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Cystic Fibrosis in the Leeds Region: Incidence and Life Expectancy

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Incidence

A prospective survey of the incidence of cystic fibrosis among the 3 million population served by the Leeds Regional Hospital Board (L.R.H.B.) was begun in 1952 by all the paediatricians concerned. New births from January 1, 1952, to December 31, 1962, were the subjects of the study, and every effort was made to discover and record patients with cystic fibrosis who evolved from this group (Table I).

During the 11 years 132 patients with cystic fibrosis were confirmed among the 546,764 births; an incidence of 1/4142 or 0·241 per 1000.

TABLE I

Number of Births 1952-1962 and Number With Cystic Fibrosis

Year	Births in L.R.H.B. Area	No. with Cystic Fibrosis
1952	46,866	5
1953	47,632	10
1954	45,971	12
1955	45,776	8
1956	48,211	8
1957	50,413	16
1958	49,825	15
1959	49,824	10
1960	52,388	20
1961	53,817	17
1962	56,041	11
1952-1962	546,764	132

If the area is considered in three parts, located in the three Ridings of Yorkshire, a relatively lower incidence is found in the West Riding portion, which may indicate less complete ascertainment and suggest that the higher figure of 1/3000 births prevailing in the East and North Ridings is more accurate (Table II).

The figure of 1 in 3000 will inevitably be an underestimate, since only cases in which the diagnosis was certain are included, and a few patients born after 1960 may yet be recognized to be affected.

TABLE II

Regional Variation in Incidence

	Births 1952-1962	No. with Cystic Fibrosis
Part of West Riding in L.R.H.B. area	417,739	87 (1/4800)
Part of North Riding in L.R.H.B. area	30,393	10 (1/3000)
Whole of East Riding in L.R.H.B. area	98,632	35 (1/2800)
Total	546,764	132

Survival

The most impressive data relate to the high mortality during the first year of life when 56 patients died (44 in the first 6 months). Thereafter, survivors diminished relatively slowly with 5, 5, 3, and 0 dying in the subsequent pre-school years. By July 1966, 75 of the 132 children had died, and of the 57 survivors 15 are aged 10 years or more. All require close medical supervision.

A life-table is shown in Table III. The first column shows the ages at which the diagnosis was confirmed. In many the diagnosis was made when the children were already seriously ill and so a life-table in which children were entered at age of diagnosis will overestimate mortality. If ascertainment was complete, then all patients could be entered at birth, as in column 5, and the life-table derived as shown in column 7. This will underestimate the true mortality since it is likely that the patients not ascertained will, on the whole, be those more severely affected. Of the 132 children in this series, 12 presented with meconium ileus, though only 8 required operation.

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TABLE III

Life Expectancy

1	2	3	4	5	6	7	8
	No. Entering Series During Period	No. in Series Alive at End of Survey	No. Dying During Period	No. Present in Series at Start of Period if All are Entered at Birth	No. at Risk	Mortality per 1000 in Period Shown	Proportion of 1000 Alive at Start of Each Period
Days							
0	9	0	1	132	131·5	8	1000
7	7	0	5	131	128·5	39	992
Months							
1	6	0	6	126	123·0	46	953
2	13	0	8	120	116·0	63	907
3	37	0	21	112	101·5	175	844
6	26	0	15	91	83·5	120	669
Years							
1	16	0	5	76	73·5	37	549
2	8	1	5	71	68·0	38	512
3	2	3	3	65	62·0	23	474
4	5	9	0	59	54·5	0	451
5	1	13	1	50	43·0	10	451
6	1	4	1	36	33·5	13	441
7	1	6	0	31	28·0	0	428
8	0	5	2	25	21·5	40	428
9	0	1	0	18	17·5	0	388
10	0	2	1	17	15·5	25	388

TABLE IV

Population Surveys

Authors	Incidence
Steinberg and Brown (1960), Ohio, U.S.A.	0·25-0·29/1000
Selander (1962), Sweden	0·11-0·13/1000
Kramm, Crane, Sirken, and Brown (1962), Massachusetts, New Hampshire, and Vermont, U.S.A.	0·42+/1000
Danks, Allan, and Anderson (1965), Melbourne, Australia	0·41/1000

Comment

Early estimates using post-mortem figures gave a high incidence, for example 1 in 600 (Andersen and Hodges, 1946), perhaps because of selective referral. Estimates based on hospital admissions related to child population served give probably better estimates but with a wide range of error, for example 0·1 to 1·0 per 1000 (Carter, 1952). These estimates draw attention to the importance of the condition in the community.

More accurate results have come from population surveys and some of these are listed in Table IV.

Our figure of 0·34/1000 for East and North Ridings is close to, though a little lower than, that of the last two surveys listed above.

The above account has been collated by R. J. Pugh and J. D. Pickup from details supplied by the members of the Leeds Regional Paediatric Club.

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