Perioperative Infusion Management for Adhesive Bowel Obstruction with Congenital Nephrogenic Diabetes Insipidus: A Case Report

Wataru Miyauchi,* Tomoyuki Matsunaga,* Yu Sakano,* Masahiro Makinoya,* Shota Shimizu,* Kozo Miyatani,* Yuji Shishido,* Teruhisa Sakamoto* and Yoshiyuki Fujiwara*

**Division of Gastrointestinal and Pediatric Surgery, Department of Surgery, School of Medicine, Faculty of Medicine, Tottori University, Yonago 683-8504, Japan*

ABSTRACT

Congenital nephrogenic diabetes insipidus (CNDI) is a rare disease that results in polyuria due to decreased responsiveness to the antidiuretic hormone in the collecting ducts of the kidney. Without compensation by drinking large amounts of water, dehydration and hypernatremia can rapidly develop. We present a case of a patient originally diagnosed with CNDI who required surgery and a fasting period due to adhesive bowel obstruction. The patient was a 46-year-old man who was originally diagnosed with CNDI. He was prescribed trichlormethiazide but self-discontinued treatment in the process. His normal urine output was about 7,000–8,000 mL/day. He underwent robot-assisted radical cystectomy and uretero-cutaneostomy for bladder cancer. Two years later, he was hospitalized due to adhesive bowel obstruction. A 5% glucose solution was infused, and the dose was adjusted according to the urine volume and electrolytes. An adhesiotomy was performed due to recurrent bowel obstruction in a short period of time. A 5% glucose solution was used as the main infusion during the perioperative period. Once drinking water was resumed after surgery, urinary output and electrolytes were easily controlled. In conclusion, patients with CNDI should be given a 5% glucose solution as the primary infusion, and the infusion volume should be adjusted by monitoring daily urine output, electrolytes, and blood glucose levels. Infusion management is easier if oral intake is initiated as early as possible.

Key words diabetes insipidus; nephrogenic; infusions; intravenous; perioperative period

Diabetes insipidus is a disorder of the synthesis and secretion of antidiuretic hormones or their action in the kidneys, resulting in large amounts of diluted urine.[1] In central diabetes insipidus, the secretion of the antidiuretic hormone from the pituitary gland is inhibited,[2] In nephrogenic diabetes insipidus, the response of renal collecting duct cells to the antidiuretic hormone is impaired.[3, 4] Nephrogenic diabetes insipidus can be classified as congenital or acquired. Most are acquired due to lithium treatment.[4] Congenital nephrogenic diabetes insipidus (CNDI) is a rare genetic disorder, with a report of 8.8 cases of X-linked recessive form of the disease per million male births.[5] Nephrogenic diabetes insipidus treatment includes thiazide diuretics, potassium-sparing diuretics, nonsteroidal anti-inflammatory drugs, and a low-sodium diet, all of which are expected to slightly decrease urine output. However, no specific treatment is currently available for this disease.[6] Without compensation by drinking large amounts of water, dehydration and hypernatremia can rapidly develop. However, few reports are available on patients with CNDI who, for whatever reason, are unable to rehydrate by oral intake.[7] We report a case of a patient originally diagnosed with CNDI who required surgery and a fasting period due to adhesive bowel obstruction with a review of the literature.

PATIENT REPORT

The patient was a 46-year-old man who was originally diagnosed with CNDI. He was prescribed trichlormethiazide in the past but self-discontinued treatment in the process. His normal urine output was about 7,000–8,000 mL/day. He underwent robot-assisted cystectomy and ureterostomy for bladder cancer at the age of 44. He had no other previous disease and no history of alcohol or allergic episodes. He has an older brother and a younger brother with diabetes insipidus.

The patient presented to the emergency room with complaints of abdominal pain and vomiting two years after the cystectomy. His abdomen was distended but soft and without rebound tenderness. The bowel sounds were weakly audible. Computed tomography showed bowel dilatation and caliber changes in the pelvic floor, with no closed loop (Fig. 1).

Blood tests showed only a mildly elevated inflammatory response and no other pathologically abnormal

Corresponding author: Tomoyuki Matsunaga, MD, PhD
matut0m0@tottori-u.ac.jp
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Abbreviations: CNDI, congenital nephrogenic diabetes insipidus; NG tube, nasogastric tube; POD, postoperative day

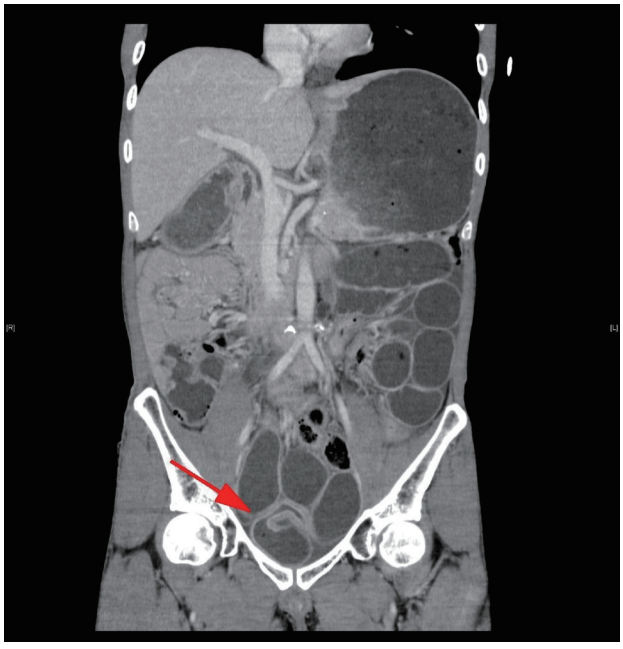

Fig. 1. Computed tomography findings at the time of consultation. Computed tomography showed bowel dilatation and caliber changes in the pelvic floor (red arrow), with no closed loop.

findings. Based on these findings, he was diagnosed with adhesive small bowel obstruction, and a conservative treatment policy was adopted.

For CNDI, the patient was referred to a nephrologist for consultation, and hydrochlorothiazide and desmopressin were decided to be used. Upon nephrology treatment resumption, blood osmolality and urine osmolality tests were performed, which showed diluted urine as expected. The test results showed urine sodium 10 mmol/L, urine potassium 9.3 mmol/L, urine chlorine 10 mmol/L, fractional excretion of sodium 0.3, fractional excretion of urea nitrogen 11.3, urine osmolality 103 mOsm/KgH2O. Vital signs and weight on admission were as follows. Body temperature 37.5 °C, heart rate 98 /min, blood pressure 120/82 mmHg, SpO2 94% (room air), weight 51.6 kg

Summary of posthospitalization progress

The nausea disappeared immediately after inserting a nasogastric tube (NG tube) to drain the stomach contents. As the symptoms of bowel obstruction disappeared over time with conservative treatment, feeding was resumed on day 9 of hospitalization, and the intravenous infusion volume gradually decreased. Hydrochlorothiazide tablets were administered after the start of oral intake.

On day 18 of hospitalization, bowel obstruction symptoms recurred. Because of the recurrence in a short period of time, an adhesiotomy was performed on day 20 of hospitalization. When oral intake was tried on day 23 of hospitalization, i.e., postoperative day (POD) 3, bowel obstruction symptoms appeared. Therefore, the patient had a sufficient intestinal rest period and resumed eating on day 30 of hospitalization (POD 10). After that, no obstruction was observed, and stable oral intake was possible. The intravenous infusion volume could be gradually reduced (Fig. 2). The patient was discharged from the hospital on day 52 of hospitalization (POD 32) because the urine volume was stable at the pre-hospital level without any electrolyte abnormalities even without infusion (Fig. 3).

Infusion management

Considering the patient's original urine output of 7,000–8,000 mL and the estimated insensible excretion and drainage from the NG tube, we first set the target infusion volume at about 10,000 mL per day. Based on previous literature,[8] a 5% glucose solution was mainly used as the infusion solution. Extracellular fluid and potassium preparations were used as needed. Each day, the infusion volume was adjusted based on urine output, NG tube drainage, and electrolytes. During the course of treatment, the maximum concentrations of electrolytes added for electrolyte level compensation were sodium 45 mEq and potassium 54 mEq. The goal of urine volume control is to achieve the same level of urine volume as in normal times. Once it was determined that oral intake was possible, in addition to the above, the infusion volume for the next day was set considering the amount of water consumed that day. As the amount of drinking water increased, the infusion volume slowly decreased.

Surgical findings of adhesiotomy

An adhesiotomy was performed through a lower abdominal midline incision. An obstruction due to adhesion of the ileum was found near the pelvic floor, and this adhesion was released. No resection of the intestine was performed. The operative time was 73 min. Intraoperative infusion volume was 1,700 mL, and urine volume was 1,100 mL. Almost no bleeding was observed.

DISCUSSION

This case was originally diagnosed with CNDI and required multiple fasting periods of approximately one week due to adhesive bowel obstruction. CNDI is a very rare disease,[5] Cases of neurological sequelae have been reported, but recently, more and more patients have been diagnosed early in life and have grown up without

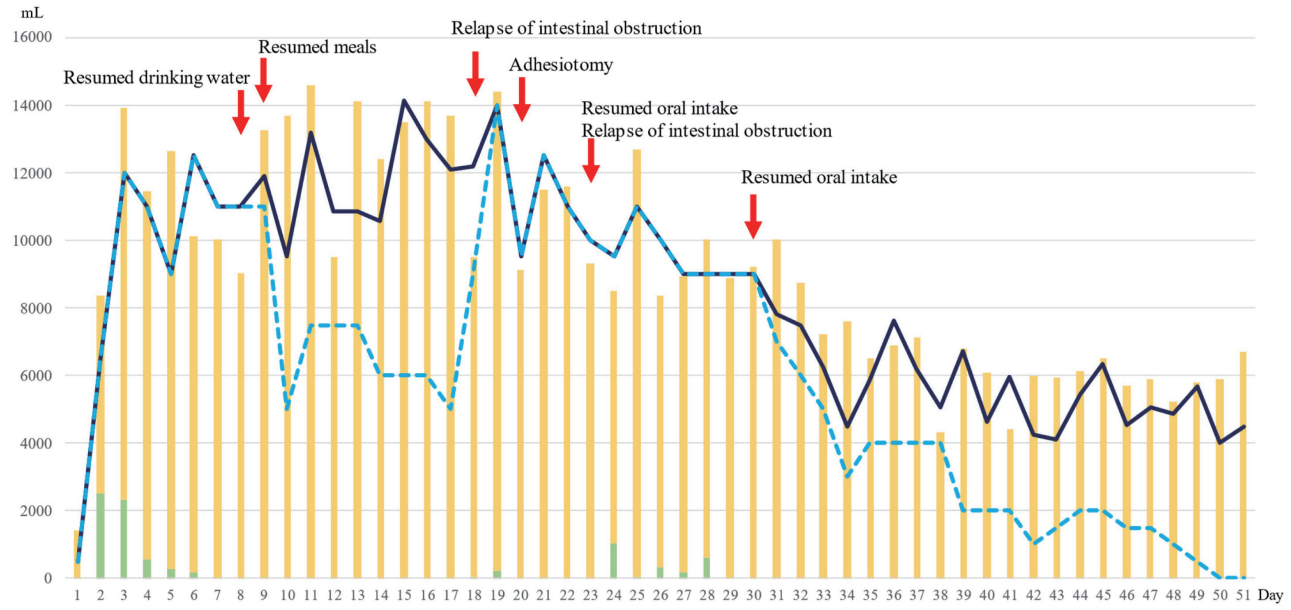

Fig. 2. Treatment progress and fluid IN/OUT balance. The solid line represents the total water supply, and the dotted line represents the infusion volume. The difference between the solid and dotted lines is the drinking water volume. The solid bar represents urine volume, and the shaded bar represents the amount of fluid drained from the nasogastric tube (NG tube). All units are in "mL".

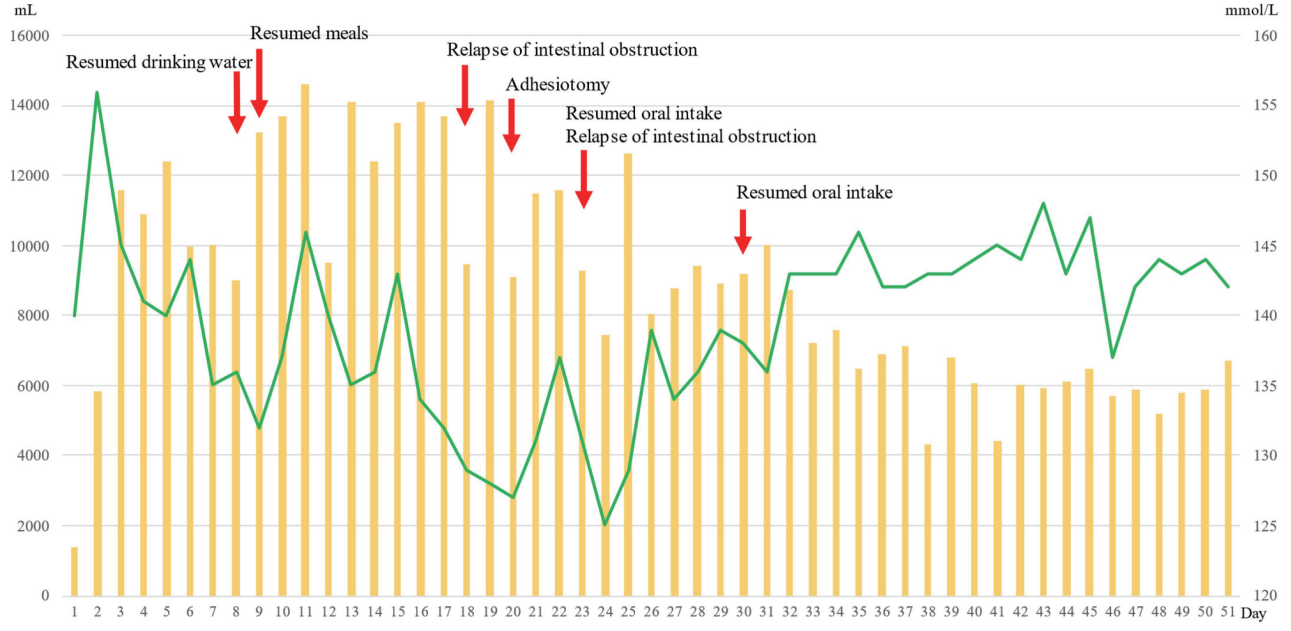

Fig. 3. Transition of urine volume and serum sodium. Bars and lines indicate the daily volume of urine output (mL) and serum sodium (mmol/L).

sequelae.[1] Since it was anticipated that physicians who are not nephrologists would increasingly treat patients with CNDI for other diseases, it is important to know how to manage fluid delivery and blood electrolyte levels during emergency hospitalization and other situations.

Table 1 shows a summary of perioperative infusions in patients with CNDI in the case report literature. Overall, five cases have been reported: two were diagnosed preoperatively with CNDI,[9, 10] and three were diagnosed following postoperative polyuria or hypernatremia.[11–13]

Table 1. Summary of perioperative infusion information for patients with CNDI

Case	Year	Author	Age	Sex	Disease	Treatment	When CNDI was diagnosed	Main infusion solution	Period of inability to take oral intake
1	2001	Mizushima et al.[9]	2	Female	Abnormal upper lip frenulum	Frenectomy	Before surgery	5% glucose solution	Unknown
2	2011	Sugiyama et al.[11]	27	Male	Open left tibial fracture	Osteosynthesis	After surgery	5% glucose solution	Unknown
3	2015	Zaha et al.[12]	56	Female	Abdominal incisional hernia	Laparoscopic incisional hernia repair	After surgery	5% glucose solution	Unknown
4	2016	Gokon et al.[10]	16	Male	Acute appendicitis	Laparoscopic appendectomy	Before surgery	5% glucose solution	1 day
5	2020	Hanada et al.[13]	70	Female	Renal pelvis rupture	Urethral catheterization	After treatment	Unknown	Unknown
Our case	2023	Miyauchi et al.	46	Male	Adhesive bowel obstruction	Adhesiotomy	Before surgery	5% glucose solution	7 days

CNDI, congenital nephrogenic diabetes insipidus.

There is no absolute standard for infusion volume in the conservative treatment of intestinal obstruction. The volume of infusion should be adjusted to avoid dehydration, taking into account the amount of fluid loss, including urine and gastrointestinal fluid drainage, and electrolytes should be corrected as needed to maintain within reference values.[14, 15]

Regarding the volume of infusion for patients with renal enuresis, Moug et al. recommend infusion with a 5% glucose solution at a volume equal to the water loss plus 50–100 mL/h.[8] In the reports summarized in Table 1, in all but one unknown case, a 5% glucose solution was used as the primary infusion after the diagnosis of CNDI. In our case, in addition to the patient's original urine output of 7,000–8,000 ml, the amount of water and electrolyte mass of the infusion was adjusted with reference to the urine output and electrolytes, taking into account the amount of water loss due to bowel obstruction and insensible excretion, with a target of 10,000 ml/day. Once it was determined that oral intake was possible, the infusion volume was reduced according to the amount of water consumed. Postoperative infusion management was also performed in the same manner as above. The urine output and total water supply during the treatment period were almost identical (Fig. 2). Hypernatremia was observed soon after admission due to dehydration, but sodium levels were subsequently corrected with large-volume infusions, mainly of a 5% glucose solution. When sodium and potassium levels became low, adjustments were made to maintain electrolyte levels by adding maintenance fluid infusion and potassium preparations as required. Once the patient was able to take oral intake, the electrolyte trend stabilized (Fig. 3), so it was advisable to resume drinking as soon as possible when drinking was deemed possible.

Useful indicators of fluid management include monitoring blood pressure, urine output, drainage from the NG tube, and, if possible, central venous pressure. Electrolyte trends are also important information. If the patient is hyponatremia, the amount of glucose solution infused may have been excessive, and conversely, if the patient is hypernatremia, the amount of glucose solution infused may have been low.

In acute hypernatremia, the rapid decrease in brain volume can lead to collapse of the brain's blood vessels, resulting in intracranial hemorrhage and irreversible sequelae.[16] Symptoms such as impaired consciousness and convulsions occur when blood sodium concentrations suddenly rise above 158 mEq/L, with fatalities occurring at levels above 180 mEq/L.[17] In such cases, rapid correction of blood sodium concentrations is necessary, using a 5% glucose solution at a rate of 1–2 mEq/L per hour to bring blood sodium concentrations within reference values within 24 hours.[18] In this case, hypernatremia occurred rapidly due to inadequate fluid replacement for dehydration caused by intestinal obstruction in the early stages of hospitalization. The patient did not show any symptoms such as impaired consciousness, and the infusion of 5% glucose solution corrected the hypernatremia to within the reference level. On the other hand, during the course of his hospitalization, his blood sodium level temporarily fell below the reference level. In the treatment of hyponatremia,

one should pay attention to osmotic demyelination syndrome caused by rapid correction of blood sodium concentration, but this is unlikely to occur in the acute phase within 48 hours of onset of hyponatremia.[19] Methods of correction of serum sodium concentration in hyponaemia include fluid restriction, oral administration of salt, and 3% saline infusion.[19] Since the patient had an underlying CNDI and it was anticipated that administration of 3% saline would easily induce hypernatremia, an infusion product with a sodium concentration of 45 mEq/L was used to correct the hyponatremia.

The lowest blood sodium level was found on POD 4. This was thought to be due to the higher infusion of a 5% glucose solution compared to the postoperative decrease in urine volume and the increased sodium excretion from the NG tube due to recurrent bowel obstruction. Since a 5% glucose solution is the main infusion in patients with CNDI, the possibility of hyponatremia should be considered if the infusion volume is excessive.

The patient was given about 10,000 mL/day of a 5% glucose solution, resulting in a total daily caloric intake of about 2,000 kcal and a glucose dosage of about 500 g. In this patient, the maximum random blood glucose level was 187 mg/dL, which did not cause hyperglycemia requiring therapeutic intervention. However, 20,000 mL/day of a 5% glucose solution has been reported to cause hyperglycemia, which was treated with insulin.[12] Therefore, it is necessary to carefully monitor not only electrolytes but also blood glucose levels.

Desmopressin was also administered in this case. Desmopressin is a treatment for central diabetes insipidus, but it has been reported that desmopressin is effective in reducing urine output in mild cases of nephrogenic diabetes insipidus.[20] In this case, since the patient was expected to require long-term abstinence from food and drink, and since there was a possibility of surgery for intestinal obstruction, it was desirable to control urine output as soon as possible, desmopressin was also used in the hope that it would be effective.

This patient was originally diagnosed with CNDI at our hospital and was known to require careful management of infusions. Although the infusion of mainly a 5% glucose solution prevented dehydration and prolonged hypernatremia in the emergency setting, depending on the patient's level of consciousness and severity of illness, it is possible that treatment may have to be initiated without a full history being obtained. If patients are unaware that they have CNDI and are continued on extracellular fluid replacement, hypernatremia can easily result, which can be difficult to diagnose and delay the electrolyte correction with appropriate infusions. CNDI is a very rare disease, and a few cases of patients who are unable to take oral fluids have been reported. This case had the longest period of inability to ingest as far as we could determine and may serve as a reference for future infusion management in patients with CNDI.

CNDI is a disease that should be considered when treating a diverse group of patients without sufficient information, as it can sometimes be difficult to diagnose. In addition, patients with CNDI may have more difficulty than non-CNDI patients in controlling fluid volume and electrolytes when free drinking is not possible, so prevention of postoperative adhesive bowel obstruction is important and the use of anti-adhesive agents should be considered at the time of initial surgery.

In conclusion, patients with CNDI should be given a 5% glucose solution as the primary infusion, and the infusion volume should be adjusted by monitoring daily urine output, electrolytes, and blood glucose levels. Infusion management is easier if oral intake is initiated as early as possible.

The authors declare no conflicts of interest.

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