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Formation of Paediatric Endocrine Society of Bangladesh (PESB)

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Health care services in Bangladesh have seen a remarkable improvement in the last few years with dramatic fall in communicable diseases due largely to the hugely successful EPI programme. With this reduction noncommunicable diseases (NCDs) have emerged as a big threat in the form of diabetes mellitus, hypertension, cardiac disease, cancer. Lifestyle changes have brought about an alarming rise in obesity and its consequences: impaired glucose tolerance (IGT) and type 2 diabetes mellitus (T2DM). A large number of children suffer from various endocrine problems like diabetes, hypothyroidism, short stature, obesity etc in our country. More than four thousand children and adolescents attend BIRDEM hospital with various endocrine problems every year and about six thousand children and adolescents with diabetes are registered in this hospital, which is the largest referral centre for endocrine disorders in our country. This number may not reflect the total burden of childhood endocrine problems in the community.

There is a dearth of trained manpower and service in the field of paediatric endocrinology in our country. With the thought of forming a common platform for paediatric endocrinology in Bangladesh, a meeting was held on January 7, 2009 in BIRDEM Hospital, Shahbag in presence of Prof Nazmun Nahar, Prof Kishwar Azad, Prof Tahmina Begam, Dr. Shahida Akhter, Dr. Fauzia Mohsin, Dr Jebun Nahar and Dr. Bedowra Zabeen from Dept pf Paediatrics, BIRDEM Hospital. Subsequent meetings were held in presence of paediatricians from various institutions and finally in presence of 33 members from various institutions, in Auditorium, BIRDEM Hospital. Paediatric Endocrine Society of Bangladesh (PESB) was formed on 26th September 2011 with an objective to promote and improve overall care in the field of paediatric endocrinology (including paediatric and adolescent diabetes) in the country through

a. promoting better care and services for patients with various endocrine disorders.
b. providing and encouraging training and continuing medical education on paediatric endocrinology.
c. facilitating basic and clinical research in the field of paediatric endocrinology.
d. developing relevant manpower.

Inside the Issue

Paediatric Endocrine Society of Bangladesh (PESB)
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Till date the Society has organized Updates on Paediatric Endocrinology in collaboration with Asia Pacific Pediatric Endocrine Society (APPES), several CME programmes on various endocrine topics throughout the country. Its First National Conference and First International conference were successfully held in April, 2015 and April, 2017 respectively and the 2nd International Conference will be held on 29 April, 2019 in Dhaka. This is our first newsletter and we plan to publish newsletter at least twice yearly. Currently we are preparing a guideline on 'Common Endocrine Problems in Children" which is well under progress and we should be able to release it soon. It may be noted that in recent times physicians from different Institutions have taken training on paediatric endocrinology in BIRDEM hospital. As a prerequisite to start FCPS course on Paediatric Endocrinology we have applied to BCPS for developing a curriculum on Paediatric Endocrinology. As a society PESB is still young and have a long way to go and we hope that together we will make this happen.

ISPAD Clinical Practice Consensus Guidelines 2018: What is new in diabetes care?

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Since the first version of the guidelines was published in 1995, evidence has accumulated demonstrating the lasting benefits of near normal glycemia. Intensive management of all aspects of diabetes, especially glycemic control, is now the international gold-standard in children, adolescents, and young adults. The release of the 2018 ISPAD guidelines comes at a time when type 1 diabetes represents an increasingly major burden in both adults and children.[1,2] Each chapter is organized as follows: what is new, executive summary, and recommendations; main body of the chapter; references. Three new chapters have been added to the 2018 guidelines. One of these provides recommendations on the use of technology in children, adolescents, and young adults with diabetes and appraises the pros and cons, as well as costs, of pumps, sensors, and automated insulin dosing devices.[3] Continuous subcutaneous insulin infusion (CSII) pump therapy can be used safely and effectively in youth with type 1 diabetes (T1D) to assist with achieving targeted glycemic control and is appropriate for youth with diabetes, regardless of age . Insulin pump therapy can assist with reducing episodes of hypoglycemia . Insulin pumps reduce chronic complications of T1D in youth, even when compared to those with similar hemoglobin A1c (HbA1c) levels on multiple daily injection (MDI) therapy.[4] The number of young people with diabetes attending school is increasing, placing a significant burden on families, health care systems, and schools. Children may spend more than 30 hours per week in the school environment. Irrespective of age and ability, all students with diabetes at school must receive the support, encouragement, and supervision of school personnel. Optimal management of diabetes at school is a prerequisite for optimal school performance, including learning , and for the avoidance of diabetes-related complications. Maintaining normoglycemia during school hours is important and day to- day glycemic targets should not differ from any other setting. The type of insulin regimen used at school should be tailored to the needs, ability, and wishes of the child/family and should not be dictated by the school resources. "Glycemic control targets and glucose monitoring" chapter has been updated to reflect the major advances that have occurred regarding blood glucose monitoring, and technology.[5] An individualized approach to the patient is emphasized and a decrease in "target" HbA1c to <7% is recommended for those using the new technologies consistent with the goal for children, adolescents, and young adults. Advances in the genetic diagnosis of atypical diabetes have guided decisions regarding the best treatment for many children.[6] Type 2 diabetes in youth, a consequence of the obesity epidemic, has become widespread in many regions of the world and the evidence base for treatment of this disorder has expanded significantly since 2014.[7] Moreover, pathophysiology-based treatment with glucagonlike peptide 1 (GLP-1) agonists and Inhibitors of dipeptidyl peptidase 4 (DPP-4) and sodium-glucose co-transporter 2 (SGLT2) inhibitors is being investigated and will likely increase the pharmaceutical options available for adolescents with type 2 diabetes in the next few years.

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Human growth hormone supplementation in Children

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Growth hormone is essential for normal growth in children. It increases growth by a direct action on the growth plates and by production of insulin-like growth factors, especially IGF-1. Growth hormone has important effects on the metabolism of proteins, lipids and carbohydrates, not only during childhood, but also throughout adult life.

Since 1986 only available GH was from cadaveric donor. In 1985 and thereafter, Creutzfeldt-Jakob disease, a degenerative neurologic disease was diagnosed in some patients who had received natural hGH. Because of the possibility that prior contaminating donor pituitary glands were transmitted to the GH-deficient patients, causing their deaths, natural GH was removed from distribution. Recombinant hGH called somatropinnow accounts for the world's current supply.

FDA-Approved Indications for GH Therapy: 1. Childhood GH deficiency (1985) 2. Turner syndrome 1(996-1997) 3. Noonan syndrome 4. Chronic renal insufficiency (1993). 5. Small for gestational age (SGA)(2001) 6. Idiopathic short stature. 7. SHOX gene mutation 8. Prader-Willi syndrome (2000) 9. Acquired immune deficiency syndrome (AIDS) (catabolic state)

Monitoring: 1. Growth with gender and age reference charts for population. 2. Pubertal staging. 3. Bone age. 4. Serum IGF-1 level, glucose, HbA1c, TSH, FT4. 5. Symptoms of benign intracranial hypertension, slipped capital femoral epiphysis.

Treatment with rGH should be discontinued if any of the following apply:

1. Growth velocity is less than 2 cm/year.
2. Reaching an acceptable height close to target height.
3. Bone age reaching 16 years in boys and 14 years in girls.

Side Effects of GH therapy: Headache, visual problem, fluid retention, weight gain, glucose intolerance, diabetes mellitus, arthralgia, myalgia, benign intracranial hypertension, kyphosis, scoliosis and sipped capital femoral epiphysis.

Contraindication of GH therapy: Active Malignancy, pre-proliferative/ proliferative retinopathy in diabetes mellitus.

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Supplementation of vitamin D in infants- should we practice?

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Vitamin D and calcium deficiencies are common worldwide, causing nutritional rickets and osteomalacia,which have a major impact on health, growth and development of infants, children, and adolescents; the consequences can last into adulthood.

Rickets has become common in some parts of Bangladesh during the last decade and one study even claimed that there were 5,000,000 affected children in Bangladesh[1].

In another study, in rural Sylhet, Bangladesh, found that the vitamin D status in young infants were poorer than that might be expected on that geographical location[2].

While breastfeeding is the recommended method of infant feeding and provides infants with necessary nutrients and immune factors but breast milk alone does not provide adequate amount of vitamin D. Human milk typically contains vitamin D concentration of 25 IU or less per liter. Although vitamin D deficiency rickets among breastfed infants is rare, but it can occur if an infant does not receive additional vitamin D from supplement or from adequate exposure to sunlight[3].

Classically, nutritional rickets presents after 6 months of age. In a report with relatively large patient populations, the mean age at diagnosis was 14.6 months and the youngest patients was 4 months old (4). In Dhaka Shishu Hospital, we also recently observed a significant number of the patients presented with hypocalcaemic seizure within the first 3 months of life and was found to be associated with vitamin D deficiency (5).

American Academy of Pediatrics in November, 2008 recommended a supplement of 400 IU per day of vitamin D for all breastfed infants, which is Recomended Dietary Allowance for this nutrient during infancy. Recently, Global Consensus Recommendations on Prevention and Management of Nutritional Rickets (6) also recommended 400 IU/day of Vitamin-D to prevent rickets for all infants from birth to 12 months of age, independently of their mode of feeding.

In conclusion, we may consider practicing global recommendation of vitamin D supplementation among our infants of Bangladesh.

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Rabson-Mendenhall syndrome: a case report

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Introduction: Rabson-Mendenhall syndrome (RMS) is a rare genetic disorder that was first described by Rabson et al in 1956[1]. This insulin resistant syndrome occurs due to autosomal recessive mutation of the insulin receptor gene (INSR)[2].

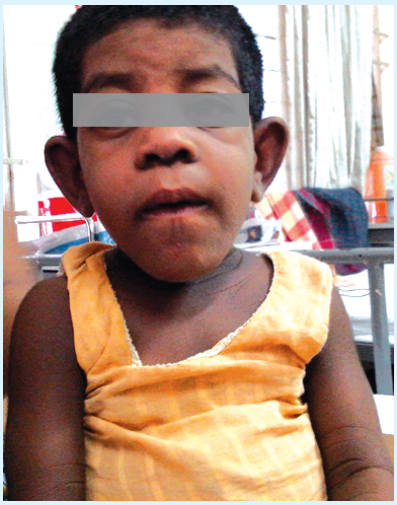

Fig 1: dysmorphic facial feature

The manifestations of severe insulin resistance includehyperinsulinaemia, post prandial hyperglycaemia, paradoxical fasting hypoglycaemia, acanthosis nigricans, lipoatrophy, growth retardation, and clitoral enlargement in female. The characteristic facial features include dysmorphic coarse facies with a prognathic jawline, fissured tongue, with dental precocity, hyperplasia and caries. Cutaneous findings include hypertrichosis, skin tags, lichenified skin, and onychauxis and protuberant abdomen. Although insulin levels are extremely elevated initially and then decrease with age, they remain higher than normal values. There is a paradoxical fasting hypoglycemia and postprandial hyperglycemia early in life, followed by constant hyperglycemia (by four years of age) and constant intractable ketoacidosis by six years of age. Patients with RMS need high doses of insulin and insulin-sensitizing drugs such as metformin and glitazones[3].

Case report: Here, we describe a 3 -year-old girl of consanguineous parents, a known case of diabetes mellitus for one year,whopresented with uncontrolled hyperglycaemia and dysmorphic facial features. Her elder sister, who had similar clinical features died at 1 year of age.

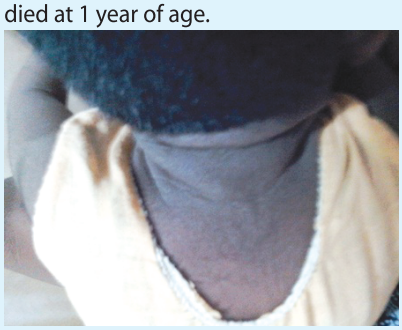

Fig 2: severe acanthosis nigricans

Our patient had coarse facial features, broad nose, prominent orbital ridge,macroglossia,dental dysplasia, dry rough skin, thick scalp hair, excessive body hair (fig 1). She had severe acanthosis nigricans. (fig 2). She also had growth retardation, developmental delay and precocious puberty.Her blood glucose profile ranges from 10-28mmol/l, HbA1c was 14.1%. Her serum Insulin level was grossly elevated (>300mIU/l). She needed large doses of insulin (18unit/kg/day) along with metformin to control her diabetes.

Conclusion: Rabson Mendenhall syndrome, Leprechaunism, Lipodystrophy all are insulin resistance syndrome. Diagnosis of RMS is done by its classic clinical features and high blood glucose and insulin level. The Pro220Leu mutation of the INSR gene was found in patients with this severe insulin resistance cases[4]. There is no complete cure for RMS. They have poor prognosis and survive usually less than 20 years of age.

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Celebration of World Growth Awareness Day jointly organized by PESB,CDiC and Novo Nordisk

A Rally on World Diabetes Day