British Paediatric Surveillance Unit

Fifth Annual Report 1990

BRITISH PAEDIATRIC SURVEILLANCE UNIT

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Fifth ANNUAL REPORT 1990

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1 FOREWORD

July 1991 marks the Fifth anniversary of the British Paediatric Surveillance Unit (BPSU). The BPSU has now come to be recognised as an important organisation for the gathering of data on rare childhood disorders. The success of the Unit can be seen in the increased number of applications for studies over the past year. There has also been continuing interest from a number of European countries who are considering the development of similar surveillance schemes. I can report that the BPSU is at the forefront of a plan to set up a network of surveillance units throughout Europe under the auspices of the Union of National European Paediatric Societies and Associations.

Six papers in which the BPSU was central to the methodology were presented at the British Paediatric Association's Annual Scientific Meeting in 1991; these studies reported on Congenital Toxoplasmosis, Galactosaemia, (plenary sessions), AIDS/HIV, Kawasaki Disease, Drowning and Diabetes (group sessions). The scientific value of the BPSU's work is reflected in the publication of BPSU papers in the British Medical Journal and the increasing number of references in other peer review medical journals and the medical press.

I would like to thank the staff of the Unit and members of the BPSU Executive Committee (BEC), who have met regularly to supervise the day-to-day running of the Unit. I would especially like to thank Sir Peter Tizard who after six years, (three years as BEC Chairman) is retiring from the BEC. Without Sir Peter's hard work in those early testing years I am sure the BPSU would not be the success it is today. Thanks should also go to Professor David Baum who shortly completes his three year term as BEC Chairman, to be replaced by Professor Euan Ross. Professor Baum has increased the profile of the BPSU such that it is recognised as the leading force in paediatric surveillance in Europe.

I should like to express my thanks to the officers and staff of the BPA for their continued support, to those bodies listed later in this report who have continued to give much needed financial support, and not least to the members of the British Paediatric Association and the Faculty of Paediatrics of the Royal College of Physicians of Ireland, whose continuing participation makes possible the BPSU's contribution to the health of children.

Finally I must remind you that the BPSU is here to respond to your surveillance needs and if you feel the BPSU can help facilitate a research interest which you may have, please do not hesitate to get in touch.

Sir Cyril Clarke
Chairman

MAY 1991

2 INTRODUCTION

The BPSU began operations in June 1986 as a national unit covering UK and Ireland enabling 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) Communicable Disease Surveillance Centre (CDSC), and the Department of Epidemiology at the Institute of Child Health, 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, and most importantly, still returning the card even if no cases have been seen (compliance with the scheme can thus be monitored). Following a case report, the BPSU office informs the relevant researcher who contacts the reporting paediatrician for further information in accordance with the study protocol for that condition. The research worker reports back to the BPSU on the outcome of their 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 Tables)

3.1 AIDS and HIV

Paediatric AIDS has been included in the BPSU reporting scheme since June 1986. In January 1990 the reporting definition was extended to include all children born to HIV positive mothers, and others infected by blood products. Paediatricians were also asked to report retrospectively any cases they had seen prior to January 1990. This scheme is the most important source of data on paediatric AIDS and HIV infection in the British Isles.

By March 1991, 431 cases had been reported through this scheme. Of these, 325 have been confirmed, 65 were duplicate reports, 31 were reporting errors, and 10 are awaiting confirmation.

TABLE 1

Exposure Category	AIDS cases (deaths)	Total HIV Seropositive
Haemophilia	15 (11)	41
Blood Transfusion	5 (3)	8
Child of HIV positive mother:		
Infected	47 (21)	95
Indeterminate		68
Uninfected		113
Total	67 (35)	325

All children reported are being followed-up annually to ascertain their infection and clinical status.

We are aware of a further 95 children of HIV positive mothers, and 189 haemophiliacs and 11 blood transfusion recipients who acquired HIV infection under the age of 15, but have not yet been reported through the BPSU.

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3.2 NEONATAL HERPES (HSV)

Between July 1986 and December 1990 there have been 112 notifications of suspected neonatal HSV infection, and 62 of these have been confirmed as cases. Of the remainder, nine were duplicates, 30 were excluded after further investigation, seven have been lost to follow-up and two are outstanding at the end of March 1991. The diagnosis could neither be confirmed nor excluded for the remaining two who were treated prophylactically but from whom the virus was not isolated.

Status of Notification	Number
confirmed cases	62
possible cases*	9
duplicate reports	9
not neonatal herpes	30
outstanding	2
TOTAL	**112**

* unable to confirm; two treated prophylactically; seven lost to follow-up

Among the 62 confirmed cases, three infants were born in the second half of 1986, 19 in 1987, 8 in 1988, 19 in 1989 and 13 in 1990.

There is a high case fatality rate: 17 (27%) infants died within a month of birth. Type 1 infections are as common as type 2, and deaths have occurred in the infants with type 1 infection (5/21) as frequently as amongst those with type 2 (6/20). Virus was not typed in 21 infants, six of whom died.

There was a history of maternal genital herpes, or evidence of a recurrent or primary infection in pregnancy in less than one-third of the cases. In most cases the maternal infection was diagnosed retrospectively, after diagnosis in the infant. HSV-1 and HSV-2 are equally represented among those infants whose mothers have evidence of genital herpes infection.

There were 11 cases where a possible postnatal source of infection was identified (parent or relative with cold sores around the time of delivery); in five of the 11 infants HSV-1 was isolated, in the remaining six virus type was unknown.

In 24% of infants no typical skin lesions or vesicles were noted; nine of these 15 infants died and in six the diagnosis was only made at post-mortem.

Further information is now being sought on those children who survived and are aged 18 months and over. Notifying paediatricians have received a brief report of the study and have been asked to provide details of any physical or mental impairment or any other condition which has been identified in the child. Follow-up continues and the data will be analysed and reported at a later date.

Paediatricians are asked to continue to notify all suspected cases of neonatal HSV **including those where infection is thought to have been acquired postnatally**. It is important that as far as possible all virus isolates are typed. However, it is not always possible to isolate virus from the infant, particularly once anti-viral treatment is underway. We are accepting as confirmed cases:

(a) infants from whom HSV has been isolated in the first month
(b) infants who have HSV - specific IgM in the first month
(c) infants who have typical clinical manifestations if maternal infection has been confirmed by either seroconversion or virus isolation around the time of delivery.

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3.3 REYE'S SYNDROME

Surveillance began as a joint BPA-CDSC venture in August 1981 and case ascertainment was transferred to the BPSU in July 1986. Annual totals of reports for Reye's Syndrome surveillance years (1 August-31 July) 1981/2 - 1988/9 were: 39, 60, 90, 61, 50, 47, 44, 31. A total 24 reports were received between 1 August 1989 and 31 July 1990. Of these, eight (33%) later had their diagnosis revised and for one case no further information was received.

Of the remaining 15 cases, the male to female ratio was 7:8. The ages ranged from 3.4 months - 15.5 years, with mean and median ages of 39.6 months and 8.4 months respectively. Outcomes were as follows: seven patients died, giving a case fatality rate of 47% (identical to that of 1988/9); four survived with neurological sequelae; three survived apparently normal and the outcome of one case is unknown.

Pre-admission medications were reported in five of the 15 patients: three had been given paracetamol, one paracetamol and metoclopramide and one an over the counter teething preparation. No patients had a history of pre-admission exposure to aspirin.

Fourteen patients resided in England and Wales and one in Scotland, with no confirmed cases coming from either Northern Ireland or the Republic of Ireland. No marked seasonal distribution could be discerned due to the small number of reports.

Among the eight cases in whom the diagnosis was revised the most common revision made was to one of the inborn errors of metabolism (five cases, mean age 12.7 months): fructose 1:6 diphosphate enzyme deficiency, lactic acidemia, long chain acylocoenzyme A dehydrogenase deficiency, one unspecified inborn error of organic acid metabolism and one unspecified metabolic disorder. The other revised diagnoses were: viral encephalitis with pneumonitis; haemorrhagic shock encephalopathy syndrome; myocarditis.

The annual number of cases of RS continues to decline. The median age of patients has remained constant at 8.4 months (eight months in 1988/90) and once again this may be due to an increasing proportion of patients with unrecognised inborn errors of metabolism. The 1990/91 surveillance questionnaire has incorporated additional questions to determine the extent to which patients are investigated for inborn errors of metabolism. To date (May 1991) 17 reports have been received for 1990/91 and of the 12 cases with follow-up information, six patients have revised diagnoses.

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3.4 KAWASAKI DISEASE

Kawasaki Disease (KD) surveillance began in 1983 as a joint BPA-CDSC venture and was transferred to the BPSU in 1986. In 1990 surveillance was extended to collect information on methods of investigation and treatment regimens, in order to provide background information for possible future treatment trials.

A total 183 reports of KD were received in 1990. This is the highest annual total since surveillance began and compares with 112 in 1989. At the time of writing detailed information had been received for 158 cases: the following preliminary analysis includes the first 140 of these.

105 (75%) cases came from England, with the greatest number of reports being received from the Northern and West Midlands Regions (14 each). Reports were also received from Wales (1), Scotland (15), Northern Ireland (6) and the Republic of Ireland (11). One case came from the Channel Isles.

82 of the 140 patients were male, (a male to female ratio as in previous years of 1.4:1). The mean and median ages were 37 months and 26 months respectively (range seven weeks - 15 years) and 82% of cases were aged under five years compared to 70% in this age group in 1989. As in previous years, white Caucasians were somewhat under-represented: 118 (86%) of 138 patients for whom ethnic background was described belonged to this ethnic group compared to 91% of the under 15 population as a whole. The remainder were: Indian (5%); Afro/Caribbean (4%); Oriental (2%); and one each (3%) were mixed race (white/Jamaican), Arab and Oriental (Filippino).

112 (80%) were "typical" cases (five or all of the six criteria of the KD case definition) and 28 (20%) were "atypical" (19 had four criteria and nine had three criteria). 113 patients recovered fully and six patients died, giving a case fatality ratio of 4% (compared to 0.2% in previous years). In four of the six deaths the diagnosis was first made at autopsy. The remaining patients recovered with sequelae, of which the most frequent was persistence of coronary artery aneurysms.

Cardiac involvement was reported in 40 cases, of whom 34 had coronary artery abnormalities including aneurysms (18 cases) and dilatation (11 cases).

125 (91%) of 137 patients with information on cardiological investigation underwent at least one echocardiagram and 115 had at least one electrocardiagram (ECG). The sensitivity of ECGs in detecting coronary artery abnormalities was, however, poor.

There were 137 patients with information on treatment. Eighty-one cases received specific KD treatment (aspirin and high dose intravenous gammaglobulin (IVGG)), 36 received only aspirin, two patients were given only IVGG and 18 cases received no specific KD therapy.

A 12 month follow-up study of 1990 cases is currently underway to provide information mainly on outcome, investigations and treatment following discharge.

The upsurge of cases seen in 1990 may represent the intermittent, usually triennial, epidemicity of KD observed in Japan and elsewhere. It is disturbing that it was associated with an increase in case fatality rate. Most patients received appropriate timely treatment but in some this was precluded by a delay in diagnosis, often at primary care level.

There is a need for heightened diagnostic awareness of KD by both GPs and paediatricians as it may be confused with measles and scarlet fever. Early institution of treatment with aspirin and IVGG has been shown to reduce the risk of coronary artery involvement in studies overseas. The efficacy of these regimens, in particular the optimum dose of aspirin, have not been determined in the British Isles. Treatment trials are, therefore, planned for 1992.

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3.5 CONGENITAL RUBELLA

The National Congenital Rubella Surveillance Programme was set up in 1971, one year after the introduction of the Rubella immunisation programme. During the eighties the impact of rubella immunisation became apparent as the number of cases of Congenital Rubella reported annually decreased; by 1989 about 1000 cases had been registered with the Surveillance Programme. Congenital Rubella is now a rare condition and has been included in the BPSU scheme since January 1990.

From January 1990, to April 1991 there have been 23 reports. 13 have been confirmed, nine of whom had rubella defects, including two who died in the first year of life and one who was stillborn; four children had no defect at the time of notification. The year of birth of these children is shown in the table below:-

TABLE 2

Year of Birth	1987	1988	1989	1990
Cases reported to BPSU	1	3	2	7
Cases reported/other sources	39	17	10	1
Total cases	40	20	12	8

Of the remaining 10 notifications, two were withdrawn, three were duplicates, one was excluded as the child was born abroad, one could not be confirmed as the child had received MMR vaccine before serological tests had been carried out and three are awaiting further information at the time of writing.

Since January 1990, an additional five cases have been reported from other sources.

Paediatricians are asked to continue to report all suspected cases of congenital rubella infection whether or not associated with typical rubella defects.

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3.6 SUBACUTE SCLEROSING PANENCEPHALITIS

Since the last report, 14 further cases resident in England and Wales have been ascertained via the BPSU. Five reports were duplicates, two were in error and in one case the diagnosis was uncertain. One case occurred in a child with acute lymphatic leukaemia. The remaining five cases were in previously healthy children, and have been entered into the main SSPE register. In three cases, the BPSU was the only source of notification; the other two were also reported to CDSC by a microbiologist.

The total number of cases on the register is now 296, of whom 246 have died. A recent noteworthy change is the proportion of cases occuring in Asian children. In the period 1962-1987, 15 of 278 (5.4%) children on the register were Asian; during 1988-1990 this increased to six of 18 (33%) cases reported.

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3.7 CONGENITAL TOXOPLASMOSIS

In view of the current debate about whether there should be a prenatal screening programme for toxoplasmosis in the UK, a survey to detemine the 'size of the problem' of diagnosed congenital toxoplasomosis (CT) was undertaken 1 June 1989 - 31 May 1990. Cases were ascertained both via the BPSU and via laboratory reports to the Public Health Laboratory Service Communicable Disease Surveillance Centre (CDSC).

A total 86 cases were reported (BPSU 40, CDSC 52, six common to both). Of the 86, 44 were rejected: 22 were not eligible because they were too old or had postnatal infection; two had clinical features suggesting CT, but no toxoplasma antibody; 20 showed a decline in antibody titres over the first year of life which was considered by clinicians to be consistent with maternal antibody rather than reflecting intra-uterine infection. However, not all were followed until the antibody titres were negative. None of these 20 had clinical features of CT and 17 were asymptomatic.

Of the 42 definite/probable cases, 17 belonged to the 12 month birth cohort of the study period and 25 were born before May 1989, but first diagnosed in the study year. Among the 17 cases born in the study year, four (three from England, one from the Republic of Ireland) had the 'classic triad' of CT; two had retinochoroiditis; six had other non specific symptoms (eg neonatal jaundice, failure to thrive, hypoglycaemia); four died in utero (microbiological confirmation of fetal infection in only one) and one was asymptomatic.

Among the 25 patients born before the study year (three via BPSU, 22 via CDSC) there were 19 children with isolated retinochoroiditis. Of these, four were aged under 18 months and at least three had been seen by a paediatrician; 15 were aged between six and 15 years (median 12) and were under the care of ophthalmologists.

Conclusions:

1 The observed "size of the problem" of symptomatic CT in the 89/90 England and Wales birth cohort, 14, was substantially less than the often quoted estimate of 60-70 used to justify a perceived need to introduce prenatal screening.

2 It is likely that children with CT who have retinochoroiditis are often under the care of ophthalmologists rather than paediatricians. This requires confirmation and further study to determine whether referral to a paediatrician would improve management and outcome.

3 Both the diagnosis and the management of all but the 'classic' cases of CT were problematic. The former reflected the limitations of the existing microbiological tests and the latter the absence of any properly controlled trial of treatment.

Practical Messages from this Study

1 There is a need for a heighted diagnostic vigilance for CT in order to ensure that the observed-expected difference in diagnosed numbers is not due to under-recognition.

2 Once suspected, either because of maternal infection or neonatal/infant symptoms, CT can only be ruled out by following the child's IgG antibody titres until they become negative using a reliable test, preferably the Sabin Feldman dye test. This decline may take over a year.

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3.8 RETT SYNDROME

Based in Glasgow, the Rett Syndrome survey was carried out against a background of eight years' research into the disorder. It was a joint initiative with colleagues in Gothenberg and Cardiff Universities and full cooperation from the National and UK Rett Syndrome Associations who recently produced and circulated teaching videos. The chief aim of the survey was to identify associations which might assist genetic investigation. A description of the classic syndrome was sent to paediatricians in the UK and the Republic of Ireland, followed by the survey protocol. Doctors were invited to report classic and atypical cases, male or female, born on or after 1 January 1975.

104 paediatricians reported 225 identified cases with 22 further reports still to be confirmed. Ten cases were born before 1975 leaving 215 cases so far identified within the invited age limits. Among 169 who were sufficiently described, classical Rett syndrome accounted for 150 (88%). 114 family histories have been received. After five years of age, when the diagnosis usually became evident, numbers reported in each year fluctuated between 12 and 25, with 17 born in 1975. Patchy reporting and a steady flow of new reports suggest that many cases are still being overlooked.

This survey has so far increased to 383 the total number of cases known at all ages in the UK and the Republic of Ireland. 91 were born before 1975, the oldest being 42 years. Among 152 family trees, associations have already been identified which are helping to direct genetic research. These include concordant monozygotic twins (one pair), discordant dizygotic twins, male/female (one pair) and female/female (one pair). One classic Rett syndrome girl is reported to have an atypical but Rett-like maternal aunt (manuscript in preparation, A. Clarke, Cardiff). No male has been found with classic Rett Syndrome but several families with profoundly handicapped, non-Rett children of both sexes are being investigated. Recurring schizophrenia and psychosis have been reported in a proportion of families. The early results of the survey indicate the prolonged healthy survival to be expected in Rett Syndrome and suggest a prevalence in the order of 1 in 10,000 girls, or 1 in 10 girls and women suffering from profound mental and physical disability. The genetic origin of the disorder and the exclusive appearance of this phenotype in females is indicated.

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3.9 ACUTE RHEUMATIC FEVER

The incidence of Acute Rheumatic Fever (ARF) has fallen dramatically in recent decades and the disorder is now rarely seen. However, during the mid-1980s there were reports of outbreaks of ARF in several areas of the United States and subsequently similar reports emerged from Europe. In January 1988, under the auspices of the BPSU, all paediatricians in the British Isles were asked to report any new cases of ARF seen during 1987; only nine were reported. Case ascertainment via such retrospective reporting was felt to be unsatisfactory so a 12 month prospective study was undertaken during 1990 with the main aim of determining the annual incidence of ARF in children less than 16 years in the British Isles.

A total 33 initial reports were received and at the time of writing further information had been provided on 29 of these. In a preliminary analysis using the revised Jones criteria, 16 children were categorised as 'most likely' ARF, a further five being possible cases. Four were considered not likely and the status of one was indeterminate. There were three duplicates. Of the 16 'most likely' cases, eight had carditis (diagnosis based on clinical findings only) seven had polyarthritis and five had chorea.

On receipt of data from all reported cases, the study will describe ARF reported to the BPSU by paediatricians in the British Isles in 1990 in terms of the numbers, age, sex, ethnicity of those affected, also clinical characteristics and those of the presumed antecedent streptococcal infection.

It is hoped that a comparison of the reported cases with the returns from the Korner data sets will provide an index of the usefulness of the latter as a source of these data. The preliminary analysis suggested that there are a number of cases in whom there is an overlap between 'classic' ARF and the increasingly recognised entity of post-streptococcal reactive arthritis. The investigators hope to review the applicability of the Jones criteria in the light of the cases reported and to draw attention to the broader spectrum of post-streptococcal disease.

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3.10 GALACTOSAEMIA

Between January 1988 and December 1990, 59 new cases of classical galactosaemia were confirmed (58 alive, one dead), see **TABLE 3** overleaf.

This gives an incidence of 1 in 45,000 births for UK and 1 in 23,500 for Eire. Scotland and Eire currently have screening programmes using blood spot measurements of GAL-1-PUT though England, Wales and Northern Ireland do not.

TABLE 3

Year	Total Cases	England/Wales/N.Ireland	Scotland	Eire
1988	25	17	2	6
1989	19	16	3	0
1990	15	12	2	1
		45 - unscreened	14 - screened	

Within the unscreened group, the diagnosis was made clinically in 37, nine (20%) of whom were actually unwell with encephalopathy, coagulopathy, septicaemia or jaundice requiring exchange transfusion. 4/14 (30%) in the screened group were actually unwell with three early clinical diagnoses and one false negative screen.

Excluding those with a positive family history of galactosaemia, 35/39 (90%) of the unscreened group were diagnosed clinically by one month, 29/39 (75%) by three weeks and 26/39 (66%) by two weeks. There were two diagnoses after two months when the additional delay might be expected to further compromise neurodevelopmental outcome.

Two older siblings had died from galactosaemia, both deaths occuring within the screened group at a time when screening was taking place. There were no other sibling deaths in either group.

We conclude that the introduction of new screening programmes for galactosaemia is not justified. Clinical vigilance should be increased by drawing attention to the variable clinical presentation where 'jaundice' is not invariable and to the relative frequency of galactosaemia within traveller' families, (1:7).

In view of the increasing concern about the long term implications (learning difficulties, speech disorder, ovarian failure and growth problems) and controversy over treatment regimes, the reporting period for galactosaemia has been extended into 1991, while a national cohort study is planned.

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3.11 MMR MENINGOENCEPHALITIS

Reporting of this condition began in February 1990. In the first year of the surveillance, 42 cases were notified to the BPSU. In two cases, the interval between vaccination and onset of symptoms was greater than six weeks. These cases, which did not meet the reporting criteria, were excluded. Of the remaining 40 cases, seven were classified as definitely vaccine related (vaccine-like virus recovered from CSF), 17 were probably related (no virus but onset 12-28 days after vaccination) and 12 were unrelated. Sufficient data for classifying the remaining four cases is currently unavailable.

The estimated incidence of vaccine-associated mumps meningitis was 14.5 per million doses distributed (definite and probable cases) and 4.2 per million doses (definite cases only). All cases reported to the BPSU scheme will be followed up a year after acute illness. The follow-up will include a general neurological examination, a Denver developmental assessment and a hearing test.

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4 FUTURE DEVELOPMENTS

It is hoped the forthcoming year will see a further increase in the number of studies applying to the BPSU for inclusion on the reporting card. A study examining the prevalence of toy chemistry set poisoning began in January 1991. This is an example of one paediatrician's particular interest being developed by the BPSU as a national survey.

In July 1991 a WHO inspired survey looking into acute flaccid paralysis will begin. It is hoped that this study will confirm the eradication of 'wild poliomyelitis' in the UK and thus enable the UK to receive a certification of eradication by the WHO.

Two further studies are imminent for 1991; these are Androgen Insensitivity Syndrome (AIS) and Haemophagocytic Lymphohistiocytosis (HLH). These two studies mark a departure for the BPSU in that the surveys will be incorporating respondents from other specialties; in the case of AIS - geneticists and in the case of HLH - pathologists. If successful the BPSU hopes to widen its database incorporating respondents from other specialties, as appropriate, for individual studies.

1992 could see the BPSU's involvement with therapeutic trials. Two such trials are currently under consideration, the first looking at high and low dosage of aspirin in the treatment of Kawasaki Disease; the second, at the administration of uridine in the treatment of galactosaemia .

5 CASES REPORTED TO THE BPSU FOR THOSE DISORDERS EXAMINED UP TO 1990

(excluding completed studies)

5.1 TOTAL REPORTS

The number of cases reported to the end of 1990 are shown in **TABLE 4** overleaf. In each column the figure under "A" is the total number of reports received and the figure under "B" is the corrected (confirmed) figure excluding cases not yet followed-up, cases reported in error and those double reported within the BPSU system. Numbers of cases here may differ slightly from the preceeding section for reasons of definition, including ascertainment from sources other than the BPSU.

TABLE 4 - CASES REPORTED: 1986, 1987, 1989 and 1990

	1986 June-Dec		1987 Jan-Dec		1988 Jan-Dec		1989 Jan-Dec		1990 Jan-Dec	
CONDITION	A	B	A	B	A	B	A	B	A	B
AIDS/HIV	25	19	35	19	47	33	30	22	205	158
Neonatal herpes	17	8	29	18	27	9	27	19	13	12
Reye	35	15	45	24	47	25	22	16	19	8
Kawasaki	84	72	106	84	129	104	142	114	195	152
SSPE	23	14	27	18	13	6	21	12	25	13
HDN	-	-	-	-	38	14	35	14	4	0
Galactosaemia	-	-	-	-	41	22	24	15	39	15
Toxoplasmosis	-	-	-	-	-	-	28	14	12	4
ARF	-	-	-	-	-	-	-	-	33	16
MMR-M	-	-	-	-	-	-	-	-	39	22
Rett	-	-	-	-	-	-	-	-	287	207
Rubella	-	-	-	-	-	-	-	-	22	14
ALL	184	128	242	163	342	213	329	226	893	621

A: All reports received B: Cases confirmed at 15/7/91

NOTES

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

Subacute Sclerosing Panencephalitis (SSPE) :

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.

Haemorrhagic Disease of the Newborn (HDN): Reporting began in March 1988 .

Congenital Toxoplasmosis: Reporting began in June 1989.

Rett Syndrome: Three month study to note all previous cases seen.

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

Acute Rheumatic Fever (ARF): Reporting began January 1990.

Congenital Rubella: Reporting began January 1990.

5.2 FOLLOW-UP OF 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 categories 1a or 1b). **TABLE 5** shows the outcome of follow-up, by the appropriate research worker, of all cases reported up to the end of 1990. The possible outcomes are explained below the table.

TABLE 5 - OUTCOME OF FOLLOW-UP
(cases reported to end of 1990 at 15/7/91)

CONDITION	VALID Ia	VALID Ib	INVALID IIa	INVALID IIb	NYK III	TOTAL	PERCENT IN EACH OF: I	II	III
HIV/AIDS	251	0	60	29	2	342	73	26	1
Neonatal herpes	66	0	8	39	0	113	59	41	0
Reye	84	4	23	50	7	168	52	43	4
Kawasaki	515	11	50	53	27	656	80	16	4
SSPE	39	24	14	12	20	109	58	24	18
Galactosaemia	52	0	26	18	8	104	50	42	8
HDN	27	1	2	39	8	77	36	54	10
Toxoplasmosis	18	0	0	21	1	40	45	52	3
ARF	16	0	3	10	4	33	48	40	12
Rett	207	0	17	38	25	287	72	19	9
MMR-M	19	3	2	13	2	39	56	38	5
Rubella	13	1	2	1	5	22	64	14	23
ALL	1307	44	207	323	109	1990	68	26	6

OUTCOMES

1 VALID REPORT:

1a: Case followed-up and confirmed by research worker as both unique and satisfying the diagnostic criteria.

1b: Case confirmed, but already known to research worker from another source (not a duplicate within the BPSU scheme).

2 INVALID REPORT:

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

3 NOT YET KNOWN:

Details not yet received by reseach worker 15/7/91

6 PARTICIPATION

The number of paediatricians participating in the scheme ranged in 1990 from 829 to 871. The increase is due to the expansion in new consultant posts and the inclusion of consultants who were not previously identified 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 89.7% overall; a monthly and quarterly breakdown is given in **TABLE 6**.

TABLE 6 - RESPONSE RATE BY MONTH, 1990

MONTH	CARDS SENT	RETURNED	RESPONSE RATE	AVERAGE FOR QTR
January	828	738	89.1%	
February	827	757	91.5%	
March	829	717	86.5%	89.1%
April	835	747	89.5%	
May	834	751	90.0%	
June	836	757	90.6%	90.0%
July	850	772	90.8%	
August	859	776	90.3%	
September	870	789	90.7%	90.6%
October	873	785	89.9%	
November	871	766	87.9%	
December	871	776	89.1%	89.0%

TABLE 7 shows for each Region the average monthly number of members, card returns and response rate for 1990. 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 1990 was 96% (East Anglia) and the lowest was 84% (North East Thames).

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

TABLE 7 - AVERAGE MONTHLY RESPONSE RATE BY REGION 1990

MONTH	CARDS SENT	RETURNED	RESPONSE RATE	RANKING (%)
Northern	46	42	91.3	8
Yorkshire	49	45	91.8	5
Trent	56	51	91.0	10
E Anglia	23	22	95.6	1
NW Thames	54	46	85.1	19
NE Thames	74	62	83.7	20
SE Thames	64	58	90.6	11
SW Thames	39	34	87.1	18
Wessex	34	31	91.1	9
Oxford	33	29	87.8	17
S Western	38	35	92.1	4
W Midlands	74	66	89.2	14
Mersey	30	27	90.0	12
N Western	53	49	92.4	3
Welsh	35	32	91.4	7
N Scotland	18	17	94.4	2
S Scotland	24	22	91.6	6
W Scotland	37	33	89.1	15
N Ireland	17	15	88.2	16
Irish Republic	50	45	90.0	12
TOTAL	849	761	89.7	

7 PUBLICATIONS 1990/1

The following reports have been published:

British Paediatric Surveillance Unit: Eighth Summary Report.
Communicable Disease Report 90/29

Report from the British Paediatric Surveillance Unit.
Hall S M, Glickman M. Archives of Disease in Childhood 1990.65:807-809

Kawasaki Disease: Lessons for Britain.
Bissenden JG, Hall S M. Brit Med J 1990. 300:1025-1026

Trends in Reye's Syndrome and Aspirin Use. Porter J D H, Robinson P H, Glasgow J F T, Banks J H, Hall S M. Archives of Disease in Childhood 1990. 65:826-829

Haemolytic Uraemic Syndromes in the British Isles 1985-88; Association with Verocytotoxin-Producing E coli. Milford D V, Taylor D M, Guttridge B, Hall S M, Rowe B, Kleanthaus H. Archives of Disease in Childhood 1990. 65:716-721

Kawasaki Disease Surveillance: Communicable Disease Report 90/41

Reye's Syndrome 1988/89 Update: Communicable Disease Report 90/09 and 90/45

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

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

Rubella Surveillance to December 1990: Miller E, Waight P A, Uurdien J E et al:
Communicable Disease Report 1:R4, 33-37

8 PRESENTATIONS

BPA Annual General Meeting 1990
Plenary Presentations:

The Outcome for Children Who Nearly Drown - A M Kemp, J R Sibert.

Vitamin K Prophylaxis and Haemorrhagic Disease of the Newborn -
A W McNinch, J H Tripp, J Handel.

The Epidemiology of Subacute Sclerosing Panencephalitis in England and Wales 1970-89.
C L Miller, P Farrington, K Rowe.

Group Presentations:

Drowning and Near Drowning in Children in the British Isles: Lessons for Prevention.
A M Kemp, J R Sibert (Community Paediatric Group).

Trends in Reye's Syndrome and in Aspirin Use. J Porter, P Robinson, J Glasgow, S Hall J Banks, (British Paediatric Immunology and Infectious Disease Group)

Modern Management of Infertility and Higher Order Births in the UK.
M I Levene, J Wild. P Steer. (British Association for Perinatal Medicine).

9 OVERSEAS CONTACTS

1990 has seen a positive commitment from the EEC to develop a network of Paediatric Surveillance Units in Europe. So far twelve countries have shown an interest in developing such units. To this end three preliminary discussions took place in 1990 under the auspices of the Union of National European Paediatric Societies and Association convened by Professor Eberhardt Schmidt from Dusseldorf: the first at the editorial meeting of 'Paediatrics in Europe' held in Dusseldorf during February; the second at the seventh European Paediatric Conference held in Budapest in April; and the third alongside the meeting of the European Society for Paediatric Research held in Vienna in September 1990.

An EEC funded meeting to formally initiate a European network will be held in September 1991, with the BPSU being prominent in its representation.

10 FUNDING

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. We would like to express our thanks to the donor who kindly supported the BPSU through its initial years. We are also grateful to Allen & Hanbury for their support in printing the protocol cards.

The unit is currently receiving a substantial grant from **Children Nationwide**, paid through the Royal College of Physicians Appeal. This funding will continue to September 1992.

The research workers are now paying the contribution requested by the Unit, which in 1990/91 was £95 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. In future it is planned that the contributions from research workers will amount to 30% of the cost of running the BPSU.

11 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 Road, Sutton Coldfield, W Midlands B75 7EZ

Congenital Rubella -

a) National Rubella Council, 33-39 Pancras Road, London NW1 2QB;
b) SENSE (Deaf/Blind Rubella Handicaps) 311Grays Inn Road, London WC1X 8PT

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

Congenital Toxoplasmosis -

a) The Toxoplasmosis Trust, Camden Studios, 11-15 Betterton Street, London WC2H 9BP;
b) Community Hygiene Concern, 32 Crane Avenue, Isleworth, Middlesex TW7 7JL.

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