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## 50 Years Ago in *The Journal of Pediatrics*

# Exocrine Pancreas Insufficiency: Supplementing Our Way to Success

Haworth JC. Malabsorption and Growth Failure Due to Intestinal Enterokinase Deficiency. J Pediatr 1971;78:481-90.

The article by Haworth et al described 1 patient's clinical course, which seemed consistent with pancreatic insufficiency, but with normal constitution of duodenal fluid (pancreatic enzymes). Growth velocity was normal until a dietary transition from maternal breast milk to cow's milk at 5 months of age. Symptoms (vomiting, irritability, loose stools) improved on a partially hydrolyzed formula. However, diet expansion led to a return of symptoms, including poor appetite, abdominal distension, muscle wasting, loose stools, delayed bone age, hypoalbuminemia, anemia, and hepatomegaly (from protein malnutrition). A thorough workup included abdominal radiographs, liver biopsy, and jejunal biopsies along with dietary modifications and several hospital admissions. The diagnosis was made when no trypsin or chymotrypsin activity was detected on duodenal aspirate, indicating enterokinase deficiency. A diet supplemented with pancreatic extract lead to the cessation of diarrhea, resolution of hepatomegaly, improved mood, and increased growth.

Since first being described in 1969, only 13 cases of enterokinase deficiency have been reported. Consistent symptoms included vomiting, diarrhea, hypoproteinemia, and failure to thrive, but these reported patients became symptomatic shortly after birth.[1-3] It is likely that he improved with a partially hydrolyzed formula owing to higher content of medium-chain triacylglycerol oil, which does not depend on pancreatic digestion for absorption. Diagnosis has become swifter with the advancement of fecal testing for elastase, which has become more widespread with improved sensitivity and specificity and with the ease of endoscopy.[4,5] The prognosis is good with appropriate enzyme supplementation, which has been the most impactful advancement.[1,3] Pancreatic enzyme replacement therapy now comes in a variety of delivery mechanisms. Delayed release capsules minimally effect gastric transit and are meticulously titrated to fit the needs of patients.[5,6] Further innovations include Relizorb, which uses a single-use cartridge form of lipase connected in line with existing enteral tube feed sets to facilitate digestion.

In conclusion, enterokinase deficiency is a rare disease under the umbrella of exocrine pancreatic insufficiency. Improved diagnostic techniques, including highly specific genetic testing, has expanded our list of known causes well beyond cystic fibrosis. Early diagnosis allows the early establishment of appropriate treatment to prevent severe, long-term effects of malnutrition. Advancements in pancreatic enzyme supplementation allow for these patients to grow and develop without significant symptoms of disease or side effects of medication.

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