Coeliac Disease

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In the following article I shall attempt to briefly outline the condition known as "Coeliac Disease". The disease is characterized clinically with loose fatty stools, from time to time, with improvement between attacks. Relapses occur and are increasingly severe. The condition progresses to marked malnutrition, a distended abdomen and a peevish, fretful but often precocious child. The stools are never normal; they are large, grayish in colour, frothy and often foul. Growth of the child suffers and is below the normal range.

Many factors have been blamed for this condition. Gee recognized early that some food in the diet was at fault and, because of the fatty stools, it was assumed that fat digestion was at fault. Investigation revealed that bile secretion was normal. Experimental work revealed that protein was well tolerated, fats moderately well, and carbohydrates very very poorly tolerated. This led to the use of high protein diets, often with striking improvement, with remissions occuring as soon as carbohydrate was added beyond very small amounts. The role of the pancreas was investigated with no significant results. And so the controversy raged, with the only factors agreed upon being the value of the protein diet and the uniformly poor prognosis.

Incidence:

It occurs mainly in the age group 1 - 5, but older children and young adults are not immune. With better diagnostic methods the disease is recognized more frequently. It is no respector of economic groups, and males and females are equally affected. More than one case in the same family is found infrequently, so that a familial tendency is not marked.

Etiology:

Despite the amount of research in the past, the understanding of the cause of coeliac disease is as confused as ever. Each hypothesis only leads farther along a confusing path. As previously stated the cause has been hypothesized as (1) a "disturbance of digestion", no cause ventured, by Gee; (2) because of fatty stools, faulty fat digestion was implicated; chemical analysis revealed chiefly split fat; (3) deficiency of bile salts was eliminated as a cause when normal amounts were found in gastric contents; (4) the pH was also found to be normal; (5) "nervous factors" have also been eliminated as a cause; (6) only, concerning abnormalities of pancreatic secretions, has there been any consistency in findings reported, several cases showing abnormal pancreatic secretions; (7) abnormal endocrine function has been shown to be a secondary condition; (8) "constitutional weakness" means nothing more than "cause unknown." (9) "Basic Constitutional Disease" is a non-descript term, allowing a wide field in which new theories may roam. In summary, the cause is still unknow.

Clinical Symptoms:

(1) Diarrhea

Although diarrhea is a classical sign, in some cases constipation has been reported. However, the stools are generally bulky and grayish. Often there is a particular foul odour. The stool is frothy and greasy, with 4 - 5 stools per day being common.

(2) Physiological state

Apathy, irritability and obvious unhappiness of the child are also classical signs. The child is neither interested in food or in normal activities. Under the circumstances can you blame him?

(3) Appetite

The psychological state seems to have a marked influence. Generally the appetite is poor, but often shows strange and peculiar cravings, for example: wall plaster, coal sand. Some lose all desire for food, amounting to complete anorexia with fits of rage at the sight of food. This condition is quickly cured by treatment using proper diet.

(4) Growth and Weight

Failure to grow is a marked feature of the coeliac child. Even with recovery the children often remain frail. There are periods of improvement, then relapse accompanied with weight loss. With proper treatment and vitamin supplements, this phase of the condition has been largely rectified. Emaciation and weakness accompany weight loss. Classically the patient wastes more in the limbs than in the face. Muscular weakness and tenderness is often marked. These are secondary symptoms from poor nutrition.

(5) Abdomen

Classically it is supposed to protrude; many investigators report cases in which this stigma was not marked. It seems to outlast the other symptoms when it occurs, the patient "growing up to his abdomen."

(6) Anemia

This is universally mentioned as a symptom; there may be clear evidence of haemoglobin deficiency, but this is secondary. It seems to be a hypochromic anemia due to failure of absorption of blood-forming elements.

(7) Temperature

A fever may be present, but is is believed due to a secondary infection. Respiratory infections often precipitate coeliac symptoms.

(8) Pain and Vomiting

Abdominal pain is infrequent. There may be cramps and intestinal spasms. Vomiting is very frequent.

(9) Urine

Nitrogen content is high in proportion to protein intake. Acetone is rarely present but uric acid content is high.

(10) Skeletal System

Rickets is a common complication attributed to coeliac disease and calcium deficiency. Osteoporosis is said to be the finding in coeliac disease. There may be delayed ossification, correctable with treatment. There appears, in some cases, to be defective calcium and phosphorus metabolism. With a diet containing adequate fats and vitamin supplements A and D, the condition is cured. Tetany may occur, but with vitamin therapy this is rarely seen.

Other symptoms as photophobia, enlarged heart, haemorrhagic states, and pareses are secondary to the condition of coeliac disease itself.

In most cases the disease has a gradual onset but may occur as a sudden diarrhea or after another acute disease such pertussis, or even after vaccination. The onset varies from vomiting, loss of appetite, and loss of weight, to anorexia and diarrhea. There may be intervals of weeks or months in which the child does fairly well, but there is no overall gain in weight or normal growth.

Pathology

Here, again, there is great disagreement over pathological findings. In general, no changes have been observed in the digestive organs at post mortem except for several cases of pathology in the pancreas. Other reports are contradictory and the reports are so conflicting that a definite statement cannot be made.

A similarly confusing picture is presented concerning bacteriology. For many years at B. infantilis was incriminated, and later B. coli. Nothing was proved concerning their part in the etiology of coeliac disease.

Digestion and Absorption

There have been reports of poor absorption of nitrogen, sulphur, phosphorus, calcium and magnesium. There is a general mineral loss, but calcium absorption is dependent on the general metabolic picture and is absorbed in a proper diet.

Because of fatty stools it was at first assumed that fat digestion was at fault. Analysis showed no unsplit fat present and bile secretions were found to be normal. It was thus eliminated as the basic digestive disturbance.

It was seen by early observers, substantiated by later investigators, that protein was well tolerated, even in large amounts. This led to the use of the high protein, low fat diet, until investigation showed fat metabolism to be normal.

It was observed by Marriott in 1922 that increased sugar in the diet caused an immediate increase in frequency and size of the stools. This he blamed on a substance formed which irritated the intestine, increasing peristalsis.

Investigation showed enzymes for carbohydrate, fat, and protein metabolism to be produced normally, the fault lying in absorption. Glucose given intravenously was well tolerated, but not orally. These patients

are insulin sensitive, with a flat blood sugar curve. Failure of phosphorylation due to vitamin B complex deficiency was blamed for poor absorption. It appears that the invert sugars are better tolerated than dextrorotory sugars, for example: cane sugar. From numerous investigations it appears that intermediate metabolism is normal, the failure of absorption of carbohydrate producing the condition of coeliac disease. Two findings agreed on are: (1) the blood sugar curve is a secondary effect, and is not a test of assimilation. (2) it has been shown clinically that monosaccharides are better absorbed than polysaccharides. This has led to exclusion, or diminution of carbohydrate in the diet of coeliac disease patients.

Vitamin deficiency occurs during a critical attack, but represents only a multiple deficiency state which is reversible under proper treatment.

Digestive Juices and Enzymes:

Study has shown that in general there is no decrease in the digestive juices or enzymes. Bile secretion has been shown to be normal. Deficient pancreatic secretion has been observed, but may have been due to fibrocystic disease of the pancreas, rather than to a true coeliac disease. These two conditions are often confused.

There is little evidence pointing to neurogenic, psychogenic or endocrine factors as a cause of coeliac disease. Emotional disturbances, it is generally agreed, are secondary rather than primary. Allergy, if present, is more likely a coincidence, rather than a cause.

Diagnosis:

The disease occurs at all ages of childhood. There are large, loose stools from time to time with weight loss. There may be improvement between attacks, with subsequent relapse and additional weight loss. Relapses become more severe. There is a subsequent malnutrition with a peevish, fretful, often precocious, child. The abdomen is distended, at first intermittently, then constantly. Stools are never normal, even between attacks, being light gray, frothy, and foul. Constipation may alternate with diarrhea. Gas is passed in large amounts and causes distention. Growth suffers in proportion to the duration of symptoms, the child being well below average height

and weight. Intercurrent infection, especially intestinal, is often a danger. Low blood sugar curves are often present. Normal pancreatic secretions, by duodenal intubation, differentiates the condition from fibrocystic disease. Anemia, avitaminosis, hydrolability, emaciation, personality disturbances, etc., are secondary and of questionable diagnostic value.

Treatment:

This consists of a proper diet for a sufficiently long time. No food may be ingested by the coeliac patient that contains an appreciable amount of carbohydrate other than that found in fruits and to a lesser extent in vegetables and in protein milk (Haas). The basic carbohydrates used are ripe banana and protein milk. Other fruits must be avoided during the period of diarrhea. To this diet, protein in any form and fats. in moderation, are added. Cheese is widely used. Gelatine, honey dates and raisins are used since they are chiefly monosaccharides. Eggs are also given, along with vegetables, when the diarrhea lessens. Fats need not be restricted beyond those given to healthy children. This type of diet must be continued for long periods with added vitamin A and D supplements.

In a coeliac diet, restricted foods must be emphasized. Any cereal grain is strictly forbidden. Sugar is forbidden in candy, etc., and milk other than protein milk. Strictness of diet cannot be overemphasized. The diet must be followed for at least a year. Most cases improve immediately, especially the child's disposition. There should be a cure within 18 months. When cure is obtained there should be no relapse. In milder cases careless use of carbohydrate is tolerated but prolongs the period necessary for cure.

Vitamin B complex and liver extract have been used but are not effective. Antibiotics and sulfonamides are used to control diarrhea. Pancreatic extract has been used to advantage in some cases.

Prognosis:

When treated by a specific carbohydrate diet the prognosis is excellent from the standpoint of mortality, morbidity, and cure. Death is usually due to an intercurrent infection. On proper diet, cure is usually achieved by the end of 18 months. After treatment there is no stunting of growth, no underweight, anemia, oedema, or forms of avitaminosis such as rickets and scurvy.

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