



**TRUSTEE REPORT
EXTRACTS – 30 SEPTEMBER 1990**

Significant events

1. Medium-Chain Acyl-Coenzyme A Dehydrogenase Deficiency:

The disclosure that research into Reye-like illnesses had produced new information concerning a previously unrecognised metabolite (chemical) in medium chain acyl-coenzyme A dehydrogenase deficiency (MCADD).

2. Surveillance of Reye's Syndrome:

The publication of information indicating that in the previous year (1989) "the annual number of cases of Reye's syndrome in Britain had dropped to 27, half the figure for 1986 (*see table below*). More important, perhaps only two per cent of the children admitted to hospital with feverish illnesses had been given aspirin by their parents." Nevertheless the warning has been expressed in a recent medical article that "many parents seem unaware of the reason that they should NOT give their children aspirin: warning labels and public education measures should continue."

Whilst recognising the improvement in the situation, the trustees take note of the warning and will give urgent consideration to further practical measures that can be undertaken by the Foundation to inform the public of the danger.



APPENDIX
BRITISH PAEDIATRIC SURVEILLANCE UNIT
4th ANNUAL REPORT
1989

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 the *table below*.

Twenty-eight reports were received in the surveillance year 1988/89. 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, propionic 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 1987/88). 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 1987/88 and 19 in 1985/86. 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 1988/89 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 August 1989 and 31 March 1990, in spite of an influenza epidemic in the British Isles larger than that in any winter since 1976.



Table

Reye's syndrome surveillance

**REYE'S SYNDROME IN THE UNITED KINGDOM
AND REPUBLIC OF IRELAND
CASES WITH FOLLOW-UP REPORTED BETWEEN 1 AUGUST 1981 and 31 JULY 1989**

| 12 month period (August-July) | Total reports British Isles | Classified as Reye's syndrome No. | Rate ⁺ | Revised diagnosis |
|----------------------------------|--------------------------------|---|-------------------|----------------------|
| 1981/82 | 39 | 32 | 0.25 | 7 (18%) |
| 1982/83 | 60 | 50 | 0.39 | 10 (17%) |
| 1983/84 | 90 | 79 | 0.61 | 11 (12%) |
| 1984/85 | 61 | 53 | 0.41 | 8 (13%) |
| 1985/86 | 50 | 37 | 0.29 | 13 (26%) |
| 1986/87 | 47 | 26 | 0.20 | 21 (45%) |
| 1987/88 | 44 | 32 | 0.25 | 12 (27%) |
| 1988/89 | 28* | 19 | 0.16 | 8 (29%) |
| TOTAL | 419 | 328 | | 90 |

+ per 100,000 < 16 years of age

* detailed information not available for one case

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