British Paediatric Surveillance Unit

Sixth Annual Report 1991

Supported by Children Nationwide Medical Research Fund

BRITISH PAEDIATRIC SURVEILLANCE UNIT

5 St Andrew's Place Regents Park London NW1 4LB Tel:071-935 1866 Fax:071-486 6009

Sixth ANNUAL REPORT 1991

CONTENTS

Additional copies are available from the BPSU office, to whom any enquiries should be addressed.

BRITISH PAEDIATRIC SURVEILLANCE UNIT

Sixth ANNUAL REPORT 1991

1 FOREWORD

The BPSU is entering its sixth year and is still going from strength to strength. The scientific value of the unit is readily apparent from the rising number of publications by BPSU investigators which have appeared in peer review journals and from formal and informal references to the unit and its work in, for example, the medical and lay press and government documents. Two papers in which the BPSU was central to the methodology were presented at the BPA Annual Scientific Meeting in 1992: these studies reported on AIDS and HIV (plenary session) and Rett Syndrome (neurology group session).

The unit also plays an important role in providing information relevant to the formulation and monitoring of public health policy - two examples are the surveillance of Reye Syndrome after the warning about the use of aspirin and the surveillance of acute flaccid paralysis as a step towards eradication of wild poliomyelitis from these islands.

In 1991 the BPSU increased the size of its respondent database by improving its ascertainment of hospital and community consultant paediatricians in academic and service appointments in the UK and Ireland. Plans are also in hand in 1992 to recruit as respondents, (for those conditions relevant) non-BPA members of specialty groups who treat children. This will not only enhance case ascertainment but will also draw the attention of other specialties to the methodology of the BPSU.

Another exciting development is the increasing interest of other countries in the BPSU to the extent that some are planning to, establish their own units. Discussions have been held with a number of European countries, resulting in the formation of a Working Party to establish a European Network of National Paediatric Surveillance Units. Further afield, a steering committee under the auspice of the Australian College of Paediatrics has recently been convened in order to develop an APSU.

British and Irish paediatricians can therefore feel justly proud of themselves as pioneers and key enactors of this unique reporting system.

The success of the unit is a corporate one: it is entirely dependent on the collaboration of its respondents. I should like to thank members of the BPA and of the Faculty of Paediatrics of the Royal College of Physicians of Ireland who continue to participate in the reporting scheme by regularly returning their orange cards and by providing detailed information on reported cases.

I should like to thank the staff of the Unit and members of the BPSU Executive Committee (BEC) chaired by Professor E Ross, who have met regularly to supervise the day-to-day running of the Unit. Finally I should like to express my thanks to the officers and staff of the BPA for their continued support and to those bodies listed later in this report who have continued to give much needed financial support.

Sir Cyril Clarke
Chairman

MAY 1992

2 INTRODUCTION

The BPSU began operations in June 1986 as a national unit covering the UK and Ireland. The Unit enables paediatricians to participate in the surveillance of infections and infection-related conditions, to promote the study of uncommon disorders, and to provide a mechanism by which new diseases can be detected quickly so that early investigation can take place.

The BPSU is a collaboration of several agencies: the British Paediatric Association (BPA), the Public Health Laboratory Service (PHLS), the Communicable Disease Surveillance Centre (CDSC) and the Department of Epidemiology at the Institute of Child Health (ICH), University of London. The Unit also collaborates closely with the Communicable Diseases (Scotland) Unit in Glasgow which administers the scheme in Scotland.

The reporting system involves the mailing of a monthly card which contains the disorders currently being surveyed and a set of reporting instructions. This card is distributed to consultant paediatricians who are members of both the BPA and/or the Faculty of Paediatrics of the Royal College of Physicians of Ireland. Respondents are asked to return the card to the BPSU office each month, reporting the number of cases seen of the relevant disorder in the preceding month. Most importantly, respondents are requested to still return the card even if no case has been seen (compliance with the scheme can thus be monitored). Following a case report, the BPSU office informs the appropriate researcher who contacts the reporting paediatrician for further information in accordance with the study protocol for that condition. The research worker then reports back to the BPSU on the outcome of the follow-up, confirming cases, identifying duplicates and reporting errors.

A study is eligible for participation in the scheme if the condition of interest is a rare childhood disorder (or a rare complication of a more common disorder) of such a low incidence or prevalence as to require case ascertainment on a national scale in order to generate sufficient numbers for the study. The Executive Committee of the BPSU meets approximately once a month to review and approve applications, advise applicants on study and questionnaire design and to oversee the day-to-day running of the unit.

3 CONDITIONS INCLUDED (see also TABLES on pages 19-20)

3.1 AIDS and HIV

Paediatric AIDS has been included in the BPSU reporting system since June 1986. In January 1990, the reporting definition was extended to include all HIV seropositive children. Paediatricians were also asked to report retrospectively any cases they had seen prior to January 1990.

By January 1992, five hundred and fourteen cases had been reported through the BPSU; 377 had been confirmed, 75 were duplicate reports, and 40 were reporting errors; 22 reports awaited confirmation.

TABLE 1 - AIDS/HIV Surveillance (to end of 1991)

Source of infection	AIDS cases (Deaths)	Total HIV Seropositive
Blood Products Haemophilia Blood Transfusion	 18 (11) 6 (1)	 43 11
Child of HIV positive mother: Infected Indeterminate Unaffected	61 (29)	 116 82 125
Total	85 (41)	377

A further 121 children of HIV positive mothers, 195 haemophiliacs and 15 blood transfusion recipients who acquired HIV infection under the age of fifteen, have not yet been reported through the BPSU, but are known about through the UK Haemophilia Centre or from obstetric reports.

All children reported, are being followed-up annually so that their infection status can be determined and, if infected, whether they have developed AIDS.

A copy of 'Guidelines for Management of Children with HIV Infection' has been sent to all paediatricians who have reported a child born to an HIV positive mother.

Paediatricians who have reported children with confirmed HIV infection through the BPSU are being offered the opportunity to join a European trial of early compared with deferred use of Zidovudine (AZT) in vertically infected children with HIV infection. This is due to start in May 1992 and updates of its progress will also be sent out through the BPSU.

Dr C Davison, Miss F Holland - Department of Epidemiology and Biostatistics,
Institute of Child Health, 30 Guilford Street, London WC1N 1EH Tel: 071-829 8686
Dr A Nicoll - PHLS Communicable Disease Surveillance Centre, 61 Colindale Avenue,
London NW9 Tel: 071-200 6868

3.2 NEONATAL HERPES (HSV)

In the five and a half years between July 1986 and December 1991 there were 131 notifications of suspected neonatal HSV infection reported through the BPSU, and three from other sources. Seventy-one of these have been confirmed as cases.

A case was confirmed if:

(a) HSV was isolated, or detected by electronmicroscopy, in the first month, **or**
(b) specific IgM was present in the infant in the first month, **or**
(c) an infant had typical clinical manifestations and maternal infection was confirmed by either seroconversion or virus isolation around the time of delivery.

Of the remaining 63 notifications, 15 were duplicates, 33 were excluded after further investigation and eight could not be identified by the notifying paediatrician. The diagnosis could neither be confirmed nor excluded for the remaining seven who were treated prophylactically but did not satisfy (a), (b) or (c) above.

Less than 30% of the 71 mothers of infants with confirmed infection had a history of genital herpes, or evidence of a recurrent or primary infection in pregnancy; in most of these the maternal infection was only diagnosed retrospectively, after diagnosis in the infant.

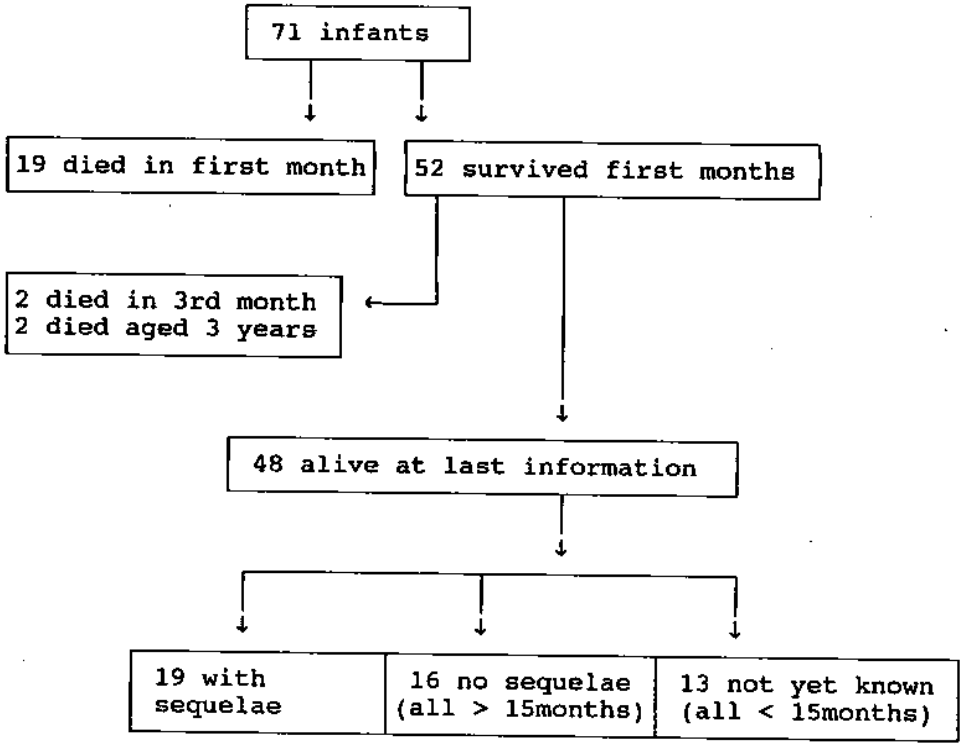
There were eleven cases where a possible postnatal source of infection was identified (five mothers with oral herpes, a whitlow or HSV eczema, three fathers and three other relatives with oral herpes); in five of the eleven infants HSV-1 was isolated, in the remaining six virus type was unknown. Among the 71 confirmed cases there was a high case fatality rate: nineteen (27%) infants died within a month of birth. Type 1 infections were common as type 2, and deaths occurred in the infants with type 1 infection (6/25) as frequently as in those with type 2 (6/24). Virus was not typed in 22 infants, seven of whom died.

Twenty-four infants (34%) presented with infection localised to the skin, eye or oral cavity, and eighteen (25%) with infection localised to the skin and CNS. Five infants (7%) presented with infection localised to the CNS alone, without any skin lesions. The remaining 24 infants (34%) had disseminated disease, including 10 with no skin lesions. Altogether 15 infants (21%) had no typical skin, oral or eye lesions; ten (66%) of these 15 infants died in the first month and in eight the diagnosis of neonatal HSV infection was only made at post-mortem. Among the 56 infants who did have skin, oral or eye lesions, nine (16%) died in the first month.

Survivors are being followed up in the second year of life, and subsequently. **(Figure 1, overleaf)**

Two children are known to have died in their third month, one was described as a 'cot death', the other, born at 29 weeks gestation, died of bronchopulmonary dysplasia. Two more severely handicapped children died before their fourth birthday. Almost all of the 19 survivors known to have sequelae have multiple handicap. Follow-up continues and the data will be analysed and reported at a later date.

FIGURE 1 - Outcome in 71 Infants with Neonatal HSV



Case ascertainment through the BPSU has now ceased. However, if any paediatrician knows of an infant with neonatal HSV (even if the infection is thought to have been acquired postnatally) who was born between 1986 and the end of 1991, please report the case to the address below. We are very grateful to all reporting paediatricians for their interest and support, and will keep them informed of the study findings.

Ms P A Tookey*, Professor C S Peckham*, Dr R Dinwiddie -
*Unit of Epidemiology and Biostatistics, Institute of Child Health, 30 Guilford Street, London WC1N 1EH Tel: 071-242 9789

3.3 REYE SYNDROME

Reye Syndrome (RS) surveillance in the British Isles has been running since August 1981. It began as a joint BPA-CDSC venture, with "passive" case ascertainment, which in 1986 transferred to the active system of the BPSU. In 1991 the surveillance questionnaire was modified to collect further information relating to the inherited metabolic disorders (IMD) which may present as a Reye-like illness.

Results 1990/91

A total 25 reports of Reye Syndrome were received in the surveillance year 1990/91 (1 August 1990 - 31 July 1991). Of these, 12 cases had their diagnosis subsequently revised to a specific alternative. **TABLE 2** (overleaf) shows annual total reports for previous years 1981/82 - 1990/91 with the breakdown of revised and non-revised cases. There was one patient in 1990/91 in whom there were atypical clinical features (see below), but no alternative diagnosis was reached; this indeterminate case has been included in the non-revised group in **TABLE 2.**

TABLE 2 - Reye Syndrome Surveillance

12 month period (August-July)	Total reports British Isles	Non-revised reports	Revised diagnosis (IMD)
1981/82	47	40	7 (3)
1982/83	69	59	10 (6)
1983/84	93	81	12 (3)
1984/85	64	56	8 (2)
1985/86	53	40	13 (4)
1986/87	47	26	21 (11)
1987/88	44	32	12 (3)
1988/89	31+	18	12 (6)
1989/90	24+	15	8 (5)
1990/91	25	13	12 (7)
TOTAL	497	380	115 (50)

+Detailed information not available for one case.

Non-revised cases

Among the 12 non-revised, non-indeterminate cases, there were five males and seven females. The ages ranged from 2.9 months - five years, with mean (SD) and median ages of 20.5 (22.8) months and 9.5 months respectively. Nine (75%) reports came from England with the remaining three cases residing in Scotland, Wales and Northern Ireland. The largest number of reports in any one month, three, occurred in January.

Four children survived apparently normal and five died, giving a case fatality rate of 42% (47% in 1989/90). Three cases survived with sequelae; hypertonia (one case), poor visual function and hypotonia (one case); one patient was reported to be unresponsive with persistent seizures and no voluntary activities.

Five patients (42%) were reported to have received pre-admission medication; paracetamol had been given in two cases; kaolin (one case); antibiotics and cough linctus (one case); one five year old child had been given "Askit" powder, which contains aspirin, one day prior to admission.

Information relating to past medical history was provided in 10 of the 12 children. In three cases, histories compatible with an underlying metabolic disorder were reported; one child had a tendency to vomit easily; another had had neonatal fits; one case had previous viral/vomiting episodes, during some of which aspirin had been taken for viral symptoms. Information on family history was provided in five cases; none included events compatible with an IMD (eg. unexplained deaths in infancy, encephalopathic illnesses).

Specific investigations for IMDs were undertaken in five of seven patients with information. All five had urine organic acids assayed; four patients had both plasma and urine amino acids measured but only in one child was urinary orotic acid investigated. In the remaining two cases no investigations were undertaken: one child died suddenly soon after admission and the other made a rapid and complete recovery within a few days.

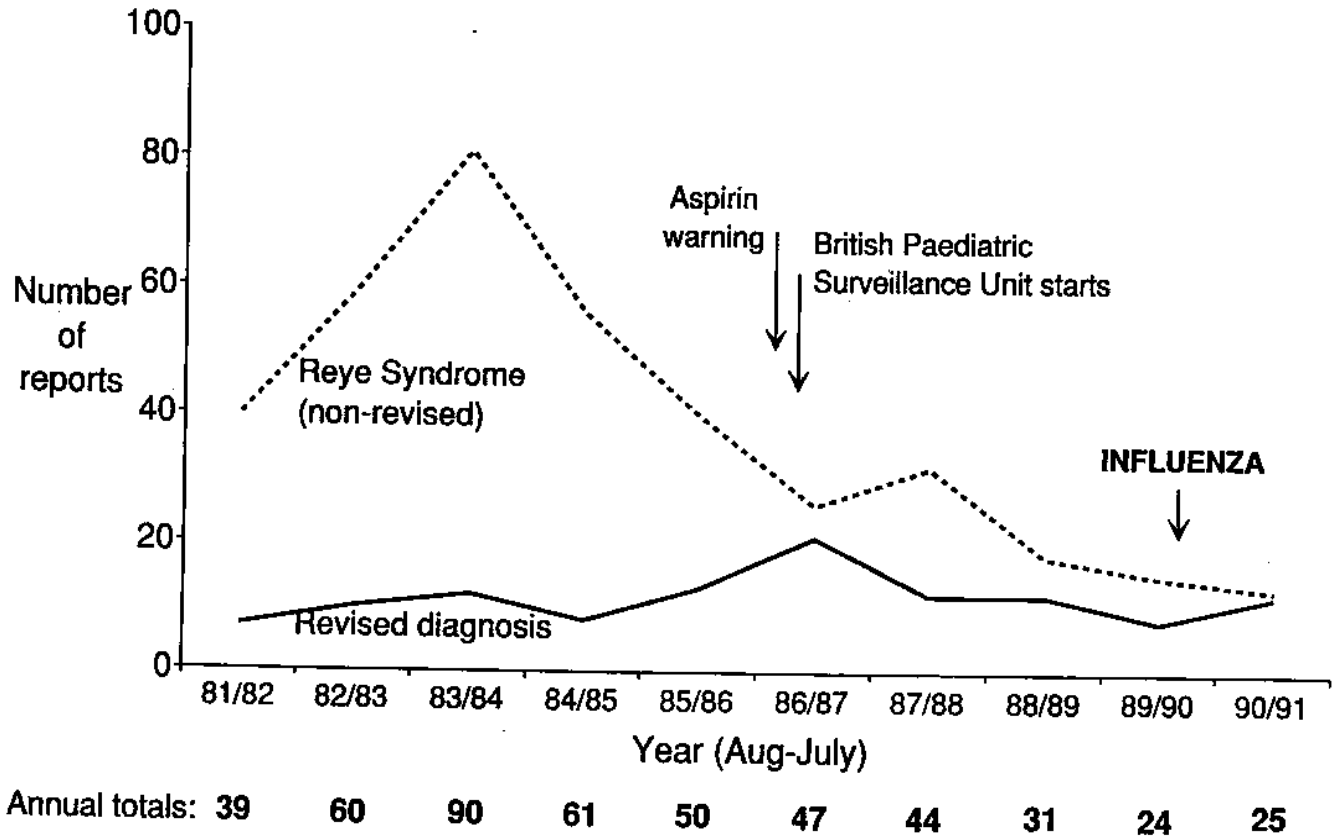
The patient with an indeterminate diagnosis was female and aged 11.9 years. Although this age is compatible with "classic" RS, a number of atypical clinical features were presented, including jaundice, an unremarkable ammonia level, grossly disordered clotting and sudden onset. Metabolic investigations were undertaken and an abnormal medium chain fatty acid result reported; however no definitive diagnosis was reached.

Revised diagnosis cases

Of the 12 patients with revised diagnoses, seven were reported to have inherited metabolic disorders (IMD). The diagnoses were as follows: Medium-chain acyl CoA dehydrogenase deficiency (MCAD) (three cases), Pyruvate dehydrogenase deficiency (one case), methylmalmonic acidaemia (one case), one unspecified defect of fatty acid oxidation and one unspecified inherited metabolic disorder. The mean (SD) and median ages of patients with IMD's were 12.4 (8.0) months and 12.5 months respectively. One five year old child with a flu-like prodrome had been treated with Anadin prior to admission.

The remaining five cases were non-metabolic revisions: Q fever, haemorrhagic shock encephalopathy syndrome, sudden infant death syndrome, pneumococcal septicaemia and viral infection with hepatic necrosis. The mean (SD) and median ages of this group were older than the IMD group, at 43.9 (54.3) months and 14 months.

FIGURE 2 - Reye Syndrome in the British Isles 1981-91



Annual totals: **39 60 90 61 50 47 44 31 24 25**

Comment

The reported incidence of RS continues to decline: the largest epidemic of influenza for 13 years in the winter of 1989/90 had no apparent effect. **(Figure 2)** The proportion of patients reported with RS, who subsequently are shown to have an IMD, has shown a steady increase from 5% of total reports in 1981/82 to 27% of total reports in 1990/91, reflecting increased awareness of these conditions among paediatricians. The positive responses to the question about investigations for these conditions provides supportive evidence for this. **However it is still likely that the patients whose diagnoses were not revised, had one of these disorders especially as the median age was so young.**

It is of concern that two patients reported with RS in 1990/91 had pre-admission exposure to aspirin; none were reported with aspirin ingestion in 1989/90. There may be a need for a renewed campaign to raise parental awareness of the dangers of aspirin given to children with feverish illnesses.

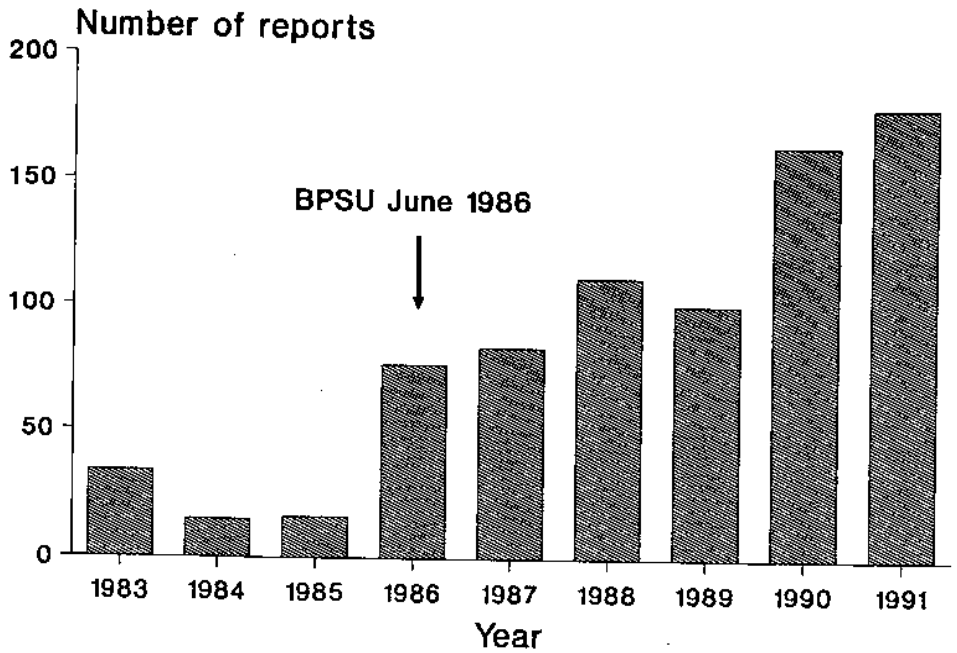
Dr S Hall, Ms L Newton - PHLS Communicable Disease Surveillance Centre,
61 Colindale Avenue, London NW9 5EQ Tel: 081-200 6868

3.4 KAWASAKI DISEASE

Kawasaki Disease (KD) surveillance began in 1983 as a joint BPA-CDSC venture and since 1986 ascertainment has been via the BPSU "active" reporting scheme.

Annual total reports of KD have been steadily increasing. **(Figure 3)** Totals for years 1983-90 were 34, 15, 16, 76, 83, 112, 100, 163. Two hundred and one reports were received in 1991, once again an increase on the previous years' total. Of these, 175 were confirmed as KD, six reports were later revised and, at the time of writing, further information had yet to be received for 20 cases.

FIGURE 3 - Kawasaki Disease Annual Total Reports 1983-91



One hundred and forty-one cases (80%) came from England with the greatest regional number of reports coming from the North East Thames region (19). Reports were also received from the following regions: Wales (five), Scotland (ten), Northern Ireland (eleven) and Irish Republic (seven). January, with 25 cases, was the peak month of onset.

As in previous years there was a male excess (M:F ratio 1.6:1). Ages ranged from two weeks to 13.5 years with mean (SD) and median ages of 37.7 (33.2) months and 30.6 months respectively. Eighty per cent of children were under five years of age and 25% were under 12 months.

Information on ethnic group was provided in 170 cases. Again as in previous years white caucasians were under represented with 135 (79%) cases, but there was a slight increase in the proportion of mixed-race patients, eight cases, (5%). The remaining cases were: Afro/Caribbean - eight (5%); Indian sub-continent - eleven (6%); Oriental - eight (5%).

154 (88%) children had "typical" KD with five or all six criteria of the case definition and 26 were "atypical" (three or four criteria).

127 (73%) children recovered fully from their illness and one child died, giving a case fatality rate of 0.6% which is substantially lower than the 3.7% seen in 1990. Thirty-three children recovered with sequelae; twenty-six had persisting cardiac involvement and the remainder suffered non-cardiac sequelae of which the most common was arthralgia/arthritis. Outcome for 13 children was unclear at the time of proforma completion.

Cardiac complications were reported in 62 (37%) children. Of these, 43 had coronary artery abnormalities including coronary artery aneurysms in 22 cases and dilation in 21 cases. Other complications were pericardial effusions (10 cases) and congestive cardiac failure (two cases). Two-thirds of patients with cardiac complications were male.

A variety of non cardiac complications were reported. The most common were as follows: athralgia/arthritis (14 cases); involvement of the liver (21 cases, including seven cases of jaundice and two cases of hepatitis; anaemia (six cases); respiratory tract complications (three cases) and diarrhoea (seven cases). A more unusual complication, gangrene of the fingers and toes, was reported in two children. The extreme irritability and misery often seen in KD was reported in 11 children.

1991 saw a further increase in the annual number of KD reports, however, this was accompanied by a marked decrease in the case fatality rate. The reason for this decrease is still unclear and may be multifactorial, perhaps a reporting phenomenon or perhaps in part related to increasing use of intravenous immunoglobulin. However the continuing high rate of reported coronary artery involvement mitigates against this explanation.

The one year follow-up study of 1990 cases, providing further information on the patient's clinical status and management following discharge, is now almost complete. To date 142 (87%) proformas have been returned and we are most grateful to colleagues for their kind collaboration with this and the main surveillance scheme.

Dr S Hall, Ms L Newton - PHLS Communicable Disease Surveillance Centre,
61 Colindale Avenue, London NW9 5EQ Tel: 081-200 6868

3.5 CONGENITAL RUBELLA

Congenital rubella was included in the BPSU reporting scheme in January 1990, since when 33 cases have been reported. Of these, 19 were confirmed cases, five were duplicates, two were not confirmed, one was a reporting error and six are still being followed up. Two of the confirmed cases have not been included in the figures of the National Congenital Rubella Surveillance Programme because the children were born outside the UK.

TABLE 3 - Cases of Congenital Rubella by Year and Source by Notification

Year of birth	1987	1988	1989	1990	1991	Total
BPSU	1	4	2	9	1	17
Other sources	39	18	10	2	1	70
Total	40	22	12	11	2	87

Paediatricians are asked to continue to report all suspected cases of congenital rubella infection whether or not associated with typical rubella defects. We are most grateful for their kind participation.

Ms Gill Jones, Ms Pat Tookey, Professor C S Peckham - Unit of Paediatric Epidemiology and Biostatistics, Institute of Child Health, 30 Guilford Street, London WC1N 1EH
Tel: 071-242 9789 ext 2606

3.6 SUBACUTE SCLEROSING PANENCEPHALITIS

Since the last summary in 1990, twenty-one cases have been reported to the BPSU. Seven of these reports were from Eire or Northern Ireland and follow-up information is not available. The remaining 14 reports were for cases in England and Wales, of which seven were confirmed and added to the national register and one was a notification of the death of a known case. Three were duplicates and three are still being followed up.

In four of the seven confirmed cases, the BPSU was the only source of notification. The other three were also reported by microbiologists. The BPSU continues to be the most reliable source of notification of cases seen by paediatricians.

Dr N Begg - PHLS CDSC, 61 Colindale Avenue, London NW9 5EQ Tel: 081-200 6868

3.7 GALACTOSAEMIA

Reporting of new cases of classical galactosaemia occurred from January 1988 to September 1991. Data were analysed for the completed three-year period 1988-1990.

The results have shown that classical galactosaemia is more common than expected, with an incidence of 1:44,000 for the UK (1:23,500 for Eire). Screening by blood spot assay for Gal-1-PUT was already occurring in Scotland and Eire (14 cases) but not specifically in other parts of the UK (46 cases).

TABLE 4 - Galactosaemia Reports 1988-90

YEAR	Total Cases	England/Wales/ N Ireland*	Scotland+	Republic of Ireland
1988	25	17	2	6
1989	19	16	3	0
1990	16	13	2	1

* = 46 cases unscreened. + = 14 cases screened

We conclude that the introduction of new screening programmes across the UK is not justified for the following reasons:

1 **Screening does not prevent acute illness.**
Encephalopathy, coagulopathy, septicaemia of jaundice requiring exchange transfusion occurred in 4/14 (30%) of the screened group and in 9/46 (20%) of the unscreened group.

2 **No excess of previous sibling deaths in the unscreened group was found.**
One death from galactosaemia occurred in the eldest of five children in the unscreened group. However, two deaths consistent with galactosaemia occurred in older siblings in the screened group at a time when screening was taking place (in the 1991 data, two further sibling deaths occurred in non-caucasian children born outside the UK).

3 **Screening at 6-10 days does not prevent early death.** One case from the unscreened group died at four days from coliform septicaemia. Effective screening would need to occur earlier than currently provided by existing neonatal screening programmes, with major cost implications.

4 **The age at starting diet is not unduly delayed by clinical diagnosis.**
Excluding those with a positive family history and the one death, 35/39 cases (90%) were diagnosed clinically by one month, without screening. (75% by three weeks and 66% by two weeks) there were only two diagnoses after two months in this unscreened group where the additional delay might further compromise neurodevelopmental outcome.

5 **False negatives occur from screening.**
One case developed acute and serious illness and delay in starting treatment in spite of screening.

6 **The existing programme for PKU screening already contributes to the diagnosis by the finding of increased phenylalanine and tryosine.**
This occurred in 9/60 (15%) otherwise unscreened cases over the total reporting period.

The reporting period for galactosaemia was extended into 1991 to ensure late diagnoses for the three-year study period were not missed and to consider a further study because of the increasing concerns over the long term implications of this disorder. A further 16 cases were detected over nine months, ie. England/Wales/N.Ireland, fourteen cases; Scotland, one case; Republic of Ireland, one case. A protocol for a national cohort study based on cases detected by this survey has been submitted for funding.

Mrs A Green, Dr J Holton, Dr M Honeyman*, Professor J Leonard -
*The Child and Family Centre, 142 Maas Road, Northfield, Birmingham B31 2PR
Tel: (021) 4766969

3.8 MMR MENINGOENCEPHALITIS (MMR-M)

Active surveillance of mumps vaccine-associated meningoencephalitis was initiated in 1990 through the BPSU. Between 1 February 1990 and 31 January 1992 all consultant paediatricians in the British Isles were asked each month to report to the BPSU, cases of meningoencephalitis in children under 16 years of age occurring within six weeks of receiving the MMR vaccine. Where a case was identified from a source other than the BPSU (ie. yellow card to the Committee on Safety of Medicine or from a laboratory report), the paediatrician was contacted and asked to report the case to the BPSU. Paediatricians were encouraged to obtain CSF samples from suspected cases and to ensure that any mumps viruses isolated were sent to the National Institute for Biological Standards and Control (NIBSC) for characterisation as wild or vaccine-like.

On receipt of a BPSU report, clinical, laboratory and vaccination details were sought from the reporting paediatrician by means of a standard questionnaire. Further details eg. vaccine batch number were obtained from the patient's GP if necessary.

Twelve months after an initial case report, a follow-up examination was conducted in the patient's home. This included a developmental assessment, hearing test and general neurological examination.

Classification of cases: Cases have been classified according to the following criteria:

Definite: Vaccine-like mumps virus isolated from CSF

Probable: No virus isolated, onset 11-28 days after vaccination

Not a case: No virus isolated, onset < 11 or > 28 days after vaccination

Results

Eighty-three reports were received between 1 February 1990 and 31 January 1992, including duplicates. Sufficient information is available to classify 72 of these reported cases, of which 15 were definite, 34 were probable and 23 were not cases. Age/sex details are known for 69 cases. Forty-six (66%) were children aged 12-23 months; only five cases (7%) were children aged five years or older. There was an excess in males (M:F ratio 1.35 to 1). **TABLE 5**

TABLE 5 - Age/Sex Distribution of BPSU Reports for MMR-M (1 Feb 1990 - 31 March 1992)

AGE	MALE	FEMALE	TOTAL
1 - < 2	28	18	46
2 - < 3	4	10	5
3 - < 4	2	0	2
4 - < 5	6	5	11
5 and over	2	3	5
TOTAL	42	27	69

NB: Age/sex not known for four cases.

Cases were reported from all four countries in the UK (including every English Region) and Republic of Ireland. **(TABLE 6)** Nine of the 15 culture-positive cases were reported in two Regions ie. Trent (six cases, of which four were in Nottingham) and Wessex (three cases, all in Poole).

To date 53 of the reported cases have been followed up, including 14 of the 15 culture-positive cases. Three are definitely abnormal: two (one culture-positive) were previously abnormal with no deterioration following vaccination and one has developed an unrelated condition (astrocytoma). A further two are possibly abnormal, although one of these (who was culture-positive) has behaviour problems and is difficult to assess.

TABLE 6 - Geographical Distribution of BPSU Reports for MMR-M (1 Feb 1990 - 31 Jan 1992)

	No. OF CASES	
REGION / BOARD	TOTAL*	DEFINITE
ENGLAND		
Northern	4	0
Yorkshire	5	0
Trent	8	6
East Anglia	7	0
NW Thames	6	1
NE Thames	3	0
SE Thames	3	0
SW Thames	4	0
Wessex	7	3
Oxford	7	1
South Western	3	0
West Midlands	4	0
Mersey	3	0
North Western	4	2
WALES	2	0
SCOTLAND	2	0
NORTHERN IRELAND	3	0
REPUBLIC OF IRELAND	8	2

* includes duplicate reports

Conclusion:

The BPSU surveillance indicates that vaccine-associated mumps meningitis is a rare, and usually mild complication of MMR vaccine. While it is possible that some mild cases may not have been reported, the benefits of MMR vaccine (which has virtually eliminated natural mumps and its complications in the UK), greatly outweigh its risks.

Dr N Begg - PHLS Communicable Disease Surveillance Centre, 61 Colindale Avenue, London NW9 5EQ Tel: 081 200-6868
Dr A MacFarlane - Community Health Offices, Radcliffe Infirmary, Oxford OX2 6HE - Tel: 0865 224637

3.9 CHEMISTRY SET POISONING

To April 1992, eighteen cases have been reported; three were rejected as being outside the case definition, because in two cases the incidents occurred in school laboratories and in one case it was very doubtful whether any chemical had actually been ingested; one report was a duplicate, leaving 14 confirmed cases. The distribution of the latter were Wales (three cases), West Midlands (four cases), Yorkshire (three cases), Wessex (two cases), Northern Ireland (one case), South-East London (one case). The age at which the poisoning took place was: <2 years, (two cases), 2-3 years (four cases), 3-5 years (three cases), 5-10 years (two cases), 10-16 years (three cases, one of whom died). Nine were male and five were female. Incidents occurred in January (three cases), February (one case), March (two cases), April (three cases), August (one case), September (two case), October (one case) and November (one case). This distribution suggests that these incidents tend to occur during the colder months and that these sets tend to be given as Christmas presents - six cases (43%) were between Jan-March, and nine case between Jan-April (64%).

Substances ingested include: Copper Sulphate (five cases); Mixtures (four cases, containing Copper Sulphate, two cases and Cobalt Chloride two cases); Cobalt Chloride (one case); Tartaric acid (one case); Ammonium sulphide (one case) and Methylene blue (two cases). The source of the chemical ingested was: chemistry set, (eight cases), crystal growing set (three cases), microscope set (two cases), and stink bomb (one case). In six cases this was a solution; in three cases, a mixture of crystals and powder; in one case, crystals and four cases were not known. Packaging of the chemicals was: cellophane packet three cases (all in crystal growing sets), one case each of glass bottle, glass tube, test tube, sealed glass vial, sealed container, plastic jar, and in six cases packaging was unknown.

Risk factors were assessed as follows: nine children were considered to lack supervision ie. two took mixtures left by a brother and a visitor respectively; one took a solution belonging to a boy from another family in a hostel for the homeless and had previously been admitted for neglect and emotional abuse; three cases were due to poor supervision, one due to poor general social care and two bit the glass container. Of the remainder, one case was due to a genuine accident in which the solution was mistaken for a bedside drink in the dark, one case was due to sibling rivalry, one case was a deliberate cry for help (pregnant and in care), one case was a boy with a reactive depression who was being bullied at school and had left a suicide note and in one case no risk factor was identified.

The outcome was full recovery in 13 cases and one died; the latter was an 11 year old girl who was growing copper sulphate crystals from a saturated solution at the bedside and in the dark mistook this for her usual Ribena drink: she drank only one mouthful, but, despite immediate removal to hospital, died three hours later from irreversible cardiac arrest.

These figures show that chemistry set poisoning is a definite problem, but not a large one, with these sets containing toxic chemicals, which are freely available to the public, sometimes getting into inappropriate hands, and whose use is often unsupervised; this situation needs to be addressed by legislators and the appropriate regulatory authorities if further deaths and/or morbidity are to be prevented - even one death is one too many!

With this in mind, I would make the following recommendations:

(a) Child resistant containers should be mandatory.
(b) There must be proper toxic hazard warnings on the box and container, and in the instructions supplied with the set.
(c) The lettering of these warnings should be larger than at present.
(d) These warnings should include the phrases "keep away from very young children" and "use under ADULT supervision".
(e) The parents of the most vulnerable groups (infants in the second six months, toddlers and nursery school children) should be targeted in Health Education programmes by talks and posters in mother and child groups, schools, hospitals and clinics, and by Press and TV advertising.
(f) Restriction of the chemicals that may be included in these sets, eg. as in Australia, where asbestos, ammonium nitrate and lithium hydroxide are specifically prohibited (Ogilvie 1992, personal communications).

Dr E Mucklow, St Mary's Hospital, Newport, Isle of Wight PO30 5TG Tel: 0983 524081

3.10 ACUTE FLACCID PARALYSIS

Surveillance of this condition began in August 1991. The primary purpose of the survey is to detect cases of paralytic poliomyelitis as part of the World Health Organisation's programme for global elimination of polio by the year 2000. All cases of acute flaccid paralysis should, however, be reported, not just those due to suspected polio.

Cases are reported to the investigator by telephone in the first instance, in order that timely laboratory investigations can be initiated. The single most important investigation is viral culture of faeces. At least two samples, taken 24-48 hours apart and within a week of onset of paralysis, should be taken.

Fifty-five cases were reported up to 8 April 1992. Based on the first eight months of the survey, the annual incidence rate for acute flaccid paralysis in the British Isles is thus approximately six per million population under 16 years of age.

A final diagnosis has been provided for 21 of the cases to date. One case of possible vaccine-related polio has been detected, consistent with previous estimates of the incidence of this condition. No cases due to wild polio have been detected. The diagnosis for the remaining 20 cases are Guillain Barre syndrome (12 cases of which four followed a *Mycoplasma pneumoniae* infection and one followed glandular fever), postinfectious encephalomyelitis (two cases), acute transverse myelitis, chronic relapsing radiculopathy, ascending polyneuritis, hypokalemic paralysis, hysterical paralysis and Chinese paralytic syndrome (one case each).

It is intended to maintain surveillance for three years. If no cases of wild polio are reported during this time, one of the World Health Organisation's criteria for eradication will have been met. The survey could serve as a model for other countries that will require polio eradication certificates. It should also provide the most reliable estimate to date of the incidence and aetiology of acute flaccid paralysis in the British Isles.

Dr N Begg - PHLS Communicable Disease Surveillance Centre, 61 Colindale Avenue, London NW9 5EQ Tel: 081-200 6868

3.11 ANDROGEN INSENSITIVITY SYNDROME (AIS)

Surveillance of the androgen insensitivity syndrome (AIS) started in September 1991. Complete AIS, which is characterised by normal female external genitalia in an individual with testes and a 46XY karyotype, is straightforward for reporting purposes. Bilateral inguinal herniae are a common presenting feature and there are several reports of an older sibling becoming recognised as having complete AIS. There are also infants with abnormal external genitalia and 46XY karyotype in whom the clinical phenotype is compatible with partial AIS (micropenis/clitoromegaly, bifid scrotum, perineoscrotal hypospadias).

The survey has already highlighted diagnostic problems hampered by lack of adequate information on testicular steroidogenesis and metabolism, and lack of histology. A small number of infants with the phenotype of partial AIS have been shown to have other disorders such as true hermaphroditism, testosterone biosynthetic defects and testicular dysgenesis.

The preliminary data are not yet sufficient to provide information on the diagnosed frequency of complete and partial AIS. The survey will provide useful information to help sort out the diagnostic conundrum surrounding the clinical phenotype of partial AIS.

Professor I A Hughes - University Cambridge School of Clinical Medicine, Addenbrooke's Hospital Cambridge CB2 2QQ Tel: 0222 744035

3.12 HAEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS (HLH)

Haemophagocytic lymphohistiocytosis (HLH) is a new name for a rare, multi-system disorder of antigen-processing histiocytes first recognised in the 1950s by Drs Farquhar and Claireaux. There is an autosomal recessive familial form, usually presenting in infancy, and a sporadic form often precipitated by an infection, that can present at any age from infancy to adult life. In the 1970s and 1980s, the two disorders were distinguished by being called:

(a) familial erythrophagocytic lymphohistiocytosis (FEL) and
(b) virus-associated haemophagocytic syndrome, respectively.

These names are now considered misleading because:

(a) it is difficult to decide whether the first affected infant in a family has the sporadic form, or is the first to express a new autosomal mutation
(b) because the familial disease can be triggered by infection, and
(c) because organisms other than viruses have been implicated.

For these reasons, the term "HLH"* has been preferred by the Histiocyte Society (an International Group of clinicians and scientists focusing on Histiocytosis Research).

There is no diagnostic test for HLH but the syndrome is fairly easily recognised, so long as clinicians are aware of the condition. The principal features are: hepatosplenomegaly, fever, non-focal CNS signs (irritability, fits) and bleeding; lymphadenopathy and skin rash are much less common and bone involvement very rare indeed, making it relatively easy to distinguish between HLH and Langerhans' cell histiocytosis. Abnormal laboratory findings include pancytopenia, hypofibrinogenaemia, hyperlipidaemia, hyperferritinaemia, raised plasma levels of soluble IL2 receptors, and decreased natural killer (NK) cell activity. Cytological and histopathological studies show phagocytosis, by morphologically benign histiocytes, of white cells and platelets as well as red cells; in the liver a picture resembling chronic active hepatitis may be present.

HLH usually progresses rapidly so **early diagnosis is crucial** especially as there is a **high response rate to a steroid/etoposide combination, with intrathecal methotrexate to treat CNS disease.** Durable remissions can be obtained in this way and in older patients (those with infection-triggered disease) the treatment can be curative. **Infants with familial HLH, however, invariably relapse after 3-12 months and for these children marrow ablation with bone marrow transplantation (from a matched, related donor or possibly a matched, unrelated donor; "MUD") is the only treatment with a curative potential.** For patients without a donor, cyclosporin A may have a role in "maintenance" therapy.

* Reference Hunter et al Semin. Oncol.18 p29-33 (1991)

Why a BPSU study?

There are four principal reasons:

1 HLH is a rare multi-system disease presenting to different specialists and is often difficult to diagnose
2 there is virtually no epidemiological research in HLH
3 although the aetiology is unknown, there is evidence for a genetic predisposition in many cases and this has implications for prenatal diagnosis
4 recent advances in therapy make it a potentially curable disease.

The BPSU survey sets out to address all of these issues.

A survey in Sweden estimated an incidence of 1.2 cases per million child population per year but this study was retrospective. Because the Swedish workers found that over half their cases were diagnosed, unexpectedly, at autopsy, the BPSU is circulating all Paediatric Pathologists (whether BPA members or not) with the "orange cards" - a feature unique to this study. Once a patient has been identified, serum and tissue samples are requested and are being stored at Kings's College Hospital, London (Dr M Layton).

Planned research projects include:

1 a systematic search for viral and other pathogens
2 studies of "informative" families to identify DNA probes "linked" to the disease
3 pathological studies to see whether the sporadic and familial forms of HLH can be distinguished
4 advice on clinical management: (to date, there have been few queries).

The HLH Study Committee, (comprising clinicians, Dr M Layton - Coordinator, Dr S Strobel, Dr J Pritchard, Dr G Miele-Vergani, Dr D Heney and pathologists, Dr M Malone - Coordinator, Dr I Moore, Dr S Humphreys and Dr M Layton) meets regularly, three to four times per year to review the study and pathological samples and to promote research projects.

To May 1992, twenty-two patients have been registered in the study; all cases submitted so far have been verified by the main clinical coordinator (Dr Stephan Strobel) to whom clinical enquiries should be addressed. Other enquiries should be directed to Dr Mark Layton.

Dr S Strobel - Institute of Child Health, 30 Guilford Street, London WC1N 1EH -
Tel: 071-242 9789
Dr M Layton - King's College Hospital, Department of Haematology and Oncology, Denmark Hill, London SE5 Tel: 071-274 6222

4 FUTURE DEVELOPMENTS

The BPSU has been particularly active over the past year, handling an ever increasing number of applications. With the continued high profile of the Unit nationally, we anticipate further applications for inclusion in the reporting scheme, and we therefore expect the reporting card to be full for the foreseeable future. With this in mind, we will be considering, in consultation with our respondents, whether to increase the number of spaces on the orange card.

1992 has so far seen the introduction of five new studies; Insulin Dependent Diabetes Mellitus in Under Fives,investigator Professor J D Baum - this is an extension of the 1988 diabetes study; three month study on Long-Term Parenteral Nutrition, investigators Professor D Candy and Dr S Devane. Two additional studies are due to start soon ie. Juvenile Dermatomyositis, investigators Dr J Sills and Dr D Symmons, and Non-Accidental Poisoning/Munchausen By Proxy, investigators Professor J Sibert and Professor R Meadow; these two studies represent a departure from previous BPSU methodology. The Juvenile Dermatomyositis study will, for the first time, include a database of non-BPA members of the Rheumatology and Dermatology specialty groups. The Non-Accidental Poisoning/Munchausen by Proxy represents a first joint application for two similar surveys. Also due to start this year is a study on *Haemophilus influenzae* type b Vaccination Failure, investigators Professor E R Moxon and Dr M Slack.

5 CASES REPORTED TO THE BPSU FOR THOSE DISORDERS EXAMINED UP TO 21 July 1992

5.1 TOTAL REPORTS

The number of cases reported to the end of 1991 are shown in **TABLE 7 (overleaf)**. In each column, the figure under "A" is the total number of reports received. The figure under "B" is the corrected (confirmed) figure excluding cases not yet reported by investigastors to the BPSU as followed-up, cases reported in error and those double reported within the BPSU system. Numbers of cases here may differ slightly from the preceding section for reasons of definition, including ascertainment from sources other than the BPSU.

TABLE 7 - Cases Reported June 1986 - December 1991

	1986 Jun-Dec		1987 Jan-Dec		1988 Jan-Dec		1989 Jan-Dec		1990 Jan-Dec		1991 Jan-Dec	
CONDITION	A	B	A	B	A	B	A	B	A	B	A	B
AIDS/HIV	25	18	35	19	47	31	30	22	209	163	169	133
Neonatal Herpes	17	8	29	18	27	9	27	18	14	13	23	14
Reye	35	15	45	24	47	25	22	16	19	9	33	13
Kawasaki	84	72	106	84	129	105	142	114	195	153	225	172
SSPE	23	14	27	18	13	6	21	12	25	13	20	12
Congen. Rubella	-	-	-	-	-	-	-	-	22	14	10	4
Galactosaemia	-	-	-	-	41	23	24	15	39	16	28	16
MMR-M	-	-	-	-	-	-	-	-	39	23	44	28
Chemistry Set*	-	-	-	-	-	-	-	-	-	-	18	14
AFP	-	-	-	-	-	-	-	-	-	-	47	16
AIS	-	-	-	-	-	-	-	-	-	-	70	18
HLH	-	-	-	-	-	-	-	-	-	-	16	-
ALL	184	127	242	163	304	199	266	197	562	404	703	440

A: All reports received B: Cases confirmed 21 July 1992

Notes: These tables exclude previously completed studies (see p27)

AIDS/HIV: Reports of AIDS in June 1986 included all cases previously seen; case definition extended to include HIV infection in January 1990

Neonatal Herpes: Reports in June 1986 include all cases seen in the previous 12 months

SSPE (Subacute Sclerosing Panencephalitis):
a) Reports in June 1986 include all cases seen in the previous 12 months
b) Cases "not confirmed" include all those outside England and Wales which are not followed up by CDSC

MMR-M (Meningoencephalitis associated with MMR vaccine): reporting began January 1990.

Congenital Rubella: reporting began January 1990

Chemistry Set Poisoning: reporting began January 1991 ended in April 1992
*cases reported to April 1992

AFP (Acute Flaccid Paralysis): reporting began August 1991

AIS (Androgen Insensitivity Syndrome): reporting began September 1991

HLH (Haemophagocytic Lymphohistiocytosis): reporting began September 1991

5.2 FOLLOW-UP REPORTS

The time taken to follow-up a report varies greatly between conditions as does the "accuracy" of reporting measured by the proportion of cases confirmed (valid). Where microbiological/pathological details are needed for confirmation of a case, follow-up reports may be delayed. **TABLE 8** shows the outcome of follow-up, by the appropriate research worker, of all cases reported up to the end of 1991. The possible outcomes are explained below the table.

TABLE 8 - Outcome of Follow-up (cases reported to end of 1991 as of 21 July 1992)

CONDITION	VALID	INVALID		NYK	TOTAL	PERCENTAGE OF:		
	I	IIa	IIb	III		I	II	III
AIDS/HIV	386	80	46	3	515	75	24	1
Neonatal Herpes	80	12	45	0	137	58	42	0
Reye	102	30	65	4	201	51	47	2
Kawasaki	700	80	68	33	881	79	17	4
SSPE	75	15	16	23	129	58	24	18
Cong. Rubella	18	3	5	6	32	56	25	19
Galactosaemia	70	36	22	4	132	53	44	3
MMR-M	51	6	23	3	83	61	35	4
Chemistry Set+	14	1	3	0	18	78	22	0
AFP*	16	2	0	29	47	34	4	62
AIS	18	1	7	44	70	26	11	63
HLH*	0	0	0	16	16	0	0	100
ALL	1530	266	300	165	2261	68	25	7

* **Awaiting microbiological/pathological details**
\+ **Follow-up to April 1992**

OUTCOMES

I VALID REPORT:

Case followed up and confirmed by research worker as both unique and satisfying the diagnostic criteria, also includes cases first known to research worker from another source.

II INVALID REPORT:

IIa Duplicate report from within the BPSU scheme.
IIb Reporting error (eg. ticked wrong box, revised diagnosis, uncertain case not meeting definition, or unable to follow-up).

III NOT YET KNOWN:

Details not yet received by BPSU from research worker.

6 PARTICIPATION

The number of paediatricians participating in the scheme in 1991 ranged from 871 to 1040. The increase is due to the expansion in new consultant posts and the inclusion of consultants who were not previously identified by the unit and thus were not participating in the scheme. To ensure the mailing list is up-to-date, the BPSU office notes changes sent in by members for the BPA handbook and monitors new consultant appointments. **The average response rate for the year (calculated as the percentage of cards sent out which have been returned within 90 days after mailing) was 90.5% overall**; a monthly and quarterly breakdown is given in **TABLE 9**.

TABLE 9 - Monthly Response Rate 1991

MONTH	CARDS SENT	RETURNED	RESPONSE RATE	AVERAGE FOR QTR
January	869	797	91.7%	
February	876	802	91.5%	
March	879	793	90.2%	91.3%
April	902	818	90.7%	
May	909	821	90.3%	
June	917	824	89.9%	90.3%
July	950	855	90.0%	
August	948	844	89.0%	
September	967	859	88.8%	89.3%
October	985	897	91.1%	
November	1014	925	91.2%	
December	1018	927	91.1%	91.1%

TABLE 10 (overleaf), shows for each Region the average monthly number of members, card returns and response rate for 1991. The Republic of Ireland, Northern, Western and Southern Scotland are treated as Regions. The response rate varies considerably between regions. The highest average Regional response rate for 1991 was 100% (North and South Scotland) and the lowest average Regional response rate was 81.6% (North East Thames).

Members who have not returned their card for four consecutive months are sent a reminder letter.

TABLE 10 - Average Monthly Response Rate by Region - 1991

MONTH	CARDS SENT	RETURNED	RESPONSE RATE (%)	RANKING
Northern	54	52	96.3	3
Yorkshire	54	51	94.4	6
Trent	62	56	90.3	12
E Anglia	27	26	96.3	3
NW Thames	57	48	84.2	19
NE Thames	87	71	81.6	20
SE Thames	71	64	90.1	14
SW Thames	41	36	87.8	18
Wessex	37	33	89.2	16
Oxford	37	35	94.6	5
S Western	43	40	93.0	7
W Midlands	79	73	92.5	9
Mersey	34	30	88.2	17
N Western	59	54	91.5	10
Welsh	41	38	92.7	8
N Scotland	19	19	100.0	1
S Scotland	24	24	100.0	1
W Scotland	38	34	89.5	15
N Ireland	21	19	90.5	11
Irish Rep	51	46	90.2	13
TOTAL	936	847	90.5	

7 PUBLICATIONS 1991-2

Outcome in Children Who Nearly Drown: a British Isles Study -
Kemp A M, Sibert J R. Brit Med J 1991; 302:931-3

Drowning and Near Drowning in children in the United Kingdom: lessons for prevention -
Kemp A M, Sibert J R. Brit Med J 1992; 304:1143-5

Incidence of Insulin Dependent Diabetes in Children Aged Under 15 Years in the British Isles During 1988 -
Metcalfe M A, Baum J D. Brit Med J 1991; 302:443-7

Haemorrhagic Disease of the Newborn in British Isles: 2 year prospective study -
McNinch A W, Tripp J. Brit Med J 1991; 303:1105-1109

Meningoencephalitis associated with MMR vaccine -
Maguire H C, Begg N T, Handford S C. Communicable Disease Report, 1991; 1 (6) R57-R59

Rubella surveillance to December 1990: A joint report from the PHLS and National Congenital Rubella Surveillance Programme -
Miller E, Waight P A, Vurdien J E, White L M, Jones G, Miller B H R, et al.
Communicable Disease Report 1991; 1 (7); R86-R88

Haemorrhagic Shock Encephalopathy Syndrome in the British Isles -
Bacon C J, Hall S M. Arch Dis Child 1992; 67; 985-993

Investigation of Metabolic Disorders Resembling Reye Syndrome -
Green A, Hall S M. Arch Dis Child 1992; (in press)

Prenatal Screening for Toxoplasmosis: Report of a Multidisciplinary Working Group. London; Royal College of Obstetrics and Gynaecologists; 1992

Rett Syndrome: British Longitudinal Study (1982-1990) and 1990 Survey. In Mental retardation and Medical Care. Proceedings of the European Congress on Mental Retardation and Medical Care, 21-24 April 1991;
Roosendaal J J, Ed. Uitgeverij Kerckebosch, Zeist; 1992

8 PRESENTATIONS 1991-92

BPA Annual General Meeting 1991

Congenital Toxoplasmosis: what is the size of the problem? -
S M Hall, L Newton,
PHLS Communicable Disease Surveillance Centre (CDSC)

BPSU Galactosaemia Survey - M M Honeyman, A Green, J B Holton, J V Leonard,
Selly Oak Hospital and the Children's Hospital Birmingham; Southmead Hospital, Bristol;
Institute of Child Health, London

Epilepsy and Drowning - A M Kemp, J R Sibert, Department of Child Health,
University of Wales College of Medicine, Llandough Hospital Penarth, South Glamorgan

Family characteristics of Childhood-Onset Insulin-Dependent Diabetes -
M A Metcalfe, J D Baum, Institute of Child Health, University of Bristol

Kawasaki Disease (KD): a descriptive survey of methods of cardiac investigation and treatment in the British Isles in 1990 - R S Dhillon*, P T Rudd*, L Newton+, S Hall+ -
*Bath Unit for Research into Paediatrics, Royal United Hospital, Bath
+PHLS Communicable Disease Surveillance Centre (CDSC) London

BPA Annual General Meeting 1992

British Isles Rett Syndrome Survey - A M Kerr, J B P Stephenson,
Quarrier's Monitoring Unit, Royal Hospital for Sick Children, Glasgow

HIV and AIDS Surveillance in the British Isles: how big is the problem? -
C Davison, F Holland, C Duggan, M L Newell, A Nicoll, D Gibb,
Unit of Paediatric Epidemiology and Biostatistics, Institute of Child Health, London WC1,
PHLS AIDS Centre, London NW9

Other Meetings

Outcome of BPSU Survey - Galactosaemia Workshop, (Abstract)
British Inherited Metabolic Diseases Group, Birmingham 31 January 1992. Honeyman M M,
Green A, Holton J B, Leonard J V.

BPSU (British Paediatric Surveillance Unit) Galactosaemia Survey 8th International Screening Symposium and Inaugural Meeting of the International Society for Neonatal Screening,
12-15 November 1991, Australia (Oral presentation and publication in Proceedings Report).
Green A, Honeyman M M, Holton J B, Leonard J V.

Galactosaemia Present and Future. International Workshop - Metabolic Information Network.
A satellite meeting to the 8th International Congress of Human Genetics, Washington DC,
5 October 1991; Holton, J B.

9 OVERSEAS CONTACTS

Following preliminary discussions with delegates from several European countries, the first meeting of a Working Group for the establishment of a European Surveillance Network (Chairman Professor E Schmidt) was held during October 1991.

It was encouraging that delegates from the following European and Scandinavian countries attended, ie. Austria, Belgium, France, Germany, Greece, Holland, Spain, Denmark and Finland.

The Working Party has drawn up a Declaration of Intent which has been submitted to the EC to gain financial support for the development of an administrative structure. It is hoped that a core of five disorders, i.e. Haemorrhagic Disease of the Newborn, Diabetes in Under Fives, Reye Syndrome, Kawasaki Disease and Haemolytic Uraemic Syndrome will be included on the surveillance card of each country.

This development has the support of the Union of National European Paediatric Societies and Associations (UNEPSA), the organisation which speaks on behalf of the national paediatric associations of European Member States. It also has the support of the European Society of Paediatric Research which serves as the leading society for the presentation and review of paediatric research, particularly relating to paediatric epidemiology in Europe.

Further afield, the BPSU has also stimulated interest in Australia and New Zealand. Following a visit to Australia, our Medical Coordinator Dr Sue Hall reports that an interest was raised to the point that an Australian Surveillance Unit is now in its embryonic stages. It is hoped that, with the support of both the Australian College of Paediatrics and the BPSU, an 'APSU' will be active in the forthcoming year.

We have also heard from Professor R Dagan, of the University of Beer-Sheva in Israel about a reporting scheme in that country for certain serious invasive bacterial infections. Although the methodology is not exactly the same, the scheme was inspired by and modelled on the BPSU and has been running successfully since 1988.

10 FUNDING

The BPSU is currently receiving a substantial grant from the Medical Research Fund of Children Nationwide, paid through the Royal College of Physicians Appeal. This funding, due to end in September 1992, has been extended to the end of the year, pending the outcome of a further grant application.

The BPSU, in its early years, was supported by a donation from an anonymous Trust received by the BPA through the Royal College of Physicians of London. This donation has now ceased and we would like to express our thanks to the donor who kindly supported the BPSU through its initial years. In addition, we are grateful to Allen & Hanburys for their support in printing the booklets for the protocol cards.

The research workers are now paying the contribution requested by the Unit, which in 1990/91 was £120 per month; this covers the start up cost of a study, the printing and distribution of the cards and the administration. Due to inflationary pressures, this contribution is currently being reassessed and in future it is planned that the contributions from research workers will amount to at least 30% of the running cost of the Unit.

11 PREVIOUS BPSU SURVEYS

There have been eleven other surveys which have been completed. Information on these surveys is included in past BPSU Annual Reports, which are available free of charge from the BPSU office. Below is a listing of these studies and the principle investigator for each.

1 X-linked Anhydrotic Ectodermal Dysplasia (June 1986 - August 1986),
principle investigator, Dr A Clarke,
University Hospital, Wales

2 Lowe Syndrome (June 1986 - February 1988),
principle investigator, Dr C McKeown,
Department of Medical Genetics, St Mary's Hospital, Manchester 13 OJH

3 Insulin Dependent Diabetes in Under Fifteens (January 1988 - December 1988),
principle investigator, Professor J D Baum,
Institute of Child Health, Royal Hospital for Sick Children, Bristol

4 Drowning and Near Drowning (January 1988 - December 1989),
principle investigator, Professor J Sibert,
Department of Child Health, LLandough Hospital, Penarth, South Glamorgan CT6 1X

5 Higher Order Births (January 1989 - December 1989),
principle investigator, Professor M Levene,
Leeds General Infirmary, Belmont Grove, Leeds LS2 9NS

6 Haemorrhagic Disease of the Newborn (March 1988 - February 1990),
principle investigators, Dr A W McNinch, Dr H Tripp,
Department of Child Health, Royal Devon & Exeter Hospital, Barrack Road, Exeter EX2 5DW

7 Haemorrhagic Shock Encephalopathy Syndrome (HSES) (June 1986 - December 1988),

8 Haemolytic Uraemic Syndrome (HUS) (June 1986 - December 1989),

9 Congenital Toxoplasmosis (June 1989 - May 1990),
principle investigator for studies 7, 8 & 9 - Dr S Hall,
PHLS Communicable Disease Surveillance Centre, 61 Colindale Avenue, London, NW9 5EQ

10 Acute Rheumatic Fever (January 1990 - December 1990),
principle investigator, Dr C Boyd-Scobie,
c/o Dr S Hall PHLS Communicable Disease Surveillance Centre, 61 Colindale Avenue,
London NW9 5EQ

11 Rett Syndrome (April 1990 - June 1990),
principle investigator, Dr A Kerr,
Quarrier's Homes, Bridge of Weir, Renfrewshire, PA1 3SA

12 SUPPORT GROUPS FOR CURRENT AND RECENT CONDITIONS ON THE BPSU CARD

Reye Syndrome - Reye Syndrome Foundation of the UK
Mrs G Denney 15 Nicholas Gardens, Pyrford, Woking, Surrey GU22 8SD

Neonatal Herpes - Herpes Association, 41 North Road, London N7 9DP

Galactosaemia - Galactosaemia Support Group
Mrs S Bevington 18 Nuthurst, off Reddicap Heath Rd, Sutton Coldfield, W Mids B75

Congenital Rubella -
1) National Rubella Council, 33-39 Pancras Road, London NW1 2QB
2) SENSE (Deaf/Blind Rubella Handicaps) 31 Grays Inn Road, London WC1X 8PT

Encephalitis Effects -
'Contacts' Mrs O Fogg, 8 Seaside Lane South, Easington Colliery, Co.Durham SR8 3PF

Rett Syndrome - The Rett Syndrome Support Group
Mrs Y Milne, Heartpool, Golden Valley, Castlemorton, Malvern, Worcestershire WR13 6AA

For information on a variety of rare childhood disorders, a directory of support groups and their addresses has been produced by: **"Contact a Family"**, 16 Strutton Ground, London SW1P 2HP