

An epidemic of pseudo-Bartter syndrome in cystic fibrosis patients

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Introduction

Pseudo-Bartter syndrome (PBS) is characterized by hyponatremic, hypochloremic metabolic alkalosis that mimicks Bartter syndrome but with no pathology in the renal tubules [5]. This article reports the manifestation of PBS in nine cystic fibrosis (CF) patients diagnosed within a 15-day-period in the hottest summer since 1975 in Ankara, Turkey.

Methods

Nine children participated in the study; all were diagnosed with CF-PBS between August 15 and August 31, 2006 in the Hacettepe University Pediatric Pulmonary Diseases Unit. The medical records of CF patients between January 1989 and November 2005 were retrospectively investigated. The main air temperatures of all Augusts from 1975 to 2006 were also obtained.

Results

A total of 57 episodes of PBS were found in 43 CF patients between January 1989 and November 2005 in our center [4, 6]; of these, nine occurred between August 15 and August

31, 2006. The demographic features and laboratory characteristics of these nine CF-PBS patients are given in Table 1. All patients, with the exception of patient no. 8, who was newly diagnosed, had been admitted prior to these dates with PBS. Eight patients were using oral salts, as recommended. A 23-month-old-boy died 6 h after admission despite intensive therapy.

While between 1975–2005 the mean air temperature in August in the Ankara region was 22.9°C (range: 7.2–37.2°C), in August 2006, it was 27.2°C (range: 14.6°C–38°C). August 2006 was the hottest August since 1975 (<http://www.meteor.gov.tr>).

Discussion

Nine CF patients were admitted to the Hacettepe University Pediatric Pulmonary Diseases Unit with clinical features of PBS that were ascribed to the extreme warm meteorological conditions in the Ankara region during a 15-day period. This is a very high incidence when compared to the total of 57 PBS episodes that were recorded in our center since 1989. PBS is a frequently reported complication of CF in infants and children. Predisposing conditions include infancy, severe pulmonary involvement, severe pancreatic insufficiency, genetic variation, and profuse sweating [1, 2]. The association between summer heat waves and CF-PBS was first reported a half century ago [3]. Eight of the patients admitted with signs of PBS had been diagnosed earlier and were using oral salts, as recommended. The manifestation of PBS in these patients is assumed to result from the extremely warm weather: August 2006 was the hottest August in 31 years.

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Table 1 Demographic features and initial laboratory findings on admission of the patients

	Patient no.								
	1	2	3	4	5	6 ^a	7	8	9
Sex	Male	Male	Male	Male	Male	Male	Male	Male	Female
Age ^b	13 y	16 y	14 y	8 m	6.5 y	23 m	9 m	4 m	4 y 9 m
Genetic	F508+/-	F508+/-	F508+/+	F508+/-	F508+/+	G542X+/+	F508+/-	-/-	G542X+/-
Sodium (mEq/l)	122	132	132	129	111	120	124	127	133
Potassium (mEq/l)	2.5	3.0	3.7	3.1	2.8	2.7	2.3	2.7	4.0
Chloride (mEq/l)	62	86	83	71	56	63	62	70	90
Blood urea nitrogen (mg/dl)	29.4	57.6	57	23	96	-	20	53	29
Creatinine (mg/dl)	0.45	1.21	1.38	0.67	2.22	-	0.35	0.63	0.44
Uric acid (mg/dl)	10.08	14.83	13.4	8.82	23.17	-	6.28	19.07	6.53
pH	7.63	7.45	7.50	7.44	7.49	7.5	7.61	7.58	7.64
HCO ₃	43	35.5	29	36	34	29	54	35.5	37
Urine chloride (mEq/l)	15	19	7	10	8	-	14	12	11
Hospitalization time (days)	6	8	8	7	8	6 hours	7	15	3

^a Died 6 h following admission

^b y, Year; m, month

An interesting finding was that five of our patients were older than 4 years. Between 1989–2005 we observed only two children with PBS aged >4 years in our center. The prevalence of PBS is known to be as high as 20% in infancy [2]. The predominance of adolescent patients in this period might have been due to the extra exposure to heat, as the adolescent patients spent more time outside. Conversely, infant patients might have been protected from exposure to the midday sun by their parents.

This condition must be diagnosed early because tetany, seizures, hypoventilation, decrease in cardiac output, and cardiac arrhythmias can occur in PBS [6]. Unfortunately our 23-month-old CF patient died despite intensive therapy.

In conclusion, in extra warm days CF patients ought to be warned to take extra oral salts and water to avoid developing PBS.

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