptomatic population. Histopathology was used as the gold standard. This false negative rate compares well with previous studies reported in the literature. Johnson *et al* [5] report a false negative rate of 4.7% using DCBE and Beggs *et al* [6] a false negative rate of 8.6%. These were both retrospective studies. A prospective study by Fork *et al* [7] to assess the accuracy of DCBE in the diagnosis of colorectal carcinoma identified a 99.3% accuracy in a patient cohort followed-up for four years. We do accept that the nature of our study, as with similar retrospective studies, has an element of selection bias, due to the fact that some patients with normal barium enemas during the study period may not have undergone further evaluation and may have eventually been diagnosed at another centre.

Our study looked at the detection of colorectal carcinoma in a symptomatic population, however there is an additional issue to be addressed when considering screening in an asymptomatic population. Although the detection of early colorectal cancers is important, the detection of adenomatous polyps is also required, as it is now accepted that these are the precursors of colorectal carcinomas. Studies have shown that colonoscopy may be superior to DCBE in the detection of polyps, with DCBE having an estimated sensitivity of 75-90% for adenomas greater than 1 cm in size, as opposed to colonoscopy with an estimated sensitivity of approximately 90%.[3] Polyps less than 1 cm in size have a low risk of malignant potential (less than 1%) and therefore if proposed screening with DCBE was performed every 5 years, then there is a reasonable window for subsequent detection without detrimentally affecting patient outcome. For the polyps greater than 1 cm in size a trade off must be made between the apparent greater sensitivity of colonoscopy and the increased risks and costs associated with this procedure.

The reasons for failing to identify adenomas or early carcinomas on barium enema examination are either due to perceptive errors by the radiologist and/or due to technical inadequacies of the examination. Both these factors were present in our series with suboptimal bowel preparation being a factor in two of the cases. Other technical considerations include incomplete air distension, insufficient tube angulation and overexposed films.[8] Concomitant diverticulosis was also present in two of the cases increasing the likelihood of a perceptive error. These may also occur due to missing the lesion in the barium pool, or missing the lesion en face or in overlapping loops.[9] Radiographic findings in overlooked colorectal carcinomas include subtle changes in normal colon architecture.[8] These include concave or irregular mucosal barium margins, convergence of inter-haustral folds, flat or missing haustra, reduced caecal volume and the presence of locally fixed debris like lesions.

CONCLUSION

Both the colonoscopic and DCBE methods of visualising the colon are in worldwide usage. Both are well-validated techniques, and each has their proponents. Colonoscopy offers the advantage of direct visualisation, biopsy and excision of lesions. It has however, a recognised technical failure rate, which may not be insignificant, can be technically challenging, and has a small risk of associated morbidity.

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The double contrast barium enema is a safe, inexpensive alternative. Failure to visualise the entire colon is rare, and the DCBE has the added advantage of producing a standardised film series, which can be retrospectively scrutinised, as in our series. This advantage has implications for clinical audit, and quality assurance in a screening program.

We conclude that the DCBE, with its widespread availability, relative inexpense and high sensitivity, should merit serious consideration as an appropriate imaging method for colonic evaluation in a future colorectal screening program.

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The Ulster Medical Journal, Volume 70, No. 1, pp. 19-21, May 2001.

Ionising radiation (medical exposure) regulations (Northern Ireland) 2000 and their implications for Accident and Emergency (A&E) doctors in training

R E Bell, R E McLaughlin

Accepted 1 February 2001

SUMMARY

With the introduction of the Ionising Radiation (Medical Exposure) Regulations (Northern Ireland) 2000 (IRMER) the medical practitioner faces greater accountability when requesting radiological investigations. The referrer (usually a doctor or dentist) must supply sufficient medical data to justify radiation exposure to a patient. These regulations can lead to criminal prosecution if breached. Our objectives were to identify the level of unjustified requests for plain abdominal radiography among A&E doctors and whether there is a statistically significant difference in the justification of request between doctors of differing experience.

We reviewed and prepared statistical analysis of 100 A&E request forms for plain abdominal radiography. Royal College of Radiologist Guidelines were used as a "Gold standard" for justification of the investigation.

A&E doctors of less than six months experience are at greater risk of breaching these regulations when requesting plain abdominal films, when compared to more experienced doctors.

This is a serious issue which should be addressed at undergraduate and pre-registration level in addition to ongoing audit.

INTRODUCTION

Plain abdominal films (PAFs) in Accident and Emergency Departments (AEDs) have been shown to be of low diagnostic yield.[1] Despite Royal College of Radiologists guidelines [2] (tables I & II) PAFs are still over utilized in AEDs for a variety of condition.[1] With the introduction of the Ionising Radiation (Medical Exposure) Regulations [3] (Northern Ireland) 2000 (IRMER) the medical practitioner faces greater accountability when requesting radiological investigations. These regulations define four main duty holders: employer, practitioner, operator and referrer. A referrer is a health care professional who requests a radiological investigation or treatment. The referrer (usually a doctor or dentist) must supply sufficient medical data to justify radiation exposure to a patient. These regulations can lead to criminal prosecution if breached. Previous studies have been done on variation of PAF interpretation [4] but not on variation of justification with respect to clinical experience.

Our aims were to identify the level of unjustified requests for plain abdominal radiography among AED doctors and to determine whether there was a statistically significant difference in the justification of requests between doctors of differing experience.

METHODS

Over a six-week period, a list of PAFs requested by the AED of Belfast City Hospital was obtained from the Radiology department. The clinical information in the notes was scrutinised to determine whether a request was justified. The criteria for justification were obtained from the RCR working party booklet "Making the best use

Belfast City Hospital.

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TABLE I

Main indications for Plain Abdominal Radiography using Royal College of Radiologists guidelines.

Suspected small or large bowel obstruction.
Acute flare of inflammatory bowel disease.
Acute abdominal pain requiring admission and surgical consideration.
Sharp or toxic swallowed foreign body.
Intussusception
Urology (Belfast City Hospital local policy to use "Kidney, ureter & bladder" views for work up of urological complaints).

TABLE II

Conditions which Royal College of Radiologists guidelines specifically mention as not needing Plain Abdominal Radiography as part of initial clinical assessment.

Appendicitis
Acute pancreatitis
Abdominal mass
Swallowed coins (Indicated if coin not passed at 6 days or suspected obstruction)
Swallowed teeth
Constipation (Adult or child)

of a Department of clinical radiology". This booklet is issued to all doctors when they take up post in this particular Accident & Emergency department. Doctors were subdivided by experience as follows: Group 1 (less than 6 months full time AED work), Group 2 (greater than 6 months full time AED work).

Group 1 contained 5 full time senior house officers (SHO) with no AED experience.

Group 2 contained 12 doctors: 2 SHOs, 2 Registrars, 2 Consultants, 1 Staff Grade and 5 sessional hospital practitioners all with at least 6 months' full time AED experience.

RESULTS

Over 6 weeks 100 PAFs were ordered, representing roughly 2% total new attenders (5274 patients). 62 patients were female and 38 were male. There was a wide age range of patients x-rayed: 13 patients less than 10 years and 5 patients over 80 years. Abdominal pain was the commonest presenting feature (Table III). Group 1 saw a total of 2217 new attendances in this period and group 2 saw 2511 new patients. This leaves a shortfall of 546 patients which represents those who were seen by nurse practitioners, ward doctors or those who did not wait.

Overall, 58% of PAFs were not justified. Group 1 ordered 42 PAFs of which 29 were not justified (69%)(Table IV). Group 2 ordered 58 PAFs of which 29 were not justified (50%). Chi squared testing of Group 1 (13 justified/42) versus Group 2 (29 justified/58) gives a p-value of 0.09 with Yates correction.

TABLE III

Presenting Features in Patients sent for Plain Abdominal Radiography

Presenting Feature	*Number of Patients (Total 100)*
Abdominal Pain	41
Simple Constipation	19
Obstructive Symptoms	9
Lower Urinary Tract Infection Symptoms	9
Ingested Foreign Body	8
Gastrointestinal Bleed	7
Others	7

Table IV

Relative Proportion of Justified to Not Justified Plain Abdominal Radiographs for Groups 1 & 2

A&E Experience	*Justified*	*Not Justified*	*Total*
< 6 Months	13 films 31%	29 films 69%	42 films 100%
> 6 Months	29 films 50%	29 films 50%	58 films 100%

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CONCLUSIONS

Most PAFs ordered were not indicated. There was more appropriate requesting of PAFs by doctors of greater then 6 months AED experience than those with less. We are aware that the RCR guidelines are simply that, and a senior clinician may wish to disregard them in certain cases where personal clinical experience is at odds with the protocol. However it is the experience of the senior doctor which allows them to override these guidelines. We believe that inexperienced doctors are simply not applying the guidelines due to lack of awareness and over-investigating patients due to a fear of missing serious pathology. Junior doctors working in AEDs are therefore putting themselves at risk of breaching the IRMER regulations. These regulations have the force of criminal law and can result in prosecution if breached.

We believe that following 6 months full time experience in an AED there is a significant improvement in the justification of requested x-ray. Unfortunately once this experience is gained many SHOs will leave to work in other specialties where they will face a new set of clinical challenges. Equally inexperienced staff then replaces these SHOs and the cycle of over-investigation of AED patients continues.

We believe that measures must be taken to protect inexperienced AED SHOs from breaching these regulations and yet at the same time allow enough patient interaction to facilitate training. These measures could include ongoing audit of radiological investigation requests jointly performed by Radiology and Accident and Emergency Departments with active participation by all medical staff. Increasing the number of consultants in AEDs could improve the quality of both requesting of radiography and the supervision of inexperienced SHOs. These issues should have a higher profile in undergraduate and pre-registration training than is currently the case.

ACKNOWLEDGEMENT

The authors would like to thank Dr Gordon Cran for his help with the statistical analysis.

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The Ulster Medical Journal, Volume 70, No. 1, pp. 22-24, May 2001.

Absence of the palmaris longus muscle: a population study

N W Thompson, B J Mockford, G W Cran

Accepted 20 March 2001

SUMMARY

We examined 300 Caucasian subjects (150 males, 150 females) aged 18-40 years to assess the incidence of palmaris longus absence. The presence or absence of palmaris longus was assessed by clinical inspection.
Forty-nine subjects had unilateral absence of palmaris longus (16%). The tendon was absent bilaterally in 26 subjects (9%).
Unilateral and bilateral absence was more common in males, however this was not statistically significant (p= 0.25 and 0.56 respectively).
In those subjects with unilateral absence, the right side was found to be more commonly affected however no statistical significance was evident (p= 0.25).

INTRODUCTION

It is well known that individuals may have unilateral or bilateral absence of palmaris longus, a structure often used in reconstructive plastic surgery mainly in the setting of tendon grafting, although it has also been used for a wide variety of procedures including lip augmentation,[1] ptosis correction[2,3] and in the management of facial paralysis.[4]

The aim of this study was to determine the incidence of unilateral and bilateral absence of palmaris longus for the Caucasian population of Northern Ireland.

PATIENTS AND METHODS

For the purpose of this study, 300 Caucasian subjects (150 males, 150 females) aged between 18 and 40 years, were randomly selected by the principal author and examined. Individuals with a history of injury or abnormality of the upper extremities were excluded. Hand dominance was recorded for each subject.

The examination entailed observation of the volar aspect of the wrist, looking for the palmaris longus tendon in its usual anatomical position just ulnar to the flexor carpi radialis tendon. If the tendon was not visible, the patient was asked to oppose his or her thumb to the little finger and flex at the wrist (Figure 1). If all of the above

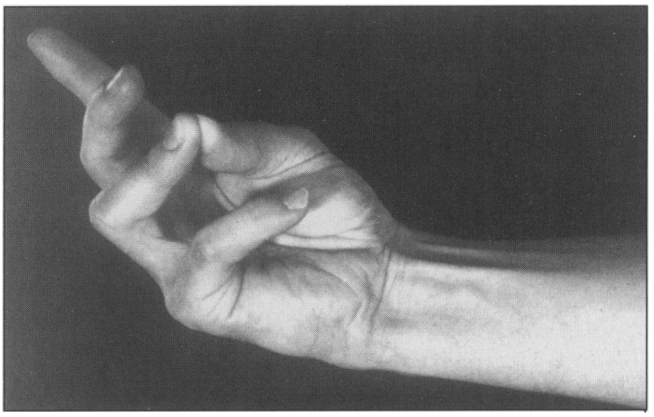

Fig 1. Subject demonstrating the presence of palmaris longus on clinical testing.

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failed to demonstrate a palmaris longus tendon, it was considered absent (Figure 2). The presence or absence of the palmaris longus tendon was recorded for both sides. Relationships between tendon absence, hand dominance and gender were analysed using the Chi-squared goodness-of-fit test.

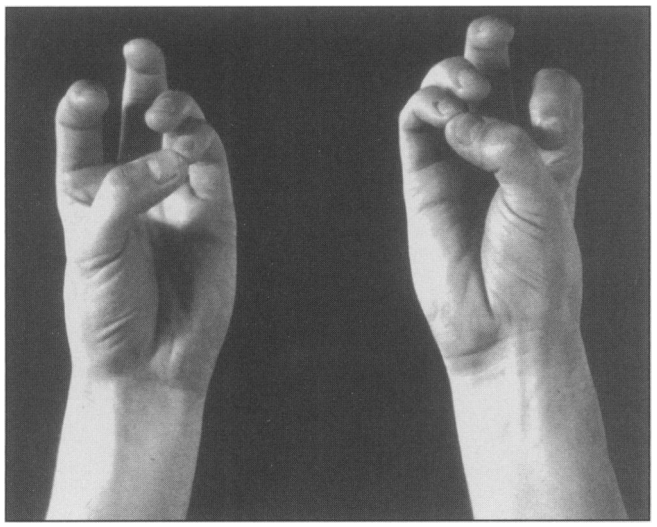

Fig 2. Subject demonstrating unilateral absence of palmaris longus (left).

RESULTS

Of the study population, twenty-one subjects were left-hand dominant (7%). Average age was 29 years (range, 18-40 years).

Twenty-six subjects (9%) were found to have bilateral absence of the palmaris longus tendon (95% CI, 5-12%). Forty-nine subjects (16%) had a unilateral absence of the tendon (95% CI, 12-20%). Of this group, the tendon was absent on the right side in 29 subjects. Twenty subjects had absence of the tendon on the left. Of the subjects with bilateral absence of palmaris longus, 15 were male and 11 female. Of those with unilateral absence, 29 male subjects and 20 female subjects were affected (Table 1).

TABLE I

Results after clinical evaluation of 300 subjects for the presence or absence of palmaris longus.

Tendon absence	*Males*	*Females*	*Right* side	*Left* side
Unilateral	29	20	29	20
Bilateral	15	11	#	#

DISCUSSION

Tendon grafts are frequently needed in reconstructive surgery on the hand. Many surgeons agree that the palmaris longus tendon is the first choice as a donor tendon because it fulfils the necessary requirements of length, diameter and availability, and can be used without producing any functional deformity.[5] The palmaris longus tendon is often considered the ideal donor for tendon grafts for replacement of the long flexors of the fingers, and of the flexor pollicis longus tendon.[6]

Palmaris longus is often described as one of the most variable muscles in the human body and is classified as a phylogenetically retrogressive muscle i.e. a short belly with a long tendon.[7] In vertebrates it is found only in mammals and is best developed in those where the forelimb is used for ambulation.[8] For example, the palmaris longus is always present in the orangutan [9] but is variably absent in higher apes such as chimpanzees and gorillas.[8] In humans the absence of palmaris longus appears to be hereditary but its genetic transmission is not clear.[9]

From the results of numerous previous studies investigating the incidence of palmaris longus absence it has been reported that bilateral absence occurs in 8% to 16% of individuals, with unilateral absence occurring in 4% to 14%.[8] Our figures compare favourably (bilateral absence, 9%; unilateral absence, 16%), and like most of the previous studies, reflects the incidence of palmaris longus absence in Caucasian individuals.[8,9] Racial variations are however well recognised.[5,9]

Previous studies have conflicted with regard to the incidence of palmaris longus absence in relation to gender and body side.[9-11] In our study, males were found to have a higher incidence of both bilateral and unilateral absence of palmaris longus, however this was not statistically significant (p= 0.56 and 0.25 respectively).

Furthermore, although the right side was affected more commonly in unilateral absence this also did not prove to be statistically significant (p= 0.25). In summary, the palmaris longus tendon is often regarded as the ideal tendon donor. Clinical testing revealed an incidence of unilateral absence of 16% and a bilateral absence of 9% in the Northern Ireland population. No statistically significant correlation was found between tendon absence and gender or body side.

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The Ulster Medical Journal, Volume 70, No. 1, pp. 25-30, May 2001.

Abnormal uterine bleeding: an evaluation endometrial biopsy, vaginal ultrasound and outpatient hysteroscopy

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Accepted 26 March 2001

SUMMARY

Abnormal uterine bleeding is a common gynaecological symptom. Whilst most patients have benign disease, thorough investigation is necessary, particularly in the peri- and post- menopausal woman. Hysteroscopy with directed biopsy of suspicious lesions is the gold standard investigation but it is invasive and is not offered in all units as an outpatient procedure. Ultrasound and outpatient biopsy techniques may allow patients to be triaged to select those who require formal evaluation by hysteroscopy.

We assessed the records of 100 consecutive referrals to the outpatient hysteroscopy clinic at the Royal Maternity Hospital, Belfast. In order to develop a nomogram for the investigation of women with abnormal bleeding patterns, we compared ultrasound/biopsy diagnoses with hysteroscopy/biopsy diagnosis. Pre- and peri- menopausal patients, and post-menopausal patients taking hormone replacement therapy should be investigated further if the endometrial thickness is more than 10 mm, if endometrial biopsy is abnormal, or if symptoms are recurrent. Similarly, post- menopausal patients not taking hormone replacement therapy with an endometrial thickness of 5 mm or more should be referred for hysteroscopy.

INTRODUCTION

Abnormal uterine bleeding is a common gynaecological symptom. In peri- and post-menopausal women it is essential to exclude endometrial carcinoma although the incidence is only 6.96 per 1000 women with post menopausal bleeding.[1] In younger women, endometrial hyperplasia and anatomical anomalies, such as uterine fibroids, comprise the main pathology.[2]

The recommendation regarding investigation of abnormal uterine bleeding from the Royal College of Obstetricians and Gynaecologists is that women, over the age of 45, should be investigated with hysteroscopy and endometrial biopsy.[3] Frequently, this is performed as an inpatient procedure with the biopsy being obtained by uterine curettage (D&C). Blind sharp curettage covers as much as 60% of the cavity but may miss polyps as they recoil from the passing curette.[4] However, inpatient investigations are costly and there are risks associated with the use of general anaesthesia.

Outpatient alternatives to D&C include microhysteroscopy (either flexible or rigid), transvaginal ultrasonography, with or without saline infusion (sonohysterography) and endometrial biopsy. Transvaginal ultrasonography is useful in determining endometrial thickness and morphology as well as the regularity of the endo/myometrial border.[5] Whilst the procedure is well tolerated by patients, sessile or pedunculated lesions of the endometrium and malignant disease cannot be definitively excluded.[6,7] However, it has been reported that post-menopausal malignant disease is very unlikely when the endometrial thickness is less than 5 mm and the patient is not taking hormone replacement therapy (HRT).[8] Vaginal sonohysterography is a modification of vaginal ultrasound in which a small volume of saline is injected into the uterine cavity during ultrasound examination.[9] This enables irregularities of the

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endo/myometrial border and the free endometrial border to be more easily visualised.[10]

Outpatient endometrial sampling devices have a narrow bore (typically about 3 mm external diameter) and sample the endometrium by aspiration. Although they pass relatively easily through the cervical canal, there may be associated pain when the sample is harvested and, unlike directed biopsy, the sample is obtained blind. As little as 4% of the cavity may be sampled and polypoid lesions are unlikely to be removed.[11] However, the reported sensitivity for detecting endometrial abnormality is approximately 85%.[12, 13]

Outpatient hysteroscopy is both feasible and highly acceptable in the majority of patients, giving a high detection rate for intrauterine pathology.[14, 15] It allows the operator to take a directed biopsy and is more specific and sensitive than transvaginal ultrasound or blind endometrial sampling. As an investigation for abnormal uterine bleeding it was recommended by both the Royal College of Obstetricians and Gynaecologists and the American College of Obstetricians and Gynecologists in 1994.[3, 16] However, it is significantly less easily performed than either ultrasound or biopsy. Therefore, a means of selecting patients by, for example, menopausal status, symptomatology and associated clinical findings might lead to a more efficient use of hysteroscopy than a blanket recommendation that all cases of abnormal bleeding should undergo the procedure.

Our aim was to develop a nomogram using historical information, ultrasound and biopsy results to select those women who should have a formal hysteroscopic evaluation.

METHODS

The charts of 100 consecutive referrals for outpatient hysteroscopy from the general gynaecology clinics at the Royal Group of Hospitals' Trust, Belfast, were reviewed. The decision to refer was made by the patient's consultant: there were no defined guidelines for referral: a number of patients had vaginal ultrasound scan and endometrial biopsy at the outpatient clinic prior to referral (these procedures were repeated at the outpatient hysteroscopy clinic). The indication for the procedure, the subject's age, parity, menstrual status and outcome of the investigation were recorded. All of the procedures were performed by a single operator. Whilst the data was recorded prospectively at the time of consultation it was analysed retrospectively.

The examination of each subject followed a standard protocol. The patient was placed in the lithotomy position with her ankles supported by stirrups. A bimanual vaginal examination was performed to note the size, position and shape of the uterus and the presence or absence of adnexal pathology. Vaginal ultrasound examination using a 5 MHz probe (FF Sonic, Fukuda Denshi, Japan) was performed to determine the dual endometrial thickness (ET), that is, the perpendicular distance from one endo/myometrial border to the other at its widest part in longitudinal section, and to identify endometrial polypoid lesions, free fluid or fibroids.

For the hysteroscopy a Simm's speculum was inserted into the vagina and the cervix grasped with a single toothed tenaculum forceps. A 3.4 mm external diameter Hamou Microhysteroscope (Karl Storz, Tuttlingen, Germany) was passed through the cervix, under direct vision, into the uterine cavity which was distended by carbon dioxide via a hysteroflator (Karl Storz) set at maximum pressure 75 mmHg and flow rate of 200 mls/minute. The cavity was illuminated by a Coldlight Fountain (Karl Storz) and the images displayed on a Sony Trinitron monitor using a Telecam Pal single chip camera system (Karl Storz). The endometrium was serially inspected for pathology. When the inspection of the cavity was complete the gas and scope were removed from the uterus.

Finally, a Pipelle endometrial sampler was used to obtain a biopsy of the endometrium using no more than two attempts at sample aspiration. The specimen was placed in formalin and sent for histopathological analysis. Neither analgesia or anaesthesia was routinely used and was not required by any patient in this series.

The histological and ultrasound diagnoses were combined to formulate a working ultrasound/biopsy diagnosis and this was compared with a similarly combined hysteroscopy/biopsy diagnosis.

RESULTS

The average age of the patients referred was 48.5 years (29-78). Six subjects were nulliparous, six were grand multiparas and the remainder had had

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Table I

Comparison of hysteroscopy/biopsy and transvaginal scanning/biopsy diagnoses in postmenopausal patients

Hysteroscopy/biopsy diagnosis	*Ultrasound/biopsy diagnosis concordant*	*Ultrasound/biopsy diagnosis discordant*
Normal	16	0
Atrophy	16	1 endometrial polyp
Endometrial polyp	2	2 normal cavity
Endocervical polyp	0	3 normal uterus
Neoplasm	1	0

between one and five vaginal deliveries. Of the 100 patients, it was not possible to perform outpatient hysteroscopy in two cases. Both were post-menopausal and subsequently underwent hysteroscopy under general anaesthesia. In one of these patients the uterus was anteverted and acutely anteflexed, in the other, access to the vaginal vault and cervix was impossible and a diagnosis of erosive vaginal lichen planus was eventually made. These two patients were not considered further in the analysis.

Ninety two (94%) patients had endometrial biopsy samples which were adequate for histological diagnosis. The six patients whose biopsies were not adequate for diagnosis had atrophic endometrium diagnosed both by hysteroscopy and scanning.

Forty-one (41.8%) of the patients were post-menopausal and, of these, 25 (61%) were using hormone replacement therapy. In 35 of the 41 subjects (85.4%) the hysteroscopic diagnosis was in agreement with the ultrasound diagnosis. Thirty-two (78%) had a diagnosis of a normal cavity or of atrophy, two (4.9%) of benign endometrial polyps and one (2.4%) of endometrial cancer (endometrial thickness 20 mm on scanning and copious blood clot within the cavity on hysteroscopy). Of the remaining six (14.6%), three had normal ultrasound and biopsy but endocervical polyps were noted at hysteroscopy (two HRT users), one of whom also had a small endometrial polyp (endometrial thickness >5 mm in all three). Three further subjects had endometrial polyps in isolation noted on hysteroscopy (one HRT user), two of whom had an endometrial thickness >6 mm (summarised in Table I) and one a diagnosis of atrophy on the basis of the ultrasound biopsy findings (endometrial thickness 3 mm, HRT user). Use of HRT was associated with a non-significant increase in ET on scanning (average ET: no HRT 3.9 mm, HRT 5.8 mm, p=0.23) and patients were no more likely to have endometrial or endocervical pathology if using HRT.

Of the 57 peri- and pre-menopausal patients the ultrasound/biopsy and hysteroscopy/biopsy diagnoses were concordant in 52 (91%). There were 50 (87%) cases with a normal cavity, atrophy or small fibroids not causing significant distortion of the cavity, one (2%) case of benign hyperplasia and one (2%) endometrial polyp.

Two further cases of endocervical polyps (both with ET >10 mm), one (2%) sub-septate uterus and one (2%) endometrial polyp (ET 12 mm) were diagnosed with hysteroscopy but not with ultrasound and one (2%) case of suspected neoplasm on ultrasound (but not on biopsy) was found to have a benign fibroid at hysteroscopy Table II). No endometrial or endocervical pathology was detected in any pre- or peri-menopausal patient with an ET of less than 11 mm. However, in postmenopausal patients with an ET of 5 mm or less one patient had a small endometrial polyp noted at hysteroscopy.

In total, the diagnoses were concordant in 87 (89%) cases. Five cases of endocervical and four cases of endometrial polyps were noted at hysteroscopy but not at ultrasound/biopsy. If hysteroscopy/biopsy is taken as the gold standard the overall sensitivity of transvaginal scanning combined with biopsy is 75% for endometrial pathology with a specificity of 90% (positive predictive value 40%, negative predictive value

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Table II

Comparison of hysteroscopy/biopsy and transvaginal scanning/biopsy diagnoses in pre- and perimenopausal patients

Hysteroscopy/biopsy diagnosis	*Ultrasound/biopsy diagnosis concordant*	*Ultrasound/biopsy diagnosis discordant*
Normal	40	0
Fibroids	9	1 suspected neoplasm
Endometrial polyp	1	1 normal uterus
Endocervical polyp	0	2 normal uterus
Huperplasia	1	0
Septum	0	1 normal uterus
Atrophy	1	0

98%). When a cut-off value of 5 mm for ET in post-menopausal women not taking HRT is taken, vaginal ultrasound/biopsy achieved a sensitivity of 100%. The 10 mm ET cut off for pre-, peri- and postmenopausal women taking HRT, has a lower sensitivity, however, the positive predictive value is increased (i.e. when pathology was diagnosed, it was more often present).

DISCUSSION

This is the first prospective comparison of transvaginal ultrasound with biopsy versus outpatient hysteroscopy with biopsy in a patient population with both pre- and postmenopausal patients. A single operator who had considerable experience in each technique performed all of the procedures. The subject body constituted 100 consecutive referrals from seven gynaecologists: the decision to refer was taken by the clinicians on an individual basis and no specific criteria were set. The patients included were those whom the referring gynaecologists felt were likely to have pathology and to require a more thorough evaluation than could be offered by ultrasound and biopsy alone. As the purpose of this study was to develop a nomogram for evaluation of patients with abnormal pre- or postmenopausal bleeding, we feel that this enhanced the power of the study by including only those likely to have pathology.

In the literature to date, there have been numerous reports examining the role of ultrasound or hysteroscopy but without biopsy. Biopsy is likely to miss polyps and fibroids whilst ultrasound is likely to identify intramural fibroids but be unable to differentiate between endometrial hyperplasia, endometrial polyps and early cancers. Hysteroscopy can identify discrete lesions but does not give a histological diagnosis. Therefore, any effective clinical service is likely to require all three methods of investigation. However, hysteroscopy is expensive in comparison and requires considerable operator skill.

In this study, the evaluation of the post-menopausal group suggests that if the endometrium is thin (<5 mm) the likely diagnosis is atrophy and this can be confirmed by endometrial sampling although caution must be exercised when an inadequate biopsy is reported.[17] If atrophy is diagnosed, oestrogen replacement may alleviate symptoms. If bleeding recurs hysteroscopy should be performed. By contrast, if the ET is >5 mm then formal hysteroscopic evaluation is indicated.

In the group of postmenopausal women using HRT, an ET of more than 10 mm (even with a normal biopsy) is an indication for hysteroscopy. In women with normal biopsy and ET 10 mm or less a period of observation is acceptable but if further bleeding occurs, referral for hysteroscopy should be made. The advantage of the raise in limit for those taking HRT is that the number of unnecessary hysteroscopies will be decreased.

For pre-menopausal patients changes in endometrial thickness throughout the menstrual cycle may cause confusion. Although ET commonly exceeds 10 mm in normal uteri,

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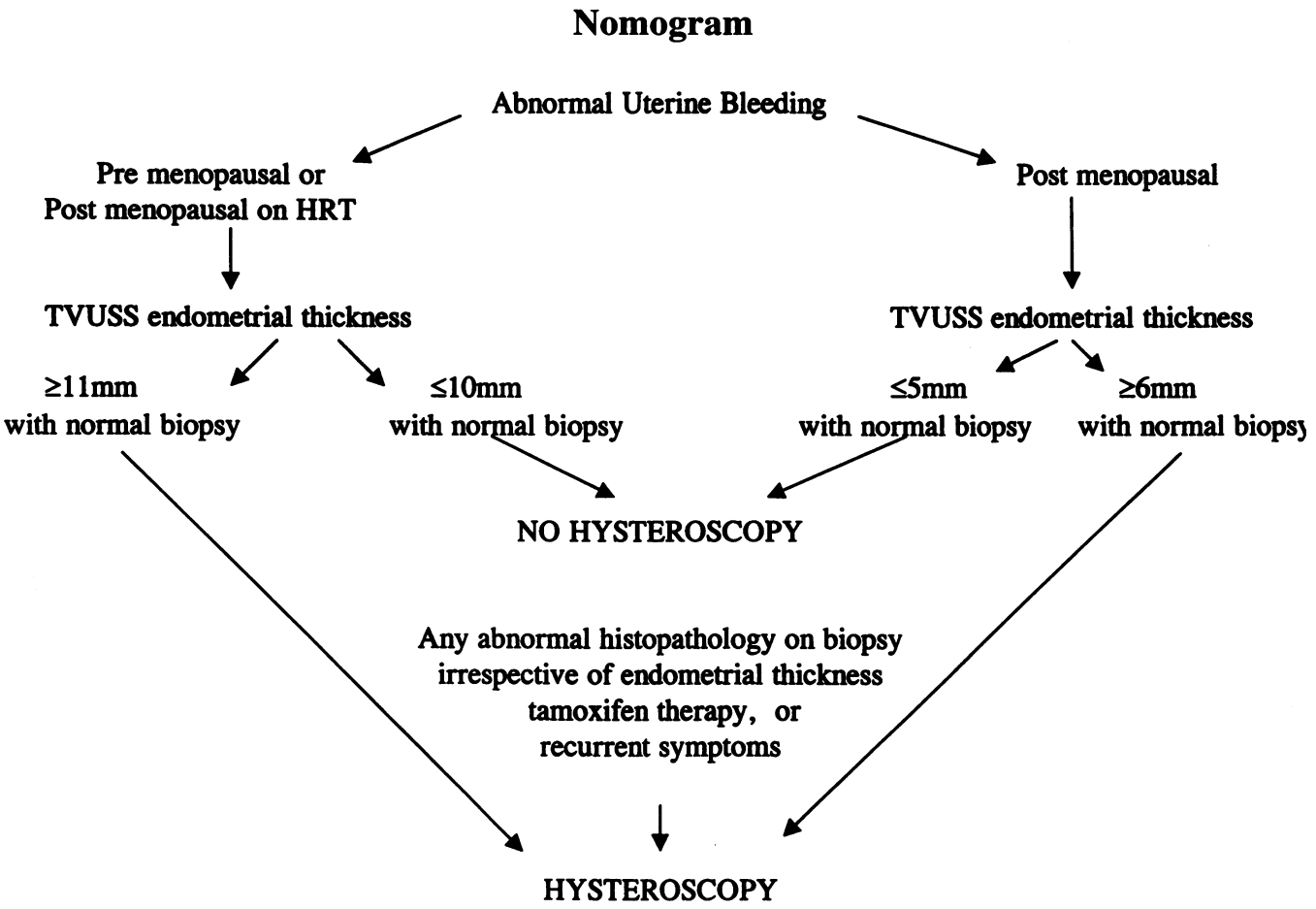
because of the difficulty in differentiating between polyps, hyperplasia and fibroid polyps, we believe that ET greater than 10 mm should lead to referral for hysteroscopy in women with abnormal uterine bleeding. If, at ultrasound, the cavity appears distorted by submucosal fibroids, the only indication for hysteroscopy is to assess their suitability for hysteroscopic resection.

Our findings largely support those previously reported.[18] For example, Guidot [12] and Schei [4] have shown that micro-curettage alone is not a safe alternative to hysteroscopy as polyps are often missed and may contain focal hyperplastic or malignant change. We have refined this further to demonstrate that, in combination with ultrasound and with the provisos given above, hysteroscopy can often be safely avoided. However, an abnormal histological report on a biopsy specimen in a woman of any age, or persistent abnormal uterine bleeding, should prompt urgent referral for hysteroscopy.

Whilst Naegele *et all* [4] suggested that outpatient hysteroscopy may become a routine procedure (as ultrasound presently is in some units) the financial implications of such a shift in clinical practice would be huge and a large number of unnecessary procedures would be performed. The guidelines of both the American and Royal Colleges of Obstetricians and Gynaecologists are less radical in their enthusiasm for hysteroscopy and suggest appropriate patient selection. Guidelines tor such patient selection have yet to be drawn up at a national level and we believe that a significant number of procedures are being performed without adequate indication. Hysteroscopy is well tolerated by women and is an important adjunct to clinical examination, transvaginal ultrasound scanning and endometrial biopsy. However, as the most invasive of the procedures it should be reserved for those cases where it is clearly indicated rather than applied to all cases.



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The Ulster Medical Journal, Volume 70, No. 1, pp. 31-35, May 2001.

Idiopathic intracranial hypertension; incidence, presenting features and outcome in Northern Ireland (1991-1995)

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Accepted 27 October 2000

SUMMARY

Objectives: **to determine the age and sex specific incidence rates, presenting features, and visual outcome of idiopathic intracranial hypertension in Northern Ireland.**

Methods: **A case-note review of all patients with idiopathic intracranial hypertension, diagnosed at the Royal Victoria Hospital, Belfast between 1991 and 1995.**

Results: **Forty-two patients were identified corresponding to an average annual incidence rate per 100,000 persons of 0.5 for the total and 0.9 for the female population. The commonest presenting symptoms were headache (84%), transient visual obscurations (61%) and sustained visual loss (34%). Impaired Snellen visual acuity and visual field loss were documented in 21% and 62% of patients respectively at presentation, and in 24% and 39% at last follow-up. One patient suffered deterioration in visual functioning sufficient to interfere with normal daily activities. Conclusions: The age and sex specific incidence rates of IIH in Northern Ireland are lower than have been reported in previous population-based series. Disabling visual loss occurs in a small number of patients despite all interventions.**

INTRODUCTION

Idiopathic intracranial hypertension (IIH) is a condition of unknown aetiology characterized by symptoms and signs of increased intracranial pressure in the absence of an intracranial lesion or hydrocephalus. All neuroscience units manage IIH, yet information on epidemiology is very limited and is based on studies that have involved either small numbers of patients or short periods of follow-up.[1-3] Figures for European populations are unknown and some reports have suggested that IIH is less common in Europe.[4,5]

Visual loss is the most important complication of IIH. Previous studies have suggested that 96% of patients have visual abnormalities at some time,[6] and up to 10% develop bilateral blindness.[7] There are no controlled trials of treatments in this condition, and despite all therapeutic interventions some patients develop progressive blindness.

The aims of our study were: (1) to determine the population-based incidence rates of IIH in Northern Ireland, over a five year period (1991-1995), (2) to provide clinical details of cases of IIH diagnosed in our locality over this period, and (3) to determine the frequency of visual complications in our patients.

CASES AND METHODS

Cases were identified by a computer search of the medical records for all patients treated at the Royal Victoria Hospital (RVH), Belfast between the 1st of January 1991 and 31st of December 1995 for whom the following diagnoses were made; pseudotumour cerebri, benign intracranial hypertension or IIH. The RVH is a university teaching hospital, and houses the only departments of neurology and neurosurgery and the main department of ophthalmology that serve Northern

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Ireland (Population 1.64 million). All patients with IIH would be expected to be seen in this hospital at some stage. Inclusion in the study was based on the Modified Dandy Criteria for IIH (Table 1). To be considered, the diagnosis of IIH should have been made between 1st January 1991 and the 31st of December 1995 and residency in Northern Ireland had to be established at least 1 year before diagnosis.

TABLE I

Modified Dandy Criteria for the diagnosis of idiopathic intracranial hypertension.[8]

1. Signs and symptoms of increased intracranial pressure.
2. Awake and alert patient,
3. No abnormal neurological findings except papilloedema or a sixth nerve palsy.
4. Normal CT/MRI except for empty sella syndrome or small ventricles.
5. Documented increased CSF opening pressure (>200 mm of water in non-obese and >250 mm of water in obese patient), with normal CSF composition.
6. No other known cause of raised intracranial pressure.

Age and sex specific incidence rates were calculated by dividing the number of incident cases by the proper denominator, gained from census data for Northern Ireland 1993. Data were independently collected from the case-notes by two of the authors (JC + DM). The follow-up period was to the 31st May 1996. Variables collected were symptoms, and examination findings, recorded at presentation and on each visit.

RESULTS

Forty-two cases, with a mean age of 29 years at diagnosis, were identified in the five-year study period. Average annual incidence rates per 100,000 persons for IIH by age and sex are shown in table II. The average annual incidence rates per 100,000 were 0.6 for the total and 0.9 for the female population. The female-to-male ratio of incidence rates was 5.7:1.0.

Symptoms (Table III) Duration of symptoms prior to diagnosis varied from one to 208 weeks. Headache was the presenting symptom in 32 cases and occurred daily in almost two-thirds. Transient visual obscurations (TVOs) were reported by five of the six cases that did not have headache. Half reported weight gain in the year before diagnosis.

Eleven (26%) patients had magnetic resonance (MR) imaging of brain with MR venography. These were reported as normal for all cases.

Table II

Age and sex specific average annual incidence rates of idiopathic intracranial hypertension per 100,000 population in Northern Ireland, 1991 to 1995.

	Females		*Males*		*Total*	
Age Group, years	*No.*	*Rate*	*No.*	*Rate*	*No.*	*Rate*
0-14	2	0.20	1	0.10	3	0.10
15-24	12	1.98	0	0.00	12	0.96
24-35	10	1.56	2	0.32	12	0.93
35-44	7	1.33	1	0.19	8	0.77
45+	5	0.35	2	0.16	7	0.26
All age groups	36	0.86	6	0.15	42	0.51

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TABLE III

Presenting symptoms

Symptom	%
Headache	84
Transient visual obscurations	61
Sustained visual loss	34
Scintillations	18
Diplopia	11
Intracranial noises	8
Retrobulbar pain	5
Deafness	3
Weight gain	50

Ophthalmological Examination All patients had optic disc swelling at presentation (Figure 1). In eleven cases it was mild and in 21 moderate to severe. Corrected Snellen visual acuity at presentation was worse than 6/9 in at least one eye in eight (21%) patients. An abnormality other than enlargement of the blind spot was recorded in 18 (62%) of the 29 who had perimetric assessment of visual fields performed at presentation. The commonest field abnormalities noted were generalized constriction and infero-nasal field defects (Figure 2).

Sixteen (42%) cases had persistent disc swelling (ten mild, six chronic) at last follow-up. Corrected Snellen visual acuity was worse than 6/9 in at least one eye in nine (24%) cases. Four of these nine were cases whose visual acuity had been normal at presentation. None had deterioration of visual acuity sufficient to interfere with normal daily activities. Seven (39%) of the 18 cases who had visual field loss documented at presentation had complete resolution of their defects. One case had a worsening of field loss with marked field constriction to within 20 degrees of fixation, which interfered with activities of daily living. The rest maintained persistent mild defects that did not interfere with visual functioning.

Patients were treated with acetazolamide or diuretics initially. Analgesic preparations taken on a regular basis by almost three-quarters of the patients were never effective at relieving headache. Over half also had repeated lumbar punctures. Weight loss was documented in eight

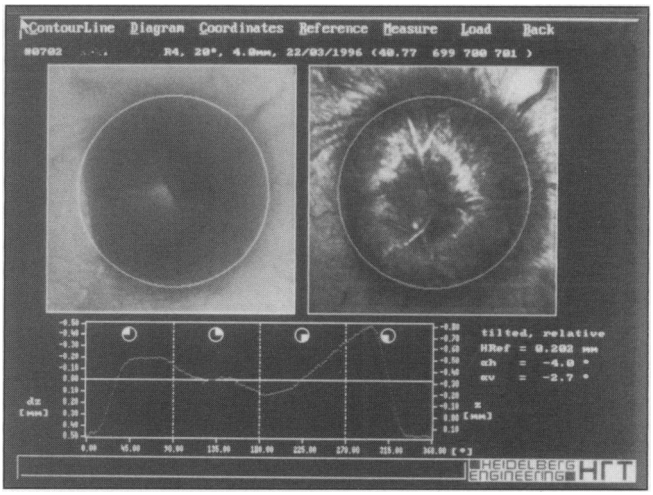

Fig 1. Confocal laser tomogram of right eye of patient showing significant disc swelling.

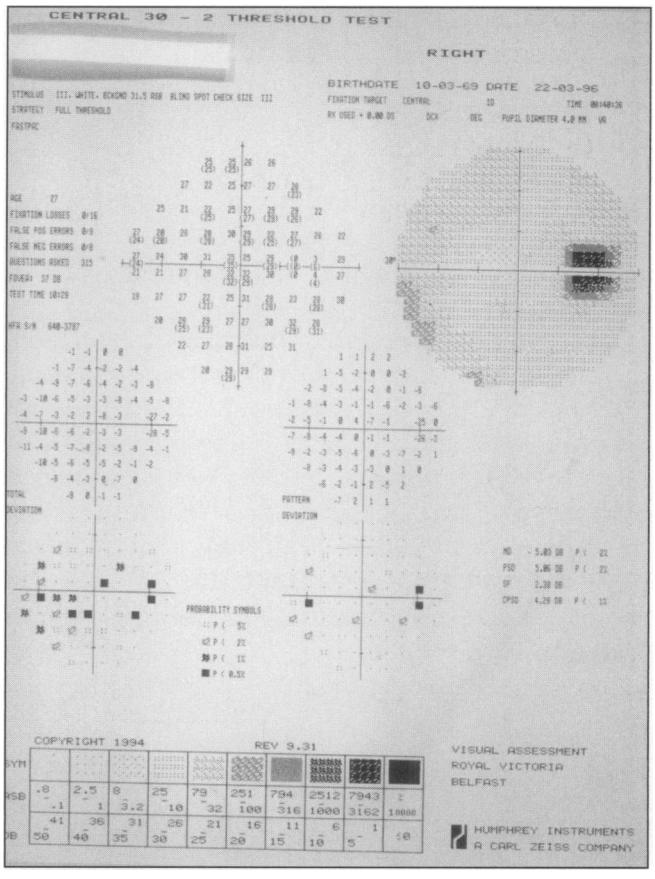

Fig 2. Humphrey visual field of central 30° of vision of right eye of patient showing slight enlargement of the blind spot and early infero-nasal field loss, which is often reversible.

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of the 21 patients reporting significant weight gain prior to symptom onset. This was sustained in four, with three having sustained symptomatic improvement and reduced CSF opening pressure. Seven cases (18%) had a theco-peritoneal shunt inserted. Optic nerve sheath fenestration was not performed in any of the cases. Symptomatic improvement occurred in all patients who had a shunting procedure, which was maintained for a mean of 16 months. All patients who had surgical management developed post-operative problems, including a return of symptoms of raised intracranial pressure in six patients; one of these patients also had a shunt infection. The other patient developed chronic headache due to persistently low cerebrospinal fluid pressure. Further surgery, amounting to 15 additional procedures, was necessary in four of these six patients.

DISCUSSION

The average annual incidence rates of symptomatic IIH in Northern Ireland per 100,000 persons at 0.5 for the total population and 0.9 for females are lower than those reported in the three previously conducted population based studies evaluating the incidence of this condition.[1-3] In these studies the incidences per 100,000 ranged from 0.9 to 1.7 for the total population and 1.6 to 3.6 for females. Whether our results are due to incomplete ascertainment or that the incidence of IIH is lower in our locality, due to such factors as the higher frequency of obesity in the populations previously studied, is unknown. Patients with papilloedema and headache without tumour will be referred to our single neuroscience centre; diagnosis is precise and our population denominator is stable. Thus we feel that the observed rates are representative of the true incidence rates of symptomatic IIH in Northern Ireland.

Headache was less often the presenting symptom in our cases. Most of those who did not have headache had TVOs. TVOs without headache have been documented in IIH[9] but never as frequently as in our series. This highlights the importance of establishing whether papilloedema and raised intracranial pressure are present in patients with TVOs.

Visual acuity and visual fields were impaired in a substantial number of patients at presentation reflecting the delay between symptom onset and diagnosis. Visual loss severe enough to interfere with activities of daily living was however uncommon at any stage in our cases. In addition the abnormalities found on testing visual functioning often improved. Our results are in keeping with those of the population-based series from Rochester, Minnesota[3] where nine patients identified over a 15 year period from a population of 70,000 were followed up for a median period of 2.7 years. Out of 18 eyes only three developed visual impairment. This was mild in all cases and did not interfere with everyday visual functioning. Such findings are at odds with the commonly held belief that disabling visual loss is a frequent result of IIH,[6] and might be explained by the relatively short follow-up period of our cases and those from Rochester. However, some of the difference could also be accounted for by selection bias in non-population-based studies with more severely affected cases being studied.

Efficacy is claimed for medical treatments used in IIH.[6, 10-14] There are however no controlled trials of any of them and patients are therefore managed according to personal or local preferences. This is also the case for the surgical procedures used. There is no doubt that surgical intervention has saved vision in some patients with IIH[15-17] but the effectiveness of either shunting procedures or optic nerve sheath fenestration have never been studied in a controlled way, and it is unclear when surgical intervention should be undertaken. Both types of procedure are also not infrequently associated with post-operative complications;[18-22] some patients may therefore be having an unnecessary intervention with attendant significant morbidity.

In conclusion, we found that the age and sex specific incidence rates of IIH are lower in Northern Ireland than in previous population-based studies and that visual loss sufficient to interfere with normal activities was uncommon. In keeping with the results from Rochester, Minnesota, this raises the possibility that the prognosis for vision in IIH is better than previously suggested from the results of non-population based series.

ACKNOWLEDGEMENTS

We are grateful to our colleagues for permitting us to review their patients.

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The Ulster Medical Journal, Volume 70, No. 1, pp. 36-39, May 2001.

The Presidential Regalia of the Ulster Medical Society

R W Stout

Accepted 1 March 2001

The Ulster Medical Society is fortunate in having some beautiful regalia for the President and Vice-Presidents. The Chain of Office is worn by the President on only two occasions during the year, the Presidential Address and the Annual Dinner. During the remainder of the year the President wears a badge and ribbon. There are also two Vice-Presidential Badges and Ribbons.

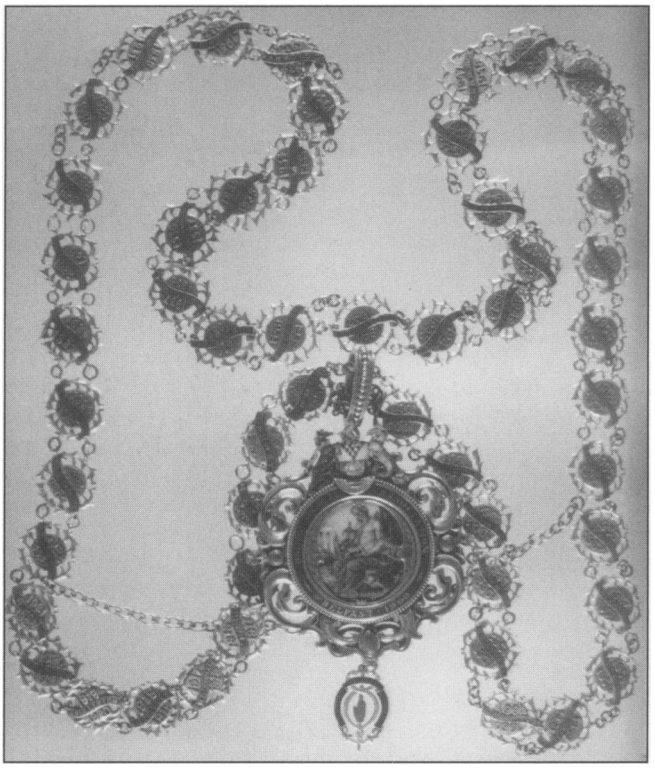

Fig 1. The presidential badge and chain..

Fig 2. The presidential badge.

THE PRESIDENTIAL CHAIN AND BADGES

The formal Presidential regalia is an ornate chain from which is suspended the badge of the British Medical Association and below it the badge of the Society (Figure 1). The large badge is engraved on the front 'British Medical Association Belfast 1909' (Figure 2). It was presented by the medical profession of Ulster to Sir William Whitla who was President of the BMA when it met in Belfast in that year (Figure 3). 1909 was the year in which

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Queen's University first took students as a university, having previously been a College of the Queen's University of Ireland. Sir William Whitla recognised this fact in his Presidential Address in which he described new facets of medical education in the new university.[1] Sir William donated the badge to the Ulster Medical Society as the Presidential badge in 1912 (Figure 3). The chain consists of a series of linked enamelled badges, each containing the name of a President and his/her dates of office. Presidents from 1912-13 until 1965-66 are recorded on the chain (Table 1). There is one absentee: Mr J A Craig was President in 1925-26 but has no link on the chain. There is no mention in the minute books of the Society of why his name has not been included. It may be that the link has been lost. Mr T S S Holmes was President for two successive years, 1939-40 and 1940-41. This was related to the onset of the Second World War (recorded in AGM minutes 30th May 1940). He is the only President since the institution of the Chain who has held office twice. Others in the past who have been President on two occasions have been James Cuming 1868-69 and 1881-82, Professor Robert F Dill 1879-80 and 1883-84, John Fagan 1884-85 and 1885-86, Sir William Whitla 1886-87 and 1901-02, John Campbell 1902-03 and 1903-04, William Calwell 1904-05 and 1905-06 and J J Austin 1909-10 and 1910-11.[2]

TABLE I

The Presidents of the Ulster Medical Society whose names are engraved on the obverse of the presidentail chain of office.

1912-13	R W Leslie	1939-41	T S S Holmes
1913-14	A B Mitchell	1941-42	G G Lyttle
1914-15	J S Morrow	1942-43	R Marshall
1915-16	A G Robb	1943-44	W Dickey
1916-17	Robt Campbell	1944-45	W A Anderson
1917-18	W D Donnan	1945-46	H P Hall
1918-19	J Colville	1946-47	J R Gillespie
1919-20	A Fullerton	1947-48	G R B Purce
1920-21	T Houston	1948-49	S Barron
1921-22	R Hall	1949-50	A J Dempsey
1922-23	R J Johnston	1950-51	Robin Hall
1923-24	W StC Symmers	1951-52	J G Johnston
1924-25	J S Darling	1952-53	J C Robb
1926-27	M J Nolan	1953-54	W G Frackleton
1927-28	J C Rankin	1954-55	J A Smyth
1928-29	T H Milroy	1955-56	F M B Allen
1929-30	Howard Stevenson	1956-57	G D F McFaddan
1930-31	Henry Hanna	1957-58	Olive Anderson
1931-32	S T Irwin	1958-59	C H G Macafee
1932-33	C G Lowry	1959-60	J C Smyth
1933-34	W J Wilson	1960-61	J A L Johnston
1934-35	S R Hunter	1961-62	F A MacLaughlin
1935-36	Foster Coates	1962-63	C W Kidd
1936-37	P T Crymble	1963-64	J R Wheeler
1937-38	W W D Thomson	1964-65	Kathleen M Cathcart
1938-39	J M McCloy	1965-66	J S Loughridge

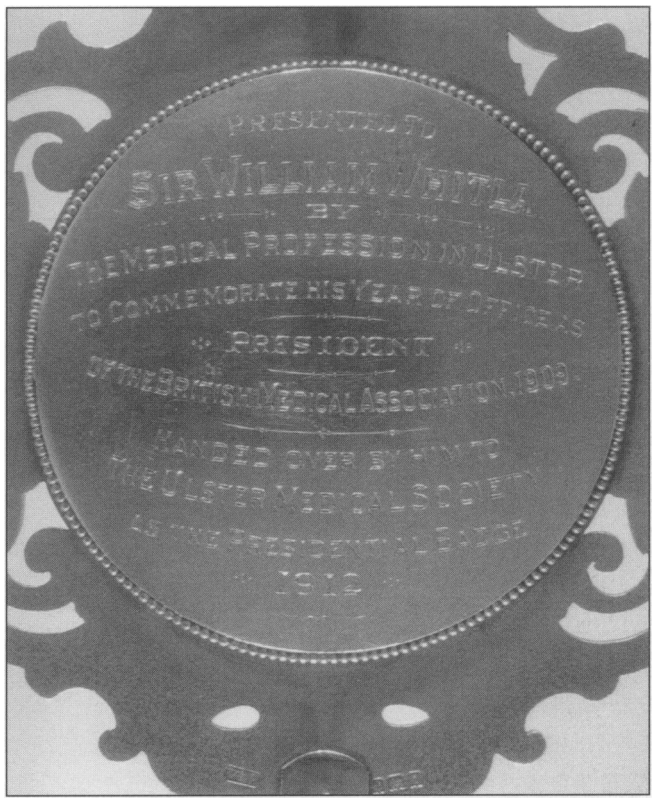

Fig 3. The reverse of the presidential badge.

The chain is long and heavy and when the present author became Honorary Treasurer of the Society, it was in two parts with about 20 of the links in a separate section which was not worn. The chain was restored to its original length at that time. It is understood that the practice of recording the name of the President on an enamel link in the chain had to be discontinued after 1966 due to its cost. Since that time the names of the Presidents have been engraved on the reverse of the links of the chain (Table 2). There are 52 links in the chain and 34 have already been engraved on the reverse. In 18 years time the Society will have to decide how to record the names of future Presidents. A complete list of Presidents and other office bearers from the beginning of the Society until 1967, was published in Dr R W M Strain's history of the Ulster Medical Society.[2]

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TABLE II

The Presidents of the Ulster Medical Society whose names are engraved on the reverse of the presidential chain of office

1966-67	J A Price	1983-84	J M G Harley
1967-68	Sir Ian Fraser	1984-85	J A Weaver
1968-69	R W M Strain	1985-86	M G McGeown
1969-70	R S Allison	1986-87	W G Irwin
1970-71	Kirk Forsythe	1987-88	D S Gordon
1971-72	Sir John Biggart	1988-89	A G McKnight
1972-73	J A McVicker	1989-90	J F O'Sullivan
1973-74	D H Craig	1990-91	Herbert Baird
1974-75	J E Morison	1991-92	Hume Logan
1975-76	D A D Montgomery	1992-93	J F McKenna
1976-77	G T C Hamilton	1993-94	R G Shanks
1977-78	H W Gallagher	1994-95	P M Reilly
1978-79	D M Bell	1995-96	D R Hadden
1979-80	E J Miller	1996-97	R L Miller
1980-81	Margaret Haire	1997-98	J W Calderwood
1981-82	T T Fulton	1998-99	J R Hayes
1982-83	M F Russell	1999-2000	R W Stout
		2000-01	R J Harland

Fig 4. The informal presidential badge.

THE PRESIDENTIAL BADGE

The Presidential Badge (Figure 4) is worn with a blue ribbon. It consists of the crest of the Ulster Medical Society, a red hand on a white background imposed on a staff of Aesculapius in gold, surrounded by the name of the Society in gold on a blue background, with four tips of a red cross. The badge is not engraved and there is no indication of when it came into the possession of the Society.

Fig 5. A vice-presidential badge.

VICE-PRESIDENTIAL BADGES

Two badges to be worn by the Vice-Presidents were presented by the 1979-80 President of the Society, Dr Eddie Millar. These are similar to the Presidential Badge, although more modern in style, and consist of a red hand on a white background, against a background of the staff of Aesculapius with the twin serpents on a red cross (Figure 5). The Vice-Presidential badges are worn with maroon ribbons. There are two Vice-Presidents of the Society, each serving a term of two years, with one changing each year.

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APPENDIX

The History of the Ulster Medical Society[2] lists the officer-bearers from the beginning of the Society to 1967. Subsequent Presidents to date are shown in table 2. For completeness, the names of the other major office-bearers from 1967-2001 are listed here.

Honorary Secretary	
1966-67	J A Weaver/ D M Bell
1967-70	J A Weaver
1970-73	J M G Harley
1973-76	D R Hadden
1976-81	M E Scott
1981-85	P M Reilly
1985-88	D D Boyle
1988-93	J I Logan
1993-96	Carol M Wilson
1996-99	D R McCance
1999-	G McVeigh

Honorary Treasurer	
1966-68	W Bingham
1968-69	J C Hewitt
1969-75	N C Nevin
1975-78	J D Biggart
1978-83	R W Stout
1983-88	S A Hawkins
1988-92	M E Callendar
1992-97	M J J Gormley
1997-	Margaret E Graham

Honorary Editor	
-74	J E Morison
1974-83	J E Morison / D A D Montaomery
1983-95	D R Hadden
1995-	J M Gibson

Honorary Archivist	
1998-	J I Logan

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The Ulster Medical Journal, Volume 70, No. 1, pp. 40-46, May 2001.

In search of optimism

Annual Oration: Royal Victoria Hospital, Belfast, 5th October 2000

A Jennifer Adgey

It gives me great pleasure to welcome the new students to this first academic term in medicine.

As Sir William Osler – a very eminent physician once said – "To study the phenomenon of disease without books is to sail an uncharted sea, while to study books without patients is not to go to sea at all". There is no substitution for the study of a disease process than the observations made at the bedside of a sick patient. I thoroughly recommend your apprenticeship because without it you will never experience the challenge of diagnosis and treatment balanced with the tragedy for those in whom your help is limited. Despite technological advances, the key to a successful diagnosis still depends mainly on the physician's judgement based on experience and diagnostic techniques.

Cox in 1999 recognised the limitations of the scientific method in clinical decision-making.[1] The scientific method deals with linear cause and effect sequences, but decisions of both doctor and patient are not linear. The choices and factors have to be matched against numerical comparisons.

A Chinese philosopher once said "Human mind like a parachute, works best when open". With open minds medicine is a thinking, feeling and doing profession, where science, technology and art converge. A place where physicians are people as well as clinician-scientists; an area where doctors are challenged to think outside the medical model. Welcome to the patient world.

"In search of optimism" I have chosen for this, the 117th, oration.

In 1804 the world's population was one billion – 100 years later it was two billion. In 2004 it is estimated that it will exceed six billion and by 2025 eight billion. Currently global life expectancy is 66 years and this will continue to increase to probably 73 years in 2025.

GENERAL DISPENSARY

In 1792 the purpose of the General Dispensary which preceded the Royal Victoria Hospital indicated that the prevention of smallpox and the recovery of persons apparently dead were objects contemplated in the plan. (Prospectus of the general dispensary, Belfast Newsletter, 13 April 1792).[2] Yet in this city, 172 years later (1964) the first person apparently dead as a result of cardiac arrest (VF) was resuscitated. However as Belfast had a considerable amount of water around it the apparently dead was referring mainly to drowning – "the scheme for the recovery of persons apparently dead from suffocation, drowning and other causes, might also be usefully united with this institution." (Prospectus of the General Dispensary, Belfast Newsletter, 13 April 1792).[3]

ROYAL VICTORIA HOSPITAL

In 1896, 12 acres of land were set aside for the erection of the Royal Victoria Hospital and in 1903 the hospital opened at a cost of £300 per bed. The Rt Hon W J and Mrs Pirrie, the Lord and Lady Mayoress of Belfast contributed £100,000 to the building fund; this allowed the hospital to open free of debt. Philanthropy thus operated well in Victorian times. The Belfast Newsletter in 18 September 1903 paid tribute to the work of the old Royal Victoria Hospital on transfer to a new building – "Its path was never an easy one. It was often cramped by lack of funds, accommodation and medical appliances and yet it never wavered from its high ideal. It faced and conquered difficulties which were apparently insuperable and it could boast that no really deserving case was ever turned from its doors. It was not merely a public institution but was bound up inseparably with the history of the city. By the citizens it was regarded as something peculiarly their own, and one and all took a personal pride in its success". I think we could write the same for today's hospital. In the Nursing Committee minutes of 1902 we read "If Sisters are thought to be expensive they may be replaced by Staff Nurses

A J Adgey, Consultant Cardiologist, Royal Victoria Hospital, Belfast.

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but capable women are difficult to get in this country and it may be necessary to start the work with Sisters even if advisable to change afterwards". – This is not too dissimilar to what is happening today.

CARDIOLOGISTS

John E MacIlwaine was the first physician in the Royal Victoria Hospital to devote himself especially to the diseases of the heart. (Figure 1) He was first appointed in 1910 and delivered the oration one year later at the age of 37 years.

The electrocardiogram was first applied to human subjects by Professor A D Waller in 1887 and this had been established as a practical clinical method following the invention by Einthoven in 1903 of the string galvanometer. The first ECG machine was purchased for the Royal Victoria Hospital in 1913 and was first used by Dr J E MacIlwaine (Figure 2). It was obviously a very sizeable piece of equipment; both arms of the patient and one leg had to be immersed in saline. The Irish News, November 13th, 1913 announcing the introduction of the electrocardiograph into the Royal Victoria Hospital stated "Electrocardiography is in its infancy and no mind can foresee what the study of these electro pulsations may evolve". The ECG equipment had been bought by the generosity of Mr James Mackie of the Albert Foundry. The Royal Victoria Hospital was one of the first hospitals in the UK to have an ECG instrument. Indeed Sir Thomas Lewis working at University College Hospital in London, the doyen of ECG development, had only purchased an ECG machine in the preceding year.

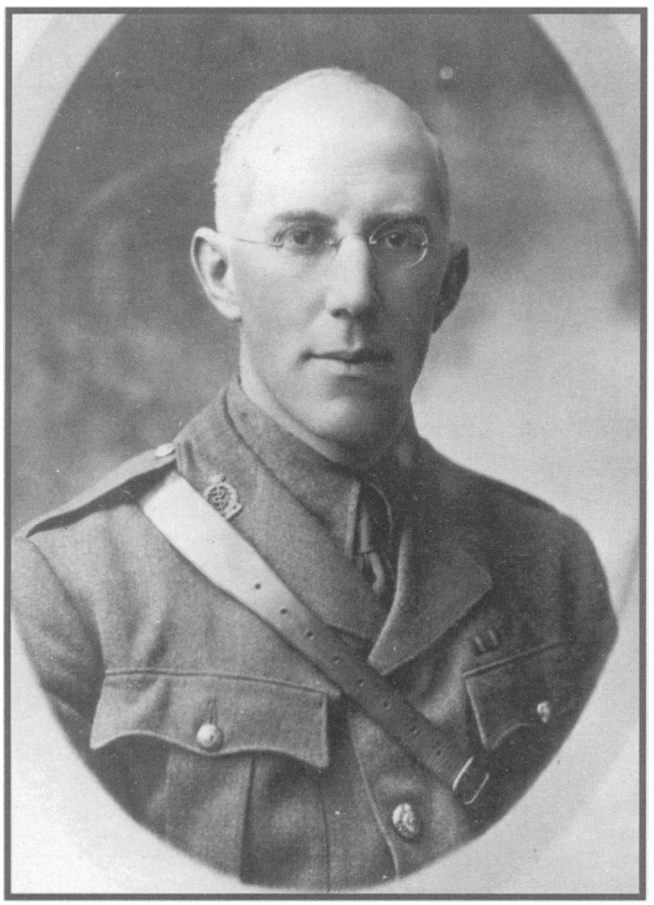

Fig 1. Dr J E MacIlwaine

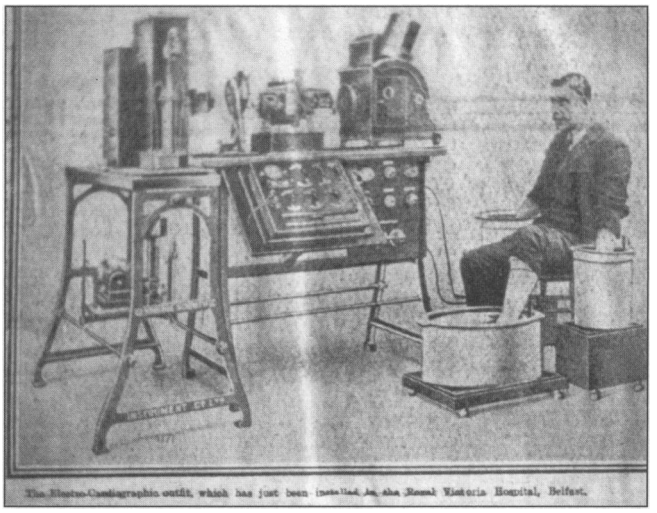

Fig 2. First ECG equipment in the Royal Victoria Hospital, Belfast.

Following the introduction of the ECG equipment in 1913, the annual report of the Royal Victoria Hospital stated that "this instrument has already more than justified its purchase and promises to revolutionise the diagnosis and treatment of heart affections".

In 1929 Dr J E MacIlwaine obtained permission to retire from his wards in order to devote himself entirely to cardiology. He was the first clinician to restrict his work to one medical speciality. During the 1914-1918 war, he had taken the hospital ECG apparatus to France for the investigation of cardiac conditions in soldiers. He became one of the original members of the Cardiac Club, now the British Cardiac Society.

Sir Henry H. Dale in 1950 said "I think that our successors, viewing the times in which we live from a long perspective in history, are likely to recognise the first half of the 20th Century as the period in which civilisation began to feel for good or ill the full impact of progress in the natural sciences. The richness of the 19th Century's closing decade in major discoveries of the kind from which science advances with a

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fresh impetus and in new directions seems to have caused a sudden acceleration of this process to begin about the turn of the century. In no department of knowledge and practice has this change since 1900 been more conspicuous or more rapidly progressive than in the general field of medicine." (Advances in Medicinal Therapeutics, Br Med J, Jan, 1950).

Dr Samuel Boyd Campbell was appointed in 1921 and he took over the ECG Department in 1930 following the tragic death of Dr MacIlwaine. In Figure 3 are seen Dr Campbell and Sr McMath with the very sizeable ECG equipment.

The next appointee to the Royal Victoria Hospital whose major interest was cardiology was Robert Marshall. He was appointed in 1924. His

Fig 3. Dr S B Campbell and Sr McMath and ECG Equipment.

application form for Consultant status is shown in Figure 4. It amounted to 2 pages which is a far cry from the present-day application.

After 1945 saw the break-up of general medicine into many sub-divisions, cardiologists were not classified as a separate group of physicians for many years. Indeed in 1948 there is a letter from Dr Marshall to Dr Sidney Allison, then the Honorary Secretary for Medical Staff, putting forward a proposal for the establishment of a Cardiological Department in the Royal Victoria Hospital.

The next appointees to the Royal Victoria Hospital with a major interest in cardiology were Howard Crozier, appointed initially in 1945, who continued mainly with the ECG Department and Professor Pantridge appointed in 1951. The first mitral valvotomy (1950) was carried out in Ireland by Mr T B Smiley who was then a Surgical

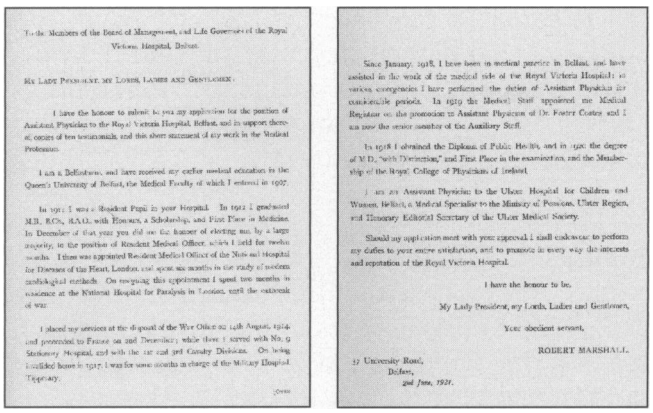

Fig 4. Dr Robert Marshall's Consultant Application.

Registrar, in conjunction with Professor J F Pantridge who at that time was a Medical Registrar – I wonder what the JCHMT would have to say about that today!

In 1950 the first cardiac catheterization was carried out in the Musgrave and Clark Clinic (right heart catheterization) by Professor J F Pantridge; in 1963 he introduced the DC Defibrillator for conversion of atrial fibrillation to sinus rhythm. As shown in Figure 5 it was a very large device which had to be placed on a trolley, otherwise it was not easily mobile. In 1964 the Coronary Care Unit was established in

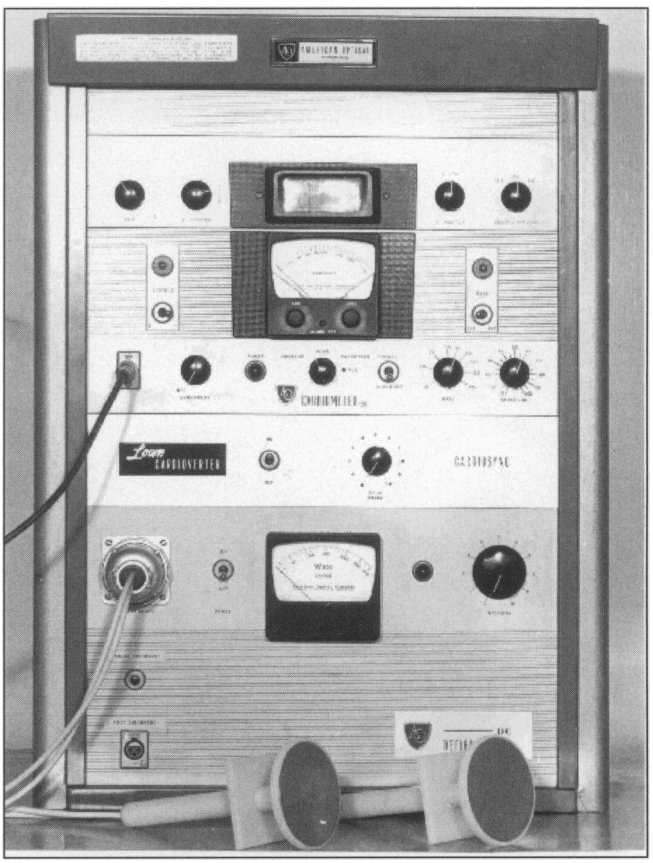

Fig 5. First DC defibrillator.

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the hospital by Professor Pantridge and Dr Geddes and in that year the first survival from ventricular fibrillation or cardiac arrest was recorded using DC defibrillation. Shortly afterwards, a cardiac arrest team centred in Wards 5/6 was established to attend those with sudden collapse in the hospital. All necessary equipment and drugs for correction of cardiac arrest were placed on a special trolley for rapid deployment to the patient.

In January 1966 the world's first pre-hospital or mobile coronary care unit was launched from the Royal Victoria Hospital by Professor Pantridge and Dr Geddes. One of the early mobile coronary care units pictured outside the Ambulance Depot is shown in Figure 6.

To complete the team of specialists in Cardiology, Dr Denis Boyle was appointed in 1965; he developed the Mobile Coronary Care concept in the Ulster Hospital, Dundonald. Dr George Patterson was appointed as a Consultant in 1969. His major development was in the cardiac catheterization laboratory. He carried out the first coronary angiogram in the Royal Victoria Hospital in 1970. Dr J S Geddes was appointed in 1971. In conjunction with Professor Pantridge he helped to establish the Coronary Care Unit, the resuscitation service in the hospital, and the Mobile Coronary Care Unit.

When I joined the Unit in 1965 cardiac catheterization was still in its infancy. Coronary arteriography was not undertaken and we were in the early stages of establishing open heart surgery. The plethora of cases in those days which filled the cardiac wards were patients with chronic rheumatic heart disease who needed valve replacement. The Mobile Coronary Care Unit had heralded the management of heart attacks outside hospital for the first time. Throughout the past three decades there has been a phenomenal development in techniques for studying heart structure in great detail and hence better overall management of cases.

Fig 6. Dr J S Geddes and Staff with one of the early mobile coronary care units.

BASIC, CLINICAL AND POPULATION BASED CARDIOLOGY

Man is a complex machine. Disease is a fault and it is the aim of medicine to repair or prevent it. This is the "mechanical model" of disease. It can at the end of the 20th century still be said to pervade scientific thinking in medicine.

The integration of discoveries between the bench and the bedside, the bridge building between basic, clinical and population based sciences with the developing collaborations with industry and governments have allowed us to take part in many of the world's multi centre trials in order to bring some of the best treatments which otherwise would not have been afforded here but eventually sets the scene for future proof of not only causation but function. To have dared to dream more than others have thought practical and care more than others thought wise, the choices have been many but have also involved travelling a well worn road and doing it better than others thought possible. Thus as we celebrate the new Millennium we are referring to the world as one civilisation.

From the age-adjusted death rates for coronary artery disease in 1996, it can be seen that Northern Ireland has still one of the highest incidences in the world for both sexes. At present in the UK some two million people suffer from angina and there are approximately 300,000 deaths from cardiovascular disease along with 380,000 hospital admissions per year. Thus in NHS costs and loss to the economy in terms of lost productivity, cardiovascular disease, particularly coronary artery disease plays a major role; although the rates for coronary artery disease are falling throughout the western world, they appear to be falling less rapidly in the UK compared to many of our European neighbours.

The Dartmouth Atlas of Cardiovascular Health Care looks at the distribution of cardiologists in the USA which ranges from 2.7 to 11.3 per 100,000 of the population. In Northern Ireland taking all who practise primarily cardiology and those with an interest in cardiology, the ratio is approximately 1.8/100,000 of the population –

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this does not reach the lower limit for the United States.

George Bernard Shaw in the preface to his play "The Doctor's Dilemma" wrote that all professions are a conspiracy against the laity. Medicine has become a very significant part of society and we are held to account. Peer review, guidelines, national standards, bureaucracy and budgets have severely restricted our activities made worse by society continuing to expect more with increased accountability and audit. The clamour of discontent – litigation, demands to cut waiting lists etc is many decibels louder than murmuring from patients directly. But unchanging will be their need for perseverance, courage, hope, trust, and the care of a sympathetic physician. In 1903 Sir William Osler stated that "Half of us are blind, few of us feel and we are all deaf". Nevertheless, from the achievements in surgery and cardiology many diseases have been treated effectively or prevented with improved prognosis.

From the estimated US population in 2025, those aged 65 or greater will be 18.3%, with increasing millions suffering heart disease. In the absence of a cure, the patient hopes for a caring doctor. Our working week is taken-up with doing, and thus to find the time to reflect on what we do is difficult but it behoves us to remember that the patient's independence is lost during illness and therefore comfort and care are major factors in our work. Sir Theodore Fox, one of the famous editors of the Lancet wrote that "Lack of time made us all bad doctors".

Medicine should not be considered a commodity for the marketplace. Yet the total expenditure on healthcare keeps appearing as a proportion of the GDP. In 1998 at 6.9% we are still lagging behind other European countries (eg France, Germany) Canada, United States and even New Zealand. (8%) I have seen the system for financing and delivering medical care undergo a series of cataclysmic upheavals and re-alignments. I have also seen the painful evolution of the doctor/ patient relationship from a largely private affair to one embedded in the "healthcare industry" (a term and concept unheard of in 1979). With all the new managerial roles that a doctor is being asked to cover, I doubt if patients would want to be treated by a health economist than by a practising physician. At times I feel the gridlock of bureaucratic resistance where a great deal of ingenious effort by a few disappears into the black hole of bureaucracy in which oases of excellence risk being overwhelmed by a desert of indifference and mistrust. I feel at times that we are expected to try an unprecedented triple somersault full twist, quite against the laws of nature. Despite all the clinical governance, peer review and guidelines I think it is important to remember Darwin's inversion which suggests that varieties of excellence, worth and purpose can emerge, bubbling up out of "mindless purposeless forces".

It was David Pyke,[4] who as ex-registrar of the Royal College of Physicians of London, in reviewing a 100 years of clinical research in Britain described the many routes to significant discoveries "they come from minds prepared and from minds unprepared which by observing what others had seen but not noticed, turned weakness into strength." Good research is about the chase of ideas. Individuals with good clinical ideas are a neglected group.

Although over the past three decades there have been significant improvements in our diagnostic and therapeutic capabilities, we can very rarely cure our patients but are frequently able to improve the quality and length of life.

In this century we hope to see a gradual replacement of palliative treatment by curative therapy, and ultimately a marked reduction and disappearance of cardiovascular diseases. Following the completion of the map of the human genome it will take some time before genetic therapy has a major effect on our therapeutic armamentarium and even longer before we have an animal with our own genetic footprint able to replace our organs when needed.

In the coming two decades we will still have to concentrate on improving our diagnostic techniques along with therapy in order to increase palliation whilst accepting the new information at the cellular and genetic level. Our basic non-invasive tests in order for the diagnosis of cardiac disease – echocardiography, nuclear magnetic resonance imaging, positron emission tomography, electron beam tomography – will undergo further improvement. More catheter-based interventions will be developed. With control of thrombotic and vascular processes in the vessel wall, and the development of new blood vessels (angiogenesis) plus the introduction of new cardiomyocytes into the failing heart we

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may succeed in prolonging life. When rejection is controlled and infection prevented cardiac xenotransplantation will become possible. The problem of sudden death outside hospital will lead to the wide application of devices able to detect and correct life threatening arrhythmias.

In the future the major challenge for cardiologists will be to take the knowledge both of basic and clinical aspects of cardiology and apply these clinically. As we discover more and more with regard to function and disease process, brings us to another paradox – the more we know the more we need to learn.

Rabbi Julia Neuberger,[5] Chancellor of the University of Ulster, once compared the NHS to a theological institution and indicated that the NHS seemed to be becoming less of a church and more of a garage giving consumers (not citizens) whatever care they can get within the limits of medical science. She felt that the NHS of the future will have to adjust to a new relationship with society and equated doctors to priests of the theological institution, and that the quality of care provided must fit those concepts of compassion and fairness written into the original National Health Service of 1948. She felt that individual temples that are unwelcoming must be reformed. Priests who misbehave must be disciplined, new ways of praying – at easy access, walk-in services that complement the continuing relationship with a priest – must be welcomed. The belief is in universal availability – no one wants to wait two weeks to pray!!

As she points out in modern Britain, the faith may appear in new ways – from the evangelical (otherwise known as NHS direct) to the fundamentalist which includes integrated care, holistic approaches and complementary medicine. She believed that faith remains the key to good health. She points out that recognising that the NHS is a creature of faith, an institution in which people have faith, is essential.

One doctor [6] indicated that "it is all very well congregations are losing their faith but priests have pride too. Many are fed up with trying to provide ever increasing miracles to an avaricious public while their church and "god" do not provide them with the resources. The health service is its staff. Increasingly staff see little reason to continue believing in their church. Many priests and servers might prefer to work in a garage where duties are clearly understood and adequate resources are available to do the job properly."

For practitioners the difficulties of keeping up with the sheer volume of the medical literature are immense. George Lundberg [7] has estimated that 2 million biomedical articles are published every year and that to keep up with all this work a diligent reader has two options – namely to read two articles a day, knowing that within a year he/she will be 60 centuries behind, or to read 6000 articles per day. Eighty to ninety percent of all scientists who have ever lived are alive today;[7] the medical published record doubles every 12 – 15 years. Thus Index Medicus, published in annual volumes maintained a sylph-like figure of 2 kg per year from its foundation in 1879 until the late 1940s. By the end of the 1970s it weighed an unhealthy 14 kg, despite being softbound and being printed on thinner paper, in a smaller font and with narrower margins. Its electronic counterpart, Medline, currently abstracts nearly 4,000 journals and adds over 400,000 references to its database each year.

Sir William Osler once said "a rare and precious gift is the art of detachment by which a man may so separate himself from a life-long environment as to take a panoramic view of the conditions under which he has lived and moved: it frees him from Plato's den long enough to see the realities as they are, the shadows as they appear. Could a Physician attain to such an art he would find in the state of his profession a theme calling as well for the exercise of the highest faculties of description and imagination as for the deepest philosophic insight". A pattern-recognition school of thought sums up our medical training and our everyday practice. William Blake's Newton (1795)(Tate Gallery, London) thought that the whole of life could be measured by dividers but this is a very simplistic view.

Oliver Wendell Holmes once said "When I want to understand what is happening today or try to decide what will happen tomorrow, I look back." Within medicine there is an enormous talent and genuine commitment to the NHS. How much better the service could be if the fear of failure and of sticking your head above the parapet were to be swept away and intelligence, truth and humanity allowed to develop. Aldous Huxley said that fear "casts out intelligence, casts out goodness, casts out all thought of truth . . . in the end fear casts out even a man's humanity".

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And now for the future. The Human Genome Project has provided the scientific community with a map of the entire collection of 100,000 or more human genes and 3 billion letters of DNA encoding these genes. The function of these genes and how they are regulated in health and disease still has to be worked out. Defining diseases by their biochemical mechanisms instead of their pathogenesis is going to alter the way physicians diagnose conditions and prescribe medication.

By the end of this decade predictive genetic screening will probably be coupled to the taking of a history and physical examination. Physicians may be able therefore to move away from what has generally been "one-size-fits-all" prevention efforts and move towards the patients' specific genetic risks. Genetic screening will become less expensive with the emergence of DNA microchip-array technology. This would be "nouvelle medicine" that replaces our halfway technology.

Physicians will be required to understand the kinds of tests available, which test is most applicable, with the interpretation and implementation of the results. They will need to recommend tests to patients, to explain the possible advantages and the limitations of these tests and to interpret the results in lay terms. This kind of information cannot be imparted in the usual 10 minute outpatient visit or a cursory telephone call. Informing patients of a genetic risk will require significant sensitivity and understanding.

Education of the public about genetic screening will undoubtedly take place on the internet and physicians must help to drive this. But with this knowledge goes confidentiality. When you are able to define an individual's genetic make-up and make reliable predictions, this is enormously powerful information.

It is awesome now to contemplate how the next wave of progress in this field will influence all aspects of life in the new millennium.

CONCLUSIONS

The writer E M Forster once said "How do I know what I feel, until I've seen what I've said?". And so I too needed to write these words in order to articulate my feelings. Medicine is one of the few spheres of human activity in which the purposes are unambiguously altruistic – in itself, a remarkable achievement.

History shows goodness follows badness follows goodness. The pessimist turns that around and concludes the end is nigh. This negative often outsells the positive. But the ace in this circular game is so far still held by the optimists – for all the centuries of naysaying and nighsaying, the sun still rises. There is a crack in everything. That's how the light gets in.

When we look at the young men and women who are thronging about us, we feel that "close on our heels a fresh perfection treads, born of us and fated to excel us". Our faith in the future surges up again and the torch gleams.[8]

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The Ulster Medical Journal, Volume 70, No. 1, pp. 47-50, May 2001.

Case Report

Familial craniosynostosis due to Pro250Arg mutation in the Fibroblast Growth Factor Receptor 3 gene

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Accepted 10 January 2001

The craniosynostoses, the premature closure of the cranial sutures, are a common heterogeneous group of disorders, affecting about 1 in 2000 children at birth. About 20% have a distinct syndrome defined on clinical and family grounds. The delineation of these syndromes has become more precise with molecular analysis. Mutations in the fibroblast growth factor receptor 1, 2, 3 loci have been identified in craniosynostosis syndromes such as Pfeiffer, Apert, Crouzon, Crouzon with acanthosis nigricans, Jackson-Weiss and Beare-Stevenson. The remainder is isolated non-syndromic craniosynostosis. Recently, a mutation in fibroblast growth factor receptor 3 (FGFR3) gene at chromosome 4 (4p16) has been described in individuals with a variable picture of craniosynostosis.[1] The mutation is at C749G predicting a proline 250 arginine aminoacid substitution in the extracellular domain between the second and third immunoglobulin-like loops. The clinical phenotype of this mutation is variable, involving unilateral and bilateral craniosynostosis. Most affected individuals have normal-appearing hands and feet, but on radiological investigations may have short, broad middle phalanges of the fingers, absent or hypoplastic middle phalanges of the toes, carpal and tarsal fusion and cone-shaped epiphyses. Some authors have described mental retardation, apparently unrelated to the management of the craniosynostosis.[2] This communication describes a family with the FGFR3 Pro250Arg mutation, in which the clinical phenotype encompasses craniosynostosis, macrocephaly, deafness, delayed development, short fingers and toes and proptosis of the eyes.

CASE REPORT A 10 month old girl (Fig.1: V-1) with an "odd-shaped" skull was referred by her

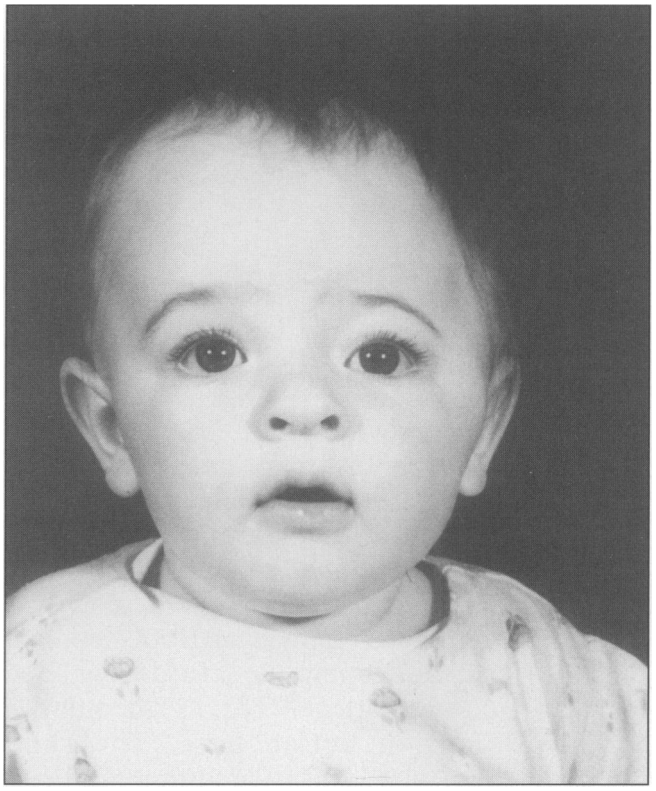

Fig 1. Patient age 10 months showing flattening of left forehead, prominence of left side of face, and left palpebral fissure lower than the right.

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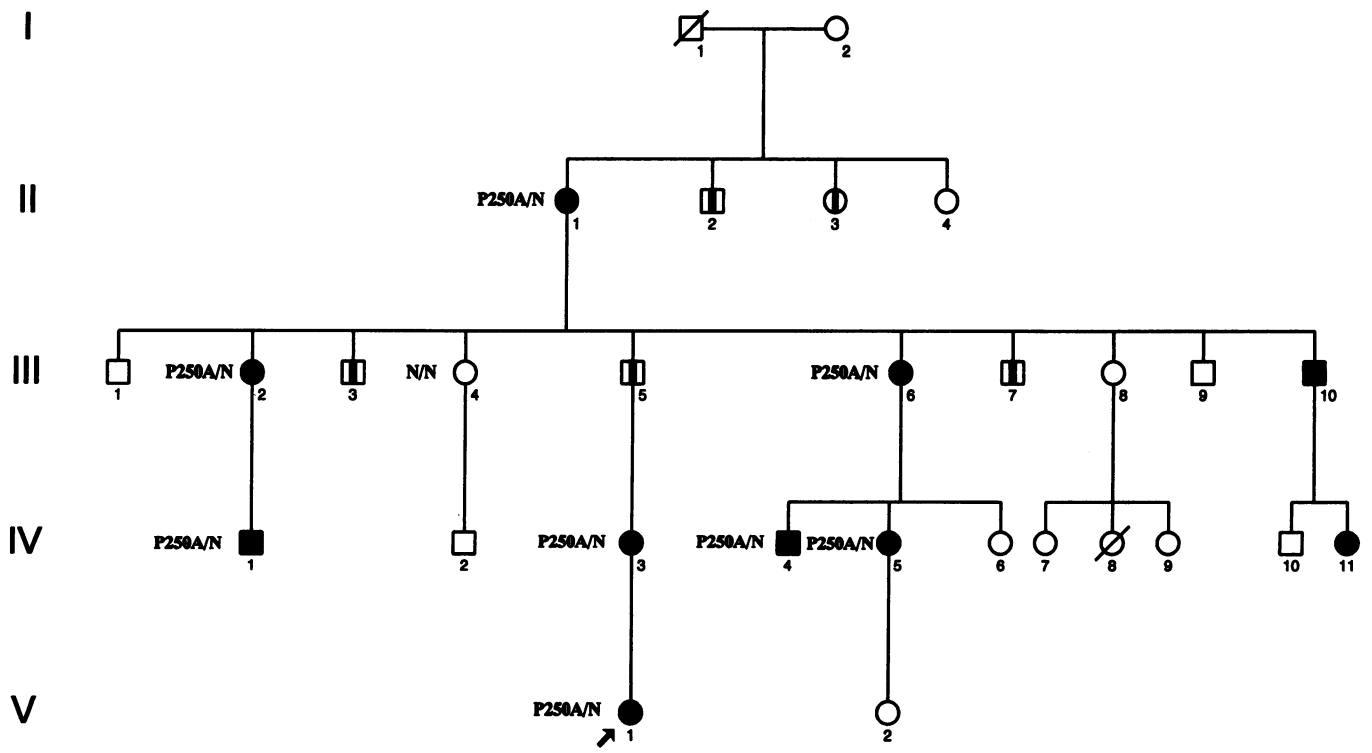

Fig 2. Pedigree of the family (□○ = unaffected male and female; ■● = affected; ◫◑ = not examined but reported affected; P250A = mutant allele; N = normal allele).

family practitioner to the Medical Genetics Clinic because of a family history of craniosynostosis. The mother had noted that her left eye was below the level of that on the right, that the left forehead was flattened and that the left lateral side of the face was more prominent. There were no concerns about hearing or her growth and development patterns. On examination, the left forehead and temple region was flattened (Fig. 1). The anterior fontanelle was of normal size. The skull circumference was 47 cms (91 centile). The tip of the nose was hooked with scalloping of the alae nasi. Fundoscopy was normal, in particular there was no papilloedema. The fingers and toes were short. The x-ray of the skull showed a left coronal synostosis. The anterior-posterior length of the skull is increased. The x-ray of the hands showed shortening of the middle phalanges and to a lesser extent the proximal phalanges of the fingers and thumb. The 4th and 5th metacarpals are short. The changes are symmetrical. The x-rays of the feet showed similar changes to those seen in the hands. There is brachyphalangy with the middle phalanges being most affected. The changes in the distal phalanges are more marked in the feet than the hands. The changes are symmetrical. The patient's mother (IV-3) had no abnormal features apart from slightly proptosed eyes and a similar shaped nose to that of her daughter. The skull circumference was just below 98 centile. Fingers and toes were short. X-ray of her skull showed no craniosynostosis but the x-ray of her hands and feet were similar to those of her daughter.

FAMILY HISTORY

Two relatives had craniosynostoses requiring neurosurgical intervention. The patient's mother (IV-3) had two paternal first cousins, a male (IV-1) with a right coronal synostosis and small auricular appendages and a female (IV-11) with bilateral coronal synostosis, mid-facial hypoplasia and choanal atresia. The Table shows the clinical features of individuals in the family.

MOLECULAR INVESTIGATIONS

Peripheral blood samples were available from nine relatives. The DNA was extracted from lymphocytes and screened using PCR for the FGFR3 Pro250Arg mutation. The eight affected relatives tested proved positive for the mutation. One relative (III,4) who developed breast cancer at age 40 years was negative for the mutation.

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TABLE

Clinical Features of affected individuals in the family

RELATIVE (Pedigree ref)	*CLINICAL FEATURE*	*FGFR3 Pro250Arg mutation*
II-1	Mid facial hypoplasia, scoliosis thoracic spine	+
II-2	Facial asymmetry, learning disability	NE
II-3	Facial asymmetry, breast cancer (onset early 40s)	NE
II-4	Breast cancer (onset early 60s)	NE
III-2	Facial asymmetry, flattened R. forehead	+
III-3	Learning disability, deafness	+
III-4	Breast cancer (onset age 40)	–
III-5	Macrocephaly, proptosis	NE
III-6	Macrocephaly, mild proptosis, nerve deafness, short fingers	+
III-7	Macrocephaly, learning disability	NE
III-10	Depressed nasal bridge, treated surgically	NE
IV-I	R. coronal synostosis, small accessory auricles, treated neurosurgically	+
IV-3	Mild proptosis, hook shaped tip of nose, short fingers and toes	+
IV-4	Macrocephaly, sensor – neural deafness	+
IV-5	Macrocephaly, short fingers	+
IV-I I	Fusion coronal sutures, mid-face hypoplasia, hypertelorism, choanal atresia	NE
V-I	L. coronal synostosis, short fingers and toes	NE

+ = mutation present; – = mutation absent; NE = not examined

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DISCUSSION

The phenotype of individuals in our family with a FGFR3 Pro250Arg mutation showed wide variability of clinical features (Table). Some individuals had craniosynostosis requiring neurosurgical interventions, whereas others only had minor features such as mild ocular proptosis. It is important to recognise the spectrum of clinical variability as the mutation is transmitted in an autosomal dominant manner.

Mutational analysis of patients with craniosynostosis is not only essential for diagnosis but also is a prerequisite for providing accurate genetic counselling. An individual with the FGFR3 Pro250Arg mutation with a mild clinical expression has a 50% risk of transmitting the gene mutation to children, who may be more severely affected. One report describes a normal individual with FGFR3 Pro250Arg mutation who had a child with bilateral coronal craniosynostosis.[3] Thus with reduced penetrance in some individuals not manifesting the disorder, it is important that both parents of patients with isolated sporadic craniosynostosis due to a mutation should be tested regardless of their clinical status. Any individual with neurosurgical repair of craniosynostosis contemplating having children also should be tested for the mutation in order to estimate the risk.

Mental retardation unrelated to the craniosynostosis has been described with this condition.[2] Deafness has been reported in 33% of affected individuals.[4] The explanation for the learning disability and the hearing loss is unclear. Individuals with this mutation often have macrocephaly.

In our family, four individuals had macrocephaly. Most affected individuals had normal hands and feet on clinical examination but show on x-ray short or broad middle phalanges of fingers and absent or hypoplastic middle phalanges of the toes.[4] Cone shaped epiphyses are also characteristic. The typical appearance of the hands and feet, even in the absence of craniosynostosis should suggest investigation of FGFR3 Pro250Arg mutation. Our patient and her mother had typical finger and toe anomalies. It has been suggested that this syndrome should be designated 'coronal craniosynostosis with brachydactyly and carpal/tarsal coalition due to Pro250Arg mutation in the FGFR3 gene'.[4] The mutation rate at C749G nucleotide which leads to the Pro250Arg change is one of the highest mutation rates in the human genome and makes this an extremely common form of craniosynostosis.[5]

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The Ulster Medical Journal, Volume 70, No. 1, pp. 51-53, May 2001.

Case Report

Endovascular repair for acute symptomatic and ruptured abdominal aortic aneurysms

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Accepted 7 April 2001

Since the introduction of endovascular repair of abdominal aortic aneurysm, an increasing proportion of elective asymptomatic patients have been found to be suitable for endovascular repair despite uncertainties over long-term durability and cost benefit. However, one group of patients whose recovery would most likely be improved and in whom postoperative hospital stay would be reduced by this technique are those who present with an acutely symptomatic or leaking abdominal aortic aneurysm. Conventional urgent and emergency open repair of these patients are associated with significant mortality rates of approximately 20% and 50% respectively.[1] Those who survive may have a protracted recovery, requiring prolonged management in an intensive care unit.

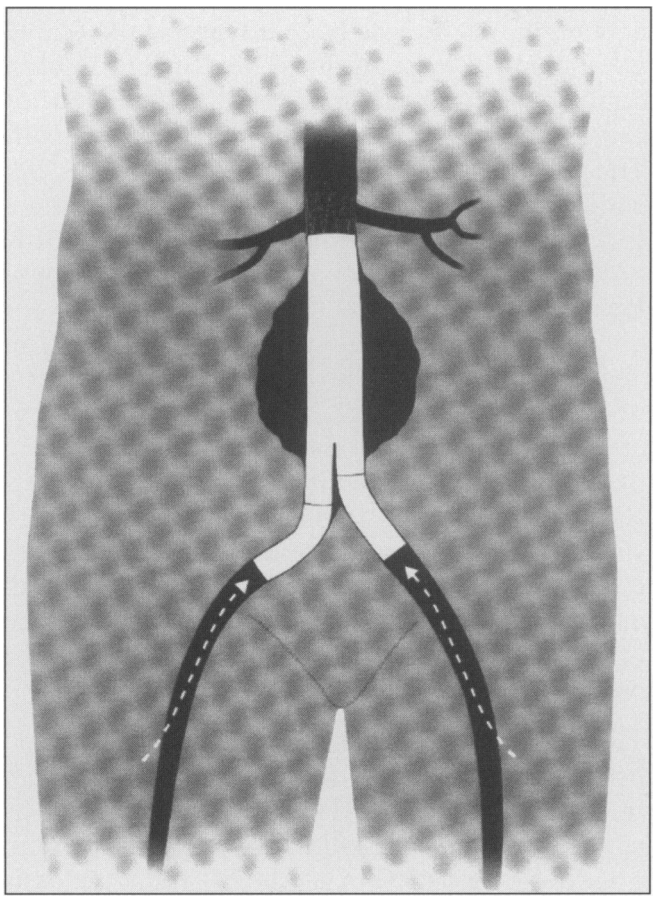

Fig 1. Diagrammatic representation of insertion of stent graft (Zenith device (William Cook Europe A/S, Bjaeverskov, Denmark)).

The endovascular technique avoids an abdominal incision and involves the intraluminal placement of stem-grafts via surgically exposed femoral arteries. The graft, which is preloaded in a plastic sheath, is advanced over guidewires up the femoral arteries until it is immediately distal to the lower of the two renal arteries. Once in position, the graft is released from the sheath and extensions into the iliac arteries are then attached to the main body of the graft through additional delivery sheaths (Figure 1). The stem-grafts are self-expanding and will fill to exclude the aneurysm.

Here we report three patients, who presented with acute symptomatic infrarenal abdominal aortic aneurysms, one of which was confirmed to have a contained leak on CT scan.

CASE REPORT The first patient was a 76 year old man who presented to a district general hospital with pain in the left flank radiating to the groin. He had a documented history of valvular and ischaemic heart disease, atrial fibrillation

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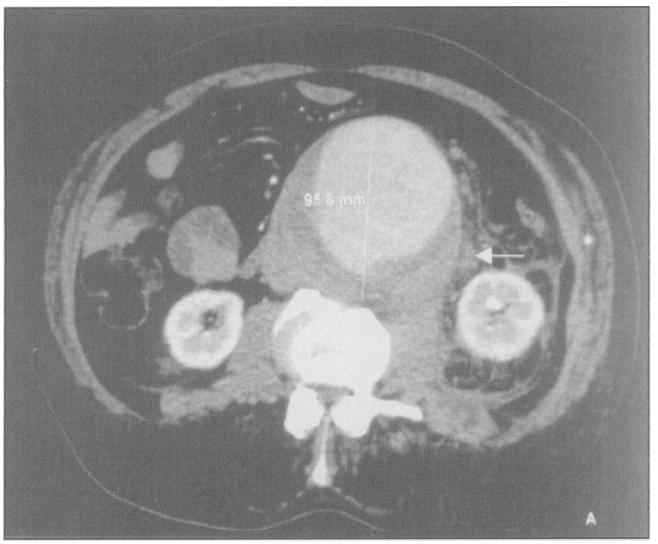

Fig 2. CT scan abdomen showing abdominal aortic aneurysm with contained leak posteriorly (Arrow showing site of contained leak).

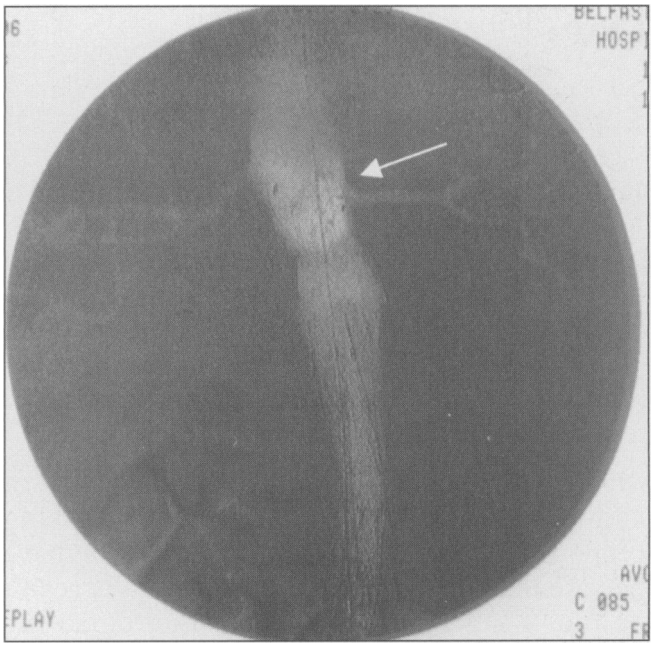

Fig 3. Completion angiogram showing exclusion of the aneurysm sac by stent graft (arrow showing Renal arteries).

and left ventricular failure. On examination, he was found to have a pulsatile mass in the epigastrium. This was confirmed on both ultrasound and CT scan to be a leaking infrarenal abdominal aortic aneurysm measuring 7.5 cm (Figure 2). He was haemodynamically stable and evaluation was made following transfer for possible endovascular repair. Measurements from CT scans were used to select a suitable off-the-shelf stem-graft (Talent, Kimal plc, London, UK). Two hours after arrival to the Belfast City hospital, the patient was anaesthetised in theatre, and femoral arteries exposed. This facilitated the potential use of an intra aortic occlusion device, which in this case was not required. A bifurcated system was used and deployed through the femoral arteries. Total anaesthetic time was $4^1/_2$ hours and blood loss was estimated to be 1700 ml. On-table completion angiography confirmed exclusion of the aneurysm by the stem-graft with no evidence of an endoleak (Figure 3). Post-operatively the patient was transferred to the intensive care unit where he stayed for approximately 12 hours. He was allowed to eat and drink on the first post-operative day and was transferred to the vascular ward where he commenced mobilisation on the second post-operative day. His discharge from hospital was delayed to three weeks because of social circumstances, although it was assessed that he was fit for discharge on the seventh postoperative day.

The second patient was an 82 year old man who was known to have an infrarenal abdominal aortic aneurysm. He presented with pain in the abdomen radiating into his back and tenderness over the aneurysm. As he was haemodynamically stable an urgent CT scan was performed which confirmed a 7 cm aneurysm without evidence of rupture that was amenable to endovascular repair. The patient was haemodynamically stable and, endovascular repair was carried out 48 hours following his admission to hospital. On this occasion, a bifurcated device (Zenith, Cook, Bjaererskov, Denmark), which was pre-made for another patient, was utilised. Completion angiography revealed no endoleak and exclusion of the aneurysm sac. Post-operatively his epigastric pain resolved and he was cared for in the high dependency unit for less than 24 hours, after which he was transferred to the vascular surgical ward to resume oral diet. He developed chest infection, which rapidly resolved with antibiotics. Check CT scan revealed no evidence of endoleak and the patient was discharged home on the twelfth post-operative day.

The third patient was an 83 year old man who had been under outpatient surveillance for a 4.6 cm infrarenal abdominal aortic aneurysm. He was admitted to the ward following presentation with a sudden onset of pain in his left flank and was found to be tender over the aneurysm. CT scan revealed an infrarenal aneurysm with a 'thin irregular' posterior wall. As no leak was evident

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on the scan, investigations to exclude other possible cause for his pain were undertaken. A barium enema and oesophagogastroduodenoscopy were performed and found to be normal. Again the CT scan showed the aneurysm to be anatomically suitable for endovascular repair. An off-the-shelf commercially available bifurcated stem-graft (Talent, Kimal, London, UK) was used to exclude the aneurysm. The operation itself was uneventful, but postoperatively, he suffered a left hemispheric stroke. He made good early recovery with minimal residual weakness. Fifteen days following the stroke the patient developed several episodes of melaena. Endoscopy revealed a bleeding duodenal ulcer, which responded to injection sclerotherapy and triple therapy for Helicobacter pylori. He made a slow recovery but was eventually well enough to be discharged to a rehabilitation unit on the twenty-eighth post operative day following a check CT scan which showed no endoleak.

DISCUSSION

The morbidity and mortality of ruptured and symptomatic abdominal aortic aneurysm repair remains high. Many of the survivors require prolonged periods of recovery in the Intensive Care unit, often needing a protracted period of rehabilitation prior to achieving independence and discharge from hospital. Endovascular repair of abdominal aortic aneurysms has already been well described for the repair of asymptomatic elective cases. The technique is certainly feasible, but as yet the available data have not shown significant survival advantage over conventional open repair for elective cases.[2,3] The emergency and urgent repair of abdominal aneurysm is however, still associated with a high perioperative mortality of up to 60%. It is anticipated that this is the area where an endovascular technique could significantly reduce mortality if it were technically and logistically possible. Some preliminary studies from pioneering units have suggested feasibility, albeit with substantial logistical support.

Our report provides further evidence that endovascular repair for patients with acute symptomatic or stable leaking abdominal aortic aneurysm is possible, utilising commercially available 'off the shelf' devices. In all three patients, postoperative ICU stay was short and hospital recovery time relatively low in comparison to the conventional open technique. However, this report must be treated with caution. These patients deemed suitable for the endovascular repair, were haemodynamically stable and would tend to have a better prognosis than unstable patients, even with open repair. Hardman *et al*[4] demonstrated that the mortality rate with open repair in patients who are haemodynamically stable was approximately 20%. Therefore, although savings in ICU stay may be incurred by endovascular repair of acute abdominal aortic aneurysm, the potential reduction of morbidity and mortality has yet to be proven. This may only be determined by a well conducted multicentre randomised study.

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The Ulster Medical Journal, Volume 70, No. 1, pp. 54-55, May 2001.

Case Report

Gastric variceal haemorrhage successfully managed by splenectomy – a case report and literature review

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Accepted

INTRODUCTION

Isolated gastric varices secondary to left-sided portal hypertension are a rare but important cause of upper gastrointestinal blood loss. Diagnosis is a challenging problem; however appropriate treatment should prevent further bleeding. A case is reported and the recent literature reviewed.

CASE REPORT A 57 year old lady presented following upper gastrointestinal bleeding. She had a past medical history of retroperitoneal fibrosis and had incidentally been noted to have a cyst at the splenic hilum associated with splenomegaly. Gastroscopy revealed altered blood in the lumen of the stomach but no bleeding source. Following a rebleed the lady underwent laparotomy. Prominent veins around the gastric fundus and splenomegaly were noted. An anterior gastrotomy revealed two large bleeding gastric varices which were oversewn. A total of 30 units of blood had been transfused to this point. On the tenth post operative day she had a further significant bleed and was transferred to our unit for further management. A second laparotomy was performed and the previously noted gastric fundal venous dilatation and splenomegaly were confirmed. A cystic inflammatory mass was identified in the tail of the pancreas while the liver appeared grossly normal. The pancreatic cyst was deroofed and a splenectomy performed. Histology of the cyst revealed chronic inflammation and no evidence of tumour. She has remained well without further bleeding.

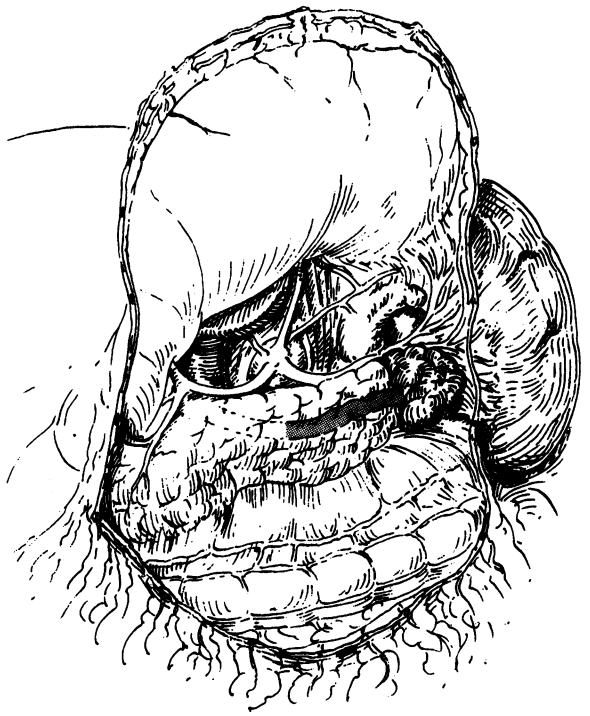

Figure Diagrammatic representation of operative findings demonstrating a 6 cm inflammatory mass in the pancreatic tail with gastric vein dilatation.

DISCUSSION

Sinistral or left-sided portal hypertension is a localised form of extrahepatic portal hypertension that develops after splenic vein thrombosis and may result in gastric varices. Anatomically the splenic vein is vulnerable to disease affecting the pancreas. Conditions associated with splenic vein thrombosis therefore include acute or chronic pancreatitis, pancreatic pseudocysts and pancreatic neoplasms.[1-4] It is felt that this lady had a pancreatic pseudocyst secondary to subclinical pancreatitis. However retroperitoneal disease is also an aetiological factor.

Following splenic vein thrombosis collateral circulation develops via the short gastric,

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gastroepiploic and left gastric veins (see figure). The resulting venous hypertension leads to the formation of gastric varices along the greater curvature and fundus of the stomach.[5]

Presentation is typically with G.I. blood loss either as anaemia or haematemesis and melaena. Abdominal pain is also common with splenomegaly documented in 60-70% of cases.[1,2,6] Isolated gastric varices with splenomegaly, a history of pancreatic disease and absence of liver disease is almost pathognomic of sinistral portal hypertension. The incidence of haemorrhage from gastric varices ranges from 10-70%.[7] In general, gastric varices bleed less frequently than oesophageal varices, however haemorrhage is often of a more life-threatening nature.[8-10]

Diagnosis of gastric varices at gastroscopy or on barium studies is often difficult with a reported accuracy of 14-74%. The gold standard for identifying the thrombosed splenic vein is mesenteric angiography with venous phase imaging but ultrasound and CT may also be usefull.[11]

Treatment of gastric varices is difficult as they are poorly controlled by balloon tamponade and there is a high incidence of complications with injection sclerotherapy. The treatment of choice is splenectomy as first described by Greenwald in 1939.[12]

This procedure decompresses the short gastric vessels by halting inflow from the splenic circulation. In the emergency situation gastric varices may also be oversewn.[13,14] It is now advocated that patients without prior haemorrhage or significant bleeding should initially be observed, as splenectomy may be associated with significant morbidity and mortality.[15] Additionally there are a number of cases of spontaneous resolution probably due to recanalization of the vein.[9]

In summary, sinistral portal hypertension and isolated splenic vein thrombosis are a rare but important cause of gastric variceal bleeding. This can be very successfully treated by splenectomy.

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The Ulster Medical Journal, Volume 70, No. 1, pp. 56-58, May 2001.

Case Report

Cronkhite-Canada Syndrome

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Accepted 20 November 2000

Cronkhite-Canada syndrome is one of the rare causes of multiple polyposis, characterised by generalised gastrointestinal polyposis, cutaneous hyperpigmentation, alopecia, and nail dystrophy.[1] Although Cronkhite and Canada described it for the first time in 1955, little is known about its aetiology and the prognosis remains poor. We describe a case of Cronkhite-Canada syndrome in a 79-year-old Japanese man, which illustrates the typical features of the condition.

CASE REPORT A 79 year old man presented with a history of weight loss of ten kilogrlms over a period of 2 months, associated with diminished taste and watery diarrhoea (4-5 times per day). He had pigmentation of the skin, especially on the hands, as well as dystrophic nail changes and alopecia with loss of8scalp and facial hair.

Barium enema showed an unusual polypoid appearance throughout the colon with relative sparing of the ascending colon and caecum (figure 1). Colonoscopy revealed numerous polypoid lesions of the colonic and rectal mucosa. Gastroscopy revealed multiple polyps in the stomach and duodenum similar in appearance to those seen in colon and rectum. A small bowel series showed multiple polyps throughout the small bowel.

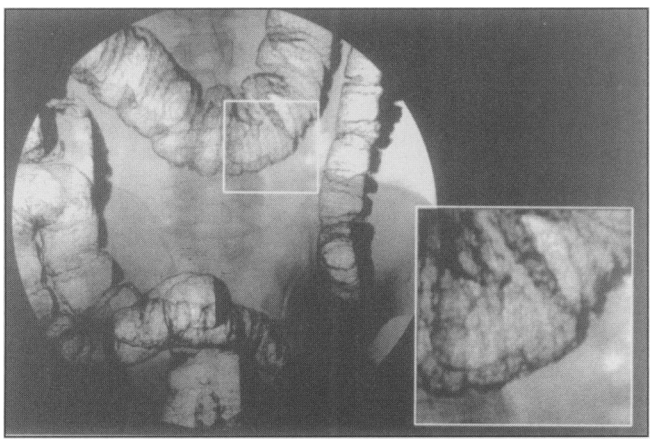

Fig 1. Barium enema showing multiple polyps throughout the colon.

Biopsies taken from stomach and colon showed small hamartomatous mucosal polyps. In both sites the mucosa was oedematous and showed cystic dilatation of foveolae in the stomach and of mucosal glands in the colon (figure 2). The cysts were lined by normal or flattened epithelial and mucus secreting cells and there was no evidence of dysplasia or malignancy in either site. The histological features were similar to those of juvenile polyps although eosinophils were less prominent in the stroma.

The histological features, the wide distribution of the polyps together with the skin changes described above pointed to the diagnosis of Cronkbite-Canada syndrome.

Nutritional support was instituted with enteral feeding both orally and through a nasogastric tube. Despite this, his serum albumin continued to fall. From a level of 28 gm/l on admission this fell to 23 gm/l in 3 weeks and further fell to 15 gm/l within the next six weeks. In parallel with this he developed gross peripheral oedema and his general health rapidly deteriorated. He died two and half months after presentation.

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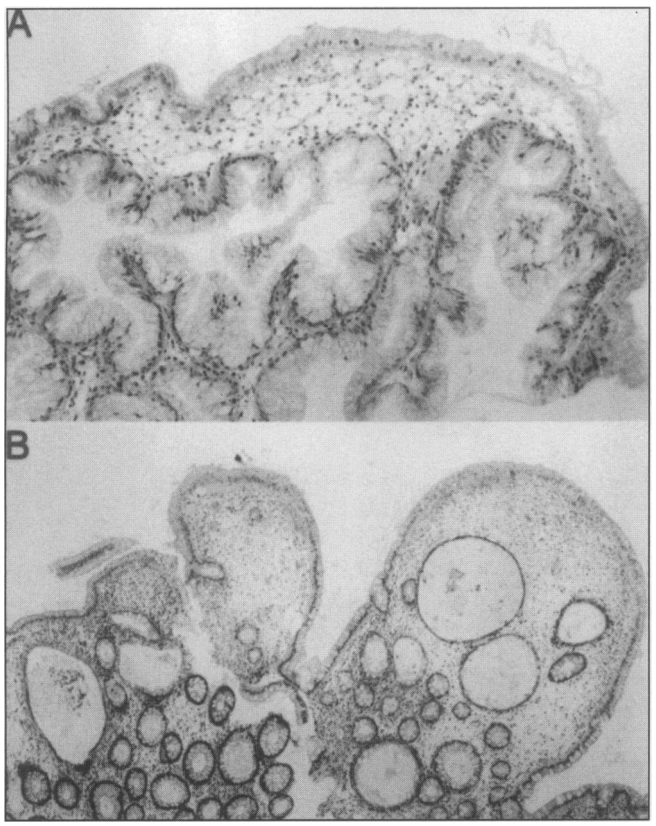

Fig 2. Oedematous mucosa with cystic dilatation of the gastric foveolae (A) and mucosal glands in the colon (B).

DISCUSSION

Cronkhite and Canada first described a syndrome of generalised gastrointestinal polyposis associated with cutaneous pigmentation in 1955.[1] The cardinal features of this syndrome are: (1) the presence of hamartomatous polyps of the juvenile (retention) type throughout the stomach and intestines, (2) ectodermal changes consisting of alopecia, onychodystrophy and hyperpigmentation, (3) the absence of family history, (4) adult onset, (5) the eventual development of diarrhoea and weight loss. The coexistence of all these features separates Cronkhite-Canada syndrome from other polyposis syndromes,[2] however, multiple regenerative polyps localised to the stomach, ascending colon and transverse colon, has been described by Hanzawa et al.[3]

Although Cronkhite-Canada syndrome and Menetrier's [4-6] disease have similar gastric morphology and both are associated with protein-losing enteropathy, Menetrier's disease is confined to gastric mucosa and is not associated with ectodermal changes.

The ectodermal changes seen in Cronkhite-Canada syndrome are not exclusive to the condition. Similar changes are seen in hypoparathyroidism[2] and kwashiorkor.[2,7] The hyperpigmentation seen in Cronkhite-Canada syndrome is not believed to be secondary to hormonal changes.[2]

The polyps found in Cronkhite-Canada syndrome cannot easily be distinguished from other causes of colonic polyps. They resemble juvenile polyps found in stomach and colon and hyperplastic polyps in the stomach. The most striking histological feature is the intense oedema of the lamina propria of the mucosa. In addition gastric foveolae and colonic mucosal glands are cystically dilated. In the presence of surface erosion, the lamina propria may be inflamed, but in general inflammatory cells which include plasma cells and eosinophils are widely separated by oedema. Since in the stomach, the histological features are indistinguishable from juvenile or hyperplastic polyps the diagnosis depends on the presence of additional clinical features. In the colon, juvenile polyps have a smooth rounded surface and are pedunculated, while the polyps of Cronkhite-Canada syndrome are sessile and have an irregular, multilobulated surface.[8] Adenomatous and carcinomatous changes have been reported in Cronkhite-Canada polyps,[9,10] but this is rare, and dysplasia is not normally a feature. Similar polyps occur in the duodenum although they were not biopsied in this case.

The pathogenesis of the condition is obscure but interruption of the normal maturation of undifferentiated crypt cells to epithelial cells has been suggested.[11] The protein loss could be due to excessive mucus secretion by the crypt cells, along with malabsorption due to the villous atrophy. An improvement in the diarrhoea has been reported on administration of lactase in a single case,[12] suggesting that the diarrhoea could be partly caused by lactase deficiency.

The prognosis in Cronkhite-Canada syndrome is very poor, with few patients surviving more than a few months. Where the disease process seems localised, resection has been attempted with anecdotal reports of symptomatic remission. Steroids do not modify the course of the disease. Patients are usually resistant to enteral supplementation although two of the 55 patients reviewed by Daniel *et al*, showed complete symptomatic remission with resolution of all of the ectodermal aberrations.[2]

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In summary, this case history illustrates one of the many rarer polyposis syndromes. In particular, it illustrates the devastating degree of protein losing enteropathy, which can occur when the small bowel is involved.

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The Ulster Medical Journal, Volume 70, No. 1, pp. 59-60, May 2001.

Case Report

Cystic dilatation of the rete testis

M A Fawzy, B E Kelly, S R Johnston*

Accepted

Sonography is the primary imaging technique used for the assessment of scrotal swellings.

Seminiferous tubular ectasia at the level of the mediastinum testis is a recently recognized rare benign condition. It is often bilateral, and usually occurs in elderly men. It is commonly associated with epididymal abnormalities (e.g. spermatocele, epididymal cysts or epididymitis), and it shares a common location (within the mediastinum testis) with cystic dysplasia of the testis and testicular cysts. Obstruction caused by trauma or inflammation is thought to be a causative factor in seminiferous tubular ectasia and testicular cysts.

The sonographic appearance may be misinterpreted as a testicular neoplasm; however it has characteristic features which if recognized by the sonographer should result in avoiding unnecessary orchidectomy or testicular biopsy.

CASE REPORT A 72 year old man was admitted to hospital with sudden onset of urinary retention.

On examination, the patient had an enlarged prostatic gland. An incidental finding was a palpable lump in the left testis. The patient declared that he had had the lump for 5 years, and it had not changed in size.

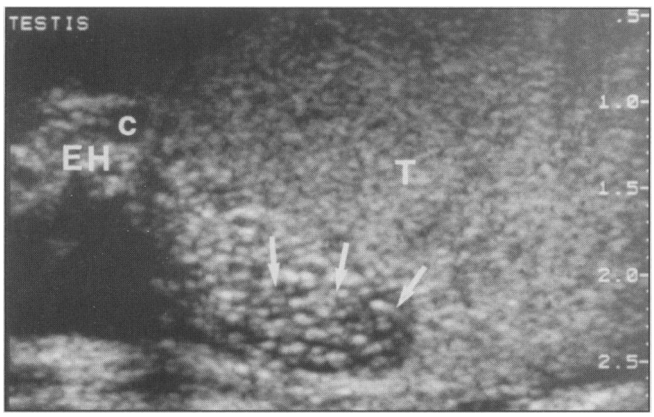

Fig 1. U.S. of the left testis showing cystic dilatation of the rete testis (arrows). (T.- testis, EH.-epididymal head, simple C.-epididymal cyst).

The patient underwent scrotal ultrasound (U.S.) examination (Figure 1), which revealed a left sided epididymal cyst measuring 2 x 0.6 cm, a larger one in the right side measuring 2.6 x 2.7 cm, and a large right hydrocele. There were also bilateral numerous small tubular and rounded anechoic cystic lesions within the rete testis. A diagnosis of dilatation of the rete testis was made.

A follow-up scan 25 months later showed no significant interval change, particularly no increase in size of the rete testis lesion, confirming the diagnosis.

DISCUSSION

Eleven patients with seminiferous tubular ectasia diagnosed with U.S. were reported by Weingarton.[1] The abnormality was frequently bilateral (8 out of 11) and associated with ipsilateral spermatocele in 10.

A large study by Brown,[2] described the sonographic findings in 31 cases with dilated rete testis. The median age of patients was 62 years. The abnormality was bilateral in nine patients. 34 of the 40 testicles involved had co-existing epididymal abnormalities, 32 with epididymal cysts and two with epididymitis.

Pavlica,[3] found that seminiferous tubular ectasia is always localized in the mediastinum testis, was bilateral in three patients, was associated with spermatocele in five, and with testicular cysts in

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four patients out of the 10 included in that study (all were over 50 years).

Nistal,[4] reviewed the testicular and epididymal specimens obtained from autopsies (1,798 men) or surgery (518 patients). Cystic transformation of the rete testis was found in 0.022% of the autopsies (all of them were elderly, and 85% of these were bilateral), and in 3.8% of the surgical specimens. They suggested that the mechanism leading to cystic transformation of the rete testis might be mechanical (compression of the epididymis by an epididymal tumor or spermatic cord tumour, or the result of a long standing epididymitis or traumatic haematocele), ischaemic (autopsied elderly men), hormonal (cirrhotic patients), malformative (cryptochidism), or unknown.

The condition has also been rarely described in children,[5,6] and associated with agenesis of the ipsilateral kidney in three cases.

We present a 72 year old man whose demographics are similar to those reported by Brown, Pavlica, and Nistal.[2,3,4] The abnormality was bilateral as described in other studies.[1,4] The sonographic examination also showed bilateral epididymal cysts and a large hydrocele on the right side. The presence of co-existent epididymal abnormality is described in several other studies.[2,3,4,7,8]

Brown,[2] confirmed the diagnosis of seminiferous tubular ectasia surgically and histologically in only one patient, and relied on the follow up U.S. scan (in up to 4.5 years time) to confirm their diagnosis in five out of their 31 cases.

Pavlica,[3] also did a follow up scan for five (out of 10) of his cases, over a period ranging from four to 19 months, which also showed no change in lesion size and structure.

One of the seven patients reported by Tartar,[7] had a histologically proven tubular ectasia when he had orchidectomy for a suspected testicular tumour. Spermatocelectomy was performed in another man whose testis was normal on surgical inspection. The diagnosis in all other patients relied only on the typical sonographic appearance. Tarta also described the MRI appearance of tubular ectasia in 1993 showing a homogenous signal similar to that of the co-existing spermatocele in all pulse sequences (hypo-intense relative to the testis on T1 and proton density weighted images, and, unlike tumors, were not visible on T2). Also follow up scans over a period ranging from eight to 36 months showed no change in the appearance.

In our patient we also relied on the typical sonographic appearance as well as the results of the follow up scan done after a period of 25 months to make the diagnosis and confirm the benign nature of the lesion and to eliminate the need for surgical intervention.

CONCLUSION

Tubular ectasia of the rete testis is an uncommon entity that is usually discovered incidentally during the U.S. examination for another scrotal abnormality.

The sonographic appearance, particularly the location of the lesion in the rete testis, the frequent presence of associated epididymal abnormalities (particularly spermatocele), the age of the patient, lack of interval change in the follow up scans, and the presence of bilateral lesions, should suggest the diagnosis, thus avoiding surgical intervention.

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The Ulster Medical Journal, Volume 70, No. 1, pp. 61-63, May 2001.

Case Report

A fatal case of carcinoma arising from a pilonidal sinus tract

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Accepted 7 March 2001

ABSTRACT

We report a male patient with carcinoma arising on the basis of neglected sacrococcygeal pilonidal sinus disease. Following initial operation, performed without suspicion of malignancy, histology demonstrated cellular atypia and an increased mitotic rate. A second, wider tissue excision was recommended but the patient declined further surgery. Two years later, he presented with fungating carcinoma involving the rectum but again declined surgery. This rare case demonstrates that the presence of carcinoma should be suspected in long-standing, although innocent-looking, pilonidal sinus disease. In the circumstance of uncertain histologic diagnosis, more generous surgical sampling is required. Every effort must be made to overcome patient's reluctance to accept a second, possibly life-saving procedure.

INTRODUCTION

Carcinoma arising from a pilonidal sinus tract is rare: only approximately 44 cases have been reported.[1-5] The incidence of the condition is estimated to be 0,1%.[2] Surgery brings the only hope of cure: wide excision of the lesion vvith tumour-free margins yields a disease-free, five years survival in ony 55% of patients.[1]

CASE REPORT A 49-year-old man had a 22-year history of a chronic, draining sacrococcygeal pilonidal sinus. He had had multiple pilonidal abscesses drained and innumerable episodes of sinus tract drainage. He gave no history of persisting pain and/or bleeding. Past medical history was insignificant; regarding the sacrococcygeal disease, the patient had deliberately avoided medical or surgical consultation. Physical examination was unremarkable except for the sacrococcygeal area

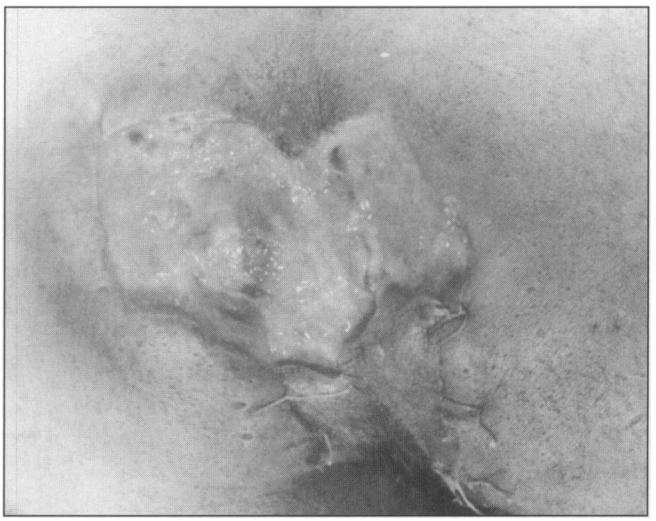

Fig 1. External appearance of the wound during the early postoperative period.

where multiple sinus tracts actively extruding purulent material were noted. The entire area was moderately indurated, without ulcerations. Inguinal lymph nodes were normal on palpation. Laboratory data and lumbosacral and coccyx spine films were all normal. The wound cultured *staphylococcus aureus* and pseudomonas aeruoinosa. After treatment with parenteral

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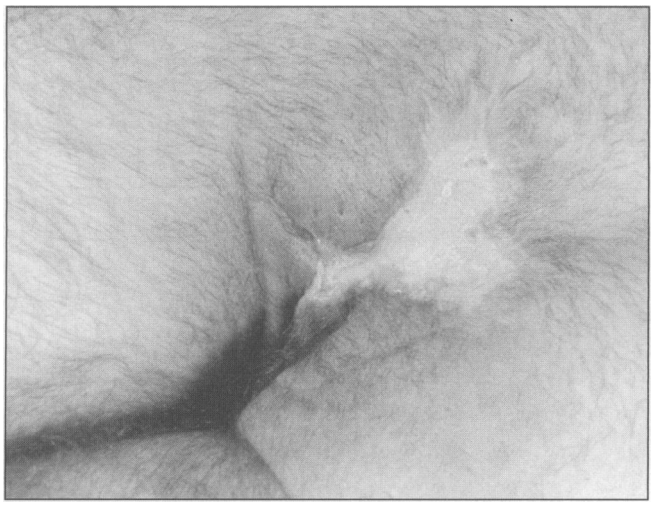

Fig 2. Wound left to heal secondarily.

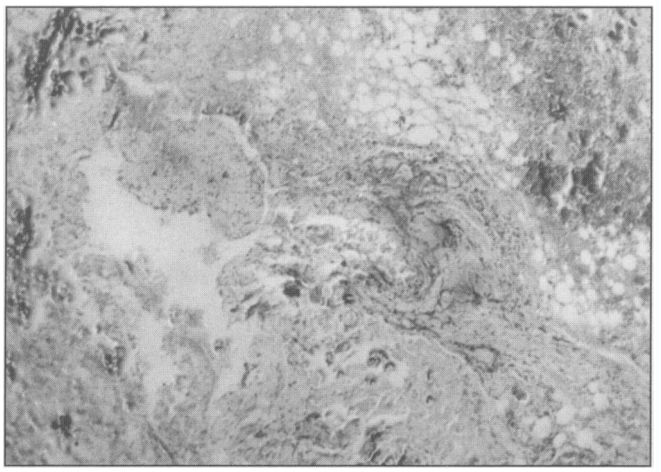

Fig 3. Photomicrograph of the excised specimen demonstrating severe inflammatory changes, pseudocarcinomatous hyperplasia, some cellulas atypia, an increased mitotic rate, but no overt carcinoma. Hematoxylin & eosin, x 80.

antibiotics and local wound care for 10 days, a significant decrease in inflammatory reaction was noted. Thereafter, complete excision of the lesion was performed (Figure 1) and the wound was left to partially heal by secondary intent (Figure 2). Pathologic studies revealed sinus tracts with severe inflammatory changes, and hyperplasia of squamous epithelium with some cellular atypia and an increased mitotic rate (Figure 3). A second wide local excision was recommended by the patologist because it was felt impossible to definitively exclude or confirm squamous cell carcinoma in the specimen, despite of the fact that the margin of surgically excised tissue was free of atypical cells. However, the patient declined further surgery. The wound healed well

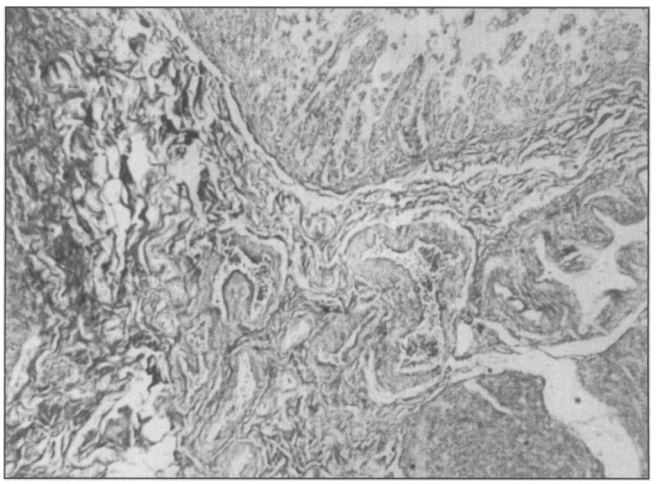

Fig 4. Photomicrograph of the biopsied sacrococcygeal lesion demonstrating squamous carcinoma. Hematoxylin & eosin, x 80.

and he was without evidence of disease one year after surgery. Two years following discharge from the hospital, the patient presented again. He had a fungating lesion 15 x 20 centimetres, extending deep to the coccyx inferiorly with involvement of the rectum on palpation; inguinal lymph nodes were markedly enlarged. Biopsy from the sacrococcygeal lesion demonstrated squamous carcinoma (Figure 4); biopsies were taken from the rectum and from an enlarged inguinal lymph node which again demonstrated infiltration by squamous carcinoma. The patient declined further treatment despite careful explanation of both his condition and prognosis of the disease. He was known to have died from metastatic disease at his home one month later.

DISCUSSION

The patient has been of interest because at initial presentation there were no macroscopic stimata inclicating malignancy but only those of innocent-looking draining sinus tracts. To our knowledge, this has been the first case with squamous carcinoma arising in a pilonidal sinus tract which presented initially without a fungating and/or ulcerating lesion. This rare presentation deserves special consideration from two aspects.

First, the incidence of malignancy is far too small for pilonidal sinus to be considered as carrying a significant malignant potential;[6] however, from the study of our patient it becomes evident that even in such an innocent-looking though long-standing pilonidal sinus the presence of a hidden carcinoma should be suspected and actively searched for. Among 3,734 primary operations

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for chronic pilonidal disease performed at our Institution between 1975 and 1999, this has been the only case of carcinoma, giving an incidence of 0,027%. Among all patients, 3,009 (80.58%) had had more than three episodes of recurrent inflammation and repeat purulent discharge, and 684 (18.31%) gave more than a 5-year-history of persisting discharging fistulae. Of all 3,734 patients, only 471 (12.61%) have been considered suspicious enough to deserve histologic investigation of the excised lesion, and from the 684 patients with long-standing disease, only 314 (45.91%) specimens have been subjected to microscopic examination. It seems wise to ask ourselves: in how many of these patients had there been in-situ carcinomas for which the excision performed had proved curative? We believe, as do others,[7] that all pilonidal sinus lesions should be sent for pathologic examination. It must be kept in mind that, possibly, specimens submitted for this are not always adequately studied.[8]

A second important aspect of this case that deserves mention is that at delailed histologic examination of the specimen there was no absolute certainty regarding the presence of carcinoma. The most important consideration in the histologic differential diagnosis of pilonidal carcinoma is pseudocarcinomatous hyperplasia of squamous epithelium, which may in fact be in association with any severe inflammatory process.[2,5] Since some cellular atypia and an increased mitotic rate (as found in our patient) may also be observed in pseudocarcinomatous hyperplasia, more generous tissue sampling may be required to establish the final diagnosis. In the present case, the histologic results were inconclusive initially and unfortunately the patient declined radical surgery. Taking into account the still very low survival rate (55%) and the very high recurrence rate (50%) in patient who have pilonidal carcinoma,[1-4] we believe that any histologic suspicion should prompt a repeat wide local excision with subsequent detailed pathologic examination, thus pursuing a definitive diagnosis and a better chance of cure. Finally, every effort must be made to encourage the patient to undergo a second surgical procedure which may be life-saving.

In conclusion, this report adds an interesting and instructive case to the scant number of patients affected by this rare entity.

ACKNOWLEDGEMENT

The authors thank Mahmoud N Kulaylat, MB, CHB, FACS, FASCRS, for supplying most of the articles necessary for the literature review.

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The Ulster Medical Journal, Volume 70, No. 1, pp. 64-66, May 2001.

Case Report

Dysphagia due to an aberrant left subclavian artery in a right-sided aortic arch

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Accepted 4 April 2001

In 1794, Bayford described the post-mortem findings of a woman with life-long dysphagia who eventually died of starvation, caused by oesophageal obstruction.[1] At post-mortem a right subclavian artery was identified passing aberrantly from a left-sided aortic arch behind the oesophagus, causing the woman's dysphagia. Dr Bayford referred to this extraordinary disposition of the right subclavian artery as, *lusus nature* or sport of nature. Since then the term 'dysphagia lusoria' has been used to refer to all aortic root anomalies causing oesophageal dysphagia.2 We report a case of dysphagia lusoria caused by a right aortic arch with a large diverticulum (diverticulum of Kommerell3) at the origin of an aberrant left subclavian artery.

CASE REPORT A 45 year old lady presented with a 2 month history of fatigue, shortness of breath, and a feeling of tightness at the lower end of her sternum associated with occasional episodes of dysphagia. Physical examination was unremarkable. Her ECG was normal. A chest radiograph demonstrated a right sided aortic arch. A subsequent barium meal demonstrated an area of constriction in the upper third of the thoracic oesophagus with an indentation arising posteriorly causing compression and contrast hold up. (Fig 1). The indentation corresponded to the level of the aortic arch and the radiological appearances suggested this was due to an aberrant vessel. Echocardiography was normal but subsequent

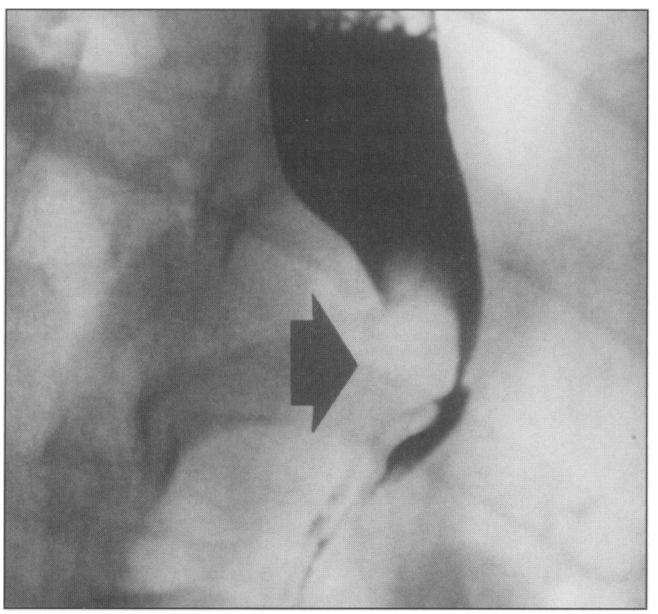

Fig 1. Barium meal image showing compression of the oesophagus by an aberrant vessel.

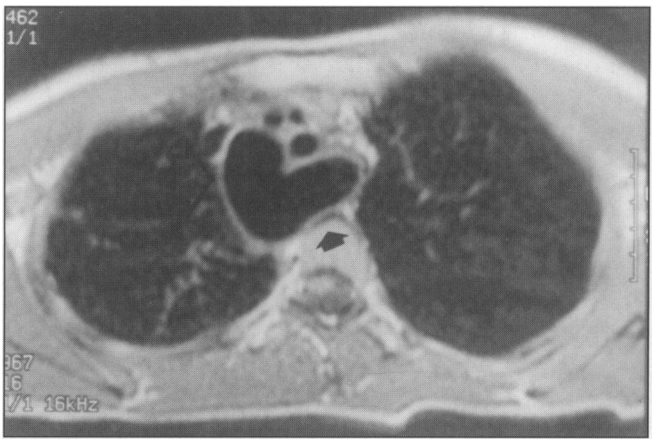

Fig 2. Axial T1-Weighted MR image through the thorax shows the right-sided aortic arch (open arrow) and the aberrant left subclavian artery with the wide mouthed diverticulum of Kommerell at its origin (closed arrow).

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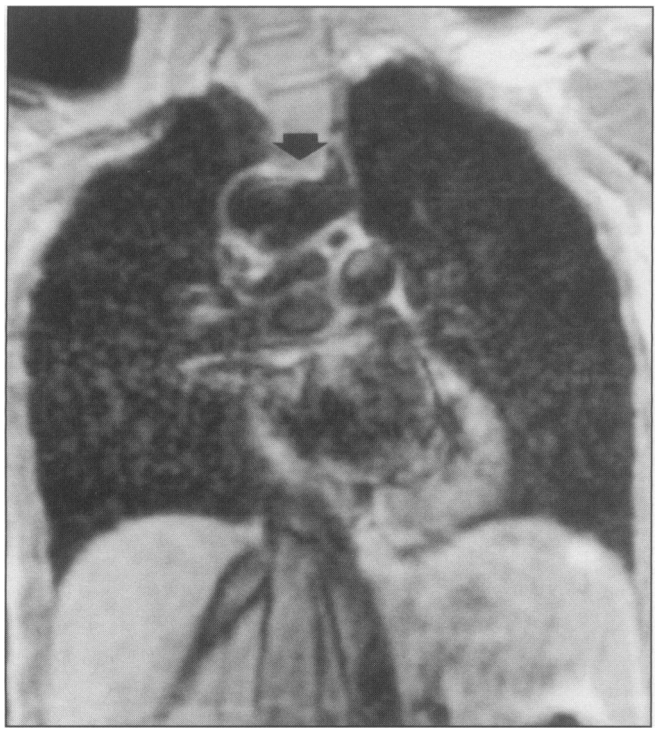

Fig 3. Coronal T1-Weighted MR image illustrating the diverticulum at the origin of the aberrant vessel.

cardiac catheterisation confirmed the right sided aortic arch and also a diverticulum, at the origin of an aberrant left subclavian artery, indenting the oesophagus. To further define the anatomy a magnetic resonance scan (MR) was performed (Fig 2 and Fig 3) which demonstrates the aberrant vessel and the diverticulum.

Surgery was deemed inappropriate given the mild symptoms and a conservative approach was adopted.

DISCUSSION

Developmental anomalies of the aortic arch and its major branches are relatively common representing 3% in post mortem series,[4] but they are usually asymptomatic. Aortic arch anomalies become symptomatic when they completely 'ring' the trachea and oesophagus or when coincident congenital heart defects occur.

Symptomatic aortic arch anomalies clinically present in a bimodal fashion. The trachea is compressible during infancy and these patients therefore typically present with respiratory symptoms: stridor, wheezing, cyanosis, or recurrent pneumonia when solid foods are introduced.[5] In adults, as the trachea is rigid, the oesophagus is more likely to be compressed, resulting in dysphagia.

During embryologic development the aortic arch begins as a duplicated system. As normal embryologic development continues, the right arch atrophies beyond the origin of the right common carotid and subclavian arteries. In 70% of the population the right subclavian and right common carotid merge to form the right innominate artery, the first branch of the normal left arch. In our patient the right arch persisted during embryologic development, while the left arch became vestigial between the origin of the left common carotid and left subclavian arteries. The diverticulum which develops from a vestigial remnant of the distal, embryologic left arch and gives off an aberrant left subclavian artery has come to be known as the diverticulum of Kommerell. A persistent right aortic arch with a diverticulum of Kommerell and aberrant left subclavian artery is an uncommon anomaly, estimated to occur in 1 in 1000 individuals. To actually see attributable symptoms associated with this anomaly is rare, as the ring that is formed, is generally loose.[6] Importantly, 5% of patients with this aortic root anomaly will have an associated congenital heart defect and 2% of patients with tetralogy of F allot will have this aortic arch abnormality. Echocardiography in this patient revealed no such abnormality.

The diagnosis of dysphagia lusoria can be elusive. It is not uncommon for patients to undergo repeated investigations for dysphagia that are unrevealing.[7] In cases where dysphagia lusoria is undiagnosed, patients have commonly been prescribed sedatives and tranquillisers for a presumed psychosomatic cause of their dysphagia.

The upper gastrointestinal barium study is an excellent method for the evaluation of dysphagia lusoria, but the diagnosis can easily be missed if the high thoracic oesophagus is not carefully examined, and if lateral or oblique projections of the oesophagus are not obtained. Endoscopy has the advantage of excluding other potential causes of dysphagia but it has been reported that it has a false negative rate of up to 50% in patients with dysphagia lusoria.[8]

Both computerised tomography (CT) and MR of the chest are helpful in evaluating the patient with suspected dysphagia lusoria.[9] MR is particularly sensitive in the evaluation of the mediastinal vasculature and can graphically demonstrate aortic arch anomalies. With its inherent advantages of multiplanar imaging and

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non ionising radiation, MR is rapidly becoming the imaging modality first employed. Angiography is still needed for pre-operative assessment.

Management of dysphagia lusoria is dependent upon the severity of the symptoms.[10] In our patient, the symptoms were relatively mild and slight dietary modification and explanation of the cause of the symptoms has been adequate. She has undergone review at outpatients and a second MR scan performed five years after the initial study has shown no change in the size of the diverticulum. In patients with increasing dysphagia or weight loss, surgical cure is possible by ligation and resection of the aberrant vessel.

CONCLUSIONS

The diagnosis of dysphagia lusoria requires a high index of suspicion. These patients present with symptoms of intermittent dysphagia and a mediastinal abnormality seen on a chest radiograph. Non invasive imaging of the chest with either CT or MR are excellent methods for evaluating the mediastinum for solid tumours or vascular anomalies that can cause extrinsic oesophageal compression. Dysphagia lusoria caused by a persistent right embryologic aortic arch and diverticulum of Kommerell with an aberrant left subclavian artery may be managed by dietary modification and follow-up when the symptoms are mild.

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The Ulster Medical Journal, Volume 70, No. 1, pp. 67-71, May 2001.

Association of Clinical Pathologists: Autumn Meeting 2000

NASAL PARAGANGLIOMA: A CASE REPORT

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Paraganglioma of the head and neck is an uncommon entity; only twelve such tumours arising in the nasal mucosa have been reported. As with all neuroendocrine tumours, it can be very difficult to predict the biological behaviour of these lesions. We describe a case of nasal paraganglioma and discuss the utility of immunohistochemistry in prognostic evaluation.

A twenty-four year old male presented with unilateral nasal obstruction and epistaxis; anterior rhinoscopy revealed the presence of a vascular polypoid lesion high in the left nasal cavity. Following resection by lateral rhinotomy, histology demonstrated a neuroendocrine tumour consisting of chief cells arranged in *zellballen*, surrounded by S100-positive sustentacular cells consistent with a paraganglioma; electron microscopy provided supportive ultrastructural evidence.

Paragangliomata may exhibit cytological atypia and mitotic activity but the vast majority are clinically benign – only four malignant variants have been described. Since morphology alone cannot be used as the sole determinant of benignity, S100 and Ki-67 immunohistochemistry have become a valuable adjunct; the presence of S100-positive sustentacular cells surrounding nests of chief cells is a reassuring feature. In the present case, Ki-67 immunoreactivity was demonstrated in 18% of the tumour cells, a greater state of cellular proliferation than one might expect in a benign lesion. Although numerous sustentacular cells were also identified, the degree of cellular proliferation is worrying and suggests a high potential for local recurrence.

Although paragangliomata can provide both diagnostic and prognostic difficulties, immunohistochemistry is a useful adjunct to routine histology and can reinforce the need for vigilant follow-up.

ROLE OF NESTED POLYMERASE CHAIN REACTION FOR DETECTION OF RESPIRATORY PATHOGENS IN CHILDREN WITH BRONCHITIS.

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Influenza A(H3N2) and respiratory syncytial virus (RSV) co-circulated during the winter months of November 1999 - January 2000. Nested polymerase chain reaction (nPCR) was compared with immunofluorescence (IF) for the detection of RSV serotype A and B in children with suspected bronchiolitis. Coinfection with influenza A(H3N2), Chlamydia spp. and enterovirus/rhinovirus was also investigated using molecular techniques.

A total of 50 nasopharyngeal secretions collected from babies with bronchiolitis in the month of January 2000 comprising IF RSV positive (n=27) and RSV negative (n=23) specimens were tested for both RSV serotypes, influenza A(H3N2), Chlamydia spp. and enterovirus/rhinovirus by nPCR. All specimens were also negative by IF for influenza A, influenza B, adenovirus and parainfluenza virus types 1, 2 and 3.

Twenty-six specimens were positive for RSV by both methods. Two secretions which were negative for RSV by IF were positive for RSV A and B respectively by nPCR. Chlamydia spp. (identified as C.trachomatis) was present in one infant with RSV B and who gave a recent history of treated *C.trachomatis* conjunctivitis. Enterovirus was found in 11/50 secretions of which 9 were speciated as rhinovirus, 3 had RSV or influenza A coinfection (RSV A=1, RSV B=1, H3N2=1) detected by nPCR and one had RSV by IF.

nPCR proved more sensitive and specific than IF in detecting RSV and Influenza A(H3N2). The presence of influenza A infection in secretions reported negative using conventional techniques highlights infection control issues in the cohorting of bronchiolitis patients in hospital. The demonstration of RSV and C.trachomatis infection addressed the contribution of the latter

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to bronchiolitis and the efficacy of treatment in eradicating Chlamydia carriage in this child.

CORRELATION BETWEEN SERUM IGA ANTIENDOMYSIAL ANTIBODY AND DUODENAL MORPHOLOGY IN COELIAC DISEASE.

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The objective of this study was to correlate the serum IgA antiendomysial antibody (EmA) status and titre with the severity of the small bowel enteropathy as assessed by light microscopy.

Three groups of twelve patients were reviewed retrospectively; all patients had a diagnosis of small bowel enteropathy by light microscopy. Group 1 had a positive serum IgA EmA, Group 2 had a weak positive serum IgA EmA and Group 3 had a negative serum IgA EmA. Repeat biopsies together with contemporaneous IgA EmAs and clinical details were reviewed.

The degree of enteropathy as assessed by villus crypt ratio, intraepithelial lymphocyte count (IEL) and inflammatory infiltrate in the lamina propria was most severe in the Group 1 patients and declined progressively through Group 2 and 3. This was statistically significant with regards to the mean villus crypt ratio, p=0.0007 (Mann Whitney U test) and the mean IEL count, p=0.049 (Student t test). The titre of EmA also correlated positively with the severity of enteropathy. Repeat duodenal biopsy, on a gluten free diet confirmed the presence of improved duodenal morphology. Similarly the EmA status reverted to the negative in the previously positive group.

In conclusion the presence and titre of EmA correlates positively with the severity of the small bowel enteropathy in Coeliac disease but is not 100% sensitive for its diagnosis.

MANTLE CELL IYMPHOMA PRESENTING AS A BREAST MASS

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Breast lymphoma accounts for less than 1% of all non-Hodgkin's lymphoma (NHL) and approximately 0.1% of all breast neoplasms. Most breast lymphomas are classified as diffuse large B-cell or MALT lymphomas according to the REAL classification.

A case of a 53 year old lady presenting with a breast mass and found to have a peripheral blood (PB) lymphocytosis is described. Core biopsy of the breast lesion showed a B-cell NHL, probably of large-cell type and of high grade. Morphologic and immunohistochemical analysis of PB and bone marrow (BM) suggested a mantle cell lymphoma (MCL). The diagnosis of MCL and the clonal origin of the lymphoma was confirmed by detection of t(11;14) in BM and breast tissue by polymerase chain reaction (PCR) analysis.

The patient received six cycles of CHOP chemotherapy following which there was no evidence of the lesion on mammography but residual disease remained in the bone marrow trephine biopsy. In view of this the patient proceeded to high dose chemotherapy with autologous peripheral blood stem cell transplantation. Subsequent BM trephine biopsy examinations have shown evidence of residual disease after transplantation.

To our knowledge there have been no previous reports of confirmed MCL presenting as a breast mass. As a diagnosis of MCL has therapeutic and prognostic implications for the patient, cyclin D1 immunohistochemistry and PCR analysis are necessary for its diagnosis. Haematologists and pathologists should also be aware that MCL can present in this way.

TRENDS IN BACTERAEMIA ON THE HAEMATOLOGY AND ONCOLOGY UNITS IN A UK TERTIARY REFERRAL HOSPITAL, 1994/5, 1998/9 AND 1999/00

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This is a retrospective study of bacteriologically confirmed bacteraemia/fungaemia on the haematology and oncology units at Belfast City Hospital Trust (BCHT) between September 1994 and August 1995, and September 1998 and August 2000 looking at trends in aetiology and sensitivity profiles. BCHT is a tertiary referral hospital with an 11 bed haematology unit and an oncology unit

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that has increased in size from 10 beds in 1994/5 to 18 beds in 1999. The trends in bacteraemia are consistent with the findings of the European Organisation for Research and Treatment of Cancer (EORTC) trials.[1] Coagulase negative staphylococci (CNS) and *Escherichia coli* remain the leading causes of monomicrobial bacteraemia on both the haematology and oncology units. On the haematology unit enterococci have emerged as an important cause of monomicrobial and polymicrobial bacteraemia increasing from one isolate in 1994/5 to 14 isolates in 1999/00. *Stenotrophomonas maltophilia* together with other non fermenting Gram negative rods are an increasingly common cause of bacteraemia on the haematology unit.

There were no isolates of glycopeptide resistant enterococci (GRE) or methicillin resistant *Staphylococcus aureus* (MRSA) in blood on the oncology unit. GRE were first isolated from blood on the haematology unit in 1997 and between September 1998 and August 2000 nine of 25 (36%) enterococcal isolates in seven patients on the haematology unit were glycopeptide resistant. During the same time period three of 19 (16%) isolates of *Staphylococcus aureus* in blood on the haematology unit were methicillin resistant. Only one of 50 isolates of *E. coli* from both units was resistant to ciprofloxacin.

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SOFT TISSUE SARCOMAS - THE BELFAST EXPERIENCE

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Soft tissue sarcomas tend to be difficult to manage due to their multitudinous anatomical sites with local recurrence presenting a particular problem. Given the rarity of these tumours, multidisciplinary management by specialist orthopaedic and plastic surgeons, radiotherapists, oncologists and pathologists is necessary for optimal results. The recent UKCCCR meeting on soft tissue sarcomas indicated that too many surgeons from various specialties are dealing with these tumours. We decided to both quantify the number of soft tissue sarcomas diagnosed in Belfast over a twelve-year period (1988 to 1999 inclusive) and to ascertain which surgical specialties have excised these tumours.

The histopathology reports of all patients with soft tissue sarcomas diagnosed in Belfast during the aforementioned period were retrospectively reviewed with regard to the tumour type and the referring specialty. Two hundred and thirty-four soft tissue sarcomas of 19 different histological types were treated by 35 individual surgeons from 9 specialties. Ninety-four tumours (40.2%) were excised by surgeons in disciplines other than orthopaedic or plastic surgery; 87 cases (37.1%) were operated on by orthopaedic surgeons and 53 cases (22.7%) were managed by plastic surgeons. Since the establishment of a sarcoma team in Northern Ireland in the mid- 1990s, the proportion of tumours excised by non-sarcoma specialists has halved, falling from 50% in 1995 to 24% in 1999.

Too many soft tissue sarcomas are still being managed by non-specialists. All surgeons in Northern Ireland and throughout the UK should be aware of the need for centralisation of soft tissue sarcoma management.

MOLECULAR DETECTION OF THE T(2;5) TRANSLOCATION BY REVERSE TRANSCRIPTASE-POLYMERASE CHAIN REACTION

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Anaplastic large cell lymphoma (ALCL) is a high grade lymphoma which usually shows expression of the Ki-1 (CD30) antigen and the presence of a specific translocation t(2;5)(p23;q35) which involves the genes coding nucleophosmin (NPM), a non-ribosomal nucleolar phosphoprotein on chromosome 5 and a novel tyrosine specific protein kinase called anaplastic lymphoma kinase (ALK).

This translocation leads to the production of a NPM/ALK fusion mRNA and protein. The presence of the products of the fusion gene can be detected by reverse transcriptase-polymerase chain reaction (RT-PCR). The t(2;5) has not been described in non-haematolymphoid malignancies using classical cytogenetics.

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Methodologically, molecular demonstration of the t(2;5) is problematical. RT-PCR is used to amplify the NPM/ALK chimeric message which is difficult to detect when RNA is extracted from formaldehyde-fixed paraffin embedded tissue.

Paraffin section immunohistochemistry on a lymph node from the left side of neck showed tumour cells staining with T cell antibodies and CD30. Staining for CD20 was negative and CD15 focal. Total RNA was extracted from formaldehyde-fixed paraffin wax embedded tissue biopsies using a guanidinium thiocyanate based method. RT-PCR was performed using oligonucleotide primers specific for chimeric mRNA encoding for the fused NPM/ALK transcript.

The presence of a NPM/ALK chimeric message was demonstrated using the method described. This approach should provide a reliable means of confirming t(2;5) in equivocal cases. Recognition of this NPM/ALK transcript is both diagnostically and prognostically significant since t(2;5) cases of ALCL represent a group of good prognosis neoplasms.

HOTSPOT MICROVESSEL COUNT, PLATELET COUNT AND PLASMA VASCULAR ENDOTHELIAL GROWTH FACTOR IN NON-SMALL CELL LUNG CARCINOMA

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The aim of this project was to evaluate the prognostic significance of, and any possible correlation between MVC, plasma vascular endothelial growth factor (VEGF) and platelet count in patients with non-small cell carcinoma.

The patient cohort studied consisted of 47 patients who had resection of non-small cell carcinoma of the lung. Microvessel counts were evaluated using the "vascular hotspots" technique, interactive digital analysis and anti CD34 antibody. Plasma VEGF and the peripheral blood platelet count were measured pre-operatively. The median follow-up was 18 months.

There was a positive correlation between plasma VEGF levels and the platelet counts. VEGF levels and platelet counts were inversely related to cancer-free survival. Raised microvessel counts suggested a poor prognosis but the findings did not reach statistical significance.

PRIMARY ADENOCARCINOMA ARISING FROM AN ILEOSTOMY

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Adenocarcinoma arising from an ileostomy site is rare with only 30 cases previously reported.

A 69-year-old female presented in 1999 to surgical outpatients with a six month history of a bleeding lesion at the verge of her stoma. She had undergone panproctocolectomy in 1965 for familial adenomatous polyposis (FAP). Local excision of the lesion was performed and histological examination revealed a well-differentiated adenocarcinoma.

The first such case was reported in 1969, and the mean interval between ileostomy formation and diagnosis of adenocarcinoma is 24.5 years. All the cases have been associated with FAP, ulcerative colitis or Crohn's disease.

The pathogenesis of an ileostomy adenocarcinoma is speculative. A feature present in many of the previous reported cases is colonic metaplasia of the ileostomy mucosa, and it is often postulated that this potentiates malignant degeneration of the stoma via either the dysplasia-carcinoma or adenoma-carcinoma sequence.

The lesion excised in this case was stained using high iron diamine-alcian blue and showed no evidence of black-staining sulfomucins which are only present in large bowel mucosa. This suggested that the tumour was of small bowel phenotype and did not arise on a background of colonic metaplasia.

Regardless of the pathogenesis, the clinical implications are that patients with longstanding ileostomies are at a small risk of developing stomal tumours, and with a long latency period the incidence is likely to increase.

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HER-2/NEU OVER EXPRESSION IN BREAST CARCINOMA: QUANTIFICATION AND CORRELATION WITH OTHER PROGNOSTIC VARIABLES.

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Her-2/neu is a transmembrane receptor the overexpression of which is an independent indicator of poor prognosis in breast carcinoma. Recently anti-Her-2 antibody has become available for therapeutic use in Her-2 positive tumours.

The purpose of this study was to evaluate the level of Her-2/neu overexpression in breast carcinoma and to correlate it with other routinely measured prognostic indicators.

A series of 80 cases of primary invasive breast carcinoma was immunohistochemically tested for Her-2/neu overexpression (Herceptest,™ DAKO). Also analysed were the patients' age, tumour type, tumour size, tumour grade, oestrogen and progesterone receptor status.

17 (21%) showed strong positive (3+) staining for Her-2/neu; a further 10 (12%) showed weak positive (2+) staining and the remaining 53 (66%) were negative. 9/17 (53%) of tumours showing strong positive staining were oestrogen receptor (ER) negative as opposed to 21/80 (27%) in the entire series. 5/17 (29%) were grade 3 tumours and 3/17 (18%) were grade 1 tumours, compared to 22% and 30% respectively for the series. The mean tumour size (29 mm) and the mean patient age (59 years) were essentially similar in Her-2/neu positive and negative cases.

Her-2/neu status is found to be an independent variable in breast carcinoma which correlates negatively with ER status. Positive Her-2/neu staining is associated with high tumour grade to a less pronounced extent. No relationship was found between Her-2/neu overexpression and either tumour size or patient age.

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The Ulster Medical Journal, Volume 70, No. 1, pp. 72-78, May 2001.

Northern Ireland Surgical Registrars' Prize Meeting

The first Northern Ireland Surgical Registrars' Prize Meeting was held in the Royal Victoria Hospital on Friday, 10th November 2000. All Basic and Higher Surgical Trainees within the Northern Ireland Surgical Training Schemes had been invited to submit abstracts for consideration for presentation at the meeting. From a submission of 26 abstracts, ten papers were selected for presentation.

Following the meeting, the John Templeton Prize for the outstanding presentation was awarded to Mr. Kevin McCallion for his paper entitled "Neurosurgical patients admitted to Intensive Care Units exhibit a systemic inflammatory response which is distinct from the mixed antagonist response syndrome exhibited by emergency surgical/trauma patients". Two additional prizes, the Ethicon and the Janssen Cilag awards, were given to Miss Jan Bingham and Mr. Robert Kennedy for their respective papers.

The following ten abstracts were presented at the meeting:

AN EVALUATION OF SERIAL MEASUREMENTS OF CEA AND CA 15-3 IN THE DETECTION OF PRE-CLINICAL RECURRENCE IN BREAST CANCER

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INTRODUCTION

An elevation in the level of tumour markers, CEA and CA 15-3, in blood has been positively correlated with the clinical stage of breast cancer. However, their use in the monitoring of disease recurrence remains unclear in the clinical setting. Combining the use of CEA and CA 15-3 has been reported to have a 64%-94% sensitivity for detecting disease recurrence, and a normal level suggests an absence of metastatic disease. However the specificity of these human markers and the senior level above which they are indicative of disease recurrence remains to be elucidated.

AIM

We aim to determine the sensitivity and specificity, and the role of serial measurements of CEA and C^a 15-3 in the detection of early recurrences in breast cancer, prior to the development of clinical signs or symptoms.

METHODS

Sequential measurements of CEA and Ca 15-3 were performed in the follow-up blood samples of 115 breast cancer patients who presented between 1995 to 2000, after treatment for their primary disease. The levels of these tumour markers were correlated with the clinical data. The cut-off values for serum CEA was compared between 2 ng/ml and 4 ng/ml, and for CA 15-3 was at 25 U/ml.

RESULTS

During follow-up, 28 patients (24.3%) developed recurrent disease. The sensitivity of serum CEA for detecting recurrent breast cancer was 37% (10/27), and specificity 83% using a cut off level >4 ng/ml. The sensitivity increased to 66% (18/27) when a CEA cut-off level >2 ng/ml was used, but the specificity dropped to 50%. For CA 15-3, the sensitivity was 57.7% and specificity also 83%. Combining both CEA (cut-off 2 ng/ml and 4 ng/ml) and CA 15-3, the sensitivity was found to be 69.2% and 84.6%, and the specificity 65.9% and 36.4% respectively.

The CEA and CA 15-3 levels were raised in 69.2% (18/26) using CEA >4 ng/ml as cut-off: they were raised in 88.5% (22/26) using CEA >2 ng/ml as cut-off. The time between a rise in the serum level of these tumour markers and the appearance of clinically detectable disease was between 2 to 44 months. Six patients developed symptomatic disease recurrence. They were further investigated radiologically, resulting in the administration of a different adjuvant therapy in 5 patients.

CONCLUSION

Combining the use of 2 tumour markers, CEA and CA 15-3, enabled early detection of breast cancer recurrence prior to the development of clinically apparent disease. A high sensitivity

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and specificity was achieved using CEA >4 ng/ml and CA 15-3 >25 U/ml to indicate disease recurrence. We propose that serial measurements of CEA and Ca 15-3 may be incorporated into the routine follow-up assessment of breast cancer patients.

THE USE OF URINARY TNF RECEPTOR CONCENTRATION AS AN INFLAMMATORY MARKER AND DIAGNOSTIC TOOL IN GASTROINTESTINAL DISEASE

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Soluble tumour necrosis factor receptors (TNFr) are released in sepsis. Urinary concentrations of TNFr have been shown to correlate with disease activity in patients with IBD. In this study, urinary concentrations of TNFr were measured along with recognised markers of inflammation in patients with one of four gastrointestinal conditions [colorectal carcinoma, acute diverticulitis, ulcerative colitis (UC) and Crohn's disease (CD)] and in healthy controls. We hypothesised firstly that urinary TNFr concentration would correlate with other markers of inflammation in gastrointestinal disease and secondly that TNFr measurement might be used to differentiate between UC and CD.

METHODS

Forty patients were recruited to the trial. Ten healthy controls were also enrolled. Erythrocyte sedimentation rate (ESR), C-reactive protein and alpha-1 acid glycoprotein were measured using standard laboratory techniques. Urine was also collected for measurement of p75 TNFr concentrations. Spearman's rank coefficients were used to assess correlation. The Mann-Whitney U test compared the ulcerative colitis and Crohn's disease results.

Urinary TNFr concentrations correlated positively with each of the other markers in each condition ($P<0.001$). There was no significant difference between TNFr concentrations in UC and CD ($P=0.257$).

The non-invasive measurement of urinary TNFr concentration may, in the future, be a useful, cost-effective marker of inflammation in patients with gastrointestinal inflammation but is unlikely to differentiate between ulcerative colitis and Crohn's disease.

RESULTS ARE EXPRESSED AS MEDIAN (INTER-QUARTILE RANGE)

	n=	*ESR (mm/hr)*	*C-reactive protein (mg/L)*	*α-1 acid glycoprotein (g/L)*	*Urinary TNFr (ng/ml)*
Colonic carcinoma	10	43(30-98)	48(38-242)	1.9(1.3-2.2)	22.8 (5.1-31.7)
Acute diverticulitis	10	79(46-111)	53(9-79)	2.0(1.1-2.1)	30.8(21.1-60.3)
Ulcerative colitis	10	15(12-26)	3(0-10)	1.0(0.6-1.2)	8.3(4.2-10.6)
Crohn's disease	10	25(16-50)	8(3-28)	1.6(1.2-2.6)	9.9(7.9-12.4)
Controls	10	14(13-19)	0(0-0)	0.5(0.3-0.6)	3.7(3.0-5.2)

NEUROSURGICAL PATIENTS (NSX) ADMITTED TO INTENSIVE CARE UNITS (ICU) EXHIBIT A SYSTEMIC INFLAMMATORY RESPONSE WHICH IS DISTINCT FROM THE MIXED ANTAGONIST RESPONSE SYNDROME (MARS) EXHIBITED BY EMERGENCY SURGICAL/ TRAUMA PATIENTS (TSX)

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INTRODUCTION

It was hypothesised that NSx patients requiring ICU admission would exhibit evidence of a systemic inflammatory response, explaining their risk of multi-organ dysfunction.

METHOD

NSx (elective and emergency, n=15) and TSx (n=10) patients admitted to a regional ICU had blood taken on the 3rd day of admission. Blood was also taken from controls (n=24). Maximal neutrophil (PMN) respiratory burst activity was assayed over 17.3 mins using a BioOrbit 1251 Luminometer to detect whole blood chemiluminescence in the presence of luminol, phorbol 1,2- myristate 1,3-acetate and tumour necrosis factor. The peak signal was obtained and divided by the white cell count ($x10^{-7}$mV/white cell). Soluble p55 TNF receptor (anti-inflammatory marker) and interleukin 6 (IL6, pro-inflammatory marker) were measured by ELISA and bioassay respectively.

RESULTS (MEDIAN [Q1,Q3])

TSx patients exhibited a MARS on day 3, with elevated IL6 (547 [232, 720] pg/ml *vs* undetectable levels in controls, $p<0.001$), elevated p55TNF (22 [15, 36] ng/ml *vs* 8 [5, 13] ng/ml in controls, $p=0.007$) and elevated PMN activity (61 [26, 94] *vs* 14 [11, 19] in controls, $p=0.001$). NSx patients exhibited a different inflammatory pattern, with

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elevated IL6 (217 [48, 280] pg/ml *vs* undetectable levels in controls, p=0.030), no rise in p55TNF (9 [7, 13] ng/ml *vs* 8 [5, 13] ng/ml in controls, p=0.662) and a non-significant elevation in PMN activity (30 [7, 79] *vs* 14 [11, 19] in controls, p=0.107).

DISCUSSION

The concept of a balance between pro- and anti-inflammatory mediators is attractive, explaining why measurements of a single mediator may be misleading. As PMN are subject to stimulation by many such mediators, assays of respiratory burst activity may be useful in determining the status of such a balance.

CONCLUSION

Patients admitted to ICU with neurosurgical insults exhibit a systemic inflammatory response which is distinct from that exhibited by emergency surgical/trauma patients.

EUROPEAN QUESTIONNAIRE SURVEY OF SURGICAL STRATEGIES FOR THE MANAGEMENT OF SEVERE ACUTE PANCREATITIS.

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OBJECTIVES

Although contemporary guidelines provide a framework for the initial management of severe acute pancreatitis, there remains no consensus opinion regarding the optimal surgical strategies for local complications. The objective of this study was to survey surgeons in hepatobiliary units across Europe to establish an overview of the current surgical management of severe acute pancreatitis.

METHODS

A mailshot questionnaire survey was sent to 866 members of the European Chapter of the International Hepatobiliary Association (IHPBA). A total of 331 questionnaires were returned, but only 327 (38%) were suitable for analysis (4 clinicians retired).

RESULTS

The most common (127 [38.8%]) number of patients with severe acute pancreatitis treated per year was 11 to 20. The commonest severity stratification system used was Ranson's criteria (168 [51.4%]). CRP was used as a serum marker of disease severity by 149 (45%) respondents, however 180 [55%] respondents used clinical assessment also. Prophylactic antibiotic therapy in severe acute pancreatitis was used by 229 (70%) of respondents, the commonest antibiotics used being third generation cephalosporins and imipenem. The majority of respondents (210 [64.2%]) used an early (within 72 hours) CT with 284 (86.9%) using intravenous contrast. 168 (51.4%) respondents use FNA of pancreatic necrosis for the diagnosis of local infection, with 99 (30.3%) of these stating they would always operate on a positive FNA. There was no general consensus as to the optimal timing of surgical intervention. The commonest procedure carried out for pancreatic abscess was percutaneous drainage (154 [47. 1 %]). The commonest procedure performed for pancreatic necrosis was surgical drainage, followed by closed irrigation (142 [43.4%]), then staged surgical resection (necrosectomy with skin closure and planned re-operation) in 94 (28.7%) respondents.

CONCLUSIONS

This study highlights the widely disparate protocols currently employed in the management of severe acute pancreatitis, advocating the implementation of standardised guidelines to encourage more consistency in the management of severe acute pancreatitis.

CIRCULATING LEVELS OF VASCULAR ENDOTHELIAL GROWTH FACTOR FOLLOWING OESOPHAGEAL CANCER RESECTION – RELATIONSHIP TO PLATELET COUNT

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BACKGROUND

It has been proposed that serum (S-VEGF) or plasma (P-VEGF) Vascular Endothelial Growth Factor levels may have prognostic significance in patients following cancer resection. However the

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potential confounding effect of surgical trauma is unknown The aim of this study was to investigate the degree and duration of alteration in circulating VEGF concentration and platelet count following oesophageal cancer resection.

METHODS

S-VEGF, P-VEGF and platelet counts were measured in 23 patients undergoing oesophageal cancer resection. Samples were taken preoperatively and at 6 weeks following surgery. Seven patients were also sampled on postoperative days 1, 5 and 10. VEGF levels were assayed using an Enzyme Linked Immunosorbent Assay (ELISA) kit specific for Human VEGF (Quantikine human VEGF, R&D Systems, Minneapolis, Minnesota.).

RESULTS

Circulating levels of VEGF were elevated postoperatively. However by 6 weeks S-VEGF had fallen to baseline but P-VEGF remained elevated ($p<0.05$, Wilcoxon Signed Ranks). Platelet counts mirrored changes in circulating VEGF levels, correlating closely with S-VEGF (rho=0.281; $p<0.05$, Spearman's Rank) and P-VEGF (rho=0.330; $p<0.01$, Spearman's Rank).

CONCLUSIONS

Surgery causes comparable elevations in platelet count and circulating VEGF postoperatively. S-VEGF and P-VEGF are closely associated with platelet count. The prognostic significance of circulating VEGF independent of platelet count remains to be confirmed.

RECURRENT SYMPTOMS AND FAECAL INCONTINENCE FOLLOWING HAEMORRHOIDECTOMY

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INTRODUCTION

Haemorrhoidectomy has been the gold standard for the treatment of significant haemorrhoids for many years. However the long-term outcome and the incidence of faecal incontinence following surgery is largely unquantified.

METHODS

The case notes of all patients who underwent a haemorrhoidectomy from January 1988 to January 1999 were reviewed. Demographic and operative data were retrieved. All patients were sent a postal questionnaire concerning recurrent symptoms, faecal incontinence, GP attendance and overall satisfaction.

RESULTS

164 patients (81 male; 83 female) received a questionnaire of which 120 (73.2%) responded. Three patients were deceased, 1 had a permanent ileostomy, and 40 were lost to follow up. The majority had elective surgery (n=112). The median age at the tine of surgery was 53 years (44-63) years and the median follow-up was 70 months (45-95).

Chart review suggested that 92 (76.5%) patients had not incurred a complication. Only 5 (4.2%) patients were recorded as having any degree of faecal incontinence post-operatively. Questionnaire results showed that 44 (36.7%) patients were symptom free whereas 28 (23.3%) patients had symptoms more than once/week. Twenty eight (23.3%) patients had consulted their GP and 30 (25%) had used further treatment. Bleeding was better controlled than itch/discomfort (no bleeding = 73; no itch/discomfort = 59). Only 45 (37.5%) patients were completely continent with 9 (7.5%) patients complaining of incontinence of solids, liquids, and gas more than once/week. A significantly higher proportion of patients who had undergone anal stretch at the time of surgery (n=29) experienced incontinence compared with those who did not have this additional procedure (69% *vs* 61%; p=0.05)*. Overall patient satisfaction was high with 91 (75.8%) grading their outcome as excellent or good.

CONCLUSIONS

Although surgery is considered as the definitive treatment for haemorrhoids, a significant number of patients have recurrent symptoms or faecal incontinence, which they may not report. Anal stretch at the time of surgery results in a higher incidence of faecal incontinence.

* Fisher's exact

CAN POOR OUTCOME AFTER CARDIAC SURGERY BE PREDICTED?

Quality of life and survival in patients after a long stay in cardiac surgical ICU following cardiac surgery

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Long stay in the ICU following cardiac surgery has major implications regarding manpower and resource utilisation. Majority of cardiac surgical procedures have a low risk with high probability of relief of symptoms and improved expectation of quality of life. A predictive model that identified patients who are certain to die would spare suffering and free resources. The aim of this study was to check the accuracy of the current risk scoring algorithms available to the clinician in predicting- poor outcome after cardiac surgery.

In the calendar year 1998, 1,006 adult patients underwent cardiac surgical procedures, only 34 (3.37%) spent more than 96 hours in the cardiac surgical ICU, the average length of stay excluding the long stay patients was 24 hours. Patients who had a protracted stay the mean was 631 hours (26 days), range (96-2,158 hours). Data was collected on these 34 long stay patients retrospectively and preoperative estimation of Parsonnet and Euroscore was done, postoperative estimation of APACHE II score and mortality prediction was performed. In hospital mortality in this group was 26.47% (9 deaths). At the end of one and half years there were 23 survivors, (11 deaths), telephonic administration of quality of life Short Form (SF 36v2™) questionnaire was done, 20 patients answered the questionnaire, two patients could not be traced while one refused to answer.

The sensitivity and specificity was calculated for the preoperative risk scores and postoperative APACHE II score for mortality prediction. Parsonnet score had (sensitivity=0.20, specificity=0.95, overall accuracy=0.72), Euroscore (sensitivity=0.45, specificity=0.91, overall accuracy=0.76), APACHE II score at 32 hours post op (sensitivity=0.82, specificity=0.76, overall accuracy=0.78), the predicted death rate calculated from APACHE II score (sensitivity=0.82, specificity=0.83, overall accuracy=0.82). Quality of life was severely affected with physical status score of 39.08 (range 22.21-57.09), however the average mental status score of 50.034 (range 22.75-63.31) represented only mild affection. The maximum number of potential ICU beds that could be freed was 61 days (16.71%).

Poor outcome after cardiac surgery can be predicted. The existing preoperative risk scores work well for the low risk patients but their accuracy is less in high risk patients. APACHE II score is designed for the noncardiac surgical patients, but it works well in high-risk cardiac surgical patients. The quality of life is significantly affected in the long stay patients after cardiac surgery. An improved risk algorithm needs to be developed which takes in to account the preoperative, intra operative and post operative factors in determining the morbidity and mortality after cardiac surgery.

BILIARY SYSTEM COMPLICATIONS FOLLOWING LAPAROSCOPIC CHOLECYSTECTOMY – IMPROVED OUTCOME WITH EARLY REFERRAL

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Laparoscopic cholecystectomy is now widely accepted as the 'gold standard' for treatment of symptomatic gallstones, however concerns remain about the incidence of bile duct injury. We review a series of patients referred to a hepatobiliary unit with biliary complications following laparoscopic cholecystectomy.

32 patients were referred over a 7-year period with complications following laparoscopic cholecystectomy, 5 male and 27 female with a median age of 58. Injuries were categorised according to Strasberg's classification.[1]

7 patients were referred immediately, 19 early (within 6 weeks) and 6 late (after 6 weeks). 12 patients sustained 'minor' bile duct injury (types A, B and C), 19 receiving more major type D and E injuries. 87% of patients required surgical intervention for definitive management. There were 2 deaths and 28 of the remaining 30 patients have normal liver function tests at follow-up.

STRASBERG CLASSIFICATION OF BILIARY INJURIES

Type	*Description*	*No. Patients*
A	Injury to bile duct (in continuity with CBD)	8
B	Partial occlusion of biliary tree	1
C	Injury to bile duct (not in continuity with CBD)	3
D	Laceration to extra-hepatic biliary tree	9
E2	Occlusion of biliary tree < 2 cm from confluence	10

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A successful outcome can be obtained for the majority of patients who sustain biliary injuries if they are promptly referred to a specialist centre. Late referral and previous inappropriate attempts at management can be fatal.

LAPAROSCOPY IN THE STAGING OF UPPER GASTROINTESTINAL MALIGNANCY

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BACKGROUND

Optimal management of cancer of the upper gastrointestinal tract requires accurate pre-operative staging to facilitate multimodality therapy planning and to identify those who would benefit from potentially curative surgery. Studies have suggested that staging laparoscopy is a useful adjunct in this process, reducing the number of unnecessary laparotomies in patients with advanced disease. The aim of this retrospective study was to compare staging laparoscopy with CT scanning in the preoperative staging of patients with upper gastrointestinal malignancy in the Royal Victoria Hospital over the last 2 years.

METHODS

Since 1998 laparoscopy has been used in combination with CT scanning in the staging of patients with upper gastrointestinal malignancy in this hospital. The medical charts of 49 patients in which staging laparoscopy was performed were reviewed retrospectively.

RESULTS

Over the study period staging laparoscopy was performed in 33 patients with gastric carcinoma, 10 with pancreatic/periampullary carcinoma, 3 with carcinoid, 1 with a gastrinoma and 2 with benign disease. Compared with CT scanning, laparoscopy confirmed ascites in 11 patients compared to 4. Five patients had peritoneal involvement that was not identified on CT. Furthermore; CT scanning suggested liver metastases in 2 patients that were not confirmed by laparoscopy. In addition laparoscopy identified a further 2 cases of liver metastases, which the CT scan had not picked up. There was no difference in the observation of lymphadenopathy or mass detection using the two modalities. Cytology of peritoneal washings at laparoscopy confirmed malignancy in 6 patients who had features of advanced disease.

CONCLUSIONS

This small study has demonstrated staging laparoscopy to be an important adjunct in the staging of upper gastrointestinal malignancy as it identified advanced disease that CT failed to demonstrate. This reduces the morbidity and mortality in patients who would formerly have had an unnecessary laparotomy. Peritoneal washout cytology does not appear to contribute significantly to the pre-operative staging of these patients.

ISCHAEMIA REPERFUSION INJURY IS EXAGGERATED BY DIABETES

W K Edrees, S E Refsum, I S Young, B J Rowlands, C V Soong

This study investigates whether diabetes mellitus accentuates hind limb ischaemia reperfusion injury and leads to greater muscle injury.

METHOD

Male Wistar rats rendered diabetic (n=40) following injection of streptozotocin were compared to non-diabetic control rats (n=30). Each group was divided into sham, 4h of hind limb ischaemia, 4h of ischaemia followed by 10 min, 30 min or 60 min of reperfusion. Blood was taken from the inferior vena cava for measurement of plasma concentration of the end product of lipid peroxidation (malondialdehyde (MDA)) and antioxidants vitamins A & E. Transmembrane potential (TMP) of the gastrocnemius muscle of the ischaemic limb was also measured.

RESULTS

Mean (s.e.m.)

CONTROL

	sham	ischaemia	10 min rep.	30 min rep.	60 min rep.
MDA (umol/l)	1.4(0.1)	1.1(0.1)	1.6(0.1)*	2.0(0.2)*	1.4(0.1)
Vit.A (umol/1)	1.5(0.1)	1.5(0.I)	1.5	1.4	1.3*
Vit.E (umol/1)	28.0(0.7)	28.0(0.9)	27.4(0.8)	25.4(1.0)	24.9(1.0)*
TMP (mV)	79.0(1.9)	63.0(2.2)	58.2(1.6)	53.5(2.6)	62 2(2.9)

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DIABETIC

	sham	*ischaemia*	*10 min rep.*	*30 min rep.*	*60 min rep.*
MDA	4.6(0.3)†	2.2(0.4)†	3.0(0.3)†	4.3(0.4)†	2.4(0.2)*†
Vit.A	1.0†	0.9†	0.8*†	0.7*†	07*†
Vit.E	26.9(0.8)	27.9(0.8)	27.7(1.5)	24.1(1.1)*	21.9(0.9)*
TMP	74.4(3.4)	56.0(2.2)	51.5(0.8)†	46.4(1.4)*	57.3(3.0)

* $p < 0.05$ vs ischaemia, †$p < 0.05$ vs control

CONCLUSION

These results indicate that oxidative stress following reperfusion injury is greater in the presence of diabetes mellitus. This may influence the recovery of diabetic patients following lower limb bypass surgery.

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