important to avoid long-term neurological sequelae. We aimed to describe the presentation, clinical and genetic outcomes in a series of infants with CHI.

Methods: Retrospective case series of twenty-two patients diagnosed with CHI between 1992 and 2010 at The Children's University Hospital, Temple Street.

Results: Fifteen of the twenty-two patients were male. Median age at presentation was day 2 (range: day 1- 18 months). Seizure was the most common presentation occurring in nine patients (40%). Mean glucose requirements to maintain euglycaemia were 14.6 mg/kg/min. First line treatment with diazoxide was commenced in all patients of whom twelve (54%) responded. Of those that did not respond to diazoxide, three were stabilized on octreotide and seven required surgery. One of these patients had a focal laparoscopic pancreatic resection, following pre-operative 18F-Fluorodopa

positron emission tomography (18F-L -dopa PET). Genetic testing was performed on fifteen patients (68%). Five had a mutation(s) in the *ABCC8* gene and two had mutations in the *HNF4A* gene. Twelve patients (54%) had a normal neurological outcome. Of the remaining ten, three patients had severe global

developmental delay, three had moderate motor impairment and the remaining four patients had mild impairment.

Conclusions: No clinical parameter significantly predicted outcome. Advances in both molecular genetics and 18F-L -dopa PET scanning will likely improve outcome for children diagnosed with CHI in the future.

1101

NEONATAL HYPERINSULINAEMIC HYPOGLYCAEMIA : AUDIT ON HYPOGLYCAEMIA SCREEN

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Background: Hyperinsulinaemic hypoglycaemia (HH) is a major cause of recurrent and persistent hypoglycaemia in infancy and childhood. HH can be congenital, associated with developmental syndromes or secondary to risk factors, also called

as transient HH. Prompt recognition and early management are essential, to avoid brain injury

Objective: To assess the adherence to hypoglycaemia screen protocol in terms of diagnosis, management and short term follow up (off medication). Design and

Methods: Retrospective audit to analyses all hypoglycemic screens carried between April 2008 to December 2009. Data from the proforma was collated to Excel and was analysed using simple statistical methods and percentage calculations.

Results: 17 babies were identified who had glucose requirement higher than 8mg/kg/minute. 65% of them were term and 35% preterm. 59% had birth weight less than 10th centile of which half of them were asymmetrical IUGR. 12% babies were born to mother with gestational diabetes mellitus. All babies were born with Apgars >5 @ 5minutes with lactate ranging from 1.8 to 17mmol/L. One baby was diagnosed with genetic abnormality. Inappropriately raised levels of insulin (6.3 to 226 pmol/L) in the context of low blood sugar (0.0 to 3.1 mmol/L) were noted in 70% babies. Pharmacological intervention were needed in 41% and remaining were managed with increased calories by adding glucose polymer.

Conclusion: All babies had transient HH secondary to risk factors. All of them responded appropriately and were able to come off pharmacological interventions. Transient Hyperinsulinaemic hypoglycaemia appears to be fairly common and resolves spontaneously.

1102

EVALUATION OF INCIDENCE AND ETIOLOGY OF NEONATAL HYPOGLYCEMIA IN EMAMREZA HOSPITAL NICU

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Introduction: Hypoglycemia is a common problem in neonatal period and can cause many undesirable neurological effects such as : Seizures , Psychomotor retardation and decreased IQ . the goal of this study is to determine the incidence and etiology of hypoglycemia in neonataes .

Methods: this discriptional study was performed on 927 neonates admitted in Emam Reza hospital NICU over a one year period. After admission in

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

1103

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.

1104

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