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EDITORIAL

Care of people with cystic fibrosis: What is the role of specialists in Endocrinology and Nutrition?☆

Atención a personas con fibrosis quística: ¿cuál es el papel de los especialistas en Endocrinología y Nutrición?

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Cystic fibrosis (CF) is a disease caused by the alteration of a single gene located on the long arm of chromosome 7 (CFTR gene, cystic fibrosis transmembrane conductance regulator). The protein encoding the CFTR gene behaves like a chloride channel and the mutations lead to a defect in chloride transport in the epithelial cells of the respiratory, hepatobiliary, gastrointestinal and reproductive systems, as well as in the pancreas and the sweat glands. Due to the multitude of organs and systems it affects, CF is a complex disease that requires an integrated approach in specialised units. The estimated incidence is one per 2,500—6,000 births, depending on the population studied.[1]

Until very recently, all articles published on CF began with ''it is the most common fatal recessive Mendelian inheritance disease in the Caucasian population''. In the 1950s, the average life expectancy was a few months, with recurrent respiratory infections, meconium ileus and malnutrition secondary to pancreatic malabsorption being the main causes of death. Today the patient profile has changed radically, and what was once a ''fatal paediatric disease'' has become a ''chronic, multisystemic complex disease of children and adults'', which requires an integrated multidisciplinary approach in specialised units.

With regard to the standards of care for people with CF, the main scientific societies insist that the multidisciplinary team should include a dietician or equivalent specialist in nutrition, who should be responsible for nutrition education and support.[2–4] Notwithstanding the fact that CF Units should have their own dietitians-nutritionists, in Spain the training received by endocrinology and nutrition specialists makes them the healthcare professionals most qualified to holistically coordinate nutritional and endocrine care for adult patients with CF.

Over the last 30 years, advances in the treatment of CF have contributed to improved survival, with a median age of around 40 years, and adult specialists have been integrated into patient care.[5] This radical change is the result of mul-

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tiple factors, which include the following: a) Improvement in its diagnosis, both in the paediatric population (due to increased awareness and neonatal screening) and in adults (with a greater diagnosis of mild forms with little or no gastrointestinal involvement); b) Advances in antibiotic therapy (oral, inhaled and intravenous); c) The incorporation of ''acid-resistant'' pancreatic enzymes in the 1980s, along with adequate dietary and nutritional monitoring; d) A comprehensive approach to comorbidities (such as CF-related diabetes and psychological symptoms such as depression and anxiety); e) Pulmonary rehabilitation and physiotherapy; f) Transplantation; and g) Treatment and follow-up in multidisciplinary CF units.

In Spain, it is endocrinology and nutrition specialists who mostly see adults with CF-related malnutrition in the Clinical Nutrition and Dietetic Units (Unidades de Nutrición Clínica y Dietética, UNCYDs), covering a range of areas from diet therapy (basic and advanced) to supplementation, enteral tube nutrition and home parenteral nutrition. Malnutrition in CF is an independent risk factor for predicting morbidity and mortality. The interaction between nutrition and lung function is therefore highly relevant as, by deteriorating in parallel to each other, they affect quality of life and survival prognosis.[4,6] Exocrine pancreatic insufficiency (EPI) occurs in 70–90% of CF patients and the correlation between genotype and phenotype is high. Most patients with EPI tolerate a high-fat diet (which allows them to meet the necessary calorie requirements) if they are treated with adequate doses of pancreatic enzymes. Although the prevalence of malnutrition has markedly decreased in recent years, figures close to 25% continue to be reported in both children and adults with CF based on body mass index (BMI). More and more patients have lean body mass malnutrition with apparently normal weight (or even in the overweight and obesity range).[7] It is therefore essential to properly categorise patients according to their morpho-functional features in order to intervene, not only in relation to their BMI, but also based on their muscle mass and function. In addition to improving nutritional parameters, dietary-nutritional intervention supported by pulmonary rehabilitation programmes can slow down the progressive decline in lung function. In this context, it should be the endocrinology and nutrition specialist who coordinates comprehensive nutrition education and care in adults: complete nutritional assessment including intake estimation, anthropometric measurements and body composition (fat-free mass, muscle function parameters), nutrition education and treatment, and assessment of pancreatic function and intestinal absorption, vitamin and other micronutrient status, and pubertal development. Therapeutic education promoting healthy eating habits, but adapted to individual energy-protein requirements (high-calorie in patients undernourished or at risk of malnutrition and normal or low-calorie in overweight and obese patients) forms the essential foundations of the nutritional therapy approach. In addition to dietary recommendations, a not-insignificant percentage of patients require oral nutritional supplements and up to 5–10% of the series report the need for home enteral nutrition by gastrostomy tube.[8–10] The inclusion of advanced practice nurses specialising in nutritional support and dieticians-nutritionists in the UNCYDs team complements the multidisciplinary approach for these patients.

The prevalence of CF-related diabetes (CFRD) increases with age and peaks at around 50% of patients in their 40 s. CFRD is associated with an increase in morbidity and mortality, and is another marker of disease progression. Systematic screening is therefore recommended from the age of 10 so that appropriate treatment can be started at an early stage. The treatment of choice for CFRD is insulin, generally in a basal-bolus regimen and with self-monitoring of blood glucose and carbohydrate counts.[11,12] Once again, it is the endocrinology and nutrition teams that possess the experience and training in this field to meet the needs of patients, including the incorporation of new technologies and advanced therapeutic education by expert nurses. The inclusion of continuous blood glucose monitoring systems on the list of funded devices from 2021 will undoubtedly be a turning point that will improve patient follow-up.[13] The prescribing of these devices falls to the endocrinology and nutrition specialists.

Approximately two-thirds of adults with CF could have osteopenia and osteoporosis. People with CF have multiple risk factors for inadequate bone mineralisation, including malnutrition, pancreatic insufficiency and malabsorption, the use of corticosteroids, vitamin D and K and calcium deficiencies, hypogonadism and delayed puberty, diabetes mellitus and chronic infection, with increased inflammatory cytokines, to name but a few.[14] Once again, the experience amassed by endocrinology and clinical nutrition teams in the prevention, approach, treatment and monitoring of secondary osteoporosis places us in a key position within the multidisciplinary teams for CF care.

The assessment and monitoring, in coordination with other healthcare professionals, of other complications and comorbidities such as liver involvement, gastrooesophageal reflux, pre- and postoperative management (transplantation, intestinal resections, etc.), bacterial overgrowth, distal intestinal obstruction syndrome, fibrosing colonopathy, etc. are also a routine part of patient care for specialists treating people with CF.

Lastly, it is important to highlight the role that endocrinologists play within CF teams in promoting teaching and translational biomedical research, to enable new scientific knowledge (both preclinical and clinical) to be rapidly transferred to care practice and, ultimately, to help resolve patients' problems.

The advent of new CFTR protein repair drugs (potentiators and correctors) points to a very promising future, as we can look forward to a radical change in the natural history of the disease.[15] However, while we await the arrival of a full cure, the role of the endocrinologist is essential in multidisciplinary CF units in Spain.

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