

# A Rare Cause of Hyponatremia in a Child with Hirschsprung Disease Operated with an Ileostomy

## INTRODUCTION

Pseudohypoaldosteronism (PHA) is a rare condition characterized by hyponatremia, hyperkalemia, metabolic acidosis and high plasma aldosterone level. PHA secondary to gastrointestinal losses is under-reported in the pediatric population

## AIM

We present a rare case with secondary PHA due to excessive gastrointestinal losses through ileostomy. Additionally, we present the unique association of PHA with thrombocytosis, which may be used as a useful predictor of an impending hypovolemic shock.

## METHOD

A 1.5 year-old-male child, who was diagnosed with Hirschsprung disease at the age of 2 months with ileostomy, presented 2 weeks postoperatively with hypovolemic shock. This was preceded by excess gastrointestinal fluid losses through ileostomy. Laboratory tests showed picture of PHA. Hormonal work up was sent and patient was started on hydrocortisone, sodium supplement with intravenous fluid and sodium polystyrene for hyperkalemia.

## RESULTS

Hormonal work up showed picture of PHA (table 1). Hydrocortisone was tapered and discontinued. Depletion of sodium and water, leads to activation of renin-angiotensin system, decreasing urinary sodium excretion. Subsequently, aldosterone can no longer facilitate potassium excretion in presence of excess sodium loss; leading to hyperkalemia. It was noticed that patient developed thrombocytosis at presentation. Activation of sympathetic nervous system during hypovolemic shock maintains vascular stability and causes thrombocytosis. At the age of 14 months, Duhamel pull-through operation was done with closure of ileostomy, with maintenance of normal electrolytes and normal aldosterone level.

	During episode of salt-losing crisis	After closure of ileostomy	Normal range
Na (mEq/L)	126	137	135-145
K (mEq/L)	7.0	5.1	3.5-5.8
Aldosterone (ng/dL)	573.6	86	7-99
Renin (ng/mL/hr)	5.8	-	2.4-37
Platelets (k/mm <sup>3</sup> )	819	297	150-400
17-OH-progesterone (nmol/L)	3.5	-	< 10
Cortisol (nmol/L)	407.9	-	81.9-957.9
ACTH (pg/mL)	7.3	-	5-46

Table 1: Laboratory results for patient during hyponatremia and after closure of ileostomy

## CONCLUSIONS

Pseudohypoaldosteronism should be included in the differential diagnosis when evaluating hyponatremia especially in those with excessive fluid losses. Another interesting finding is the presence of thrombocytosis, which can be used as an important predictor of an impending hypovolemic shock.

## REFERENCES

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## ACKNOWLEDGEMENTS

None.

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