British Paediatric Association

Minutes of the Annual General Meeting

The Annual General Meeting of the British Paediatric Association was held on Thursday, 18 April, 1985 at the University of York. The President, Professor Sir Peter Tizard, took the chair.

1 OBITUARY
Members of the Association stood in memory of those Members (all Honorary) who had died since the last meeting: Professor J L Henderson (Past President), Professor O Kerpel-Fronius, Dr G H Newns, Dr R A Shanks, and Dr A White Franklin (Past President).
The President reported that the widows of three recently deceased members had accepted invitations to attend the 1985 Annual Dinner.

2 MINUTES
The Minutes of the last meeting held on 12 April, 1984, which had been circulated to members in March in the Annual Report, were accepted and signed as a correct record.

3 ELECTION OF HONORARY OFFICER
The Meeting ratified Council’s election of Dr R MacFaul to serve as Honorary Assistant Secretary 1985–8.

4 ELECTION OF NEW MEMBERS
Dr M W Artherton, Professor P T Bray, Professor F S W Brimblecombe, Dr D Clark, Dr Christine Cooper, Dr R Davie, Dr R H Jackson, Dr H R Jolly, Professor Noboru Kobayashi, Lady Lovell-Davis, Dr T P Mann, Dr D Morris, Mr H H Nixon, Dr G Jackson Rees, Professor P P Rickham, and Dr B S B Wood had been nominated as Honorary Members by Council, together with 125 Ordinary Members and 10 Associate Members. The Meeting approved the nominations and the new Members were duly elected. The President pointed out that although the number of Honorary Members elected this year was greater than usual, the total number of Honorary Members of the Association remained within the agreed ratio of Honorary to Ordinary Members.

5 TRANSFER OF MEMBERSHIP
The Meeting ratified Council’s approval of the transfer of 87 Senior Clinical Medical Officers from Associate Membership to Ordinary Membership.

6 PRESIDENT’S REPORT
The President reported on the following matters:

BPA headquarters. The move to St Andrews Place was due to take place in October 1985. The accommodation would be double the size of that at Queen Square.

Royal College of Physicians of London Appeal. Although the target for the Royal College of Physicians of London Appeal had not yet been reached, the Association had benefited greatly from several large donations for paediatric research projects. In addition the College had agreed to give the Association the sum of £15 000 annually for four years, and this would offset some of the increased outgoings involved in the Association’s occupancy of the new premises.

Consultation. A large and increased number of requests for comments from statutory and other organisations had been received by the Association. Many were dealt with directly by the Honorary and Executive Secretaries, some were compiled by the Officers following consultation with informed members of the Association, and others were sent to the appropriate Standing Committee or Specialty Group for comment. Documents compiled from these comments are approved by the Officers and sent to the consulting body or approved by the Executive Committee and Council.

The recently published limited list of prescribable drugs was discussed. Members had expressed concern about specific omissions from the list, the absence of a mechanism for appeal, or for inclusion of new drugs on the list. Members with particular concerns were asked to write to the Honorary Secretary. The Royal College of Physicians of London had approved the list in principle with the following provisos: (a) all clinical needs should be met; (b) the standards and purity of generic equivalents must be guaranteed; (c) a review procedure and (d) an appeals mechanism should be established.
Concern was expressed about the failure to consult the Association prior to publication of the limited list. The President pointed out that no medical organisation had been so consulted. In general the frequency with which the DHSS consulted the Association had greatly increased during the past two years.

**BPA policies and publications.** A number of important documents had been published during the year, some of which were circulated to members with the Newsletter.

The President thanked Professor A D Milner and Dr N R C Roberton for their work on the preparation of the Paediatric Equipment Report. Some 209 Members had requested copies.

Child health and the integration of the child health services had been a major topic of attention. It was regretted that the principle of *ad hominem* promotion to consultant rank for those SCMOs carrying out consultant work had not yet been put into effect, although the principles had been accepted by the Joint Paediatric Committee, Joint Consultants Committee, and the Central Committee for Hospital Medical Services. A document prepared by Dr W J Appleyard on the mechanism for the integration of the child health services would be circulated with the Newsletter. The continuing number of new posts of Consultant Paediatricians with a special interest in Community Child Health was welcomed.

**Relationships with other organisations.** The Association had continued to develop links with other organisations, the work of which influenced the health and welfare of children: these include the National Association of Health Authorities, the National Association for the Welfare of Children in Hospital, the Foundation for the Study of Infant Deaths, and the Casualty Surgeons’ Association. The Association’s comments had been incorporated in a document on severely handicapped infants produced by the Association of Directors of Social Services. It was hoped that the final document would be circulated to Members.

A joint working party had been established between the BPA and the British Association of Otolaryngologists. The agreement reached in 1973 between the Associations was still recognised policy. It was hoped that a new document being prepared as a result of current discussions would be presented to the Executive Committee and Council in the summer.

The Officers of the Association met the Chief Nursing Officer at the DHSS. The problems of paediatric nurse training and staffing were discussed. The Association’s view that training for sick children’s nurses should be on a regional basis had not been accepted.

**Links with industry.** The Honorary Secretary was serving on a Royal College of Physicians working party on the ethics of the relationship between physicians and the pharmaceutical industry. The Association benefited from the support of a wide number of commercial companies. The funding obtained was used for specific purposes and not for the general running of the Association.

Sponsorship was being sought for fellowship for paediatricians from third world countries similar to the Heinz Fellowships available to paediatricians in the Commonwealth.

**Research.** The Association’s research capabilities were being enhanced by the move to St Andrews Place and by generous donations. A Steering Group had been established to coordinate investigation on the surveillance of rare communicable and non-communicable disease in childhood.

**Archives of Disease in Childhood.** Professor S R Meadow expressed the appreciation of the Editors of the Archives for the assistance of the members of the Editorial Board. Some 1000 manuscripts were submitted each year, only one third of which could be published. Members were urged to send only their best papers and help to ensure that the journal represented the best of British paediatrics.

**Joint Paediatric Committee of the Royal Colleges of Physicians in the UK and the BPA.** Professor J O Forfar reported that over the last year the Joint Paediatric Committee had continued to seek the support of the Royal Colleges for proposals and suggestions put forward by the BPA. The views of the Association were voiced on certain national bodies through representation effected through the Joint Paediatric Committee. The Colleges had generally supported the Association’s views and efforts were made to ensure that a unified view is put forward by the paediatric committees of the Royal Colleges. A document on training for community health doctors had been approved. Professor C E Stroud had replaced Professor Forfar as Chairman. The President expressed the Association’s debt of gratitude to Professor Forfar for his chairmanship of the Joint Committee since its inception in 1979.

**Diamond jubilee 1988.** It was the view of Council that the Association’s Diamond jubilee should be recognised. A Standing Committee has been established under the convenership of Professor J O
Forfar. Membership also included Dr J D Baum, Sir Alan Ray, Professor C E Stroud, and the Honorary Secretary. The Committee’s first meeting had taken place. It was seen as appropriate that the occasion provided an opportunity for celebration, the bringing up to date of the History of the Association and the production of a major report on a number of questions of importance in paediatrics and child health. Suggestions from members would be welcomed by the Committee. Professor S D M Court requested that an updated History of the Association should contain an account of the substantial improvement in child health over the period covered by the publication.

7 TREASURER’S REPORT
The Treasurer reported on the Accounts for the period 1 January to 31 December 1984, which had been circulated to members with the March Newsletter and had subsequently been audited without alteration. During the last year the financial policy described at the last Annual General Meeting had been put into effect. The Annual Meeting was self financing, and the Association’s other activities were covered by members’ subscriptions and investment income. Reserves had been built up to a level equal to one year’s turnover. Profits on sales of the Archives were to be used for specific projects, including the move to St Andrews Place. The Association was indebted to the Archives for its financial contribution to the work of the BPA.

The outcome of discussions with HM Customs and Excise had been that the Association was not required to register for VAT.

The members’ subscriptions were virtually paid up to date and Mrs Doreen Cope was thanked for her persistence in achieving this satisfactory position. The Association’s investments had been kept in liquid form to take advantage of high interest rates on longer term deposit accounts. The Association would take further advice on its investments.

The Honorary Treasurer thanked Mr Michael Ray, the Association’s financial adviser, and the Secretariat for their hard work in preparing the accounts.

The Honorary Treasurer’s verbal account and the printed Accounts for 1984 were accepted.

8 MEMBERS’ SUBSCRIPTIONS
The Meeting agreed that the annual subscription rate for Ordinary Members should be increased to a point between £85 and £90 from 1 January 1986, with increases pro rata for the lower rates; the Honorary Treasurer would present the final figure decided upon to Council for ratification at its autumn meeting.

9 RULES OF THE ASSOCIATION
The Meeting considered amendments to the Rules of the Association which had been approved by solicitors and the Charities Commissioners. After full discussion:
(a) A proposed amendment 2(c) Termination of Membership was accepted.
(b) A category of Senior Membership was approved. It was agreed that only Ordinary Members should in future be eligible to vote and to stand for election as Honorary Officers (with the exception of the office of President).

10 REPORT OF COUNCIL
The Reports of Council and of the Academic Board which had been circulated to Members in March 1985 were received. The President thanked members of these two Committees for their hard work during the year.

11 ANNUAL REPORTS OF STANDING COMMITTEES, WORKING PARTIES, PAEDIATRIC SPECIALTY GROUPS, AND OUTSIDE BODIES 1984-85
The Meeting received reports which had been circulated in the Annual Report to Members in March 1985. All those who had served were thanked for their valuable contribution to the work of the Association.

12 ANNUAL MEETING, 1990
The Honorary Secretary reported that the University of York could not accommodate the BPA Annual Meeting in 1990 on dates convenient to the Association. The Honorary Secretary illustrated the alternative facilities which had been investigated and the Meeting agreed that the 1990 Annual Meeting should be held at the University of Warwick.

13 CONSUMER QUESTIONNAIRE
Dr J D Baum urged Members who had not already done so to complete the questionnaire about the Annual Meeting which had been prepared by the Wyeth Research Secretary.

Dr J Martin expressed the gratitude of the Association to Sir Peter Tizard on his retirement as President. During his term of office Sir Peter had been an outstanding leader; at Council and regional meetings alike he had always put the aims of the Association first and throughout his Presidency had been an outstanding representative of the interests of children in this country.

The meeting closed with a standing ovation for the retiring President.

1 OBITUARY
Professor J L Henderson (Honorary Member since 1945 and Past President); Professor O Kerpel-Fronius (Honorary Member since 1958); Dr G H Newns (Honorary Member since 1945); Dr R A Shanks (Honorary Member since 1951); Dr A W Franklin (Honorary Member since 1938 and Past President).

2 COUNCIL
The following members of the Association have served on Council during 1984–5: Sir Peter Tizard (President); Dr D P Addy; Dr Margaret Anderson (Associate Members’ Representative); Dr Patricia Anderson (Associate Members’ Representative); Dr W J Appleyard (Honorary Treasurer); Dr J D Baum (Chairman to the Academic Board); Dr J K Brown; Dr J L Burn; Dr T L Chambers (Honorary Secretary); Dr R W I Cooke (Secretary to the Academic Board); Dr D W Fielding (Honorary Assistant Secretary); Professor Emeritus J O Forfar (President Elect); Dr J F T Glasgow; Dr G W Hatcher; Dr Lilian Jones; Dr J A Kuzemko; Dr I J Lewis (Junior Paediatric Staff Representative); Dr R MacFaul; Dr E McKay; Dr J Martin; Dr S T Meller; Dr D H Mellor; Dr Marion Miles (Honorary Assistant Secretary); Dr C L Newman; Dr R E Oliver; Dr E M Ross; Dr D C L Savage; Dr A L Speirs; Dr E Tempany; Dr R G Welch; Dr V L Woolley.

Observers. The Association is grateful to the following for assistance and advice both at Council and in many other ways:
Dr Margaret Bell (Department of Health and Social Services, Northern Ireland); Mr D M Forrest, (President, British Association of Paediatric Surgeons); Dr D M Gambier (Welsh Office); Dr J R Harper (Central Committee for Hospital Medical Services); Professor S R Meadow (Editor of the Archives of Disease in Childhood); Dr J S Metters (Department of Health and Social Security); Professor T E Oppé (Adviser in Child Health to the Chief Medical Officer, DHSS); Mr J E S Scott (Past President, British Association of Paediatric Surgeons); Professor H Simpson (Association of Clinical Professors and Heads of Departments of Paediatrics); Professor C E Stroud (Standing Medical Advisory Committee); Dr O A Thores (Scottish Home and Health Department); Dr C Waine (Royal College of General Practitioners); Dr A Yarrow (Department of Health and Social Security).


3 MATTERS CONCERNING THE ADMINISTRATION OF THE ASSOCIATION
The Association’s staff comprises an Executive Secretary, a Senior Assistant Secretary, an Assistant Administrator (Membership), two Assistant Secretaries, the Wyeth Research Secretary, a part time Librarian, and a Junior Clerical Assistant.

It is expected that the Association’s offices will move to 5 St Andrew’s Place, Regent’s Park, during the course of 1985.

The BPA would again like to record its debt of gratitude to the Dean of the Institute of Child Health for the use of facilities.

4 ACADEMIC BOARD
The 19th Annual Report of the Academic Board has been received by Council and is published as an Appendix.

5 AWARDS OF THE ASSOCIATION
James Spence Medal. Council was pleased to nominate Dr N S Gordon to receive the medal in 1985.

Donald Paterson Prize. The decision of the adjudicating panel was that Dr P T Rudd should be awarded the Prize in 1985 for his paper on ‘A prospective study of chlamydial, mycoplasmal, and viral infections in a neonatal intensive care unit’.

Heinz Fellowships of the BPA. Heinz Fellowships for 1984–5 have been awarded to Dr S H Ahmad (India) and Dr A M Ramal (Jamaica)—Fellowship A. No C Fellowships were awarded for 1984–5. The BPA continues to be grateful to the Heinz Company for providing funds for these valuable Fellowships.

6 FINANCE AND ALLIED MATTERS
The Association and paediatrics have benefited greatly from the Royal College of Physicians of London Appeal. The National Medical Research Fund has granted £250 000 to establish and fund two Research Fellowships in Paediatrics, to be jointly administered by the RCP and the Association. An anonymous donation of £92 000 over four years towards the research activities of the Association and a donation of £7500 earmarked for paediatrics from The Mercers’ Company have also been received.

The Association is indebted to Cow & Gate Ltd for meeting the costs of the programme, abstract forms and stationery for the 1985 Annual Meeting and the Directors of Unigate Ltd have again donated their annual travel grant (£250).
The Association is extremely grateful for the donation of £4000 from the Trustees of the Moor-gate Trust Fund.

Other donations were received from Delta plc (£100) and the Hunting Group Charitable Trust (£60).

The BPA continues to benefit from a share of the profits of the sale of the *Archives of Disease in Childhood*.

7 MEETINGS OF THE ASSOCIATION
The 56th Annual Meeting of the Association was held at York University in April 1984. Some 945 members and guests attended. Council thanks the Academic Board for organising the scientific programme and the many members and guests who submitted papers. Council also wishes to thank the conveners and members of the paediatric specialty groups (13 of which held sessions during the meeting) for their contributions.

8 STANDING COMMITTEES AND WORKING PARTIES OF THE ASSOCIATION
The following were established during the year:

Standing Committee on Manpower;
Diamond Jubilee Committee (the Association’s Diamond Jubilee will be celebrated in 1988);
Working Party in Liver Transplantation in Children;
Working Party on Prescribing for Children;
Working Party on Intensive Care for Infants and Older Children in the UK.

9 MATTERS CONCERNING GOVERNMENT DEPARTMENTS AND OTHER BODIES
A meeting between the Officers of the BPA and Mr John Patten MP, Parliamentary Under Secretary at the Department of Health and Social Security, was held at the DHSS on 21 May 1984. Comments on or to the following have been submitted:

Maternity Services Advisory Committee
Second Report of the Maternity Services Advisory Committee: ‘Maternity Care in Action’. Part II: Care During Childbirth (Intrapartum Care).

House of Lords
The Association was represented at a meeting of the All Party Parliamentary Group for Children held at the House of Lords to discuss ‘Mothers and babies in prison’.

House of Commons

Social Services Committee Report on ‘Children in Care’ (April 1984) and the Government’s Response (July 1984).

Standing Intercollegiate Committee on Oncology
Appendix to the 3rd Report of the Standing Intercollegiate Committee on Oncology on ‘Professional Recognition of Training in Terminal Care of Cancer Patients’.

Department of Health and Social Security
Report of the Committee of Inquiry into Human Fertilisation and Embryology; Draft Code on Confidentiality of Personal Health Data; Working Group on Performance Indicators; Proposals to Limit the Range of Prescribable NHS Drugs.

Standing Medical Advisory Committee
Working Group on Acute Services for Cancer.

Royal College of Obstetricians and Gynaecologists
Guidance Notes on the Management of Perinatal Deaths; Recommendations for Confidential Regional Reviews into Perinatal Mortality.

Royal College of Obstetricians/BPA/Royal College of General Practitioners Joint Working Party

Association of Directors of Social Services
Social Work and Severely Handicapped Infants.

Royal College of Physicians of London/Faculty of Community Medicine
Training for Community Child Health.

10 BPA DOCUMENTS APPROVED BY COUNCIL
Paediatric Manpower Towards the 21st Century; The Integration of Child Health Services; Report based on Survey of Children’s Attendants at Accident and Emergency Departments; Guidelines for the Workload of Consultant Paediatricians; Mechanism for Integrating the Child Health Services: A Policy Statement.

Council is grateful to the members who have served on committees and working parties, and also to those who have represented the Association on both statutory and voluntary bodies. Many individual members have helped the Association by their advice, suggestions, and criticisms.

Council again wishes to record its special appreciation of the work of the Editors of the *Archives of Disease in Childhood*, the journal continues to be of the greatest value to British paediatrics.

Finally Council would like to express its thanks to the Secretariat for their loyal service to, and continued hard work for, the Association.
Scientific proceedings

Oral presentations


Since December 1983, in an extension of an adult liver transplantation programme, 14 children with terminal liver disease have received orthotopic grafts. There were nine girls and five boys, aged from 7 months to 17 years; six had biliary atresia, three alpha-1 antitrypsin deficiency, two hepatic tumours, one chronic active hepatitis, one post-viral hepatic necrosis, and one Budd Chiari syndrome.

Donors were matched only for size and major blood groups. Recipients received prednisolone and azathioprine, a 10 day course of an anti-T cell monoclonal antibody (Campath I) and later introduction of cyclosporin A.

There were no operative deaths but two children died in the early postoperative period. One child died at five months from metastatic hepatocellular carcinoma. Two children had second transplants, one at five days for graft infarction and one at six months for rejection, both are doing well.

Eleven children are alive 10 days to 12 months after operation. Complications have included problems with biliary drainage, infection, and rejection but nine are currently well.

Liver transplantation presents formidable problems but offers the only therapeutic option for a small number of children. The programme is critically dependent on the supply of donor organs of suitable size.

Thirty eight different metabolic errors (previously fatal) have been corrected by displacement bone marrow transplantation. J R Hobbs (London).

The birth of some 80% of children with metabolic errors is not preventable. Elective transplants into fit recipients from matched sibling donors have for four years achieved 95% survival, but in emergency, or 'end of the line' transplants, only 60% survive. This emphasises the importance of early diagnosis and identification of such a possible donor.

Half-matched relatives (sharing one genetic haplotype) can be used as donors with overall survival around 50%, but improving to up to 68%. In this situation, cyclosporin-A together with donor marrow processing has the best results, and possible advantage is a non-carrier donor. For such transplants, aseptic isolation is mandatory due to the longer and more severe immunosuppression required. Patients should be referred where possible before they have acquired infections, especially of the herpes group of viruses, which can break out and complicate the issue. Sensitisation of the recipient should be avoided as secondary antibody responses cause major problems.

Diseases where successful correction has been achieved do not include the mucopolysaccharidoses (still being assessed), and a further 40 different diseases.


In 1980 the United Kingdom Children’s Cancer Study Group (UKCCSG) started a new treatment programme for patients with Wilms’ tumour in which patients were stratified by histology as well as stage. For patients with localised disease and favourable histology (FH) the accent was on treatment refinement: stage I patients received no radiotherapy (RT) and six months of Vincristine (Vcr) alone; patients with stage II and III received abdominal RT 20 Gy and combination chemotherapy with Vcr and Actinomycin D (stage II) together with Adriamycin (stage III). Patients with metastatic disease (stage IV) and all those (15%) with unfavourable histology (UH), of whatever stage, received these three drugs plus Cyclophosphamide; radiotherapy for UH patients was reserved for those with residual disease at second look laparotomy. One hundred and fifty patients have now been off treatment for more than one year and results are encouraging for patients with FH and local disease with actuarial survival at 96% for stage I, 91% for stage II, and 81% for stage III; there have been no flank relapses in stage I and only one in stage II. Survival in stage IV is disappointing (47%), while the survival of 55% for all cases with UH is probably an improvement over the previous MRC study. We conclude that reduction of treatment for children with stages I–III and favourable histology has not compromised efficacy but that new approaches are needed for the minority of patients with stage IV and unfavourable histology.

The natural history of ventricular septal defects—a long term prospective study. G R Sutherland, M J Godman, B R Keeton, and J E Burns (Southampton and Edinburgh).
Studies into the natural history of ventricular septal defects (VSD) have been limited by the lack of a non-invasive method whereby defects could be identified and classified, and closure mechanisms determined. 2-D echo can potentially supply this information. Since June 1981, 354 consecutive infants, with clinical evidence of VSD, have been enrolled into a prospective patients echo study. In 135 a VSD was not visualised. The following VSDs were visualised and identified—perimembranous inlet (PMI) 64; perimembranous outlet (PMO) 28; perimembranous confluent (PMC) 22; muscular inlet (MI) 7; muscular outlet (MO) 13; trabecular (T) 85 (small 66, large 19).

Serial follow-up (mean=1.9 years) showed 47 defects closed spontaneously (10 T; 5 PMI; 2 PMO; 1 MO and 29 presumed small T defects). Overall spontaneous closure rate/year (year 1=9.7% year 2=7.3%) was calculated. Thirty three defects required surgical closure (11 PMO; 12 PMC; 2 MI; 1 MO; 3 PMI and 4 large T). Analysis of the findings indicated that differing VSDs have differing rates and mechanisms of closure. Perimembranous VSDs with an inlet component closed by involving accessory tricuspid tissue. Trabecular defects closed by direct muscle ingrowth. Size reduction was clearly related to defect type but not defect size.

In conclusion: 2-D echo not only identifies and classifies virtually all haemodynamically significant VSDs but also determines specific closure rates and closure mechanisms for each type of VSD.

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Almost all the paediatricians circulated have returned the BPA questionnaire enquiring about cystic fibrosis (CF) patients under their care. Questionnaires have also been returned from members of the British Thoracic Association. Information has been obtained about approximately 4500 patients. Ascertaining is less complete for adult patients but the preliminary data suggest an 80% survival to age 13 years and 50% survival to age 20 years or more. Mortality is higher in females. It is estimated that approximately 1000 additional patients will enter the ‘adult’ (15+ years) age group during the next 5 years. Epidemiological data showed regional variations in incidence and prevalence of CF, and the locations of large CF clinics. Continuation of the survey for a further 2–3 years will allow accurate calculations of age at diagnosis, life expectancy, and differential mortality.

The Working Party has also considered data from other countries. It concludes that there is a need for properly established and funded referral centres for CF, although patterns of care will vary in different localities. The structure and functions of such centres will be discussed.

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Children's care by their parents in hospital. C P Q Sainsbury, J Cleary, M Davies, and O P Gray (Cardiff).

Care by parent units have been working successfully in North America for 15 years, and claim to reduce stress for children and their resident parents. Improved health education and parental confidence shortens the duration of hospitalisation and reduces the readmission rate.

A care by parent system, requiring no additional finance, accommodation or staff, was introduced into a general paediatric ward. Resident parents were given the option of assuming full responsibility for their children's care, including measuring vital signs, charting observations, and giving treatment. The nursing staff taught, supervised, and supported the parents.

The scheme was evaluated by observational studies, and questionnaires which were completed by the parents, medical, and nursing staff. The vast majority of parents coped very successfully, to the satisfaction of nursing and medical staff, and the children's benefit. Analyses of the observations of the children and parents activities and the accomplishments gained throughout the hospitalisation showed that this system offered advantages, and could become more generally used in paediatric wards in Great Britain.

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Classification of deaths in the multicentre study of post-neonatal mortality. J Knowelden, J Keeling, and J Nicholl (Sheffield).

The deaths were classified by three systems, the certified cause of death, the clinical presentation, and the necropsy findings. A review of all the available information was the best indicator of whether the death was entirely unexpected or occurred in a child who had been recognisably ill. A detailed necropsy was the chief source for determining how far the death was explained. Whereas only a few of the infants with no pathological evidence of terminal disease had clinically identified diseases,
the majority of children whose deaths were clinically unexpected had pathological lesions of terminal disease.

The entries on the death certificates were often in conflict with the clinical and pathological findings; they exaggerated the frequency of sudden unexpected death, or were poor indicators of the explanation for the deaths. The use of the terms 'cot death' or 'sudden unexpected death' as causes on death certificates needs to be reviewed. Post-neonatal deaths should be classified on two separate axes, a primarily clinical one describing how the event presented, and a primarily pathological one indicating how far the lesions discovered were thought to have contributed to the death. The term 'sudden infant death syndrome' (SIDS) should be reconsidered as it does not describe consistently the degree to which death was unexpected or how far it was unexplained.

**Principles of genetic counselling for the prevention of neural tube defects and other disorders.** K M Laurence (Cardiff).

Women (and their partners) known to be at increased risk for neural tube defect should be counselled to enable them to plan their future pregnancies, to improve their diet, to consider preconceptional supplementation with folic acid, to attend early for antenatal care and to have diagnostic tests. Counselling can be carried out by any clinician with the time, the inclination, and the knowledge but preferably separate from the general management consultation. Generally, cases for counselling which involve Mendelian inheritance with variable dominance, X linked disorders or consanguinity, rare genetic disorders or undiagnosed syndromes, carrier detection and prenatal diagnosis are perhaps better referred to the regional genetics centre.

**A clinical approach to the diagnosis of dysmorphic syndrome.** D Donnai (Manchester).

After the birth of a baby with single or multiple birth defects, it is important to make an accurate diagnosis in order to institute appropriate medical management and give accurate information about prognosis and recurrence risks to the family. Since some babies with defects are stillborn or die soon after birth, a systematic and rapid scheme for gathering clinical and laboratory information is necessary. Such a scheme includes history of family and pregnancy, examination and measurements, photographic and radiographic records, and specific laboratory investigations. The use of correct terminology is vital. Identification of rare dysmorphic syndromes is time consuming and an approach to diagnosis and literature search is needed.

**The genetics of non-dysmorphic mental retardation.**
M Baraitser (London).

Non-dysmorphic mental retardation, also called non-specific or 'pure' mental retardation, includes all who are retarded but are without specific clinical features. It contributes about 10% to severe retardation. A major inroad into the male contribution to this group has been made by the identification of the fragile X syndrome. This contributes 6% to severe retardation in males, has a low fresh mutation rate, and has a birth frequency of 1 in 2000.

Now that a laboratory test is available (chromosome analysis after culture in a folate deficient medium), it is now evident that there are recognisable dysmorphic features. These features are minor but lead to the recognition of those retarded males who would need special chromosomal analysis. Similarly, other conditions thought to be non-dysmorphic could, when an appropriate test is available, show recognisble clinical patterns that we are, as yet, unable to perceive.

**Clinical application of direct DNA analysis.**
E Pembrey (London).

At present the main clinical application of direct DNA analysis is genetic prediction within families with simply inherited disorders (eg, haemophilias A and B, Duchenne MD, OCT deficiency). Benefits derive from more reliable carrier detection, and in particular carrier exclusion; plus earlier prenatal diagnosis based on chorionic villus sampling. There are two fundamentally different approaches:

Gene tracking uses gene specific or chromosome-region-specific DNA probes to ask the question 'has the fetus (or relative) inherited the same relevant chromosome region as a previously affected family member?' There are variations to this question, but they all use a genetic linkage analysis within the family to make the prediction. Gene tracking is dependent on being able to distinguish each chromosome of the relevant pair, and naturally occurring variation in DNA sequence in intergenic DNA is exploited for this purpose. The great advantage of gene tracking over gene detection (see below) is that it is independent of the particular mutation that is disrupting the gene in question. The
disadvantage is that not all families have an informative DNA polymorphism to distinguish the homologous chromosomes.

Gene detection requires a gene-specific probe and knowledge of the particular mutation that is segregating within that particular family. This latter point is a major problem, since in most disorders we can expect different point mutations in different families with the same disease.


Beta thalassaemia trait is common throughout the Mediterranean area, and in some places thalassaemia is an important public health problem. Programmes for the prevention of thalassaemia major by heterozygote screening and counselling, together with mid-trimester fetal blood sampling and selective abortion of affected fetuses have been established in several such areas. Careful monitoring has shown 50–90% reduction in births of affected children since the inception of these programmes.

Childhood onset anorexia nervosa. B Lask, R Bryant, A Fosson, and J Knibbs (London).

Very little is known about the aetiology, presentation, course or outcome of childhood-onset anorexia nervosa. The aim of this study is to provide information on these aspects of the condition.

Case note analysis of all children seen at the hospital between 1960 and 1984 diagnosed as having anorexia nervosa revealed that 51 children fulfilled our diagnostic criteria. The age range was 7–15 years (mean age 11–8), with a sex ratio of 3–1, (girls to boys). Tanner-Whitehouse standards for nutritional status showed 85% had a weight for height ratio of under 80%, indicating severe malnutrition.

Common features of the illness included moderate to severe depression (55%), distorted body image (45%), fear of fatness (45%), and physical symptoms such as abdominal pain, nausea, vomiting, dysphagia or feelings of fullness (45%). There was a striking variation in symptomatology during the course of the illness in 33% of the sample, and the diagnosis was frequently missed. Marked family pathology was noted in half the sample.

It is clear that anorexia nervosa does occur in childhood, is difficult to diagnose, and often causes severe malnutrition.

Haemorrhagic disease of the newborn and the use of prophylactic vitamin K. D Garrow, M Chisholm, and M Radford (Southampton).

Haemorrhagic disease of the newborn results from a deficiency of vitamin K and the vitamin K dependent clotting factors, but controversy still exists as to whether healthy newborn babies should be given the vitamin. Hospitals have adopted varying policies.

The vitamin K dependent clotting factors were measured by Thrombotest on day 2 of life in 48 breast fed babies and 24 bottle fed babies. Half of each group had been given vitamin K at birth.

The untreated, breast fed babies had significantly lower Thrombotest values than the treated, breast fed babies, or either group of the bottle fed babies. Nine of the 24 untreated, bottle fed babies showed Thrombotest values below 10%, when bleeding may occur, yet none of the babies showed any haemorrhagic disease.

Administration of vitamin K did not affect the Thrombotest values in bottle fed babies.

Prophylactic use of vitamin K is considered justified in breast fed babies but is probably unnecessary in bottle fed babies.


We report a double blind, cross over, placebo controlled trial of reintroduction of provoking foods, following successful oligoantigenic diet treatment, in 28 children with hyperkinetic syndrome. Symptoms returned or were exacerbated significantly more frequently following active material than placebo. Of 76 patients so treated, 62 improved, of whom 21 were regarded as having recovered. Many had associated symptoms (headaches, abdominal pain, fits, etc) which also resolved. Forty eight foods were incriminated (nearly all children reacted to several, and two reacted to 30). Artificial colours and preservatives were the commonest provoking factor, but no child responded to these alone.


Acute encephalopathy in childhood is a major cause of handicap in the community, but the cause in previous studies has been ascertainable in less than a third of cases.
We studied prospectively and followed for 6–9 months 29 children presenting with acute encephalopathies. Using virus culture, serology, and interferon assays we were able to establish the presence of active virus infection in almost 90% of otherwise unexplained cases including 20 of 22 cases with a clinical diagnosis of encephalitis. Among these, approximately half had evidence of virus infection of the CNS and half evidence of blood-brain barrier breakdown. There was no evidence of involvement of complement, immune complexes, or autoimmune responses against neural proteins in the pathogenesis of these illnesses. Age <3 years, involvement of organs other than the CNS, depth of coma, and the absence of brain stem or cerebellar signs were significantly predictive of residual intellectual and physical handicap.

Rapid and appropriate investigation can increase the understanding of these illnesses.

**Referral and outcome in <1000 g infants. R W I Cooke (Liverpool).**

Although several series of under 1000 g (ELBW) survivors have recently been reported, most are small, from ‘tertiary’ centres, and do not take referral patterns into account.

Some 232 ELBW infants were admitted to Mersey Regional NICU over a 4 year period (January 1980–December 1983). Altogether 109 were discharged home, and represent 90% of recorded survivors <1000 g in Merseyside and North Wales over this period.

When survival was examined in relation to place of booking, ELBW infants whose mothers booked at the Regional Unit, had the best chance of survival. Survival was related in referring districts to their referral policies. Some 19% of survivors followed up for between 1 and 5 years were impaired. Although the rate of impairment varied little between modes of referral, the pattern of impairment was markedly different. Cerebral palsy was commonest in infants transferred in utero, and severe retinopathy occurred only in postnatal transfers.

Differences between regions in referral patterns for ELBW infants influence the outcome in this high risk group.

**The maturation of renal function and water excretion in preterm babies. M Coulthard and E Hey (Newcastle upon Tyne).**

Ten healthy preterm babies (mean gestation 32 weeks) were studied during alternating periods of low (96 ml/kg/day) and high (200) total water intake over the first week of life. Glomerular filtration (GFR) and urine flow rates were measured daily by constant inulin infusion. From day three water and electrolyte balance was maintained appropriately, regardless of intake. The higher water intake did not affect GFR or sodium excretion, but the proportion of filtrate that was excreted rose from 7-4% to 13-1% (P<0.001) (both high compared to adult values).

We measured GFR in 39 babies of various gestational and postnatal ages, and reviewed the data from 14 published studies. GFR increased logarithmically with postconceptional age, and did not mature suddenly after birth. The impression that it does so has arisen because GFR has been expressed per unit body size, and early postnatal growth is usually slower than in utero growth.

These data indicate that from the third day of life healthy preterm babies can cope with fluid intakes of 200 ml/kg/day and that the excretion of drugs such as gentamicin will not depend on postnatal age. They do not lend support to current practices of gradually increasing fluid intake and gentamicin dosage with increasing postnatal age.

**Elevated levels of immunoreactive prostacyclin metabolite in babies who develop intraventricular haemorrhage. J M Rennie, J Doyle, and R W I Cooke (Liverpool).**

Prostacyclin is an extremely potent vasodilator and disaggregates platelets. High levels of the stable metabolite of prostacyclin, 6-keto prostaglandin F1 alpha (6-keto PGF1 alpha) have been demonstrated in infants with respiratory distress syndrome (RDS). Measurement of 6-keto PGF1 alpha was made in a group of 35 low birthweight infants (LBWI) at risk of intraventricular haemorrhage (IVH) who were not receiving any drugs known to interfere with the prostaglandin pathway.

The groups were well matched for birthweight, gestational age, and mode of delivery. There were more infants with acidosis and hypercarbia in the IVH group, confirming the previously noted associations of these factors with IVH.

The babies who developed IVH had significantly higher peak levels of 6-keto PGF1 alpha than those who did not, and failed to show the falling levels seen over the first three days of life in the non-IVH group. It is suggested that high levels of prostacyclin in babies with RDS may be a factor which contributes to the alteration of cerebral blood flow and capillary bleeding time with subsequent development of IVH.

With increasing pressures to economise on health care costs, it is important to be aware of the actual expenditure incurred in neonatal intensive care and the factors which influence it.

An economic study of the costs of neonatal intensive care in Mersey Region NICU in 1983 was made. Overall annual expenditure exceeded £1 million. Nursing costs (36%), laboratory tests (17%), overheads (16%), and drugs and disposables (18%), formed the greatest part of costs. Medical staff (8%), and capital equipment costs (5%) were relatively low.

Costs per day were for ventilator care £250, special/intensive care £104, and nursery care £63 (exclusive of laboratory tests). Very low birthweight infants formed 32% of admissions but used 63% of inpatient days and 71% of ventilator days.

Small changes in policies such as nurse/patient ratios or the proportion of small infants admitted may have major cost implications.


Intraventricular haemorrhage (IVH) is the most common neurological disorder in preterm infants and has been associated with defects in haemostasis. We have conducted a randomised prospective controlled trial using fresh frozen plasma (FFP) to investigate its effectiveness in preventing IVH.

Between December 1983 and August 1984, 85 neonates ≤1500 g and/or ≤32 weeks' gestation were admitted to our unit. Altogether 73 fulfilled the study criteria. The treatment group received 10 ml/kg of FFP on admission and at age 24 hours. The control and treatment groups were well matched for birthweight, gestation, sex, and five minute apgar scores, and other perinatal factors that have previously been associated with IVH. Fifteen out of 37 (41%) control infants sustained IVH compared with five of 36 (14%) of the treatment group (x² 6.52 P<0.02).

We were unable to show any significant effect of FFP on the clotting studies performed at age 48 hours. FFP appears to have a beneficial effect in preventing IVH though its action may not necessarily be mediated through the coagulation cascade.

Atypical mycobacterial lymphadenitis in childhood. M P White, H Bangash, K M Goel, and P A Jenkins (Glasgow and Cardiff).

The majority of cases of mycobacterial lymphadenitis in children are caused by atypical mycobacteria (AMB). Despite this there is little consensus of opinion on management.

We reviewed 19 children with AMB lymphadenitis to define likely presenting features, helpful diagnostic measurements, and optimum management.

There were 12 boys and seven girls. Mean age at diagnosis 5-2 years. The majority had no systemic upset and clear chest x rays.

Cervical nodes were the commonest affected and enlargement was usually unilateral. Mean duration of swelling at diagnosis was 6-6 weeks. Some 63% of nodes had an appearance suggestive of cold abscess.

Routine haematology was unhelpful, and standard tuberculin testing, performed in 47%, was negative in two thirds. Two cases had differential Mantoux testing with human PPD and Avian-intracellular antigen. This indicated AMB infection in both. This was the sole proof of AMB infection in one case, the remaining 18 having bacteriological proof usually M avium-intracellular.

Antituberculous drugs given to 78-9% for a mean of 7 months appeared ineffective. The organism was usually highly resistant.

Altogether 94-7% had surgical intervention. The treatment of choice for AMB lymphadenitis is total excision. Antituberculous drugs are unnecessary.

Insulin dependent diabetes mellitus diagnosed under the age of 5 years. I G Jefferson, M A Smith, and J D Baum (Oxford).

The incidence and clinical characteristics of insulin dependent diabetes mellitus diagnosed under the age of 5 years (DUAFY) (n=62), are compared with those of children diagnosed aged 5 to 10 years (n=75), within the Oxford district over the past 15 years. Mean (SD) yearly, age specific incidence per 100 000 for DUAFY children was as follows:1969-73, 8.9 (2.1); 1974-78, 7.4 (2.2); 1979-83, 13.6 (3.7); and for children diagnosed aged 5–10 years it was: 1969–73, 11.0 (6.4); 1974–78, 16.5 (4.3); 1979–83, 13.8 (3.8). The two DUAFY children presenting aged <1 year were both diagnosed in the last five years. DUAFY incidence shows a winter/spring peak in contrast to the autumn/winter peak seen in those diagnosed aged 5–10 years. A positive, first degree family history for IDDM was present in 16% of DUAFY children compared with 10% of the 5–10 year group; in none of these cases was the first degree relative the mother. In DUAFY children at
presentation duration of symptoms was less than 2 weeks in 30% compared with 12% for the 5–10 year group (P<0.05). In DUAFY children 27% presented in ketoacidosis compared with 15% for the 5–10 year group (NS).

DUAFY is not rare and may show an increasing incidence. It seems to show some clinical and epidemiological features which are different from diabetes in older children.

The steroid responsive nephrotic syndrome: a generalised disorder of membrane negative charge.
M Levin, C Smith, S Walters, P Gascoine, and T M Barratt (London).

The normal glomerulus functions both as a size and as a charge selective filter, restricting the passage of negatively charged macromolecules to a greater extent than neutral or positively charged molecules of similar size. The charge selective function is due to the presence of fixed anionic sites composed of sialic acid and glycosaminoglycans on the glomerular capillary wall (CGW). Similar charged groups are also present on other cell membranes such as red blood cells and platelets. In the steroid responsive nephrotic syndrome (SRNS) there is a reduction in anionic sites on the GCW and a selective increase in glomerular permeability to negatively charged proteins such as albumin, suggesting a basic defect in the charge-selective filtration barrier.

We have developed an assay for quantifying cell membrane negative charge based on the binding of the cationic dye Alcian blue. We have found a significant reduction in surface charge of erythrocytes and platelets from children with SRNS compared with controls. However, the sialic acid content of the red cell membrane, measured chemically, was normal. These results suggest that there is a generalised reduction of cell membrane negative charge in SRNS, which may be responsible for the increased glomerular permeability to anionic proteins. This reduction in cell membrane charge is probably due to neutralisation of charged groups by a cationic substance, rather than a deficiency of anionic groups.

The use of ovarian ultrasound to assess gonadotrophin secretion in disorders of sexual maturation.

We have performed pelvic ultrasound examinations in 40 normal girls aged 6 months to 14 years. From the age of 8 years, a multicystic ovarian appearance on ultrasound (more than six follicles in each ovary of 4 mm or greater in diameter) was seen. We have used the pulsatile administration of gonadotrophin releasing hormone to induce puberty in 12 girls with hypogonadotrophic hypogonadism and have followed the changes in gonadotrophin pulsatility and pelvic ultrasound during treatment. We have been able to mimic the clinical, endocrine, and ultrasound changes of normal puberty. The multicystic appearance seen in normal children is the ovarian morphological response to gonadotrophin pulsatility. Abnormalities of gonadotrophin pulse amplitude or frequency (both high and low) may impair pubertal development, which is why the gonadotrophin response to an acute injection of gonadotrophin releasing hormone is of limited diagnostic use.

An identical multicystic ultrasound appearance was seen in 15 girls with central precocious puberty, and was easily distinguishable from the appearances seen in eight patients with isolated premature thelarche.

Pelvic ultrasound is a non-invasive, in vivo method which has enabled us to assess gonadotrophin secretion without the requirement for invasive and complex endocrine investigation.

Effectiveness of bag and mask resuscitation at birth.
D Field, A D Milner, and I E Hopkin (Nottingham).

We have assessed the effectiveness of three of the most widely used bag and mask systems for neonatal resuscitation at birth. These were the Laerdal neonatal, the Laerdal paediatric, and the Ambu. All were used with an Ambu circular mask which we have previously found to be the most efficient. A pressure transducer inside the face mask allowed inflation pressure to be measured and a pneumotachograph fixed between the resuscitation bag and the face mask allowed us to measure expiratory flow (ie, that returned passively from the baby). Each bag was used to resuscitate 10 term babies born by elective caesarean section who required supplemental respiratory support. On all occasions, the resuscitation was carried out by the same person.

Results of mean tidal volume achieved by each system for the first three inflations are as follows: Laerdal neonatal 5.47 ml; Laerdal paediatric 10.4 ml; Ambu 15.4 ml.

It would appear that both the latter two bags are producing effective ventilation. However, if a tidal volume of three times dead space is used as a measure of adequate ventilation, then this occurred on no occasion with the Laerdal neonatal bag and on three occasions only for each of the other two bags.

Our results indicate that current bag and mask systems are not satisfactory for routine neonatal resuscitation.