Conference report

Sixteenth annual meeting of the European Working Group for Cystic Fibrosis

The 16th annual meeting of the European Working Group for Cystic Fibrosis was held in Prague in June 1989. There were 400 delegates from Europe, the United States, Canada, and Australia. Keynote lectures reviewed the immunology of *Pseudomonas aeruginosa* infection, the evaluation of antibiotic treatment in such infections, the importance of gastrointestinal function in cystic fibrosis, heart-lung transplantation, and recent advances in the identification of the cystic fibrosis gene. Spoken paper and poster sessions devoted to specific topics were also held.

A major session of the conference related to the role of heart-lung transplantation in cystic fibrosis. It was reported from the Brompton Hospital that 56 such transplants had been carried out in the United Kingdom and that over 40 recipients were alive; at the time of the conference the longest survivor had had the transplantation 3·75 years before. The major criteria used to match donors and recipients were ABO blood groups, the size of the recipient's thorax, and whether he or she had been infected with cytomegalovirus. There appeared to be few absolute contraindications to surgery apart from extensive thoracic or pleural surgery in the past and the presence of aspergillomas, though even these were not absolute contraindications. The United Kingdom figures included patients transplanted in London and Cambridge, where a one year survival rate of 80% had been achieved. Early reports from France suggest that a similar success rate is likely there.

Dr Rosalind Smyth reported the experience in Cambridge, where 14 patients had had heart-lung transplants. Their criteria for transplantation included an FEV_1 below 30% predicted, a 12 minute walking distance of under 500 m, ventilatory failure, right ventricular failure, and frequent or prolonged admissions with a failure to gain weight after exacerbations. Of the 14 patients receiving a heart-lung transplant, 12 were alive three to 42 months after surgery. Two patients had died, one from fungal sepsis and one from bleeding and the adult respiratory distress syndrome. Of 23 patients accepted for transplantation 11 had died while on the waiting list. After transplantation there were on average two rejection episodes per patient in the first three months and about 0·2 rejection episodes after one year. With regard to infection, cytomegalovirus appeared to be important, with five episodes of either reactivation or reinfection. Herpes simplex, *Pneumocystis carinii* infection, and invasive aspergillosis also occurred. An attempt to measure the quality of life after transplantation in a small number of patients by means of the Nottingham health profile showed a substantial improvement after transplantation in terms of physical disability and lack of energy.

A large proportion of the meeting related to the problem of lung destruction secondary to chronic *Pseudomonas aeruginosa* infection. The consensus view was that bacterial products probably caused only minor local injury, the immediate inflammatory and chronic immune response of the host being the main causes of lung destruction. There was increasing interest in the development of methods to detect and quantify the injury process as a means of monitoring lung destruction and the effectiveness of antimicrobial treatment. Professor Niels Høiby (Copenhagen) reviewed the immunology of *Pseudomonas aeruginosa* infection in cystic fibrosis, and described the pattern of the host antibody response to various cell wall constituents and exoproducts of *P aeruginosa*. It is possible that early in the course of infection such antibodies have a protective effect, but that later the formation of immune complexes may trigger a greater host response and in chronic infection lead to more substantial lung destruction. Relating to this important topic, studies reported from Hanover by Dr B Tummler and from London by Pauline Nicholson indicated that there is very little patient to patient transmission of *P aeruginosa* at these centres, despite a high degree of sharing of facilities by patients in the latter case. Dr Tummler also reported that despite phenotypic variation in antibiotic resistance little genotypic change in *P aeruginosa* occurs within an individual colonised patient, as shown by genomic finger printing.

Much interest was shown in the reports relating to the basic molecular genetics of cystic fibrosis. It was clear from reports presented at the meeting in June 1989 that the molecular geneticists were close to identifying the gene responsible for cystic fibrosis. There was evidence from several centres that the severity and pattern of disease may vary and that this may reflect different defects in the cystic fibrosis gene. This expectation of a breakthrough was confirmed in September with the report of the structure of the cystic

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fibrosis gene and a common deletion.

This gathering, although called the European Working Group for Cystic Fibrosis, provided a valuable forum for a wide range of research workers from around the world. The combination of keynote review lectures and presentation of recent research provided delegates with an up to date review of clinical approaches in cystic fibrosis as well as details of current research. The detailed abstracts will be published in the Proceedings of Charles University.

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EDITOR'S NOTE

Thorax will from time to time publish reports on conferences such as this meeting of the European Working Group for Cystic Fibrosis. We would particularly aim to cover meetings on specialised topics which would be of interest to *Thorax* readers but where most readers would not be attending the meeting and the main contents of the meeting would not be published in abstract or other form in a widely distributed journal. Readers who are organising or attending such a meeting and think that the proceedings would be of interest to *Thorax* readers are invited to drop the editor a line with details of the meeting. The topic of the meeting should be of general interest; non-promotional meetings on the development of individual drugs, for example, would not be considered appropriate.