

### 228 Pulmonary and nutritional outcomes in children with cystic fibrosis diagnosed by meconium ileus in Argentina

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**Introduction:** Early diagnosis in Cystic Fibrosis (CF) still remains a challenge, because it allows the implementation of appropriate therapeutic strategies. It can reach through neonatal screening (NS) an early detection or by meconium ileus a premature clinical manifestation associated with poorer growth.

**Objective:** To compare clinical status and lung function in children with CF diagnosed with meconium ileus and those detected by NS.

**Methods:** We compare two groups: one detected by NS and one diagnosed by meconium ileus (MI) since 1995 (beginning of the program of NS in the state of Buenos Aires). We assessed: Z score Weight/age, Z score Height/age from 6 months up to 6 years of age, Forced Expiratory Volume in 1<sup>st</sup> (FEV<sub>1</sub>). We also registered first isolation of *Pseudomonas aeruginosa* (Pa). Student's t-test was performed to compare the means of the indicators.

**Results:** We included 26 patients in NS group and 31 in MI, all with pancreatic insufficiency. p.PHE508del/p.PHE508del was present in 52% of NS and 41.4% in MI. In MI group were four deaths and one in NS. The NS group showed better nutritional parameters than MI, as seen in the table. We found no significant differences in first isolation of Pa (p=0.55) and FEV<sub>1</sub> (p=0.11) between the two groups.

Table: Nutritional parameters in NS and MI groups

	Z score		1 year*		3 years		6 years	
	6 months*		W/A	H/A	W/A	H/A	W/A	H/A
	W/A	H/A						
NS	-0.53	-0.77	-0.52	-0.62	0.28	0.22	0.27	0.25
MI	-1.63	-1.85	-1.33	-1.77	-0.60	-0.50	-0.62	-0.68

\*p=0.05.

**Conclusion:** Although the results suggest better nutritional status and lung function in the NS group, advances in non surgical techniques as well as nutritional support have achieved better growth parameters within the MI group.

### 229 Prucalopride: a review of the first 12 months of use

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**Objectives:** Constipation is common in CF. In 2010 NICE recommended prucalopride, a selective serotonin receptor agonist, for the treatment of chronic constipation in women who had failed conventional therapy. Patients with severe lung disease were excluded from pivotal trials.

**Methods:** Data were retrospectively collected from patient records. Laxative treatment and admission frequency were compared pre and post prucalopride. Side-effects and patients' perceptions were noted.

**Results:** Four pancreatic-insufficient females with a history of refractory constipation/DIOS were commenced on oral prucalopride (Table 1).

Table 1. Patient demographics

Patient (age [yrs])	Genetics	FEV1 (% predicted)	BMI	Diabetic
1 (28)	ΔF508/ΔF508	59	23.8	No
2 (31)	ΔF508/ΔF508	37	19.8	Yes
3 (38)	ΔF508/ΔF508	33	20.7	Yes
4 (41)	ΔF508/Unknown	27	21.1	Yes

Prucalopride was well tolerated. Patient 1 developed diarrhoea on initiating treatment which spontaneously resolved. Patient 3 reported a headache that resolved within 14 days. Particular benefit was noticed by Patients 1, 3 and 4. Patient 1 has reduced her regular laxative therapy, has not required admission nor any additional laxatives since starting treatment. Patient 2 reported minimal response. Patients 3 and 4 have reduced their use of gastrograffin (Table 2).

Table 2. Treatment and clinical response

Pt	Pre-prucalopride		Prucalopride			
	Regular laxatives	Additional laxatives in last 12 months	Admissions, constipation/ DIOS	Months of prucalopride in 2011	Additional laxatives	Admissions, constipation/ DIOS
1	Docusate Lactulose Senna N-Acetylcysteine	Klean prep ×2 courses (variable dose) Gastrograffin ×10 (3 day course)	2	10	None. Senna discontinued & lactulose dose decreased	0
2	Senna Lactulose Movicol Domperidone	Gastrograffin ×2 (3 day course)	0	6	Picolax ×1	0
3	Senna Movicol Docusate	Gastrograffin ×16 (3 day course)	0	3	Gastrograffin ×2 (3 day course)	0
4	Senna Movicol Docusate	Gastrograffin weekly (3 day course)	0	2	1 dose of gastrograffin weekly	1

In this small number of patients prucalopride has provided symptomatic benefit and reduced the need for additional treatments.

### 230 Review of bowel preparation in adults with cystic fibrosis requiring colonoscopy

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An increased risk of gastrointestinal (GI) tract carcinoma has been reported in the CF population. GI investigation necessitates patients with CF require a colonoscopy procedure. Lack of adequate bowel preparation (prep) and the difficulty in cleansing a "CF bowel" may result in misdiagnosis.

**Objectives:** To review the effectiveness of standard bowel prep versus CF specific bowel prep for colonoscopy.

**Methods:** A retrospective chart audit was performed on CF patients with colonoscopy procedures performed from March 2000 to March 2011. Between March 2000 and October 2009 patients received standard bowel preparation, and from October 2009 to March 2011 a CF specific bowel preparation was introduced.

**Results:** Sixty patients (53% male) had a colonoscopy procedure between March 2000 and March 2011. 55 total procedures were reviewed. Results indicated, standard bowel prep provided effective clear out in 11 patients (32%) and CF specific bowel prep provided effective clear out in 18 patients (86%). A Fisher's Exact statistical analysis demonstrated a significant difference (p < 0.001), favouring effectiveness of the CF specific bowel prep for colonoscopy. Results from colonoscopy procedures identified colonic carcinoma in 3 (5.5%) patients and occurrence of polyps in 14 (25%) patients.

**Conclusions:** Previous reports have highlighted a higher incidence and earlier age of diagnosis of GI carcinoma in CF compared to the general population. The introduction of a CF specific bowel prep has demonstrated a more effective cleanse, to visualise the lower GI tract and assist investigation.

### 231 Helicobacter pylori infection in CF patients

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**Objectives:** The purpose of the work was the assessment of infection with *Helicobacter pylori* (Hp) and adjacent pathology (ulcer, reflux esophagitis, ulcers) in children and young people with CF, compared with those without CF.

**Methods:** Two study groups were considered for a transversal study. Group I included 20 CF patients (9 male and 11 female), mean age 14.3 years and group II – 24 patients (pts) with Hp infection (14 male and 10 female), median age 13.6 years. Selection criteria: presence of gastric symptoms. Quantitative detection of Hp antigen, esophagogastroduodenoscopy with histopathology examination of gastric biopsy were performed. Gastritis were classified according to Sydney criteria; Savary and Miller criteria were used for reflux esophagitis. All CF pts had physiotherapy lying in special anti reflux positions.

**Results:** Half of CF patients had Hp infection associating chronic gastritis, 80% with nodular lesions. Esophagitis was diagnosed in 7 patients (70%). No ulcer was found in CF patients nor in controls. In controls, 58.3% had Hp infection with nodular gastritis in 13 cases (92.8%). 71.4% of controls presented esophagitis lesions.

**Conclusion:** No significant differences in the incidence of Hp infection in children and young people with CF compared to controls with similar median age at Hp infection and resemblance pathological features. In CF patients esophagitis seems to be mainly linked to Hp infection and less related to physiotherapy position.