Co-Morbidity in a Cystic Fibrosis Population Attending a Regional Clinic.

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Abstract
Pulmonary disease remains the major cause of morbidity in patients with cystic fibrosis (CF). However, of 115 patients attending a regional CF clinic we noted 16 cases (14%) with co-morbid conditions. Of this group, 4 of 115 patients (3.5%) had renal problems including both structural and functional defects and 4 (3.5%) had neurological disorders, 3 of which were types of epilepsy. Notably, 3 of 115 patients (2.6%) had different forms of neoplasia, all of which required surgical and/or chemotherapeutic intervention. There is now increasing evidence of the association between digestive tract malignancy and CF, which further complicates management of these already complex cases.

Introduction
Cystic fibrosis (CF) is a chronic, progressive disease exhibiting multiple complications related to viscous mucus, malabsorption and infection. We noted a significant number of CF patients attending a regional clinic had additional co-morbid conditions, thus adding to their complex management.

Methods
We reviewed all 115 patients attending CF clinics at the Mid-Western Regional Hospital - 42 adults and 73 children. We reviewed each case individually, by retrospective chart review, assessing the prevalence of additional medical diagnoses. As this was a study of co-morbidity, we excluded any of the recognised complications of CF including diabetes mellitus, CF liver disease, arthropathy and sinus disease.

Results
We found 16 cases (14%) with co-morbid conditions (Table 1). Most notably, 3 (2.6%) had neoplasia, 4 (3.5%) had renal problems and 4 (3.5%) had neurological disorders. Of the neoplasia group, a 19-year-old male had been diagnosed with poorly differentiated adenocarcinoma of the ileocaecal valve, grade T3N2Mx. Management involved surgical resection followed by chemotherapy. The second neoplasia case was of stage 2 testicular teratoma in a 24-year-old male. Treatment was with radical orchidectomy and chemotherapy without bleomycin or radiation. This patient achieved full remission with < 1% risk of relapse. The third case of neoplasia was in a 4-year-old girl diagnosed with ovarian teratoma after routine abdominal ultrasound. Management involved left oophorectomy and histology revealed a mature cystic teratoma.

The 4 cases of renal co-morbidity included a 16-year-old female with pelvi-ureteric junction obstruction. There were 2 separate cases of duplex kidney, one with an obstructed upper pole secondary to ureterocele. The fourth case was a 13-year-old male who presented with Henoch Schonlein Purpura requiring dialysis for renal failure. He was subsequently diagnosed with IgA nephropathy after a renal biopsy. The 4 neurology cases included 3 of 115 patients (2.6%) with epilepsy (Table 1). The fourth case was a female with a history of right middle cerebral artery infarct in the neonatal period resulting in left sided hemiplegia.

Discussion
Approximately 3.1% of the Irish population have a diagnosis of cancer, compared to 2.6% of the CF population in this report. To date there have been few reports of malignant tumours in patients with CF. This finding is attributed to the fact that the majority of patients with CF die at a young age as a result of chronic pulmonary infection with impaired nutrition and resistance to infection. However, as in our case with adenocarcinoma, malignancy is beginning to impact on prognosis and life expectancy. A recent study has also demonstrated an increased risk of digestive tract cancers among adult CF patients, particularly in the small bowel, colon and biliary tract. This increased risk was even more pronounced in those patients who had an organ transplant. In our report 0.86% of the CF population had a large intestinal malignancy compared to a prevalence rate of 0.45% for the Irish population.

Recognised renal complications of CF include nephrolithiasis due to hyperoxaluria, hyperuricosuria and lack of colonization with Oxalobacter formigenes. There is also accumulating evidence that vigorous use of intravenous aminoglycosides in CF management is a significant cause of acute renal failure. Chloride channelopathies, defined as disorders caused by mutations in genes encoding for ion channels, can cause diseases as diverse as CF to forms of epilepsy. However, there is no known association between epilepsy and CF. Furthermore, the rate of epilepsy in our study group was 0.82%, which is similar to the national prevalence rate of 0.9%. CF, a complex autosomal recessive disorder, is the most common lethal genetic disorder in Ireland with a carrier frequency of 1 in 19. Pulmonary disease remains the major cause of morbidity in patients with CF. However of 115 patients attending a regional CF clinic, a significant number have co-morbid conditions including neoplasia. This has an additional impact on their complex management. Physicians, patients and families must work together to maintain an optimistic but aggressive approach to life and treatment.
References

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