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Recurrent tetany in the course of metabolic alkalosis in patient with short bowel syndrome

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Occurrence of tetany often requires consultation by an endocrinology specialist. We would like to present a case of a patient whose symptoms where just "tip of the iceberg" for underlying, very serious disorders, which had their beginning many years earlier when she was diagnosed with thrombophilia.

A 35-year-old woman was admitted to the emergency department due to tetany and dehydration symptoms. She had been previously diagnosed with idiopathic thrombocytosis, which was complicated 1 year prior with portal vein thrombosis. She was treated with thrombolytic therapy (alteplase) but eventually underwent surgery due to intestinal necrosis. The caecum and the distant part of the ileum were resected and an ileostomy was formed. Shortly after surgery she required parenteral nutrition, but by the time of her discharge oral nutrition was introduced.

During her ER consultation the patient admitted that she had excessive ileostomy drainage, which she evaluated for 3500 mL per day, and that she was unable to compensate it accordingly.

On admission the laboratory tests showed increased creatinine concentration, hypochloraemia, hyponatraemia, and the level of calcium ion (Ca^{2+}) decreased to 0.9 mmol/L with metabolic alkalosis (Tab.1 — No. 1). After treatment with intravenous calcium, and repletion of fluids and other electrolytes, the patient obtained both clinical and biochemical improvement (Tab. 1 — No. 2, 3).

She was discharged with a diagnosis of acute kidney injury, dehydration, dyselectrolitaemia, metabolic alkalosis, and tetany in the course of short bowel syndrome (SBS).

Despite some modifications in her treatment, within 5 days the patient was once more admitted to the emergency department with tetany and alkalosis (Tab. 1 — No. 4, 5). After symptomatic treatment, she was transferred to the surgery department and qualified to ileostomy closure and ileocolic anastomosis. As a result, her condition gradually improved, and by the time of discharge dyselectrolitaemia, acid-base disorders and kidney insufficiency were no longer present (Tab. 1 — No. 6, 7).

Short bowel syndrome is a cause of intestinal insufficiency or intestinal failure, which leads to malabsorption, diarrhoea, and dehydration. If left untreated, it can cause severe complications, including death [1]. In Poland it affects around 6 people per million.

The most common causes of SBS are mesenteric ischaemia, intestine resections in the course of other conditions (inflammatory bowel diseases, neoplasms, injuries), intestinal fistulas, and severe malabsorption syndrome caused by celiac disease or different enteritis.

There are few reports on metabolic alkalosis in patients with short bowel syndrome, although these patients are likely to develop electrolyte and acid-base disorders especially when increased ileostomy drainage is present [2, 3].

Metabolic alkalosis is a disorder defined by increased blood pH (> 7.43) combined with high biocarbonate (HCO_3^-) (> 26 mmol/L) concentration. Its pathophysiology requires specific clinical settings: primary base increase ($HCO3^-$) and/or acid loss (H^+) combined with renal failure to excrete HCO_3^- [3, 4].

In the case shown above, excessive water and electrolyte loss caused not only dehydration with acute kidney injury but also sodium chloride (NaCl) deple-

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Table 1. *Laboratory test results*

	No. 1	No. 2	No. 3	No. 4	No. 5	No. 6 (post-surgery)	No. 7 (discharge)	Reference range
pH	7.72	7.49	7.42	7.59	7.58	7.32		7.32–7.43
pO_2 [mm Hg]	26.1	40.6	25.3	38.7	15.8	17.1		75–100
pCO_2	35.2	55.3	40.1	45.0	56.4	64.6		35–45
K^+ [mmol/L]	4.0	3.0	3.9	2.7	3.8	5.0	3.5	3.5–5.5
Na^+ [mmol/L]	131	133	132.4	137	137	137	136	136–146
Ca^{2+}[mmol/L]	0.91	1.33	1.23	0.99	1.21	1.22		1.15–1.33
Cl^- [mmol/L]	71	76	88	–	71	102	107	98–107
HCO_3^-[mmol/L]	48.1	40.1	38.3	43.8	51.1	33.2		21–25
BE [mmol/L]			11,2					–2.5–2.5
Creatinine [umol/L]	442	427	347	328	328	150	54	50–98

tion, which is crucial for alkalosis persistence. A protein called pendrin, a Cl-HCO_3 transporter in the distal part of the nephron, is responsible for HCO_3^- secretion. However, a shortage of Cl^- (that was already absorbed in the Henle loop) in the collecting duct prevented Cl^--HCO_3^- exchange and impaired HCO_3^- elimination. In such cases it is crucial to correct the Cl^- deficit to restore bicarbonate excretion and normalise blood pH [4, 5].

Other potential pathomechanisms of acquiring and sustaining metabolic alkalosis include the following: gastric acid loss with intestinal fluid [3], activation of renin–aldosterone–angiotensin (RAA) axis by hyponatraemia and low blood pressure, hypokalaemia, and hypomagnesaemia. Aldosterone enhances the reabsorption of sodium by replacing it with kalium and hydrogen ions. That not only creates alkalosis by eliminating hydrogen ions but also aggravates it by causing hypokalaemia. Hydrogen ions from extracellular fluid are exchanged for potassium, which results in the state of intracellular acidosis. In addition, it increases renal ammoniogenesis and reabsorption of bicarbonates, which further consolidates alkalosis [4].

This entity may have a poor prognosis depending on its cause and underlying disorders. It leads to hypoventilation, hypoxia, decreased cerebral blood flow, decreased myocardial contractility, arrhythmias, and increased neuromuscular excitability such as tetany episodes.

Tetany, a condition of involuntary muscle spasm caused by a deficiency of calcium ions (Ca^{2+}), can affect all muscle groups, causing painful cramps, but it can also lead to life-threatening bronchi and laryngospasm. In differential diagnosis we must consider all causes of calcium depletion, such as malabsorption or high urine calcium excretion, which include: hypoparathyroidism, deficiency or disorders of vitamin D3 metabolism, use of loop diuretics, calcium deposition in soft tissues (acute pancreatitis) or bones (hungry bone syndrome, use of bisphosphonates or denosumab), as well as conditions with hyperphosphataemia (tumour lysis syndrome, high-phosphate diet, Mg deficiency).

In this patient's case, tetany was due to alkalosis, which reduced the concentration of Ca^{2+} in favour of albumin-bound calcium causing neuromuscular irritability. Understanding the complexity and background of this disorder allowed to implement successful symptomatic and causal treatment that led to final recovery.

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Conflict of interest
None declared.

References

1. Pironi L. Definitions of intestinal failure and the short bowel syndrome. Best Pract Res Clin Gastroenterol. 2016; 30(2): 173–185, doi: 10.1016/j.bpg.2016.02.011, indexed in Pubmed: 27086884.
2. Weise WJ, Serrano FA, Fought J, et al. Acute electrolyte and acid-base disorders in patients with ileostomies: a case series. Am J Kidney Dis. 2008; 52(3): 494–500, doi: 10.1053/j.ajkd.2008.04.015, indexed in Pubmed: 18562060.
3. Masuyama S, Nagatoya K, Kawaoka T, et al. Metabolic alkalosis due to short bowel syndrome in a hemodialysis patient. CEN Case Rep. 2020; 9(2): 162–164, doi: 10.1007/s13730-020-00445-1, indexed in Pubmed: 31955390.
4. Gillion V, Jadoul M, Devuyst O, et al. The patient with metabolic alkalosis. Acta Clin Belg. 2019; 74(1): 34–40, doi: 10.1080/17843286.2018.1539373, indexed in Pubmed: 30369299.
5. Wall SM, Lazo-Fernandez Y, Wall SM, et al. The role of pendrin in renal physiology. Annu Rev Physiol. 2015; 77: 363–378, doi: 10.1146/annurev-physiol-021014-071854, indexed in Pubmed: 25668022.