Introduction
PHHI is the most common and severe forms of hyperinsulinemic hypoglycemia in neonates and infants. PHHI is a genetic disorder with familial and sporadic forms, both of which are characterized by dysregulation of insulin secretion occur in the neonates. In recent study described genetic abnormalities in nine genes (ABC8, KCNJ11, GCK, SLC16A1, SUR, HNF1A, HNF4A, and UCP2) that lead to the congenital forms of HH (1). The most severe forms of CHH are due to defects in the genes (ABC8 and KCNJ11) (2,3). In PHHI the histological abnormalities in pancreatic structure grouped into two categories: The focal lesion has abnormally islet cells; the majority of focal CHH due to heterozygous paternally inherited mutation in the ABC8 or KCNJ11 gene, account for almost 30–40% of all CHH cases. Focal CHH is usually confirmed by a fluorine-18 dihydroxyphenylalanine–position emission tomography (18F-DOPA-PET) scanning. Surgical resection of the lesion usually resolves HH. Diffuse CHH, the diffuse pancreas affects all the pancreatic β-cells. Patients with diffuse CHH either have a homozygous recessive or a compound heterozygous mutation in their KATP channel genes. This form of CHH accounts for 60–70% of all CHH cases, require a near total pancreatectomy (4).

RESULTS
• Mean birth weight 4 kg.
• Mean age of presentation 3 weeks except one patient was presented at 6 year.
• 10 pts (71%) having first cousin of consanguinity.
• 3 patients have had family history of siblings died from neonatal hypoglycemia.
• Initial Blood sugar 2mg–2.2mg/dl.
• Mean insulin levels 21.6 µu/ml in presence of hypoglycemia.
• Insulin: glucose ratio was 0.75–8.
• All patients underwent subtotal to near-total pancreatectomy except one patient.
• Histological Findings:
  • Focal form in 6 patients (43%) with insulin level > 20 µu/ml. Blood sugar ≤2mg/dl.
  • Diffuse form in 8 patients (57%) with insulin level ≤20 µu/ml and Blood sugar >12mg/dl.
  • Focal (3 M, 3 F), Ratio 1:1. Diffuse (6 M, 2 F), Ratio 3:1. Overall (9 M, 5 F) with male to female ratio was (1.8:1).
  • Large size baby with focal lesion and small or average sized baby with diffuse hyperplasia.
  • Focal lesion is more severe than the diffuse one.
• Two male infants presented with severe form of hypoglycemia earlier age 1–7 days of life with blood sugar 2–4 mg/dl, and had greater birth weight (4.5 kg & 5.7kg) their mothers were not suffering from gestational diabetes, they didn’t respond to medical therapy they found to have had multifocal adenomatosis in their pancreases.
• Diabetes Mellitus developed in 2 cases (14%), 1 focal and other one diffuse, with mean age 5 year.
• Unfavorable neurological outcome in 2 pts due to late intervention, because their family rejected operation.
• Neurologic & Psychomotor retardation in 4 patients (29%) & ADHD in one patient.

CONCLUSIONS
• Early recognition, diagnosis & treatment to prevent or minimize neurological damage.
• Fetal education & long term Follow-up to detect recurrence and detection of DM.
• A genetic studies considered as a useful parameter to determine cause and for genetic counseling.
• New drugs should be available.
• Preoperative investigations will limit pancreatectomy & prevent post operative diabetes.

METHODS
We have reviewed 14 infants (9 male & 5 female) who presented with severe recurrent non ketotic hypoglycemia, in the period between (1996–2013), the mean age of presentation 3 weeks (1 days-3 months) except one patient was diagnosed at 6 years of his age. The diagnosis of primary form of congenital hyperinsulinemic hypoglycemia was confirmed by laboratory investigations. Analysis of data regarding the time & mode of presentation, birth history, family history, consanguinity, Initial blood sugar levels, Insulin levels. Insulin to glucose ratio, genetic analysis, management, histopathology & outcome of the patients were studied.

Table: clinical evaluation of the patients

<table>
<thead>
<tr>
<th>PT NO</th>
<th>SEX</th>
<th>BW(kg)</th>
<th>Age at onset</th>
<th>Current age</th>
<th>Insulin µu/ml</th>
<th>Initial BS mg%</th>
<th>Insulin/glucose ratio</th>
<th>Age at operation</th>
<th>Histopathology</th>
<th>Mental status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>3.3</td>
<td>6yr</td>
<td>21yr</td>
<td>26</td>
<td>14</td>
<td>1.85</td>
<td>7yr</td>
<td>Fo</td>
<td>MR</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>3.8</td>
<td>3yr</td>
<td>12yr</td>
<td>26</td>
<td>18</td>
<td>1.85</td>
<td>9 M</td>
<td>Fo</td>
<td>MR</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>5.7</td>
<td>4yr</td>
<td>21yr</td>
<td>17</td>
<td>12</td>
<td>1.85</td>
<td>9 M</td>
<td>Fo</td>
<td>MR</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>4.2</td>
<td>2yr</td>
<td>12yr</td>
<td>26</td>
<td>18</td>
<td>1.85</td>
<td>2 D</td>
<td>Fo</td>
<td>MR</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>1.5</td>
<td>3yr</td>
<td>12yr</td>
<td>26</td>
<td>18</td>
<td>1.85</td>
<td>9 M</td>
<td>Fo</td>
<td>MR</td>
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Figure a: male to female, focal to diffuse & over all ratios

Histopathology

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<thead>
<tr>
<th>Focal</th>
<th>Diffuse</th>
<th>Overall Mean F to M ratio</th>
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<td>60%</td>
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References