NICU blood sugar was tested with glucometer and if it was less than 40mg/dl and if this low level was confirmed by laboratory, the patient entered this study. For the determination of etiology of hypoglycemia more diagnostic tests were performed and according to history, physical examination and laboratory findings the etiology of hypoglycemia was defined and finally all these information were recorded in the questionnaire.

Results: According to this study, 7.5% of neonates that admitted in Emam Reza hospital NICU had hypoglycemia and the most common Etiologies for hypoglycemia included: prematurity, Asphyxia and SGA.

Conclusion: Incidence of neonatal hypoglycemia in our NICU is identical that of western countries and is less than the incidence of hypoglycemia in developing world. and because the most common etiology of neonatal hypoglycemia is prematurity, SGA and Asphyxia, we can decrease its incidence through better control of preterm deliveries and through preventing preterm labor and also institution of good resuscitation measures when needed.

Keywords: Hypoglycemia, Prematurity, Asphyxia

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PREGNANCY INDUCED HYPERTENSION AND SIGNS OF DECREASED INSULIN SENSITIVITY IN THE NEONATES

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Aim: To determine the impact of maternal pregnancy induced hypertension (PIH) on insulin sensitivity in term neonates.

Methods: The study group consisted of 130 term neonates without major malformations, whose mothers did not have chronic hypertension or intrauterine infections. Neonates were divided into two groups according to the maternal presence or absence of PIH. Anthropometric measurements were performed at birth. Fasting glucose and insulin levels were measured on the 3rd day after birth. Lipids were determined from blood samples obtained by venipuncture from 72 neonates.

Results: The 24 neonates born to mothers with PIH had significantly lower birth weight, body length and chest circumference ($p < 0.05$) than

106 neonates born to mothers without PIH. Insulin levels and insulin:glucose ratio in neonates born to mothers with PIH were significantly higher than that in controls (mean $\pm$ SE 15.98 $\pm$ 2.18 vs 10.33 $\pm$ 1.04 μ IU/ml, $p < 0.05$; 4.15 $\pm$ 0.57 vs 2.52 $\pm$ 0.27, $p < 0.05$, respectively), despite similar glucose levels (mean $\pm$ SE 4.07 $\pm$ 0.26 vs 4.45 $\pm$ 0.14 mmol/l, $p = 0.265$). This association was still observed after adjusting for birth weight. Furthermore, 12 neonates of mothers with PIH had significantly higher mean triglyceride levels, and lower mean high-density lipoprotein cholesterol levels than 60 neonates from mothers without PIH (mean $\pm$ SE 1.98 $\pm$ 0.24 vs 1.36 $\pm$ 0.11 mmol/l, $p < 0.05$; 0.51 $\pm$ 0.11 vs 0.81 $\pm$ 0.05 mmol/l $p < 0.05$ respectively).

Conclusion: Intrauterine environment may be a major determinant in neonatal metabolism; maternal PIH may influence birth weight and insulin sensitivity of term neonates.

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PEDIATRIC ADRENOCORTICAL TUMORS. TWENTY FIVE YEARS EXPERIENCE IN A UNIVERSITY HOSPITAL IN TEHRAN, IRAN

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Adrenocortical tumors are rare childhood and adolescent neoplasms. More than 95% of these tumors are functional and present with virilization, Cushing's syndrome, hypertension or hyperestrogenism. During 25 years of experience in Taleghani hospital, Tehran, Iran, we could find 7 cases of such tumors. Patients were 2 males and 5 females, aged 2-16 years. Five patients presented with virilization, one with a mixed picture of acute psychosis, Cushing's syndrome and feminization and one with Cushing's syndrome. Duration of the disease was between 4 months to 5 years and 3 patients were erroneously diagnosed and treated as congenital adrenal hyperplasia. Diagnosis was confirmed with basal hormonal evaluation, suppression tests and appropriate imaging. All patients underwent open surgery. During surgery a blood sample was drawn from the adrenal vein of the affected side and its steroid contents were compared with the data obtained from the peripheral blood during and after surgery. Adrenal vein evaluation revealed that cortisol, testosterone and other hormones were directly secreted from the tumor and peripheral conversion had a limited

contributory effect on plasma levels. The tumors weighed between 36 to 103 grams and while in cases #1&3 pleomorphic atypical nuclei with capsular and vascular invasion were reported, the patients had no signs or symptoms of relapse so far. Duration of follow up is 4 to 16 years and no case of relapse has been seen.

Conclusion: Pediatric adrenocortical tumors are multi - hormone secreting neoplasms and have a more favorable prognosis in comparison to adults.

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THYROID HORMONE RESISTANCE IN AN IRANIAN FAMILY PRESENTING WITH GOITER AND NEUROLOGIC ABNORMALITIES: REPORT OF THE MUTATION

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Resistance to thyroid hormones (RTH), an inherited syndrome of reduced tissue sensitivity to thyroid hormones is a well known although rare disorder and has not been reported in Iran so far.

Most of the reports concerning the genetic characteristics of the disorder belong to western countries and there are limited reports on thyroid hormone gene study on patients with RTH in Middle East region. Since 1986 we follow the first five patients belonging to a single family with RTH in Iran and reported their clinical pictures and also the effects of short- term treatment with graded dose of T3 on pituitary and peripheral tissue markers. Here we report the result of the gene study on this family.

DNA was extracted from peripheral white blood cells of the family in Tehran, Iran and transported to the Thyroid Study Unit at the University of Chicago. Sequencing of the TR beta gene disclosed a point mutation in ligand binding domain with substitution of Aspartic Acid instead of Glycine at position 345. Same mutation has been reported previously in limited number of patients with similar manifestations.

Conclusion:

1, Glycine to Aspartic Acid substitution is associated with a rather sever form of RTH presenting with big goiter and neurologic manifestations,

2, paucity of the reports on RTH in Asian and African countries are due to the lack of attention to this syndrome and we propose that the diagnosis of RTH should be kept in mind in differential diagnosis of patients who are presumed to be thyrotoxic.

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HYPEROSMOLAR, HYPERGLYCAEMIC, NON-KETOTIC ACIDOSIS IN A 17 MONTH OLD - A CASE REPORT AND LITERATURE REVIEW

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Background: Hperosmolar, hyperglycaemic, non-ketotic acidosis (HONK) is a rare initial presentation of diabetes in childhood. It is increasingly reported in the literature in older children with type II diabetes but there are few case reports of younger children with insulin dependant diabetes (IDDM). It is associated with higher morbidity and mortality than diabetic keto-acidosis (DKA), probably secondary to cerebral oedema.

Case: We report a case of this disease in a 17 month old boy with Down syndrome who presented to the ward with a decreased level of consciousness following a short history of being generally unwell. Initial investigation revealed a finger prick glucose of "Hi" with a pH of 7.24. A presumptive diagnosis of DKA as first presentation of IDDM was made. However, subsequent investigations revealed a plasma glucose of 95 g/dl, serum osmolality 400 mosm/kg, with no urinary ketones and the patient was diagnosed with HONK. We discuss the difficulties we encountered in avoiding a precipitous fall in his plasma glucose and in managing his hypernatraemia, which peaked at 171 mmol/l.

Conclusion: HONK is a rare but serious first presentation of diabetes in children. Conventional DKA management is likely to result in a precipitous fall in plasma glucose and exacerbate electrolyte disturbances, increasing the risk of cerebral oedema. Current management is based on data extrapolated from adult studies and a few case reports and involves much lower doses of insulin with cautious fluid replacement. Knowledge of this condition should reduce the risk of serious complications.