

THE JOHNS HOPKINS UNIVERSITY  
SCHOOL OF MEDICINE

*Silverstein  
Development  
of GI  
Lymphatics*

DEPARTMENT OF MEDICINE  
Division of Gastroenterology

Please address reply care of  
THE JOHNS HOPKINS HOSPITAL  
BALTIMORE, MARYLAND 21205

February 12, 1971

Dr. Victor A. McKusick  
Moore Clinic  
Johns Hopkins Hospital

Re: Fourth annual conference on  
clinical delineation of birth  
defects, June, 1971

Dear Dr. McKusick:

I will be pleased to participate in your conference.

You had asked for suggestions as to topics or participants for the area of digestive and absorptive functions of the GI tract such as alterations in the digestive ability of the small intestine mucosa. You are covering this with the disaccharidase deficiencies. Some interests in this area include fetal development and changes in lactase levels after birth as well as the prevalence in adults. There is also some recently accumulating information on peptidase deficiencies and defects in amino acid absorption. Along the lines of absorptive defects, the glucose-galactose malabsorption is an excellent example. In terms of defects in mucosal synthesis,  $\alpha$ -Beta lipoproteinemia is a good example. If you wish I could certainly cover all of these topics. Another area would be familial chloridorrhea. These patients are unable to reabsorb chloride and have a severe diarrhea and electrolyte imbalance. In terms of the pancreas, there are some children with hereditary pancreatitis and some with absence of the pancreas. I think that this could easily be covered by your cystic fibrosis people. The area of the development of the stomach is being explored by Dr. Julius Deren of the University of Pennsylvania. There are no major clinical problems that come to mind, so this area is probably not vital. It could also be covered very briefly in a discussion of the small bowel development.

Dr. Deren is also interested in the small bowel. There are a number of other people who could easily speak on the small bowel, but I think that most of this could be included in the talk on disaccharidases.

As I'm writing this, I wonder if celiac disease should not be mentioned because this certainly seems to be an inherited disorder and the defect is probably present at birth, but does not become clinically

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apparent until the child is about 8 months of age when he begins to consume large amounts of wheat, rye and oats. The current going thoughts are that this might be due to a peptidase deficiency and inadequate digestion of some of the wheat and peptides. This has not been proven but it does make a nice story. Obviously Tom Hendrix or I could talk about this. Dr. Dowd Kowlessar-Jefferson in Philadelphia is extremely interested in the peptide but has been unable to come up with a specific peptidase deficiency at this time.

Pardon my rambling. I hope this is of some help to you.

Sincerely,

A handwritten signature in black ink, appearing to read 'Ted', written over the word 'Sincerely,'.

Theodore M. Bayless, M.D.  
Associate Professor of Medicine

TMB/ert