232 Gastrointestinal cancer survey at the Stockholm CF Center

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Introduction: The average age of the adult CF population is constantly increasing and might imply a rise in number of malignancy cases in the adult CF population. The previous studies on cancer incidence in CF adults emphasize a higher risk of gastrointestinal (GI) cancer and to a lower extent other forms of cancer and malignancies.
The aim of the study was to survey the number of cancer cases as well as other malignancies in nontransplanted adults at the Stockholm CF center.
Results: The retrospective case analysis performed in 136 CF adult patients reveals 4 cases of GI cancer within the last decades. Two patients (at diagnosis 39 and 65 years old, respectively) have had high differentiated adenocarcinoma of the colon, which was surgically removed. One of these patients was later lung transplanted and up to now is doing well. The other one died but not due to cancer. The third patient (age 44) had a cholangiocarcinoma and underwent radical surgery. Nevertheless she died 7 months later due further progress of the tumour. The last patient (age 49 years old) has a suspect GIST (gastrointestinal stromal tumor). Its histological verification is in progress. One case of acute lymphatic leukemia was also observed. This patient (age 27) is doing well after allogenic stem cell transplantation.
Conclusions: The data from the Stockholms CF center demonstrates an increased cancer risk in the CF adult population in concert with the data from others. Since the previous studies are from 1990s it would be of interest to disclose whether the cancer incidence is increasing further due to the aging of the CF population. Time for GI cancer screening program?

233 A real-life glucose tolerance test in children to screen for CFRD

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Introduction and Aim: A yearly oral glucose tolerance test (OGTT) is advised to diagnose CF-related diabetes (CFRD), from the age of 10 yrs and up. This test does not reflect a real-life situation, is disliked by patients and not fully adhered to. The aim of this study is to test the acceptance and results of a real-life glucose tolerance test (RLGTT) and compare positive outcomes with the OGTT, from Jan 2010 to Dec 2011.
Materials and Methods: For the yearly check up 33 children with CF of 10–17 yrs, not already diagnosed with CFRD, were admitted. The carbohydrates of the meal are registered according to the Dutch Food Composition Table 2006. The RLGTT consists of a 4 hour fasting period followed by eating a minimum of 1.25 g carbohydrates per kg bodyweight in a regular meal, with a maximum of 1.75 g/kg or 75 g carbohydrates. A glucose sample is taken before and 2 hours after the meal. A 2 hour glucose of >9 mmol/L after 1.50–1.75 g/kg or >8 mmol/L after 1.25–1.50 g/kg is considered abnormal. Stricter cut-off values in comparison to OGTT were set, because of the use of a meal instead of glucose.
Results: We did 47 tests in 33 patients. All patients easily accepted the test in this design. An impaired fasting glycemia was found 2 times. In 7/47 tests (7/33 patients) results were abnormal with a mean postprandial glucose of 11.0 mmol/L (8.2 to 11.1 mmol/L). In 6 patients an OGTT pointed out CFRD. They all started with insulin once a day.
Conclusion: The RLGTT is a good predictor of CFRD, is easily accepted and implemented in regular care. An abnormal RLGTT should be followed by intensive monitoring of blood glucose.

234 Glucose abnormalities in children with cystic fibrosis (CF)

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Objectives: CF-related diabetes (CFRD) is related to worsening pulmonary function and increased mortality and its prevalence is significantly high beyond the first decade of life. Current guidelines recommend annual OGTT as screening method of CFRD for all CF patients ⩾10 years. We evaluated glucose abnormalities among patients ⩽10 years regularly attending the CF Regional Centre of Naples.
Methods: From 2004 to 2010 67 patients ⩽10 years (30 F) without CFRD were screened for glucose abnormalities by fasting 2 h–1.75g/kg OGTT and classified as NGT, IGT, INDET, CFRD according to current guidelines [1]. The inclusion criteria were: pancreatic insufficiency, no acute pulmonary exacerbation and no current steroid therapy. Mean age at the time of OGTT was 80.4 months (range 30–118), median age 83 months. We identified 18 NGT (26.8%), 4 INDET (6%), 11 IGT (16.4%), 4 CFRD (6%). 30 patients (44.7%) resulted with glucose levels ⩾140 mg/dl in the middle of OGTT.
Conclusions: More CF patients ⩽10 years had glucose abnormalities than previous reports. Our data suggest that also patients ⩽10 years, have to be early screened for CFRD. Larger prospective studies need to define the prognostic role of INDET and IGT status, as well as to assess the prognostic role of abnormal glucose tolerance (AGT – glucose level ⩾140 mg/dl in the middle of OGTT) in CF patients.

Reference(s)

[1] Moran A, et al. Clinical care guidelines for cystic fibrosis-related diabetes. Diabetes Care 2010; 33(12): 2697.

235 Using HbA1c and random blood glucose to screen for cystic fibrosis related diabetes (CFRD)

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Introduction: UKCF Trust guidelines recommend all adult CF patients should be screened annually for CFRD by oral glucose tolerance test (OGTT). This is time consuming, inconvenient and disliked by some patients. We have previously shown that using a combination of HbA1c, random glucose (RBG), annual FEV_1 decline and annual weight loss to screen for CFRD may be an effective alternative approach. To validate this in the wider CF population we performed the current study to determine the most accurate criteria for predicting patients with a positive OGTT requiring treatment.
Method: Prospective study of consecutive adult CF patients (not known to have CFRD) attending an annual review assessment between March 2009 and December 2011. All patients had an OGTT. In parallel they had: HbA1c, RBG, and calculation of annual %predicted FEV_1 and weight decline.
Results: Data for 323 patients (mean±SD age = 31±10, 42% female, 76% pancreatic insufficient) were collected. Mean±SD RBG = 5.40±1.6; HbA1c = 5.4±0.6; annual %predicted FEV1 change –0.6%±2.7; annual %weight change +1%±3. OGTT at 120 min 7.8–11.0 mmol/l, n = 43; at 120 min >11, n = 17. By systematic approach the optimal criteria for predicting a positive OGTT was: HbA1c ⩾5.8% (n = 171) and/or RBG >11 mmol/l (n = 6). This provided a test accuracy of 94.1% sensitivity and 49.3% specificity. The one patient missed by these criteria is clinically well and not currently requiring insulin.
Conclusion: This large study confirms the validity of performing an OGTT selectively. The use of two biochemical markers – HbA1c ⩾5.8% and RBG >11 mmol/l – provides an excellent test accuracy for CFRD screening, reducing the need for an OGTT by nearly 50%.