National Reyes Syndrome Foundation UK

TRUSTEES REPORT
EXTRACTS – 30th SEPTEMBER 1993

Significant events

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

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 ended 1994 at the Queen's University of Belfast. Following the appointment of a new member of staff the project is now making progress.

2. Pilot study into Medium Chain Acyl-CoA Dehydrogenase Deficiency:

During the year the Foundation commenced part funding of a pilot study into medium chain Acyl-CoA dehydrogenase deficiency (MCAD) – a Reye-like illness, which is being undertaken at Birmingham Children's Hospital.

3. Reye's Syndrome in the British Isles – report for 1990/91 and the first decade of surveillance:

1993 has seen the publication of the PHLS Communicable Disease Surveillance Centre report 'Reye's Syndrome in the British Isles; report for 1990/91 and the first decade of surveillance' which was circulated as part of the 1993 newsletter. The report states inter-alia that during the *ten* years ended 1990/91 a total of 497 cases of Reye's syndrome have been reported since surveillance began; of these 115 cases (23%) have had their diagnosis subsequently revised. *(see table below)*

It was with sincere regret that the trustees learnt that Dr Susan Hall was to relinquish her responsibility for the surveillance scheme in September 1993. However they were pleased to learn that the work of surveillance will continue under the direction of Dr Michael Catchpole, a consultant epidemiologist at CDSC. Lisa Newton, a scientist at CDSC who has been running the day to day work of the surveillance scheme for the past three years, will continue to manage it.

4. Current Problems in Pharmacovigilance:

In February 1993 The Committee on Safety of Medicines and the Medicines Control Agency in their publication 'Current Problems in Pharmacovigilance' issued a reminder entitled "Reye's Syndrome and aspirin in children". The statement stressed that "parents may still be unaware of the risk associated with the use of aspirin in children. Doctors and pharmacists should remind parents not to give aspirin to children. Nurses and health visitors have an equally important role in putting this message across".

5. Medical and Scientific Advisory Board - appointments

The trustees welcome the appointment of Prof. A Aynsley-Green, MA, DPhil, MB BS, FRCP, FRCP (Edin) and Mr. Charles Pascall, MB BS, FRCS to the Foundation's Medical and Scientific Advisory Board

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APPENDIX
BRITISH PAEDIATRIC SURVEILLANCE UNIT
7th ANNUAL REPORT

Reye's syndrome

Reye's syndrome (RS) surveillance has been included in the BPSU reporting system since June 1986. Prior to this, ascertainment was via a joint BPA-CDSC venture which involved "passive" ascertainment. A total of 521 cases of RS have been reported since surveillance began in 1981/82. Of these, 121 (23%) have had their diagnosis subsequently revised (*see table below)*

Reports received for surveillance year ended 31st July 1992

Twenty four initial reports of Reye's syndrome were received in 1991/92. At the time of writing further information had been received for 21 of the patients.

Patients whose diagnosis was not revised (15 cases)

Twelve (80%) reports came from England; the remainder were from Northern Ireland (two) and the Republic of Ireland (one). Reports were highest during the spring months (February – April); seven (47%) patients had their onset during this period. There were six males and nine females. The mean (SD) and median ages were 23.3 (29.8) months and 16.5 months respectively (range 1.6 – 10 years).

Six patients survived apparently normal and six died, giving a case fatality rate of 40% which is slightly higher than that seen in 1990/91 (38%). Two cases survived with neurological sequelae; one child was freported to be "stiff and unresponsive"; the other recovered with recurrent myoclonic seizures and poor visual and social response. The outcome for one patient was unknown.

Nine children were reported to have received pre-admission medication: three had been given paracetamol; four, antibiotics, and one, oral rehydration therapy. One seven month old infant had recently been treated with half tablets of adult "Disprin" over a two week period, although none had been given in the five days preceding hospital admission.

Three patients were reported with a significant past medical or family history. One five month old infant had suffered two previous possible encephalopathic episodes during the month before admission; another had "developmental delay and growth retardation associated with a chromosome abnormality". The third patient had a cousin who had died suddenly and unexpectedly during infancy.

Investigations for inherited metabolic disorders (IMD's) were undertaken in seven of the fifteen patients. Five had plasma amino acids measured, six urine amino acids and four urine organic acids. Among the three with a significant history only one was fully investigated for an IMD.

Patients whose diagnosis was revised (six cases)

Six children (40%) had their diagnosis subsequently revised, a trend in keeping with previous years (*see table below).* Five were to an inherited metabolic disorder; MCAD (three cases); unspecified disorder of fatty oxidation (one) and isolated gluco-corticoid deficiency with no adrenoleukodystrophy (one). The mean (SD) and median ages of these cases were 24.9 (35.0) months and 13.6 months respectively.

The remaining revision, in a thirteen year old female, was to hyperammonaemic encephalopathy associated with a complication of total parenteral nutrition.

Comment

Following the peak of 81 cases of RS in 1983/84, annual totals have shown a gradual decline and for the last three years of surveillance have remained constant.

The median age of patients, which since 1988/89 has been under ten months, increased to 16.5 months in 1991/92. With such small numbers this may be a chance variation or it may represent awareness among paediatricians of IMD,s in very young infants presenting with a Reye-like illness (they would therefore not report the patient if the alternative diagnosis was made quickly). However, it is clear that not all such patients are being investigated; three children in 1991/92 were reported to have had no metabolic investigations undertaken and all were under three years of age. A further two with histories suggestive of an IMD were not comprehensively investigated.

Pre-admission exposure to aspirin was reported in one child this year (two children were reported with aspirin ingestion in 1990/91) and once again emphasises the need to maintain parental awareness of the dangers of giving aspirin to children with feverish illnesses.

Although the epidemiology of RS has changed little in the past three years, its surveillance will continue: no public health action which results in the decline of a disease should result in complacency about surveillance and any upsurge in association with major influenza epidemic and warning public memory about the risks of aspirin requires early detection. The surveillance proforma will be simplified from 1993/94 onwards with the cessation of the survey of investigation for IMD's.

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Table – *Reye Syndrome Surveillance*

12 month period (August-July)	Total reports British Isles	Non-revised reports	Deaths No. (CFR%)	Revised diagnosis (IMD)
1981/82	47	40	26 (65)	7
1982/83	69	59	33 (56)	10
1983/84	93	81	36 (44)	12
1984/85	64	56	32 (57)	8
1985/86	53	40	22 (55)	13
1986/87	47	26	13 (50)	21
1987/88	44	32	19 (59)	12
1988/89	31[+]	18	9 (50)	12
1989/90	24[+]	15	7 (47)	8
1990/91	25 ..497	13	5 (38)	12..115
1991/92	24[*]	15	6 (40)	6
TOTAL	521	395	208 (53)	121

[+] Detailed information not available for one case

[*] Follow-up not yet received for three cases.

CFR: Case fatality rate

Dr. S. Hall, Ms L Newton
PHLS Communicable Disease Surveillance Centre

(NB from 1st September 1993 Dr M Catchpole (also based at PHLS CDSC) will take over from Dr S Hall)