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(Citation)
Pediatrics and Neonatology,62(1):119-121

(Issue Date)
2021-01

(Resource Type)
journal article

(Version)
Version of Record

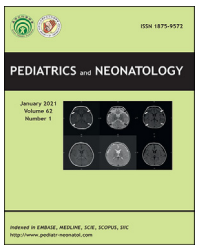
(URL)
https://hdl.handle.net/20.500.14094/90008067

Pediatrics and Neonatology (2021) 62, 119—121

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: http://www.pediatr-neonatol.com



Letter to the Editor

Three cases of pseudohypoaldosteronism following ileostomy in preterm infants

Dear Editor,

Pseudohypoaldosteronism (PHA) is a disorder caused by renal tubular resistance to aldosterone and is characterized by hyperkalemia, hyponatremia, metabolic acidosis, and high plasma renin and aldosterone levels.[1] In general, PHA is classified as "primary" when it is caused by genetic mutations and "secondary" when it is associated with urinary tract malformations and/or urinary infections. Secondary PHA usually occurs in infants.[2] A few adult cases of secondary PHA caused by ileum resection have been reported in the literature,[3,4] but none of the newborns have reported ileum resection—induced PHA. Herein, we describe three preterm infant cases of secondary PHA following ileostomy without renal complications.

The gestational age and birthweight of the patients ranged from 24 to 29 weeks and 568—706 g, respectively. Ileostomy was performed on days 6, 9, and 3 for cases 1, 2, and 3, respectively. PHA occurred following possible trigger events associated with the decrease of Na supply or volume depletion on the days 21, 144, and 74 in cases 1, 2, and 3, respectively. The onset of PHA with initial symptom for each case was on day 22 with oliguria, day 145 with cardiac arrest, and day 96 with no symptoms, respectively. Blood testing at the onset revealed hyponatremia, hyperkalemia, and high plasma renin and aldosterone concentrations in all the cases. Abdominal ultrasonography and urinalysis revealed no renal abnormalities or urinary tract infections in all the cases. Additionally, congenital adrenal hyperplasia was excluded because of the negative results of the mass screening tests. Hyponatremia/hyperkalemia was normalized immediately after the bolus infusion of normal saline with/without glucose-insulin therapy, which was replaced by the oral supplementation of sodium chloride (NaCl). In cases 1 and 3, NaCl supplementation could be completed one month after the ileostomy closure, and hyponatremia did not relapse afterward. In case 2, NaCl supplementation was continued until two years of age at the discretion of the attending physician, and genetic analysis of the causative genes of primary PHA was performed; however, no mutation was found (Table 1). Based on the above clinical description and blood test results, we diagnosed all the cases with secondary PHA following ileostomy; however, the late status of severe hypovolemia—induced secondary hyperaldosteronism, resulting in hyperkalemia and persistent metabolic alkalosis, should also be considered in the cases resembling cases 1 and 3.

Regarding the mechanism of secondary PHA following ileostomy in adults, Vantyghem et al. hypothesized that excessive fecal sodium losses result in chronic Na depletion with the contraction of the plasma volume and severe secondary hyperaldosteronism, which might trigger PHA.[3] Conversely, Niyazov et al. speculated that a chronic increase in the aldosterone levels may induce the downregulation of the mineralocorticoid receptor, thus contributing to the development of secondary PHA.[4] Furthermore, preterm infants have been reported to be susceptible to hyponatremia despite their increased renin—angiotensin—aldosterone system, possibly because of tubular immaturity.[5] Therefore, we consider that preterm infants with ileostomy potentially have renal tubular resistance to aldosterone and are at a risk of developing secondary PHA triggered by profound volume depletion.

In conclusion, careful observation is necessary for the preterm infants who undergo ileostomy, as they might be at a risk of developing secondary PHA, even in the absence of renal complications.

https://doi.org/10.1016/j.pedneo.2020.09.006

Table 1 Clinical characteristics of the three cases.

	Case 1	Case 2	Case 3
Clinical background and ileostomy management			
Sex	Female	Male	Male
Gestational age (weeks)	24 4/7 weeks	24 6/7 weeks	29 6/7 weeks
Birthweight (g)	568	706	658
APGAR Scores at 1 min/5 min	3/5	5/9	6/8
Intestinal complications	Idiopathic pneumoperitoneum	Ileoileal intussusception	Meconium ileus
Ileostomy operation (day)	6	9	3
Start of enteral feeding (day)	12	16	7
Achievement of full feeding (day)	23	29	15
Ileostomy closure (day)	131	194	128
BUN before onset (mg/dL)	25.5 on day 17	12 on day 144	2 on day 77
Creatinine before onset (mg/dL)	0.67 on day 17	0.25 on day 144	0.32 on day 77
Characteristics and management of PHA			
Onset (day)	22	145	96
Possible trigger events	Reduction of intravenous Na supplementation on day 21	Prolapse of intestinal fistula on day 144	End of oral Na supplementation on day 74
Initial symptoms	oliguria	cardiac arrest	none
Treatment	Na supplementation	Na supplementation with GI therapy	Na supplementation with GI therapy
End of Na supplementation (days)	165	1020	157
Duration of Na supplementation (months)	5	26	2
Renal complications	None	None	None
Genetic examinations	Not performed	No mutations in NR3C2	Not performed
Blood test results at the onset of PHA			
Na (mEq/L)	124	133	121
K (mEq/L)	6.4	9.8	7.3
Cl (mEq/L)	94	99	86
Plasma renin levels	27,100 (pg/mL)	>20 (ng/mL/h)	>20 (ng/mL/h)
Serum aldosterone levels (pg/mL)	4480	9190	15,000
BUN (mg/dL)	36.2	16.0	17.0
Creatinine (mg/dL)	1.02	0.64	0.26
pH	7.425	7.045	7.422
Base excess (mmol/L)	3.4	−10.8	3.4

PHA; Pseudohypoaldosteronism, GI therapy; glucose-insulin therapy, BUN; blood urea nitrogen.

Declaration of competing interest

None.

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Jul 9, 2020