



**Fig. 1.** (A) Chest computed axial tomography slice, showing areas of parenchymal consolidation with air bronchogram and ground glass opacities in the upper right lobe. (B) Histology study ( $\times 40$ ) of the lung biopsy showing multinucleated macrophages with foreign body vacuoles (silicone).

intracytoplasmic lipid inclusions in the bronchoalveolar lavage is characteristic of silicone pneumonitis, and may obviate the need for a biopsy when clinical suspicion is well-founded. Acute phase treatment entails respiratory support, with administration of high-flow oxygen and mechanical ventilation in more severe cases.

In conclusion, silicone pneumonitis is a rare and potentially severe complication that may occur after the application of silicone for cosmetic purposes. This complication occurring in association with a gel prosthesis is even rarer, although it is important to include it in the differential diagnosis of patients with breast implants who present inflammatory pulmonary processes.

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## Dehydration in Adult Cystic Fibrosis Patients<sup>\*</sup>



### *Deshidratación en pacientes adultos con fibrosis quística*

To the Editor:

Cystic fibrosis (CF) affects the eccrine and exocrine epithelial cells, causing pulmonary disorders, abnormally concentrated sweat, and pancreatic failure. The genetic alteration responsible for this disease is a mutation of the cystic fibrosis transmembrane conductance regulator (CFTR) protein, which acts as a chloride channel, controlling ion transport through the apical membrane.<sup>1</sup> Hot weather, intense physical exercise, stress, fever, vomiting, diarrhea or overdressing<sup>2</sup> can cause patients to present chloride, sodium and potassium depletion, due to a failure to replace loss of salts.<sup>3</sup> Since survival of patients with this disease has increased in recent decades, CF is no longer an exclusively pediatric condition, so it is important to know that other non-respiratory complications exist that may present as a medical emergency.<sup>2</sup>

We report 3 cases of adults with CF, who, despite the usual recommendations, presented severe dehydration syndrome during the summer.

A 19-year-old man, transport driver, presented in the emergency room with a clinical picture of intense asthenia and some vomiting. He had been working under exposure to high temperatures for several hours. He presented in the emergency room with blood pressure 137/71 mmHg, normal body temperature, mucocutaneous pallor, and sunken eyes.

A 25-year-old woman with CF, summer camp monitor, presented in the emergency room with a 24-h history of vomiting (about 20 episodes) and inability to take anything by mouth. She also reported reduced urine output. On arrival, she had arterial hypotension (85/52 mmHg).

A 28-year-old man, employee in a mechanical workshop, presented in the emergency room with cramps, generalized muscle pain, and reduced urine output. The day before, he had been working in the sun for a long period.

The patients' laboratory tests results on admission and discharge are shown in Table 1. All patients received serum replacement therapy, leading to improved biochemical parameters.

Under normal conditions, chloride and sodium are reabsorbed from sweat via the CFTR channel in the sweat glands. When

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**Table 1**  
Clinical Laboratory Tests on Admission and Arterial Blood Gases.

|        | Hb (g/dl)<br>ER | Hct (%)<br>ER | Urea<br>(mg/dl)<br>ER | Cr<br>(mg/dl)<br>ER | Na<br>(mEq/dl)<br>ER | K<br>(mEq/dl)<br>ER | Urea<br>(mg/dl)<br>Discharge | Cr (mg/dl)<br>Discharge | Na<br>(mEq/dl)<br>Discharge | K (mEq/dl)<br>Discharge | pH                | pCO <sub>2</sub><br>(mmHg) | Bicarbonate<br>(mMol/l) | Base<br>Excess<br>(mMol/l) |
|--------|-----------------|---------------|-----------------------|---------------------|----------------------|---------------------|------------------------------|-------------------------|-----------------------------|-------------------------|-------------------|----------------------------|-------------------------|----------------------------|
| Case 1 | 17.2            | 47.5          | 88                    | 0.91                | 118                  | 2.81                | 28                           | 0.65                    | 140                         | 3.1                     | 7.61              | 46                         | 46.2                    | 24.8                       |
| Case 2 | 16.8            | 49            | 127                   | 2.86                | 128                  | 2                   | 39                           | 1.43                    | 140                         | 5.4                     | 7.47 <sup>a</sup> | 45.90 <sup>a</sup>         | 32.7 <sup>a</sup>       | 7.7 <sup>a</sup>           |
| Case 3 | 18.5            | 53.8          | 94                    | 1.43                | 131                  | 4.36                | 56                           | 1.05                    | 134                         | 3.91                    | 7.48              | 33                         | 24.3                    |                            |

Cr, creatinine; ER, emergency room; HB, hemoglobin; Hct, hematocrit; K, potassium; Na, sodium.

<sup>a</sup> Venous blood gases.

CF patients sweat excessively, this reabsorption fails to occur, leading to excretion of large amounts of sodium chloride and decreased blood levels of these ions.<sup>3</sup> This induces secondary hyperaldosteronism with metabolic alkalosis due to increased bicarbonate reabsorption and low blood potassium caused by potassium secretion from the collecting tubule.<sup>4</sup> Moreover, the loss of extracellular fluid lowers the glomerular filtration rate and bicarbonate filtration.<sup>1</sup> This condition is described as “pseudo-Bartter” syndrome, and is characterized by metabolic alkalosis, hyponatremia with hypochloremia, with no renal tubule involvement.<sup>1</sup> It is more common in pediatric patients, and occurs only exceptionally in adolescents and adults; it is sometimes the presenting feature of a CF diagnosis.<sup>5</sup> In view of the potential seriousness of these ion alterations, including the risk of arrhythmias with cardiac arrest, muscle paralysis with involvement of the respiratory muscles or laryngospasm, tetany, and metabolic alkalosis convulsions,<sup>1</sup> it is important that certain recommendations are followed. Treatment is based on appropriate fluid replacement and correction of the electrolyte deficit, with high sodium, chloride and potassium supplements to correct alkalosis; appropriate prevention, with the addition of salt to the diet (1–4 g/day, according to patient age); avoidance of situations leading to excess sweating; and the administration of appropriate supplements during strenuous physical activity.<sup>2</sup>

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## Recurrent Respiratory Infections in a Patient With Chronic Diarrhea<sup>☆</sup>



### *Infecciones respiratorias de repetición en paciente con diarrea crónica*

To the Editor:

Good's syndrome (GS) is a primary immunodeficiency characterized by thymoma and humoral immunodeficiency. It is the most unusual form of the parathymic syndrome, after far behind *myasthenia gravis* or pure red cell aplasia.<sup>1</sup> The most common forms of clinical presentation are recurrent infections, hematological changes, and chronic diarrhea.<sup>1,2</sup>

We report the case of a 76-year-old man with a history of arterial hypertension, a former smoker of 20 pack-years, with chronic diarrhea which yielded *Campylobacter coli* on culture. He was referred to the respiratory medicine department for repeated respiratory infections. Forced spirometry showed mild obstruction: FEV<sub>1</sub>/FVC 0.67, FEV<sub>1</sub> 2.1 l (87%), FVC 3.17 l (98%). Skin prick tests for airborne allergens were negative. Clinical laboratory tests

showed hemoglobin 11.7 g/dl with normal corpuscular volume, with no impact on platelet levels, and markedly reduced CD19 lymphocytes, with a CD4/CD8 ratio of 1.04. Immunoglobulin levels were low: IgA <5 mg/dl, IgG <74 mg/dl, IgM <5.3 mg/dl. Methicillin-resistant *Staphylococcus aureus* was isolated from repeated sputum cultures. Computed tomography (CT) of the paranasal sinuses showed occupation of the maxillary sinuses, while the chest CT revealed mild bronchiectasis in the middle lobe, lingula and both lower lobes, and a solid multilobulated mass in the anterior mediastinum suggestive of thymoma. VATS was performed, confirming the histological diagnosis of polygonal cell cortical thymoma. With these findings, the patient was determined to have GS, and immunoglobulin replacement therapy was started. He showed good progress and the number of infections fell.

GS mostly appears in patients in their 30s or 40s (unlike our patient), and affects men and women equally.<sup>1</sup> This immunodeficiency is caused by an antibody deficit, and is currently classified as a different entity to common variable immunodeficiency (CVID).<sup>1</sup> It accounts for 2% of cases of primary antibody deficiency treated with immunoglobulin replacement therapy.<sup>3</sup>

The most common clinical manifestation is recurrent respiratory infection, and the major pathogens are *Haemophilus influenzae* and *Pseudomonas* spp. The chronic diarrhea presented by 50% of patients appears to have an autoimmune basis, and the isolation of pathogenic agents is anecdotal.<sup>3</sup>

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