Methods: During August and October 2009, Samples of umbilical cord blood was collected from 200 full term newborns (89 males and 111 females) to be used to determine lipid profile levels: The analysis was performed using Selectra 2 auto analyzer (vital scientific, spankeren, Netherlands). Total cholesterol (TC) and triglycerides(TG) levels were assayed with a sensitivity of 5 mg/dl using enzymatic colorimetric tests with cholesterol esterase and cholesterol oxidase and glycerol phosphateoxidase respectively (ParsAzmon kits, Iran). High-density lipoprotein cholesterol (HDL-c) was measured after precipitation of the Apo lipoproteins with phosphotungistic acid. Low density lipoprotein cholesterol (LDL-c) was calculated from serum TC, TG and HDL-c using Fried Wald formula. It was not calculated when TG concentrations were more than 400 mg/dl. and Non-HDL cholesterol was calculated.

Results: Mean TC: 74.58, Mean TGs: 96.59, Mean HDL-C:28.68, Mean LDL-C: 29.25 and Mean Non-HDL cholesterol: 45.90. Biochemical factors which been studied had not significantly difference between genders.

Conclusions: Our findings show that cord blood TGs level in neonates of our center are higher than other countries that have been studied previously, no significant difference be observed in others biochemical factors. More detail researches of predisposing factors of cardiovascular system in Iranian population is highly suggested.

1099

NEURO-PSYCHOMOTOR DEVELOPMENT IN CHILDREN WITH CONGENITAL HYPOTHYROIDISM

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Aims: Neuro-psychomotor development follows different evolutionary phases that successive one another with different phases in each child. The achievement of a normal psycho-intellectual development is the main goal of the treatment of Congenital hypothyroidism(CH).

The objective of our study was to evaluate the neuro-psychomotor development and intellectual ability in a group of 127 (74F, 53M) children with permanent CH.

Methods: CH children have carried out: 62.2% assessment of Global Functioning, with analysis of functional acquisitions; 34.6% test with Brunet-Lezine test in order to evaluate the quotient of motor skills, correlated to chronological age; 33.8% a proper reagent Level for assessing the standard of their Intellectual

Results: 7,6% of children that had assessment of Global Functioning showed an initial slowing of the neuro-muscular development that was confirmed in 2.5% of this children. To the children that had valuation with Brunet-Lezine test 2.3% had moderate delay and 2.3% mild delay; this date was confirmed in 2.3% of children. 4.6% of children had mild delay at the valuation with reactive intellectual level, confirmed in all children. There was no difference between genders.

Conclusions: Most of our patients presented psychomotor and intellectual development in the normal range, only 7.8% of CH children had initial inadequate levels of development (mild or moderate) that was confirmed, at follow-up, only in 3.9% of children, all mild delay. These data represent the clear advantage given by early diagnosis and subsequent therapy in children with CH, which ensures complete somatic and neuro-psychic-motor development of the children with this endocrine disorder.

1100

PRESENTATION, CLINICAL AND GENETIC OUTCOMES IN A SERIES OF INFANTS WITH CONGENITAL HYPERINSULINISM

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Background & aims: Congenital hyperinsulinism (CHI) is a rare condition but a significant cause of recurrent hypoglycaemia in infancy and childhood. Prompt recognition and appropriate management is

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