

Modified oral rehydration therapy in a case with cystic fibrosis

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Infants with cystic fibrosis can develop episodes of hyponatremic, hypochloremic dehydration with metabolic alkalosis, and management is difficult. In this paper, we present a nine-month-old case with cystic fibrosis with pseudo-Bartter syndrome, who was rehydrated with two types of modified oral rehydration solution. Intravenous rehydration was impossible due to inaccessibility of route. With this oral rehydration treatment, her purging rate decreased, and serum sodium levels improved and remained stable. In cases with impossible intravenous route, one of these modified rehydration solutions could be given.

Key words: cystic fibrosis, dehydration, metabolic alkalosis, management, oral rehydration.

Cystic fibrosis is a disease caused by defective cyclic AMP-dependent cystic fibrosis transmembrane conductance regulator (CFTR) chloride channels, leading to failure in chloride and water transport¹. The patients may present with attacks of diarrhea leading to hyponatremic, hypochloremic metabolic alkalosis in the infancy period²⁻⁵. The treatment of choice is intravenous replacement of fluid and electrolytes¹⁻⁵. Sometimes, in such cases, intravascular application may be difficult, due to vascular collapse, and is thus time-consuming, with risk of infectious diseases. However, oral hydration is always safe and easier to apply⁶. Therefore, prompt restoration of intravascular volume in cases with cystic fibrosis could be achieved either by the oral or intravenous route, depending upon conditions. In cases with acute diarrhea, oral rehydration therapy (ORT) has been a major medical advance that has been life-saving in many cases. In addition, ORT provides clear and practical methods for replacement of fluid and electrolyte losses during acute diarrhea with mild or moderate dehydration. However, the standard World Health Organization (WHO) Oral Rehydration Solution (ORS) consists of 3.5 g/L NaCl, 1.5 g/L potassium chloride (KCl), and 2.5 g/L sodium bicarbonate (NaHCO₃), which may

be inappropriate for a cystic fibrosis patient with metabolic alkalosis and hyponatremia⁶. To our knowledge, there are no published studies about oral hydration of cases with cystic fibrosis. In this paper, we report a case with cystic fibrosis who was rehydrated with two types of modified ORS. In this case, intravenous rehydration was impossible due to inaccessibility of route.

Case Report

We present a nine-month-old cystic fibrosis patient who admitted with pneumonia to our clinic and then developed diarrhea leading to moderate dehydration and hyponatremic metabolic alkalosis (serum Na: 131 mEq/L, Cl: 90 mEq/L, pH: 7.501, HCO₃: 31.5 mmol/L, Fig. 1). Intravenous fluid with sodium deficit (2500 ml/m² containing 50 mEq/L Na, with 8 mEq/kg Na added) was given; however, when there was no adequate improvement in the serum sodium concentration (Na: 135 mEq/L, Cl: 98 mEq/L, pH: 7.43, HCO₃: 30.0 mmol/L), the intravenous line was impaired and we could not access a new vascular route, peripherally or centrally. Previously, she had been hospitalized at another clinic for about three months before she was referred to our clinic, with the same problems of intravenous

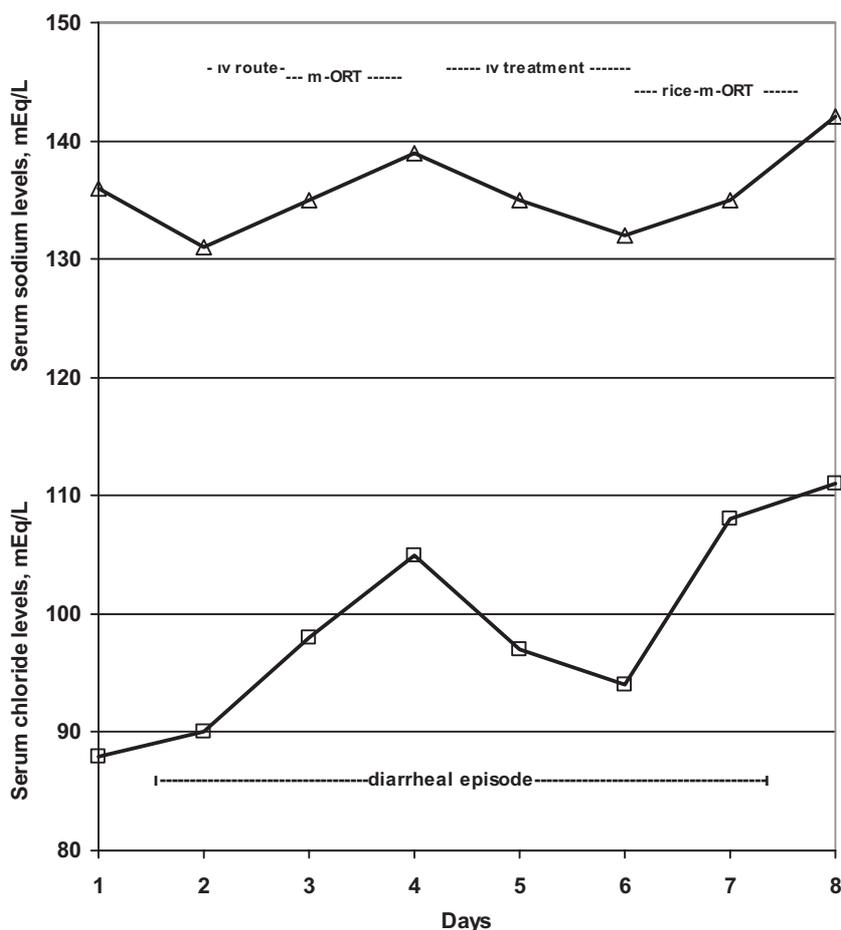


Fig. 1. Serum sodium and chloride levels during the follow-up period.

line application. As her watery, loose diarrhea (8 times/day) and fluid loss continued, we decided to give her modified ORS (6 g/L NaCl, 1.5 g/L KCl, 111 mmol/L glucose) consisting of extra added sodium chloride and no sodium bicarbonate due to the metabolic alkalosis state. We applied a nasogastric feeding tube, and gave 100 ml/kg modified ORS for six hours. She was fed at the beginning of the therapy and then every three hours. Maintenance therapy was then started, with 100 ml modified solution given after each stool output for six hours. She was also fed every 2 to 3 hours during the maintenance treatment. With this treatment, the serum sodium concentration increased to normal ranges (Na: 139 mEq/L, Cl: 105 mEq/L, pH: 7.40, HCO₃: 30.8 mmol/L) at the end of 12 hours of intervention. After this treatment, with restoration of intravascular space, we could easily apply an intravenous line, and classic intravenous fluid treatment was started and

given for two days due to the high purging rate; nevertheless, hyponatremia developed again (Na: 132 mEq/L, Cl: 94 mEq/L, pH: 7.46, HCO₃: 32.4 mmol/L). Finally, we applied a nasogastric feeding tube, and administered 100 ml/kg modified rice ORS (containing 6 g/L NaCl, 1.5 g/L KCl, 50 g/L cooked rice powder) for six hours. Maintenance therapy was then initiated as 100 ml modified rice solution given after each stool output for 24 hours. Her serum sodium concentration increased to normal levels (Na: 142 mEq/L, Cl: 111 mEq/L). With this treatment, her purging rate decreased and serum sodium levels remained stable.

Discussion

Cases with cystic fibrosis may present with electrolyte imbalances and metabolic alkalosis, either due to excessive sweating, vomiting or diarrhea³⁻⁵. The treatment is appropriate

intravenous fluid administration, which may sometimes be difficult to apply or ineffective, as in our case, especially when purging rate is high. In this cystic fibrosis case with hyoelectrolytemia and metabolic alkalosis, modified ORS was given safely.

The cystic fibrosis transmembrane conductance regulator (CFTR) chloride channel controls salt and water transport across epithelial tissues. Mutations in CFTR lead to defective epithelial fluid and electrolyte transport. However, the exact mechanisms and proper treatment of metabolic alkalosis and hyponatremia and the role of CFTR are still unclear¹⁻⁵. Recently, Gawenis et al.⁷ reported that cell shrinkage induced by hypertonic medium had inhibited Na(+)/H(+) exchanger-mediated Na(+) absorption in the jejunum of both CFTR(+) and CFTR(-) mice. It might be also speculated that this modified ORT constituted the hypertonic medium.

Here, we offer two types of modified ORT, which are both easier to apply and more physiological. This treatment might be primarily supportive and directed at treating dehydration and hyponatremia. Further clinical studies are

necessary to detect the effect of modified ORT and to find appropriate solutions for cases with cystic fibrosis.

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