mal right side liver and normal kidneys on both sides. Hemogram and peripheral smear did not reveal any abnormality.

Discussion

Alfred Poland in 1841 described the association of aplasia/hypoplasia of the sternal head of the pectoralis major muscle with ipsilateral symbrachydactyly(2,4), Since then several cases of Poland Syndrome have been reported with varying associations like ipsilateral Moebius syndrome(4,5), absent kidney and upper limb musculature(2,4), rib deformities, Sprengel's deformity, hemivertebrae and radio-ulnar synostosis and axillary webs(6,7).

Dextrocardia has been reported only with two cases of isolated Poland Syndrome, one had associated acute lymphatic leukemia(2) and other had absent incisors(2). Dextrocardia in association with Poland and Moebitis syndrome has been reported twice(2). Our patient was isolated case of Poland's anomaly with dextrocardia without any evidence of leukemia at the time of diagnosis.

Embryogenesis of this anomaly is unknown but it has been suggested that primary defect may be in the development of the proximal subclavian artery with early deficit of blood flow to the distal limb and pectoral region(1,8,9).

Surgical management of Poland Anomaly in females includes latissimus dorsi augmentation to anterior chest with silicone gel prosthesis placement in submuscular position to augment breast movement. In males transfer of latissimus dorsi is done if abduction of involved shoulder is severely affected(1,3).

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Celiac Disease in Insulin Dependent Diabetes Mellitus

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Diarrhea and failure to thrive in a patient with diabetes mellitus may often be

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dismissed as an integral part of the disease. That this may not always be the case, has been amply demonstrated by reports of the occurrence of celiac disease with Insulin Dependent Diabetes Mellitus (IDDM)(1). The purpose of this report is to record the rare occurrence of celiac disease, in association with glycogen storage in the liver (Mauriac's syndrome) in a young girl with IDDM.

Case Report

A 9-year-old girl, who was diagnosed to have diabetes mellitus at the age of 18 months, was referred to the Pediatric services of this Institute with failure to thrive, progressive distension of the abdomen and diarrhea for the preceding 4 years. The stools were pale, offensive and bulky suggesting a malabsorptive state. She had poorly controlled diabetes and had been receiving (irregular) single daily injections of lente insulin. The doses had been regulated on the basis of urinary qualitative change in color of Benedict's reagent. Diabetic ketoacidosis had necessitated admission, once, in a peripheral hospital. She had severe growth retardation with a weight of 15 kg and height of 108 cm only. She had a moon like facies and there was generalised muscle wasting. The abdomen was protruberant with a firm moderate hepatomegaly (liver span of 12 cm). There was no hypertension or clinical evidence of peripheral neuropathy.

The hemoglobin was 7 g/dl and white blood cell count 7.2×10^9/L. The anemia was predominantly microcytic, hypochromic in nature. The screening tests of coagulation revealed no hemostatic abnormality. A skiagram of the chest was normal. The results of blood biochemistry at the time of admission were as follows: Blood sugar 400 mg/dl; serum bilirubin 0.5 mg/dl; serum proteins 7 g/dl albumin 3.8 g/dl and globulin 3.2 g/dl, serum alkaline phosphates 24 KA units, SGOT14–IU and SGPT–16IU.

The-24 hour fecal fat was more than 10% of the total intake. The D-xylose test was 2 g/5 g/5 hours. Biopsy of the small intestinal mucosa showed flattening of the mucosa with loss of villous configuration over most areas. The lamina propria showed a diffuse excess of lymphoplasmacytic cells whilst the crypts showed hyperplastic changes.

A percutaneous trucut liver biopsy showed normal morphology with hepatocytes full of glycogen. Gradual increases in lente and plain insulin given in twice daily doses and a gluten free diabetic diet resulted in steady improvement. A diagnosis of IDDM with Mauriac's syndrome and celiac disease was thus established.

Her diarrhea subsided within a fortnight. She was discharged once satisfactory diabetic control had been attained. She maintains good progress and has demonstrated a weight gain of over 2 kg in the first month.

Discussion

The diagnosis of celiac disease in the index patient is based on a clinical picture of malabsorption and initial flat jejunal mucosa and significant symptomatic improvement on a gluten free diet. The first biopsy confirmed case of celiac disease in a diabetic was documented by Ellenburg and Bookman(2) in 1960. Since then several authors have shown that diabetes mellitus coexists with celiac disease much more frequently than the reported figures for the prevalence of these diseases in the general population(3,4). It is estimated that 1 to 1.5% of children with IDDM have celiac disease as well whilst, diabetes mellitus oc-

curs in 4-6% of patients with celiac disease(3,4).

The control of diabetes in patients with coexisting celiac disease is usually difficult and episodes of symptomatic hypoglycemia are fairly frequent, especially during exacerbations of diarrheal symptoms. This could be one of the reasons for poor control in our patient, besides the fact that she also had Mauriac's syndrome. Her insulin requirement increased once she was started on a gluten free diet. A similar phenomenon has been described earlier by Thain *et al.* and by Komrower(3,5). It is probable that fluctuations in control of diabetes are associated with episodic alterations of carbohydrate absorption and improved carbohydrate absorption following gluten restriction may result in an increased insulin requirement.

Diarrhea, which is the commonest symptom of celiac disease, may also be a feature of diabetes. It often, does not, attract the attention that it deserves. It is re-commended that the presence of short sta-ture, anemia, clubbing, edema, abdominal distension and the absence of neuropathy should arouse the suspicion that diarrhea is not of diabetic origin. Similarly, a history of repeated episodes of hypoglycemia, particularly when the diarrhea is troublesome, warrants appropriate investigations for coexisting celiac disease(1). A jejunal biopsy in such circumstances provides diagnostic evidence. In diabetic diarrhea the intestinal mucosa is normal whilst there is partial or severe villous atrophy in celiac disease(1).

In the majority of cases, diabetes has preceded the onset of symptoms of celiac disease by several months or years(3,4). It is speculated that genetic factors are important in the etiology of both diabetes mellitus and celiac disease(1,3). A higher frequency of the histocompatibility antigens HLA B8 has been demonstrated in patients with celiac disease(6) and in IDDM(7). However, other factors, probably environmental, must be operative since celiac disease coexists with diabetes mellitus relatively infrequently.

Our patient also had increased glycogen stores in the liver and short stature features that are consistent with Mauriac's syndrome(8). This syndrome was first described in 1930 in a 10-year-old poorly controlled diabetic girl with hepatomegaly, protuberant abdomen, dwarfism, moon shaped face and, fat deposition around the shoulders and on the abdomen. Mauriac had suggested that the drug itself might well be the cause of these curious mainfestations. High levels of blood glucose have since been shown to increase liver glycogen content and this has been confirmed by liver biopsies(9). Hansen has shown that improvement in diabetic control decreased the size of the enlarged liver(9).

This report heightens awareness of coexisting celiac disease in IDDM. It is recommended that intestinal mucosal biopsies be resorted to more frequently in the diabetic with recurrent or chronic diarrhea. A prompt diagnosis of celiac disease and consequent introduction of a gluten free diet would mean a greater stability in diabetic control and a healthier patient.

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Lethal Multiple Pterygeum Syndrome

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Lethal multiple pterygeum syndrome (LMPS) manifests in the newborn with multiple contractures and have been subclassified based on associated features(1). They are often referred to the geneticists as cases of arthrogryposis multiplex congenita (AMC). Identification of this group with pterygeum is important for management and counselling. We present here four cases because of the rarity of this condition and importance of recognition as a separate entity, as less than 50 cases have been reported so far in the world literature(2).

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Case Reports

Four cases being reported were referred to the Genetics Department in 1990, as cases of AMC for evaluation and counselling. The details of the four cases are given in *Table I.*

Discussion

All the above four cases were diagnosed as multiple pterygeum syndrome. All were male. The male:female ratio as per literature is 1:1. Two of the four couples were consanguinous and one was advanced in age. There was no recurrence or family history of similar defect. All the four presented as extended breech, probably due to reduced motility of the fetus due to physical defect. Intrinsic neuromuscular abnormalities of the fetus may prevent the fetus from undergoing normal version, hence the association between breech presentation and fetal abnormalities(3). Only one child is alive and under follow-up.

The abnormalities reported in literature are–popliteal web, toe nail hypoplasia, cleft lip with or without cleft palate and lower lip pits(2). The occasional findings reported are: oral frenula, cutaneous webs between eyelids, intercrural pterygeum, syndactyly, hypoplasia of digits, bifid or absent patellae, scoliosis, cryptorchidism, ambiguous external genitalia and inguinal