Diagnosis and treatment of persistent hyperkalemia in newborn twins
----Rare case report of Gordon syndrome

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Background: hyperkalemia is one of the clinical critical situations. Neonatal hyperkalemia is common within 3 days after birth especially in the preterm neonate whose gestational age is less than 32 weeks or birth weight is less than 1.5 kg. But persistent hyperkalemia is rare in neonate. Gordon syndrome, also known as pseudo-hypoaldosteronism type II, is one of the cause of persistent hyperkalemia. The onset of it is also rare in the neonatal stage.
Objectives: To summarize the diagnosis and treatment course of hyperkalemia in twins, review the diagnosis approach of hyperkalemia in neonate and guide clinical practice.
Methods: The clinical manifestation, laboratory examination, diagnosis and treatment of the two cases were summarized. Review the literatures of the diagnostic process of hyperkalemia in neonate.
Results: A pair of twin girls, 38 days after birth， came to our department because of “Hyperkalemia more than one month”. The twins were MCDA, with the gestational age 37 weeks. The birth weight of B1 and B2 was 1.95 kg and 1.59 kg respectively. Postnatal routine biochemical examination showed that increasing of serum potassium was found. After the treatment of furosemide diuretic, alkali supplementation, intravenous insulin and glucose, serum potassium decreased temporarily, but increased again after withdrawan. Laboratory test in local hospital showed that arterial blood gas analysis, urine routine and renal function were normal(both creatinine and urea nitrogen), Doppler ultrasound of kidney and adrenal gland were normal. For further diagnosis and treatment, the twins were administrated in NICU of our hospital. Since the onset of the disease, no vomiting or diarrhea.
Mother of the twins was found high blood pressure and elevated blood potassium at the time of pregnancy without any examination or treatment.
When admitted to our hospital, the physical examination showed that B1： Wt 3.05kg， BP 93/46mmHg， B2:Wt 2.73kg， BP93/61mmHg. According to the family history of hyperkalemia and no dehydration, the twins gained 1.1kg and 1.2 kg body weight on 38 days after birth, their blood pressure increased, serum renin decreased obviously, so they were clinically diagnosed as Gordon syndrome (pseudo-hypoaldosteronism type II). B1 and parents' KLHL3 gene sequence were analyzed simultaneously. B1 and mother had a same heterozygous c. 230c > a mutation in exon 3 of the KLHL3 gene in chromosome 5, conforming the diagnosis of Gordon syndrome. The results are shown in figure 1. Twins were treated with hydrochlorothiazide 2mg/kg.d orally. Serum potassium and blood pressure decreased gradually .The onset of most of the Gordon syndrome is scholar age to adolescence, while that of our two cases is the neonatal stage, which widened the clinical manifestation spectrum of Gordon syndrome.

Table 1 Results of laboratory examinations of B1 and B2 (2017-11, our hospital)

Content	Serum potassium （mmol/L）	Serum sodium (mmol/L)	Serum chlorine (mmol/L)	Serum calcium (mmol/L)	Serum magnesium (mmol/L)	Creatinine (umol/L)	Blood PH	BE (mmol/L)
B1	6.41	138	111	2.4	0.73	24	7.38	-3.2
B2	6.48	140	115	2.34	0.70	28	7.37	-4.5

Content	Urine PH	Renin (pg/ml) (4-24)	Aldosterone (pg/ml) (10-160)	ACTH (pmol/L)	Cortisol (ug/dl)	17（OH）P (ng/ml)	Andros -tendione (nmol/L)	DHEA (umol/L)
B1	7.5	1.7	156	12.1	17.9	3.93	5.53	23.97
B2	7.0	2.9	167	13.5	18.0	10.8	23.60	29.16

Figure 1 gene sequences of B1 and parents