Evaluation of fecal pancreatic elastase 1 as a measure of pancreatic exocrine function in children with cystic fibrosis

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Pancreatic elastase 1 (EL-1) is a specific human protease synthesised by the acinar cells. It is stable, unaffected by exogenous pancreatic enzyme treatment, and correlates well with stimulated pancreatic function tests. We report our experience of EL-1 measurements in 142 patients from a large cystic fibrosis (CF) clinic. The median patient age was 7.7 years (range 0.1-20.8 years), 93 were homozygous and 38 heterozygous for DeltaF508, and 11 had other or unidentified mutations. There were 85 non-CF control subjects. Seven were pancreatic sufficient (PS). The median (quartile 1-quartile 3) fecal El-1 of the 135 pancreatic insufficient (PI) patients was 10 µg/g stool (2.5-33); of the 7 PS patients, 698 µg/g stool (400.5-824.5), and of the non-CF controls, 615 µg/g stool (420-773). Using the Mann-Whitney U test, there was a statistically signi-ficant difference for fecal EL-1 activity between the PS and PI patients ($P = 0.0001$) and the PI and control group ($P < 0.0001$), but not between the control and PS groups ($P = 0.63$). Median (quartile 1-quartile 3) fecal EL-1 in the pancreatic insufficient DeltaF508 homo-zygotes was 10 µg/g stool (2-33), and in the heterozygotes 12 µg/g stool (4-39) (not significant, $P = 0.62$). We now use fecal EL-1 as evidence of PI in screened CF infants (reliable over the age of 2 weeks); in older CF patients at diagnosis; for confirming the need for pancreatic enzymes in patients referred to the clinic already taking enzymes; for annual monitoring of PS patients to detect the onset of PI; and as supporting evidence when excluding the diagnosis of CF in patients attending the pediatric gastroenterology clinic. The low values in the first 2 weeks in some normal and premature infants, and the persisting normal values in PS infants, make the fecal EL-1 test unsuitable for neonatal CF screening.

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