British Paediatric Association

Report from the British Paediatric Surveillance Unit

This is the second report on the activities of the British Paediatric Surveillance Unit (BPSU) to be published in this journal. The first described the background, methodology, and results of the first 12 months of operation (July 1986–June 1987).¹ This report summarises the results of mailings between July 1986 and January 1988.

Cases reported

The table shows, for each of the conditions, the total number of reports received in months 1–19, and the proportion of cases which on detailed follow up (as at 7 April 1988) satisfied the diagnostic criteria. The cases not confirmed include duplicates, reporting errors, and patients who either did not strictly meet the studies' diagnostic criteria or who had their diagnosis revised as a result of further investigation. The last two categories were particularly prevalent among reports of Reye's syndrome and of paediatric AIDS. Among the former nearly half of the cases reported with onsets in the surveillance year

1 August 1986–31 July 1987 had a later diagnostic revision, most frequently to one of the inborn errors of metabolism that mimic Reye's syndrome, such as fatty acid oxidation defects or urea cycle disorders. In the AIDS/HIV illness category, children were reported who had both serological evidence of HIV infection and were symptomatic but whose clinical condition did not meet the United States Centers for Disease Control case definition for paediatric AIDS.²

These observations illustrate the need for meticulous and often detailed follow up of case reports. Surveillance of very rare disorders, unlike that of common ones such as measles or pertussis, requires a high order of sensitivity and specificity of case ascertainment and case definition if accurate epidemiological inferences are to be drawn. Follow up is often demanding of the time of the reporting clinician: the success of the BPSU and the acquisition of knowledge about the rare disorders shown in the table is attributable to the goodwill and cooperation of British Paediatric Association (BPA) and

Table BPSU: reports received July 1986–Jan 1988 (months 1–19)

Condition	Total reports	Outcome of follow up by investigator		
		Outcome known		Not yet known‡
		Report confirmed* (% of known)	Report not confirmed†	
AIDS in childhood	60	20 (40)§	30	10
Neonatal herpes	46	24 (63)	14	8
Reye's syndrome	80	35 (57)	26	19
Kawasaki disease	190	155 (86)	26	9
Haemolytic uraemic syndrome	98	74 (91)	7	17
Haemorrhagic shock				
encephalopathy syndrome	27	14 (74)	5	8
Subacute sclerosing				
panencephalitis	50	43 (86)	7	0
Lowe's syndrome	19	9 (64)	5	5

*Meets case criteria.
†Reporting error, duplicate, revised diagnosis.
‡Still being followed up by the investigator at 7 April 1988.
§Seventeen of these, as well as two cases known to the Communicable Disease Surveillance Centre before BPSU reporting began, were included in *HIV infection in the UK:2*. The three exclusions were: one Irish case; one information incomplete; one report received too late for inclusion.
||Includes 14 cases who are HIV positive (10 symptomatic) but who do not meet the case definition for AIDS.

Irish Paediatric Association (IPA) members with the scheme, for which the Unit and the investigators are extremely grateful.

Workload of respondents

Sensitive to retaining this goodwill, the BPSU conducted an analysis of workload as measured by the number of cases (all conditions) reported by each respondent for the period July 1986 to September 1987.

About two thirds of respondents had reported no cases, so their BPSU time commitment was restricted to ticking 'nothing to report' on the monthly card (the success of the scheme of course depends on the nil returns). Two thirds of those who did report notified only one case. The highest number, eight cases, was reported by only one paediatrician. These figures suggest that most respondents were not excessively burdened by the BPSU.

New conditions

The Unit will, however, continue to monitor workload and respondents' comments in 1988 because further reportable conditions have been added this year, including (for one year only) diabetes, which has engendered a relatively high rate of positive returns. The other new conditions are: haemorrhagic disease of the newborn; galactosaemia; drowning and near-miss drowning, rheumatic fever; and congenital rubella. The last two were retrospective studies each on the report card for one month only. Lowe's syndrome reporting was discontinued from April 1988 when that study was completed.

Response rate

The response rate (proportion of cards returned by 90 days after each mailing) rose steadily from an average of 81% for the first three mailings in 1986 to 90% for the last three in 1987. An average of over 90% for the whole period was achieved by respondents in 10 regions: seven in England, one in Scotland, and Northern Ireland, and Wales. The lowest rate, 77%, was from the Republic of Ireland.

Comment

The BPSU has progressed from the phase of being an experimental venture to that of consolidation. The operation is running smoothly with regular updating of the mailing list, monitoring of response rates and identification of, and contact with the few persistent non-respondents. A recent local effort to increase response rates from the Republic of Ireland has been most successful. The Unit has provided help and advice in the design of protocols and of proformas for data collection for which research workers have expressed appreciation. Several respondents have also commented on the value of the summary protocol cards as an educational aid and stimulus to heightened diagnostic awareness of these disorders.

All but one of the 12 spaces on the report card were filled by April 1988 and the Scientific Advisory Committee had continued to receive further applications. If accepted, these studies will be deferred until 1989 when space on the card will again be available. In view of the demand from research workers, measures to determine paediatricians' views on the optimum number of reportable conditions are currently in progress.

Funding of the BPSU for three years from 1989 has recently been secured by a generous donation from Children Nationwide. Thus the early promise of the Unit in facilitating research in rare childhood disorders continues to be realised. This success is entirely due to the enthusiasm and support of BPA and IPA members.

References

- ¹ Hall SM, Glickman M. The British Paediatric Surveillance Unit. *Arch Dis Child* 1988;63:344-6.
- ² PHLS Communicable Disease Surveillance Centre and Communicable Diseases (Scotland) Unit. HIV Infection in the UK:2. *Journal of Infection* 1988;17:71-82.

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