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Lung ultrasound evaluation in people with cystic fibrosis: A new approach in the pulmonology outpatient clinic

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Abstract

Background: Cystic fibrosis (CF) is a genetic disease that causes progressive lung disease with major impact on the quality of life. Lung ultrasound (LUS) allows to assess the lung involvement through the artefacts analysis and is increasingly used in children but is not yet used to monitor people with CF(pwCF). The main aim of this study was to describe the LUS pattern of pwCF during their routinary check-up visit. The secondary objective was to correlate the LUS findings with pulmonary function indices.

Methods: We performed a cross-sectional observational study, enrolling adolescents and young adults with CF. Each patient underwent clinical assessment, measurement of SpO2, assessment of lung function by spirometry and LUS.

Results: Twenty-nine subjects with CF were included. The most frequent alterations were consolidations (72.4%) located in the left apical anterior and right apical posterior regions followed by interstitial syndrome (65.5%). The 41.4% of cases presented the lingula involvement, characterized by a consolidation with static air bronchogram, and 55.2% showed pleural irregularity mainly in the posterior apical regions. A significant correlation was found between the LUS total score and spirometric indices: FEV_1 ($p = .003$), FVC ($p = .002$), Tiffenau Index <80% ($p = .014$), and FEF 25–75 ($p = .004$).

Conclusions: Our study describes LUS findings in pwCF. It also showed a correlation between LUS score and the patients' lung function measured by spirometric indices. We conclude that LUS may be useful in routine monitoring of pwCF in combination with clinical and spirometric assessment.

KEYWORDS

children, cystic fibrosis, lung ultrasound, personalized medicine, pulmonary disease

1 | INTRODUCTION

Cystic fibrosis (CF) is a common autosomal recessive genetic disease, with an estimated prevalence of 9.36/100,000 and 9.79/100,000 residents in Italy in 2019 and in 2020, respectively.[1,2] It is caused by pathogenic variants of the cystic fibrosis transmembrane conductance regulator (CFTR) gene,[3] which codes for an anion channel protein found in various exocrine-secreting cells, such as those of lung, liver, pancreas, and sweat glands.[3,4] Absent or defective CFTR protein produces dehydrated and viscous secretions that can damage many organs.[4]

Currently, respiratory disease is the leading cause of morbidity and mortality in CF: alterations in mucociliary clearance lead to a cycle of repeated infection, airway inflammation and obstruction resulting in the development of bronchiectasis,[5] progressive lung tissue destruction and evolution to chronic respiratory failure. People with CF (pwCF) suffer during their lifetime intermittent pulmonary exacerbations (PEx), characterized by increased cough, increased sputum production and/or change in its appearance/consistency, tiredness, exercise intolerance, weight loss, dyspnea and decreased peripheral oxygen saturation (SpO2), haemoptysis, decrease in pulmonary function by 10% or more from a previously recorded value and change in physical examination of the chest.[6] For these reasons, the chronic pulmonary disease results the main cause of morbidity, having a major impact on the quality of life.[5]

Several tools are used to assess the anatomical and functional lung damage in pwCF. Spirometry is a common, and relatively easy, noninvasive, test used to monitor pulmonary function by measuring the quantity (volume) and velocity (flow) of inhaled and exhaled air. Moreover, the most common imaging techniques for lung anatomical assessment are chest x-ray (CXR) and high-resolution computed tomography (HRCT). CXR is fast and inexpensive, but it is insensitive to the early changes of CF, such as air trapping and primary bronchiectasis.[7] HRCT is considered the gold standard for quantitative and qualitative evaluation of the lung and it is recommended every 2 years in the follow-up of the patient.[8] Nevertheless, the radiation exposure, the necessity of anesthesia in younger children and its high cost, limit its use in pwCF.[9] Recently, magnetic resonance imaging (MRI) has been proposed for imaging surveillance of PwCF. It is free of ionizing radiation but is less sensitive than HRCT to identify specific imaging features, such as distal bronchiectasis.[8,9]

In recent years, lung ultrasound (LUS) has become increasingly popular, especially in the pediatric age, for the diagnosis and follow-up of several parenchymal and pleural diseases.[10] It is safe, noninvasive, cost-effective, reduces radiation exposure, does not require sedation and can be performed at the patient's bedside.[10] These characteristics have encouraged training and studies on the use of LUS also in CF.

Currently, only a few studies assessed the applicability of ultrasound for the evaluation of CF lung disease.[11–14] In these studies, authors found that LUS results were superior to CXR and comparable with HRCT for the evaluation of CF pulmonary exacerbation.[11–14] Hassanzad M. et al., showed that LUS in PwCF for the identification of consolidations had a sensitivity of 94.7%, a specificity of 90% and a positive predictive value of 94.7%. In addition, LUS was 100% sensitive for the detection of the airway bronchogram and 88.9% for the identification of B-lines.[14]

2 | MATERIALS AND METHODS

2.1 | Study design

A cross-sectional observational study was conducted at the University teaching tertiary children's hospital, Institute for Maternal and Child Health–IRCCS Burlo Garofolo of Trieste (Italy), involving adolescents and young adults affected by CF admitted to the Pulmonology outpatient Clinic for a routine diagnostic check-up.

We approached all eligible children over 10 years of age, with diagnosis of CF confirmed by genetic studies and that consent to participate in the study. Exclusion criteria included: children under 10 years old, patients undergoing immunosuppressive therapy after lung transplantation, ongoing PEx and a lack of consent by patients or parents. We chose to include children over 10 years old and adults both because they were more cooperative in the examinations performed (especially spirometry) and because their anatomical and functional lung characteristics are similar.

The main aim of our study was to describe the LUS pattern of pwCF during their routinary check-up visit. The secondary objective was to correlate the LUS findings with pulmonary function indices in these subjects. The study period was from May 2021 to October 2021.

During the routinary visit each patient underwent clinical evaluation, measurement of SpO2, assessment of lung function by spirometry and LUS. General features including age, gender, body mass index (BMI, kg/m^2), genetic mutations, diabetes and/or pancreatic insufficiency, microorganism associated with CF and therapies were recorded for each patient. Written parental informed consent as well as written child assent was obtained from all participants. The study was approved by the Institutional Review Board and Ethic Committee of University of Trieste.

2.2 | Pulmonary function

To assess the basic lung function, a spirometry was conducted at rest using the Vmax™ Encore system (Viasys Healthcare). The spirometry was performed in accordance with ATS/ERS criteria. The following indices were recorded and evaluated: forced vital capacity (FVC, %), corresponding to the maximum volume of air exhaled from a maximum inhalation (normal value > 80%); forced expiratory volume in 1 s (FEV_1, %) that is the exhaled air volume during the first second of FVC (normal value > 80%); Tiffenau Index, which corresponds to the ratio between FEV_1 and FVC and it is reduced in obstructive pulmonary disease (pathological value < 80%); Forced Expiratory Flow (FEF 25–75, %), the effort independent part of FVC,

represented by the airflow eliminated during the central part of FVC and is a sensitive indicator of small airway function (normal value > 70%); Peak Expiratory Flow (PEF, %), that is the maximal airflow speed achieved during the maximally forced exhalation. All values reported were determined by the instrument having as reference the predicted values of spirometry according to ERS1993 Update + Zapletal in the Caucasian population.

2.3 | Ultrasound imaging

LUS was performed by a certified pediatrician (AC), who participated a completed training in point-of-care ultrasound and chest ultrasound. The Esaote MyLab 40 device was used for LUS with a high-frequency 12 MHz linear probe and, depending on the patient's age, a convex probe of 4–9 MHz frequency. We established a depth of 4 cm and the focus was always positioned at the level of the pleural line. Each patient was evaluated in the supine and prone decubitus, studying the anterior, lateral and posterior chest walls. Chest wall was examined in 12 anatomical regions (anterior apical-medium and lower, lateral apical-medium and lower, and so on) performing two scan for each area (longitudinal and transverse), according to the "2012 International Consensus Conference on Lung Ultrasound".[15] Longitudinal scans were obtained by placing the probe along the midclavicular line, the midaxillary line, the scapular line and the paravertebral line and by moving it in a cranio-caudal direction; transverse scans were performed by placing the probe in all intercostal spaces. The scans were performed to assess the presence of A-pattern, B-pattern (interstitial syndrome), consolidations, combined pattern (A-pattern + B-pattern) and any new findings. The A-pattern was represented by horizontal hyperechogenic lines (A-lines), due to reverberation artefacts and expressing the air content of the lung.[14] The B-pattern consisted of multiple diffuse B-lines; they were laser-like vertical hyperechoic reverberation artifacts extending from the pleural line to the bottom of the screen and moving synchronously with lung sliding.[14] Consolidations were subpleural hypoechogenic areas with irregular margins (shred sign), which based on air loss and the predominance of fluid could take on an ultrasound aspect like the liver (hepatic lung).[14] Each lung area was scored with values ranging from 0 to 3 points, based on the presence of B-pattern and consolidations: a region with A-pattern scored 0, a region with B-pattern scored 1 point and with a consolidation 2 points.[11,15] Therefore, each anatomical region could achieve a maximum score of 3 points and all chest a maximum score of 36.[11,15] In addition, we described also for each region the presence of pleural irregularities, pleural effusion and the lingula involvement.

2.4 | Statistical analysis

Categorical variables are reported as counts and percentages. Normality of distribution of continuous variables was tested by means of Shapiro Wilk test. Continuous variables are expressed as means and standard deviations (SD) or as median and inter-quartile ranges (IQR), if not normally distributed. Differences in not normally distributed continuous variables were tested by Mann-Whitney U test. Kruskal-Wallis test was used to compare continuous variables not normally distributed between more than two groups. Spearman's Rho partial correlation was used to describe the strength of the correlation between two variables. A two-sided p value < .05 was considered as statistically significant. All data analyzes were performed using the Statistical Package for the Social Sciences (SPSS for Windows; version 25.0; SPSS Inc.).

3 | RESULTS

The study was initially proposed to 45 pwCF, aged over 10 years, admitted to our Pulmonology outpatient Clinic. Of these, 10 refused to participate, 5 were excluded from the analysis due to PEx, and 1 for incomplete ultrasound images (Figure 1). Finally, 29 pwCF were included of which 14 male and the mean age was 27.07 ± 9.12 years. Of all the patients included, 22 had a mutation of the delta F508 allele in homozygosity or heterozygosity. Among the nine patients under 20 years of age, BMI was within 25%–50% in six of them, within 50%–75% in two and within 75%–90% in one. The mean BMI of the remaining 20 pwCF aged over 20 years was 22 ± 3.1 kg/m^2. Out of the total sample, the pulmonary involvement evidenced by an altered FEV_1 (pathological value < 80%) was observed in 21 of them. More detailed demographical and clinical features of the study population are summarized in Table 1.

Therapies prescribed at home included: inhaled hypertonic saline in 12 (41.4%) patients and rh-DNase in 9 (31%). In addition, inhaled antibiotic therapy was prescribed continuously (ON/ON) in 21 (72.4%) patients, while every other month (ON/OFF) in 8 (27.6%). Twenty-four (82.8%) of enrolled subjects underwent daily Chest Physiotherapy (CPT), periodically followed by a experienced physiotherapist. In particular, 13 (44.8%) patients used the application of positive end-expiratory pressure through a face mask (PEP-mask), 3 (10.3%) performed autogenic drainage (AD), 6 (20.7%) used SpiroTiger(®) device, 1 (3.4%) used PEP-mask followed by AD and 1 (3.4%) used PEP-mask combined with SpiroTiger(®) device. In addition, in our study population, 11 (37.9%) patients treated with CFTR modulator therapy according to approval for age and CFTR gene variant: Lumacaftor/Ivacaftor (Orkambi®) was prescribed in 7 (24.1%) cases, Tezacaftor/Ivacaftor (Symkevi®) in 1 (3.5%), Ivacaftor/Tezacaftor/Elexacaftor (Kaftrio®) in other 2 (6.9%) and only Ivacaftor (Kalydeco®) in 1 (3.5%).

All patients included in the study underwent LUS during their routine visit. Twenty-six (89.7%) subjects had at least one alteration revealed by lung ultrasonography: consolidations were found in 21 (72.4%) subjects, interstitial syndrome in 19 (65.5%) and combined pattern in 16 (55.2%) (Table 2). Consolidations were found more in the left anterior superior (33.3%) and right posterior superior posterior (23.8%) regions; in 13 (62%) patients these lung consolidations were >1 cm, while in 8 (38%) they were ≤1 cm. In particular, 12 (41.4%) cases

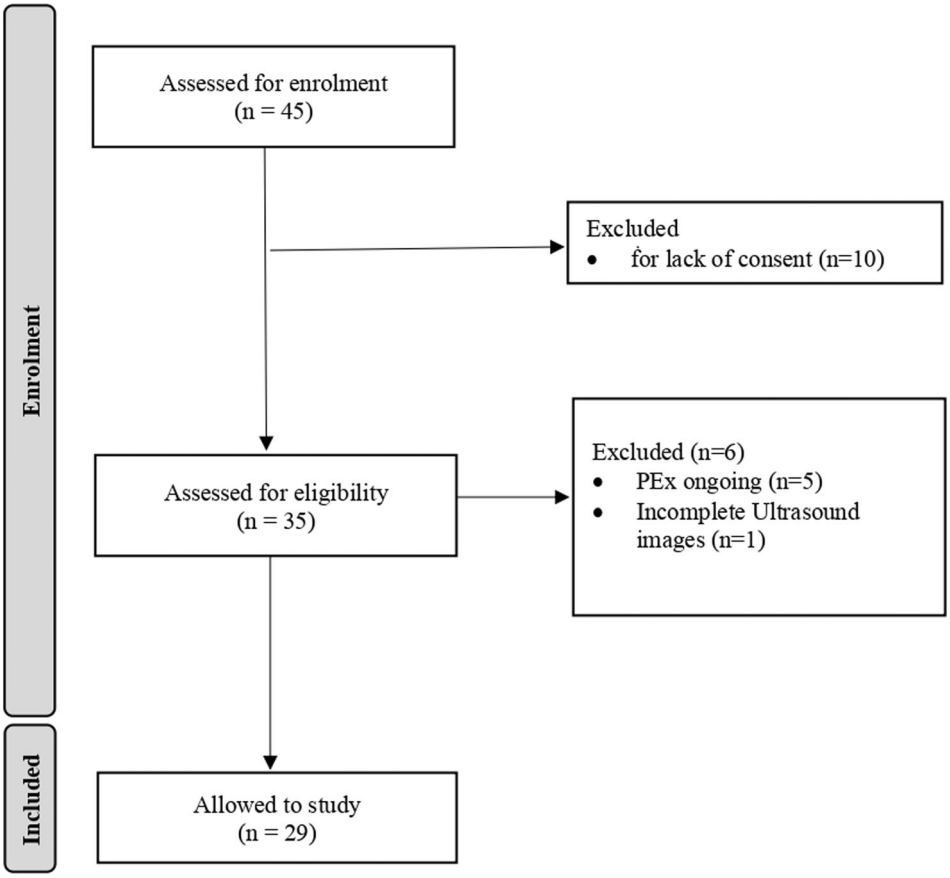

FIGURE 1 Study flowchart.

presented the lingula involvement, characterized by the presence of a consolidation with static air bronchogram (Figure 2A, E-video 1). This ultrasound finding was present in 11 (91.7%) adult patients over the age of 18, while only one (8.3%) 16-year-old adolescent presented this alteration. In addition, 16 (55.2%) patients showed pleural irregularity on LUS mainly in the posterior superior regions (Figure 2B) and a mild pleural effusion was found in only one patient (3.4%). The LUS total score ranged between 1 and 17 out of a total of 36, with a median value of 6 (IQR 2–11) (Table 2). As regards the LUS total score no statistically significant difference emerged according to the presence of other comorbidities, such as diabetes and pancreatic insufficiency, or ongoing therapies. Furthermore, no correlation was found between LUS total score and the age, weight, hight and BMI.

For the secondary objective of the study, we analyzed the correlation between LUS total score and pulmonary function data, such as SpO2 and spirometric indices. No statistically significant correlation was observed between LUS total score and SpO2 ($p = .463$; rho = −0.142). Instead, a LUS total score higher was found in those patients presenting with $FEV_1 < 80\%$ (U = 23.5, $p = .003$), FVC < 80% (U = 25.5, $p = .002$), Tiffenau Index <80% (U = 48.0, $p = .014$) and FEF 25–75 < 70% (U = 11, $p = .004$) (Figure 3). No statistically significant difference was observed as for PEF value ($p = .078$). Analyzing more detailed LUS score in different explored lung regions we observed that this was higher in the right anterior superior region of patients with pathological FEV_1, FVC, Tiffenau Index and FEF 25–75 values ($p = .031$, $p = .012$, $p = .040$ and $p = .036$ respectively). Yet the score was higher in left anterior and lateral basal region in patients with pathological FEV_1 ($p = .053$, $p = .020$) and in right posterior superior region for pathological FEF 25–75 ($p = .051$) and Tiffenau index ($p = .026$). Moreover, the LUS total score showed a partial correlation controlled by age and BMI with pulmonary function as shown in Table 3.

4 | DISCUSSION

Our study describes the LUS characteristics in adolescents and adults affected by CF and shows that the most subjects enrolled (89.7%) had at least one ultrasound abnormality; in particular, the most observed alterations were consolidations followed by interstitial

TABLE 1 Demographical and clinical characteristics of patients with cystic fibrosis.

Total number of patients	29
Male, *n* (%)	14 (48.3%)
Age, years	27.07 ± 9.12
Weight, kg	59.9 ± 11.7
Height, cm	166 ± 8.6
BMI, kg/m^2	21.5 ± 2.8
Diabetes mellitus, *n* (%)	10 (34.5%)
Pancreatic insufficiency, *n* (%)	27 (93.1%)
CFTR mutations	
DeltaF508 heterozygous	12/29 (41.4%)
DeltaF508 homozygous	10/29 (34.5%)
2183AA > G/3276 C > A	1/29 (3.4%)
621 + 1G- > T/2347delG	1/29 (3.4%)
NI303K/G542X	1/29 (3.4%)
G542X/G542X	1/29 (3.4%)
G542X/I507del	1/29 (3.4%)
1717G-D homozygous	1/29 (3.4%)
R31C/UN	1/29 (3.4%)
Bacterial colonization	
Pseudomonas aeruginosa	15/29 (51.7%)
Staphylococcus aureus	11/29 (37.9%)
Achromobacter xilosoxidans	4/29 (13.8%)
Burkholderia cepacia	2/29 (6.9%)
Klebsiella	1/29 (3.4%)

Abbreviations: BMI, body mass index; CFTR, cystic fibrosis transmembrane regulator; UN, unknown

TABLE 2 Main findings in lung ultrasound and spirometry in patients with cystic fibrosis.

Lung Ultrasound (LUS) features (*N* = 29)	
LUS total score	6.00 (2.00–11.00)
A-pattern	29/29 (100%)
B-pattern (interstitial syndrome)	19/29 (65.5%)
Consolidations	21/29 (72.4%)
Combined pattern	16/29 (55.2%)
Pleural irregularity	16/29 (55.2%)
Lingula involvement	12/29 (41.4%)
Pleural effusion	1/29 (3.4%)
Spirometric indices	
FVC (%)	90.00 (78.00 - 102.50)
FEV_1 (%)	75.00 (55.00 – 87.50)
Tiffenau index (%)	82.05 (71.11 – 92.68)
FEF 25–75 (%)	38.00 (21.50 – 57.00)
PEF (%)	86.00 (66.50 – 103.50)

Abbreviations: FVC, forced vital capacity; FEV1, forced expiratory volume in one second; FEF 25–75, forced expiratory flow; PEF, peak expiratory flow.

syndrome. Furthermore, it shows a correlation between LUS score and spirometric values. To date, there are few studies in the literature describing the Ultrasound features of pwCF. CXR, HRCT and MRI are the most widely used imaging techniques for these patients.

LUS in recent years has become widely used for the identification of acute lung diseases both in adult patients and in pediatric age.[16–18] In particular, during the recent pandemic crisis, LUS has proved to be highly sensitive for the identification of pulmonary involvement in Coronavirus disease 19 and also in the referral of dyspnoic patients to intensive care.[19,20]

In the field of CF, previous studies had compared LUS with CXR and with HCRT. Strzelczuk–Judka et al. compared LUS to CXR, creating a LUS score - CF Ultrasound Score and the results showed that this correlated with the conventional modified X-ray Chrispin-Norman score.[4] Peixoto et al., showed that LUS findings were comparable to HRCT results assessed by changes in Bhalla score.[11] Finally, Hassanzad et al. in a recent study highlighted that LUS was superior to CXR and comparable to HRCT for identifying air bronchograms and consolidations.[14]

Analyzing the specific LUS features in our population, consolidations were the most detected alterations (72.4%) in agreement with previous reports.[12,14] Interstitial syndrome, characterized by the B-pattern, was observed in 65.5% of cases in contrast to previous studies who described the B-pattern as the most represented in their patients.[11,14] These different findings could be linked to a higher average age of our patients, who therefore had more damaged lungs.

A notable feature of the LUS in our pwCF was the presence of a consolidation with a static air bronchogram involving the lingula (41.4% of cases), which was predominantly present in adult patients over 18 years of age. Indeed, adults with CF usually showed more damaged lungs than younger patients. To our knowledge, this ultrasound characteristic has never been described in the literature. Previously, Mason et al. in a study that analyzed the type and distribution of bronchiectasis in adults with CF using Computed Tomography showed that these lesions were more represented at the level of the upper lobes and the lingula.[21] In accordance with previous studies, we revealed that more than half of our patients had pleural irregularity at LUS mainly in the posterior superior regions, while pleural effusion was found in only one case.[11] LUS is known to have a high sensitivity for the study of the pleura and pleural cavity, in fact the diagnosis of pleural effusion was its first application.[22] The alterations in the pleural line observed in our study included irregularities, fragmentations, thickness variations and small subpleural consolidations, described also by previous authors.[4,11]

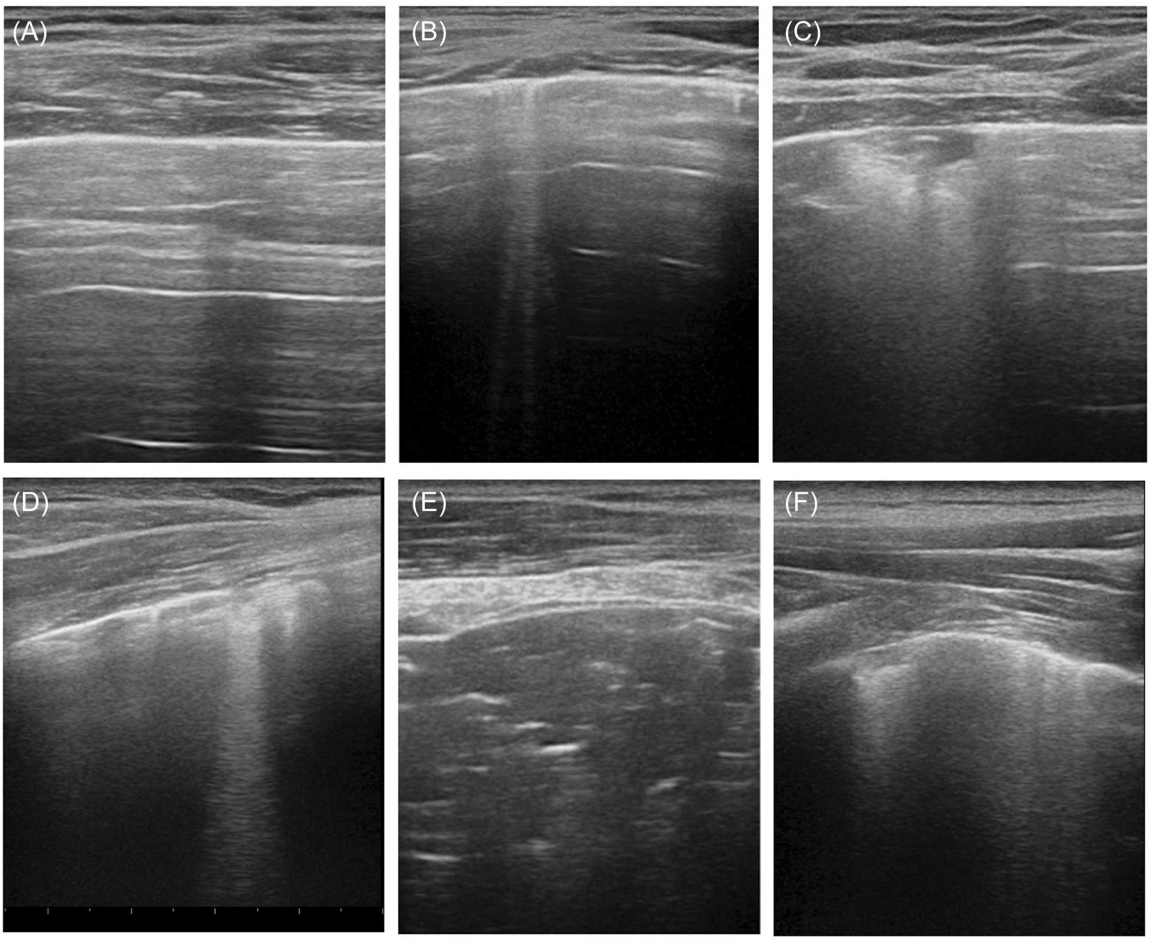

FIGURE 2 (A) A-pattern, represented by horizontal hyperechogenic lines (A-lines), due to reverberation artefacts and expressing the air content of the lung. (B) B-lines, laser-like vertical hyperechoic artifacts extending from the pleural line to the bottom of the screen. (C) Consolidation: area with the solid organ appearance, indistinct margins, with the possibility of air bronchogram. (D) Combined pattern (B-lines + consolidations). (E) Lingula involvement: a large area of consolidation with deep static air bronchogram involving the lingula. (F) Pleural irregularity in association with short B-lines and small areas of subpleural subcentimetric consolidation.

Regarding the secondary objective of the study, we compared the LUS features of pwCF with the respiratory function indices of these subjects (SpO2 and spirometry data). In contrast to a previous study, our data did not show a correlation between LUS findings and SpO2.[11] Instead, consistent with the same authors we observed a negative correlation of the LUS score with the following spirometric indices: FVC, FEV_1, Tiffenau Index and FEF 25–75. These results were predictable, as reduced respiratory functionality is closely dependent on anatomical changes to the lung parenchyma. In literature different reports highlighted the correlation between spirometry and lung structure evaluated by CXR and HRCT.[23,24] Most studies including children and adolescents with CF admitted to the outpatient clinic showed a good correlation between HRCT structural lung damage with spirometric indices (FVC, FEV_1, FEF 25–75, FEV_1/FVC), corroborating its usefulness in evaluation and monitoring of pwCF.[24,25]

Therefore, given the association between lung function and structure and the correlation described in previous studies between LUS score and Bhalla score of the HRCT and LUS score and Chrispin-Norman score of the CXR, we propose to extend the use of the LUS to pwCF during their routine follow-up.

As the LUS is a radiation-free and repeatable examination, it could be performed in all patients, especially pediatric ones, during pulmonary visits in association with spirometry. This imaging tool could also help the physician to detect parenchymal alteration at an early stage, such as lingula consolidations, that otherwise only HRCT could show.

In addition, as the LUS can easily identify lung areas with high secretion accumulation, it may help the physician and physiotherapist to guide the patient during the CPT.

Currently, the diagnosis of PEx is clinical and based on the patient's reported symptoms and objective chest examination. It can be supported by spirometry that in case of PEx shows a reduction in FEV_1 of 10% compared to a previous measure.[26] The LUS in combination with spirometric indices could help the physician make a diagnosis of PEx. Some Brazilian authors, in a previous report describing two cases of PEx in young adults, suggested that LUS could be a useful technique to assess changes

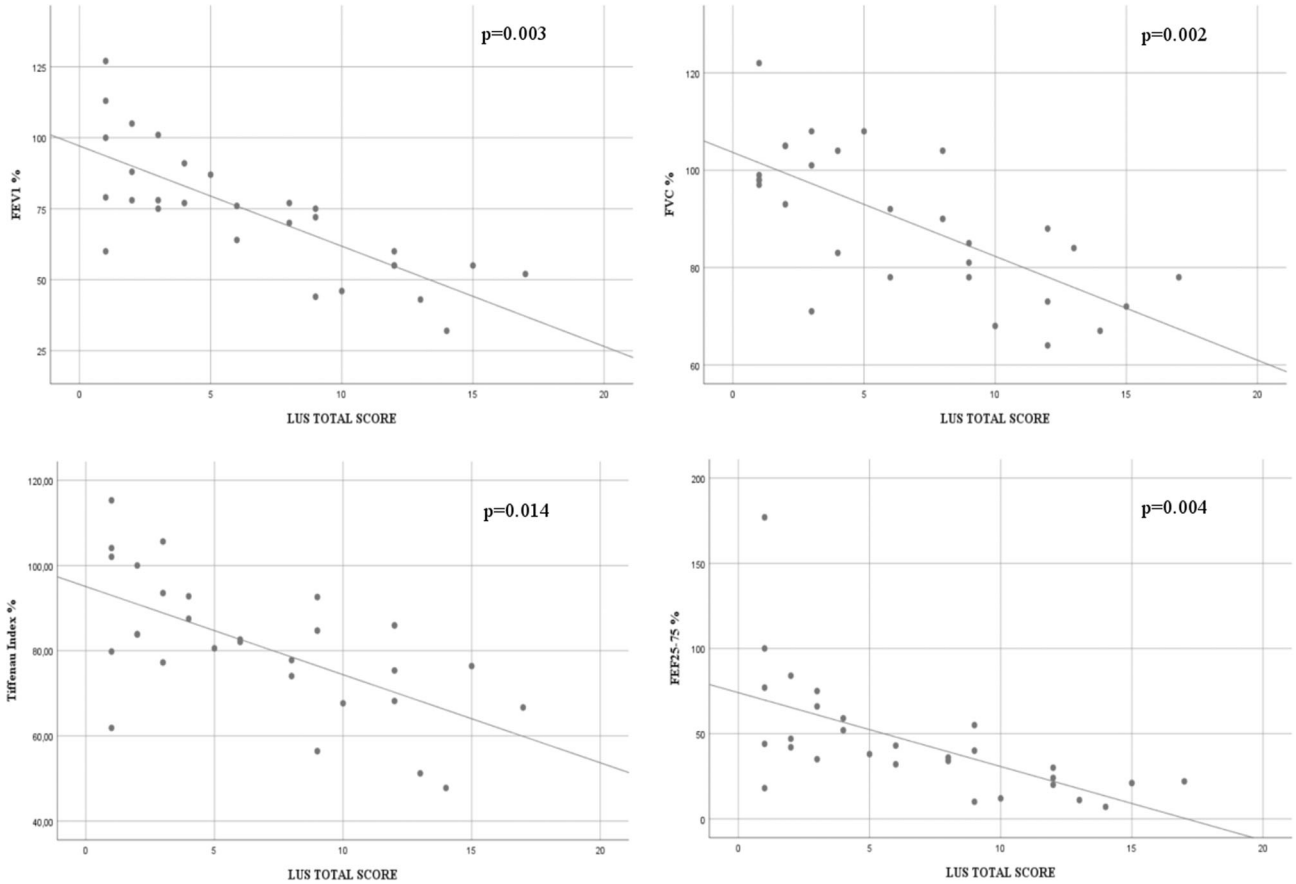

FIGURE 3 Correlation between LUS total score and pulmonary function data measured by spirometry. FEV_1, forced expiratory volume in one second; FVC, forced vital capacity; FEF 25–75, forced expiratory flow.

TABLE 3 Partial correlation controlled by age and by BMI between LUS total score and pulmonary function in subject with cystic fibrosis.

	FVC	FEV_1	Tiffenau Index	FEF 25–75
LUS total score correlation controlled by age	−0.707	−0.775	−0.626	−0.629
p	.000	.000	.000	.000
LUS total score correlation controlled by BMI	−0.716	−0.778	−0.627	−0.619
p	.000	.000	.000	.000

Abbreviations: FVC, forced vital capacity; FEV1, forced expiratory volume in one second; FEF 25–75, forced expiratory flow.

in lung parenchyma during exacerbations and also to monitor the response to antibiotic therapy.[6]

This study has limits. First the monocentric nature and the small number of patients enrolled. Secondary, the heterogeneity of the patients included, both in terms of age - ranging from adolescence to adulthood - and the therapy performed; we recruited patients taking new drugs and those following conventional therapies. Finally, LUS is easy-to-perform technique, but operator-dependent; it must therefore be performed by a certified operator expert in this field to obtain reliable results.

In conclusion, our study describes LUS findings (consolidations and interstitial syndrome) in pwCF. It also showed a correlation between LUS score and the pwCF' lung function measured by spirometric indices. LUS may be useful in routine monitoring of pwCF. Further studies with larger participation and that include younger children are needed to confirm and expand our observations.

AUTHOR CONTRIBUTIONS

Federica Corona: Data curation (equal); Writing—original draft (equal). **Domenica Squillaci**: Data curation (equal); Writing—original draft (equal). **Alessia Saccari**: Investigation (equal); Supervision (equal). **Antonio Chiaretti**: Investigation (equal); Supervision (equal). **Egidio Barbi**: Conceptualization (equal); Writing—review & editing (equal). **Massimo Maschio**: Conceptualization (equal); Writing—review & editing (equal).

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

DATA AVAILABILITY STATEMENT

Data not available.

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SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

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