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British Paediatric Surveillance Unit

First Annual Report 1985/86

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1985/86

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BPSU 1987

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Numbers of cases are unconfirmed reports unless otherwise stated and should not be taken to indicate actual numbers of patients.

1 FOREWORD

This is the British Paediatric Surveillance Unit's first annual report, and I am pleased to say that this unique initiative has got off to a most impressive start. Two years from the first tentative proposals for a scheme to improve the efficiency of surveillance of rare childhood diseases, one three-month survey has already been completed and eight continuing studies are in progress.

We are grateful for generous financial and practical support which has helped to make this possible, from John Wiley & Son Ltd, Duphar Laboratories Ltd, Allen & Hanburys Ltd and an anonymous Trust.

I should like to thank the Scientific Advisory Committee, and in particular its Chairman, Sir Peter Tizard, who bear the main burden of the running of the Unit. I should also like to thank the staff of the Unit, Dr Susan Hall and Mr Myer Glickman, for their work during the year.

Sir Cyril Clarke
Chairman of Steering Committee

2 INTRODUCTION

The British Paediatric Surveillance Unit (BPSU) was established in 1985 with the aim of involving paediatricians nationally in the surveillance of rare childhood disorders, making possible the ascertainment of cases on the scale needed for both clinical and epidemiological study of such uncommon diseases as Reye's syndrome, neonatal herpes and subacute sclerosing panencephalitis (SSPE). The Unit is a joint venture of the British Paediatric Association (BPA), the Communicable Disease Surveillance Centre of the Public Health Laboratory Service, and the Department of Epidemiology at the University of London Institute of Child Health.

The basis of the BPSU reporting scheme is the mailing of a monthly report card to all consultant paediatrician members of the British and Irish Paediatric Associations. Respondents return the card to the BPSU office in London, marking the number of cases seen against of specific conditions or ticking a "nothing to report" box. For respondents in Scotland, the scheme operates via the Communicable Diseases (Scotland) Unit in Glasgow.

Notification of a case is forwarded to a research worker studying that particular condition, who sends a short questionnaire (which has been approved by the BPSU) to the reporting consultant or requests a loan of the case notes. The researcher subsequently notifies to the BPSU the number of confirmed cases, and for certain conditions gives basic epidemiological data.

The full implementation of the scheme was preceded by a pilot mailing to a sample of BPA members and a "pre-mailing" of information to all Ordinary Members. The first full mailing took place in July 1986.

3 PILOT MAILING

A pilot mailing was carried out in November 1985 - March 1986 to obtain the views and comments of a sample of BPA members on the concept and operation of the reporting scheme.

The sample consisted of 49 consultant paediatricians from all Health Regions in the British Isles, the 18 Convenors of BPA specialty groups, and two other members, making a total of 69. Members of the sample were sent a draft Introductory Booklet giving general information on the scheme, a report card on which 10 "dummy" reportable conditions were listed, and a set of reporting instructions. They were asked to comment on the design and content of all of these and on whether a "reply paid" card would achieve a better response rate.

Answers were received from 66 of the 69 members selected (96%). Many respondents volunteered their enthusiasm and support for the BPSU. Only one respondent anticipated a poor response in general and resolved not to co-operate with the scheme.

Most respondents found the design of the documents clear and easy to use. Some minor modifications to the report card and Introductory Booklet were made as a result of respondents' ccomments. The most important points made were: that follow-up questionnaires must be short and simple to complete; and that the procedure for deciding which conditions to include in the scheme must be clear and be seen to be fair.

On "reply paid" postage, a majority of respondents (62% of those answering the question) did not think that a "reply paid" card would achieve a better response, while 16 (38% of those answering) felt that it would. The remaining 24 made no comment on postage. In view of the limited budget of the BPSU it was therefore decided to start the mailing using cards which were pre-addressed but not "reply paid".

4 INCLUSION OF STUDIES

4.1 Studies included in the scheme to date

The following studies are or have been included in the scheme:

AIDS in childhood

Principal research workers: Dr A Ellam & Dr M McEvoy
Research base: CDSC
Duration of study: June 1986 for 3 years (then subject to review)

Childhood onset diabetes

Principal research worker: Professor J D Baum
Research base: BPSU
Duration of study: September 1986 for three months
This is a pilot survey in the South West health region only

Neonatal herpes

Principal research workers: Dr R Dinwiddie & Professor C Peckham
Research base: Institute of Child Health
Duration of study: June 1986 for 1 year (then subject to review)

Reye's syndrome
Kawasaki disease
Haemolytic uraemic syndrome (HUS)
Haemorrhagic shock encephalopathy syndrome (HSES)

Principal research worker: Dr S Hall
Research base: BPSU/CDSC
Duration of studies: June 1986 for 3 years

Subacute sclerosing panencephalitis (SSPE)

Principal research worker: Dr C Miller
Research base: CDSC
Duration of study: June 1986 for 3 years (then subject to review)

X-linked anhidrotic ectodermal dysplasia (XLAED)

Principal research worker: Dr A Clarke
Research base: University Hospital of Wales
Duration of study: June 1986 for 3 months
This survey has now been completed

4.2 Studies under consideration

The decision whether or nor to add a condition to the list of reportable diseases is taken by the Unit's Scientific Advisory Committee. The Committee meets monthly to consider proposals for and protocols of new studies.

Proposals under consideration or which have been accepted for future
inclusion in the scheme are:

Accidental drowning and near-drowning
Atlanto-axial dislocation in Down's syndrome
Chronic liver disease
Lowe's syndrome
Meningococcal septicaemia
Recurrent or persistent hyperinsulinaemic hypoglycaemia

4.3 Studies not included in the scheme

An important aim of the BPSU is to improve efficiency of ascertainment by including in a unified system as many reporting schemes as possible. However, after careful discussion with project directors it was thought best that two major national reporting schemes, the Karim Centre meningitis study and the congenital rubella register, should remain separate from the BPSU. A comprehensive list is being compiled of all other national or regional enquiries to which paediatricians are currently asked to contribute.

After extensive discussion, a proposal for surveillance of Glucose-6-phosphate dehydrogenase (G6PD) deficiency was declined.

Considerations which might result in the exclusion of a particular study from the surveillance system include: the involvement of other specialties in case ascertainment; a need for immediate specimen collection; or the condition being too common to come within the sccope of the scheme. To accept a study, the Scentific Advisory Committee must be convinced that it is methodologically sound, will represent a valuable contribution to medical knowledge and will not create an unreasonable addition to the workload of those who report cases. The Committee must also be assured that adequate funding has been secured for the completion of the research.

5 PROGRESS OF THE SCHEME

5.1 Response rate

Over the first three months of the reporting scheme, on average over 80% of members on the mailing list returned their card, as shown in Table 1 below.

Table 1: RESPONSE TO MAILING - FIRST QUARTER

	June	July	August
Date of mailing:	16/7/86	5/8/86	8/8/86
Total cards sent:	824	821	820
Total cards returned:	603	681	704
Cards returned as %age of total sent:	73.2%	83.0%	85.9%
Total cases reported:	49	22	37
Cases reported as %age of cards returned:	8.1%	3.2%	5.3%
Cases reported as %age of total sent:	6.0%	2.7%	4.5%

(correct at 19/11/86)

The number of cases reported in the first month was greater than that in succeeding months because the reporting instructions for that month for some of the conditions requested all cases which had been seen over the previous twelve months, or all cases known to the respondent.

Small variations in the size of the mailing list result from identification of omissions from the original list, from new appointments, retirements, changes of post or address and exclusion of those whose work is non-clinical.

5.2 Cases reported

The scheme has provided a marked improvement in case ascertainment for some of the conditions which were previously the subject of surveillance schemes with a different methodology. Results have been particularly notable for AIDS in childhood, Kawasaki disease and XLAED.

Except where stated, the figures in Table 2 (overleaf) include only confirmed cases; further cases have been reported but not yet confirmed.

Table 2: CASES REPORTED - FIRST QUARTER

Condition	Confirmed cases			
	June	July	August	Total
AIDS	8 [1]	-	-	8
Herpes	6* [2]	-	1*	7*
Reye's	6	1	2	9
Kawasaki	7	9	10	26
HUS	1	5	4	10
HSES	1	-	-	1
SSPE	3 [2]	-	3	6
XLAED	8* [1]	-	5*	13*

(correct at 19/11/86)

* Includes unconfirmed cases
[1] Month 1 figures included all cases known to respondents
[2] Month 1 figures included cases seen in past 12 months

Of cases reported to date, only one has been "double reported" within the scheme. Nine have been "parallel reported" with other methods of ascertainment such as laboratory reporting.

6 CHARITABLE & COMMERCIAL SUPPORT

The setting-up of the Unit was made possible by a donation from an anonymous Trust and by a donation form a research fund received by the BPA through the Royal College of Physicians of London.

The organisers of the Harley Street Ball, an annual charitable event, have kindly donated the proceeds of the first Ball, held in September 1986, to the BPSU.

Generous practical support from industry and commerce has included printing of the report cards by John Wiley & Son Ltd; printing of the Introductory Booklet by Duphar Laboratories Ltd; and production of the study protocols in permanent reference form by Allen & Hanburys Ltd.

Approved by Steering Committee 19/11/86

BRITISH PAEDIATRIC SURVEILLANCE UNIT : FIRST QUARTERLY SUMMARY REPORT

The British Paediatric Surveillance Unit (BPSU), described in CDR 86/28, sent out the first of its monthly cards to all hospital consultant paediatricians in the British Isles in July (returns from Scottish paediatricians are received via the CD (Scotland) Unit). This report summarises the Unit's activities and results for the first 3 months' mailings. During this period the reportable conditions were: AIDS in childhood; neonatal herpes; Reye's syndrome; Kawasaki disease; haemolytic uraemic syndrome; haemorrhagic shock encephalopathy syndrome; subacute sclerosing panencephalitis.

Response rate

Because paediatricians are asked not only to report cases but also to make a nil return if no patients with any of the reportable conditions have been seen in the past month, general compliance with the scheme can be measured from the proportion of total cards returned. Table 1 shows that this has been high, increasing to nearly 85% of respondents by the third month.

Table 1 Overall response rate (at 5.11.86)

	Reporting month			Total
	June Number (%)	July Number (%)	August Number (%)	
Total cards sent†	824 (100)	821 (100)	820 (100)	-
Total cards returned†	603 (73.2)	680 (82.8)	698 (85.1)	-
Total cases reported*	49	22	37	108

† Includes those sent by, and returned to, Communicable Diseases (Scotland) Unit
* All conditions as listed in table 2

The number of cases reported for June was greater than in succeeding months because the reporting instructions for that month for some of the conditions (see table 2), requested all cases who had been seen up to a year before. Small variations in the size of the mailing list result from new appointments, retirements and changes of post and address.

Table 2 Cases reported (at 19.11.86)

Condition	Reporting month			Total
	June	July	August	
AIDS in childhood†*	12 (8)	- (-)	- (-)	12 (8)
Neonatal herpes§	6 (-)	- (-)	1 (-)	7 (-)
Reye's syndrome	7 (4)	3 (1)	3 (1)	13 (6)
Kawasaki disease	8 (6)	9 (6)	15(12)	32(24)
Haemolytic uraemic syndrome*	3 (1)	9 (4)	8 (2)	20 (7)
Haemorrhagic shock encephalopathy syndrome	1 (1)	1 (-)	- (-)	2 (1)
Subacute sclerosing panencephalitis§*	4 (3)	- (-)	3 (-)	7 (3)
X-linked anhydrotic ectodermal dysplasia†*	8 (-)	- (-)	5 (3)	13 (3)
	49(23)	22(11)	37(18)	108(52)

Numbers refer to total reports received; those in brackets refer to cases followed up and confirmed by investigators.
† June figures included all cases ever seen by respondents
§ June figures included all cases seen in past 12 months
* These conditions are also ascertained by other surveillance schemes whose data are not included.

Of the paediatricians mailed there were 69 who did not return any of the first 3 cards. These non-respondents were an important indicator of possible shortcomings of the system and they were therefore asked to complete a brief proforma requesting possible reasons for non-participation. Of the 45 replies received to date (after 5 weeks), 20 were in a sub-specialty or retired or not in clinical practice and unlikely to see patients with the reportable conditions; 6 were unaware that nil returns were requested; 3 had not received the cards, 3 had been away from work; in 4 cases another paediatrician was replying on their behalf; 6 gave no reason but returned their fourth card; one had returned the cards which were therefore presumed lost in the post. Two non-respondents were dissatisfied with the system: one felt that the cost of return postage should not be borne by his health authority and the other was too under-resourced and busy to make other than 'positive' returns. The remaining non-respondents have been sent a reminder letter.

Table 2 summarises total reports of cases and those in which research workers have confirmed satisfactory follow-up. Of cases reported to date, only 1 has been 'double reported' within the BPSU system and 9 have been 'parallel reported' with other methods of ascertainment.

New conditions and changes of menu

The XLAED survey was a 3 month one so that it no longer appears on the report card and has been replaced by childhood onset diabetes, a study which is being piloted among South West Health Region paediatricians only.

Other conditions for which application has been made to the Unit's Scientific Advisory Committee for inclusion on the card are: accidental drowning and near-drowning; meningococcal septicaemia; atlanto-axial dislocation in Down's syndrome; Lowe's syndrome; recurrent or persistent hyperinsulinaemic hypoglycaemia. These proposals are currently under review.

Comment

The BPSU is a new venture and has got off to a good start. Investigators have been pleased by the response and so far there have been no serious complaints from the reporting paediatricians. There have been a few 'teething problems' due to the anonymity of the reports which has meant delay in recognising some of the double reporting. In the surveillance of rare diseases, however, over- rather than under-reporting is to be welcomed.

The response to the invitation to submit suggestions for future studies has been as encouraging as the response rate to return of the cards. This first report has been able to provide only limited figures on case numbers because of the inevitable delay in follow-up. Corrected figures will appear in the next and ensuing quarterly reports and we hope to include brief contributions from the investigators.

Reporting rates by region (averaged over the 3 months) ranged from 100% of consultants in East Anglia to 82% of those in Mersey. The proportion of cards which were returned within 2 weeks, increased over the 3 months from 59% to 70% to 75%. This is encouraging because some studies may require rapid case ascertainment in order, for example, to facilitate collection of pathological specimens.

Cases reported

The scheme has caused substantial improvement in ascertainment of some of the conditions for which there was an existing surveillance scheme, notably AIDS in childhood, Kawasaki disease and X-linked anhydrotic ectodermal dysplasia (XLAED).

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BRITISH PAEDIATRIC SURVEILLANCE UNIT : SECOND QUARTERLY SUMMARY REPORT

This report updates that of the first 3 months (CDR 86/46) and presents the results of the second 3 months' mailings (Oct-Dec 1986). During this period the reportable conditions were the same as in the first quarter except that X-linked anhydrotic ectodermal dysplasia was replaced by diabetes. This was a 3 month pilot survey with reporting restricted to consultants in the South Western region only.

The return rates of cards mailed in October, November and December were 84% (689/820), 85% (692/810) and 81% (648/804), 90, 74 and 37 days after the mailings respectively.

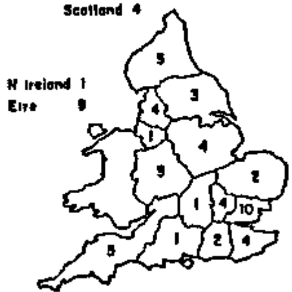
Cases reported and comment

The table summarises total reports of cases (corrected for errors and double reports), including unconfirmed cases for whom clinical information has not yet been received and, in brackets, those in whom the research workers have confirmed the diagnosis on follow-up.

Table Cases reported to BPSU June-November 1986

Condition	First quarter total	Second quarter Reporting month Sept	Oct	Nov	Total	Total first six months
AIDS in childhood	10(10)	3 (2)	3 (-)	2 (-)	8 (2)	18 (12)
Neonatal herpes	7 (2)	- (-)	2 (-)	3 (1)	5 (1)	12 (3)
Reye's syndrome	15(10)	1 (1)	4 (3)	8 (1)	13 (5)	28 (15)
Kawasaki disease	31(27)	11(10)	11 (8)	12 (7)	34(25)	65 (52)
Haemolytic uraemic syndrome	20(10)	7 (5)	4 (2)	1 (1)	12 (8)	32'(18)
Haemorrhagic shock encephalopathy syndrome	2 (1)	1 (1)	- (-)	1 (1)	2 (2)	4 (3)
Subacute sclerosing panencephalitis	8 (8)	4 (1)	5 (5)	3 (2)	12 (8)	20 (16)
X-linked anhydrotic ectodermal dysplasia	13 (2)	N/A	N/A	N/A	N/A	13 (2)
Diabetes	N/A	7 (7)	8 (8)	5 (4)	20(19)	20 (19)
All	106(70)	34(27)	37(26)	35(17)	106(70)	212(140)

The overall number of cases of the reportable conditions in the second quarter (106) was identical to that in the first. There was a slight decrease in reports of haemolytic uraemic syndrome (20 to 12) which is consistent with the seasonal distribution of this disorder. Although the first quarter's figures for SSPE and for AIDS were artificially high because of the inclusion of 'past' cases (see CDR 86/46), the number of SSPE cases increased in the second quarter from 8 to 12, and for AIDS decreased only slightly, from 10 to 8.

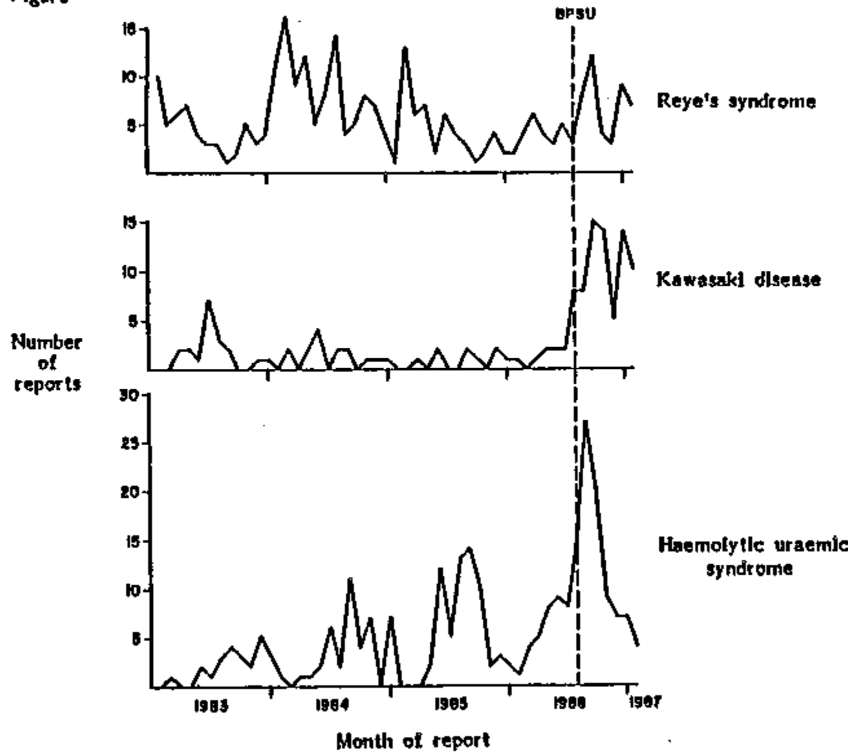


The figure illustrates the substantial increase in reporting of haemolytic uraemic syndrome (HUS), Reye's syndrome (RS) and Kawasaki disease (KD) which has occurred since the introduction of the BPSU scheme. This has been particularly notable for KD; the map shows the regional distribution of reports ranging from 0 from Wales to 10 from North East Thames. There has been no corresponding increase in reporting of haemorrhagic shock encephalopathy syndrome. Only 4 reports of this condition have been received, compared with 8 between June and November 1985 and with 3 in 1986 before the BPSU scheme started.

The feasibility of conducting a regional survey of diabetes has been confirmed by the results of a pilot study in the South Western region.

Lowe syndrome - a rare X-linked genetic disorder, is to be added to the list of conditions in the January mailing. This will be a two year study.

Figure



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Addendum to Table 1: RESPONSE TO MAILING

	June	July	Aug	Sept	Oct	Nov
Date of mailing:	16/7/86	5/8/86	8/9/86	8/10/86	5/11/86	14/12/86
Total cards sent:	824	821	820	820	810	804
Total cards returned:	603	681	706	689	700	685
Cards returned as %age of total sent:	73.2%	83.0%	86.1%	84.0%	86.4%	85.2%
Total cases reported:	47	22	35	34	37	34
Cases reported as %age of cards returned:	7.8%	3.2%	5.0%	5.0%	5.3%	5.0%
Cases reported as %age of total sent:	5.7%	2.7%	4.3%	4.2%	4.6%	4.2%

(correct at 18/3/87)

Addendum to Table 2: CASES REPORTED

Condition	Confirmed cases						
	June	July	Aug	Sept	Oct	Nov	Total
AIDS	10 [1]	-	-	2	-	-	12
Herpes	4 [2]	-	1	-	-	1	6
Reye's	5	1	3	1	2	2	14
Kawasaki	8	7	13	10	9	10	57
HUS	1	2	4	4	2	1	14
HSES	1	-	-	-	-	-	1
SSPE	4 [2]	-	1	1	3	2	11
XLAED	8* [1]	-	5*	n/a	n/a	n/a	13*
Diabetes [3]	n/a	n/a	n/a	7	8	4	19

(correct at 18/3/87)

* Includes unconfirmed cases
[1] Month 1 figures included all cases known to respondents
[2] Month 1 figures included cases seen in past 12 months
[3] Survey covering South Western Regional Health Authority only
n/a Study not in progress in this month