Overview of autopsy for CF patients: The organs weight analysis

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The 187 autopsy reports (77 males and 110 females) from 1940s to 2000s with the age distribution from 5 days to 50 years were examined in this study. We focused on the weight of the right lung, left lung, heart and liver with respect to age, body weight, height and body mass index (BMI). The Organs weight was analyzed using the statistical analysis with respect to the independent variables. The characteristics associated with the way lung weight varies with age, weight and height were quite similar for both autopsied male and female CF patients. The same also holds true for the heart and the liver weight characteristics as they relate to age, weight and height. The weight of the left lung was heavier than or equal to the weight of right lung for 20% of the female CF patients and 23% for the male patients. The polynomial curve fittings indicated larger magnitudes and variations in heart and lung weights for autopsied CF patients of all ages for both genders. The magnitude and variations in liver weight were not significantly different for CF patients and those who do not have CF with the exception of females. Our results indicated that the weight of the right and left lung in CF patients tends to increase more rapidly than the normal distribution for those not afflicted with CF in all variables of age, weight, height and BMI. This result correlates with the mucus accumulation into the bronchi and inflammation of the proximal airways. It is not easy to explain the heavier weight of the heart in CF patients, but heart condition might be associated with heart dilatation which is a secondary CFTR related disease. The heavier weight of the liver for CF patients was associated with the accumulation of fatty deposits.

Patient satisfaction in a District General Hospital cystic fibrosis service

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Background: The urban/rural Clyde area has 73600 children, from which our Cystic Fibrosis team provide a service to 27 children (25 families) aged 0–16 years, in two outpatient clinics and one in-patient facility.

Aim: To establish whether we are providing a service that meets the satisfaction of our patients, whilst upholding professional standards of care.

Methodology: A questionnaire (A) focusing on all aspects of care was devised for all 25 families. Also, 8 children over 12 years received an extra questionnaire (B).

Results: Questionnaire response rates were (A) 17 (68%) (B) 6 (75%). The quality of the service provided was rated highly. Key themes emerged; in-patient care, annual review process and psychology provision.

Inpatient stay was subject to mixed response; the majority indicated satisfaction with regard to management and treatment received. It was felt difficult to explain the variations in lung weight and height. One patient was felt to have reached end stage. The results indicated that the overall attendance was high. The majority had attended at least 12 appointments.

Conclusions: We have introduced joint CF and diabetes clinics every 3 months this allows both CF and diabetes review so is not seen as an extra clinic by patients. Patients are offered at least twice yearly appointments. Patients are now offered joint CF and diabetes clinics. 88% of patients felt they would benefit from a clinic where they could see a CF nurse and diettian with a specialist interest in diabetes. Only 22% had completed outpatient blood sugar (BS) diaries. 61% of respondents adjust their insulin to the amount of food they eat, only 22% had received training on how to do this.

Preparing practitioners to meet the challenges of a specialist service. A joint venture between Bristol regional CF centres and University West of England

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The Government recognises cystic fibrosis as a specialist service. National guidelines and standards recommend patients are cared for in specialist centres by skilled professionals or within a shared care framework affiliated to a CF centre. Commissioning specialised services aims to ensure fair access to clinically effective first class care across the country. Inherent within this plan is the need to achieve best outcomes, maintain clinical competence, sustain the training of specialist staff and make best use of scarce resources including clinical expertise. Only one university accredited CF course has been available in the UK located in London. In light of fulfilling the role of a regional centre a dedicated CF interprofessional course was commissioned to increase accessibility to specialist training in the South West. The course is a collaborative venture between the expertise of the CF centres and academic accreditation provided by the university at degree and masters level. Expert clinicians and scientists deliver the taught component whilst the university offers academic rigour and quality control. The course covers all aspects of paediatric and adult management delivered by 3 two day taught seminars over six months and supported by online learning. It also provides individual mentorship and flexibility to address specific clinical practice requirements.

Feedback from the first cohort of nurses, physiotherapists, a pharmacist and psychologist was extremely positive. Students reported increased knowledge and understanding of CF management and welcomed the opportunity to apply new skills into practice.

The 2009 course is full and applications have been received for 2010.