Early Decline of Pancreatic Function in Cystic Fibrosis Patients with Class 1 or 2 CFTR Mutations

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BACKGROUND: Most cystic fibrosis (CF) patients develop steatorrhea and require pancreatic enzyme replacement therapy. However, there are few data regarding the decline of exocrine pancreatic function within the first years of life in relation to CF genotype. We assessed the decline of pancreatic function in CF infants carrying class 1 or 2 CFTR mutations who were diagnosed in a neonatal screening program.

MATERIALS AND METHODS: Twenty-eight CF patients were included in the study and 27 completed the study. In all subjects, fecal pancreatic elastase-1 concentrations and fecal fat excretion were scheduled to be determined at diagnosis, at 6 months of age and subsequently at 6-month intervals.

RESULTS: In all CF patients, fecal pancreatic elastase-1 concentrations of the first assay after diagnosis (3 to 4 months of age) were lower than the cut-off level for normals of <200 mug/g stool. Steatorrhea was found in 81.5% of these subjects. At the age of 6 months, all screened CF subjects had fecal pancreatic elastase-1 concentrations <100 mug/g and at the age of 12 months all were pancreatic insufficient. At that time, having proved pancreatic insufficiency in all studied subjects, we stopped the scheduled further assessment.

CONCLUSION: CF patients require careful monitoring of pancreatic status from diagnosis onwards. In patients carrying class 1 or 2 CFTR mutations, pancreatic insufficiency develops in the first months of life. The proper assessment of pancreatic insufficiency and intestinal malabsorption is crucial for the early introduction of pancreatic enzymes.

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