National Reyes Syndrome Foundation UK

TRUSTEES REPORT
EXTRACTS – 30th SEPTEMBER 1992

Significant events

1. Enzymological studies into the aetiology of Reye's Syndrome

During the year the trustees have continued to fund the project "Enzymological studies into the aetiology of Reye's syndrome – the role of aspirin", which is being undertaken over a period of three years at the Queen's University of Belfast. Due to staff difficulties the progress of this project has been interrupted, but following the recent appointment of a replacement member of staff the project should now continue.

2. Surveillance of Reye's Syndrome

The publication of information by PHLS Communicable Disease Surveillance Centre indicating that in the surveillance year 1990/91 a total of 25 notifications of Reye's syndrome were received (*see table below).* The report comments inter-alia "It is of concern that two patients in 1990/91 had pre-admission exposure to aspirin; none were reported with aspirin ingestion in 1989/90. There may be a need for a renewed campaign to raise parental awareness of the dangers of aspirin given to children with feverish illness."

3. Aspirin Product Labelling

The Foundation had sought assurances from the Committee on Safety of Medicines regarding aspirin product labelling, once the United Kingdom becomes a full member of the European Community on 1st January 1993.

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APPENDIX
BRITISH PAEDIATRIC SURVEILLANCE UNIT
6th ANNUAL REPORT

Reye's syndrome

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

Results 1990/91

A total 25 reports of Reye syndrome were received in the surveillance year 1990/91 (1 August 1990 – 31 July 1991). Of these, 12 cases had their diagnosis subsequently revised to a specific alternative. The *Table below* shows annual total reports for previous years 1981/82 – 1990/91 with the breakdown of revised and non-revised cases. There was one patient in 1990/91 in whom there were atypical clinical features, but no alternative diagnosis was reached; this indeterminate case has been included in the non-revised group in *table 2 (not reproduced here)*

Non Revised cases

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

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

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

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

Specific investigations for IMD's were undertaken in five of seven patients with information. All five had urine organic acids assayed; four patients had both plasma and urine amino acids measured but only in one child was urinary orotic acid investigated. In the remaining two cases no investigations were undertaken: one child

died suddenly soon after admission and the other made a rapid and complete recovery within a few days.

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

Revised diagnosis cases

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

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

Comment

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

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

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Table – *Reye Syndrome Surveillance*

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

[+] Detailed information not available for one case

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