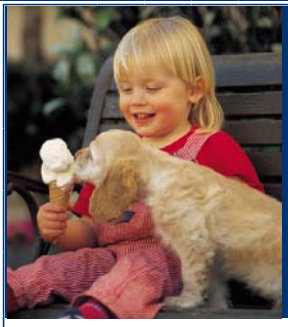

THE SUPPLEMENT

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SODIUM CHLORIDE AND CYSTIC FIBROSIS

Just as salt has had a long and influential role in world history, it also has had a long history in CF. The connection between salty skin and symptoms characteristic of pancreatic and lung disease were first noted in the literature in 1650. [1] Sodium and chloride are essential minerals for several crucial functions in the body, including fluid balance and muscle and nerve contractions. In CF, sodium and chloride play key roles in both disease presentation and disease management. Although often taken for granted, dietary salt is an essential component in the overall care of the person who has CF. This issue of The Supplement focuses on dietary salt and CF.

FEATURED PAPERS:

Changing feeding trends as a cause of electrolyte depletion in infants with cystic fibrosis. Laughlin JJ, Brady MS, Elgen H. Pediatrics. 68:203-207,1981. **Objective:** To identify the cause of electrolyte depletion in infants with CF. **Subjects:** 5 infants, less than 12 months of age; 2 infants diagnosed with CF prior to hospitalization; 3 diagnosed as a result of hospitalization. **Design:** Retrospective case review. **Results:** Baby food manufacturers removed salt from their products in 1965 resulting in a reduction of average sodium intake from 45mEq per day (1035 mg) to 15mEq (345mg) per day. **Conclusions:** Infants known to have CF require daily salt supplementation. Electrolytes should be checked in any infant experiencing anorexia and/or failure to thrive.

Metabolic alkalosis with hypoelectrolytemia in infants with cystic fibrosis. Fustik S, Pop-Jordanove N, Slaveska N, Koceva S, Efremov G. Pediatrics International. 44:289-292,2002. **Objective:** To investigate the incidence of metabolic alkalosis (MA) with hypoelectrolytemia in CF infants. **Subjects:** Records of 103 CF patients were reviewed; 17 infants with a history of MA, 10 male were identified; age 2 to 6 months; all were breast-fed; 2 had been diagnosed with CF prior to MA; 15 were identified with CF after treatment for MA; 11 were homozygous for ΔF508. **Design:** Retrospective chart review. **Results:** MA was identified in 16.5% of the patient population. Season did not influence occurrence. Early age, breast-feeding, delayed CF diagnosis, elevated ambient temperature, and presence of severe CF mutation predisposed infants to develop MA. Inadequate salt supplementation may have contributed to the occurrence of metabolic alkalosis. **Conclusions:** CF should be considered in any infant identified with MA.

Preventing dehydration in children with cystic fibrosis who exercise in the heat. Kriemler S, Wilk B, Schurer W, Wilson W, Bar-Or, O. Med Sci Sports Exerc. 31:774-779,1993. **Objective:** To identify the amount of sodium chloride required to increase voluntary fluid intake. **Subjects:** 11 subjects with CF; 5 boys; age range 10.9 – 19.5 yrs. **Design:** Convenience comparison. **Methods:** Three 3-hour intermittent exercise sessions at 35˚C (95°F), 50% relative humidity. Subjects were offered water, or 30mmol/L Na (690mg) plus 6% carbohydrate solution, or 50mEq/L Na (1150mg) plus 6% carbohydrate solution. **Results:** The higher sodium fluid induces a near significant (P=0.08) higher fluid intake and ameliorated the rate of progressive dehydration. **Conclusions:** A flavored drink with high sodium content (50mEq Na/L) enhanced drinking and attenuated voluntary dehydration.

SPECIAL POINTS OF INTEREST:

- **In CF, sodium and chloride play key roles in both disease presentation and disease management.**
- **Dietary salt is an essential component in the overall care of the person who has CF.**
- **Infants known to have CF require daily salt supplementation.**

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REVIEW

Hypoelectrolytemia (HE), a symptom of CF, was first reported by Kessler and Anderson during a heat wave of 1948. [2] Persons who have CF lose excessive sodium and chloride through their sweat, and this is the basis for the pilocarpin-stimulated sweat test to diagnosis CF. Electrolyte balance is of great importance to overall health. Thus, the body maintains a delicate system to promote either electrolyte retention or excretion. In CF, endocrine and metabolic adaptations to salt depletion have not been well studied [3] and sodium chloride lost through the sweat may inadvertently activate these systems resulting in MA in CF infants [Fustik, 2002] and MA or dehydration in older CF individuals. [4] There is some evidence that the patient's genotype, therefore severity of disease, may influence overall sodium and chloride losses in sweat thereby impacting overall dietary salt needs, [5, 6, 7,Fustik,, 2002,] but to date there is no conclusive evidence to support adjusting dietary salt prescription based on sweat test results.

Infants with CF are exquisitely sensitive to developing electrolyte abnormalities, often presenting with pseudo-Bartter syndrome (p-B). Although infants may develop HE when exposed to high ambient temperature, there is evidence that it also may occur in cooler environments [5,6,8, Fustik,2002] and may be related to fever, vomiting and/or diarrhea, or over-bundling. There are numerous papers describing HE and MA in infants with CF. [5,6,9, Fustik, 2002] These two conditions frequently are seen in infants who have yet to be diagnosed with CF, and therefore not receiving supplemental salt. However, there are reports describing infants with MA who are known to have CF, but either are not receiving supplemental salt or are receiving an insufficient amount to meet increased needs. [6,10] Infant formula is low in sodium (8.6mEq (198mg)/liter); breast milk is even lower (7.0mEq (161mg)/liter). Both contain insufficient sodium and chloride for the infant with CF. Laughlin (1981) reported an increased incidence of electrolyte abnormalities in infants with CF when manufacturers of infant foods removed salt from their products.

Children, teens and adults who have CF are instructed to consume diets containing "high" amounts of salt. As is true for infants, there is little evidence for the recommended amount of daily salt. The advice for a "high" salt diet may be sufficient in cool weather for the stable, sedentary person who has CF. However, additional salt intake probably is necessary for those who engage in exercise or physical labor in warm environments or are ill. When compared to persons without CF, those with CF have much higher concentrations of salt in their sweat, [11] resulting in lower serum osmolality during periods of excessive sweating. Normally, increased serum osmolality stimulates the trigger for thirst. Due to the increased salt in their sweat, persons who have CF may be deprived this trigger, causing lower fluid intake, referred to as "voluntary dehydration." [4] Kriemler, 1999 found that a drink containing 50mEq Na (1150mg) per liter corrected voluntary dehydration. MA has been reported as the presenting symptom in teens [12] and adults [13] subsequently diagnosed with CF. As more women with CF became pregnant and breast-fed their infants, clinicians became concerned about the sodium content of the women's breast milk. An initial report of elevated sodium in breast milk from women with CF [14] proved incorrect. [15]

CLINICAL APPLICATIONS

For people without CF the Dietary Reference Intake [16] for Adequate Intake for sodium ranges from 120mg (5.2mEq) per day for infants less than 6 months of age to 1500mg (65.2mEq) per day for adults. The exact salt prescription for persons who have CF is unknown. Growth failure has been documented in animals fed sodium-deplete chows and in premature infants fed formulas containing insufficient sodium to meet needs. [17] Chronic HE can negatively impact appetite and result in anorexia and growth failure or weight loss. Frank electrolyte depletion can cause shock and lead to death. To prevent HE, which is often present without overt symptoms, supplemental salt must be added to the diet of people who have CF.

Based on his patient care experience, Waring, [18] in the 1970s, recommended adding ¼ teaspoon of salt (26mEq/598mg sodium) daily to the diets of CF infants. Currently, one frequently accepted recommendation for CF infants is 2-4mEq (46-92mg) sodium/kg/

TO PREVENT HE, WHICH IS OFTEN PRESENT WITHOUT OVERT SYMPTOMS, SUPPLEMENTAL SALT MUST BE ADDED TO THE DIET OF PEOPLE WHO HAVE CF.

CLINICAL APPLICATIONS (CONT.)

day [19]. This is the range used to calculate the sodium content of TPN for non CF infants. Alternately, another generally accepted suggestion is adding 1/8 teaspoon of salt (13mEq (299mg) sodium) daily to the infant's diet. However, neither of these amounts may be sufficient, especially if the infant is living in a warm environment or is ill. [5,6,20] In the United States, clinicians most often recommend ⅛ teaspoon of salt daily, starting at birth. [20] Small salt packets, such as those available in fast food restaurants, contain ⅛ teaspoon of salt [21] and may be an easy way for some parents to measure recommended salt supplementation. Some Canadian CF Centers prescribe a pharmacy-prepared liquid "mineral mix,' which offers the benefit of a more precise electrolyte dose. [22] The European Nutrition Consensus report does not recommend routine salt supplementation, but notes that breast-fed infants and patients in high temperatures may require supplementation. [23]

For infants with CF over 6 months of age consideration should be given to increasing the amount of salt to ¼ teaspoon daily. Some CF Centers in Canada either increase salt supplementation at 6 months of age to ¼ teaspoon daily or increase the liquid "mineral mix" to provide 4-5 mEq Na/kg/day. [22] Prior to the reduction of salt in manufactured baby foods, infants could receive as much as ½ teaspoon of salt daily. [Laughlin, 1981] For toddlers, salt intake may be more exact if parents are given a specific amount of salt to add to food daily. Maintaining salt supplementation at ¼ teaspoon until 24 months of age, or until the RD is confident that the toddler is consuming sufficient "high" salt foods, may be indicated. Children, teens and adults who have CF are at risk for electrolyte abnormalities. Parents and patients must be alerted to optimizing salt intake in warm weather. So often "Gatoradeâ" (7.2 mEq (165 mg) sodium per12 oz) or other sports drinks are recommended and consumed by CF patients during exercise. Yet the research of Kriemler(1999) demonstrated that such beverages may not contain adequate sodium when physically active in a warm environment. To reach the equivalent of 50 mEq sodium per liter, as done in the Kriemler study, ⅛ teaspoon of salt must be added to every 12 ounces of regular Gatoradeâ.

Although patients with CF need supplemental salt, the RD must teach parents not to provide excessive salt, especially in infancy. **Hyper**electrolemia can be deleterious to health and may result from overzealous parents adding salt greater than recommended. Also, there are rare instances when individuals who have CF develop additional medical conditions, or CF complications that may require a sodium-controlled diet. In such situations the RD and patient develop a diet plan that addresses the medical condition without causing HE.

ALTHOUGH PATIENTS WITH CF NEED SUPPLEMENTAL SALT, THE RD MUST TEACH PARENTS NOT TO PROVIDE EXCESSIVE SALT, ESPECIALLY IN INFANCY.

Assessing sodium status is challenging. Serum and urine sodium are not sensitive measures of total body sodium. The body will spare sodium to maintain serum levels; therefore, once serum levels are low, the need for sodium is urgent. Normally, urinary sodium is dependent on urine output and influenced by the state of hydration. Calculating the urine sodium/creatinine ratio, particularly in infants with CF, has been suggested. [24] The Hospital for Sick Children in Toronto, Canada uses the following protocol to monitor electrolytes: "Aim to maintain urinary sodium and chloride in range of 40-60mEq/L. If the infant is becoming salt depleted, urine sodium and chloride will fall toward, or even to, zero. Alternatively, if an excess of the supplement is being given, there will be large concentrations of sodium and chloride in the urine. Only in these situations would it be necessary to check serum electrolytes. Providing there is normal renal function, urine analysis minimizes the need for blood taking for monitoring purposes." [22]

In summary, the RD is encouraged to be attentive to the salt needs of persons who have CF. Providing anticipatory guidance for salt supplementation is key. This is especially true for individuals during the first two years of life and for persons with CF when playing, exercising, or doing physical labor in warm environments, and whenever a person is ill. CF should be considered for any patient, regardless of age, admitted to the hospital for MA with HE without renal involvement.

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Conversions and Terminology

Milligrams of Na to mEq (mmol): Divide mg by the atomic wt of sodium, 23.
Grams of NaCl to Na: Multiple gm NaCl by 0.393.
mEq (mmol) NaCl to mg: Multiply mEq (mmol) by 58.5.
Dehydration: Condition resulting from excessive loss of body fluid.
Hyponatremic dehydration: Decreased amount of sodium in the blood, but a normal amount of body fluid.
Pseudo-Bartter syndrome: A condition with hyponatremic, hypochloremic metabolic alkalosis with biochemical abnormalities similar to Bartter syndrome**,** with no pathology in the renal tubules.
Metabolic alkalosis: A disturbance in which the acid-base status of the body shifts toward the alkaline side because of retention of base or loss of acids

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