Classification of Malabsorption Syndrome

Introductory Remarks

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IT IS advisable to classify the malabsorption syndrome into primary and secondary forms. The primary malabsorption syndrome includes several classic entities, celiac disease of childhood and, in the adult, tropical and non-tropical forms of sprue. The primary malabsorption syndrome must be strictly separated from the secondary malabsorption syndrome which is caused by close involvement of the small bowel by such diseases as lymphosarcoma, amyloidosis and jejunoileitis, or resulting from extensive resection of the small bowel, inadvertent gastroileostomy or from liver-pancreatic disease.

The primary malabsorption syndrome, according to our present thinking, is a genetically-transmitted complex metabolic disorder. From the clinical aspect, it is mainly characterized by intestinal malabsorption and hematologic abnormalities. It involves, however, many other metabolic disturbances and affects the metabolism of proteins, carbohydrates, lipids, electrolytes and water and may be hematopoietic, which is not necessarily related to the difficulties in absorption. Profound disturbances in genetically-controlled enzymatic chain reactions seem to affect intestinal absorption mainly in the wall of the gut as well as the metabolic pathways of many factors mentioned above.

Based on the genetic anlage, the malabsorption syndrome may become manifest at various periods of life: in infancy or childhood (as celiac disease) or later in life as tropical or non-tropical sprue. Having produced manifestations in childhood, the disorder may go underground for many years or may never again produce manifestations of malabsorption. As in many other hereditable metabolic errors, there is a steady interplay of the genetic predisposition with environmental influences which may both stimulate and mitigate the manifestations of the disorder. Distress of tropical climates, malnutrition and infections are important environmental factors which may convert a predisposed person into a patient with manifest malabsorption syndrome. Strong emotional factors may play a similar part. Whether wheat or rye gluten represents a dietary triggering mechanism or is more profoundly involved in the basic metabolic defect (pterylglutamate complex) of primary malabsorption remains to be established.

In our climate, we are mostly concerned with one clinical entity of the primary malabsorption syndrome: non-tropical sprue, which will be the main topic of discussion. Reference will be made to the tropical variety of sprue and to the celiac syndrome of childhood.

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