British paediatrics

Working party on cystic fibrosis

The British Paediatric Association (BPA) has recently published the report of a working party set up in 1982 to assess the advantages and disadvantages of regional centres for cystic fibrosis.

The report quotes several papers published over the last four years, which indicate considerable differences in survival between different countries and different centres. In England and Wales as a whole 80% of children with cystic fibrosis survive to the age of 9 years, whereas in a special clinic in Birmingham 78% survive to 16 years. In Denmark the National Cystic Fibrosis Centre has an 80% survival rate to the age of 12 compared with a survival rate of 80% to age 10 for the country as a whole. Among the best results are those from Australia where, for example, the special clinic in Melbourne reports an 80% survival to the age of 20.

The working party carried out a survey of the whole of the United Kingdom by questionnaire to paediatricians, of whom 99% responded. Members of the British Thoracic Association were also asked to report on their adult patients with cystic fibrosis, but these returns are not yet complete. Data were collected for all live patients and those who have died since 1977, a total of 4557 patients. There are 16 centres in the UK that treat more than 50 patients each, and 1800 patients (46.5%) are currently on the records of these centres. Although figures are not quoted, the working party found that survival rates are better at large centres than in the country as a whole.

In addition to improved survival rates other possible advantages of cystic fibrosis centres are clinical and psychological benefits to the patients, additional experience and expertise for the staff, and better opportunities for research from access to larger numbers of patients. The possible disadvantages are the inconvenience and expense of travel when centres are remote from the patients' homes and the limited experience and interest in managing patients with cystic fibrosis in local hospitals. Furthermore, when emergency treatment is required at

the local hospital the staff will be unfamiliar with the patients.

The working party supports the principle of specialised centres for cystic fibrosis, which has been accepted jointly by the World Health Organisation and the International Cystic Fibrosis Association. However, it recommends that local considerations should determine whether patients should receive all their care at a cystic fibrosis centre, which would be possible in large conurbations, or be managed jointly between a local paediatrician with an interest in cystic fibrosis and the regional centre. In the latter case day to day care would be the responsibility of the local paediatrician, perhaps with visits by clinicians from the centre, and annual assessments could be carried out by the centre.

It is proposed that there should be one centre with from 50 to 100 patients in most regions, and the following minimum staffing levels are suggested for a clinic with 50 patients and three to four inpatient beds:

- One consultant paediatrician with a major commitment to cystic fibrosis (three to four sessions);
- One other senior doctor in support (two sessions);
- Junior medical staff (at least five sessions);
- Physiotherapists (two whole time);
- Nursing sister/coordinator (one whole time);
- Laboratory technician (one whole time);
- Dietitian (two-three sessions);
- Social worker (three sessions);
- Secretary (two sessions).

An adult physician should work closely with the paediatricians at the centre, sharing joint clinics for adolescents, taking over patients when they become adults, and providing support for colleagues in adult medicine in the region.

The working party finally recommends that cystic fibrosis centres should cooperate with each other to pool data, conduct clinical trials, and collaborate in research projects.

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