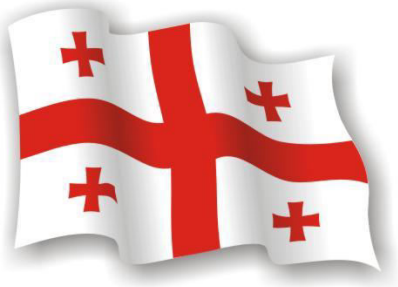

Patients with Congenital Hyperinsulinism in resource limited settings – Challenges for the physicians

Nino Kheladze.MD
M. Iashvili Children's Central Hospital
Tbilisi. Georgia

Patient No 1, Nini 2008

- Episodes of stillness, eye fixation, mainly during or after feeding since the age of 40 days.
- Hypoglycemia conformed and hospitalized with seizures at the age of 3.5 month.
- Blood glucose levels from un-measurable to 50 mg/dl, Insulin – 17. 82 mIU/ml
- Had several different regimes of feeding and infusion (40 % glucose) for maintaining glucose concentration in blood over 50mg/dl.
- Diazoxide, glucagon and octreotide were not used, because they were not available.

Nini 2008

- Went for further treatment in Charite-Berlin Medical University, Institute of Pediatric Endocrinology
- Had PET-Scan and surgery for focal form of hyperinsulinism in the head of pancreas.
- Now is free of symptoms, attending school and has age appropriate and good quality of life.

Patient No 2, Valeri. May 2015

Valeri 9 month old

- Seizures and hypoglycemia form 7 week of life
- Hospitalized twice, seen by multiple doctors including Pediatric Endocrinologist, “Critical Sample” not obtained.
- Very frequent feeding, “addicted” to juice, overweight
- Non ketotic, recurrent hypoglycemia, with altered consciousness and seizures, increased frequency of episodes, “normal” insulin with low BG.
- Genetic testing performed in Exeter University- No mutation (ABCC8, KCNJ11)

- Originally got Proglycem MSD 25 mg from patient who no longer needed it(had transitory HI)
- Later was supplied by Georgian doctor working in USA (Manufactured by TEVA)
- Currently needs – 30 mg /3 times per day. Has about 20 days supply left.
- No official and constant way to get more medicine.
- No glucagon-for emergency

Patient No 3,Nene April 2016

Nene April 2016

- Severe hypoglycemia from day two in otherwise healthy newborn
- Hospitalized in the NICU, multiple complications, including sepsis and NEC
- Not adequate response on Diazoxide (borrowed form Valeri), some response on Octreotide + continuous glucose infusion
- No glucagon, had several episodes of immeasurable blood glucose level.

And miracles do happen

- Genetic Testing in Exeter - ABCC8 mutation inherited from her father
- Advise from Julie Raskin- referral to Dr. Henrik Christesen
- PET-Scan, treatment and Surgery in Odense University, Denmark.
- Nene is cured, so far with no signs of neurological problems, growing well.

Many thanks and Best Regards from Nene and her family

Patient No 4,Nino July 2016

Patient No 4,Nino July 2016

- Symptoms of severe hypoglycemia from day 1.
- Diagnosis of hyperinsulinism – 1.5 month old
- Good response on Diazoxide.
- Getting Proglycem/ MSD 100 mg over the counter from Russia, later from Italy. Now needs a new source.
- Has clinical and MRI sings of brain damage
- No Glucagon
- Genetic Testing in Exeter- negative for GLUD1, ABCC8, KCNJ11

Main Problems - 2016

- Need for increased Awareness
- Availability of resources, mainly essential medications
- Clear guidance documents for medical specialist
- Need for established reference centers and specialists
- Financial considerations

Now - 2018

- All 5 patients are quite well, 3 are receiving moderate doses of diazoxide
- Essential medications still not available
- Still super hard to get diazoxide
- No new cases – good or bad?
- Still low awareness
- But we know where to get great help!

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Thank you!