158 *Celiac-sprue Syndrome: Etiologic Considerations.* MICHAEL I. COHEN, HELEN MCNAMARA, OLGA BLUMENFELD and IRWIN M. ARIAS, Dept. of Ped., Montefiore Hosp. and Depts. of Ped., Biochem. and Med., Albert Einstein Coll. of Med., Bronx, N.Y. (introduced by Laurence Finberg).

Gamma-glutamyl transpeptidase (γGTP), an intestinal mucosal enzyme, hydrolyzes γ glutamic carboxyl amide bonds. Subfractions of gluten were obtained by a modification of the hydrolytic scheme of Frazer. A high glutamic acid concentration characterizes gluten (38%) with enrichment of this amino acid in the residue following hydrolysis (68–70%). A γ-carboxyl peptide configuration appears in 2 of the subfractions. An *in vitro* assay system utilizing the concept of enzyme inhibition showed the specific hydrolytic action of γGTP to be the γ-glutamic acid site in gluten and its subfractions. When γ-glutamyl beta naphtylamine served as substrate, with guinea pig intestinal mucosal scrapings supplying the enzyme, several of the subfractions acted as potential inhibitor compounds.

Intestinal γGTP was determined in jejunal biopsies from 10 adult human subjects, 4 controls and 6 patients with small intestinal pathology including 4 with celiac-sprue. Enzyme activity was found to be approximately half that of the normal in 2 sprue patients in clinical remission (normal fecal fat balance, normal morphological appearance of the jejunal biopsy and normal intestinal maltase activity). In the other 2 sprue patients the subjects were not in remission and the γGTP levels were even lower. These preliminary data suggest γGTP may be required to hydrolyze the γ glutamyl moieties in gluten and that a deficiency of this enzyme may result in the clinical entity of celiac-sprue. Whether there is a genetic-molecular basis for celiac-sprue, an acquired deficiency of enzyme, or both, will be answered when additional data become available.

159 *Cyst(e)ine Dependency Presenting with Growth Failure and Steatorrhea.* ALBERT JOHN SCHNEIDER, Upstate Med. Center, Syracuse, N.Y. (introduced by Wm. H. Bergstrom).

Initial study of a first-born male Caucasian child with classical celiac syndrome failed to reveal the cause of his growth failure: birth wt. 3.0 kg, age 12 weeks 3.3 kg. At age 16 weeks the child was admitted to hospital with an acute bout of diarrhea and dehydration. On fat-free intake, steatorrhea disappeared but hyperphagia and watery diarrhea with sugar-free stools persisted. Weight gain was less than normal for age. Following oral feeding of Amigen with glucose, alopecia occurred, and blood levels of methionine, ½ cystine, and taurine were >115, <1.5, and 3.0 μM/dl respectively. Homocystine in blood and urine, and cystathionine in blood, were undetectable by ion-exchange chromatography. Cystathionine was excreted in urine at less than 15 mg daily, dietary methionine averaging 6.5 μM daily. These findings suggested a block in the conversion of methionine to homocysteine.

The essential nature of cystine for growth was documented in a series of feeding experiments. Three basic diets were used, providing the percentage distribution of calories as shown in the table. Caloric intake averaged 150 cal/kg/d. Weight gain was calculated from the average weight of the day before, day of, and day after a diet change. Diets were fed at least 6 days without significant change.

	Basic diet			
	A		B	
%Cal: prot/fat/CHO	15/0/85		15/40/45	
Diet supplement:	None	L-cyst.	None	L-cyst.
Average wt. gain (gm/d)	18.7	35.0	6.3	17.5

	C		
%Cal: prot/fat/CHO	5/10/85		
Diet supplement:	None	L-cyst.	DL-homocyst.
Average wt. gain (gm/d)	25.4	45.4	42.4

Catch-up growth in body length and head circumference has occurred with dietary supplementation. Mental development seems normal but physical weakness persists.

160 *Malabsorption and Growth Failure Due to Intestinal Enterokinase Deficiency.* JAMES C. HAWORTH and BETH GOURLEY, Winnipeg Children's Hosp. and Dept. of Ped., Univ. of Manibota, Canada (introduced by H. Medovy).

Recently HADORN *et al.* [Lancet *i:* 812, 1969] published the first description of a patient with intestinal enterokinase deficiency. A similar abnormality has been defined in a boy who showed growth failure and chronic diarrhea from 5 months of age. From 1–3 years of age he grew fairly well when receiving Nutramigen. When seen at 6 ½ years of age he appeared very malnourished, had a distended abdomen, and a greatly enlarged liver. He had hypoproteinemia, a bone age of 3 years, and mild steatorrhea. Biopsy of the liver showed generalized fatty infiltration. Cystic fibrosis was excluded. The jejunum showed a normal histological appearance. Various dietar yregimes during the next 1 ½ years did not improve his condition. When he was aged 8 years his duodenal juice was found to contain no tryptic activity. He was treated with pancreatin and rapidly became asymptomatic, the liver became normal in size and he showed a greatly increased rate of growth. A secretin-pancreozymin test for pancreatic function was performed recently when he was 12 years old. The duodenal juice was normal in volume and bicarbonate content. Activity of amylase was normal; lipase activity was low. Trypsin and chymotrypsin activities were very low. The *in vitro* addition of either crystalline enterokinase or of a homogenate of duodenal mucosa from another subject to the duodenal juice of the patient resulted in rapid appearance of normal activities of trypsin and chymotrypsin. It was concluded that in the patient, the pancreatic proteolytic enzyme precursors were secreted normally into the duodenum but could not be activated due to deficiency of enterokinase.

161 *Hydrogen Peroxide Hemolysis: A Test for Biliary Obstruction.* BERTRAM H. LUBIN, ROBERT L. BAEHNER, ELIAS SCHWARTZ, STEPHEN B. SHOHET and DAVID G. NATHAN, Children's Hosp. Med. Center, Boston, Mass. (introduced by Charles A. Janeway).

To detect the presence of complete biliary obstruction in the neonatal period, we have utilized the red cell hydrogen peroxide hemolysis (PH) test. This simple procedure is based on the fact that vitamin E, a fat soluble vitamin, is dependent upon bile salts in the intestine for absorption. Red blood cells hemolyze in the presence of hydrogen peroxide when cellular vitamin E is depleted. Hence, in severe biliary obstruc-