Poster P2-078

A case of an infant with congenital hyperinsulinism complicated by diabetic ketoacidosis during treatment

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INTRODUCTION

Congenital hyperinsulinism (CHI) is a disorder causing persistent hypoglycemia due to oversecretion of insulin. Diazoxide, a KATP channel opener in pancreatic beta cells is the treatment of choice, however, the glucose level should be monitored carefully. We report here a case of an infant girl with CHI who was complicated by diabetic ketoacidosis (DKA) during acute febrile illness.

CASE DESCRIPTION

INITIAL VISIT

- **Age/Sex :** 15-month/Female
- **Chief complain** : Recurrent seizure
- **Birth history** : 38weeks, 3100g
- **Past history**
  - Recurrent afebrile seizures since 4 months of age
  - Delayed development with unknown cause
    (Gross motor: 9-month, Fine motor/Adaptive: 8-month,
    Language: 9-month, Personal-Social: 12-month)
- **Family history** (-)
- **Physical examination**
  - **Weight/Height** : 10.5 kg/78.5 cm (both 50-75 percentile)
- **Evaluation** for delayed development
  - Brain magnetic resonance imaging (MRI) : Normal
  - Thyroid function test (TFT) : Normal
  - Tandem mass screening : Normal
- **Laboratory Test**

Serum glucose (mg/dL)	**Insulin** (uIU/mL)	**C-peptide** (ng/mL)
< 30	8.36	1.97

- **Diagnostic criteria** of CHI

✓ **Glucose infusion rate (GIR)** to maintain euglycemia
: **8-10 mg/kg/min**

✓ **Glucagon stimulation test**

	Before glucagon	**After glucagon**
Serum glucose (mg/dL)	38	**76**
Insulin (uIU/mL)	7.6	4.4

➢ **Diagnosis : Congenital Hyperinsulinism (CHI)**

- Further evaluation
- **Abdominal ultrasonography** : Normal
- **Targeted gene pannel including** KCJ11, *ABCC8, KCNJ11, GLUD1, HNF4A, GCK, HADH,* and *UCP2* : all negative
- Whole Exome Sequencing (WES) : negative
  → Under consideration of the 2nd inspection

➢ **Treatment :** Diazoxide (3mg/kg/day)

After discharge, her self-monitored blood glucose level was well-controlled on follow up visit.

AFTER 4 WEEKS

- **Chief complain** : 2 days of fever and hyperglycemia
- Taking diazoxide was skipped during 2 days due to fever
- **Laboratory Test**

Serum glucose (mg/dL)	**Blood gas analysis** [pH-pCO2-HCO3-(BE)]	**Serum ketone**
398	**7.293-16.6-7.9-(-16.6)**	**+**

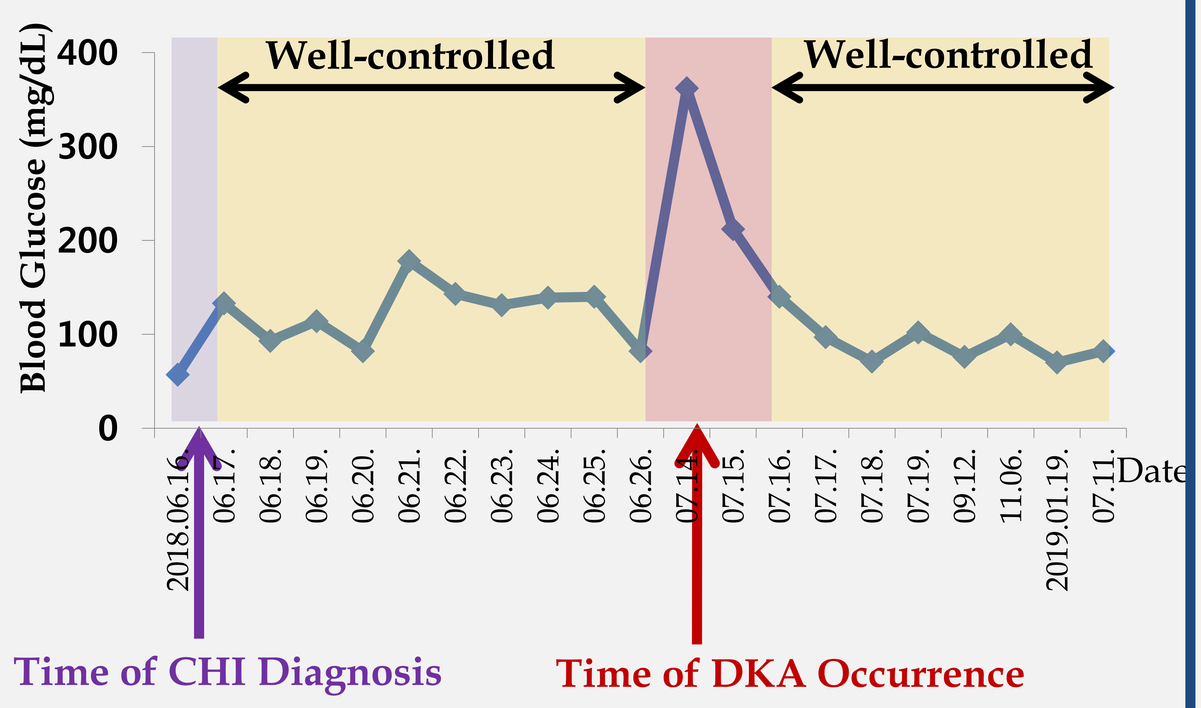
➢ **Diagnosis : Diabetic Ketoacidosis (DKA)**

➢ **Treatment :** Continuous intravenous insulin infusion with the rate of 0.05 IU/hr/kg

AFTER RECOVERY FROM DKA

- **Under the same dosage of diazoxide,**
  - Well-controlled serum glucose level
  - Improved developmental delay with rehabilitation therapy
    (Gross motor: Standing with holding bar is possible,
    Fine motor: minimum 8-month ~ maximum 18-month,
    Mental evaluation on Bayley test : 13-month)

TREND OF BLOOD GLUCOSE LEVEL



CONCLUSIONS

We present a rare case of CHI complicated by DKA during treatment. If the patients with taking diazoxide are under febrile condition, their blood glucose level and status should be monitored more carefully and tightly than usual.