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The Ulster Medical Journal, Volume 56, No. 2. October 1987

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CASE REPORTS

The Ulster Medical Journal, Volume 56, No. 2, October 1987.

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The Ulster Medical Journal, Volume 56, No. 2, October 1987.

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External fixation in contemporary fracture management

G F McCoy, J F Orr, J Templeton

Accepted 30 June 1987.

SUMMARY

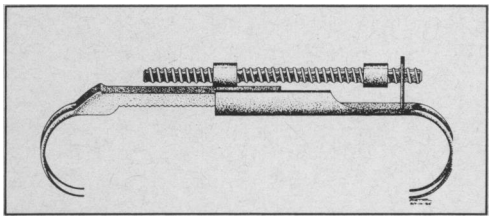
Important advances have been made within the last two decades in the field of fracture management. The development of the AO internal fixation system and the advances in cast bracing techniques are but two of the improvements worthy of mention. It is, however, in the field of external fixation of fractures that the greatest advances have been made. This paper traces the history of external fixation up to the present day and discusses, with examples, the application of external fixation in the management of complex limb fractures.

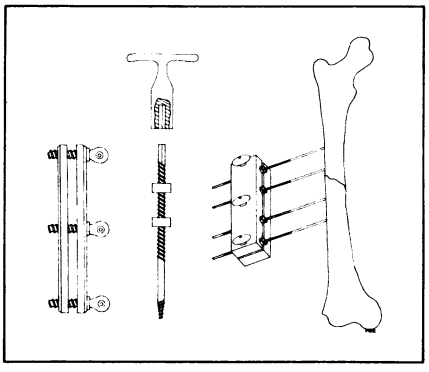
INTRODUCTION

More than a century ago, Malgaigne reported the first use of external fixation when he described a bone clamp (Fig 1) which percutaneously gripped patellar fragments and fixed them in the reduced position.[1] This initial report was followed in the succeeding decades by others.[2, 3, 4] Modern external fixation devices, however, began with Lambotte who, in 1902, described an apparatus used in the treatment of diaphyseal fractures of long bones and consisting of four iron screws clamped together between two plates (Fig 2).[5] With further experience of external fixation came the first reports of associated problems, i.e. pin tract infection, inadequate fixation and difficulties with realignment after application of the fixator.[6, 7, 8]

Fig 1 (below). The original bone clamp described by Malgaigne.

Fig 2 (right). Lambotte's external fixation device (1902), the forerunner of all the modern single-sided devices.





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In 1931, Boever summarised the indications for this technique, maintaining that it was the most aseptic method of osteosynthesis and, once a fixator was applied, movements of adjacent joints could be resumed.[9] However, despite the considerable contributions of Anderson,[10, 11, 12] external fixation gradually fell into disrepute, especially in the United States. The recurring problems of lack of rigid fixation and of pin track infection led the American Academy of Orthopaedic Surgeons to conclude that external fixation was of limited value in the treatment of fractures.

In Europe, however, developments continued with the work of Hoffmann,[13] who produced the first commercially available external fixator, a device consisting of two clamp units with universal ball joints and a connecting bar. This flexibility permitted reduction of the fracture even after the application of the fixator, a process which Hoffmann termed 'osteotaxis'. The unilateral nature of the fixator compromised its rigidity and limited its overall usefulness. The bilateral frame and clamp described by Charnley in 1948 was very much more stable and, although popularised as a device for knee arthrodesis, it was responsible for a revival of interest in external fixation.[14]

The original one-sided Hoffmann device was modified by Vidal [15] and Adrey [16] to a bicortical apparatus, greatly increasing its stability and widening the indications for its use. Jorgensen,[17] Olerud [18] and others [19, 20] further documented its role in a series of compound and complicated fractures. Great stability was a feature of the devices described by Ilisarov [21] and Wagner.[22] The latter device, although introduced as a leg lengthening apparatus, became widely used as an external fixator. As a single-sided device, the Wagner apparatus could be applied to the subcutaneous border of the tibia without impaling the anterior compartment musculature. This overcame the tethering effect of double-sided frames which frequently resulted in residual equinus deformity of the foot.

Burny [23] recognised the shortcomings associated with rigid double-sided fixation and proposed a single-sided bar and pin system, thereby introducing a degree of elasticity to trigger bulkier callus and more rapid fracture union. De Bastiani et al [24, 25] further developed the concept of elasticity at the fracture site with their introduction of the Dynamic Axial Fixator (DAF) which had a telescopic facility to allow for conversion to 'dynamic fixation' once callus formation had commenced, a process which they termed 'dynamisation'. In reporting their results in 288 patients, De Bastiani et al claimed a success rate of 94% with an average time to union of under five months.[24] Many reports now attest to the efficacy of external fixation in the treatment of complex limb fractures [26–28] and it is likely that the indications for, and the use of, external fixation in the treatment of such fractures will continue to increase.

THE CURRENT ROLE OF EXTERNAL FIXATION

It is now generally accepted that external fixation has a major role to play in fracture management, especially where other forms of immobilisation are either inappropriate or impracticable. The most common indication therefore is in severe open fractures where treatment by cast or traction methods would not permit sufficient soft tissue access. In addition, with injuries of such severity, the exposure necessary to implant an internal device may contaminate larger areas and might significantly increase the risk of infection or loss of the limb itself. With these grossly compound wounds, where the fracture site itself is exposed, an anatomical reduction can be achieved and maintained using an external fixator (Fig 3).

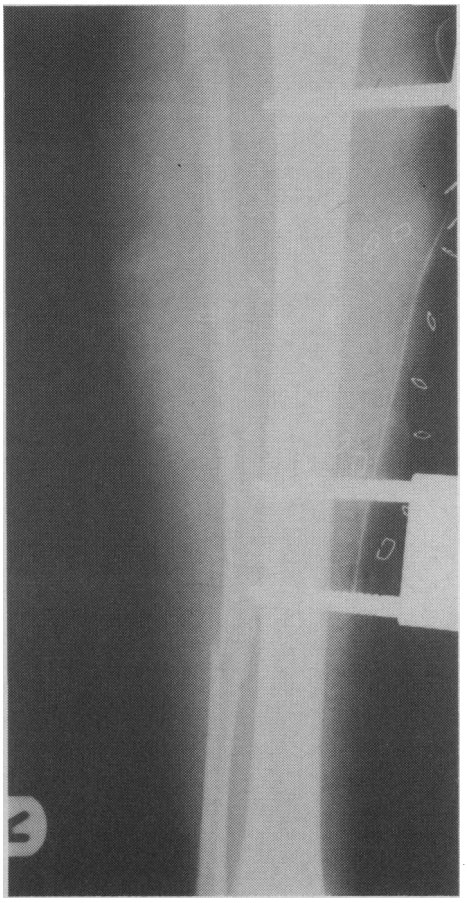

Fig 3. External fixation device in use with a compound tibial fracture. The fracture line is virtually invisible.

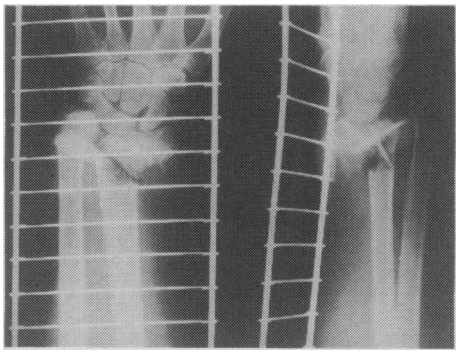

Fig 4. Comminuted fracture of the distal radius with considerable radial shortening.

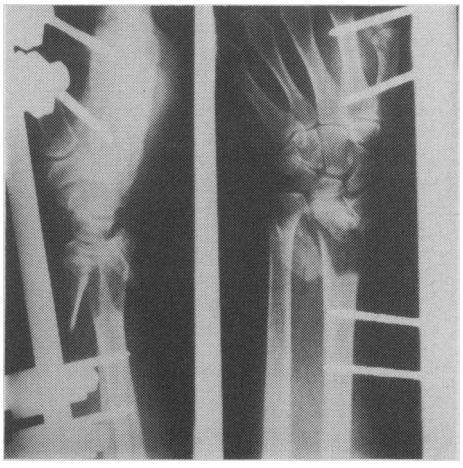

Fig 5. Radial length restored using a unilateral fixator.

The indications for external fixation are constantly being extended. Fractures associated with burns are optimally treated by this method, allowing wound toilet, dressing changes and skin grafting to be performed without disturbing the fracture alignment. Thus, rigid external fixation allows for aggressive and simultaneous management of the bone and soft tissue injuries. Where bone loss is present, especially in one of paired bones, external devices can be applied to restore and maintain bone length (Figs 4 and 5). In such cases, bone grafting can also be applied at the time of fixator application. Where there are vascular or nerve lesions in association with fractures, rapid fracture stabilisation can reduce the time taken to restore effective circulation and lower the incidence of limb loss (Fig 6). External fixation has undoubtedly saved many limbs which would previously have been lost.

Many closed fractures are now being treated by external fixation. The difficult spiral fractures of the distal tibia have a justified reputation for shortening and malunion (Fig 7). Treatment previously involved skeletal traction through a calcaneal pin which, although it improved fracture alignment, often compromised

subtalor movement. The application of an external fixation device improves and maintains alignment (Fig 8) while permitting immediate movement of the proximal and distal joints. This early mobilisation facilitates the reduction of oedema and limits capsular fibrosis, muscle atrophy and disuse osteoporosis.

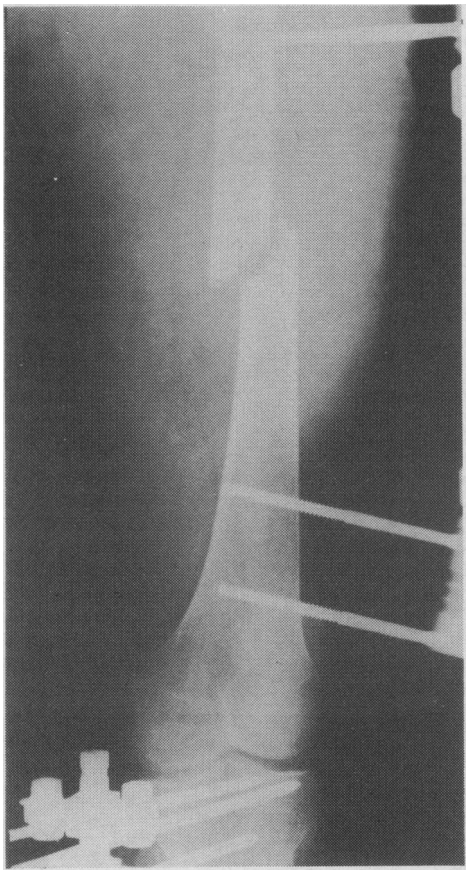

Fig 6 (left). Femoral fractures can be reduced and rigidly held within a few minutes, allowing the vascular surgeons a stable field in which to restore the circulation. In the example shown, the tibia has also been externally fixed.

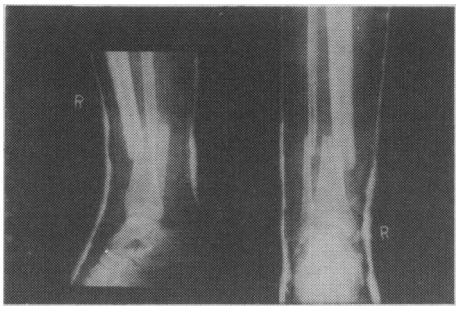

Fig 7. Closed spiral fracture of the distal tibia which has shortened and become displaced in plaster.

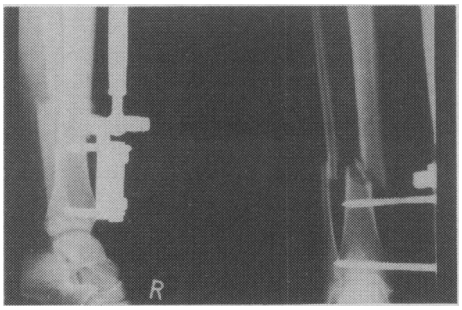

Fig 8. Alignment and length restored after application of an external fixation device.

External fixation is also used extensively in limb lengthening, arthrodesis and in the treatment of infected fractures and non-unions. In limb lengthening two techniques are in common usage. The first involves distraction at the osteotomy site once callus is radiologically apparent, a process which has become known as 'callustasi'. The second involves distraction at the growth rate towards the end of skeletal growth with encouraging early results.[29] It was Charnley who described the first rigid external device used for arthrodesis.[14] Originally proposed as a primary treatment for osteoarthritis of the knee, its indication in this area has been removed by the increasing acceptability of total knee replacement. Arthrodesis is however the salvage procedure for failed total knee replacement and, in the presence of infection, is ideally achieved using external fixation devices.[30]

Some debate exists as to whether bilateral or unilateral fixation should be used in severe limb fractures. Without doubt, the greatest stability is afforded by the two-sided quadrilateral frame popularised by Adrey.[16] This stability is gained,

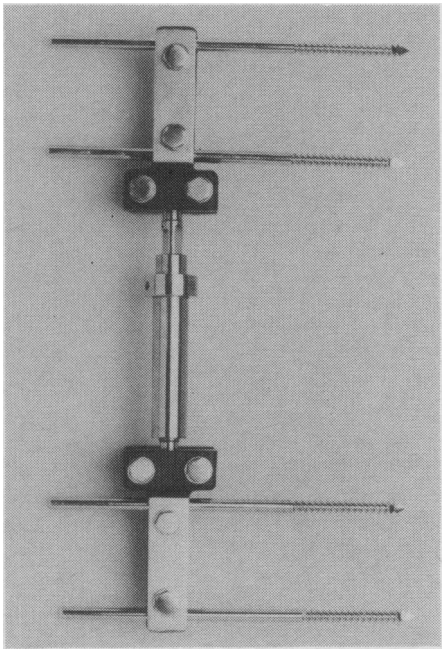

Fig 9. The Belfast external fixator in current usage.

however, at the cost of transfixing the adjacent musculature, which may cause restriction of movements at the distal joints. This is observed in tibial fractures where transfixion of the anterior compartment may lead to fixed equinus deformity of the foot. The rigidity afforded by the latest unilateral devices, such as the Orthofix or the Belfast (Fig 9), greatly reduces the indications for bilateral fixation, and many are now of the opinion that unilateral fixation is adequate for almost all limb fractures.

Every surgical technique has its complications, but, adherence to basic surgical principles can minimise their incidence. The chief complications of external fixation are those of pin loosening and pin tract infection. Pre-drilling of the bone and the use of wide-bore Schanz screws have greatly reduced both problems. With tibial fractures, the pins are inserted into the subcutaneous (antero-medial) border, thus avoiding completely the anterior compartment musculature. In fractures of the femur and the humerus, the fixator is generally applied from the lateral side, while in forearm fractures the device is applied from the radial side. In siting the fixators thus, movements of the knee, elbow and wrist are largely unrestricted. Obviously, a good knowledge of the cross-sectional anatomy of the limb is required — in particular, the safe zones for pin insertion. The radial nerve in the distal half of the arm and proximal third of the forearm, the dorsal sensory nerve just above the wrist and, the anterior tibial artery and deep peroneal nerve in the distal third of the leg are the structures most at risk of impalement. Vascular penetration, thrombosis, late erosions and the formation of arteriovenous fistulas and false aneurysms have also been observed.

Compartment syndromes have also been reported. It is likely that this is more a pure association than a direct consequence of transfixion or transfixation of bone. Anatomical fracture reduction may increase compartment pressure by reducing the volume available to accommodate soft tissue swelling. Therefore, in applying an external fixator, the surgeon must be especially careful to guard against a compartment syndrome by clinically assessing for and, if necessary, taking action to reduce, the intrinsic soft tissue pressure.

The more rigid forms of external fixation may 'unload' the bone at the fracture site with consequent demineralisation and weakening of the cortex, similar to that observed with internal rigid compression plate fixation. The callus produced is entirely endosteal, and delayed union rates of 20 – 30% have been reported.[31] Rigid fixation, whether external or internal, is attended by a risk of refracture after removal of the device. Bony union which is the result of rigid fixation is endosteal, with very little peripheral callus formation and thus very little intrinsic 'splintage'. In addition, the demineralisation resulting from rigid fixation leads to a form of

disuse osteoporosis, and the risk of refracture is increased unless the limb is adequately protected until remineralisation has taken place.

External fixation of fractures is a generally safe technique and its availability has saved many limbs which previously would have been amputated. The complications which have been discussed are much rarer than formerly, especially with the increasing use of single-sided fixators. The indications for, and potential of, external fixation are things with which every fracture surgeon should be familiar.

LOCAL EXPERIENCE WITH EXTERNAL FIXATION

Advances in other fields, such as vascular and reconstructive/plastic surgery, combined with an increasing number of severe limb injuries, led to a requirement among local orthopaedic surgeons to develop experience with external fixation techniques. From 1977, when the first fixators were applied, until the present, there have been more than 200 cases in which external fixators have been used. The earliest experiences were gained with the Hoffmann/Vidal and Wagner devices. Our experiences with the former confirmed the reports of other workers. Following application of these bilateral devices there was often difficulty with soft tissue access. In addition, there was a not inconsiderable incidence of residual equinus deformity following removal of the fixator. The Wagner device required an almost perfect reduction of the fracture prior to its application and, because of its rigidity, was associated with an increased risk of delayed union and of non-union.

Aware of the shortcomings imposed by existing devices, the senior author (JT) investigated the feasibility of developing a new single-sided fixator. The design incorporated a single external (outrigger) bar permitting maximal access for soft tissue procedures and twin clamps at either end of the outrigger to accommodate 6mm Schanz screws, these clamps being attached to the outrigger by means of universal joints which permitted adjustment of the fracture once the device had been applied. The device was engineered in such a way as to reduce weight and costs to a minimum. The outrigger bar permitted length adjustment during fracture reduction and could be locked using two 'Allen' grub screws which when unlocked allowed the fracture to 'dynamise'. A line diagram of the current Belfast fixator is illustrated in Fig 10.

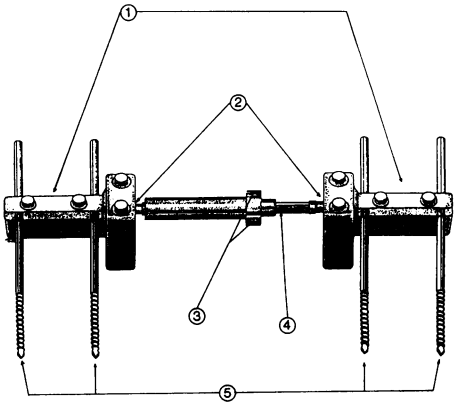

Fig 10. Line diagram of the Belfast fixator.

Commencing in 1981, when the first model became available, the Belfast external fixator was used in severely compound fractures of the tibia where maximal soft tissue access was required. With experience, it could be applied in under 15 minutes and, with open fractures, an anatomical reduction could be achieved under direct vision. The ease of application of the fixator and general satisfaction with the early results led to a general widening of the indications for its use. Eventually, almost all compound tibias and closed fractures where

satisfactory reduction could not be obtained or maintained were treated by external fixation.

From October 1981 until April 1986, 42 patients with fractures of the tibia were treated using the Belfast external fixator. Twenty-eight cases involved compound injuries, the remainder closed tibial injuries. The majority of these injuries resulted from road traffic accidents, particularly cases involving motorcyclists. Many patients had multiple limb injuries and some had severe head injuries. The majority of patients were young males in the age-group 18 – 35 years. Patients were kept in hospital so that any problems relating to the apparatus could be identified and dealt with immediately. When the fracture showed radiological evidence of callus formation, the fixator was removed, a cast applied and the patient discharged from hospital. On average, the fixator remained *in situ* for six weeks and a further eight weeks was required in cast.

Thirty-nine of the 42 patients treated using the Belfast external fixator were available for follow-up. Seven patients required remanipulation of the fracture as in-patients, but, because of the flexibility of the device, this was achieved without having to remove or re-site the fixator. Five patients had a pin tract ooze for variable periods and in one of these cases repositioning of the pin was required. None of these became persistent pin tract infections. Three patients developed non-union necessitating bone grafting and/or plating. There were three cases of delayed union in this series. In one patient, fracture healing was achieved with an unacceptable degree of shortening (in excess of two centimetres), whilst another patient was left with a residual equinus deformity which required subsequent corrective surgery. Of the 39 patients available for review, 30 achieved a good result, this being defined as healing in good alignment with less than one centimetre of shortening and within six months of injury. Although this might appear a high rate of indifferent results (23%), it must be acknowledged that all these fractures were complicated and often attended by risk of limb loss. Results would certainly have been much worse without the option of external fixation.

The more than 200 cases of complex limb fractures in which external fixation devices have been used have confirmed our belief that this method of fracture treatment will have a major and increasing role to play in the future. No longer reserved for the complicated and compound fractures, external devices are now being routinely applied to closed fractures which are reducible but likely to displace in plaster. Increasingly, it is the single-sided devices which are being used, thus avoiding the problems inherent in the bilateral fixators. Apart from the Belfast fixator, the results of which have been described, other devices commonly in use include the Hughes, AO and Wagner devices and, latterly, the excellent Orthofix external fixator. A programme is now under way to standardise external fixation in all the Belfast trauma centres, reducing the large variety of devices in use to two or three (the Belfast, Orthofix and possibly, the AO) but increasing their general availability.

FUTURE DEVELOPMENTS

The future for external fixation is very exciting. The increasing frequency of severely injured limbs as a result of high speed accidents has stimulated and maintained a general interest in its use. In some centres, external fixation has already become the mode of treatment for almost all major fresh fractures.[24] Whilst we would not advocate such general use of the technique, with increasingly reliable fixators, more and more fresh fractures will come to be treated by this

method. In the area of fractures involving the joint surfaces, the concept of 'ligamentotaxis' is being increasingly reported in the European literature. This permits the reduction of comminuted epiphyseal fractures by creating strong distraction on both sides of the joint, placing tension on the capsuloligamentous structures and aligning the fracture fragments.[32] By combining this with a hinged external device, continuous active movement is permitted at the joint surfaces allowing congruity to be restored and preventing joint stiffness. A similar hinged external fixation apparatus has been described for interposition arthroplasty.[33]

Undoubtedly, as the physiology of fracture healing becomes better understood, the design of fixators will further improve. The concept of fixators made from elastic materials is being investigated.[23] The dynamisation feature of modern fixators is a response to the observation that micromovement at a fracture site produces bulkier callus and therefore a reduced likelihood of delayed or non-union. Our early experience with these dynamising fixators generally supports this hypothesis.

External fixation also has a significant and increasing role outside the field of fracture treatment. Its role in arthrodesis following failed total joint replacement has already been mentioned. It is, however, in the difficult clinical area of limb lengthening that the potential is greatest. The earlier work of Wagner[22] and the more recent work of De Bastiani and his colleagues[29, 34] suggest that this is indeed the case. In the latter, De Bastiani et al report their results with 100 limbs lengthened using the Orthofix device. Increases in limb length of up to 65% were reported with no nerve or vascular lesions and no bony infections. No case required bone grafting and pin tract complications occurred in only 1.5% of pin sites. Not only are these exceptionally good results, but the lengthening was achieved within an acceptable time period and the child was able to walk, attend school and enjoy many normal activities while the fixator was *in situ.*

As it is now generally accepted as a major treatment mode in fracture management, the technique of external fixation is a skill which all who deal with bony trauma must master. Many limbs have been saved which previously would have been amputated. Reliable and safe external fixation of fractures has been ranked with arthroscopy and total joint replacement as a revolutionary advance in the field of orthopaedics and traumatology.

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Risk of recurrence after treatment of severe intraepithelial neoplasia of the cervix. A follow-up of 896 patients

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SUMMARY

Eight hundred and ninety-six patients were followed up cytologically for up to 21 years following treatment for a CIN III lesion of the cervix. The recurrence rate (8.8%) was lower after hysterectomy than after treatment which preserved the cervix (23%). Long-term yearly follow-up is not required as all recurrences were detected by annual smears for a seven-year period after treatment in both groups. It is important to keep patients under cytological review following hysterectomy because of the appreciable recurrence rate and also evidence that intraepithelial lesions of the vaginal vault behave in an aggressive fashion.

INTRODUCTION

Following treatment of cervical intraepithelial neoplasia (CIN), periodic cervical smears are required to ensure that the lesion has been completely eradicated. Long-term follow-up has been advocated.[1, 2] In Northern Ireland, patients are not uncommonly advised to have an annual smear until the age of sixty. There would seem to be grounds for this practice. Surgical excision of the lesion is often found on histological examination to be incomplete and, even in apparently normal epithelium surrounding the lesion, there is often evidence of papillomavirus infection which is thought to be associated with the development of CIN.[3, 4] Recent suggestions that carcinoma of the cervix has become more aggressive have also served to increase caution in the management of these patients. Frequent follow-up has disadvantages for both the patient and the laboratory service. A requirement for annual smears tends to set a woman apart from her fellows, continually focuses attention on the cervix and may well engender cancer-phobia. At the laboratory, cervical smears from these patients require to be examined by senior staff and, since the Belfast City Hospital has now been engaged in community screening for 22 years, a considerable number of patients are undergoing frequent follow-up and are straining limited resources. It is therefore important to determine safe guidelines for the management of these

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patients after treatment. At the Belfast City Hospital we were able to attempt this because all patients found to have abnormal smears during our 22 years of operation have been recorded on computer together with their biopsy results and their subsequent history obtained by the follow-up programme which the laboratory has operated.

METHOD

For this study we selected from our records all those women who had had treatment for a histologically proven severe cervical intraepithelial neoplasia (CIN III lesion) between 1965 and 1983 and on whom we had follow-up cytology smears and clinical information until September 1986. Most of the patients had had annual smears following their treatment. The patients were divided into two groups: those who had had cone biopsies (even though these often resulted in apparently incomplete excisions) and those who were treated by hysterectomy. Some of the biopsy group may have had subsequent cautery to the cervix. For each of these groups we determined from our computer records the pattern of recurrence of abnormal smears over a period extending to 21 years. We also examined the biopsy reports of 100 of those patients who were treated by excisional biopsy to obtain an indication of the proportion of patients in whom it was felt that the lesion was probably not completely excised. In order to assess their risk of invasive cancer we compared a list of those eventually lost to follow-up with a register of cases of invasive cancer in Northern Ireland from 1965 to 1987 previously compiled by the laboratory.

RESULTS

There were 896 women in our records who had been kept under clinical review with cervical smears following treatment of a histologically proven CIN III lesion of the cervix. Of these, 215 had had a hysterectomy following the initial biopsy, and in 681 the lesion had been treated by excisional biopsy. The percentage of patients subsequently having normal cervical smears in these two groups is shown in life table form in the Figure, which illustrates the first 12 years of follow-up. As expected, recurrence of an abnormal smear was much less common in patients treated by hysterectomy, 19 (8.8%) of whom showed evidence of recurrence in the vaginal vault. Of those in whom the lesion had been excised, 157 (23%) had relapse in the cervix. This recurrence rate in those treated by cone excision was, however, much lower than predicted by our review of the histology, which showed that in 50% of the patients it did not appear that the whole of the lesion had been removed. A striking feature is the similarity in the time of the recurrence, whether the patient was treated by hysterectomy or excisional biopsy. Most recurrences took place during the first two years with smaller numbers relapsing until the sixth and seventh years after surgery. Thereafter we found no further relapses even on prolonged follow-up to 21 years. The life table shows the recurrence rates as determined by a further positive smear without regard to the severity of the subsequent lesion as shown by biopsy. Our records show that in all cases the recurrence was still apparently intra-epithelial when first detected. However, in 12 cases invasive cancer eventually developed. Five of these arose in the vaginal vault after hysterectomy. Three were patients in their sixth decade who had abnormal vault smears after hysterectomy,

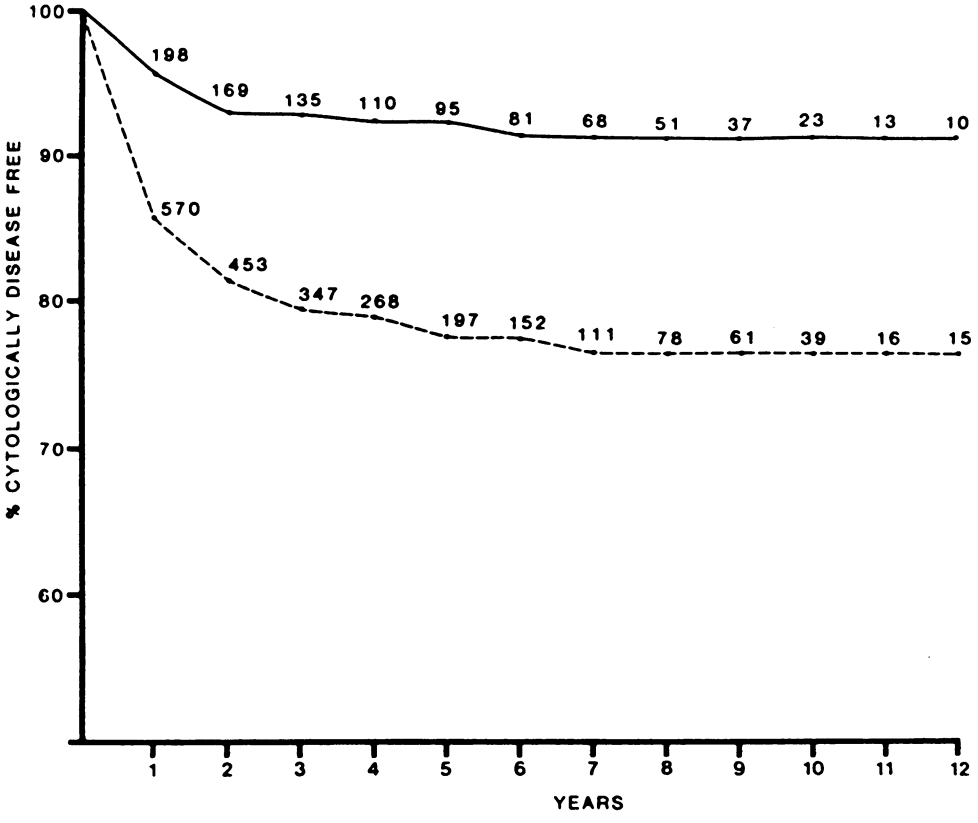

Figure. Follow-up cervical smear of patients with CIN III after hysterectomy (————, 215 patients) or local treatment to the cervix (— — — —, 681 patients). The results are given in life table form. Years 12 – 21 are not shown, there being no further relapses.

invasion occurring two years after operation. In a further two patients invasive cancer was found eight years after operation although in each case the first smear taken from the vault after hysterectomy was positive, there being no smears taken in the intervening period. Two further vaginal carcinomas arose in patients who had had their initial cervical lesion locally excised. Both had persistently positive smears after operation, the invasive lesion developing three years later in one and seven years later in the other, despite subsequent hysterectomy. There were five invasive cervical cancers, three arising within the first two years of operation. The other two developed the disease after eight and fifteen years respectively. In both it was associated with inadequate follow-up although one had a positive smear soon after cone biopsy.

DISCUSSION

Recurrence of cervical intraepithelial neoplasia (CIN) during the first two years after treatment very probably reflects incomplete excision of the lesion in the cervix or extension of the original lesion to the vaginal vault in those treated by hysterectomy. The slower relapse rate for the next five years may result from the

progressive development of epithelial changes which were only at an early stage of evolution at the time of operation, perhaps from epithelium remaining in the cervix showing no abnormality but already infected by the papillomavirus, currently thought to be closely associated with mutagenesis. Reinfection with this virus is also thought to occur frequently and could add to the relapse rate. Although in a number of patients the lesion may have been eradicated by subsequent cautery it is striking that only 23% patients relapsed. This contrasts with our biopsy review which indicated that, even on histological criteria, which probably underestimates the extent of epithelium infected by papillomavirus, about 50% of the lesions were likely to recur.

A low rate of recurrence of severe dysplasia has been noted by others.[1, 5] These surveys were, however, performed before the marked increase in prevalence of both papillomavirus infections and CIN lesions of the cervix accompanied by fears that cervical cancer was occurring in a more aggressive form. In Northern Ireland there has been a threefold increase in the prevalence of positive smears commencing in 1976 and it is reassuring that in our review, which includes patients treated between 1976 and 1983, relapse rates including mild CIN lesions remain low.

Although frequently commented upon,[6, 7] it has not been satisfactorily explained why CIN lesions may regress after partial removal. It seems possible that immunological factors are involved. There are many types of human papillomavirus some of which infect the skin producing the common wart. It has been shown that mechanical or chemical damage to the skin lesion can induce immunologic reaction with regression to the wart.[8] Conversely, in immunosuppressed patients, as following renal transplantation, both papillomavirus infection of the skin and also CIN lesions of the cervix are increased in incidence.[9] It would not be unexpected therefore if biopsy of the cervix had a similar effect in promoting immunologic resistance to the virus in the cervix.

Following excision of intraepithelial lesions of the cervix, follow-up with annual cervical smears is frequently advised for many years. Our results suggest that this is unnecessary and that after seven negative annual smears it is safe to assume that the lesion has been eradicated, further smears being taken at the normal screening interval. In contrast, after hysterectomy it is the experience of this laboratory that many clinicians are reluctant to arrange for follow-up smears in the belief that recurrence is unlikely and in the desire to spare patients the lengthy follow-up practised for women who have retained their cervix. However, an 8.8% recurrence rate is appreciable, and our results show that in this group too a seven-year follow-up should be sufficient.

A further finding of interest in this review was that of the twelve cases of invasive carcinoma known to us, seven arose in the vaginal epithelium. This suggests that intraepithelial lesions are more aggressive in this site, and indeed Koss also comments that in his experience vaginal dysplasias have a tendency to rapid progression and should not be left unattended.[10] This adds emphasis to our recommendation for review of patients treated by hysterectomy.

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Immune response to multiple skin test antigens in haemophiliacs

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SUMMARY

Using seven skin test antigens the cell-mediated immune response was evaluated in 20 haemophiliacs, 10 human immunodeficiency virus (HIV) antibody-positive and 10 antibody-negative. Response rates were compared with 75 healthy males of similar age range. All haemophiliac patients displayed significant impairment of cell-mediated reactivity to the test antigens; however, there was no apparent correlation with HIV antibody status.

INTRODUCTION

The acquired immune deficiency syndrome (AIDS) results from destruction of a subset of cells (helper T-cells) which regulate the cell-mediated immune system. The causative agent was found to be an RNA retrovirus designated human immunodeficiency virus (HIV). Infection with this agent may result in only partial damage of the immune system and patients may, therefore, remain completely asympotamic. However, approximately 30% eventually develop the complete immunodeficiency syndrome. While virtually all subjects (>95%) who have encountered the virus develop antibodies and became HIV antibody-positive, there are as yet no methods of predicting which individuals will progress to the state of profound immunodeficiency.

Before the causative agent of AIDS was discovered, infective blood products were accidentally used to treat patients with bleeding disorders, including haemophilia, and many haemophiliacs are now HIV antibody-positive.[1]

We studied these patients to determine whether *in vivo* testing of the cell-mediated immune system could identify those who had been exposed to the HIV agent. Some workers have shown that haemophiliacs, whether HIV antibody-positive or -negative, exhibit impaired cell-mediated immune response to a specific challenge with a single antigen not previously encountered,[2] although this type of testing has a number of disadvantages.[3] Assessment of *in vivo* reactivity of the cell-mediated immune system to multiple antigens, using delayed cutaneous hypersensitivity (DCH), overcomes many of these disadvantages[4] and

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also provides information regarding the recall of immune response to antigens previously encountered by individuals as a result of infection and/or vaccination programmes. The Multitest CMI system[5] was used to study 20 haemophiliac patients, 10 of whom were HIV antibody-positive and 10 antibody-negative and 75 healthy males of similar age range.

MATERIALS AND METHODS

Study population

Twenty haemophiliac patients, age range 19 – 64 years (mean 31 years) were tested using the Multitest CMI system. Nineteen patients were suffering from Haemophilia A, while one patient had Haemophilia B (Christmas disease). All patients were asymptomatic at the time of study with none displaying any clinical evidence of immunodeficiency. The testing was performed by a single investigator (RA McI) to avoid observer error; the HIV antibody status and T lymphocyte subsets results were determined independently. Results were compared with 75 healthy males, age range 19 – 65 years (mean 36 years) who were randomly selected from the general population. Persons with concurrent or recent illness were excluded as were any subjects taking drug therapy known to cause immunosuppression. Written informed consent was obtained from all subjects.

Multitest CMI and scoring system

The test system consists of a plastic disposable multiple puncture device capable of simultaneously applying eight test materials.[5] The antigens are applied by two rows of four puncture heads. The heads are 20mm apart so that positive reactions do not interfere with each other. Application was made to the volar surface of the forearm and skin induration was measured at 48 hours. The size of the inflammatory response was taken as the average of measurements made in two perpendicular directions. Reactions were considered positive if >2mm. The test materials include seven delayed type hypersensitivity antigens and a glycerin/saline diluent (negative control). The antigens used were two toxoids, tetanus and diphtheria, three bacterial antigens, Streptococcus, Tuberculin and Proteus, and two fungal antigens, Candida and Trichophyton.[6]

The scoring system which was used reflected the overall DCH reactivity of an individual. The score consisted of two parts: the first was the sum of the average millimetre induration for all positive responses; the second was the number of positive antigen responses out of the possible seven.

T lymphocyte subsets

At the time of the multitest CMI testing, blood was withdrawn to enumerate the proportion and absolute numbers of T lymphocyte subsets in all haemophiliac subjects. Mononuclear cells were separated by Ficoll hypaque density gradient centrifugation and the cells identified by an indirect immunofluorescent technique using monoclonal antibodies to OKT3, OKT4 and OKT8 phenotype markers for pan-T cells, helper-inducer and suppressor cytotoxic subsets respectively.

HIV antibody status

Blood was withdrawn and the presence of antibodies to the HIV virus was determined using an enzyme-linked immunosorbent assay (ORGANON).

Statistical analysis to DATA

The Mann Whitney U test was used to compare the frequency of positive reactions for each of the seven test antigens and the total millimetres of induration for each individual.

RESULTS

For the control population, the total inflammatory response ranged from 0–33mm with a median of 15mm. In haemophiliac patients, the range was 0–14mm with a median of 3mm. This was statistically significant at the 0.1% level. The number of positive responses to the antigens tested also showed a significant reduction in the haemophiliacs with a median value of 2 in this group compared with 3 for the control population; this again was significant at the 0.1% level. Overall, 99% of normal individuals reacted to one or more of the test antigens compared with 75% of the haemophiliacs, while positive reactions to two or more antigens were found in 95% and 50% respectively (Table I).

TABLE I

Number of control subjects and haemophiliacs with positive responses to test antigens

No. of positive reactions	*75 Controls*	*20 Haemophiliacs*
0	1 (1%)	5 (25%)
⩾ 1	74 (99%)	15 (75%)
⩾ 2	71 (95%)	10 (50%)
⩾ 3	57 (76%)	4 (20%)
⩾ 4	35 (47%)	1 (5%)
⩾ 5	18 (24%)	1 (5%)
⩾ 6	4 (5%)	0
⩾ 7	1 (1%)	0

Only one of the 75 normal individuals failed to respond to any of the antigens (1.3%) while five of the 20 haemophiliacs (25%) were completely anergic. However, analysis of data comparing haemophiliacs who are HIV antibody-positive with those HIV antibody-negative showed no statistical difference for either total inflammatory response or number of positive responses. Analysis of the data for reactivity towards individual test antigens showed a marked impairment of response towards the tuberculin reagent in the haemophiliac population with only one of the 20 showing a positive response compared with 58 of the 75 normal subjects (Table II). The mean number of helper T-cells in the patients positive for antibody to HIV was 675 (range 410–1180) cells/mm^3 compared with a mean value of 725 (range 410–1010) cells/mm^3 in the HIV antibody-negative patients. This difference was not statistically significant.

All the haemophilia A patients had received cryoprecipitate and Factor VIII concentrate. The mean quantity used per patient was 51,439 units. No correlation was found between the quantity of blood product given and HIV status or degree of suppression of the cell-mediated immune response. The patient who had

received the maximum quantity of Factor VIII (347,000 units) was HIV-negative with normal CMI response, while the patient with haemophilia B had received no Factor VIII or cryoprecipitate but became HIV-positive after plasma infusion.

TABLE II

Number of control subjects and haemophiliacs with positive DCH responses to test antigens

Test antigen	*75 Controls*	*20 Haemophiliacs*
Tuberculin	58 (77%)	1 (5%)
Candida	56 (75%)	13 (65%)
Tetanus	51 (68%)	10 (50%)
Diphtheria	37 (49%)	4 (20%)
Proteus	27 (36%)	0
Streptococcus	23 (31%)	3 (15%)
Tricophyton	10 (13%)	0

DISCUSSION

The Multitest CMI system evaluates the competence of the cellular immune system *in vivo,* and in particular its ability to respond to antigens previously encountered by the individual. Our results show that all haemophiliac patients treated with blood Factor VIII concentrates have significantly impaired responses compared with a healthy age/sex matched population. No significantly detectable difference in the responses of haemophiliacs HIV antibody-positive or HIV antibody-negative could be shown and, therefore, this method of testing would not be helpful in screening haemophiliacs for possible previous exposure to the AIDS virus. There was no apparent correlation between reduced cell-mediated immune response and quantity of Factor VIII used or the presence or absence of inhibitors.

This study would confirm the findings of other workers,[1] that haemophiliac patients display impaired cell-mediated immunity. While Madhok et al[2] have recently shown this phenomenon using a single test antigen (DNCB), our data would indicate that, despite presumably similar exposure to infectious agents and vaccination programmes, haemophiliacs become anergic to previously encountered antigens or fail to develop normal recall for antigens to which they have been exposed. The lack of correlation between depressed reactivity and HIV antibody status or quantity of Factor VIII treatment would suggest that the state of impaired reactivity may not be iatrogenic, but rather an inherent defect in haemophiliacs. However, it may indicate exposure of these patients to immunosuppressive agents other than HIV. While our study shows that haemophiliac patients fail to react positively to the tuberculin antigen, the clinical significance remains unclear. These patients were not discouraged from receiving BCG vaccination and we know of no increased incidence of tuberculosis in haemophiliacs compared with the general population.

We wish to acknowledge the help of Dr J Connolly who performed the HIV antibody testing.

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Henoch-Schönlein purpura: problems in surgical diagnosis and management

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SUMMARY

The clinico-pathological features of 133 consecutive cases of Henoch-Schönlein purpura are presented, with emphasis on the gastrointestinal manifestations. The potential pitfalls of contrast radiography are underlined with respect to management of intussusception and a plea is made to re-establish clinical assessment of the abdomen as the prime indicator in deciding to undertake laparotomy.

INTRODUCTION

In 1837 Schönlein described what he called 'peliosis rheumatica', a purpuric rash associated with arthritis, and 37 years later Henoch added his classic description of a rash associated with colicky abdominal pain, gastrointestinal haemorrhage and arthritis. Now, almost 150 years later, Henoch-Schönlein purpura is the term given to a syndrome characterised by the typical purpuric rash affecting the lower extremities associated with gastrointestinal, joint and renal involvement. The aetiology is unknown, and, although allergies to drugs (particularly antibiotics), some foods, or a hypersensitivity reaction to the beta-haemolytic streptococcus have been suggested, nothing has been proven. Pathologically the characteristic lesion is a vasculitis affecting the small vessels and this gives rise to the various manifestations of the syndrome.

Surgical intervention is required only in a minority of cases even when gastrointestinal symptoms are severe. However, the decision to operate or to continue with conservative management is often difficult. This review of cases at the Royal Belfast Hospital for Sick Children was undertaken to clarify and correlate the spectrum of gastrointestinal symptoms and pathology.

MATERIALS AND METHODS

Only those cases where the consultant in charge had classified the final diagnosis as Henoch-Schönlein purpura were included in this study. The patients' age, sex, date of admission, point of onset of illness and discharge were noted. The presence or absence of the typical skin rash involving the buttocks and the extensor surfaces of the lower limbs, joint pain and swelling, haematuria or proteinuria were recorded, and also the past medical history, family history and

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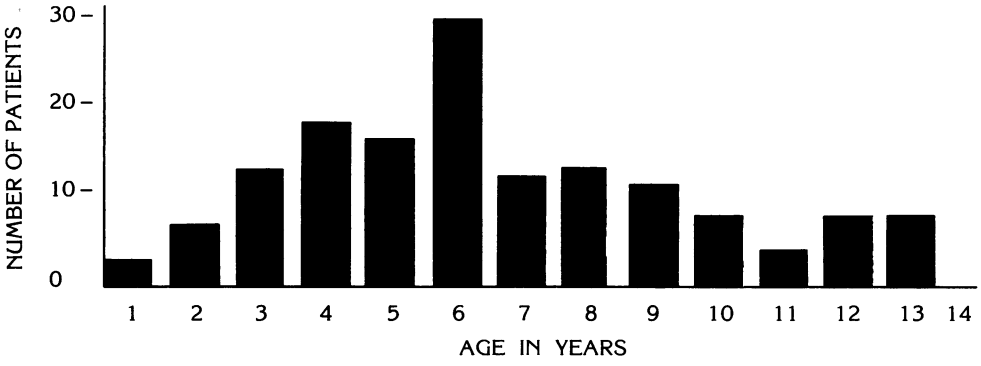
details of recent antibiotic administration. The specific gastrointestinal symptoms of abdominal pain, vomiting, diarrhoea and gastrointestinal bleeding were detailed.

RESULTS

Data were collected for the period 1972 – 1985 inclusive. A total of 133 patients were studied. There were 80 males and 53 females, a male : female ratio of 3 : 2. Ages ranged from one to 13 years with a mean of six years. The age incidence is presented in the Figure. Length of hospital stay was less than one week in 60 (45%) cases, 1 – 2 weeks in 35 (26%) cases and more than two weeks in 37 (28%) cases.

FIGURE — AGE INCIDENCE



In the two weeks prior to admission 53 (40%) of cases gave a history of upper respiratory tract infection and 33 (25%) cases had been prescribed antibiotics, 82% of which were of the penicillin type. The typical rash was present in 129 (97%) cases, an atypical rash in three, and in one there was no rash. In 47 (36%) cases, the rash appeared after the onset of abdominal pain. Joint pain and/or swelling was recorded in 107 patients (80%). In 73 (55%) only the lower limbs were affected. Isolated upper limb involvement was present in seven (5%), and in 53 (40%) both upper and lower limbs were involved.

Haematuria (overt or microscopic) or proteinuria were taken as evidence of renal involvement, and 52 patients (40%) showed these features. Haematuria was present in 23 (17%), proteinuria in 33 (25%) and 77 (58%) had both. In 84 (63%) of these patients, renal manifestations had resolved by the time of discharge, but, in the remainder, haematuria and/or proteinuria persisted for a variable period. Three patients developed renal failure. One, a six-year-old boy three months after the acute illness, did not require dialysis but he still shows biochemical evidence of renal impairment six years after the acute illness. Another, a 10-year-old boy, required dialysis and had a successful renal transplant. A third died with a nephrotic syndrome three months after his acute illness.

The profile of abdominal symptoms is presented in the Table. Abdominal pain and vomiting were by far the most common gastrointestinal problems. Melaena or haematemesis were uncommon, occurring in 26 (20%) cases, and 47 (35%) had other abdominal signs and symptoms without any evidence of gastrointestinal

TABLE

Gastrointestinal tract symptoms

Symptoms	*Number of patients*
Pain	58 (44%)
Vomiting	46 (35%)
Melaena	14 (10%)
Haematemesis	12 (9%)
Diarrhoea	8 (6%)

Total number of patients with gastrointestinal symptoms 73 (56%).

bleeding. Three children developed signs of intestinal obstruction and required laparotomy. Two were aged six years and one aged three years. They all had moderate to severe abdominal pain with vomiting; two had melaena, none had haematemesis. At operation two were found to have ileo-ileal intussusception and the third had an ileal submucosal haematoma. No other cases of intussusception were identified and no other children required surgery. Only three children received steroids as part of their therapy.

There were two deaths, but only one was directly related to Henoch-Schönlein purpura. The other occurred many years later from an unrelated cause.

DISCUSSION

Henoch-Schönlein purpura as an entity is essentially a clinical syndrome, there being no diagnostic laboratory test. The characteristic rash usually precedes abdominal pain but in our series the rash appeared afterwards in 36% of cases giving rise to obvious diagnostic difficulties. Despite this, no patient had an unnecessary laparotomy.

The cause of abdominal pain in this condition has always been assumed to be from involvement of the gastrointestinal tract. However, whilst 45% of those with abdominal pain had no evidence of kidney involvement, 73% of those with renal manifestations had abdominal pain without obvious gastrointestinal disease. Involvement of the kidneys may be responsible, at least in part, for the 'abdominal pain' associated with Henoch-Schönlein purpura in a significant percentage of cases.

The basic pathological process is a vasculitis resulting in perivascular oedema and haemorrhage. Involvement of the bowel, particularly the small bowel, may be of sufficient degree to cause a haematoma in the wall which may then give rise to perforation, subacute obstruction, intussusception or haemorrhage into the lumen.[1-7] The incidence of obstruction and intussusception is difficult to assess from the literature as so few reported series are on unselected patients. In this study, there were two cases (approx. 2%), as compared with reported incidences of up to 10%.[1, 5, 6]

Severe abdominal pain was present in 15% of cases in this study. Only 2% came to laparotomy, so pain *per se* is not necessarily an indication for surgery. More important are the signs and symptoms of intestinal obstruction or of peritonitis, which are relatively uncommon in this disease.[5]

A straight radiograph of the abdomen is mandatory in all children with evidence of intra-abdominal pathology. However, diagnostically and therapeutically, barium studies in Henoch-Schönlein purpura are of limited value, as 50% of intussusceptions in this condition are ileo-ileal.[3, 4, 5] This contrasts with 90% of all childhood intussusceptions which are ileo-colic, and although it is possible to reduce an ileo-ileal intussusception hydrostatically, we feel that it would probably be inappropriate to attempt this manoeuvre in the presence of obstruction and co-existing vasculitis of the gastrointestinal tract. Computerised axial tomography has also been used in the evaluation of the abdomen in Henoch-Schönlein purpura, and perhaps its most valuable future role would be accurately to distinguish a submucosal haematoma from an intussusception.[8] As yet, computerised tomography has not superseded the existing methods of assessment.

Earlier series reported an unacceptably high mortality and morbidity associated with surgical intervention.[4, 6] This was probably caused by delay in laparotomy, which almost certainly stemmed from a reluctance to operate: firstly, because surgical complications were relatively common; and, secondly, because steroid therapy theoretically places patients at an increased risk of infection and poor wound healing.[6] The role of steroids in contemporary management is now limited and their use should not cause any delay in the decision to operate. Although the incidence of significant surgical lesions in Henoch-Schönlein purpura is less than 5%, the morbidity in this group is high; we feel that with careful attention to clinical detail, it should not be necessary to perform laparotomy in the 10 – 15 per cent of those with more severe abdominal pain which has been suggested by other authors.[5]

Contrast abdominal radiography may be misleading or inconclusive in Henoch-Schönlein purpura, and we recommend that laparotomy should be undertaken without delay when a child with this condition develops clinical signs which would normally indicate surgery. Attributing such signs to an 'ileus' or submucosal haematoma could have disastrous consequences in allowing intestinal gangrene, perforation or septicaemia to develop.

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Anogenital warts: epidemiology, treatment and association with cervical atypia

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SUMMARY

In Northern Ireland in 1984 anogenital warts were diagnosed in 592 (352 male, 240 female) genitourinary medicine clinic attenders. Of these patients, 561 were heterosexual, 28 homosexual and three male bisexual. In the male patients 290 had penile warts, 67 meatal warts, 59 perianal warts, 25 anal canal warts and five rectal warts. In the female patients, 193 had vulval warts, 27 vaginal warts, 25 cervical warts, 107 perianal warts, 30 anal canal warts and one a rectal wart. Sexual partners were brought to the clinic by 345 patients and of these 93 male and 100 female partners had genital warts.

The mean time from exposure to development of warts was 17 weeks SE ± 1.5 (range 1 week – 12 months). As treatment, podophyllin 25% was used alone in 218 patients, and 132 were known to have had clearance of warts. At least one other sexually transmissible infection was found in 407 (69%) of patients. Cervical smears were taken in 164 women and were abnormal in 40 (24%). Cervical cytology was recorded in 89 regular sexual partners of male patients and was abnormal in 23 (26%).

INTRODUCTION

The study of human papilloma viruses (HPV) has been constrained by a number of problems, the principal one being the lack of an *in vitro* technique for propagating the virus. The virus is easily seen by electron microscopy in common skin warts and was first identified in 1949 [1] but it was not until 1968 [2] that similar particles were seen in extracts from anogenital warts. The use of molecular biological techniques has, in recent years, led to the identification of 46 types of HPV DNA,[3] these being enumerated in the order in which they have been identified. The common anogenital wart contains HPV types 6 and 11 but the main interest is the presence of DNA from HPV types 16 , 18, 31, 33 and 35 in 80 – 90% of cervical carcinoma with integration of the cellular genome in the cervical-carcinoma tissue.[3-14] No method exists by which these DNA types can be identified on a routine basis, and until this becomes available all persons with genital warts and their contacts need adequate investigation and follow-up for identification of potentially malignant lesions. In addition, such patients require

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investigation for other sexually transmitted diseases (STD) with which there is a high association. Traditional treatment methods for anogenital warts are generally considered unsatisfactory [15] but there is scant data available.

The aim of the present study was to review in retrospect all of the patients attending our clinic in 1984 with a diagnosis of anogenital warts, to analyse the clinical characteristics, rates of HPV infection, efficacy of treatments, occurrence of other sexually transmitted diseases, frequency of anogenital warts in sexual contacts and the result of cervical cytology in female patients.

METHODS

All patients who had the diagnosis of anogenital warts in 1984 confirmed at any of the three genitourinary medicine clinics in Northern Ireland (Belfast, Coleraine and Londonderry) had their medical records examined retrospectively. Age, sex, sexual orientation, month of diagnosis, duration of infection, site of warts, treatment given and duration of treatment were noted. Other sexually transmitted diseases found in these patients were recorded. Sexual contacts were traced from hospital records. Cervical cytology was recorded both in female patients with warts and female sexual partners of males with warts. The treatment in use during the study period was podophyllin 25% initially, except in pregnancy, or for flat keratinised and intrameatal warts. Concentrated trichloroacetic acid and cryotherapy (Spembly cryoprobe No. SRIR) were used in the latter and in all patients unresponsive to podophyllin.

RESULTS

In 1984 in Northern Ireland anogenital warts were diagnosed in 592 clinic attenders (352 male, 240 female). Of these patients, 561 (95%) were heterosexual, 28 (5%) were male homosexual and three (0.5%) were male bisexual. There was no clear pattern in the monthly rate of diagnosis of anogenital warts (the range was 36 – 63 cases per month). Ages ranged from two to 68 years, mean 25 years ± SE 0.3 (0 – 15 years — four patients; 16 – 19 years — 90 patients; 20 – 24 years — 240 patients; 25 – 29 years — 136 patients; 30 – 34 years — 66 patients; 35 – 39 years — 29 patients, and over 40 years — 27 patients).

Of the whole group 401 were new patients with a first infection of anogenital warts, 13 were new patients with a known history of anogenital warts treated in another hospital, 119 were previous clinic attenders with a first presentation of anogenital warts and 59 had a relapse or new infection of anogenital warts.

Sites of infection. In the 352 male patients, 290 (82%) had penile warts, 67 (19%) had meatal warts, 59 (17%) had perianal warts, 25 (7%) had anal canal warts and five (1%) had rectal warts. In the 240 female patients, 193 (80%) had vulval warts, 27 (11%) had vaginal barrel warts, 25 (10%) had cervical warts, 107 (45%) had perianal warts, 30 (12%) had anal canal warts and one (0.4%) had a rectal wart.

Sexual partners. Of the 592 patients, 345 (58%) brought a total of 392 sexual partners to the clinic. One partner was brought by 311 patients, two partners by 26 patients, three partners by six patients, five partners by one patient and six partners by one patient. Proportionately more female patients, 163 (68%) than male patients, 182 (52%) brought partners to the clinic ($p < 0.0005$). Of the 392 contacts who attended the clinic, 193 (49%) had anogenital warts — 93 (52%) male and 100 (48%) female.

Incubation. The time between first exposure to infection from a partner known to have anogenital warts and the development of anogenital warts was clearly inferred in only 67 patients. This was less than one month in six patients (9%), less than two months in 20 patients (30%), less than three months in 35 patients (52%) and less than six months in 55 patients (82%). All patients had developed warts by 12 months. The mean time to development of warts was 17 weeks ± SE 1.5 and the median time was 12 weeks.

The period of time the patient had noted warts before diagnosis was given by 326 patients: 156 patients (48%) had warts for 0 – 4 weeks, 72 (22%) for five to eight weeks, 32 (10%) for nine to 13 weeks, 34 (10%) for 14 – 26 weeks and 32 (10%) for more than 26 weeks.

Treatment. Podophyllin 25% was used in 493 patients and was the only treatment in 218 patients. In this latter group, 119 (55%) had one application only, 44 (20%) had two applications, 29 (13%) had three applications, nine (4%) had four applications, nine (4%) had five applications and eight (4%) had six or more applications. In this group of 218 patients, after treatment 132 were known to have had clearance of their warts (had had one visit to the clinic after their last treatment when they were observed to have no warts present) and the other 86 were clinic defaulters. Trichloroacetic acid was used in 245 patients, 160 (65%) had two applications, 51 (21%) had four applications, 10 (4%) had six applications and 24 (10%) had more than six applications. It was the sole treatment in only 16 patients. Cryotherapy was used to treat warts of the urethral meatus in 60 patients with 32 (53%) having one treatment only, 15 (25%) having two treatments, eight (13%) having three treatments and five (8%) having four or more treatments. Cryotherapy was used in sites other than the urethral meatus in 159 patients: 92 (58%) having one application, 33 (21%) having two applications, 22 (14%) having three or four applications, six (4%) having five or six applications and six (4%) having seven or more applications.

Clinic attendances. Attendances before cure or default were one or two visits — 287 (48%); three or four visits — 110 (19%); five or six visits — 65 (11%); seven or eight visits — 38 (6%) and nine or more visits — 92 (16%) patients. The duration of treatment was up to two weeks in 257 (43%) patients, and over 26 weeks in 51 (9%) patients. The overall known cure rate was 53% (314 patients). Relapses occurred in 63 patients (11%) after eight weeks clear of warts and, of these patients, 14 (2%) had a relapse after six months clear of warts.

Other infections. A total of 407 patients (69%) had other sexually transmissible diseases present. These diseases were: non-specific genital infection — 308 patients (52%); Gardnerella vaginalis — 119 patients (20%); candidiasis — 102 patients (17%); gonorrhoea — 45 patients (8%); pediculosis pubis — 15 patients (2.5%); trichomonas — 12 patients (2%); herpes — 10 patients (2%), and scabies in two patients (0.3%).

Cervical cytology. Cervical smears were taken from 164 women with anogenital warts and, of these, 40 (24.4%) were reported as abnormal. The abnormalities reported were koilocytosis in one case, dysplasia in 31, cervical intraepithelial neoplasia III in seven and carcinoma-in-situ in one. In the group with cervical warts (25 patients), 20 had cervical cytology, with 12 (60%) having abnormality, and eight (40%) normal smears. Cervical cytology was recorded in 89 regular sexual partners of male patients with genital warts, and was abnormal in 23 patients (26%). Of this latter group, anogenital warts were recorded in only 12

patients (52%), including one patients with cervical warts. The cervical cytology abnormalities noted were dysplasia in 19 cases and CIN III in four.

DISCUSSION

The epidemiology of anogenital warts behaving as a sexually transmitted disease has already been established in studies by Oriel.[16, 17] In this study the findings are as would be expected from a sexually transmitted infection, with the male-to-female ratio reflecting that of our overall clinic attenders, the predominant age group affected being the 16–30-year-old group and sexual contacts being affected in 49% of cases. In Oriel's first study,[16] 64% of contacts were seen to have warts; our study, however, included both casual and regular partners, whereas his looked at regular partners only and consequently his subjects would have had an increased exposure to infection. The association with other sexually transmitted diseases in 69% of cases is characteristic and underlines the importance of adequate screening.

The retrospective nature of this study makes it difficult to obtain accurate information regarding the incubation period of warts. Our finding of a mean incubation period of 17 weeks in the 67 patients whom we thought could be accurately assessed was longer than the 2.8 months noted by Oriel.[16] A presumed incubation period of up to one year was noted in some of our cases, whereas Oriel gave a maximum of nine months. This variable latency period in wart virus infection suggests that contact tracing should in some cases be extended up to one year if there is no definite history of contact within this period.

Previous reports suggest that males are less likely than females to bring contacts to the clinic;[18] this is reflected in the present study, and is particularly worrying in view of the association between HPV infection and cervical dysplasia and malignancy.[3–14] The response rate to contact tracing in this study of 58% of patients bringing partners for examination left many people untraced. While there is some doubt about the carcinogenic role of HPV,[13, 14] there is strong evidence in support of a link between HPV and cervical dysplasia and therefore it was not surprising that, of women with anogenital warts who had cervical smears, 24% were abnormal and, in the subgroup of women with cervical warts, 60% had cytological abnormality on smear testing. It was also worrying that in the female partners of men with anogenital warts, cervical abnormality was reported in 26% of contacts while less than half of these patients actually had clinical warts. Seventy-six women did not have smear testing during this study period because it was routine to check cytology after treatment and they defaulted. All female patients now have cytology at their first visit. While only one patient in our cohort had carcinoma-in-situ, it appears likely from the known tendency of cervical dysplasia to progress to more serious lesions that a similar progression may occur in this group.[19] In the present state of knowledge, it would appear that both women with anogenital warts and the female partners of men with anogenital warts should have at least annual cervical cytological screening as part of regular health care with further investigation of cytological abnormality.

The treatment of warts continues to be problematical. In this series 218 patients had 25% podophyllin applied as the sole treatment; of these 75% either responded completely or defaulted after one or two treatments, and a further 17% did so after three treatments. Because trichloracetic acid was used as an adjunct to podophyllin at our clinic, it was impossible to assess its use as an individual agent, but, when used in combination with podophyllin or cryotherapy,

86% of patients had four or less treatments. Cryotherapy similarly was an adjunct to therapy but was used four or less times in 93% of patients. It appears that successful treatment of warts is achieved in the great majority of patients with four treatments with these conventional agents and that further treatment beyond this point is often frustrating for the physician and futile for the patient. It was disappointing that cryotherapy of the urethral meatus was unsuccessful in 47% of patients after one treatment. This is in contrast with the high success rates of Ghosh who reports 20 of 28 patients (71%) with meatal warts cured by one application of cryotherapy alone.[20] This difference is unexplained, since a similar cryotherapy technique is used in both centres.

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Death rates from diabetes mellitus in Ireland 1833 – 1983: a historical commentary

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SUMMARY

A world-wide increase in diabetic deaths and a varying rate of increase between one country and another over the past hundred years has long been recognised. During the nineteenth century, the incidence of diabetes was low in Ireland as measured by mortality. Nevertheless, the rising trend found elsewhere was also apparent in Ireland. Recorded deaths were 0.22/100,000 of the population in 1840, rising to 13.2 by 1972. Most of the increase occurred between the 1880s and 1911, but only 15% of this can be accounted for by an ageing population. It is, therefore, necessary to seek other explanations. During the period, sugar and fat consumption in Ireland rose sharply. It has not been possible precisely to relate dietary causes to the incidence of diabetes, but the Irish experience suggests that such a link may exist.

INTRODUCTION

Sir William Wilde in his report on deaths written for the 1841 census of Ireland stated that the prominent diabetic symptoms of increased thirst and appetite and raised output of urine were frequently mentioned in the early Irish manuscripts. Of course, it is debatable whether these symptoms really were the consequence of diabetes mellitus or of some other disorder. Nevertheless, legend provides some colourful metaphors for the disease.

> *'Aire Luachra'*, the lizard of the rushes, . . . (and it) is believed by country people to enter the throats of persons who may happen to fall asleep in the open air, and taking its abode in the stomach, it there generates by its inordinate craving for food, . . . [1]

An historical study of the occurrence of diabetes is not possible before the nineteenth century and even then is plagued with difficulties since the data consist exclusively of mortality statistics. Such material is unreliable, particularly in the case of diabetes mellitus, as often the disease was not the proximate cause of death, and there is evidence to suggest that for some time in such cases diabetes was not recorded on the death certificate. Problems may also arise out of differences in definitions of diabetes. Yet mortality statistics are all we have, and they do indicate a trend. Both Joslin et al[2, 3] and Himsworth[4] used figures of diabetic deaths in their retrospective studies.

SOURCES

For Ireland the statistics extend back to the 1830s. These are found in two sources: the decennial Irish censuses, and the Annual reports of the

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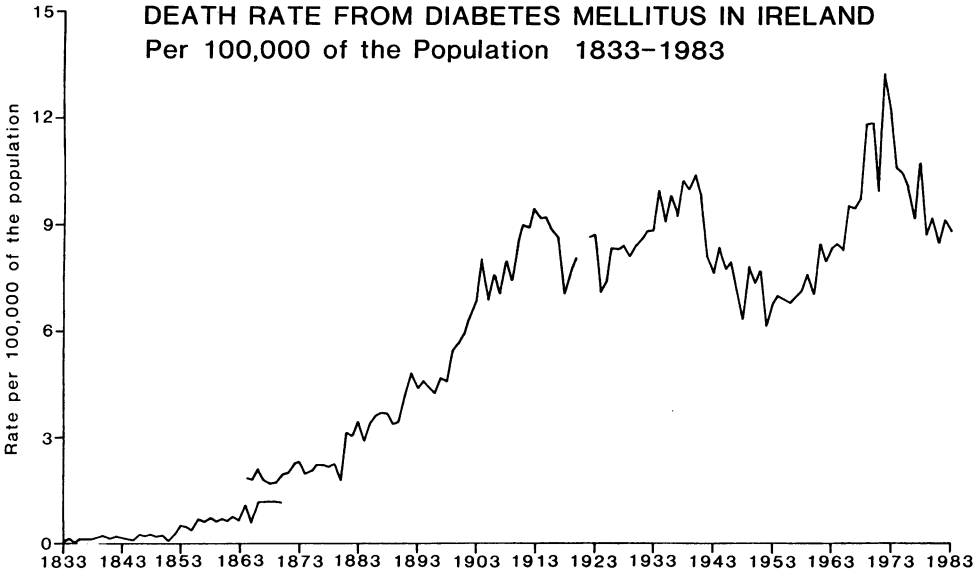
Registrar-General for Ireland. Four Irish censuses, 1841, 1851, 1861 and 1871 publish tables recording deaths from certain diseases including diabetes mellitus. (From 1881 the Census of Ireland no longer recorded number and causes of death). Since each census contained retrospective data for the preceding ten years we possess figures of diabetic deaths from 1831/2 to 1870. (In the 1841 Census of Ireland the years 1831 and 1832 were aggregated, and so they have been eliminated from the survey). From 1864 the Registrar-General was charged by statute with the responsibility for collecting mortality statistics, including causes of death, and these continue to the present day.[5] The Figure plots the death rates from diabetes mellitus in Ireland from 1833 to 1983 compiled from these two sources.

FIGURE



Before analysing the trends it is important to discuss the different ways by which these statistics were obtained. The census figures were extracted from the household returns, which were filled in by the head of the household or on his behalf. The Registrar-General's data, on the other hand, were compiled from death certificates, which had to be filled in by a medically qualified person. The different methods of acquiring the information probably explain the disparity during those years of overlap, 1864 – 1870. After 1871 the censuses no longer carried detailed information on causes of death.

TRENDS

Very low, though steadily increasing, numbers of diabetic deaths were recorded in Ireland between 1833 and 1880. Several dips in the trend are noteworthy. The drop in 1851 was probably a recording quirk in the aftermath of the Famine, and the fall in 1880 can be explained by a subsistence crisis during 1879/80 when deaths from other causes could have swamped diabetes mellitus. Between 1880 and 1881 the recorded death rate from diabetes rose sharply from 1.8 to 3.2/100,000, an increase which coincided with a change in disease classification in the Registrar-General's reports. From 1881 the diabetic death rate climbed

erratically to 9.5 in 1913. Over the next 28 years, the rising trend was interrupted at three points. The drop in 1918/19 probably reflects the influence of food shortages in the closing years of the First World War. The trough in the 1940s is for the same reason. The decline in 1924/25 is more difficult to explain. A fall also occurred in America at this time, insulin injection therapy being the cause. Whether this explanation will serve for Ireland cannot be stated with accuracy since we do not have a definite date for the first administration of insulin. But the most obvious feature of the inter-war years is the long-term stability of the diabetic death rate. The rate fell during the years of the war and post-war shortages before rising again in the late 1950s, reaching a new peak of 13.2/100,000 in 1972. Thereafter the trend has been downward with a number of small peaks in isolated years. The gap in the graph for 1921 is because no records were collected for the partition year.

The rise in the diabetic death rate between the 1860s and 1930s parallels a world-wide increase. While we do not have international statistics stretching back to the mid-nineteenth century, data from 1900 demonstrate the point (Table I).[2,4] A comparison of mortality from diabetes in Ireland with other countries shows that its position was at the bottom of the international league table. Furthermore, the pace of increase in Ireland was slow.

Diabetes is more prevalent among the elderly, and so an important factor which must be considered in our assessment of the Irish incidence is the changing age structure of the population which was ageing, principally because of emigration. In order to eliminate the effect of ageing, the rates have been standardised, using the 1861 age structure as the base. This adjustment, shown in Table II, revealed that by 1911 only 15% of the increase in deaths from diabetes mellitus can be explained by the changing age structure, leaving 85% to be explained in some other way.

TABLE I

Diabetic death rates in principal countries 1900 – 1930 (per 100,000)

	1900	*1905*	*1910*	*1915*	*1920*	*1925*	*1930*
United States	11.0	13.9	17.6	21.5	20.4	21.0	24.0
England and Wales	8.6	9.3	11.0	13.0*	10.0*	11.2	14.2
Netherlands	—	—	9.9	13.1	12.6	14.5	17.6
Italy	3.3	4.0	4.7	5.3	4.5	5.9	8.2
Australia	—	—	9.5	10.4*	11.5*	11.4	11.2
Japan	—	—	1.8	2.9	3.1	3.3	3.5
New Zealand	7.5	—	12.4	13.5	12.5	12.2	15.7
Scotland	7.2	—	9.7	11.4	9.0	9.4	12.5
Switzerland	5.6	—	—	7.6	7.7	9.2	11.0
Ireland	6.0	7.0	8.7	9.3	8.4	7.7	8.4
N. Ireland**	—	—	—	10.6	9.5	8.3	9.7
Irish Free State	—	—	—	8.9	7.6	7.4	8.0

Extracted from Joslin EP, Dublin LI, Marks HH, *Studies in diabetes mellitus* (Ref.2). p.761; Annual reports of the Registrar-General for Ireland, 1900 – 20; Annual reports of the Registrar-General for Northern Ireland, 1925 and 1930.

*Death rates based on civilian population.

**Antrim, Armagh, Down, Fermanagh, Londonderry and Tyrone, which became Northern Ireland after 1920.

TABLE II

Crude and age standardised diabetic death rates

	A. *Crude death rate*	*B.* *Age-adjusted death rate*	*Percentage A/B*
1864 – 8	1.9	1.9	100%
1869 – 73	2.1	2.0	95%
1881 – 3	3.2	3.1	97%
1889 – 93	4.2	3.9	93%
1899 – 1903	6.2	5.7	91%
1909 – 13	8.7	7.4	85%

ANALYSIS

Diet plays a major role in the treatment of diabetes mellitus,[6] but is it also a cause of the disease? Different dietary customs are found in different countries and, within countries, social classes have different food consumption patterns. Over the past half century dietary surveys have been carried out in many countries and some have been examined in an attempt to explain the diabeticity of particular nations and races. As a result a number of dietary theories have been advanced to account for the differing incidence of diabetes.[4, 7-10] One has focused attention on the fat content of the diet: the higher the level of fat the higher the incidence of diabetes.[4] Another has explored the relationship between the type of carbohydrate and diabetes, with the conclusion that increased incidence in diabetes has been the consequence of rising sugar consumption.[7] Still other research has questioned these hypotheses, and pointed instead to an excess of energy value over expenditure, causing obesity, which then predisposes towards diabetes.[10]

Can we explain the low rate of diabetic deaths in Ireland by dietary characteristics? Further, can the rising death rate from diabetes be attributed to changes in the Irish dietary pattern? Nutritional analysis of Irish diets is possible because of the existence of seven dietary surveys of labouring-class families, taken at intervals over a sixty-year period, 1839 – 1904.[11-17] Since labourers constituted a large section of the country's population and their dietary pattern mirrored on a reduced scale that of the rest of the population, these surveys provide us with a measure of the diet generally pertaining. Discussion will be restricted to the period before the First World War for it was then that the greatest increase in diabetic death rates occurred, and it was also when the most dramatic change in Irish dietary patterns took place.

Before the Great Famine, the Irish labouring classes subsisted on huge quantities of potatoes, washed down with skimmed milk, buttermilk or sometimes whole milk. Such a diet is very high in carbohydrate and exceptionally low in fat, particularly when skimmed milk or buttermilk were drunk. The Great Famine marked a distinct break with this monotonous régime, particularly for those living in the east and south-west. Greater culinary variety was established by the 1850s. Table III illustrates this increase in variety of fare by presenting typical dietary histories from two dietary surveys, one taken in 1839, the other 1863.

TABLE III

Weekly food rations of Irish labourers (Co. Tipperary) 1839 & 1863

1839 *(Per labouring man)*	*1863* *(For a seven-person family)*
63 lb. — 94½ lb. Potatoes	7 lb. Flour
21 pt. — 42 pt. Skimmed milk	56 lb. Indian meal
	140 lb. Potatoes
	1½ lb. Sugar
	2 lb. Butter
	2 lb. Meat
	56 pt. Skimmed milk
	14 Eggs

Source: *Sixth annual report of the Poor Law Commissioners, BPP1840 (245) XVII, Appendix D: 244, Sixth report of the Medical Officer of the Privy Council, BPP 1864 (3416) XXVIII: 324.*

In nutritional composition these two diets were very different. Turning to the country as a whole, Table IV provides a clear picture of the changing pattern in nutrient content which occurred in the diet of the Irish labourers between 1839 and 1904. The 1839 diet had a very high percentage of energy value (87%) derived from carbohydrate, all of which was starch, while the percentage of energy contributed by fat was exceptionally low at a mere 1%. Such a nutritional pattern closely resembles that of the Japanese,[4] and in both Ireland and Japan, when this configuration was present, the mortality rate from diabetes mellitus was low. By 1859 the energy value contribution of carbohydrate had fallen to 77%, whereas the fat contribution had increased tenfold. This trend continued so that by the opening years of the twentieth century the carbohydrate contribution had contracted even further to 66%, while the percentage of energy value obtained from fat had jumped to 24%. Ireland was then following a pattern evident elsewhere, with diabetes increasing.

TABLE IV

Nutritional analysis of the Irish labourers' diet 1839 – 1904

		Protein		*Fat*		*Carbohydrate*		*Energy value*
	No	*g.*	*% of Kcal*	*g.*	*% of Kcal*	*g.*	*% of Kcal*	*Kcal*
1839 Survey*	13	135	12	4	1	1099	87	4720
1859 Survey	161	105	12	40	10	760	77	3682
1863 Survey	52	111	11	40	9	843	79	4008
1904 Survey	190	82	10	87	24	590	66	3370

*Dietary analysis is based on a menu of potatoes and skimmed milk. When whole milk is substituted for skimmed milk the contribution of fat to energy value rises to only 5%.

Another important dietary change to occur from the closing decades of the nineteenth century was the increase in consumption of sugar among the labouring classes. The evidence comes from two sources. The dietary surveys reveal that daily adult consumption rose tenfold between 1859 and 1904. Imports of sugar into Ireland tell the same story. Published figures for trade into the principal Irish ports indicate an almost eightfold increase in sugar imports between 1889 and 1914 (See Table V).[18] If the suggested connection between rising sugar consumption and the rising incidence of diabetes is correct, then the growing wealth of nineteenth century Ireland brought to its people a disease of affluence.

TABLE V

Imports of sugar at principal Irish ports in cwts

1864 – 1868	98,073	1889 – 1893	366,362
1869 – 1873	158,322	1894 – 1898	1,020,863
1874 – 1878	188,850	1899 – 1903	1,490,866
1879 – 1883	181,211	1904 – 1908	2,691,665
1884 – 1888	143,268	1909 – 1913	2,800,701

Source: *Trade and navigation annual statistics for Ireland.*

In conclusion, deaths from diabetes mellitus rose during the second half of the nineteenth century, the greatest increase occurring during the closing decades and into the twentieth century. Parallel with this rise were significant dietary changes. Total carbohydrate intakes fell, while sugar and fat consumption increased. Which, if any, of these factors was the cause of the increased incidence of diabetes mellitus remains a major epidemiological question, and will require prospective as well as retrospective historical study.

I am indebted to Dr D R Hadden and Professor L A Clarkson for their advice in the preparation of this paper.

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Antenatal care in Belfast

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SUMMARY

A questionnaire study of mothers' views of the antenatal care provided in Belfast showed general satisfaction. Retrospective examination of their charts however showed in some cases that insufficient attention was paid to the medical and obstetric history in the selection of type of care made by the women and their doctors. Some women with high risk factors were booked for shared care and some patients at low risk were booked for total hospital care. The reasons for this are unclear.

The mothers felt that continuity of care and communication at the health centre were better than at the hospital. Analysis of the number of hospital attendances showed that shared care patients appeared to be making an excessive number of visits to hospital. Many total hospital care patients also admitted that they were attending their general practitioners. There appeared to be marked duplication of effort as a result of poor communication between patient, general practitioner and hospital.

Alternative ideas for care are suggested — a more integrated system for sharing antenatal care, and the development of general practitioner units within the specialist obstetric hospital.

INTRODUCTION

Obstetric practice in Belfast has undergone major changes in the recent past, the more important being the closure of all the general practitioner maternity units and the subsequent transfer of all confinements to specialised hospital obstetric units. General practitioners, however, continue to provide antenatal and postnatal care in a shared care system. Patients are able to choose the type of care they wish to have — total hospital care, shared antenatal care or private care — but should expect medical advice in making the choice. This study was undertaken to examine the different types of antenatal care provided in Belfast health centres and maternity hospitals, to look at the reasons for a particular choice being made and to assess the opinions of a sample of women on the present types of care available. It was part of a larger study mainly looking at health education topics.[1-4]

PATIENTS AND METHODS

One group practice from each of the eleven health centres in Belfast was invited to take part. The health centres covered both affluent and deprived areas of Belfast, thus providing a representative sample of women. The general practitioners were

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asked to identify all their pregnant patients from July 1982 and 70 per cent were randomly selected in order to produce a sample size of 500 patients. Permission for inclusion in the study was obtained prior to interview. The obstetricians in the four Belfast maternity hospitals gave permission to the author to make a historical search of the obstetric records 2 – 3 weeks after delivery.

The fieldwork research assistant (a health visitor, funded by the Friar Fund, Faculty of Medicine, QUB.) interviewed the patients in their own homes and completed questionnaires after the hospital booking visit and again approximately 8 – 10 weeks following delivery.

The information was coded and transferred to punched cards. All data processing and statistical analysis was performed using the Statistical Package for the Social Sciences on the Queen's University ICL 2900 computer. The conventional level of significance ($p<0.05$) was used for all statistical comparisons. The chi squared test was used to compare different groups.

Of the initial sample of 500 patients, 380 questionnaires were completed. Twenty-nine patients miscarried and twelve questionnaires were never completed because the patients moved away from Belfast during the study. Seventy-nine patients were unwilling to be interviewed. Forty-six of these came from two health centres situated in West Belfast. The remaining thirty-three who were unwilling to be interviewed were evenly distributed among the other nine health centres. Thus there was a potentially substantial response bias.

The first report of the Maternity Services Advisory Committee[5] identified two categories of medical risk factors. First, those women with a predicted high risk in both pregnancy and labour, (high, high risk). This might have been due to obstetric causes (history of spontaneous abortion, premature labour or low birthweight baby, stillbirth or neonatal death) or medical causes, such as diabetes or hypertension. The Committee suggested that these women needed specialist supervision of pregnancy and labour with delivery in a consultant maternity unit. The second category was women with a predicted high risk in labour (low, high risk), such as those with a small pelvis or multiple pregnancy. They suggested that these women did not need specialist antenatal care throughout pregnancy, but that specialist care should be arranged for the confinement.

The final responsibility for the type of care in Belfast rests with the consultant obstetrician after discussion with the patient and possibly the general practitioner. This medical influence on the choice of care should lead one to expect that the majority of low risk patients would have shared care, and higher risk patients total hospital care. The women in this study were placed retrospectively in the above categories according to their past history recorded in their obstetric charts.

RESULTS

Choice of care

Choice of care was defined as the type of care the patient understood she was to have after she had been to the hospital for her initial booking visit. Seventy-seven patients said they were having total hospital care, 290 were having shared care, 12 patients were attending a consultant privately and one patient had arranged to have total GP care. The latter 13 were excluded and the final comparison was confined to the two main groups — total and shared care. When the patients themselves were asked the reason for their choice of care, the majority choosing hospital care said they chose it because it was safer or more convenient; the

majority choosing shared care said it was because their GP had suggested it, it was less time consuming or more convenient. Patients themselves did not consider risk factors in the choice.

Table I shows the initial choice of care for patients in 'high, high risk', 'low, high risk', and 'low, low risk' groups. There was no significant difference in the choice of care made by the women in the three categories, so that the assumption that selection of care is made on grounds of risk appears to be unsupported in many cases.

TABLE I

Initial choice of care sub-divided retrospectively into high/high, low/high and low/low risk

	High/High	*Low/High*	*Low/Low*	*Total*
Total hospital care	26 (25.0%)	17 (19.8%)	34 (17.9%)	77
Shared care	76 (73.1%)	69 (80.2%)	145 (76.7%)	290
Private care	2 (1.9%)	0 (0.0%)	10 (5.4%)	12
TOTAL	104 (100%)	86 (100%)	189 (100%)	379

One patient (low/low risk) had total GP care.
Omitting private care $X^2 = 1.753$, $df = 2$, $0.50 > p > 0.30$.

The type of care actually received by each patient during her pregnancy was assessed retrospectively at the second interview (Table II). More patients actually received hospital care (105) than had initially chosen it (77).

TABLE II

Type of care finally received

Type of care chosen initially	*Total hospital*	*Shared*	*Private*	*Total GP*	*Total*
Total hospital	73	3	1	0	77
Shared	31	256	3	0	290
Private	1	0	11	0	12
Total GP	0	0	0	1	1
TOTAL	105	259	15	1	380

Communication

All the shared and total hospital care patients had been booked at hospital initially. They were asked if they felt that informal discussion was encouraged at the clinic, and if there was time to ask questions of both the doctor and the midwife. Eighty per cent of hospital care patients and 73 per cent of shared care patients felt that informal discussion was encouraged at the hospital booking clinic. Seventy-eight per cent of hospital care patients and 70 per cent of shared care patients felt that there was time to ask the doctor questions and somewhat higher proportions that there was time to ask the midwife questions. When the patients were asked about communication at subsequent hospital antenatal clinics, the responses

were slightly less positive, but 93 per cent of shared care patients were satisfied with the level of communication at their antenatal visits to the health centre.

The women were specifically asked if they were given explanations for certain clinical procedures which were carried out on them. From their responses it would appear that explanation of the reasons for blood tests was particularly poor for both total and shared care groups. Nearly half of the women said no reason was given. Explanation of the internal examination was better — about 70 per cent of the women were satisfied about the explanation for this procedure. The staff involved in carrying out the ultrasound examination were the most effective. They satisfactorily communicated the reason for the examination to 90 per cent of the patients.

Convenience and efficiency of clinics

At the second interview almost 90 per cent of both groups said that the hospital antenatal clinic was convenient in terms of distance and the time of appointment. However, only 33 per cent of hospital care patients and 42 per cent shared care patients said that they were seen on time at hospital.

Over 96 per cent said the health centre was convenient in terms of distance and 94 per cent that the appointment times were convenient. In contrast to the hospital, 83 per cent shared care patients said they were seen on time at the health centre.

Continuity of care

Table III shows that there is generally poor continuity of care at hospital antenatal clinics, but a greater percentage of the hospital care patients than of the shared care patients said they usually saw the same doctor. There was no difference between the groups in the percentage seeing the same midwife at each visit. Over 94 per cent of shared care patients said they usually saw their own GP at the health centre and 93 per cent of shared care patients usually saw the same midwife.

TABLE IIIa

Response to question 'Did you see the same doctor at each hospital visit?'

	Total hospital care patients	*Shared care patients*
Usually	36 (37.1%)	39 (15.1%)
Rarely	29 (29.9%)	103 (40.0%)
Never	32 (33.0%)	116 (44.9%)

TABLE IIIb

Response to question 'Did you see the same midwife at each hospital visit?'

	Total hospital care patients	*Shared care patients*
Usually	37 (35.2%)	65 (25.1%)
Rarely	36 (34.3%)	94 (36.3%)
Never	32 (30.5%)	100 (38.6%)

(Nine patients attended the midwife's couch only)

Attendance at hospital antenatal clinics

The number of hospital visits is shown in Table IV. There was a significant difference between the two groups, as expected, but 122 (47.1%) shared care patients attended hospital seven or more times and 35 (13.5%) ten or more times. Thus many shared care patients were attending hospital more frequently than the usual 5 – 6 visits. This may have been related to the development of maternal complications in 104 (40.2%) of the shared care patients. However, 47 (45.2%) shared care patients with maternal complications made less than seven visits to hospital. For patients classified as clinically 'at risk' or 'not at risk' according to the criteria mentioned previously, there was no significant difference in attendance between the groups.

TABLE IV

Number of hospital clinic attendances by patients according to the final type of care received

	Total hospital care patients	*Shared care patients*
<7	12 (11.4%)	137 (52.9%)
7 – 12	61 (58.1%)	114 (44.0%)
>12	32 (30.5%)	8 (3.1%)
TOTAL	105 (100%)	259 (100%)

$X^2 = 85.460$, df = 2, $p < 0.001$

Forty-seven hospital care patients also attended their general practitioners for antenatal care or advice; 21 women (20%) said they attended between four and eight times, and eight (7.6%) said they attended their general practitioner more than eight times.

Postnatal care

Attendance at the postnatal clinic is poor compared with antenatal attendance. Only 264 (69%) of hospital and shared care patients had attended for postnatal examination by the time they were interviewed 8 – 10 weeks after delivery and the result was similar in both groups. Sixty-four (36%) shared care patients who had a postnatal examination had attended hospital rather than their health centre for the examination and 28 (37%) of hospital care patients who had had a postnatal examination had attended their health centre rather than hospital. Of the 64 shared care patients who attended hospital, 29 (45%) had no complications which might have justified hospital follow-up. Similarly, of the 28 hospital care patients who attended the health centre, 11 (39%) had had complications which might have justified a hospital follow-up.

DISCUSSION

The majority of patients said they were satisfied with the antenatal care they received. However, there does appear to be room for improvement in communication between staff and pregnant women at hospital antenatal clinics, and an even greater need for explanation of procedures carried out. More women attending their health centres were satisfied with the time allowed for informal

discussion and questions and most of them said they were seen on time. Similar results have been found in other studies.[6-9]

An aim of this study was to examine the reasons why pregnant women made a particular choice of care and what factors might have influenced them. The analysis of these findings leads one to question the system of shared care in Belfast in its present form. There was almost no difference in medical terms between the group of patients having total hospital care and the group having shared care, and the decision to go to the general practitioner or to hospital was based on convenience, time factors and other emotional feelings with only occasional reference to possible risk factors. It is widely accepted that predicting the outcome of pregnancy is extremely difficult,[10] but more effort should be made to identify high and low risk patients both from medical and equally important social factors.[11] The results of this study suggest that the guidelines recommended in the First report of the Maternity Services Advisory Committee 1982[5] and in the Baird Report 1980[12] are not always being followed. Some women at low risk are being booked for total hospital care and then, at subsequent visits, being seen only by midwives when they could equally well be looked after by community midwives and general practitioners. More careful planning could help to reduce the overcrowding of already busy hospital antenatal clinics.

Lack of continuity of care in hospital antenatal clinics is a common criticism. The high rate of turn-over of junior hospital doctors and midwives, particularly in teaching hospitals which all these patients attended, sometimes prevents the establishment of good staff/patient *rapport,* and contributes to the lack of communication. Poor communication is a criticism which can also be directed at many general practitioners in that they often do not include in the referral letter information about the patient which is important for the obstetrician. Co-operation cards, although widely used, contain the minimum of information and are an inadequate form of communication. When shared care patients are admitted to hospital during a routine antenatal appointment and then subsequently taken over by the hospital team, the general practitioner is often not informed. The Royal Maternity Hospital has very recently started to allow its patients to carry their own antenatal records between hospital and general practitioner. It is hoped this will improve communication.

The excessive number of antenatal visits made by shared care patients and the over-subscribing of hospital postnatal clinics is another possible reflection of lack of communication and continuity of care. It could also be a reflection of lack of confidence of obstetricians in the standard of antenatal and postnatal care given by general practitioners.

Although only 10 per cent of patients said they were officially transferred to total hospital care, the amount of visits which some other shared care patients made would suggest that the number was much higher. In general practitioner units where selection policies are strictly adhered to, about 30 per cent of patients are transferred to specialist care.[13, 14, 15]

One major problem is the sheer volume of work which hospitals attract and it has been suggested that many of the criticisms of the service could be met if a proportion of the work could be undertaken elsewhere. A closer examination of the service could lead to significant improvements and greater satisfaction for the women and the professions concerned. There are many financial pressures on the National Health Service and it has been suggested that many improvements could be made by changes in attitude and reorganisation of procedures which do

not involve additional expenditure. The obstetrician is a scarce resource; the skills of the consultant team should be devoted primarily to the care of those women in greatest need of specialist advice. There is a need for greater flexibility in the use of the professions who undertake antenatal care. An integrated specialist and general practitioner service such as that described by Zander and colleagues from St Thomas's Hospital Medical School [16] or low risk obstetric care and confinement in a general practitioner maternity unit within a maternity hospital as described by Roseveare and Bull [13] could be considered.

The results of this study do not justify criticism of the medical care that individual patients received. There is no evidence here to suggest that certain patients might have done better with a different type of care from the one they received. It is criticism of the way the system is run rather than of the service provided. Shared care as it exists in Belfast cannot be tidily defined. Interpretation often rests with the obstetrician who may apply differing criteria according to personal knowledge of individual practitioners. If improvement is desired, it is up to general practitioners and the community team to meet with the obstetricians and hospital team to work out a new and better system.

I am grateful to Professor W G Irwin, Head of the Department of General Practice, for his support throughout the study and to Dr Desmond Merrett, Head of the Department of Medical Computing and Statistics, for his invaluable help with the statistical analysis of the data. I am also grateful to Mrs Nuala Dowds and Mrs Heather Stewart for interviewing the women and completing the questionnaires so accurately. I thank all the general practitioners and obstetricians who allowed us to interview their patients and gave me access to their records.

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Invited commentary.

The editor has requested this comment from Professor W Thompson in view of the controversial nature of some of the conclusions.

The main criticism of this paper is the interpretation by the author of the guidelines for shared care set down in the First report of the Maternity Services Advisory Committee published in 1982. The author has interpreted these guidelines that patients with a high-risk past history must undertake continuous surveillance by a hospital team. Most obstetricians who work in the area covered by this paper interpret the guidelines in a different manner. Patients identified as having a high-risk past history must be booked for confinement in the consultant obstetrician's unit, but their antenatal care can have intermittent surveillance by a hospital team. This makes some of the conclusions of the report difficult if not impossible to assess. I feel it would have been better for the author to have clarified the criteria for shared care from the consultants concerned prior to the study.

However, the paper is interesting in that it highlights the duplicity of examinations and investigations on antenatal patients, the poor communication at times between the general practitioner and the mother, and the occasional haphazard approach to organising antenatal visits for hospital patients.

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Radiation therapy of cancer of the uterine cervix in Northern Ireland

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SUMMARY

From 1976 to 1980, 275 patients with invasive uterine cervical cancer were treated at the Northern Ireland Radiotherapy Centre. Most patients had combined intracavitary and external radiotherapy. Only 26% presented with clinical Stage 1 disease; there were more of these patients aged 30–39.

Five-year survival was 68% for Stage 1, 48% for Stage 2, 16% for Stage 3 and 0 for Stage 4. Survival was better in the age group 30–39 (63%) than in the age group 20–29 (18%) and for those histologically graded as squamous (49%) rather than poorly differentiated (35%). Twelve patients required colostomy. Comparison of these results with other centres in the United Kingdom and the USA show that there is still room for improvement particularly in the identification of early stages of the disease.

INTRODUCTION

For the radiotherapist, cancer of the uterine cervix represents one of the major anatomical sites where treatment is primarily by radiation, and usually undertaken with curative intent. This review describes the population of patients in Northern Ireland and the outcome of treatment over the years 1976 – 80, during which time it was policy to treat invasive cervical cancer by radiation alone. Only rarely was radiation combined with surgery. When summarised along with two previous reports dealing with the years 1953 – 60 [1] and 1968 – 72,[2] the practice of almost three decades is available.

PATIENTS AND METHODS

During the study period 275 new patients were treated at the Northern Ireland Radiotherapy Centre. For three patients there was inadequate initial documentation or early loss to follow-up, and the remaining comments relate to 272 patients. In a small minority, follow-up was completed by use of a specially designed questionnaire. Staging of disease was by clinical examination, almost always under anaesthesia, complemented by intravenous pyelogram, according to the FIGO classification.

With no pretension of systematic histological grading, this study separates those cancers described simply as squamous or moderately differentiated squamous, from those specifically described as poorly differentiated, Grade 3, or anaplastic.

The vast majority of patients had combined intracavitary and external radiotherapy. Intracavitary treatment was according to the Manchester system of

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radium dosage with a central tube and paired lateral vaginal ovoids separated by a washer. No attempt at individual dosimetry was made, and dose calculations depend on the assumption of ideal geometrical distribution. In these patients the radium insertion was of 48 or 60 hours duration, corresponding to approximate Point A doses of 30Gy and 37Gy respectively. When radium alone was used for Stage 1 cases, two insertions totalling 120 hours were employed. External pelvic therapy was delivered on megavoltage equipment (Cobalt 60 or 8MeV linear accelerator). Practice varied slightly between clinicians but generally involved treating the whole pelvis by opposed anterior and posterior fields to 40Gy in 20 fractions over four weeks, or using a four-field pelvic box technique to spare the posterior rectum delivering 40Gy in 13 fractions treating three days per week. There was no uniform policy as to the order in which external and intracavitary treatments were applied. Central pelvic shielding during external therapy was used only occasionally.

RESULTS

Patient numbers and age distribution are given in Table I. Average annual numbers were higher in the period 1968–72 but no obvious explanation was available. There has been no significant change in the age distribution when compared with the two previous reports[1,2] and the national figures for 1971.[3] The increased incidence of invasive cervical cancer in the youngest age group reported in the NW Thames region[4] was not identified in this report.

TABLE I

Age distribution (per cent) of patients with cancer of the uterine cervix during three time periods in Northern Ireland. Figures for England and Wales, and for the NW Thames region are shown for comparison

	Northern Ireland			*England and Wales*	*NW Thames region*
	1953-60[1]	*1968-72*[2]	*1976-80*	*1971*[3]	*1975-78*[4]
Patient numbers	520	408	272	4090	1175
Age group					
20–29	0.6%	2.5%	11 4.0%	3.1%	13.5%
30–39	10.8%	5.6%	27 9.9%	8.8%	21.6%
40–49	28.3%	26.0%	47 17.3%	22.7%	13.6%
50–59	28.1%	31.6%	78 28.7%	28.7%	19.2%
60–69	21.4%	20.5%	70 25.7%	19.8%	17.4%
70+	10.9%	13.7%	39 14.3%	17.0%	14.3%

In the present study 71 patients had Stage 1 disease, 124 were Stage 2, 67 Stage 3 and 10 Stage 4. There has been a trend towards more patients presenting as Stage 1, from 17% in 1953–60 through 21% in 1968–72 to 26% in 1976–80, but no change in the high proportion presenting as Stage 2 and 3 disease. Within the age groups the stage distribution closely matches the overall pattern except in the 30–39 age group where 16 of 27 patients (59%) presented

as Stage 1. All the survival statistics given are mature five-year figures representing patients alive and free from disease. This allows direct comparability with previous reports. No patient in this study who was alive and disease-free at five years has subsequently relapsed or died from her disease. Overall, 118 of 272 patients (43%) were alive at five years. Results by stage groups are shown in Table II.

TABLE II

Five-year survival for the group of 272 patients diagnosed 1976 – 80, classified by stage groups

Stage	*1*	*2*	*3*	*4*	*Total*
Number of patients	71	124	67	10	272
Died from carcinoma	16	57	53	10	
Died from unrelated cause	8	7	3	0	
Five-year survivors	47	60	11	0	118
	(67.6%)	(48.4%)	(16.4%)	0	43.3%

For the 71 Stage 1 patients, 71% of 39 treated by combined therapy, and 67% of 21 receiving radium alone became long-term survivors. All three patients who had pre-operative radium insertions survived as did three out of six patients who had combined therapy post-operatively. Of the 124 Stage 2 patients, 51% of 103 patients treated with combined therapy survived five years or more. One patient treated pre-operatively survived, as did five out of eight who were suitable for radium treatment alone, but of 12 patients who were only suitable for external radiotherapy only two survived five years.

When analysed by age-group, the only improvement over the average results is in the 30 – 39 group, who had 63% survivors. This relates to the high proportion of Stage 1 cases in this group. Only two of 11 patients aged 20 – 29 survived five years.

Although the allocation of histological grade was not as consistent as would be desirable, percentage survival figures are presented in Table III. There is a difference in survival for those reported as squamous and as poorly-differentiated tumours both overall and in Stage 2. This suggests that the histological grade is of prognostic value.

TABLE III

Five-year survival by histological type

Histological type	*Overall*	*Stage 1*	*Stage 2*
Number of patients	272	71	124
Squamous	80 of 165 (49%)*	34 of 47 (72%)	40 of 72 (56%)+
Poorly differentiated	29 of 83 (35%)*	9 of 18 (50%)	15 of 40 (38%)+
Adenocarcinoma	10 of 24 (42%)	4 of 8 (50%)	5 of 12 (42%)

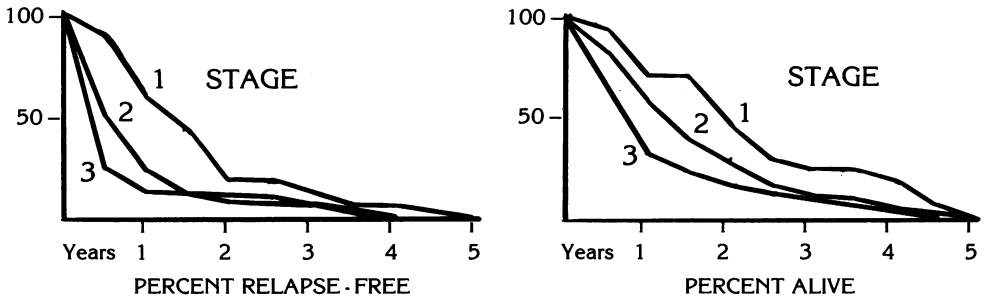
*Chi squared = 5.58, $p < 0.05$ (with Yates' correction)

+ Chi squared = 4.20, $p < 0.05$ (with Yates' correction)

The late effects of treatment have been assessed only in those patients surviving five years, to exclude confusion between treatment-related morbidity and the symptoms of recurrent or progressive disease. Eighty patients (67%) had predictable treatment morbidity which had completely settled by their first review at two months. In 17 (14%) symptoms persisted for up to one year. Fifteen of the survivors (12.6%) suffer at least occasionally from symptoms mainly of rectal irritation, and seven (5.9%) have a permanent colostomy. Within the entire group of 272 patients there were five further colostomies in patients who ultimately died from their disease, giving an overall colostomy rate of 4.4%. Of the 12 patients requiring colostomy, seven had received the higher intracavitary dose of 60 hours radium with 40Gy pelvic irradiation, three followed double intracavitary insertions and two had had pelvic irradiation by suboptimal regimens.

FIGURE

Times to relapse and death for 136 patients who died from carcinoma of the cervix



For each patient who ultimately died from her disease, the period in months to failure and to death was recorded. Failure is taken as the time of onset of symptoms due to recurrent or metastatic disease or the detection at follow-up examination of such disease. It was not possible to specify whether recurrence was primarily local, pelvic or metastatic (Figure). The pattern of relapse is predictable over the first 18 months, with less time to relapse or death with advancing stage. Beyond that, as each group yields a few late failures, the graphs tend to merge. Prognostic information can be extracted, for example 80% of Stage 2 cases, who will ultimately relapse, have done so within one year and are dead within two years.

DISCUSSION

This report demonstrates the results achieved in routine treatment of cervical cancer in Northern Ireland. While the five-year survival figures bear comparison with other UK results at the same period of time they undoubtedly fall below current expectation where the anticipated cure rates would be 80% in Stage 1, 60% in Stage 2 and 30 – 40% in Stage 3. The survival times for patients treated by radiotherapy for several major centres are shown in Table IV.

It would be unrealistic to seek increased cure rates simply by increasing the radiation dose, which would cause a disproportionately greater increase in morbidity. Policy changes incorporated in current technique will, it is hoped, result in a reduced morbidity with no loss of disease control. These changes

TABLE IV

Survival comparisons (per cent) for patients treated by radiotherapy

		Stage			
		1	*2*	*3*	*4*
Northern Ireland					
1953 – 60 [1]		57%	46%	25%	12%
1968 – 72 [2]		69%	46%	26%	
1976 – 80		68%	48%	16%	
United Kingdom					
Royal Marsden	1962 – 70 [5]	74%	46%	16%	6%
Manchester	1961 – 65 [6]	73%	52%	25%	
Cardiff Cathetron	1974 – 77 [7]	77%	50%	25%	
USA					
Patterns of Care Outcome Study [8]					
All centres		87%	66%	28%	
Major centres		92%	77%	60%	
Worldwide [9]					
118 institutions; 26 countries (1959 – 63)		77%	56%	32%	9%

include adoption of a manual afterloading system of intracavitary treatment with caesium 137 in place of radium which allows improved positioning and packing. A fairly modest Point A dose of 32Gy is given from the intracavitary treatment. External radiotherapy is given in conventional daily fractions of 2Gy and central pelvic shielding is used more often during the last quarter of the external treatment. If these improvements produce lower morbidity levels at no expense in cure rates, then increased dosage could be applied. Radical hysterectomy has been revived in the 1980s for selected Stage 1 cases in younger patients, and results of this operation will require close comparison with conventional radiotherapy.

The findings indicate that patients treated for this disease in Northern Ireland are very similar to those in the rest of the UK in terms of age, stage at presentation, histology and outcome. In the under-30 age group described here, the poor survival is closely related to relatively advanced stage at presentation and worse than average histology. A similar poor survival in young patients was also noted in patients treated on the Cathetron at the Middlesex Hospital.[10] Here relapse-free-survival in the over-35s was 87% compared with only 17% in the under-35s. Such data adds to the current debate as to how best to screen and treat these young patients who suffer from what appears to be a particularly aggressive disease.

The impact on prognosis of the histological grade, despite the absence of any systematic approach in the Northern Ireland data, suggests that this aspect should be more closely investigated.

Two major aspects of control failure are seen. First the problem of achieving local control in bulky Stage 2 and Stage 3 presentations, and the more unpredictable group of patients who can achieve local pelvic control, but whose disease behaves

with a more aggressive metastatic tendency. The first problem can be dealt with, as most commonly in the USA,[11] by considerable increase in the total pelvic treatment dose but this will produce major morbidity rates greater than 20%. Experience in the UK of acute and late complications of such increased dose render this policy largely unacceptable. The second, or indeed either group, could theoretically benefit from chemotherapy for bulk reduction or eradication of occult metastases. Experience with existing agents in advanced or recurrent disease[12] shows that only a minority of patients demonstrate any response and that very few get a complete response which even then cannot be maintained. There seems no place at present for the routine incorporation of toxic chemotherapy in the primary management of cervical cancer. More productive will be the enhancement of programmes to detect the disease at its earliest stage to ensure control by proven local measures.

Patients in this study were treated by Dr B D Burrows, Dr A R Lyons and the late Dr G A Edelstyn. The author wishes to thank Dr B D Burrows for his encouragement to undertake this project, Professor W S B Lowry for his helpful criticism, and Miss Anne Wilkie for her patient processing of the manuscript.

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Perforated duodenal ulcer: which operation?

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SUMMARY

Between January 1968 and December 1977 a total of 230 patients with a perforated duodenal ulcer underwent emergency operation in the Royal Victoria Hospital. Simple suture closure of the perforation was carried out in 205, and in the remaining 25 a definitive ulcer procedure was performed in addition. Four patients died following operation, a mortality rate of 1.7%.

During a mean follow-up period of 10.3 years at least 107 patients (52%) who had simple suture closure of their perforation developed further ulcer symptoms. Of these, seven re-perforated and a further 56 required elective definitive ulcer surgery. A strong case can be made for a definitive ulcer operation at the time of emergency surgery for a perforated chronic duodenal ulcer.

INTRODUCTION

Simple suture of the perforated duodenal ulcer has been the standard operative treatment since its acceptance as such in the early part of this century. The principle upon which this form of management is based was well described by Graham in 1937 when he stated that 'We have no responsibility to such patients but to save their lives' and 'We have no responsibility during the surgery to carry out any procedure designed to cure the patient of his original duodenal ulcer'.[1]

Since then, however, many authors have reported a high incidence of recurrent symptoms and indeed a significant incidence of life-threatening complications after simple suture of a perforated duodenal ulcer. Illingworth in 1946 reported a relapse rate of 70% and a 20% incidence of major complications in a large group of such patients followed up for five years.[2] Similar high relapse rates have more recently been confirmed by others[3,4,5] and some would now consider that a definitive procedure should be combined with simple suture as the emergency operation of choice.[6,7] It therefore seemed appropriate to review the surgical management of perforated duodenal ulcer in this hospital, to document the incidence of subsequent ulcer symptoms and, in the light of these findings, to reassess our operative strategy in these ill patients.

MATERIALS AND METHODS

During the 10-year period January 1968 to December 1977, 230 patients underwent emergency operation for perforated duodenal ulcer in the Royal Victoria Hospital. This particular time period was chosen so that, on follow-up, a minimum period of five years had elapsed since operation for each patient. The

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hospital records of these patients were obtained and clinical profiles, including pre-operative radiological findings, operative procedures performed and post-operative complications were documented. Follow-up information on patients was obtained by questionnaire, telephone interview and communication with the patients' general practitioners. Complete follow-up data were not available for 19 patients who had moved overseas, nine who declined to provide the information requested and 28 who could not be traced via their last recorded address, general practitioner or the Central Services Agency. Full information was available for 174 patients including 30 who had died during the period of follow-up.

RESULTS

Of the 230 patients in the study 200 were male and 30 were female (6.7:1). The age distribution in both sexes is shown in Fig 1. Of 217 patients for whom the information is recorded, 55 (25%) had no dyspeptic symptoms prior to perforation and a further 12 had been symptomatic for less than three months. By conventional definition, therefore, 67 patients (30.9%) perforated an acute ulcer. In the remaining 150 patients with chronic ulcers the mean duration of symptoms was 98 months (range four to 420 months). The majority (64%) of the entire patient group were regular cigarette smokers. Of 191 patients with clear documentation of pre-operative abdominal radiology, 133 (69%) had free intra-peritoneal gas on the erect film.

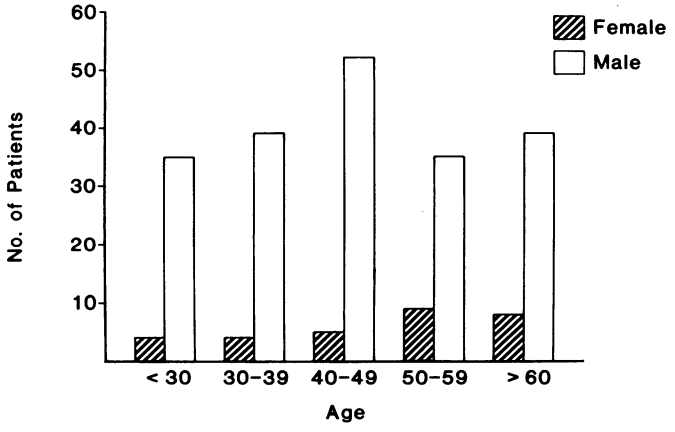

Fig 1.
Age and sex distribution of 230 patients with perforated duodenal ulcer.

The operative procedure performed in 205 patients was simple suture of the perforated duodenal ulcer, usually incorporating an omental patch. In the remaining 25, simple suture was combined with a definitive ulcer operation — truncal vagotomy and pyloroplasty in 13, truncal vagotomy and gastro-jejunostomy in 10, selective vagotomy and gastro-jejunostomy in one and truncal vagotomy without a drainage procedure in one patient.

A total of 16 patients had undergone previous peptic ulcer surgery — six had simple suture of a perforated duodenal ulcer and 10 had required definitive elective operation in the form of truncal vagotomy and gastro-jejunostomy (nine patients) and truncal vagotomy and pyloroplasty (one patient). These patients all perforated a chronic ulcer and were treated by simple suture closure with a re-vagotomy being added in one.

Post-operative complications including chest infection, wound infection, residual intra-peritoneal abscess, and thrombo-embolism were recorded for the entire patient group. The incidence of complications in the group of patients who underwent simple suture and in those who had a definitive procedure performed is recorded in the Table. Less frequent complications which arose in individual patients included bleeding per rectum (due to hypercitrataemia/hypocalcaemia), haematemesis, stomal intussussception, superficial thromboplebitis, pyloric obstruction, delirium tremens, acute urinary retention, and bacteraemia. In total 60 patients (29.3%) who had simple suture carried out and four (16.0%) of those who underwent a definitive procedure developed complications.

TABLE

	Simple suture (n=205)	*Definitive surgery (n=25)*
Chest infection	24 (11.7%)	2 (8.0%)
Wound infection	18 (8.8%)	1 (4.0%)
Wound dehisence	9 (4.4%)	0
Subphrenic abscess	6 (2.9%)	0
Pelvic abscess	4 (1.9%)	0
Thrombo-embolism	1 (0.5%)	0
Other	19 (9.3%)	2 (8.0%)

Post-operative follow-up ranged from five to 15 years, mean 10.3 years. During the follow-up period, 107 (61%) of the 174 on whom complete information was available developed further symptoms related to their duodenal ulcers. With the exception of 13 all had had symptoms for longer than three months prior to perforation. All of these 107 patients had simple suture of their perforation at the initial operation. The symptom-free period following perforation ranged from zero to 12 years but the vast majority experienced recurrent symptoms within the first three years, mean 20.6 months (Fig 2). In seven patients (4.0%) re-perforation occurred. In a further 56 patients (32.2%) persisting or recurring dyspeptic symptoms necessitated elective definitive ulcer surgery.

Four patients died immediately following operation, a mortality rate of 1.7%. All of these patients had simple suture closure of the perforation. Of the four post-operative deaths two occurred in frail elderly men with no previous

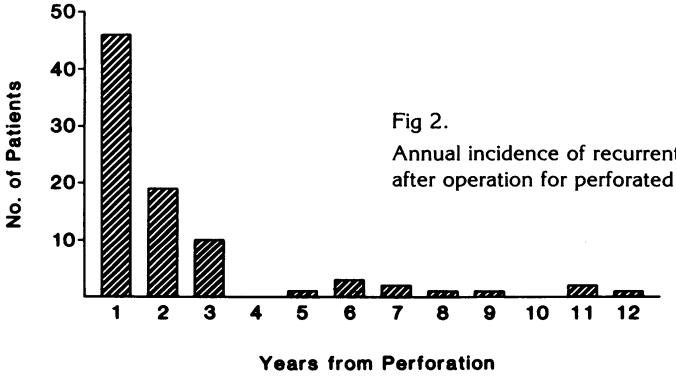

Fig 2.
Annual incidence of recurrent ulcer symptoms in patients after operation for perforated duodenal ulcer.

symptoms or precipitating factors. The third was a 64-year-old man with no previous dyspeptic history who smoked more than 30 cigarettes a day, consumed large quantities of alcohol and who developed a chest infection. The fourth was a 46-year-old woman with a dyspeptic history who was on both steroidal and non-steroidal anti-inflammatory therapy. There were no additional deaths during the follow-up period due to perforated duodenal ulcer or to related ulcer complications.

DISCUSSION

The classical presentation of perforated duodenal ulcer — sudden severe upper abdominal pain and clinical peritonitis — is well recognised. Frequently a history of peptic ulcer symptoms is given by the patient but not invariably so. Fully a quarter of our patients had no dyspeptic symptoms prior to perforation, a finding reflected in reports from other centres.[6, 8, 9] Nineteen patients were taking steroids or non-steroidal anti-inflammatory agents at the time of perforation. The proportion of patients with perforated duodenal ulcer reported to have free gas below the diaphragm on the erect abdominal radiograph is variable.[3, 10] In this group 69.6% of individuals X-rayed in the pre-operative period were found to have free sub-diaphragmatic gas. Whilst a definitive ulcer history and positive pre-operative radiological findings are strong corroborative factors, their absence in any individual patient should not deter the surgeon from making the diagnosis of perforated peptic ulcer and initiating treatment if the clinical picture is appropriate.

There is continuing debate in the literature regarding the preferred surgical procedure for the patient with a perforated duodenal ulcer. Simple suture closure of the perforation has much to commend it — the procedure itself is usually technically undemanding and can readily be performed by a relatively inexperienced surgeon, often operating on an ill patient. The vast majority of the patients in our series (205; 89%) had simple suture of their perforation carried out over a 10 year period by a large number of surgeons, virtually all of whom were in the training grades. The mortality rates of 2% for this group of individuals, and 1.7% for the entire group of 230 patients compare very favourably with those reported in other series,[3, 6, 9, 10, 11] but do not quite match the remarkable 'zero' mortality achieved by A B Mitchell, operating in the Royal Victoria Hospital in the early years of this century.[12]

During the follow-up period of our study at least 107 patients (52%) who had simple suture closure of a perforation at initial operation developed further ulcer symptoms. Of these, seven re-perforated and 56 required definitive ulcer surgery. These figures are in keeping with those documented from elsewhere.[1, 3, 4, 5, 8] It is because of this very high incidence of recurrent symptoms following simple suture of a perforated duodenal ulcer that some authors advocate the performance of a definitive operation at the time of initial surgery.[3, 6, 7, 10, 11] Clearly such a policy can only be justified if the more demanding and time-consuming operations such as proximal gastric vagotomy or truncal vagotomy and drainage can be performed in these uniformly sick patients without any increase in morbidity and mortality. In common with others,[3, 10, 11] our data suggest that this indeed may be the case although the number of patients in the definitive surgery group is too small for legitimate comparative purposes. However, prospective randomised studies by other authors, with careful follow-up of patients, do indicate that emergency definitive ulcer surgery is not associated with an increased complication rate.[13, 14]

We thus feel that a strong case can be made for performing a definitive ulcer operation at the time of emergency surgery for the chronic perforated duodenal ulcer. These patients have a high risk of developing further symptoms refractory to medical treatment. However, we do not extend this argument to the treatment of perforated acute duodenal ulcers since it would appear that the risk of recurrent symptoms in such patients is significantly less — 13 of 67 individuals (19%) during the period of our study. In general we hold the view that a dogmatic approach is not appropriate and readily acknowledge that simple suture closure of the perforation by a relatively inexperienced surgeon operating on an ill patient in the small hours of the morning is a perfectly legitimate and correct treatment.

We wish to thank the consultant surgeons at the Royal Victoria Hospital, Belfast, for allowing us to study the case notes of their patients. Thanks are also extended to Miss C Rusk and Miss D McFarland for secretarial assistance and to Mr C Patterson, Department of Medical Statistics, The Queen's University of Belfast.

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Case report

A case of ocular torticollis

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Torticollis is a relatively common condition of childhood. The principal cause is fibrous contraction of the sternomastoid muscle and a 'tumour' is often noted in the muscle. Less commonly, torticollis may be due to skeletal or ocular abnormalities. We report a case of torticollis initially diagnosed as muscular in origin but subsequently found to be secondary to an ocular muscle palsy.

CASE HISTORY

A four-month-old child with a six-week history of a head posture and transient squint was referred to a general hospital outpatient department. Obstetric and developmental history were normal. Torticollis secondary to spasm of the trapezius muscle was diagnosed and physiotherapy was instituted.

At age 14 months the head posture was still present and because of a history of squint an ophthalmic opinion was sought. On examination the head was tilted to the right (Fig 1). Upshoot of the left eye was demonstrated with the head straightened or tilted to the left side. This is a positive Bielschowsky head tilt test and is indicative of paralysis of the left superior oblique muscle.[1]

A left inferior oblique myectomy was performed and one week post-operatively the head posture had disappeared (Fig 2). Subsequently the child was knocked down by a car and the head posture returned. A right inferior rectus recession was carried out and the head posture has since straightened again.

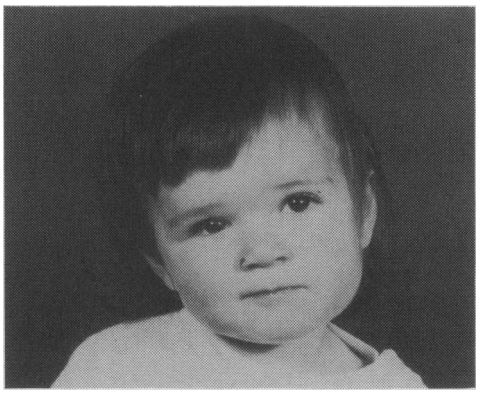

Fig 1.
Ocular torticollis, head tilted to right.

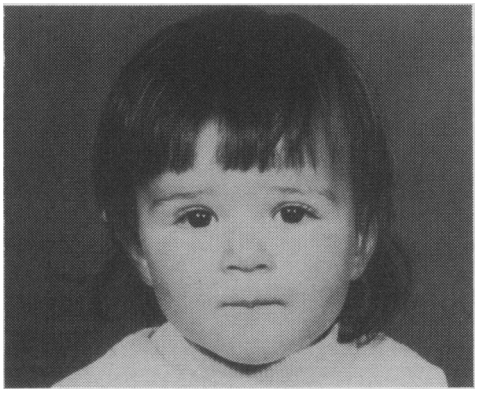

Fig 2.
One week after left inferior oblique myectomy.

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DISCUSSION

Torticollis may be muscular, skeletal or ocular in origin and occasionally no cause is found. The commonest cause is a congenital shortening of the sternocleidomastoid muscle and often there is a fibrous mass in the muscle belly. Rarely an acquired myositis can cause torticollis. Skeletal conditions associated with torticollis include occipitocervical synostosis, Klippel-Feil syndrome, atlantoaxial displacement, Sprengel's deformity and congenital deformities of cervical discs. In Sandifer's syndrome a head posture is adopted to diminish gastro-oesophageal reflux associated with hiatus hernia.[2]

Refractive errors, nystagmus and ocular muscle abnormalities can all cause ocular torticollis.[3] The head posture maintains optimum binocularity and visual acuity. With oblique astigmatism the head is held to one side to reduce distortion.[4] A child with congenital nystagmus turns or tilts his head so he can view an object at the null point where nystagmus is at a minimum. Weakness or restriction of virtually any extraocular muscle can lead to a compensatory head posture with the head characteristically held in the direction of action of the affected muscle.

In our patient an abnormal head posture was adopted to achieve binocularity and avoid diplopia caused by a superior oblique muscle palsy. The superior rectus and superior oblique muscles act as intorters of the eye, i.e. rotate it towards the nose. The inferior rectus and the inferior oblique are extorters, i.e. rotate the eye away from the nose. These movements keep the horizon level when the head is tilted. When the left superior oblique is paralysed the left inferior oblique acts unopposed to extort the eye causing diplopia. The head is tilted to the right utilising the extortion of the affected eye and causing compensatory intortion of the other eye so restoring single vision. When the head is tilted to the left the paretic superior oblique muscle cannot intort the eye and the left superior rectus acting unopposed by the normal depressing action of the oblique causes upshoot of the left eye. This is the positive Bielschowsky head tilt test demonstrated by this patient.

The head posture adopted by our patient tends to mimic that found in muscular causes of torticollis. However, passive movement of the child's head to the opposite side was unimpeded and this is unusual in cases of muscular origin. Squint surgery was successful in correcting the head posture but in long-standing cases secondary cervical musculoskeletal contractures may develop and extraocular muscle surgery alone may be ineffective. Congenital superior oblique muscle palsy may be due to birth trauma affecting the IV nerve, but there was no history of birth trauma in this case. Occasionally the nerve or muscle itself is absent.

Ocular problems are an important and treatable cause of congenital torticollis. These should be suspected in all cases where passive head movements are normal. In neglected cases with secondary contractures the exact aetiology of the condition may be difficult to elucidate.

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Case report

Renal tubular acidosis with nerve deafness

M A McShane, D J Carson, A O B Redmond, D Adams

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A child with renal tubular acidosis (RTA) and sensorineural deafness is described. This association represents a subtype of RTA and is inherited as an autosomal recessive trait. Late diagnosis of deafness occurred despite frequent follow-up, indicating the need for continual expert audiological assessment in all children with complex renal disease.

INTRODUCTION

Renal tubular acidosis (RTA) is a non-uraemic clinical syndrome of disordered renal acidification.[1] In the classic distal RTA (Type 1), subjects have hyperchloraemic acidosis, hyponatraemia and hypokalaemia. The urinary pH remains high (pH $>$ 6.2), even in the presence of systemic acidosis. Although the defect is permanent, the prognosis is good when the diagnosis, with correct management, is established early in order to prevent nephrocalcinosis and secondary renal damage.[2] Inheritance of the classic distal RTA (Type 1) is autosomal dominant. Several subtypes of distal RTA have been described such as incomplete distal RTA, distal RTA with bicarbonate wasting, transient distal RTA in infants and distal RTA with nerve deafness. We describe a patient, to our knowledge the first case from Ireland, with distal renal tubular acidosis and bilateral sensorineural deafness.

CASE HISTORY

JC was born to non-consanguineous parents following an uncomplicated pregnancy, by normal delivery at term, weighing 4.1kg. There was no family history of genetic disease or congenital abnormalities. At one month, he became dehydrated, refusing feeds and vomiting. Investigations showed hypokalaemia and hyperchloraemic metabolic acidosis. The diagnosis of renal tubular acidosis was suspected and potassium and bicarbonate replacement therapy commenced. During his first two years he required frequent hospital admissions for recurrent dehydration, vomiting, acidosis and electrolyte imbalance precipitated by minor illnesses. Delay in speech was recognised from an early age. A bilateral flat sensorineural hearing loss with thresholds of 60db was diagnosed at three years (Free Field performance test). No hearing defect was suspected at routine health visitor assessment at eight months. At four years, while receiving bicarbonate

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(1mmol/kg/day) and potassium (2mmol/kg/day) he was noted to have marked polydipsia and polyuria (>3 litres/day). The only clinical abnormalities found were short stature — height 88.8cm (3rd centile), weight 12.4kg (3rd centile) and deafness.

Initial investigations showed hypokalaemia (K 2.4 mmol/l), hyperchloraemia (Cl 117mmol/l), a partially compensated metabolic acidosis (pH 7.31, pCO_2 27mmHg, bicarbonate 16.2mmol/l, base excess 11.8mmol/l) and an alkaline urine (pH 7.4). Serum sodium, urea and creatinine were normal. After fluid restriction, urine osmolality was 148mosmol/kg and desmopressin administration did not reduce urine output or increase urine osmolality. Radiologically, there was bilateral nephrocalcinosis but no evidence of rickets. Urinary excretion of calcium was elevated, 2.79mmol/24 hours (9mg/kg/day). Chromatography showed no excess aminoaciduria.

He was discharged on an increased dose of sodium bicarbonate (5mmol/kg/day) but still required potassium supplements (2mmol/kg/day). After one month, the urinary output had decreased to 1.3l/day (mean of over three days). Serum electrolytes and acid base balance were normal and urinary calcium excretion had fallen to normal, 0.52mmol/24 hours (1.7mg/kg/day).

At the most recent review the high requirement for bicarbonate and potassium has continued (six years). The audiogram shows a 'ski slope' pattern with profound hearing loss (Figure). He now attends a school for the deaf and is making satisfactory progress. We believe this is the first report of this association in Ireland.

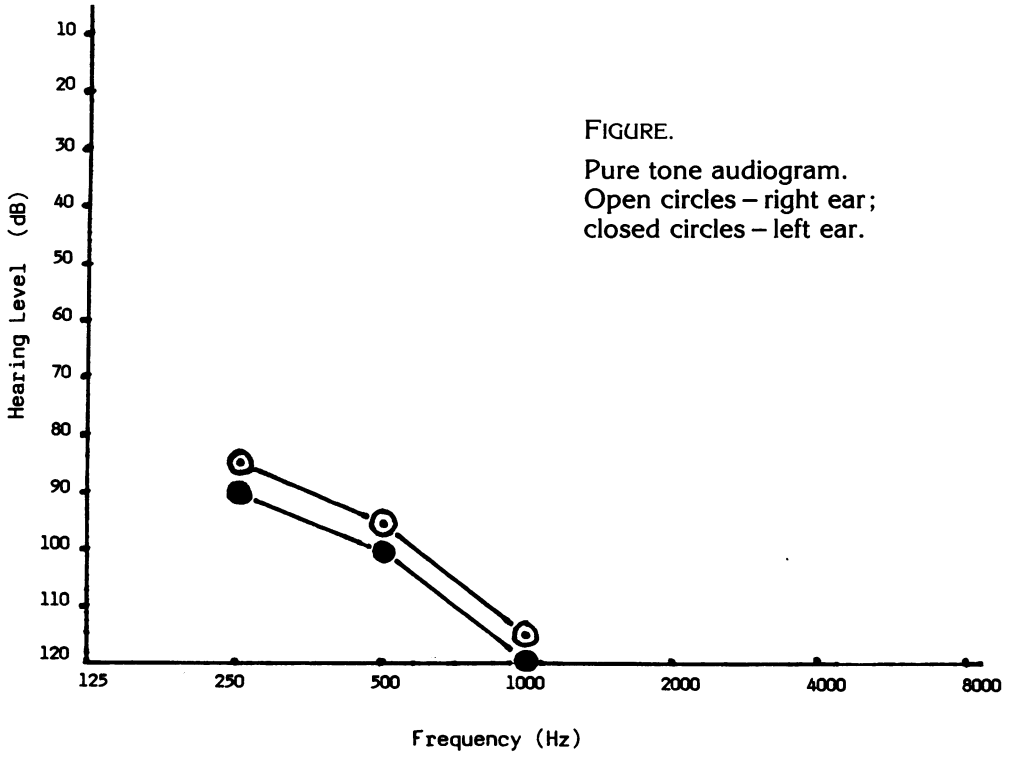

FIGURE.

Pure tone audiogram.
Open circles – right ear;
closed circles – left ear.

DISCUSSION

Our patient's initial presentation was typical of distal renal tubular acidosis. With the subsequent recognition of bilateral sensorineural deafness, it became clear that he had the subtype of RTA with deafness which is recognised as a distinct entity with an autosomal recessive mode of inheritance.[3] The associated deafness may occur at birth or later in childhood.[4] The late diagnosis of deafness in our patient and in other reports emphasises the need for early and repeated expert audiological assessment in children with RTA. Use of new techniques such as brain stem auditory evoked responses may be of help with these children.

Although the initial presentation of this case was typical, some features of the subsequent course were unexpected. Firstly, potassium supplements are not usually required to maintain normokalaemia when acidosis is corrected. In our patient their withdrawal caused a precipitous fall in serum potassium despite full bicarbonate replacement. Secondly, nephrocalcinosis developed despite bicarbonate replacement in the normal dosage of 1 – 3 mmol/kg/day from one month of age. Increasing bicarbonate therapy to 5 mmol/kg/day resulted in a reduction in urinary output, a fall in urinary calcium excretion to optimum levels (< 2 mg/kg/day), and catch-up growth (height velocity 9.6 cm/year).

Our thanks to Miss Wendy Caulfield for typing the manuscript.

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Case report

Empyema necessitatis presenting as a swelling in the right hypochondrium

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Swellings in the right hypochondrium may originate in different anatomical structures, including the gallbladder, liver, kidney, colon and the structures of the abdominal wall. We describe a case of tuberculous empyema of the pleura presenting as a swelling in the right hypochondrium.

CASE HISTORY

A 61-year-old retired insulation worker with a 30-year history of heavy alcohol intake presented to the accident and emergency department with a swelling in the right hypochondrium. Nine months previously he had complained of vague discomfort in the right side of the abdomen, but investigations including liver function tests and ultrasound scan of the abdomen were normal. The discomfort continued, and 10 days prior to this admission he noticed a swelling in the right hypochondrium, which gradually became larger.

Three years previously, pulmonary tuberculosis had been diagnosed with acid-alcohol-fast bacilli being seen in the sputum. A chest radiograph showed a right-sided pleural effusion and fluffy pulmonary infiltrates in the upper and mid zones. Antituberculous chemotherapy was instituted using streptomycin (for three months), ethambutol and pyrazinamide (substituted for isoniazid because of markedly abnormal liver function tests after starting isoniazid). After 18 months' drug therapy the pleural effusion was still present, and, since the patient admitted to poor compliance with treatment, ethambutol and pyrazinamide were continued for a further nine months.

On this admission he was apyrexial. There were signs of a right basal pleural effusion. There was a painless, fluctuant and non-reducible swelling in the right hypochondrium. Tensing of the recti resulted in incomplete disappearance of the mass, which did not move with respiration. Erythrocyte sedimentation rate was 80 mm per hour, haemoglobin 12.3 g/dl, white cell count $12.6.10^9/1$ (69% granulocytes, 22% lymphocytes). A chest radiograph showed a right pleural reaction with a component lying posteriorly suggesting an encysted effusion or pleural thickening. There were bilateral healed rib fractures, and diaphragmatic pleural calcification consistent with asbestos exposure. Abdominal ultrasound

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demonstrated an 11 × 6 × 8 cm area of decreased echogenicity separate from the gallbladder and anterior to the liver. It was mainly fluid but also contained debris, suggesting an abscess. A CT scan showed a pleural collection of fluid tracking anteriorly. A subdiaphragmatic collection of fluid was seen extraperitoneally anterior to the liver (Fig 1). There appeared to be a subcutaneous communication between the two fluid collections.

A needle was inserted into the abdominal swelling and 200 ml creamy pus was aspirated. Pleural aspiration and biopsy were performed, and 500 ml pus was removed with a reduction in size of the abdominal swelling resulting. Iopamidol (300 mg iodine/ml) was injected into the pleural space. Radiographs confirmed a superficial anterolateral communication between the pleural and abdominal collections (Fig 2). Direct microscopy of the pus was negative for tubercle bacilli. Pleural biopsy, however, showed actively inflamed granulation tissue with aggregates of histiocytes and several multinucleated giant cells. On culture of pleural fluid tubercle bacilli were grown confirming the diagnosis of tuberculous empyema. Antituberculous treatment has been recommenced with percutaneous tube drainage of the abscess.

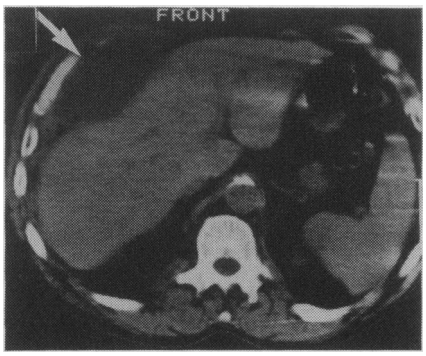

Fig 1. CT scan through the upper abdomen showing a subcutaneous abscess in front of the liver.

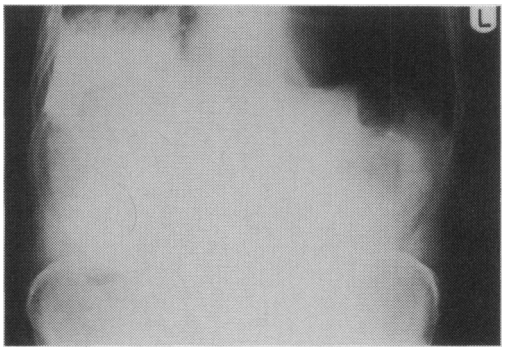

Fig 2. Iopamidol contrast study showing communication between the pleural cavity and the abdominal swelling.

COMMENT

Empyema necessitatis — the pointing of an empyema through the chest wall — is a recognised complication of an incompletely treated empyema. Standard descriptions of the condition state that the external swelling arises in the costal part of the chest wall.[1] In a comprehensive survey of the older literature, cases presenting with swellings in the loin and groin, or with rupture into the oesophagus or stomach, were noted.[2] In the present case the right hypochondrial swelling was clearly connected to a coexisting empyema of the pleura. We could not find any reference to a similar presentation complicating either tuberculous or non-tuberculous empyema.

The diagnosis of empyema necessitatis is usually clinical, but because of the unusual features in the present case, additional imaging procedures were particularly useful. The CT features of empyema necessitatis have been described recently,[3] and the CT scan in our case demonstrated a superficial anterolateral communication between the pleura and the abdominal swelling, which was confirmed by the contrast study.

Tuberculous empyema, formerly a relatively common complication of treatment by artificial pneumothorax, is now rare in this country, but remains a problem in developing countries.[4] It may follow rupture of a superficial cavity or paravertebral abscess into the pleural cavity. As in the present case, it may occasionally form at the site of a pleural effusion. Poor compliance with drug therapy and alcohol abuse[5] presumably predisposed to chronicity of infection. This case illustrates a new, or possibly neglected, complication of an old disease, and the application of modern imaging techniques in defining its unusual features.

We thank Dr J H Foster who performed the CT scan. We are grateful to Miss B Shannon for typing the manuscript.

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Case report

Meckel's diverticulum — too often forgotten in adults?

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Meckel's diverticulum is the commonest congenital anomaly of the gastro-intestinal tract, being present at 2% of autopsies.[1] Approximately 4% cause symptoms and most present during the first decade of life.[2] Hence, there is an awareness among clinicians of the possibility of the lesion presenting in infancy or childhood, but a clinical diagnosis of Meckel's diverticulum is rarely considered pre-operatively in adults. We describe three cases admitted to one surgical unit during a three-month period, which illustrate the varying presentation of the condition in adults.

CASE 1

An 83-year-old lady was admitted with a two-day history of vomiting and abdominal distension. Examination revealed increased bowel sounds and X-rays showed distended loops of bowel with fluid levels, in keeping with small bowel obstruction. Laparotomy was performed and revealed several loops of distended small bowel, with one loop entrapped behind a mesodiverticular band, which extended from the apex of a Meckel's diverticulum to the base of the small bowel mesentery. The diverticulum and band were resected and her post-operative course was uneventful. Histology revealed ischaemic necrosis of the diverticulum, but no heterotopic mucosa.

CASE 2

A 42-year-old man was admitted with a 10-hour history of severe lower abdominal pain. Abdominal examination revealed marked rebound tenderness in the left iliac fossa. However, on reassessment 30 minutes later, the point of maximal tenderness had surprisingly shifted to the right hypochondrium. Serum amylase and electrolytes were normal but the white cell count was elevated. Abdominal X-rays showed no free intraperitoneal gas. Laparotomy was undertaken and revealed a 5 cm long Meckel's diverticulum with a 6 x 4 cm gangrenous pouch at the apex. The diverticulum was resected and his post-operative course was uncomplicated. When the specimen was opened it was found to contain a 4 x 3 cm enterolith. Histology revealed ischaemic necrosis and gangrene of the diverticulum but no heterotopic mucosa.

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CASE 3

A 20-year-old man was admitted following an episode of severe crampy abdominal pain, after which he lost consciousness for a few seconds. On recovery, he passed approximately one pint of bright red blood per rectum. On admission he was clinically shocked, haemoglobin 9.2 g/dl. He was resuscitated with intravenous fluids, including four units of blood. Three years prior to this he had had a similar episode. At that time, barium meal and follow-through, barium enema, coagulation screen and tectnetium scan to detect ectopic gastric mucosa had all been normal. Gastroscopy had revealed a superficial duodenitis which was assumed to have been the source of bleeding. On this occasion, gastroscopy was normal but a small bowel series revealed a persistent shadow, consistent with a Meckel's diverticulum (Figure). This was confirmed at laparotomy when a 6cm long Meckel's diverticulum was found. The diverticulum was resected and his recovery was uneventful. Histology revealed ulcerated ileal mucosa, adjacent to an area of heterotopic gastric mucosa.

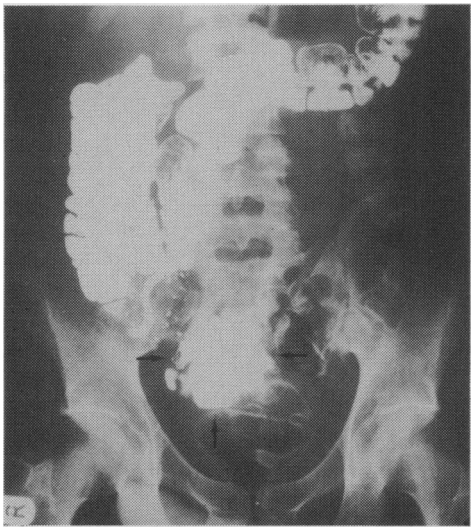

Figure. Case 3: Barium small bowel series demonstrating a Meckel's diverticulum.

DISCUSSION

Small bowel obstruction is reported in several series as the most common complication of Meckel's diverticulum in adults.[1-4] The obstruction is often caused by entrapment of bowel behind a mesodiverticular band or a persistent omphalomesenteric artery, although other mechanisms are recognised.[5,6] The estimated risk of developing such a complication in the elderly has been reported as zero,[1,3] but our 83-year-old patient serves as a reminder that despite the more frequent problems in early years, Meckel's diverticulum can cause symptoms over a wide age spectrum.

Inflammation of the diverticulum is said to be the next most common presentation of disease of Meckel's diverticulum in adults.[2,4] In this situation, a pre-operative diagnosis of acute appendicitis is usually made as the diverticulum is often located near the right iliac fossa. However, because of the mobility of the small bowel, symptoms and signs may be maximal in another area and thus mimic cholecystitis, pancreatitis, perforation of a duodenal ulcer or diverticulitis. This could account for the changing abdominal signs in our second patient. The chances of accurate pre-operative diagnosis of an inflamed Meckel's diverticulum are therefore slim, but there should be a high index of suspicion when clinical features of intra-abdominal inflammation are not indicative of any specific condition.

Rectal bleeding is the usual presentation of Meckel's diverticulum in children[5] but in adults it is much less frequent and is almost unknown in patients over 40 years of age.[2] This is a common presentation of a number of conditions and our

third patient demonstrates the difficulties in positively identifying a Meckel's diverticulum suspected as the source of gastrointestinal bleeding.

In patients who are actively bleeding, angiography or ^{99m}Tc-labelled red cells[7] may accurately locate the site of haemorrhage, but patients who are not actively bleeding often require extensive investigation. Abdominal scintigraphy will identify ectopic gastric mucosa in 90% of patients in whom it is present,[8] but in our patient this method failed to identify the diverticulum despite the presence of ectopic mucosa. The anatomical site of a Meckel's diverticulum makes visualisation with contrast media difficult and our patient had a negative barium meal and follow-through and a negative barium enema before the lesion was finally identified using a small bowel series. The chance of pre-operative diagnosis may be improved by using a barium small bowel enema, but it is clear that negative barium studies and scintigraphy do not exclude the presence of a Meckel's diverticulum.

There is still considerable debate in the surgical literature on the advisability of resecting a Meckel's diverticulum found incidentally at laparatomy. Several authors have attempted to correlate the anatomical characteristics of the diverticulum with the risk of its developing complications. It had previously been thought that a broad based diverticulum was less likely to become obstructed and was therefore relatively harmless. However, Mackey and Dineen have shown that diverticula with broad bases are no less likely to become symptomatic[2] and Leijonmark et al present evidence that these diverticula are in fact, more likely to become symptomatic.[1] Both papers suggest that the length of the diverticulum is a better indicator of the risk of complications developing.

The lifetime risk of patients developing a complication of their diverticulum has been calculated by different authors as being 4% in childhood, 3% during the teenage years and falling to zero in the elderly.[1, 3] These authors suggest that the morbidity and mortality associated with resecting an asymptomatic diverticulum is greater than the risk of developing a complication in later life. Others favour resection of a diverticulum found incidentally,[4, 6] especially in patients under 40 years of age.[5] Our experience with these patients, all of whom suffered significant morbidity and required laparatomy in adult life, would encourage us to support the latter view.

We would like to thank Mr D M Bell and Mr W A Hanna for their permission to publish these cases.

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Case report

Non-menstrual associated toxic shock syndrome

P G Murphy, W Holmes, T S Wilson, J P Alexander

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A 40-year-old female patient developed toxic shock syndrome following breast surgery. A toxin producing strain of *Staphylococcus aureus* was isolated from the post-operative wound.

INTRODUCTION

The term toxic shock syndrome (TSS) was first used in 1978[1] to describe a condition characterised by sudden onset of fever, shock, confusion, subcutaneous oedema, erythematous rash which desquamates in convalescence, abnormalities of many other systems, and negative blood culture. These effects are thought to be due to circulating staphylococcal toxins arising from a focus of staphylococcal infection. It has most commonly been associated with the use of certain types of tampons during menstruation, but is now increasingly being recognised in up to 13% of cases in a variety of clinical situations not associated with menstruation.[2] *Staphylococcus aureus* has been associated with TSS and most strains isolated from patients with this syndrome have produced enterotoxin F or pyrogenic exotoxin C, now thought to be the same toxin and renamed Toxic Shock Syndrome Toxin-1 (TSST-1). However, other strains of *Staphylococcus aureus* implicated in the syndrome have produced different toxins, particularly enterotoxins B and C[3] in non-menstrual cases. Mortality is higher in these cases with non-TSST-1 producing strains than with TSST-1 producing strains.[4] We report here a patient who fulfilled the case definition of the Center for Disease Control, Atlanta, USA,[5] and which we believe is the first case notified in Northern Ireland.[6]

CASE HISTORY

A 40-year-old woman was admitted to another hospital for removal of a breast lump. Surgery was uneventful and she was discharged the following day. The biopsy was subsequently reported as benign fibroadenosis. Two days later the patient was readmitted with pyrexia (40°C), hypotension (90/40 mmHg), and an erythematous rash on the trunk which spread rapidly to all limbs over the next two days. The patient was commenced on benzyl penicillin and gentamicin for a presumed diagnosis of septicaemia, possibly meningococcal.

Two days after this second admission the patient was transferred to the intensive care unit of this hospital. The breast wound, although not painful, exuded a slight

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discharge from which *Staphylococcus aureus* was cultured and the antibiotic therapy was changed to flucloxacillin and gentamicin on the advice of the bacteriologist.

Other clinical features included vomiting, periorbital oedema, pulmonary oedema, leg muscle myalgia, confusion, and disorientation in time and place. Laboratory investigations showed a leucocytosis ($20 \times 10^9/l$), impaired renal function (creatinine 395 μmol/l), and impaired hepatic function (bilirubin 64 μmol/l, aspartate transaminase 184 μmol/l, alanine transaminase 70 μmol/l, and lactate dehydrogenase 595 μmol/l). There was also an elevated creatinine kinase (1589 μmol/l), a decreased serum calcium (1.68 mmol/l), and an increased serum phosphate (1.6 mmol/l). The haemoglobin dropped from 12.5 g/dl to 9.2 g/dl over 48 hours without any signs of blood loss but this may have been contributed to by a dilutional effect. A coagulopathy was detected with a partial thromboplastin time of 72 seconds (control 44s), a fibrinogen degradation products level of 32 μg/ml (normal 0 – 8) and a low platelet count ($105 \times 10^9/l$). All these features are characteristic of TSS.

On the eighth post-operative day the rash had faded considerably and there had been no pyrexia for 48 hours. However, the creatinine continued to rise (418 μmol/l) and dopamine was still required to maintain the blood pressure at 100/70 mmHg. The patient was then taken back to theatre for exploration of the breast wound in which considerable necrosis was observed and *Staphylococcus aureus* was re-isolated from the wound.

The patient made a gradual recovery over the next few weeks. Desquamation occurred throughout the area of distribution of the rash starting two weeks after the rash had disappeared and continuing for three weeks into convalescence. This was followed by marked hair and nail loss with subsequent regrowth after several months. These are the convalescent features characteristic of TSS. The patient was discharged one month after admission.

BACTERIOLOGY

The strain of *Staphylococcus aureus* isolated from the wound was non-typable by phage typing at the routine test dose ×100. It was sensitive to benzyl penicillin with a minimum inhibitory concentration (MIC) of 0.125 μg/ml and a minimum bactericidal concentration (MBC) of 0.25 μg/ml, and also to flucloxacillin (MIC 0.5 μg/ml and MBC 0.5 μg/ml). Screening swabs from various carriage sites did not produce similar strains of *Staphyloccocus aureus.* A *viridans* group streptococcus was isolated from one of four blood culture bottles taken on the day of re-admission, but this was not considered clinically relevant. *Staphylococcus aureus* was not isolated from blood culture.

Toxin studies are not routinely available but tests carried out at a reference laboratory showed that this strain of *Staphylococcus aureus* did not produce TSST-1, but did produce enterotoxin C. Limited serum antitoxin studies in both acute and convalescent sera showed the presence of antibody to TSST-1 and to enterotoxin B but not against enterotoxin A. Unfortunately antibody studies against enterotoxin C and other toxins were not available in the reference laboratory.

DISCUSSION

Up to 15% of some reported series of non-menstrual TSS have followed surgery, from which a median incubation period of two days has been estimated.[2]

Typically, the wound is not painful nor clinically suspicious of being infected. Other cases have been described, associated with subcutaneous abscesses, cellulitis, infected insect bites, hydradenitis suppurativa, an infected cutaneous ulcer and an infected burn.[2]

Menstrual and non-menstrual associated TSS strains appear to have different bacteriological features. In this case, the strain of *Staphylococcus aureus* was not phage-typable. Epidemiological studies have shown that strains of *Staphylococcus aureus* in menstrual associated TSS are usually in phage group 1,[4] and non-menstrual associated TSS strains are either in other phage groups or are not phage-typable. Also, this strain did not produce TSST-1 and did produce enterotoxin C. Although TSST-1 is produced by about 90% of strains from reported cases of menstrual associated TSS, it is only associated with about 60% of non-menstrual associated cases, and it has recently been suggested that its role is not essential in the pathogenesis of TSS and that other as yet unrecognised toxins may play a part.[4]

Antibodies may have a protective role, as 88% of control group patients have antibody titres to TSST-1,[7] whereas only 18% of acute phase sera[8] and 20 – 30% of convalescent sera of all TSS patients show antibody to TSST-1.[9] Patients with TSS would appear to have a defect in the anti-TSST-1 immune response as shown by the low percentage of patients who seroconvert. The antibody titres in these patients are also much lower than those of control sera.[3] This may also explain the high recurrence rate of 28% in the same individual patients.[5] Significant antibody titres to TSST-1 are present in normal human immunoglobulin[10] and this may have a therapeutic role in the future when the causative toxin has been more clearly identified.

We wish to thank Antonnette A Wieneke of the Central Public Health Laboratory, Colindale, London, for toxin detection studies, Dr J de Azavedo of the Moyne Institute, Trinity College, Dublin, for the antibody studies, and Mr M J G O'Reilly for permission to report this case.

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Case report

Adrenomyeloneuropathy

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Addison's disease is a well recognised clinical syndrome, but the neurological syndromes which can be associated with it are more unusual.

CASE HISTORY

A 27-year-old male presented with mild unsteadiness of gait of two years' duration. Since childhood he had had enuresis and occasional faecal incontinence. From the age of eight he had had recurrent episodes of upper abdominal pain and anorexia which had been severe enough on several occasions to necessitate admission to hospital, although a precise diagnosis of the cause was never made. Between episodes, his appetite was not good and he never attained a weight greater than 120 lbs. During his teenage years he acquired a great fondness for potato crisps and salted peanuts and he often added copious amounts of salt to his food.

In his early 20s he had a particularly severe episode of anorexia, vomiting and abdominal pain while on a summer holiday in Spain. Despite feeling exceedingly weak, he made his way back to London by plane, and, without seeking medical attention, checked into a hotel where he spent his time in his room drinking a lot of fluid and eating many packets of potato crisps and peanuts, so that three days later he was able to emerge feeling more or less back to normal.

Puberty occurred normally in his mid-teens but he became aware of difficulty in penile erection; this symptom persisted together with the disturbance of bladder control which had been present since childhood. He became unsteady on his feet at about the age of 25. His neurological symptoms have not changed significantly during the three years he has been under observation. He has had no sensory symptoms in his limbs nor any visual symptoms and, in particular, colour vision is intact.

He was an adopted child and has no knowledge of his family background.

Examination revealed a thin well-tanned young man with pigmentation of the lips and buccal mucosa. Development of secondary sexual characteristics was normal. Blood pressure was 120/70 mmHg lying and standing. His speech function was normal, colour vision unimpaired and function of the cranial nerves intact. In the limbs there was a mild spasticity of arms and legs together with mild

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weakness of hip flexors and ataxia on heel-shin testing bilaterally. The tendon reflexes were generally brisk with bilateral extensor plantar responses. On sensory testing vibration sense was absent to the knees, and position sense defective at both great toes. Pin prick and light touch were unimpaired. He had a spastic ataxic gait.

Serum urea and electrolytes, blood count, sedimentation rate, serum B12 and folic acid levels were all normal. Cerebrospinal fluid analysis and myelogram were normal. Sensory evoked responses were delayed at spinal and cortical levels. Motor and sensory conduction in the peripheral nerves were normal. Visual evoked responses and auditory evoked responses were also normal. CT scan of the brain was normal.

The diagnosis of primary adrenal insufficiency was confirmed by serum cortisol levels of 150 nmol/l and 140 nmol/l at 0700 and 2300 h respectively and 140 nmol/l 30 minutes after an intravenous injection of 0.25 mg tetracosactrin (Synacthen), and a random plasma ACTH of 1150 ng/l (normal < 70). Adrenal autoantibodies were not detected.

Plasma levels of very long chain (C24 – 30) fatty acids, measured by gas liquid chromatography,[1] are shown in the Table.

After replacement therapy with hydrocortisone and fludrocortisone his appetite and weight have increased, as has his general strength, but some three years after diagnosis his neurological symptoms and signs are unchanged.

TABLE

Plasma levels of very long chain fatty acids

	Patient	*Controls**
C22% total fatty acid**	0.773	1.18 ± 0.61
C24% total fatty acid	0.963	0.78 ± 0.32
C25% total fatty acid	0.030	0.03 ± 0.03
C26% total fatty acid	0.028	0.01 ± 0.01
C26 (μg/ml)	0.773	0.33 ± 0.18
C24/C22	1.246	0.84 ± 0.08
C26/C22	0.036	0.01 ± 0.01

*From Moser et al.[1] Results as mean ± SD of 22 persons age 12 to 15 years with no known disease and 28 persons with neurologic or metabolic disorders not related to adrenoleukodystrophy or adrenomyeloneuropathy.

**C22 etc refers to the carbon chain length of the fatty acid.

DISCUSSION

The findings of primary adrenocortical failure together with a mild spastic ataxic paraparesis and an elevated plasma level of very long chain fatty acids established the diagnosis of adrenomyeloneuropathy,[2] and to our knowledge this is the first case described in Ulster.

Adrenomyeloneuropathy is a rare X-linked recessive disorder. A variant, adreno-leukodystrophy, was first described in 1923[3] and both forms may occur in one family. The first features of adrenomyeloneuropathy are symptoms of adrenal

insufficiency beginning in the first or second decade followed by the appearance of a progressive myelopathy during the third or fourth decade,[4] and often accompanied by a cerebellar ataxia and sphincter disturbance as in the present case. Features of a peripheral neuropathy may also occur. The precise aetiology of the condition is unknown but the accumulation of saturated unbranched very long chain fatty acids in tissues and body fluids is characteristic,[2] suggesting an inherited defect in some enzyme(s) involved in fatty acid metabolism. Cytoplasmic inclusion bodies, which may be detected in the brain, spinal cord, adrenal and testicular tissue are thought to represent breakdown products of very long chain fatty acids.[5]

Although the hypoadrenalism can be satisfactorily treated with corticosteroids as in Addison's disease, correction of the hypoadrenal state does not modify the progressive neurological course.[2] Perhaps the most remarkable aspect of this case has been the patient's ability to fend off any serious metabolic catastrophe for a 20-year period through copious salt ingestion. Salt hunger has long been recognised as a feature of Addison's disease[6] but such prodigious efforts must be exceptional!

Detection of 'carriers' to facilitate genetic counselling should represent an important part of the overall management of any inheritable progressive disorder lacking specific curative therapy. One report suggested that females, heterozygous for the condition, may be identified by abnormal auditory evoked responses,[7] but the finding of normal responses in our patient who has well developed features of the syndrome must cast doubt on this. A more promising approach to the detection of heterozygotes lies in the measurement of plasma very long chain fatty acids, which have been shown to be elevated in carriers as well as in patients.[1, 2]

We are indebted to Dr H W Moser, Director of the John F Kennedy Institute, Baltimore, Maryland, for measuring plasma fatty acid levels in this patient. We would like to thank Miss Angela Gibney for typing the manuscript.

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Case report

Systemic amyloidosis — three illustrative cases

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Amyloidosis is characterised by the extracellular deposition of abnormal protein fibrils. Recent advances in the study of amyloidosis have been based on the chemical analysis of these protein fibrils.[1,2] This has also led to a more rational classification of the condition (Fig 1). Further discussion of localised forms of amyloidosis is outside the scope of this article, which will be confined to systemic amyloidosis.

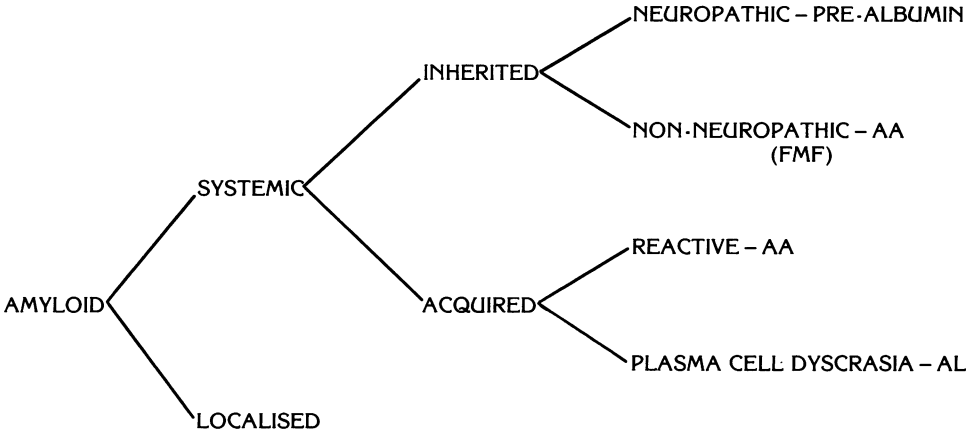

Fig 1. *Classification of amyloidosis*

Systemic amyloidosis may be either inherited or acquired. The neuropathic forms, which show an autosomal dominant inheritance, predominantly affect peripheral nerves, but may involve also the heart, kidneys and other tissues. The protein fibrils are derived from circulating prealbumin, which has an inherited abnormality in the amino acid sequence in some kindreds.[3] The most studied form of inherited non-neuropathic amyloidosis is familial Mediterranean fever (FMF). This autosomal recessive disorder is confined to populations originating on the southern and eastern coasts of the Mediterranean Sea. It is characterised

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by recurrent acute inflammatory attacks of various types (abdominal pain, polyserositis, arthritis, skin rashes) and the gradual deposition of amyloid protein, most notably in the kidneys. The amyloid fibrils in FMF are derived from a circulating acute phase reactant known as serum amyloid A (SAA) protein. Serum amyloid A is also the major precursor protein of acquired reactive systemic amyloidosis (AA amyloid). This may occur as a complication of chronic inflammatory diseases such as rheumatoid arthritis or chronic infection, and was previously classified as secondary amyloidosis.

The other main type of systemic amyloidosis is AL amyloid, which is derived largely from the variable regions of immunoglobulin light chains, secreted by a clone of plasma cells. This clone may give rise to overt myeloma with features such as lytic lesions in bone but may also present solely as amyloidosis. Such cases were previously classified as primary amyloidosis.

Amyloidosis is an uncommon condition, with a wide variety of clinical presentations, and for this reason the diagnosis may be overlooked. Three cases presented to a general medical ward within a three-month period. We describe these cases to illustrate the diverse features of the disorder.

CASE 1

A 62-year-old lady presented with swollen salivary glands and mild cardiac failure. She also complained of dysphagia, loss of hair, episodes of spontaneous bruising around the eyes, and aching muscles. She had been well until one year previously when she had complained of aching leg and back muscles on exertion and had had an operation for spinal stenosis, without improvement in her symptoms. On examination, the submandibular glands were enlarged and had a firm rubbery consistency, the tongue was grossly enlarged and showed indentations made by her teeth (Fig 2). The skeletal muscles were enlarged and indurated to give an incongrous brawny appearance (American footballer or shoulder pad sign). Biopsy of deltoid muscle showed extensive infiltration of the interstitium with amyloid which was permanganate resistant. Biopsy of the rectum contained mucosa only and did not contain any amyloid. A type G-lambda paraprotein was present in the serum (6g/l) and urine (0.7g/l). Bone marrow trephine revealed a moderate increase in plasma cells (15%). No lytic lesions were present on skeletal survey and other investigations revealed normal haemoglobin, urea and serum albumin concentrations and ESR of 25 mm/hr. An electrocardiogram (ECG) showed low voltage complexes.

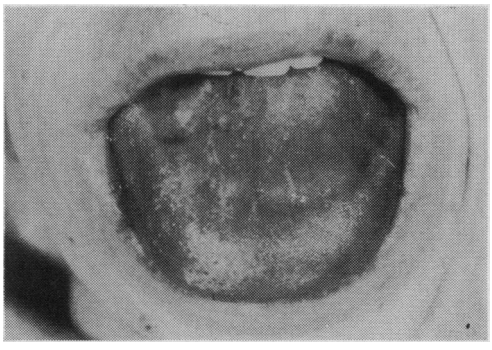

Fig 2. Case 1. Enlarged indurated tongue with indentations made by the lower teeth.

The cardiac failure was easily controlled by diuretics. A course of melphalan and prednisolone was given but there was no reduction in the serum paraprotein concentration. The most disabling symptoms were dysphagia due to progressive enlargement of the tongue and generalised muscular weakness. The patient died nine months after diagnosis from a respiratory arrest.

CASE 2

A 62-year-old man presented with oedema due to nephrotic syndrome and cardiac failure. He had been previously well. Urinary protein loss was 6g/24 hours (normal less than 0.15g) and serum albumin was reduced to 16g/litre (normal 35 – 50). However, renal function was preserved as indicated by a normal urea concentration 5.1 mmol/l (3 – 6). A type G-lambda paraprotein was present in the serum (10g/l) and urine (concentration not measured). The kidneys showed increased echogenicity on ultrasound examination. ECG showed low voltage complexes. The cardiac failure and oedema proved resistant to intensive treatment with diuretics and albumin infusions. Melphalan and prednisolone therapy was commenced, but the patient died from a bleeding duodenal ulcer five weeks after presentation. At autopsy, the heart, kidneys, spleen and liver were heavily infiltrated with amyloid which was permanganate resistant. The blood vessels in the base of the duodenal ulcer were also infiltrated with amyloid. There were large numbers of plasma cells in the bone marrow (proportion not estimated).

CASE 3

A 58-year-old man was referred for investigation of diarrhoea. Two years previously he had developed cardiac failure and was diagnosed as having amyloid heart disease by endomyocardial biopsy at the Regional Medical Cardiology Centre, Royal Victoria Hospital, Belfast. Prior to this, he had been healthy, with no history of chronic inflammatory disease, ischaemic heart disease or hypertension. He was of Anglo-Irish descent with no family history of amyloidosis. On his initial presentation he underwent extensive cardiac investigation. ECG showed sinus rhythm, some reduction in voltage of the limb lead complexes and T wave inversion in leads, 11, 111, AVF, V1 – V4. Echocardiographic appearances were consistent with a cardiac infiltration. A pyrophosphate radioisotope scan demonstrated diffuse intense uptake of the isotope by both ventricles which is a characteristic finding in cardiac amyloidosis.[4] Amyloid was demonstrated in the endomycardial biopsy stained with Congo red, and amyloid fibrils were also demonstrated by electron microscopy. At this time ESR was 8 mm/hr and there was no detectable paraprotein in serum or urine. A radiological skeletal survey and bone marrow examination were not performed. His cardiac failure was treated with Digoxin and diuretics and a course of Cyclophosphamide was given. However, over a two year period he became progressively disabled by diarrhoea, postural dizziness and muscular weakness. On examination his muscles were wasted and weak. All tendon reflexes were absent, and sensory testing of the extremities caused unpleasant dysasthesia. Blood pressure was 110/70 mmHg supine and 55/40 mmHg erect.

A barium enema was normal, small bowel X-ray series showed coarsening of the mucosal folds and was notable for the rapid transit time from mouth to caecum of 15 minutes. Nerve condition velocities were significantly slowed (median nerve 41 m/sec, posterior tibial nerve 36 m/sec). Heart rate on ECG monitoring did not alter with posture, respiration or valsalva manoeuvre consistent with an autonomic neuropathy.[5] Rectal biopsy showed amyloid in the submucosa and pre-treatment of the tissue sections with permanganate gave equivocal results. Haemoglobin, ESR and a biochemical screen for malabsorption were normal. Serum urea was 3.9 mmol/l (3 – 6) and protein loss in the urine was less than 0.1 gm/24 hours. Electrophoresis of serum and concentrated urine (×250) was repeated and combined with immunofixation techniques but still failed to demonstrate a paraprotein.

The cause of the diarrhoea was diagnosed as autonomic neuropathy due to amyloidosis. The postural hypotension has been improved by support stockings. The peripheral neuropathy persists and has progressed to the extent that the patient now requires a frame to aid walking. His cardiac failure remains well controlled by Digoxin and modest doses of diuretics.

DISCUSSION

Case 1 illustrates three clinical signs which are virtually pathognomonic of AL amyloidosis. These are the enlarged, indurated tongue, the shoulder pad or American footballer sign and spontaneous periorbital purpura. The purpura typically occurs after proctoscopy for a rectal biopsy and can be abbreviated to the more memorable PPPP (post proctoscopic periorbital purpura).[6]

Cases 1 and 2 are undoubtedly examples of AL amyloidosis since a circulating paraprotein was detected and there was an increased proportion of plasma cells in the bone marrow. In Case 3 a paraprotein was not detected which makes the classification of the amyloid type less certain. The detection of a paraprotein is not essential for the diagnosis of AL amyloidosis and cannot be detected in the serum in 32% of cases and in the urine in 28% of cases.[6] This patient presented with cardiac and neural involvement which are both common presentations of AL amyloidosis.[7] The normal urinary excretion of protein is unusual but can occur in 10% of cases.[6] Acquired reactive AA amyloidosis is excluded by the absence of underlying inflammatory disease. However, it is not possible to exclude a sporadic case of inherited amyloidosis, despite the lack of family history and the late age of onset (58 years).[8]

Histochemical analysis using a potassium permanganate technique[9] was unhelpful in this case. This technique is useful for identifying AA amyloid but does not distinguish AL amyloid from inherited amyloid consisting of prealbumin. Immunocytochemistry offers the prospect of a specific staining method. However, it has proved difficult to raise suitable antibodies to AL amyloid proteins because they are derived from the variable rather than the constant regions of the immunoglobulin light chains. Recently, Dalakas and co-workers have reported a series of 15 patients with amyloid polyneuropathy stained using antibodies to immunoglobulin light chains and prealbumin.[10] These patients had no family history of amyloidosis and no evidence of a plasma cell dyscrasia. In 12 of the 15 cases the amyloid protein was AL and in 3 it was prealbumin. We conclude that Case 3 is probably an example of AL amyloidosis but the classification will not be certain until these immunocytochemical techniques become more widely available.

In Cases 1 and 2 the immunoglobulin light chain belongs to the lambda (λ) class. This is consistent with previous observations that λ chains are found twice as frequently as kappa (κ) chains in AL amyloidosis, which is in contrast with myeloma not complicated by amyloidosis, in which κ chains are twice as common as λ.[6] It is not clear why λ chains should be more 'amyloidogenic' than κ chains.

A histological diagnosis is essential to confirm amyloidosis. Tissue sections which contain amyloid exhibit apple-green birefringence when stained with Congo red and examined under polarised light. Amyloid fibrils also have a characteristic appearance when examined by electron microscopy. The best site for initial biopsy is the rectum, which is positive in 80% of cases provided that submucosal tissue is obtained. The rectal biopsy in Case 1 was not deep enough to include

submucosa which is the most likely reason for the failure to detect amyloid. It may be necessary to biopsy other tissues if rectal biopsy is negative, although there is the risk of bleeding due to vessel wall infiltration or factor X deficiency which is a rare feature of AL amyloidosis.[11]

The treatment of AL amyloidosis is unsatisfactory. The most logical approach is to suppress light chain production using regimes such as melphalan and prednisone or cyclophosphamide which are of use for multiple myeloma. A prospective trial of melphalan and prednisone versus placebo showed that the nephrotic syndrome improved in a few patients but survival was not significantly changed.[12] Colchicine is effective in preventing amyloidosis in familial Mediterranean fever.[13] However, the survival of patients with AL amyloidosis is marginally worse when treated with colchicine as compared to melphalan and prednisone.[14]

Melphalan and prednisolone were given in Cases 1 and 2 and cyclophosphamide in Case 3. In the first case, amyloidosis progressed despite treatment. In Case 2 the patient died before any response to treatment could have been expected. In Case 3 the patient has remained relatively well following the course of cyclophosphamide, but it is not possible to know whether this can be attributed to the drug.

We thank Dr Claire Hill for advice on the histopathology of amyloidosis.

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Case report

Salmonella in a chicken hatchery

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Due to concern about increasing mortality among 7-day-old chicks in a chicken hatchery the management arranged to screen all hatchery staff for salmonella excretion. The hatchery is part of a complex involving all stages of chicken meat production but is physically separate from the broiler units and chicken processing plant. The hatchery also has its own staff, there being no exchange of staff between it and other parts of the complex. The hatchery receives eggs from a variety of local farms and the day-old chicks are subsequently distributed throughout Northern Ireland. Approximately 200,000 chicks per week are hatched in the unit.

Regular bacteriological analysis is performed on eggshell, fluff, blood and faeces found in the hatchery. This has, in the past, revealed periodic salmonella infection; usually *S typhimurium* and *S enteritidis*. Routine bacteriological monitoring of chicken feed had proved negative. The management have regarded a mortality rate of 1% among 7-day-old chicks as acceptable. Recently the mortality rate rose to 5 – 10%. To exclude the possibility that staff could be introducing salmonella into the hatchery, management decided to screen the staff for salmonella excretion.

CASE STUDY

Twenty-three staff were employed in the hatchery. None had recently been on sick leave. Twenty-one submitted specimens and 11 were found to have salmonella in their faeces. *S enteritidis* (phage type 4) was isolated from nine, one was excreting *S infantis* and one *S typhimurium* RDNC.

The specimens were cultured in a hospital laboratory and by long-standing arrangement the cases of salmonella excretion were notified by the laboratory to the community medicine department for further investigation. The hatchery staff were then interviewed by health visitors to ascertain the presence of any abdominal symptoms and to obtain details of household contacts. All staff were interviewed to emphasise the importance of personal hygiene. Until investigations were complete, household contacts who were employed as foodhandlers and children under seven were excluded from work/school. Stool samples were only obtained from contacts if they had abdominal symptoms or were in the above groups.

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All contacts were asymptomatic and their stool samples were negative for salmonella. Only one member of staff, excreting *S enteritidis,* had abdominal symptoms. He had had intermittent upper abdominal pain, vomiting and semi-formed motions for one year. His wife and two children had had several episodes of diarrhoea and vomiting during the past six months. However, his family were asymptomatic and culture negative at the time of examination.

Several months earlier the six-week-old baby girl of a local farmer had contracted *S typhimurium* RDNC. No other family members were affected. Her father had recently received 10,000 day old chicks from this hatchery. Approximately 200 of these chicks died in the first week and subsequent postmortem examination on five chicks revealed chronic yolk sac infection with *S typhimurium* RDNC.

COMMENT

Consumption of poultry products has increased dramatically over the last 10 – 15 years and this has led to the development of a highly intensive industry. Many episodes of food poisoning are due to salmonella infection and poultry is frequently the food vehicle. Modern processing plants can have a throughput of 10,000 birds per hour and hatcheries can contain thousands of birds. Thus there is a considerable risk of cross-infection occurring and once salmonella infestation occurs in a plant it is difficult to eradicate.

It is impossible to state the exact incidence of salmonella infections in chickens as often the birds are asymptomatic.[1] In Canada 15 – 39% of retail chicken carcases have been found to be contaminated with salmonella.[2] However, the current prevalence of salmonella in chicken carcases in Northern Ireland is not known. Eggs from infected flocks can be contaminated during laying, or from infected litter, dust and equipment on the production site, as motile salmonella can penetrate the shell. Contaminated poultry feed is another source of infection. Spread to healthy chickens is particularly likely to occur during hatching when chicks are breaking through the shell. Stress such as handling, transportation and overcrowding tend to increase shedding of salmonella from infected chickens.[3, 4]

Very little has been described in the literature concerning hazards to staff working in poultry plants, yet they frequently seem to work with poultry in which salmonella infestation is endemic. The organism can be cultured from fluff, floor litter, water and poultry house dust. It has been suggested that when fluff and meconium collected at the hatchery are contaminated with salmonella, it is likely that day-old chicks are infected. When such birds are processed the carcases may well be contaminated.[3]

In this hatchery 48% of staff had a positive stool culture for salmonella when first screened. Repeat specimens from this group also revealed salmonella in most instances. This pattern was more suggestive of intermittent excretion of salmonella, rather than intermittent detection. It is likely that staff acquired the infection from their work as both *S enteritidis* and *S typhimurium* had been periodically observed in the unit over the past year. Also, staff excreting *S enteritidis* had a common phage type suggesting a common source. *S infantis* had been recently noted in other parts of the complex.

There was a surprising number of staff asymptomatic despite working in an infected environment and excreting salmonella. There was no evidence to suggest spread of salmonella to close household contacts and this probably relates to good personal hygiene. None of the household contacts of hatchery

staff included a young baby. The six-week-old baby described earlier was bottle-fed and was thus probably at higher risk of gastrointestinal infection. Her father, who handled the chicks, admitted that he let the baby suck his fingers and this was probably how the baby acquired the infection.

Unfortunately because of the fluctuating mortality among the chickens it is impossible to conclude whether the health visitors' efforts in reminding the employees of the importance of personal hygiene were responsible for any reduction in mortality rates. Nearly half of the hatchery staff were excreting salmonella and it is conceivable that most staff would be salmonella excretors. This study highlights the importance of maintaining good personal hygiene while working in a salmonella contaminated environment. Salmonella are easily removed from hands by simple handwashing with soap and water.[5]

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BOOK REVIEWS

AIDS: questions and answers. 2nd ed. By V G Daniels. (pp 116. £5.95). Cambridge: Cambridge Medical Books, 1987.

This book is written in an easy to read and informative style, and is intended for non-medical readers. It covers all the social questions that are inevitably asked, and answers them clearly. The sections on the more specifically medical aspects would probably not mean much to the lay reader, and even the appendix on medical terms would not be comprehensible to most of the public. For the medical reader this section provides a useful guide to further reading which may be required.

The author has provided some very helpful information in the appendices with regard to additional reading, and the section on useful addresses would be very helpful for those worried about this condition. Overall, this is as good a book as I have seen aimed at this level of readership.

RDM

AIDS: the acquired immune deficiency syndrome. 2nd ed. By V G Daniels. (pp 188. £11.95). Lancaster: MTP, 1987.

This is a very light-weight book which appears to be cashing in on the increased awareness and interest in the acquired immune deficiency syndrome. It is aimed at the 'informed layman' rather than the medical profession. The low-key information does not add substantially to knowledge of the subject.

DIHS

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BOOK REVIEWS

AIDS: questions and answers. 2nd ed. By V G Daniels. (pp 116. £5.95). Cambridge: Cambridge Medical Books, 1987.

This book is written in an easy to read and informative style, and is intended for non-medical readers. It covers all the social questions that are inevitably asked, and answers them clearly. The sections on the more specifically medical aspects would probably not mean much to the lay reader, and even the appendix on medical terms would not be comprehensible to most of the public. For the medical reader this section provides a useful guide to further reading which may be required.

The author has provided some very helpful information in the appendices with regard to additional reading, and the section on useful addresses would be very helpful for those worried about this condition. Overall, this is as good a book as I have seen aimed at this level of readership.

RDM

AIDS: the acquired immune deficiency syndrome. 2nd ed. By V G Daniels. (pp 188. £11.95). Lancaster: MTP, 1987.

This is a very light-weight book which appears to be cashing in on the increased awareness and interest in the acquired immune deficiency syndrome. It is aimed at the 'informed layman' rather than the medical profession. The low-key information does not add substantially to knowledge of the subject.

DIHS

staff included a young baby. The six-week-old baby described earlier was bottle-fed and was thus probably at higher risk of gastrointestinal infection. Her father, who handled the chicks, admitted that he let the baby suck his fingers and this was probably how the baby acquired the infection.

Unfortunately because of the fluctuating mortality among the chickens it is impossible to conclude whether the health visitors' efforts in reminding the employees of the importance of personal hygiene were responsible for any reduction in mortality rates. Nearly half of the hatchery staff were excreting salmonella and it is conceivable that most staff would be salmonella excretors. This study highlights the importance of maintaining good personal hygiene while working in a salmonella contaminated environment. Salmonella are easily removed from hands by simple handwashing with soap and water.[5]

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DIHS

The Ulster Medical Journal, Volume 56, No. 2, pp. 160 – 162, October 1987.

Case report

Hereditary renal and retinal dysplasia — the Senior-Loken syndrome

Angela H Bell, P J McKiernen, J M Savage, M McC Reid

Accepted 30 June 1987.

The association of nephronophthisis (medullary cystic disease of the kidney) and tapeto-retinal degeneration was first described by Senior[1] and by Loken[2] in 1961. Tapeto-retinal degeneration covers several hereditary disorders characterised by degeneration of the choroid and retina leading to blindness. We present here the case of a boy known to have Leber's amaurosis, a severe form of retinal degeneration, and mental retardation, who presented with chronic renal failure. This is the first description of the condition in Ireland.

CASE HISTORY

An eleven-year-old boy was admitted to Ava Paediatric Medical Unit, Belfast City Hospital, with a three-day history of malaise, nausea, and vomiting. On the day of presentation he had a number of generalised twitching episodes.

Since the age of two years he had been recognised as being developmentally delayed. Poor vision was brought to the attention of ophthalmologists at age five years and a suspected diagnosis of Leber's amaurosis was confirmed by an electroretinogram at age eleven years. There had been no suspicion of renal disease at this time. Urinalyses performed when he was aged seven and aged nine years were normal. He was the third of six children of unrelated parents and there was no family history of renal or eye disease. He was attending a school for visually handicapped children and had profound visual and educational handicaps.

On examination he was having tetanic spasms. He was dull and lethargic. He was clinically anaemic but not dehydrated. Blood pressure was 160/110 mmHg. Visual acuity was poor, with a right-sided strabismus. Fundoscopy showed pigmentary disturbance in both macular areas with diffuse peripheral atrophy. The remaining physical examination was normal.

Investigations showed serum sodium 137 mmol/l, potassium 3.5 mmol/l, urea 62.0 mmol/l (normal range 2.5 – 6.6 mmol/l) and creatinine 836 μmol/l (20 – 80 μmol/l). Serum calcium was 1.4 mmol/l (2.2 – 2.7 mmol/l), phosphate 3.3 mmol/l (1.1 – 1.9 mmol/l) and alkaline phosphatase was $>$ 430 iu/l (56 – 190 iu/l), indicating early renal osteodystrophy. He had a normochromic

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normocytic anaemia, haemoglobin concentration 6.2 g/dl. Urinalysis showed the presence of blood and 1 g/l of protein, and culture was sterile. In a 24-hour collection of 2.4 litres, urine sodium concentration was 160 mmol/l, potassium 30 mmol/l and protein 1.43 g (normal $<$ 150 mg). Creatinine clearance was 6.6 ml/min/1.7 m^2 (normal 98 – 150 ml/min/1.7 m^2). Renal ultrasonic scan showed two kidneys of normal size with increased echogenicity and loss of definition of the cortico-medullary junction.

These findings were consistent with acute-on-chronic renal failure, with tetany secondary to the resultant hypocalcaemia. Management consisted of administration of calcium supplements, phosphate binding agents, a high fluid intake, 1-alpha hydroxycholecalciferol, nifedipine to control the hypertension and subsequently sodium bicarbonate and potassium supplements with a protein-restricted diet. With this regimen his symptoms were controlled and blood pressure, serum calcium and phosphate levels returned to normal. Serum urea fell to 26 mmol/l, but creatinine remained grossly elevated at 775 µmol/l. Urinary output continued to exceed 2 litres daily, with persistent loss of sodium and potassium. Since discharge he has been reasonably well but no further improvement in renal function has occurred. A live donor graft from a relative has been offered for him should his condition deteriorate.

DISCUSSION

The syndrome described by Senior and Loken is a rare disorder combining a renal disease resembling familial juvenile nephronophthisis (previously known as medullary cystic disease) with tapeto-retinal degeneration. The term tapeto-retinal degeneration covers a group of familial and hereditary disorders characterised by progressive degeneration of the choroid and retina but varying in nature and severity. It is inherited in an autosomal recessive manner and progresses at a variable rate. Tapeto-retinal degeneration and familial juvenile nephronophthisis have been associated with other abnormalities including peripheral dysostosis,[3] mental retardation[2] and cerebellar ataxia,[4] mental retardation being present in the case described. The heterogeneity of this condition is further evidenced by the variable age at onset of the retinal lesion. In some families this may be congenital whereas in others its onset is in childhood.[5]

The earliest presenting signs of the renal component are polyuria and polydipsia secondary to defective urinary concentrating ability. Thus in a child with nausea, vomiting and poor intake, rapid dehydration is a hazard and in this instance precipitated acute-on-chronic renal failure. The onset is insidious and most cases do not present until renal failure is advanced. Urinalysis is essentially normal with no haematuria or albuminuria until late in the illness. Invariably there is excessive urinary loss of sodium. Anaemia, hypocalcaemia and hyperphosphataemia are proportional to the degree of renal failure. Nephronophthisis will not recur after renal transplantation.

The ocular component of Senior-Loken syndrome may be either Leber's amaurosis, retinitis pigmentosa or retinitis punctata albescens.[6] The case described here had Leber's amaurosis, a severe form of tapeto-retinal degeneration leading to blindness in early infancy. In Leber's amaurosis nystagmus and visual impairment usually occur in early infancy, the severity being disproportionate to the ophthalmoscopic changes. Ophthalmoscopy shows diffuse fundal pigmentation with pallor of the optic disc and narrowing of the retinal arteries. These changes may not be apparent until the age of six despite

severe visual impairment. The electroretinogram is characteristically flat at an early stage of the disease.

Awareness of the possibility of familial juvenile nephronophthisis in patients with Leber's amaurosis is important because it usually leads to death in the first or second decade. It is now agreed that children with nephronophthisis should undergo detailed ophthalmic assessment, including an electroretinogram. Children with primary tapeto-retinal degeneration should have measurements of blood pressure, urinary concentrating ability and renal ultrasound scan. Early diagnosis, control of hypertension and restriction of protein intake may prolong the time before renal replacement therapy is necessary.

We acknowledge the assistance of Mr J Briars, FRCS, for ophthalmological diagnosis and advice, and are grateful to Lynda Thompson for typing the manuscript.

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The Ulster Medical Journal, Volume 56, No. 2, p. 163, October 1987.

Pages from the past — Dr R S Allison

Extract from the memoirs of the late Dr R S Allison, physician to the Royal Victoria Hospital. (Edited by Dr John Logan, Honorary Archivist).

Doctor R S Allison (1899-1978) was an exceptionally able and hardworking physician, on the staff of the Royal Victoria Hospital from 1930 to 1964. His first interest was in gastroenterology. This was superseded by neurology and for his neurological work he became nationally and internationally known. He was the first full-time neurologist in the Royal Victoria Hospital. It was he who built up the large and effective department which now exists. He had a life-long interest in the sea and in the Royal Navy. Among his many publications is his invaluable history of the Royal Victoria Hospital, *The seeds of time.* His success was due to his linguistic gifts, to his devotion to the study of disease and the care of patients, and to his ability to command and organise.

PRACTICE IN A SOUTH WALES VALLEY IN 1922

A J Cronin in his book *The citadel* gives a good description of the rigours of practice in the mining districts of South Wales. In the early part of 1922 I did two months' locum in the small mining town of Abertillery. There was one other assistant. The principal in the practice, an old Scotsman with a strong face and a twinkle in his eyes, was confined to bed with influenza and pneumonia. I remember that his wife was most parsimonious in the meals she prepared for us.

Abertillery itself was no health resort, being built along a cleft in the hills surrounding it, and depending on six coal pits for the employment of its work-people. In January the sun would set behind the western range of hills, leaving the valley for the long hours of succeeding darkness wreathed in mists and vapours escaping from the coalpits. Our days varied very little. Each of us was given a list of patients to call on before we set out after breakfast, our journeys being short, and involving going on foot up and down the different terraced streets composing the town, knocking on one door after another, spending only the minimum of time with each patient and rarely, if ever, making an examination. In this way we succeeded each in making thirty or forty visits between morning and afternoon surgery. This gave us little time for reflection so that we grew to loathe the place. One Saturday afternoon I was unwise enough to offer to referee a local rugby match between the town of Crosskeys and Abertillery, and became involved in a brawl which broke out between the players of the conflicting sides. In the morning and evening surgeries it was customary for many of the patients to bring their greyhounds, each on its leash, with them into the surgery. It seemed indeed that I had sunk very low and abrogated my responsibility as a keen young doctor in such connivances. There was no sorrow in my heart when the two months ended, and I was released, feeling a great sense of satisfaction at having earned for my two months' toil the goodly fee of £100.

I gained experience in doing short locums in other practices and especially memorable was one in the heart of Suffolk where I was very happy. The doctor who was away kept a well-appointed house and in autumn, when the mellowing afternoon sun lit up the fading beech leaves, the countryside was beautiful — full

Continued on page 166

Malaria. Edited by G T Strickland. (pp 279. £15.00). London: Saunders, 1986. (Clinics in tropical medicine and communicable diseases, 1: 1).

The greatest disappointment of post-war preventive health measures is that malaria is again reinfesting the areas from which it was so painstakingly driven by sustained public health effort following the Second World War, and again a majority of the world's population live under its shadow. This is explained in the excellent chapter on the epidemiology of malaria and proper attention is given to the changing ways in which the illness may be propagated, in particular the influence of its second vector, the modern jet plane.

As the clinical descriptions in this book so amply demonstrate, malaria has a very protean symptomatology but its early recognition is essential since if, unrecognised and thus untreated, Falciparum malaria can be rapidly fatal especially to those reared in non-endemic areas. The same changes responsible for the changing epidemiology and treatment of malaria have largely altered the clinical picture, outdating descriptions in the standard medical textbooks and invalidating much of what we were once taught as undergraduates. Thus, a book providing an up-to-date account of the clinical presentations, management and prophylaxis of malaria was much needed and a busy clinician will find it hard to better this monograph as an authoritative source of information on all aspects of malaria. The chapter on the current status of malaria prophylaxis and the distribution of drug resistance should be required reading for anyone offering advice to prospective travellers, while the extensive exploration of the immunopathology and pathophysiology of malaria is well worth reading to gain some idea of how far the modern understanding of an infectious disease process can be extended.

Sensibly hard-backed to stand up to the heavy use it will undoubtedly receive as a reference source on all aspects of malaria, the book should be included in any hospital or health centre library and represents such good value by modern publishing standards that its purchase can be strongly recommended to practising clinicians. As the first in a very welcome series of clinics in infectious disease topics, Malaria sets an exceptional standard of scholarship and clarity which the editors of the succeeding volumes will do well to match.

DAC

Knowledge representation in medicine and clinical behavioural science. Edited by L Kohout and W Bandler. (pp 211. £24.50). Cambridge, Mass: Abacus Press, 1986.

This is really a book for the enthusiast, although it will have some interest to most doctors. The authors examine many aspects of knowledge representation especially when it is relevant to information processing machines and to medical expert systems.

There is no doubt that computers have made a significant impact on the periphery of clinical practice. This book sets out to examine the background and problems related to future development of computers much more directly involved with clinical decisions. For this reason, the concepts are exciting as the scope involved is that much greater. The book itself is by a number of authors presenting different aspects of the analysis of problems and possible solutions. Parts of it are very technical and would, I suspect, only really interest people with a primary computer background. However, there are several challenging chapters concerning such things as the methodology of clinical decision making which should prove interesting to doctors with no computer knowledge. The authors also illustrate the considerable difficulty of reconciling the practical constraints of computing to the rather inexact science of the practice of medicine.

While the book as a whole is difficult to read, even a rather superficial examination is worthwhile in illustrating the state of the art and future lines of thought in true medical computing.

JDL

MEMOIRS OF DR R S ALLISON — Continued from page 163.

of narrow twisting lanes with high hedgerows between sleepy villages. Most of the gables then were painted pink — a survival I was told of Danish influences in the past. I only remember one patient there. She was a woman to whom I was called on a Sunday morning to find in labour, but she denied that there had been any interruption of her menstrual cycle, which had apparently continued uninterrupted during the pregnancy.

The Ulster Medical Journal, Volume 56, No. 2, pp. 164 – 166, October 1987.

Book reviews

Oxford textbook of medicine. 2nd ed. By D J Weatherall, J G G Ledingham and D A Warrell. (Vol I, Sections 1 – 12; Vol II, Sections 13 – 28. £95.00). Oxford: Oxford University Press, 1986.

The pride in British medicine that was established by this flagship — the first edition of the *Oxford textbook of medicine* — is rekindled by the second edition. Some rearrangement and expansion of sections have occurred. The first edition indicated that the book was intended for anyone studying or practising clinical medicine (as a first reference source for general practitioners and specialists) and this new edition amply fulfils that challenge. A section on primary care brings this discipline into its rightful place in such a textbook and helps to break down any artificial barrier that may exist between hospital and family practice medicine.

One looks for updating between the two editions and apart from increase in size there are 'improvements' throughout — almost completely new and recent references in the section on pituitary and hypothalmic disorders, an account of CT scanning in pituitary and adrenal disease, developments such as magnetic resonance imaging included in the cardiovascular disease section. The section on respiratory disease has illuminating descriptions of the pathophysiology of the airways and gas exchange. The clinical section on asthma is updated to modern concepts. Rheumatology and connective tissue disorders is expanded from eight to 15 subsections. AIDS, dealt with in only three pages, may (one hopes) be in the end a true perspective or, alternatively, this field is changing so rapidly that we can await a major assessment of this condition in a third edition.

Pleasure at a local author's inclusion, Professor David Simpson in Microbiology. Pleasure in Sir Douglas Black's quotation of a local poet — Louis McNeice — 'And green is life's own golden tree'. This book is surely a golden tree that will grow in future decades. I was so charmed with this book that I went out immediately to buy a copy as a present. It weighed 16 lb — but good value at £95.

JAW

Clinical medicine: a textbook for medical students and doctors. Edited by Parveen J Kumar and Michael L Clark, with the assistance of William F Jackson. (pp 1011. £14.95, paperback). London: Baillière Tindall, 1987.

The editors of this multi-author text from the St Bartholomew's Hospital Medical School admit that there must be a good reason to write a new textbook of medicine. They make the somewhat audacious claim that none of those currently available adequately conveys the detail and background needed for medical practice in the late 1980s and 1990s. It is difficult to see that the text lives up to this claim when there is, for example, no mention of ambulatory pH monitoring in suspected reflux oesophagitis or reference to branch chain amino acids, while at the same time it seems necessary to remind the reader that food is required to provide the body with energy. Nevertheless, the overall balance of the text is good. There are important chapters on infectious diseases incorporating advice on antibiotic chemotherapy which is up to date, and others on basics of genetics, molecular biology and immunology. The latter provides important background information for modern medical practice.

All the authors of the various sections were on the staff at Barts and this contributes undoubtedly to the generally high standard and uniformity of presentation of the text, sometimes not achieved in multi-author books. The text is clear and concise. It is well illustrated with line diagrams, radiographs and other imagery. There is an important mistake in the title of Fig 16.21 which may cause confusion in the diagnosis of thyroid disease. There are helpful tables for reference and particularly useful are clear descriptions of practical procedures in everyday clinical practice such as arterial cannulation, jugular venous puncture and liver biopsy. The chapters all contain references to further reading. These are largely monographs in appropriate topics.

In view of the amount of material which has been collated in this text, it must at the price represent very good value for money in its paperback form when compared with the more recent editions of established textbooks.

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Thrombosis and the vessel wall. C N Chesterman, guest editor. (pp 273 – 565. £15.00). London: Saunders, 1986. (Clinics in haematology, 15: 2).

Despite the small size of the volume, it contains a wealth of comprehensive detail on all aspects of vessel wall interaction relevant to thrombosis. There is an excellent chapter on the naturally occurring circulating anticoagulants and a good update of advances in clinical fibrinolysis with particular attention paid to the therapeutic aspects of coronary thrombolysis.

One of the most interesting chapters is related to thrombosis and immune disorders. This would be of particular interest to those practising in rheumatology or immunology. Another chapter is of more general interest to physicians, devoted to the pharmacology of platelet/vessel wall interaction. This gives a comprehensive discussion on the current anti-platelet drugs and their actions. The present state of therapy utilising these drugs is well set out. However, it has to be said that the book requires a detailed background knowledge of the present concepts of blood coagulation, in particular of the prostaglandin metabolic pathways and platelet membrane physiology and the complexities of the activation of coagulation factors.

I do not feel it is a book of widespread interest to the general physician or surgeon, but would be more useful to the postgraduate examination candidate. EM

Androgen metabolism in hirsute and normal females. R Horton and R A Lobo, guest editors. (pp 213 – 409. £15.00). London: Saunders, 1986. (Clinics in endocrinology and metabolism, 15: 2).

The editors of this book suggest that because it is multi-authored it may suffer from some overlap and different styles. In fact the sixteen authors have produced an excellent review of androgen metabolism which will be of interest to both dermatologist and endocrinologist.

The first half of the book contains an up-to-date summary of the hormonal basis for hirsutism, providing much information on ovarian and adrenal androgen metabolism that is not as yet readily available in any of the larger textbooks. The following three chapters relating to the peripheral action of androgens and the physiology of the pilosebaceous unit are comprehensive and easy to follow. They go some way to clarify why there is such a wide range of serum androgen levels in hirsute patients, in some of whom the levels are normal. The remainder of the book is devoted to therapy and, having made the point that in the majority of these patients the underlying abnormality lies in the end-organ response to androgens, the treatments covered are those which have an effect on the androgen receptors in skin, namely spironolactone and cyproterone acetate. The chapter on spironolactone is of particular interest as this treatment is not widely used in the British Isles. Little attention is paid to cosmetic means of treating hirsutism and acne and this is disappointing as such measures are very widely prescribed and practised.

The only major criticism of an otherwise excellent book is that it does not cover the subject of virilisation. This problem is seen not infrequently by those with an interest in the hirsute female, and is one requiring expert investigation and management. AIT

Intensive care: a concise textbook. By C J Hinds. (pp 378. £14.95). London: Baillière Tindall, 1987.

To be brief but comprehensive was the difficult task Dr Hinds set for himself in writing this textbook. He has undoubtedly succeeded in being comprehensive but 378 pages could hardly be described as brief. This is in no way a major criticism because there are no short cuts in the complexities of caring for the critically ill. This book is aimed at junior medical staff and nurses trying to familiarise themselves with one of the most taxing and demanding areas of medicine, but it contains much food for thought for the specialist. The author has served a long and in-depth apprenticeship in intensive care medicine, and his practical knowledge is clearly evident.

There are 17 chapters of varying lengths which encompass the main problems encountered in this specialised area. The text is clear and easy to read with a nice balance between theory and practice. I particularly liked the introduction which clarifies some of the more contentious areas such as equipment, staffing, costs and Aache scoring. The chapter on infection is one of the best and helps in understanding why infection is so prevalent in our units. The chapter on acute poisoning is somewhat disappointing, the section on paraquat poisoning being a 1976 editorial from the *Lancet.* This practice of quoting editorials is somewhat unusual and occurs in several parts of the text.

In keeping with a 1987 publication, the newer techniques such as HFJV, fibro-optic bronchoscopy, pulse oximetery, mass spectrometry and ultrafiltration are all included. This book is a good buy for all grades of staff involved in intensive care. Perhaps a 2nd edition could be briefer. DLC

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The first half of the book contains an up-to-date summary of the hormonal basis for hirsutism, providing much information on ovarian and adrenal androgen metabolism that is not as yet readily available in any of the larger textbooks. The following three chapters relating to the peripheral action of androgens and the physiology of the pilosebaceous unit are comprehensive and easy to follow. They go some way to clarify why there is such a wide range of serum androgen levels in hirsute patients, in some of whom the levels are normal. The remainder of the book is devoted to therapy and, having made the point that in the majority of these patients the underlying abnormality lies in the end-organ response to androgens, the treatments covered are those which have an effect on the androgen receptors in skin, namely spironolactone and cyproterone acetate. The chapter on spironolactone is of particular interest as this treatment is not widely used in the British Isles. Little attention is paid to cosmetic means of treating hirsutism and acne and this is disappointing as such measures are very widely prescribed and practised.

The only major criticism of an otherwise excellent book is that it does not cover the subject of virilisation. This problem is seen not infrequently by those with an interest in the hirsute female, and is one requiring expert investigation and management. AIT

Intensive care: a concise textbook. By C J Hinds. (pp 378. £14.95). London: Baillière Tindall, 1987.

To be brief but comprehensive was the difficult task Dr Hinds set for himself in writing this textbook. He has undoubtedly succeeded in being comprehensive but 378 pages could hardly be described as brief. This is in no way a major criticism because there are no short cuts in the complexities of caring for the critically ill. This book is aimed at junior medical staff and nurses trying to familiarise themselves with one of the most taxing and demanding areas of medicine, but it contains much food for thought for the specialist. The author has served a long and in-depth apprenticeship in intensive care medicine, and his practical knowledge is clearly evident.

There are 17 chapters of varying lengths which encompass the main problems encountered in this specialised area. The text is clear and easy to read with a nice balance between theory and practice. I particularly liked the introduction which clarifies some of the more contentious areas such as equipment, staffing, costs and Aache scoring. The chapter on infection is one of the best and helps in understanding why infection is so prevalent in our units. The chapter on acute poisoning is somewhat disappointing, the section on paraquat poisoning being a 1976 editorial from the *Lancet.* This practice of quoting editorials is somewhat unusual and occurs in several parts of the text.

In keeping with a 1987 publication, the newer techniques such as HFJV, fibro-optic bronchoscopy, pulse oximetery, mass spectrometry and ultrafiltration are all included. This book is a good buy for all grades of staff involved in intensive care. Perhaps a 2nd edition could be briefer. DLC

Thrombosis and the vessel wall. C N Chesterman, guest editor. (pp 273 – 565. £15.00). London: Saunders, 1986. (Clinics in haematology, 15: 2).

Despite the small size of the volume, it contains a wealth of comprehensive detail on all aspects of vessel wall interaction relevant to thrombosis. There is an excellent chapter on the naturally occurring circulating anticoagulants and a good update of advances in clinical fibrinolysis with particular attention paid to the therapeutic aspects of coronary thrombolysis.

One of the most interesting chapters is related to thrombosis and immune disorders. This would be of particular interest to those practising in rheumatology or immunology. Another chapter is of more general interest to physicians, devoted to the pharmacology of platelet/vessel wall interaction. This gives a comprehensive discussion on the current anti-platelet drugs and their actions. The present state of therapy utilising these drugs is well set out. However, it has to be said that the book requires a detailed background knowledge of the present concepts of blood coagulation, in particular of the prostaglandin metabolic pathways and platelet membrane physiology and the complexities of the activation of coagulation factors.

I do not feel it is a book of widespread interest to the general physician or surgeon, but would be more useful to the postgraduate examination candidate. EM

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Malaria. Edited by G T Strickland. (pp 279. £15.00). London: Saunders, 1986. (Clinics in tropical medicine and communicable diseases, 1: 1).

The greatest disappointment of post-war preventive health measures is that malaria is again reinfesting the areas from which it was so painstakingly driven by sustained public health effort following the Second World War, and again a majority of the world's population live under its shadow. This is explained in the excellent chapter on the epidemiology of malaria and proper attention is given to the changing ways in which the illness may be propagated, in particular the influence of its second vector, the modern jet plane.

As the clinical descriptions in this book so amply demonstrate, malaria has a very protean symptomatology but its early recognition is essential since if, unrecognised and thus untreated, Falciparum malaria can be rapidly fatal especially to those reared in non-endemic areas. The same changes responsible for the changing epidemiology and treatment of malaria have largely altered the clinical picture, outdating descriptions in the standard medical textbooks and invalidating much of what we were once taught as undergraduates. Thus, a book providing an up-to-date account of the clinical presentations, management and prophylaxis of malaria was much needed and a busy clinician will find it hard to better this monograph as an authoritative source of information on all aspects of malaria. The chapter on the current status of malaria prophylaxis and the distribution of drug resistance should be required reading for anyone offering advice to prospective travellers, while the extensive exploration of the immunopathology and pathophysiology of malaria is well worth reading to gain some idea of how far the modern understanding of an infectious disease process can be extended.

Sensibly hard-backed to stand up to the heavy use it will undoubtedly receive as a reference source on all aspects of malaria, the book should be included in any hospital or health centre library and represents such good value by modern publishing standards that its purchase can be strongly recommended to practising clinicians. As the first in a very welcome series of clinics in infectious disease topics, Malaria sets an exceptional standard of scholarship and clarity which the editors of the succeeding volumes will do well to match.

DAC

Knowledge representation in medicine and clinical behavioural science. Edited by L Kohout and W Bandler. (pp 211. £24.50). Cambridge, Mass: Abacus Press, 1986.

This is really a book for the enthusiast, although it will have some interest to most doctors. The authors examine many aspects of knowledge representation especially when it is relevant to information processing machines and to medical expert systems.

There is no doubt that computers have made a significant impact on the periphery of clinical practice. This book sets out to examine the background and problems related to future development of computers much more directly involved with clinical decisions. For this reason, the concepts are exciting as the scope involved is that much greater. The book itself is by a number of authors presenting different aspects of the analysis of problems and possible solutions. Parts of it are very technical and would, I suspect, only really interest people with a primary computer background. However, there are several challenging chapters concerning such things as the methodology of clinical decision making which should prove interesting to doctors with no computer knowledge. The authors also illustrate the considerable difficulty of reconciling the practical constraints of computing to the rather inexact science of the practice of medicine.

While the book as a whole is difficult to read, even a rather superficial examination is worthwhile in illustrating the state of the art and future lines of thought in true medical computing.

JDL

MEMOIRS OF DR R S ALLISON — Continued from page 163.

of narrow twisting lanes with high hedgerows between sleepy villages. Most of the gables then were painted pink — a survival I was told of Danish influences in the past. I only remember one patient there. She was a woman to whom I was called on a Sunday morning to find in labour, but she denied that there had been any interruption of her menstrual cycle, which had apparently continued uninterrupted during the pregnancy.

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