COELIAC DISEASE IN SUDANESE CHILDREN

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INTRODUCTION:

Coeliac disease is a malabsorption syndrome due to gluten intoleronce and is characterized by a state of malnutrition associated with crhonic diarrhoea, steatorrhoea, wasting, abdominal distention and failure of growth.

Samuel Gee (1889) wrote the classical discription of this disorder and in 1950 Dick noted that children with coeliac disease improved when wheat and rye were excluded from the diet. In 1953 Van De Kamer, had shown that the gluten fraction of wheat and rye and specially gliadin, one of its major protiens was harmfull to coeliac patients. It was suggested later that the gluten fraction of barely and possibly oates were also incriminated.

The mucosal changes in coeliac disease are characteristic with loss of the villous pattern and cellular infilteration of the lamina poropria. The mechnaism of pathogeneses of these changes have not been ellucidated, however a direct toxic effect of incompletely digested peptides and immunological reactions to the gut are considered possililities.

Coeliac disease is probably uncommon in the Sudan. The feeding habits are certainly different from the European ways and this may explain partly the low incidence of this syndrome, but constitutional, genetic, and environmontal factors may be involved. Kisra, a local bread made from Dura or Sorgham Vulgaris constitutes a stable diet in many Sudanese homes; however, bread biscuits and other foods made of wheat are also used.

MATERIALS AND METHODS:

The patients presented here represent two types of cases. In two the diagnosis was confirmed by jejunal biopsy and in two thers the diagnosis was based on clincial features, evidence of steatorrhoea and in both a good respence to gluten free diet was obtained.

CASE(1):

A female child of 18 months. She was the last of a family of 5 children. Her parents were first cousins and there were no other similar cases in the family.

Her pregnancy and delivery were uneventfull. She was breast fed for the first 40 days of her life and thereafter put on similac and cows milk. At the age of 8/12 cereals, biscuits, beans and potatoes were introduced and gradually

other types of food were offered. Her devolopment until that age was normal as she sat at the age 5 months and was able to stand unsupported.

A few weeks after introduction of solid food she developed diarroea and vomiting. Her general health started to deteriorate, she lost her appetitc and began to lose weight steadily.Her abdomen became distended and she gradually lost the ability to stand.

Efforts to treat her diarrhoea and to improve her general health here and abroad proved failure. She was labled as a case of severe marasmus and her parents were advised to put her on a high protien and carbohydate diet and at one stage antituberculous chemotherapy was given for a period of 4 months.

Examination on admission revealed her to be moderately anaemic, fretful and weak. Her weight was 5.5 kg. and her height was 70 cm.i.e. well below the 3rd percentile for age, sex. Her abdomen was markedly distended compared to her wasted face, buttocks and extremities. She had a reddish coarse hair. Her liver was just palpable but the spleen was not felt. Examination of her C.V.S. and R.S. revealed no abnormality.

Investigations done on admission were: Hb of 10.5 gms/100mls. Urea, Electrolytes and plasma protiens were normal Stools PH. was 6.5 and microscopic examination of the stools showed exess of fats but no giardia or parasites were seen and no organisms were grown on culture. Fat content of the stools in 24 hours was 12 gms. Her G.T.T. was flat and one hour blood xylose level was 12.5 mgs%. Mantoux test was negative. Barium meal and follow through showed features of malabsorption.

Her jejunal mucosa seen under the dissecting microscope was flat, and histologically there was almost total loss of villi with cellular infilteration of the lamina propria.

She was put on a gluten free diet on the 2nd of Nov. 1973. This was followed by a drammatic improvement of her general health and return of her appetite. She gained double her previous weight in 13 weeks time with a maximum spurt of growth in the first 6 weeks of treatment. Her height increased to 78 cms over the same period. Her diarrhoea stopped and her bowls opened once a day her stools were normal and contained no fats on direct microscopy. I saw her a week ago when she was walking unsupported and was feeding herself alone.

Xylose absorption test repeateed after 3 weeks of treatment was 24 mgs%.

CASE (2):

An 18/12 months male child the only product of a consanguinous marriage. His parents were first cousins and were of mixed Sudanese and Yamani blood His pregnancy and delivery were normal. He sat at 5 months and could walk supported before he devoloped his present illness.

He was breast fed for the first year of life. Solid food was introduced at 6/12 of age and at 10 months he joined his family eating a normal Sudanese diet including bread, kisra, biscuits and wheat products among other types of food.

His illness started with diarrhoea at the age of 8/12 with a frequency of 4 to 10 times per day. The stools were described as bulky, offensive, mixed with mucus and were yellowish in colour. He had a poor appetite and had been progressively losing weight over the last 9 months. His abdomen was noted to increase in size and during his illness he lost the abilty to stand or sit unsupported.

On examining him he was miserable, wasted and had carious teeth. His weight was 9 kgs and height was 30.5 cms. His abdomen was grossly distended and his buttocks were hanging in folds. His limbs were wasted and weak. His liver and spleen were not palpable and examination of other systems was unremarkable.

On admission his Hb% was 7.7 gms%, his W.B.C. was normal and E.S.R. was 30mm/hour. Urea, electrolytes, liver fuction tests were normal. However he had a flat glucose tolerence test and one hour xylose test was 12.5 mgms% His fat excretion in 24 hourss was 12.5 mgms. Barium meal and follow through showed dilatation of bowl loops with floculation and clumbing of barium. Dissecting microscopic examination of the jejunal biopsy showed convolutions and ridges; histologically no villi could be seen and there was marked cellular infilteration of the lamina propria.

He was put on a gluten free diet with rapid improvement in his general health and appetite. He added four kilos in weight in 12 weeks time and seven centimeters in height over the same period. His bowls opened regularly once a day. His xylose level at one hour rose to 20.5%.

CASE (3):

A female child aged 18 months. She was admitted to hospital in Jan. 1960 with history of chronic diarrhoea and occational attacks of vomiting for 5 months before admission. She had no appetite for food and there was a remarkable weight loss during her illness.

She was breast fed for 15 months. Suppliments with solid food were introduced at the age of one year and those consisted of rice, bread, Kisra and biscuits.

At the time of examination she was pale, wasted with thin extremities and markedly distended abdomen. Her liver and spleen were not palpable. Examination of other systems did not reveal any abnormalities.

INVESTIGATIONS:

She had a normocytic normochromic anaemia with a Hb of 40%. Her W.B.C was normal and E.S.R. of 30 mms/hour. Urea and electrolytes: normal. Plasma

protiene 6.5 Gms with an albumen level of 3 Gms%. Her tryptic activity of the duodinal juice was positive up to a dilutionof 1/512 stools fat was 10 Gms. in 24 hours. Barium meal and follow through showed dilatation of bowl loops with hoculation oclumbing of barum.

A gluten free diet was advised with very mgood responceThe child started to gain weight rapidly as a consequence of an improved appetite. Her diarrhoea stopped and she was discharged in a good health. Sehe was seen in subsequent occations and she seemed to have maintained improvement in all parameters, but unfortunately she was not followed up for a longer time to assess her clinical course any further.

CASE (4):

A male 9 mothes old, the last in a family of four children. His parents were second cousins.

He suffered from attacks of constipation alternating with diarrhoea from the age of 20 days. The mother was advised to stop breast feeding and to introduce dried milk formula and cereals at the age of 10 weeks.

Since then, the child had never been well as he continued to have diarrhoea. The stools were bulky and offensive. His condition took a progressive course and at 6 months his abdomen was noted to be distended.

Clinically he looked ill, anaemic, and wasted. His weight was 9 pounds. His abdomen was grossly distended and limbs were wasted. Liver and spleen were not felt.

Investigations done showed him to be moderately anaemic with a Hb. of 8 Gms/100 mls; Urea and electrolytes were normal. Serum albumen 2.5 gms/100 mls. Fat content in 24 hours was 14 gms and the tryptic activity in the duodinal aspirate was normal. A barium meal and follow-through showed features of malabsorption.

He was put on a gluten free diet and following which he improved tremendo-ously with return of mood, appetite and gain in weight. His bowls opened normally. Again he was followed-up for a short period.

CONCLUSION:

Coeliac disease once thought an affection of white races, have been docu mented in other ethnic groups i.e. in Indians and pakistani's. So far it has not been described in Africans nor in the chinese.

The first two cases represent the first cases of coeliac disease in Sudanese children confirmed by jejunal biopsy, low xylose absorption test and the respo nce to gluten free diet. The last two cases were diagnosed on clinical features evidence of steatorrhoea and by a favourable response to gluten free diet.

Dura or Sorgham Vulgaris could be toxic to ceoliac patients as has beeu suggested by qualitative and quantitative studies of this type of millat. (Unpublished work). It has been shown to contain a high glutamine level of it's total protiens. Clinical trials on proven cases of ceolaic disease are required to solve this question.