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.

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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.

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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