Standards of care and educational gaps in adult cystic fibrosis units: a European Respiratory Society survey

Almudena Felipe Montiel [1], Antonio Álvarez Fernández[1,2], Mario Culebras Amigo[1], Francesco Blasi[3,4], Andrea Gramegna [3,4], Stuart Elborn[5], Alexander Horsley [6], Susan Madge[7], Daiana Stolz[8], Nathalie Tabin[9], Carine Pannetier[10] and Eva Polverino[1,2]

[1]Department of Respiratory Medicine (Adult Cystic Fibrosis Unit), Vall d'Hebron University Hospital, Vall d'Hebron Research Institute (VHIR), Universitat Autònoma de Barcelona, Barcelona, Spain. [2]CIBER de Enfermedades Respiratorias, Instituto de Salud Carlos III, Madrid, Spain. [3]Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Internal Medicine Department, Respiratory Unit and Cystic Fibrosis Adult Center, Milan, Italy. [4]Department of Pathophysiology and Transplantation, Università degli Studi di Milano, Milan, Italy. [5]Medicine and Faculty of Medicine, Health and Life Sciences at Queen's University and Imperial College London, London, UK. [6]Division of Infection, Immunity and Respiratory Medicine and Adult CF Centre, Faculty of Biology Medicine and Health, University of Manchester, Manchester, UK. [7]Royal Brompton and Harefield NHS Foundation Trust, London, UK. [8]Clinic of Respiratory Medicine and Pulmonary Cell Research, University Hospital Basel, Basel, Switzerland. [9]Education Department, European Respiratory Society, Lausanne, Switzerland. [10]European Respiratory Society, Lausanne, Switzerland.

Corresponding author: Eva Polverino (eva.polverino@vhir.org)

Shareable abstract (@ERSpublications)
Cystic fibrosis (CF) has seen progress, leading to more adult patients. Ageing adds complexity, raising concerns about healthcare. This survey reveals disparities in resources and highlights the need for standardised, high-quality care for CF patients. https://bit.ly/49V8zUg

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Abstract

Background Significant progress in the field of cystic fibrosis (CF) has substantially extended the life expectancy of patients with CF (pwCF). Consequently, the population of adult pwCF has outnumbered paediatric patients in most developed countries. Ageing is a new factor that can contribute to disease complexity and can require adaptation of CF units. Therefore, the necessity for standardised, specialised and multidisciplinary care is imperative. Concerns arise regarding the adequacy of current healthcare, therapeutic and educational offerings.

Methods To address these concerns, a multinational survey was conducted to assess the current state of care in specialised multidisciplinary adult and paediatric CF units and identify areas for improvement. Responses were collected from 44 centres providing regular care to CF patients.

Results The survey unveiled considerable disparities in the availability of critical resources, including diagnostic access, supplementary testing, treatment modalities, transplant and transition programmes, and healthcare professionals' training.

Conclusion This study underscores the urgent need to standardise care across these centres in order to minimise disparities in terms of available resources and training with a particular emphasis on adult pwCF who are becoming more numerous and showing different needs with ageing. The changing landscape of CF in adulthood will require constant monitoring to ensure proper adaptation of the current model of care.

Introduction

Cystic fibrosis (CF) is a multisystemic disease that has historically been considered a childhood illness with a life expectancy not exceeding 18 years due to the high risk of early mortality [1, 2].

In recent years, there has been a favourable evolution in the prognosis of CF, which can be attributed to several factors, for example, early diagnosis, the introduction of new drugs (inhaled antibiotics and, more recently, drugs that functionally correct the defect caused by Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) gene mutations) and the implementation of multidisciplinary care within specialised centres or units [3]. As a result, in recent years the number of adult CF patients has exceeded the number

of paediatric patients [4–6]. According to data from the latest European Cystic Fibrosis Society Patient Registry (ECFSPR), in 2021 54% (n=28 986) of patients with CF (pwCF) were ⩾18 years, a figure that is consistently increasing [7]. Furthermore, the life expectancy of pwCF is steadily increasing, and predictive models based on current findings from various disease registries, both at European and UK level, suggest a shift in the age distribution of CF, with a higher proportion of pwCF reaching older ages [8, 9]. With the ageing of the pwCF population and the emergence of adulthood-related comorbidities [10–13], it has become evident that current care needs to be adjusted to meet the unique requirements of this demographic that differ from those of the paediatric age group. In this sense, the multidisciplinary care model is of paramount importance. The European Cystic Fibrosis Society (ECFS) standards of care [14–17] emphasise the importance of providing pwCF with access to multidisciplinary teams within specialised units. However, despite these recommendations, there is still a lack of uniformity in terms of the access to required services for pwCF and the training of the healthcare teams involved.

The objective of this survey was to describe the current standards of care for pwCF in specialised multidisciplinary care units and to identify potential areas for improvement.

Methods

The study is based on an online survey targeting all members of the European Respiratory Society (ERS), comprising a total of 30 558 individuals, regardless of their potential professional association with CF (see supplementary material).

The survey participation was voluntary, and individuals were invited to participate through an invitation letter containing a link to the survey. The survey consisted of 36 closed multiple-choice questions covering various aspects of care, organisation, training and research related to CF (refer to the supplementary material for details). Data collection was conducted from December 2021 to November 2022. Additionally, a descriptive analysis of the surveyed centres and the questionnaire responses was conducted.

Results

The survey received responses from a total of 44 centres (34 dedicated CF hospital units, 10 hospitals without a dedicated CF centre) across 13 different countries (35 EU and nine non-EU centres). It is important to distinguish between centres dedicated to the care of children with CF, adults with CF, and those combining care for both age groups. To avoid data duplication, survey responses from the same centre were carefully discarded. The response rate per centre ranged between 90% and 95% and all responders were physicians. Only the available responses to survey questions were counted for the analysis. Details on the distribution of responses by country and number of centres participating in the survey are given in table 1.

Care activity

72.7% (n=32 out of 44) of all the surveyed centres provided care for children and adults with CF, while 15.9% (n=7 out of 44) of the centres provided only paediatric care, and 11.4% (n=5 out of 44) only

TABLE 1 Distribution of responses by country and centre

Country	Number of centres
Countries from the EU region (nine countries, 35 centres)	
Spain	20
Germany	5
Italy	3
Macedonia	2
Bulgaria	1
Moldova	1
Portugal	1
Romania	1
Turkey	1
Countries from non-EU regions (four countries, nine centres)	
UK and Northern Ireland	6
Australia	1
Chile	1
Malaysia	1

EU: European Union.

adult care. The annual number of patients varied among centres, with only 27.7% (n=10 out of 44) providing care for >250 patients, most saw between 101 and 250 patients (29.6%; n=13 out of 44) followed by centres seeing <50 patients (27.3%; n=12 out of 44) and finally, 20.5% seeing between 50 and 100 patients (n=9 out of 44). In >80% of the centres surveyed (n=32 out of 40) the experience in terms of management of both adult and paediatric pwCF was >10 years.

Organisational activity

Resources and space

Most of the centres had specialised personnel available for pwCF, both at the outpatient and inpatient levels (table 2). The multidisciplinary team was in general quite complete in most centres with a few exceptions, such as the presence of other medical specialists (for diagnosis, follow-up and prevention of comorbidities associated with the disease) and nurse case managers (both <60%) in adult centres, pharmacists in paediatric and mixed centres (<60%), geneticists in adult and mixed centres (<50%) and research personnel such as research nurses and clinical trial coordinators in paediatric and mixed centres (<30% overall). In terms of resources the coverage was broad with the exception of neonatal screening (<60%) and availability of lung clearance index (LCI) testing (<14%) in paediatric centres. Only 58.1%

TABLE 2 Multidisciplinary teams and resources according to centre

	Paediatric centres	Adult centres	Mixed centres	Total
Centres n	7	5	32	44
Cystic fibrosis multidisciplinary team				
Paediatric pulmonologist	7 (100)	0 (0)	30 (93.8)	37 (84.1)
General paediatrician	5 (71.4)	0 (0)	11 (34.4)	16 (36.4)
Cystic fibrosis specialist pulmonologist	3 (42.9)	5 (100)	24 (75)	32 (72.7)
Other medical specialists	2 (28.6)	3 (60)	18 (56.3)	23 (52.3)
Clinical microbiologist	6 (85.7)	5 (100)	22 (68.8)	33 (75)
Medical support from trainee(s)	3 (42.9)	4 (80)	16 (50)	23 (52.3)
Clinical nurse specialist	5 (71.4)	5 (100)	23 (71.9)	33 (75)
Nurse case managers	3 (42.9)	3 (60)	6 (18.8)	12 (27.3)
Research nurse	1 (14.3)	4 (80)	7 (21.9)	12 (27.3)
Specialist physiotherapist	7 (100)	5 (100)	24 (75)	36 (81.8)
Specialist dietitian	5 (71.4)	5 (100)	22 (68.8)	32 (72.7)
Clinical psychologist	5 (71.4)	5 (100)	20 (62.5)	30 (68.2)
Social worker	7 (100)	5 (100)	16 (50)	28 (63.6)
Specialist pharmacist	4 (57.1)	4 (80)	19 (59.4)	27 (61.4)
Clinical geneticist	6 (85.7)	1 (20)	16 (50)	23 (52.3)
Database coordinator	0 (0)	3 (60)	9 (28.1)	12 (27.3)
Clinical trial coordinator	1 (14.3)	4 (80)	9 (28.1)	14 (31.8)
Resources				
Analysis of sweat	6 (85.7)	4 (80)	31 (96.9)	41 (93.2)
Glucose tolerance test	7 (100)	5 (100)	31 (96.9)	43 (97.7)
Genetics service	7 (100)	4 (80)	29 (90.6)	40 (90.9)
Neonatal screening	4 (57.1)	3 (60)	26 (81.3)	33 (75)
Microbiology laboratories	7 (100)	5 (100)	32 (100)	44 (100)
Spirometry	7 (100)	5 (100)	32 (100)	44 (100)
Plethysmography	5 (71.4)	3 (60)	27 (84.4)	35 (79.5)
Lung clearance index	1 (14.3)	3 (60)	12 (37.5)	16 (36.4)
Cardiopulmonary exercise testing	4 (57.1)	5 (100)	22 (68.8)	31 (70.5)
Walking exercise test	6 (85.7)	5 (100)	28 (87.5)	39 (88.6)
Polysomnography	5 (71.4)	5 (100)	29 (90.6)	39 (88.6)
Noninvasive ventilation inpatient	5 (71.4)	5 (100)	29 (90.6)	39 (88.6)
Home ventilation	6 (85.7)	5 (100)	26 (81.3)	37 (84.1)
Nuclear medicine service	3 (42.9)	5 (100)	26 (81.3)	34 (77.3)
Interventional radiology	4 (57.1)	5 (100)	24 (75)	33 (75)
Bronchoscopy	6 (85.7)	5 (100)	29 (90.6)	40 (90.9)
Gastrointestinal endoscopies	6 (85.7)	5 (100)	28 (87.5)	39 (88.6)
High-frequency pure tone audiometry	5 (71.4)	4 (80)	18 (56.3)	27 (61.4)
Other (oscillometry)	0 (0)	1 (20)	1 (3.1)	2 (4.5)

Data are presented as n (%).

(n=25 out of 43) and 53.5% (n=23 out of 43) had access to liver and lung transplant programmes, respectively, either at the same centre or a partner centre.

Access to and funding of the different drugs was variable. Tobramycin and DNase were the most commonly funded drugs, 97.7% (n=42 out of 43) and 93% (n=40 out of 43), respectively. At the time of survey completion, triple therapy with CFTR modulators (ivacaftor, tezacaftor and elexacaftor) was not available in 13.9% of centres (n=6 out of 43) for any reason including lack of approval or reimbursement. Ivacaftor was the most funded modulator (83.7%; n=36 out of 43), followed by the combination of lumacaftor–ivacaftor (78.6%; n=33 out of 42) and tezacaftor–ivacaftor (76.7%; n=33 out of 43). Importantly, between 2% and 7% of all centres had only partial funding for CFTR modulators (one or more). Similarly, between 2% and 4% of centres had partial funding for mannitol (28.6%; n=12 out of 42), levofloxacin (27.9%, n=12 out of 43) and inhaled aztreonam (20.9%, n=9 out of 43).

Outpatient and inpatient care

At the outpatient level, 56.8% (n=25 out of 44) had specific sites for the exclusive treatment of pwCF, whereas at the hospital level, only 43.2% of the centres surveyed (n=19 out of 44) offered exclusive hospital facilities for pwCF, while in the remaining cases healthcare was shared with other non-CF patients. Likewise, 72.1% of the centres (n=31 out of 43) had an emergency service available, and more than half of the centres (56.8%; n=25 out of 44) had care provided by a CF specialist physician 24 h a day, every day of the week.

Of the centres surveyed, 95.3% (n=41 out of 43) practised a cross-infection policy of physical segregation both inside and outside the CF unit during hospitalisation. In outpatients, 51.2% (n=22 out of 43) of the centres segregated all patients according to microbiological status, and 37.2% (n=16 out of 43) only applied segregation for certain microbiological isolates (*Pseudomonas aeruginosa*, *Burkholderia cepacia* complex, methicillin-resistant *Staphylococcus aureus* and non-tuberculous mycobacteria). At the hospital level, most patients had access to single rooms (60.5%; n=26 out of 43), and 34.9% (n=15 out of 43) shared rooms with other patients with the same microbiological isolates.

Over half of the centres surveyed (55.8%; n=24 out of 43) saw patients at least four times a year as a minimum (approximately every 3 months), although in 39.5% of the cases (n=17 out of 43), visits could be more than six per year.

The majority of centres had access to spirometry on the same day of the visit as well as the various resources considered necessary for the acute management of pwCF (table 3).

Transition and communication

A transition programme between paediatricians and pulmonologists was available in 69% of the centres (n=29 out of 42). The availability of multidisciplinary teams at the time of transition was 81.4% (n=35 out of 43).

Regarding the availability of written information (pamphlets) for patients, 59.5% of the centres (n=25 out of 42) had informative brochures. 100% of the surveyed centres had a communication channel between physician and patient in case of need. The most frequent communication channel was a land-line (80.9%; n=34 out of 42), followed by e-mail (78.6%; n=33 out of 42) and text message by phone applications (33.3%; n=14 out of 42). Some of the centres (11.9%; n=5 out of 42) confirmed that they communicated *via* mobile applications, social networks or personal phones of the different healthcare professionals.

The vast majority of the centres (95.3%; n=41 out of 43) reported collaborating with local and/or national associations and societies.

Training and education of healthcare personnel

One of the survey items was the training and education of the healthcare personnel involved in the CF units. A total of 79.5% (n=35 out of 44) of the surveyed centres claimed to have sufficient training for the

TABLE 3 On site resources at centres to manage cystic fibrosis exacerbations

Spirometry on the same day as the consultation	81.4% (n=35 out of 43)
Nebulised antibiotic treatment	74.4% (n=32 out of 43)
Intravenous antibiotic treatment	72.1% (n=31 out of 43)
Placement and maintenance of venous access	72.1% (n=31 out of 43)

management of pwCF, but only 53.5% (n=23 out of 43) of the centres had specific obligatory or optional CF rotations for residents during their academic training. Centres declaring insufficient training in CF (20.4%; n=9 out of 44) identified the need to improve knowledge about new modulators (66.7%; n=6 out of 9), current research on pathophysiology (55.5%; n=5 out of 9), antibiotic treatment (55.5%; n=5 out of 9), lung and liver transplant programmes (44.4%; n=4 out of 9) and finally, on follow-up (33.3%; n=3 out of 9) and diagnosis (22.2%; n=2 out of 9). The format chosen for training by most centres was the mixed online and face-to-face course 84.9% (n=8 out of 9), followed by monograph-type review documents in paper and/or online format in 55.5% (n=5 out of 9). Face-to-face and online courses were the least requested by respondents with 33.3% (n=3 out of 9) in both cases.

Research

Almost 70% of the centres (69.8%; n=30 out of 43) participated in the ECFSPR. Nearly 32% of centres (31.8%; n=14 out of 44) had a study coordinator and 27.3% (n=12 out of 44) had a data entry and a research nurse, more often present in adult than paediatric centres (table 2). Most centres (85.7%; n=36 out of 42) had the facility to include patients in clinical trials; however, only 19.1% (n=8 out of 42) had participated in five to 10 clinical trials in the last 5 years, while the vast majority (66.7%; n=28 out of 42) reported less than five clinical trials. Only 30.2% (n=13 out of 43) of the centres reported more than five scientific publications on CF per year.

Discussion

The results of this survey show that many of the CF centres or specialised CF units comply with most of the standards of care of the ECFS [14–16]. However, considerable gaps were observed, as well as relevant differences between centres and between paediatric cases and adults.

According to this survey, many CF centres have not adapted to the increasing proportion of adult patients, given that in 25% of mixed care centres, adult patients continue to be seen by paediatric pulmonologists. In fact, in the survey by Madge *et al.* [18] 50% of pwCF were still admitted to paediatric wards. Nevertheless, current trends show that the availability of adult physicians for pwCF is likely to increase progressively over time.

The new CF patient profile needs adjustments within the multidisciplinary team to provide comprehensive care. Encouraging the inclusion of transition protocols in all centres and of other specialist physicians to support pulmonologists in managing pwCF is crucial. However, our survey revealed that only 52.3% of centres had received support from other specialist physicians for disease diagnosis, follow-up and prevention of associated comorbidities. Additionally, access to liver and lung transplantation programmes is still limited, with only 58.1% and 53.4% of centres offering such services, respectively. These percentages are far lower than standard references and the data published by Madge *et al.* [18] in 2017. The observed findings may be the result of either a selection bias stemming from the composition of participating centres in the current survey or an actual shift in the medical care provided by current CF centres.

Access to a geneticist, present in only 52.3% of the centres surveyed, is crucial in a CF centre to provide support in the therapeutic management of selected mutations. Moreover, the geneticist is useful to complete patients' registry data in coordination with the data manager, a figure present in only 27.3% of the centres.

Pharmacists also play a crucial role in monitoring treatment compliance, as well as identifying potential interactions and adverse effects, yet only 61.4% of the centres surveyed had access to these specialists; however, this percentage has increased with respect to the 2017 study [18].

The psychosocial sphere of pwCF should be equally important. Therefore, it is of paramount importance to include psychologists and social workers in the team to help patients during transitional periods, for example, young adulthood, entering into working life, the transplant waiting list and the start of new treatments. Nevertheless, in this survey, 30–40% of the centres included did not have psychologists and social workers. Note that this percentage was even higher in CF centres or units providing both paediatric and adult care. All of this underscores the necessity for a coordinated transition between paediatric and adult care. However, transition programmes are still not implemented in 30% of the centres included in the study despite having a multidisciplinary team in >80% of cases. This striking discrepancy may reflect a heterogeneous level of implementation of standards of care across centres, and possibly a local shortage of dedicated resources in some centres.

Regarding resources, most of the centres have demonstrated that they have the diagnostic and therapeutic tests and techniques necessary for the correct management of the disease. However, the availability of LCI testing is still not integrated in disease management globally. Specifically, only 14% of the paediatric care centres participating in the survey and 60% of the adult centres have LCI testing in place. This is in contrast to the vast literature on LCI from paediatric groups. On the other hand, this finding is in line with the observation that most paediatric centres in this survey do not include dedicated research personnel in their team (research nurse, clinical trial coordinator). As expected, the availability of interventional radiology and nuclear medicine is greater in adult centres, as they typically handle more advanced cases of CF.

Over 50% of the services offered to pwCF are in the outpatient setting, and many procedures previously performed only in the hospital setting, such as the treatment of exacerbations, have now been outsourced. This model has proved to be preferable, particularly with pwCF. However, with the growing number of adult pwCF, it is likely that new approaches to care for this new, older and more complex patient profile will be needed. To fulfil these challenges, the multidisciplinary team will undoubtedly play a key role in shaping and implementing innovative care strategies.

The survey reveals that 57% of the centres have outpatient services dedicated solely to the care of pwCF and inpatient beds in only 43%. This could be insufficient in the context of a complex disease with increasing incidence of comorbidities. The number of exclusive care units is usually linked to the availability of more specialised personnel (nurses, physiotherapists, *etc*.) and a lower risk of cross-infection.

The survey also highlights that nowadays the availability of drugs with established efficacy in CF, such as mannitol, levofloxacin and aztreonam, is still very low in some cases. This generates significant disparity in access to treatment for pwCF. In addition, between 2% and 7% of the centres rely on private co-funding for CF drugs (not public/governmental entities).

A significant percentage of physicians caring for pwCF (20.4%) consider that they lack sufficient training in disease management. Additionally, only 53% of the centres include a rotation in CF units as part of their academic training. It is likely that access to academic training, particularly in postgraduate education, is influenced by the category of the hospital. Nevertheless, given the complex nature of this systemic disease, it is essential to have highly qualified personnel that are capable of managing CF effectively. All hospitals with CF units should prioritise specialised training for its healthcare professionals to ensure the proper functioning of these units and to guarantee equal access to the best possible care for pwCF at different stages of their lives.

Finally, the collaboration between CF units, patient associations and scientific societies can be vital in promoting training and facilitating better scientific programmes. Such collaborations will enable professionals to stay up to date with and well prepared for the latest advances and knowledge in the field of CF. A better education is the basis to promote scientific publications and engagement in clinical trials.

Our data clearly highlight the shortage of research personnel in CF centres, including nurses, database coordinators and study coordinators, especially in paediatric units, which limits scientific research and progress.

Limitations

The methodology employed in the present study, which relies on a survey, does come with inherent limitations, as well as potential information bias derived from individuals participating in the survey. Furthermore, the low percentage of responses from some Eastern European, Asian and American countries combined with a disproportionately high representation of centres from countries such as Spain, the UK/Ireland and Germany may significantly affect the homogeneity of the presented sample. Since the invitations to take part in the present survey were sent to ERS members, this inhomogeneous distribution of responses is likely to reflect the scarce participation of many CF centres in ERS activities. This implies the need to reinforce the scientific activities of the ERS in CF in view of the increasing need of adult CF centres after the introduction of CFTR modulators and prolonged life expectancy.

Finally, the findings of this survey may be biased towards clinical practice in Central/Western Europe, leading to some aspects being over- or underestimated. Therefore, a more ambitious project should be launched to provide a uniform representation of centres and more robust data. A strength of this survey is that it provides a clear overview of the potential gaps and challenges to be addressed in the future for better care of pwCF.

Conclusions

As the adult CF population continues to grow in disease complexity related to ageing (comorbidities, *etc.*), the need for homogeneous, specialised and multidisciplinary care becomes increasingly crucial. Various studies have consistently demonstrated that care provided in specialised centres with dedicated multidisciplinary teams leads to higher patient satisfaction and improved clinical outcomes for pwCF.

The present study reveals that the care of adult pwCF can be improved, further particularly in terms of resources (multidisciplinary care) and education. First, there is still a considerable proportion of adult patients waiting to be referred to adult centres. Second, adult CF centres often lack other specialists to support care of potential comorbidities related to ageing. Furthermore, they rarely have a geneticist in the team to support the late diagnosis of the disease (usually in milder or rarer forms of CF). Similarly, the centres' availability of plethysmography and LCI testing and access to liver and lung transplantation should be improved in view of the increasing proportion of patients with longer life expectancy. Lastly, there is a clear educational need to ensure specialised care, since rotations in CF centres are uncommon and numerous gaps were identified.

Current data strongly support specialised centres as the model of care for adult pwCF. However, there is still a pressing need to homogenise the care provided in these centres. This can be achieved by expanding the capacity of existing units and providing them with the necessary resources, training health professionals and creating new adult CF centres where needed. Another critical aspect is ensuring equal access to the drugs needed for managing the disease. This implies close collaboration between healthcare professionals and policy makers to ensure that the growing adult pwCF population continues to receive high-level care through appropriate monitoring and adaptation of the model of care.

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