349* Twenty years of care for CF adults in Czech Republic

L. Fila[1], P. Jakubec[2], J. Polak[3], R. Bittenglova[4], I. Binkova[5], J. Musil[1]. [1]*Pulmonary Department, University Hospital Motol, Prague, Czech Republic;* [2]*Department of Respiratory Medicine, University Hospital, Olomouc, Czech Republic;* [3]*Pulmonary Department, University Hospital, Hradec Kralove, Czech Republic;* [4]*Pulmonary Department, University Hospital, Plzen, Czech Republic;* [5]*Clinic of Pulmonary Diseases and Tuberculosis, Faculty Hospital, Brno, Czech Republic*

Background: first CF patient in Czech republic was diagnosed in 1946. Improving care led to improved survival and first adult CF centre was established in 1987. Today, five adult CF centres exist in Czech republic.
Methods: searching in databases and patient files for adult CF patients, CFTR mutations, pulmonary and extrapulmonary complications, and selected modalities of treatment.
Results: totally 184 adult CF patients (100M, 84F) were identified in databases. Fifty-four patients died or were lost from evidence. Current group of CF adults consists of 130 patients (68M, 62F) aged 27.5±6.3 years. 50.8% of them are colonized with *P. aeruginosa* and 35.4% with *B. cepacia*. 31.5% of patients have normal FEV1, 24.6% mild (FEV1 60–79% pred.), 23.1% moderate (FEV1 45–59% pred.) and 20.8% severe (FEV1 < 45% pred.) obstruction. 81.5% of patients have exocrine pancreatic insufficiency and 18.5% is malnourished (BMI < 18.5 kg/m^2). 23.8% of patients have liver disease, 29.2% diabetes mellitus or impaired glucose tolerance and 41.4% bone disease. 72.3% of patients recieve rhDNase, 29.2% tobramycin or colistin inhalations, 14.6% azithromycin, 40.0% nutritional support and 6.2% LTOT. Six patients underwent lung transplantation and four are on waiting list.
Conclusion: complex care based on european standards and given in specialized centres is the only possibility for further improvement of survival and quality of life of CF patients.
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350 Cystic fibrosis (CF) and the risk of pancreatic cancer (PDAC)

A. Lowenfels[1], P. Maisonneuve[2], B. Marshall[3]. [1]*Surgery, New York Medical College, Valhalla, NY, USA;* [2]*Epidemiology Unit, European Institute of Oncology, Milan, Italy;* [3]*Cystic Fibrosis Foundation, Bethesda, MD, USA*

Introduction and Aim: Previous reports have detected an excess risk of pancreas cancer in patients with pancreatitis. In CF, pathologic changes have been noted in all the digestive organs including the pancreas; at birth the pancreas of CF patients already demonstrates marked histological abnormalities. The aim of this study was to determine the overall risk of PDAC in patients with CF.
Method: We obtained information about patients with both CF and PDAC from the USA and Canadian Cystic Fibrosis Patient Registries, from previous literature reports, and by querying CF physicians and pathologists in the USA, Canada, and Europe. The expected number of patients with PDAC for non-CF persons was estimated from population-based data adjusted for age and sex. The risk of PDAC in patients with CF was determined by comparing the observed with the expected number of cases (standard incidence ratio-SIR).
Results: Over a 20 year period (1985–2004) we obtained reports of 9 patients (5 male, 4 female) with both CF and PDAC. Median age at diagnosis of PDAC was 35 years (range 18–58 years). Four patients were homozygous for the Delta F508 mutation, one patient was heterozygous. 5 of the 9 patients had mild or moderate lung disease. The expected number of PDAC patients in the non-CF population adjusted for age and sex was 1.6, yielding an SIR or risk ratio of 5.6 (95% CI = 4.1–17.1).
Conclusion: Based on data from North America and Europe, patients with CF have an increased risk of PDAC which closely resembles the increased risk for other digestive tract tumors. However compared to other causes of mortality in CF patients, the absolute risk of PDAC in this population is negligible.
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351* Employment status of cystic fibrosis adults – a 10 year improving picture

E. Gunn[1,2], A. Lyon[1], S. Henman[1], C.S. Haworth[1], D. Bilton[1]. [1]*Adult Cystic Fibrosis Centre, Papworth Hospital, Cambridge, United Kingdom;* [2]*CF Registry, Cystic Fibrosis Trust, Bromley, United Kingdom*

Advances in management have lead to increased survival in CF. It is important to establish whether this trend is associated with increased levels of employment. The aim of this study was to examine employment status at a UK adult CF clinic between 1996 and 2006.
Annual data is collected prospectively on all people with CF attending Papworth Hospital. The clinic population in 1996 was 79 and in 2006 182. All subjects were >16 years of age. The Mean FEV1% pred (±sd) in 1996 was 51.6% (±27.3) and 63.9% (±26.4)in 2006. Mean BMI (±sd) kg/m^2 in 1996 was 20.9 (±3.67) kg/m^2 and 21.5 (±2.86) kg/m^2 in 2006.
In 1996, 30 (38%) patients were in employment, 26 (33%) in fulltime employment and 4 (5%) in part-time employment. 40 (50%) were in full-time education, 6 (8%) were unemployed, and 3 (4%) were full-time homemakers.
In 2006, 96 (53%) patients were in employment, 68 (38%) in fulltime employment and 28 (15%) part-time employment. 32 (18%) were in full-time education, 46 (25%)were unemployed, and 8 (4%) were fulltime homemakers.
We compared national data for 2004 from the UK CF Registry. 2827 subjects >16 years of age had employment data. 1350 (48%) were in employment, 1002 (36%) fulltime employment, 348 (12%) part-time employment. 514 (18%)were in fulltime education, 789 (28%) unemployed, and 174 (6%) fulltime homemakers.
In conclusion we have confirmed that significantly more patients are in employment in 2006 compared to 1996, largely through an increase in part-time employment. Adolescents and adults with cystic fibrosis should receive career advice to prepare them for future employment.

352 Incidence and evolution of nasal polyposis in cystic fibrosis children and adolescents

S.A. Weber, G.F. Ferrari. *OFT/ORL/CCP, FMB-UNESP, Botucatu, São Paulo, Brazil*

Introduction: Cystic fibrosis is frequently associated to nasal polyposis.
Aims: To study the incidence of nasal polyposis in children and adolescents with cystic fibrosis, its possible association to age, gender, clinical manifestations, genotype and chlorid level, and its evolution with topical corticosteroid therapy.
Methods: Clinical symptoms (pulmonary, pancreatic insufficiency, undernutrition, nasal obstruction), sweat chlorid level, genotype and nasal endoscopic findings were studied in 23 cystic fibrosis patients. Polyposis was treated with topical corticosteroid during 6 months, followed by a second nasal endoscopy.
Results: Nasal polyposis was found in 39.1% of the patients (five bilateral, four unilateral), recurrent pneumonia in 82.6%, pancreatic insufficiency in 87% and malnutrition in 74%. No association was seen between nasal polyposis and sweat chlorid level, genotype, clinical sings of severity and nasal symptoms. Nasal polyps were found in children older than six years. Seven patients improved polyposis with corticosteroid therapy, six showed complete involution.
Conclusion: The study showed a high incidence of nasal polyposis in older children, even in the absence of clinical nasal symptoms. Patients with CF and nasal polyposis span the entire range of clinical severity. Topical corticosteroid therapy showed good results. An interaction among pediatricians and otolaryngologist is necessary.
Key-words: polyposis, cystic fibrosis, diagnosis, endoscopy, therapy.