BRITISH PAEDIATRIC SURVEILLANCE UNIT

Fourth

ANNUAL

REPORT

1989

BRITISH PAEDIATRIC SURVEILLANCE UNIT

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Please address any queries to the BPSU office.

BRITISH PAEDIATRIC SURVEILLANCE UNIT

Fourth ANNUAL REPORT 1989

1 FOREWORD

The British Paediatric Surveillance Unit has now entered its fourth successful year of monitoring the epidemiology of uncommon childhood disorders within the British Isles. The interest in the methodology of the unit from abroad has been such that similar schemes are to be developed in Europe in the near future.

The BPSU has also become recognised as an important research tool. Seven papers based on BPSU studies - Drowning and near drowning; Haemorrhagic disease of the newborn; Subacute sclerosing panencephalitis (plenary sessions); Reye's syndrome; Neonatal herpes; Higher order births; Drowning and near drowning (group sessions), were accepted for presentation at the British Paediatric Association (BPA) Annual Scientific Meeting in 1990. Several papers have also been published or are in press.

I should like to thank the staff of the Unit and members of the BPSU Executive Committee (BEC), chaired by Professor David Baum, who have met regularly to supervise the day-to-day running of the Unit. Special thanks should go to Mr Myer Glickman who, after administering the BPSU from the outset, has left to become a Research Fellow at the Institute of Child Health, London. Thanks should also go to the staff of the BPA for their assistance and to the bodies listed later in this report for their financial support. Most important of all, it is the paediatricians who spare time to participate in the reporting scheme by providing detailed information on reported cases, who ensure its continuing success.

Sir Cyril Clarke
Chairman

MAY 1990

2 INTRODUCTION

The BPSU was set up as an agency to facilitate surveys of uncommon infections and other rare childhood disorders within the child population of the UK and Ireland. The Unit is a joint project of the BPA, the Public Health Laboratory Service Communicable Disease Surveillance Centre (CDSC) and the Department of Epidemiology at the University of London Institute of Child Health, and it collaborates closely with the Communicable Diseases (Scotland) Unit in Glasgow which administers the scheme within Scotland. It began operations in June 1986. This report is concerned with the calendar year 1989.

The BPSU reporting system involves the mailing of a monthly card containing a 'menu' of reportable conditions to all consultant members of both the BPA and the Faculty of Paediatrics of the Royal College of Physicians of Ireland. Respondents return the card to the BPSU office, reporting the number of cases seen of the relevant disorders in the preceding month and - of the greatest importance - of still returning the card if there are no cases to report. When a case is reported the BPSU office informs the appropriate research worker who then contacts the reporting paediatrician for further information in accordance with the study protocol for that condition. The research worker also reports back to the BPSU on the outcome of their follow-up, confirming cases and identifying duplicate reports.

Conditions are removed from the report cards once the survey period has ended. They are replaced by new conditions once approval has been given by the BPSU Executive Committee (BEC). This committee meets monthly not only to review applications for approval but also to 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 15/16)

3.1 AIDS and/or HIV STATUS

Paediatric AIDS has been included in the BPSU reporting system since June 1986. In January 1990, AIDS surveillance was expanded to include any child found to be HIV seropositive. This was an important change as many HIV infected children may become seriously ill, or die, without ever reaching the surveillance definition of AIDS. All seropositive children are, therefore, to be monitored as well as cases of AIDS. Many of the seropositive children who are reported will not actually be infected because of persistence of maternal antibodies well into the first year of life, follow-up is required to determine their infection status.

Before the reporting instructions changed, a total 28 paediatric AIDS cases had been reported. All children born to seropositive mothers are now to be reported. If there is any doubt about whether they have previously been reported, respondents are asked to contact one of the principal investigators listed below, or to send a further report.

- Dr Clare Davison, Dept of Paediatric Epidemiology Institute of Child Health
- Tel: (071) 242 9789
- Dr Barry Evans, PHLS AIDS Centre CDSC 61 Colindale Avenue London NW9 5EQ
- Tel: (081) 200 6868.

3.2 NEONATAL HERPES

Neonatal Herpes simplex virus infection (HSV) has been included in the BPSU scheme since June 1986. In the first month retrospective notifications for the previous twelve months were requested, but thereafter paediatricians were asked to report cases on a month-by-month basis. There were eight "confirmed" cases (see p16 for definition) during the 18-month period covered by the June to December 1986 reports, five of whom died within a month of birth. These early notifications are unlikely to be complete or representative and have been excluded from the remainder of this report. Eighty five suspected cases of neonatal HSV infection were reported between January 1987 and December 1989, of which 45 have been confirmed. Of the remainder, seven were duplicates; 23 were excluded as they were either not neonates or did not have HSV, five were lost to follow-up and five are outstanding at the time of writing.

Of the confirmed cases, 19 were infants born in 1987, eight in 1988 and 18 in 1989. HSV 1 infection accounted for 40% of cases, HSV 2 for 29% and in 31% the virus was either not isolated or not typed (see Figure 1).

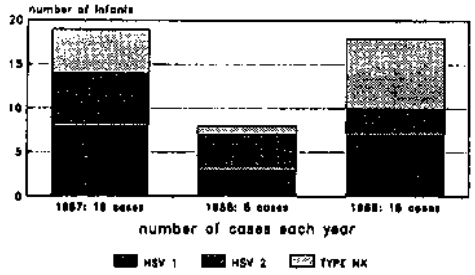

FIGURE 1 Year of Birth and Virus type

Thirteen (29%) of the 45 confirmed cases had disseminated disease, 13 (29%) had both skin and CNS manifestations; 16 (36%) had infection localised to the skin, oral cavity or eye and in three (7%) the CNS only was involved. **In ten infants, almost a quarter, no typical skin lesions or vesicles were noted; seven of these ten infants died and in five the diagnosis was only made at post mortem.**

Eleven infants (24%) died within four weeks of birth. At the initial follow-up, five of the 34 surviving infants (11% of the total) were reported to have suffered severe sequelae and **four (9%) were thought likely to have a good prognosis.** No opinion was given for 25 infants (56%). Further information on the surviving infants is being sought.

Sixteen (36%) mothers of the confirmed cases had evidence of either primary or recurrent genital infection during their pregnancy. **However, in all but one of these, maternal infection was diagnosed retrospectively after diagnosis of infection in the baby.** Three women (7%) had a history of genital herpes before the index pregnancy but in only one was this known before the diagnosis in the baby. In 26 women (58%) there was no evidence of genital herpes infection at any time before or during the index pregnancy; in seven of these 26 cases a possible postnatal source of infection was identified - three mothers, two fathers and two other relatives.

These reports to the BPSU suggest an estimated incidence of diagnosed neonatal HSV infection in the British Isles of approximately 2 per 100,000 live births, which agrees with estimates derived from laboratory reports to CDSC. However, a recent cross-check of BPSU and laboratory reports revealed that only 50% of the cases reported to the BPSU were reported to CDSC, and only about 40% of the cases reported to the CDSC were reported to the BPSU. This suggests that incidence rates derived only either from laboratory reports, or from BPSU reports may be under-estimates, not only of the true rate of neonatal infection, but even of the rate of diagnosed infection. Laboratories reporting cases to CDSC will be contacted in order to determine presenting features and outcome among patients not ascertained through the BPSU. Paediatricians are asked to continue to report all suspected cases of neonatal HSV **including those where infection is thought to have been acquired**

postnatally. It is important that all virus isolates are typed. However, it is not always possible to isolate virus from the infant, particularly once anti-viral treatment is under way. We are accepting as confirmed cases:

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

- Ms P A Tookey, Professor C S Peckham, Dr R Dinwiddie -Institute of Child Health, 30 Guilford St, London WC1N 1EH - Tel: (071) 242 9789

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. Total reports of cases of Reye's syndrome (RS) between 1 August 1981 and 31 July 1989 are shown in Table 1.

TABLE 1

REYE'S SYNDROME IN THE UNITED KINGDOM AND REPUBLIC OF IRELAND
CASES WITH FOLLOW-UP REPORTED BETWEEN 1/8/81 AND 31/7/89

12 month period (Aug-July)	Total reports British Isles	Classified as Reye's syndrome No.	Rate +	Revised diagnosis
1981/2	39	32	0.25	7 (18%)
1982/3	60	50	0.39	10 (17%)
1983/4	90	79	0.61	11 (12%)
1984/5	61	53	0.41	8 (13%)
1985/6	50	37	0.29	13 (26%)
1986/7	47	26	0.20	21 (45%)
1987/8	44	32	0.25	12 (27%)
1988/9	28*	19	0.16	8 (29%)
Total	419	328		90

\+ per 100,000 < 16 years of age
* detailed information not available for one case

Twenty-eight reports were received in the surveillance year 1988/9. Of these, eight cases initially met the case criteria, but then had the diagnosis revised to an inborn error of metabolism in three (glycerol kinase deficiency, partial ornithine transcarbamylase deficiency, proprionic acidaemia); and to haemorrhagic shock encephalopathy syndrome in two. One patient was found to have had meningoencephalitis and no conclusive alternative diagnosis was made for the last two, although metabolic disorders were suspected.

Of the remaining 19 cases with follow-up information and in whom the diagnosis was not revised, there were 11 males and 8 females. The median and mean ages were 7.7 months and 1 year 4 months respectively, with a range of 2.7 months to 6 years. Nine patients died, giving a case fatality ratio of 47% (cf 53% in 1987/8). A further three cases survived but with serious neurological sequelae. Four children who survived appeared to be normal and progress is as yet unclear on a further three. One patient had a history of pre-admission aspirin exposure. This compares with three in 1987/8 and 19 in 1985/6. No reports have been received from Northern Ireland for the second consecutive calendar year, continuing the striking decline in reports from this Region. The largest Regional number of reports, (4 cases) was from the North Western Region.

In spite of active ascertainment via the British Paediatric Surveillance Unit since 1986, Reye's Syndrome is still on the decline and the mean and median age of cases reported 1988/9 were the lowest recorded since RS surveillance began in 1981. These cases are likely to include patients with unrecognised inborn errors of metabolism, some of which are X-linked and the slight excess of males may reflect this. It is noteworthy that only 13 cases (3 with no follow-up information received at the time of writing) had been reported between 1.8.89 and 31.3.90, in spite of an influenza epidemic in the British Isles larger than that in any winter since 1976.

- Dr S M Hall - PHLS CDSC. 61 Colindale Avenue, London NW9 5EQ
- Tel: (081) 200 6868

3.4 KAWASAKI DISEASE

Surveillance of Kawasaki Disease was also a joint BPA - CDSC venture which began before the BPSU but transferred in July 1986.

A total 113 reports of Kawasaki Disease (KD) were received for cases with onset in 1989. (Annual totals for 1983-88 were 34, 15, 17, 75, 84, 111). For 14 cases in 1989 only the initial BPSU notification was received.

Reports were received from all Regions of England (except the North Western) and from Wales, Scotland, Northern Ireland and the Republic of Ireland. Reports were spread evenly over the year with the highest number in June. (Spring peaks have been described in Japan and the USA).

As in previous years there was a male excess, with a male/female ratio of 1.4:1. The mean and median ages were 43 months and 31 months respectively, range two weeks to 16 years. **70% of cases were aged under five years (22% under one year and 8% under six months).**

The ethnic group was given for 84 cases. Sixty six (79%) were white, eight (10%) were oriental/oriental-mixed race (an excess compared to the base population), and six (7%) were African/Caribbean (also an excess).

Of the 98 cases for whom detailed information was received, 95 (97%) presented with a fever persisting for more than five days; 89 cases (91%) had oral changes, 87 (89%) had a rash but only 63 cases (64%) had cervical lymphadenopathy > 1.5 cm in diameter.

Cardiac complications were reported in 30 patients (31%). Of these, 15 had aneurysms/dilatation of the coronary arteries. Seventy eight cases made a full, uncomplicated recovery and no deaths were reported in 1989. This compares with a case fatality ratio of 2% in previous years.

In 1990, Kawasaki Disease surveillance was augmented to collect data about current practice in management, in order to provide background information for possible future treatment trials.

- Dr S M Hall - PHLS CDSC, 61 Colindale Avenue, London NW9 5EQ
- Tel: (081) 200 6868
- Dr P T Rudd Royal United Hospital Bath - Tel: (0225) 823265

3.5 HAEMOLYTIC URAEMIC SYNDROME

Surveillance of haemolytic uraemic syndrome (HUS) began in 1983 as a BPA-CDSC joint venture. A total 189 reports were received for cases with onset in 1989. This is a marked increase compared to previous years. (Annual totals for 1983 - 1988 respectively were 47, 42, 83, 103, 117, 94). For 10 cases in 1989 only the initial BPSU notification was received. Reports were received from all Regions of England, from Scotland, Wales and the Republic of Ireland. The highest number of cases - 30 (17%) - was received from the Yorkshire Region. Reports were received each month of the year with a marked peak in the 'summer' months. Of those cases for whom the date of onset was known, 141 (82%) of reports occurred in the months May to October.

The mean and median ages were 53 months and 30 months respectively (range 8 - 177 months). **Five per cent of the cases were under 1 year of age, 45% were 1 - 2 years, 18% were aged 3 - 4 years,** 22% were aged 5 - 9 years and 10% were aged 10 - 16 years.
As in previous years the sex distribution showed a slight excess (52%) of females which was more marked in the older patients. Ten patients (6%) had a history of overseas travel in the month before onset. Places visited were: Pakistan (1), Spain (4), Greece (2), France (3).

In 1989 only 1 patient had no prodromal illness - an Asian male who had reflux vomiting once only. **Of those cases known to have a prodromal illness 158 (96%) had diarrhoea including 99 who had bloody diarrhoea.** 138 cases had vomiting and 17 had respiratory symptoms. The mean duration of diarrhoea was seven days, with a range of 1 - 42 days. Six cases (4%) had a non-diarrhoeal prodrome; four were female - the mean age was 67 months; Other prodromal symptoms, mostly vomiting and respiratory, were reported in all six cases.

Forty one cases (23%) reported a concurrent similar diarrhoeal illness in other members of the household. This included four sets of siblings reported with HUS at the same time, and two patients whose grandmother and mother respectively had concurrent HUS.

HUS surveillance was discontinued at the end of 1989 because of increasing laboratory based reporting of infections caused by the main associated micro-organism: verocytotoxin-producing E.Coli 0157. Paediatricians are therefore encouraged to request this investigation in patients with bloody or unexplained diarrhoea and/or HUS.

- Dr S M Hall - PHLS CDSC, 61 Colindale Avenue, London NW9 5EQ
- Tel: (081) 200 6868

3.6 SUBACUTE SCLEROSING PANENCEPHALITIS

By the end of 1989, 289 cases of subacute sclerosing panencephalitis (213 male and 76 female) were included in the main register, which began in 1970 and which is restricted to patients resident in England and Wales. After initial standard information, all cases are followed up at six monthly intervals until death. Since 1986 a total of 42 cases have been notified through the BPSU which is now the register's main source of information. Most cases are also eventually notified from other sources such as the Communicable Disease Report, but the BPSU has now been the sole notification source for five cases. The monthly return system also ensures that information about each patient can be obtained rapidly.

Recent analysis of the data shows that the distribution of age at onset of SSPE is changing. It is now more frequent in patients over the age of 15 than was the case in previous years. The explanation appears to be that these children contracted measles before the vaccination programme took full effect, while cases which would have been expected now in younger children have been prevented. There is therefore reason to hope that with the recent dramatic fall in notified measles due to better vaccine uptake, SSPE cases will show a genuine decline over the next 10 years. To monitor this, notification of every case is essential.

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

3.7 CONGENITAL TOXOPLASMOSIS

This survey began in June 1989 because of the increase in public and professional interest in this condition, caused by media attention to the findings of an antenatal serological survey in South Wales in 1987-88 which discovered an incidence of maternal 'acute' toxoplasmosis of 2 per 1000 pregnancies.

There has been pressure to introduce a national prenatal screening programme for maternal toxoplasmosis and the BPSU study was set up as one of several investigations to determine whether there is justification for such a programme.

Its main aim is to determine the annual diagnosed incidence of symptomatic congenital toxoplasmosis (CT). At the time of writing, the study, designed to run for one year, was not complete. In the first six months, 26 cases were reported for England and Wales. Of these, five were confirmed, eight cases appeared to be possible or definite CT, but were first diagnosed before 1 June 1989; eight cases did not have CT; for five cases the results of further serological investigation are awaited.

The small number (minimum 5 and possible maximum 10) of reports of newly diagnosed cases of CT in this six month period accords with previous surveys using data from the national laboratory reporting system to CDSC.

The findings also show that paediatricians often experience difficulty in making a diagnosis of CT, because of the long term serological follow-up necessary in non-specifically symptomatic infants and the unreliability of neonatal toxoplasma specific IgM as a marker of CT.

- Dr S Hall - PHLS CDSC, 61 Colindale Avenue, London NW9 5EQ
- Tel: (081) 200 6868

3.8 DROWNING AND NEAR DROWNING

Although drowning is the third most common cause of accidental death in children in Britain, there have been few studies of the outcome for nearly drowned children in this country. This survey covered a two year period from Jan 1988 to Dec 1989. Near drowning incidents were ascertained through the BPSU and further information on fatalities was supplied by the Royal Society for the Protection of Accidents (RoSPA), press cuttings and the OPCS. With the help of notifying consultants it was possible to pool a substantial amount of information regarding medical outcome and the circumstances of the incident.

At the time of writing there had been 144 reports of children admitted to hospital after nearly drowning. Fifty nine were unconscious with normally reacting pupils and all made a complete recovery - apart from two with cerebral palsy who probably died of secondary drowning. **Twenty nine children were unconscious with fixed or sluggishly reacting dilated pupils after resuscitation; of these, 13 subsequently died; of the survivors eight were profoundly handicapped with spastic quadriplegia and eight survived normal [three of whom were very hypothermic on admission].**

Drowning and near drowning cases can be divided into distinct groups characterised by the site of the accident and the age of the victim (see table). The younger age group (1-3 years) for example was involved in more incidents associated with garden/ private pools and baths than older children.

Older children were found to be involved in incidents related to rivers/lakes, sea and public pools, although in this last category only one fatality was reported.

TABLE 2
DROWNING AND NEAR DROWNING CASES FOR 1988
(England, Wales and Scotland)

	Near drowning	Drowning	Total	Average Age
Garden Pond	25	5	30	16 months
Private Pool	16	11	27	33 months
Bath	12	9	21	18 months
Public Pool	17	0	17	81 months
River/Canal/Lake	9	15	24	85 months
Sea	2	6	8	96 months
Other	4	4	8	
Unknown		3	3	
Total	85	53	138	

There is a strong case for prevention of drowning and near drowning accidents by statutory fencing of private pools. This is currently being proposed in Australia where pool incidents are significantly more frequent than in the UK. Garden ponds need to be included in building regulations and be modified to minimize the dangers of drowning to toddlers. Health visitors can also emphasize the dangers of bath drownings to mothers of young babies.

With the information collected it is hoped clear guidelines for management of children with near drowning can be produced.

- Dr Kemp, Dr J R Sibert - Dept of Child Health, Llandough Hospital, Penarth, South Glamorgan CT6 1X - Tel: (0222) 708601

3.9 HIGHER ORDER BIRTHS

In this survey, conducted Jan-Dec 1989, BPSU respondents were asked to report whenever they had been involved in the management of triplets or higher order births. Each responding paediatrician was asked to complete a brief questionnaire and to forward it on to obstetric and gynaecological colleagues in order to obtain information about the conception and the pregnancy. 113 sets of triplets, 10 sets of quadruplets and one set of quintuplets were notified. This represented a total of 384 babies and the survival figures for these babies were remarkably good. **The most important information to come out of this study was the fact that only two-thirds of all higher multiple births were associated with assisted reproduction. The commonest reason for higher multiple births was ovarian stimulation treatment followed by in vitro fertilization and then gamete intra-fallopian transfer. Of the 11 cases of quads and quins none resulted from natural fertilization; the commonest reason was ovarian stimulation.**

This information when fully collated will give important data on the contribution of modern management of the infertile couple to paediatric and neonatal services. A full report should be published later in the year.

- Professor Malcolm Levene, Dept of Paediatrics & Child Health, Leeds General Infirmary, Leeds LS2 9NS - Tel: (0532) 432799

3.10 GALACTOSAEMIA

Screening for classical galactosaemia has been introduced in many countries although the justification for it is equivocal. At the present time, Scotland and the Republic of Ireland both have screening programmes while England and Wales do not. In

order to obtain data on the incidence, clinical presentations and the start of treatment for this disorder, reports are being received via the BPSU over a three year period beginning 1 January 1988.

In 1988, 22 cases of classical galactosaemia and two Duarte variants were ascertained. Three additional cases were identified by biochemists in Ireland which had not been reported to the BPSU. Biochemists in the UK did not know of any cases not notified to the BPSU for 1988 which suggests that ascertainment is reasonably complete. In 1989 there were 14 confirmed cases in the UK with four additional cases under evaluation. This suggests that the incidence may well be higher than 1 in 70,000 births which is the generally quoted figure for the UK.

In most cases diagnosis is made as a result of clinical suspicion of galactosaemia with jaundice, failure to thrive and hepatomegaly. In 1988, 17 out of 22 cases were so diagnosed and 13 out of 14 cases in 1989. It is of diagnostic importance that jaundice was not invariable.

In 1988, 15 (94%) cases were diagnosed and treated by age 30 days and 100% of cases (14 cases) for 1989. In both 1988 and 1989, 75% of cases were diagnosed and treatment commenced by two weeks of age.

In the first year of the study there were two sibling deaths from galactosaemia but none were reported for 1989. Neonatal death of an index case from undiagnosed galactosaemia will, however, not be reported to our survey unless a presumptive diagnosis is considered post mortem on the basis of parental heterozygosity for GAL-1-PUT (galactose 1 phosphate uridyl transferase).

Paediatricians who had reported cases were contacted after the first year of the survey and all children born in 1988 were doing well. However, biochemical and clinical monitoring varied widely. Routine paediatric follow-up was a particular problem in itinerant families. **Reports of long term complications of intellectual impairment, other neurodevelopmental disorders and ovarian dysfunction suggest that the eventual outcome may be less encouraging. There is a need to standardise monitoring protocols to investigate these reports.**

Treatment with uridine, in addition to a galactose free diet, proposed on theoretical grounds and in sporadic clinical use in the USA is however of unproven effectiveness. A prospective treatment study in the UK is needed to evaluate this.

- Mrs A Green, Dr J Holton, Dr M Honeyman*, Dr J V Leonard - * The Child and Family Centre, 142 Maas Road, Northfield, Birmingham B31 2PR.
- Tel: (021) 477 0589

3.11 HAEMORRHAGIC DISEASE OF THE NEWBORN

The two year prospective study of haemorrhagic disease of the newborn has been completed. 25 unequivocal cases and two probable cases (typical history but no investigations performed) have been reported. All cases involved mature infants (above 36 weeks gestation) of normal birthweight for gestational age (2.3 - 3.8 kg).

In 17 cases no vitamin K prophylaxis had been given, in five cases oral vitamin K (0.5 - 1.0 mg), in two cases no data; one baby's notes stated that IM Vit K was given but parents denied this and it was subsequently thought to have been omitted in error. Twenty-four babies were solely breast fed, one primarily breast fed and two primarily bottle fed. Ten babies presented with intracranial haemorrhage (ICH) - two died, two required neurosurgery and there is concern about development in the eight survivors. Six babies (including four presenting with ICH) were found, after presentation, to have hepatitis.

The study confirms that HDN remains a preventable cause of mortality and morbidity in the British Isles. Babies who are solely breast fed are most susceptible and hepatitis, which may be clinically trivial, is an important additional factor in some. **No case of HDN was reported in a baby who definitely received intramuscular vitamin K. Oral vitamin K, 1 mg does not appear to protect all babies.**

- Dr A W McNinch, Dr H Tripp - Dept of Child Health, Royal Devon and Exeter Hospital, Barrack Road, Exeter EX2 5DW - Tel: (0392) 77833

4 FUTURE DEVELOPMENTS

From January 1990 the present AIDS survey will not only be monitoring the incidence of AIDS in children but also those children who are found to be HIV seropositive. It is hoped that those children who become seriously ill, or who die before being identified as an AIDS patient will be included.

A one year study of Acute Rheumatic Fever as an extension of an earlier "one-off" retrospective survey carried out by the BPSU in 1987 will also start in 1990. The study will set a baseline year for surveillance of the condition, and will allow a comparison with routine in-patient data, as well as measuring incidence and the characteristics of the condition.

Also included from January 1990 is surveillance of congenital rubella, funded by the Medical Research Council. This survey will help measure the effectiveness of the rubella immunisation programme.

A two-year survey starting in 1990 will examine the incidence of meningoencephalitis occurring within 6 weeks of a measles/mumps/rubella (MMR) vaccination.

A survey of Rett's Syndrome will commence in April 1990 and will last for three months. It is part of an established five year programme of research into profound mental handicap based at Quarrier's Monitoring Unit and the Royal Hospital for Sick Children, Glasgow. The study will examine the prevalence of Rett's syndrome, its sex association and co-inheritance with other rare genetic disorders.

5 CASES REPORTED TO THE BPSU FOR THOSE DISORDERS EXAMINED UP TO 1989 (excluding completed studies)

5.1 TOTAL REPORTS

The numbers of cases reported up to the end of 1989 are shown in Table 3. In each column the figure "A" is the total number of reports received and the figure under "B" is the corrected (confirmed) figure excluding cases not yet followed up, those 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.

TABLE 3
CASES REPORTED 1986, 1987, 1988, 1989 BY QUARTER

	1986 Jun-Dec		1987 Jan-Dec		1988 Jan-Dec		1989 Jan-Mar		Ap-Jun		Jul-Sep		Oct-Dec		Total	
CONDITION	A	B	A	B	A	B	A	B	A	B	A	B	A	B	A	B
AIDS	25	12	35	11	47	9	6	0	4	0	10	1	10	3	30	4
Neonatal Herpes	17	8	29	17	27	8	9	5	7	4	7	3	7	5	30	17
Reye	35	15	45	22	47	25	5	1	5	5	6	4	6	6	22	16
Kawasaki	84	72	106	84	129	104	46	39	41	31	26	25	29	18	142	113
HUS	35	30	63	47	86	69	13	10	77	61	73	61	44	21	207	153
SSPE	23	14	27	18	13	6	5	0	3	1	4	4	9	7	21	12
Galactossemia	-	-	-	-	41	22	4	1	5	4	9	4	6	1	24	10
Drowning	-	-	-	-	108	95	24	19	31	25	56	30	15	6	126	80
HDN	-	-	-	-	38	13	13	3	5	1	9	2	8	0	35	6
HOB	-	-	-	-	-	-	35	25	46	35	33	27	47	16	161	103
Toxoplasmosis	-	-	-	-	-	-	-	-	8	1	10	3	8	6	26	10
ALL	219	151	305	199	536	351	160	103	232	168	243	164	189	89	824	524

A: All reports received B: Cases confirmed at 24/4/90

NOTES

AIDS: a) Reports in June 1986 included all cases seen previously.
b) Cases "not confirmed" include many with HIV-related disease not meeting the strict definition of AIDS.

Neonatal herpes: Reports in June 1986 included all cases seen in the previous 12 months.

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

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

Higher Order Births (HOB): Reporting began in Jan 1989.

Congenital Toxoplasmosis: Reporting began in June 1989.

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 4** shows the outcome of follow-up by the appropriate research worker of all cases reported up to the end of 1989. The possible outcomes are explained below the table.

TABLE 4
OUTCOME OF FOLLOW-UP of cases reported to end of 1989 at 24/4/90

CONDITION	VALID Ia	VALID Ib	INVALID IIa	INVALID IIb	NYK III	TOTAL	% IN EACH OF I	% IN EACH OF II	% IN EACH OF III
AIDS	36	1	17	38	45	137	27	40	33
Neonatal herpes	50	0	8	31	14	103	49	38	14
Reye	74	4	20	45	6	149	52	44	4
Kawasaki	354	10	32	27	38	461	79	13	8
HUS	247	52	15	11	66	391	76	7	17
SSPE	29	21	11	9	14	84	60	24	17
Galactosaemia	32	0	9	11	13	65	49	31	20
Drowning	174	1	9	0	50	234	75	4	21
HDN	18	1	1	26	27	73	26	37	37
HOB	93	10	10	3	45	161	64	8	28
Toxoplasmosis	10*	0	0	8	8	26	38	31	31
ALL	1117	100	132	209	326	1884	65	18	17

* 'includes 5 possible cases'

OUTCOMES

I VALID REPORT:

Ia: Case followed up and confirmed by research worker as both unique and satisfying the diagnostic criteria.
Ib: Case confirmed, but already known to research worker from another source (not a duplicate within the BPSU scheme).

II 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.

III: NOT YET KNOWN: not yet followed up by research worker at 24/4/90.

6 PARTICIPATION

The number of paediatricians participating in the scheme ranged in 1989 from 818 to 833. To ensure that 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 the mailing) was **88.9%** overall: a breakdown by month and quarter is given in **Table 5.**

TABLE 5
RESPONSE RATE BY MONTH, 1989

MONTH	CARDS SENT	RETURNED	RESPONSE RATE	AVERAGE FOR QTR
January	831	736	88.6%	
February	830	736	88.7%	
March	833	740	88.8%	88.7%
April	830	743	89.5%	
May	828	759	91.7%	
June	824	728	88.3%	89.8%
July	822	728	88.6%	
August	820	728	88.8%	
September	818	744	91.0%	89.4%
October	819	747	91.2%	
November	818	741	90.5%	
December	824	749	90.8%	90.8%

Table 6 shows for each Region the average monthly number of members, card return and response rate for 1989. The Republic of Ireland and Northern, Western and Southern Scotland are treated as Regions. The response rate varies considerably between Regions. The highest average Regional response rate for 1989 was 100% (Northern Scotland) and the lowest was 83% (North West Thames).
Members who have not returned their card for six consecutive months are sent a reminder letter. These often turn out to be members who have retired, and who are therefore removed from the mailing list.

TABLE 6
AVERAGE RESPONSE RATE BY REGION, 1989

REGION	CARDS SENT	RETURNED	RESPONSE RATE (%)	RANKING
Northern	46	41	89.1	12
Yorkshire	46	44	95.6	2
Trent	56	50	89.2	11
EAnglia	23	22	95.6	2
NW Thames	54	45	83.3	20
NE Thames	71	60	84.5	19
SE Thames	59	53	89.8	11
SW Thames	36	32	88.8	13
Wessex	35	32	91.4	6
Oxford	33	28	84.8	18
SWestern	35	32	91.4	6
WMidlands	70	62	88.6	14
Mersey	31	27	87.1	16
Wales	53	48	90.0	9
NWestern	35	32	91.4	6
NScotland	17	17	100.0	1
SScotland	24	22	91.6	5
WScotland	34	30	88.2	15
NIreland	17	16	94.1	4
Irish Rep	53	45	84.9	17
TOTAL	825	737	89.3	

7 PUBLICATIONS 1989/90

The following reports have been published:

British Paediatric Surveillance Unit: Seventh Summary Report. Communicable Disease Report 89/15

Report from the British Paediatric Surveillance Unit.
Hall S M, Glickman M. Archives of Disease in Childhood 1989. 64:439-346

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 J G, Hall S M. Brit Med J 1990. 300:1025-1026

Trends in Reyes Syndrome and aspirin use.
Porter J D H, Robinson P H, Glasgow JFT, Banks JH, 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 C 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 90/09

BPSU/CDSC Reye Syndrome Surveillance Scheme : Communicable Disease Report 89/38

Reye Syndrome 1988/89 Update: Communicable Disease Report 90/09

Kawasaki Disease in the British Isles.
Dillon M J, Hall S M. In: Proceedings of the Third International Kawasaki Disease Conference Jap Heart Assn. Tokyo 1989. 48-51

8 PRESENTATIONS

BPA Annual General Meeting 1989

Plenary Presentations:

Paediatric AIDS and HIV infection in the UK - Dr G A Ellam
Surveillance of Kawasaki disease in the British Isles - Dr S M Hall
National survey of childhood-onset diabetes, 1988 - Professor J D Baum

Group Presentations:

British Association for Paediatric Nephrology -
The expression of blood group P1 in post enteropathic haemolytic uraemic syndrome - Dr D V Milford

Presentations on the BPSU by the Medical Co-ordinator in 1989

1) To public health physicians - 7
2) To paediatricians - 1 (Leicester "roadshow")
3) To overseas visitors to CDSC - 4 (Dept of Health Ireland; Canada; Saudi Arabia; New Zealand)

9 OVERSEAS CONTACTS

This past year has seen increasing interest in the BPSU both in this country and overseas. The development of the BPSU was discussed at the 1990 meeting of the Union of National European Paediatric Societies and Associations (UNEPSA). It is hoped that with the support of UNEPSA and with the financial backing of the EEC, similar schemes can be established within Europe, leading to collaboration of national paediatric surveillance schemes in Europe.

10 FUNDING

The BPSU is now primarily funded through a substantial grant from **Children Nationwide**, paid through the Royal College of Physicians Appeal. This funding will continue for the next three years. Previously, the BPSU was supported by a donation from an anonymous Trust received by the BPA through the Royal College of Physicians of London which 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 the printing of the protocol cards and folders.

The research workers are now paying the contribution requested by the Unit, which in 1989/90 was £95 per month. In future, it is planned that this will contribute 30% of the cost of running the BPSU.

NOTES