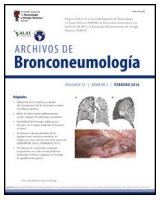
Archivos de Bronconeumología 57 (2021) 563–564

Sociedad Española de Neumología y Cirugía Torácica SEPAR

ARCHIVOS DE Bronconeumología

www.archbronconeumol.org



Editorial

Cystic fibrosis: Quality of life and radiological monitoring☆

Fibrosis quística: calidad de vida y estudio radiológico

Cystic fibrosis (CF) is a genetic disease, and while it involves multiple systems, respiratory problems are the main cause of morbidity and mortality. Recurrent respiratory infections and bronchiectasis observed on chest high resolution computed tomography (HRCT) lead to a progressive decrease in forced expiratory volume in 1 s (FEV_1). Recurrent respiratory exacerbations perpetuate the circle of inflammation and infection, contributing to the progression of bronchiectasis and consequently to loss of lung function. This, combined with the digestive involvement inherent to this disease, affects the nutritional status and worsens the prognosis of these patients.

These parameters are measurable and objective and provide relevant information on the progress of CF but discrepancies with the patient's well-being are sometimes detected. Quality of life is measured by validated questionnaires that reflect the patient's perception of the severity of their disease. The validated, CF-specific quality of life questionnaire is called the Cystic Fibrosis Questionnaire-Revised (CFQ-R). It collects 12 variables of which half address specific domains for this disease (body image, eating, treatment burden, and weight, respiratory, and digestive symptoms) while the others are similar to other quality of life questionnaires.

So what is the relationship between quality of life and these objective lung function tests and HRCT in CF?

The relationship between lung function and the CFQ-R has been evaluated. The respiratory symptoms, vitality, physical functioning and health perception domains are those most closely related to FEV_1[1].

With regard to imaging tests, chest HRCT is clearly more sensitive than chest X-ray[2], and use of the latter should be limited to the evaluation of complications such as atelectasis, new infiltrates, and pneumothorax. The modified Bhalla score in HRCT is a simple, objective tool that has shown a good degree of correlation with spirometry values[2–4].

There is controversy regarding the ideal frequency for performing chest HRCT during the clinical follow-up of CF, and whether the modified Bhalla or the Brody II is the best radiological score. The latter is more complex, and this is perhaps why it is more rarely used in routine clinical practice.

Regarding quality of life and imaging tests, Tepper et al.[5] evaluated the ability of HRCT to predict exacerbations in pediatric patients, specifically bronchiectasis and air trapping, and its relationship with quality of life. Both bronchiectasis and air trapping were related to the respiratory domain of CFQ-R, as were the extent of mucous plugging, peribronchial thickening and consolidations. The authors observed that for each point increase in the respiratory domain of CFQ-R, exacerbations in the following year decreased by 5%, and for each point decrease in the bronchiectasis domain, exacerbations in the following year increased by 10%. These results led them to conclude that the respiratory domain score of the CFQ-R could predict the number of exacerbations during follow-up.

One year later, the same authors published another study in which they reaffirmed that bronchiectasis and air trapping predicted a worse quality of life in the CFQ-R respiratory domain and supported the high sensitivity of chest HRCT for the follow-up of progression in CF[6].

Similarly, Kilcoyne et al.[7] published a study in 2016 evaluating the relationship between HRCT and the CFQ-R questionnaire in 101 CF patients over 15 years of age. They divided patients according to whether the HRCT was performed in the inpatient or the outpatient setting. As expected, the Bhalla score was significantly worse in hospitalized patients, with a significant relationship between the CFQ-R social domain and the appearance of consolidations and between the respiratory domain and air trapping observed on HRCT. In outpatients, an association emerged between consolidations and the respiratory, physical, and vitality domains, and between the severity of bronchiectasis and the vitality and body image domains. The total Bhalla score was associated with the overall treatment burden domain.

Our group was interested in exploring these areas further, and we published a multicenter study with 158 adults with CF which found not only a statistical significance between lung function tests and the modified Bhalla score, but also a significant relationship between CFQ-R domains and the extent of mucous plugging and bronchial sacculations on the modified Bhalla score[3]. Furthermore, the total modified Bhalla score was able to predict the number of future exacerbations with a worse Bhalla score being related to age, genotype, *Pseudomonas aeruginosa* bronchial infection, and pancreatic insufficiency[8]. We have also assessed the evolution of anatomical damage on HRCT measured by the Brody II and Bhalla scales and their relationship with an increased need for antibiotics[4].

The above highlights the essential role of chest HRCT in the follow-up of CF patients. We believe that it is a key tool that pro-

☆ Please cite this article as: Diab Cáceres L, Girón Moreno RM, Caballero Sánchez-Robles P. Fibrosis quística: calidad de vida y estudio radiológico. Arch Bronconeumol. 2021;57:563–564.

vides early, accurate information that relates to quality of life, lung function data, and can predict future exacerbations.

The risk of exposure to ionizing radiation should always be taken into account, although low radiation studies are now safer and these techniques can help the physician offer better follow-up care to the CF patient.

It should be noted that the quality of life of CF patients is highly impacted by respiratory comorbidity and loss of lung function and exacerbations, and by alterations observed on HRCT, primarily bronchiectasis, mucous plugging, air trapping and bronchial sacculations[3]. CFQ-R, which is safe and easy to use and can help rationalize the use of HRCT in some patients, should therefore be integrated into routine clinical practice. Obtaining CFQ-R scores could possibly be streamlined with the implementation of electronic formats or with the validation of more simplified specific questionnaires.

Funding

Financed by SEPAR. “Research Grant”.

References

1. Riekert KA. The association between depression, lung function, and health-related quality of life among adults with cystic fibrosis. Chest. 2007;132:231–7.
2. Bhalla M, Turcios N, Aponte V, Jenkins M, Leitman BS, McCauley DI, et al. Cystic fibrosis: scoring system with thin-section CT. Radiology. 1991;179:783–8.
3. Diab Cáceres L. Fibrosis quística y calidad de vida en adultos: valoración del estudio radiológico. Madrid: Universidad Autónoma de Madrid; 2020.
4. Zorzo C, Caballero P, Diab L, Pastor Sanz MT, Gomez Punter RM, Girón RM. Predictive value of computed tomography scoring systems evolution in adults with cystic fibrosis. Eur Radiol. 2020;30:3634–40.
5. Tepper LA, Utens EMWJ, Caudri D, Bos AC, Gonzalez-Graniel K, Duivenvoorden HJ, et al. Impact of bronchiectasis and trapped air on quality of life and exacerbations in cystic fibrosis. Eur Respir J. 2013;42:371–9.
6. Tepper LA, Caudri D, Utens EM, van der Wiel EC, Quittner AL, Tiddens HA. Tracking CF disease progression with CT and respiratory symptoms in a cohort of children aged 6-19 years. Pediatr Pulmonol. 2014;49:1182–9.
7. Kilcoyne A, Lavelle LP, McCarthy CJ, McEvoy SH, Fleming H, Gallagher A, et al. Chest CT abnormalities and quality of life: relationship in adult cystic fibrosis. Ann Transl Med. 2016;4:87, http://dx.doi.org/10.21037/atm.2016.03.08. PMID: 27047946; PMCID: PMC4791339.
8. Diab Cáceres L, Girón Moreno RM, Garcia Castillo E, Pastor Sanz MT, Olveira C, García Clemente M, et al. Predictive value of the modified Bhallascore for assessment of pulmonaryexacerbations in adults with cystic fibrosis. Eur Radiol. 2021;31:112–20, http://dx.doi.org/10.1007/s00330-020-07095-y.

Layla Diab Cáceres,[a,*] Rosa María Girón Moreno,[a]
Paloma Caballero Sánchez-Robles[b]
[a] *Servicio de Neumología, Hospital Universitario de La Princesa, Universidad Autónoma de Madrid, Madrid, Spain*
[b] *Servicio de Radiodiagnóstico, Hospital Universitario de La Princesa, Universidad Autónoma de Madrid, Madrid, Spain*

* Corresponding author.
E-mail address: layladch@gmail.com (L. Diab Cáceres).