333* Characteristics of visual function in adults with cystic fibrosis

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Objectives: Ocular abnormalities have been observed in CF subjects including: impaired dark adaptation, contrast sensitivity and colour vision. Previous studies are small and have included confounding factors. This study aims to compare visual function (VF) in CF subjects of known vitamin A and diabetic status, with healthy controls.
Methods: Distance and near visual acuity (DVA, NVA), contrast sensitivity (CS), colour vision (CV), dark adaptation (DA) and retinal photographs were recorded from 28 CF subjects (19m, 9f; 27±7.4 years) and matched controls.
Results: CF subjects displayed significantly reduced NVA ($p < 0.05$), CS ($p < 0.005$), DA ($p < 0.005$) and CV ($p < 0.01$) compared to controls (t-test). Considering CF subjects, diabetes ($n = 11$, 45% with diabetic retinopathy) had no significant effect on VF apart from DA ($p < 0.05$). Low vitamin A level (<1.10 μmol/L, $n = 12$) had no significant effect on VF. There was no significant correlation of vitamin A status and DA thresholds ($r = -0.282$) although one deficient subject exhibited a complete absence of the normal rod retinal photoreceptor response. Generally, VF in ΔF508 homozygotes ($n = 12$) appeared to be reduced compared to heterozygotes ($n = 11$) although this failed to reach significance.
Conclusions: VF is adversely affected in CF. CFTR has been detected in a layer of the retina known as the retinal pigment epithelium (RPE). Abnormal ion transport across the RPE is likely to impair normal VF. These results support the hypothesis that abnormal VF is a primary manifestation of CF which is exacerbated by genotype, vitamin A insufficiency and CFRD. These findings highlight the importance of regular eye examinations and education of eye care professionals to the ocular associations of CF.

334* Autonomic neuropathy in cystic fibrosis

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Cardiovascular autonomic neuropathy (CAN) is an important form of diabetic autonomic neuropathy causing increased morbidity and mortality. The prevalence of such diabetic complications in cystic fibrosis (CF) is likely to rise with increasing survival. The aim of this study was to assess the prevalence of CAN in our adult CF population.
Methods: Heart rate variability (HRV) testing during slow respiration, a valsalva manouvere and in standing from supine (measures of parasympathetic function) were performed using ECG monitoring as recommended by American Diabetic Association. Early and definite involvement of CAN was defined as 1 and 2 or more positive tests respectively. Blood pressure (BP) response to standing from supine and isometric exercise were also measured to assess sympathetic function. Vitamin E levels and complications of CF were recorded.
Results: 43 patients were screened, 27 had CF related diabetes (CFRD) (mean age 28), 5 had impaired glucose tolerance (GT) (mean age 28), and 11 had normal GT (mean age 30).
Of the 43, 25 (58%) had definite involvement, 13 (30%) had an early involvement and 5 (11%) had no evidence of CAN. There was no significant response of BP to standing in any group whilst isometric exercise was not found to be reproducible.
Conclusions: Although CAN appears prevalent in CFRD, it was also evident in our non-diabetic CF group. Vitamin E deficiency and chronic liver disease may explain such findings but this was not evident in our population. Autonomic neuropathy in CF warrants further investigation.

No. of +ve tests according to diabetic status

No.of +ve tests	CFRD (27)	CF impaired GT (5)	CF Non-Dm (11)
0 (Normal)	3 (11%)	2 (40%)	0
1 (Early involvement)	6 (22%)	1 (20%)	6 (54%)
2 (Definite)	18 (67%)	2 (40%)	5 (46%)

335 Thyroid dysfunction in newly diagnosed cystic fibrosis patients with pancreatic insufficiency and restitution with enzyme therapy

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Introduction: Thyroid function has not been adequately assessed longitudinally in cystic fibrosis patients as yet.
Aim of the study: To investigate thyroid function in newly diagnosed cystic fibrosis patients, and to follow the natural course of possible thyroid alterations with pancreatic enzyme substitution treatment.
Patients and methods: Fourteen newly diagnosed patients (7male/ 7female, mean age 5 months [range 1–19 months]), all with pancreatic insufficiency, were evaluated regarding their thyroid function at diagnosis of cystic fibrosis and for 6 months ensuing onset of pancreatic enzyme substitution therapy.
Results: All infants displayed normal TSH values in neonatal screening. Eleven out of 14 (80%) had hyperthyrotropinemia, while all had normal freeT4 and freeT3 values upon initiation of enzyme substitution therapy. Within the first month of therapy, TSH values were normalized in 60% of the cohort while the rest demonstrated a persistent TSH elevation for a longer period. At re-evaluation, after 3–6 months, 70% of the patients had normal TSH values while on pancreatic enzyme substitution even though they received no thyroxine treatment. The correction of hyperthyrotropinemia might be partly associated with restitution of defective absorption of various minerals.
Conclusions: Mild TSH elevation, reminiscent of subclinical hypothyroidism, is a frequent finding in newly diagnosed cystic fibrosis patients with pancreatic insufficiency during infancy. TSH elevation resolves in the majority of cases 3–6 months after initiation of enzyme substitution therapy.

336 Assessment of bone mineralisation in cystic fibrosis

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CF patients may develop low bone mineral density (BMD), leading to an increase in bone fragility and susceptibility to fracture. In 2007 the UK CF Trust published its consensus guidelines on Bone Mineralisation in CF. We have audited our current practice against these guidelines. The notes of 47 adult CF patients [24 female, mean (range) age 30 (17–57), mean FEV1 65% predicted (20–115%), mean BMI 22.7(16.9–32.5)] receiving full or shared care at our centre were reviewed. 35 patients (74%) were pancreatic insufficient, 2 were post transplant and 7 were on long-term (>3 months) oral corticosteroids.
14 patients (30%) had ever had a bone densitometry (DEXA) scan, of whom 11 had a scan in the last 3 years. 7 (50%) had their calcium and vitamin D levels measured in the previous 12 months. None of the patients had their CXR routinely reviewed for vertebral fractures.
Of those who had a DEXA scan, 3 were normal (Z score >–1), 3 met the criteria for CF-related low BMD (Z score of <–2) and 4 had a significantly reduced BMD (Z score –1.5-–2). 1 patient had a Z score of –1.4. The results of the 3 remaining scans were not available. Of those with CF-related low BMD the mean (range) of FEV1 was 36% predicted (20–54%) and the mean (range) BMI was 21.4 (19.8–24.3). Amongst these patients, 1 was on bisphosphonate treatment whilst completing a prolonged course of steroids.
This audit demonstrates that many of our patients are not having their bone mineralisation status assessed. Of those assessed, 21% had CF-related low BMD. Although the numbers in this patient group are small, recognition that CF-related low BMD may occur in those with above average clinical status, (as defined by FEV1 and BMI) has re-emphasised the importance of incorporating bone mineralisation measurement into our centre's annual review for all patients.