

Original

A comparative study among adolescents with cystic fibrosis and primary ciliary dyskinesia and their caregivers: psychological aspects

Selene Valero-Moreno¹, Inmaculada Montoya-Castilla², & Marián Pérez-Marín³

¹Assistant Professor, Personality, Assessment and Psychological Treatments Department, Faculty of Psychology, Universitat de Valencia

²Full Professor, Personality, Assessment and Psychological Treatments Department, Faculty of Psychology, Universitat de Valencia

³Associate Professor, Personality, Assessment and Psychological Treatments Department, Faculty of Psychology, Universitat de Valencia

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ABSTRACT

This work aimed to conduct a comparative study between cystic fibrosis and primary ciliary dyskinesia among adolescents and their caregivers to understand their emotional adaptation to the disease. Thirty-nine dyads of adolescents aged 9–18 years and their primary caregivers were assessed, 69.20% of whom had a diagnosis of cystic fibrosis. For the adolescents, anxious-depressive symptomatology was assessed using the HADS, perceived threat of disease (B-IPQ) and quality of life (CRQ-SAS). For the caregivers, emotional symptomatology (HADS) and perceived stress levels (PIP) were assessed. Mean comparisons were made according to diagnosis and severity of anxious-depressive symptomatology, relationships between variables were studied, and qualitative comparative analysis models, QCA, were performed. Higher levels of anxiety and depression were found in the caregiver than in the adolescent. Adolescents with primary ciliary dyskinesia have a more significant emotional impact than adolescents with cystic fibrosis. However, no differences were found in caregivers according to diagnosis. The adolescent's emotional state, quality of life and perceived threat of disease were related to and explained by the caregiver's emotional state, and vice versa. Therefore, there appears to be a certain degree of emotional contagion between the dyad members. This is a preliminary study that has not been carried out before and can help to understand the psychological aspects associated with these diseases to favour the patient's adjustment and thereby ensure more effective management of the disease and adherence to treatment.

Estudio comparativo entre adolescentes con fibrosis quística y discinesia ciliar primaria y sus cuidadores: aspectos psicológicos

RESUMEN

El objetivo fue realizar un estudio comparativo entre la fibrosis quística y la discinesia ciliar primaria entre adolescentes y sus cuidadores para comprender su adaptación a la enfermedad. Evaluando 39 díadas de adolescentes de entre 9 y 18 años y sus cuidadores, el 69,20% de los cuales tenían un diagnóstico de fibrosis quística. Para los adolescentes, se evaluó la sintomatología ansioso-depresiva mediante el HADS, la amenaza percibida de la enfermedad (B-IPQ) y la calidad de vida (CRQ-SAS). Para los cuidadores, se evaluó la sintomatología emocional (HADS) y los niveles de estrés percibido (PIP). Se realizaron comparaciones de medias según el diagnóstico y la sintomatología ansioso-depresiva, se estudiaron las relaciones entre variables y se realizaron modelos de análisis cualitativo comparativo. Se encontraron mayores niveles ansioso-depresivos en el cuidador que en el adolescente. Los adolescentes con discinesia ciliar primaria presentan un impacto emocional mayor que los adolescentes con fibrosis quística. No se encontraron diferencias en los cuidadores según el diagnóstico. El estado emocional, la calidad de vida y la percepción de amenaza de la enfermedad del adolescente estaban relacionados con el estado emocional del cuidador y se explicaban por él, y viceversa.

* Corresponding author.

E-mail address: marian.perez@uv.es (M. Perez-Marín).

Por lo tanto, parece existir cierto grado de contagio emocional entre los miembros de la diada. Se trata de un estudio preliminar que no se ha realizado previamente y que ayuda a comprender los aspectos psicológicos asociados a estas enfermedades favoreciendo el ajuste del paciente y asegurar así un manejo más eficaz de la enfermedad y la adherencia al tratamiento.

The World Health Organisation (WHO) notes the importance of mental health in the entire population, especially in the adolescent population (World Health Organization, 2021). Adolescence is a vulnerable stage for emotional disorders to develop and worsen. According to WHO data (World Health Organization, 2021), one in seven young people between 10 and 19 years has a mental disorder, accounting for 13% of the global disease burden in this age group (Shorey et al., 2022).

Depression, anxiety and behavioural disorders are among adolescents' leading causes of serious health problems (Shorey et al., 2022). Preventing and alleviating these problems is, therefore, a priority. Failure to address adolescent mental health disorders can lead to significant consequences that extend into adulthood (Copeland et al., 2021), harming their physical and mental health and limiting their chances of achieving a fulfilling life.

The onset of these emotional disturbances may increase when a rare disease such as cystic fibrosis (CF) or primary ciliary dyskinesia (PCD) is diagnosed. Previous studies on CF indicate that symptoms of anxiety and depression are two to three times more frequent among CF patients than in the general population (O'Hayer et al., 2021). These symptoms are associated with decreased adherence to treatment, reduced lung function (Smith et al., 2016), poorer health-related quality of life and healthcare costs (Graziano et al., 2020; Tickner et al., 2022). However, despite significant recent medical progress affecting CF, there is a lack of research on the impact on mental health in patients with CF or PCD. Studies on PCD are scarce (Carotenuto et al., 2013; Author 1, Castillo-Corullón, et al., 2020), but similar results are found with CF. Depression and anxiety rates in these diseases have ranged from 20% to more than 51% (Catastini et al., 2016; Georgiopoulos, 2020; Guta et al., 2021). Among caregivers of children with chronic respiratory diseases, there are also high rates of depression, with high percentages found above clinical thresholds (Graziano et al., 2020). Caregivers' coping with chronic illness may influence the course of the illness and the emotional state of the adolescent (Yuksekgonul et al., 2020). Caregivers of adolescents with a chronic illness showed small to moderate increases in overall stress (Casaña-Granell et al., 2021; Pinquart, 2018; Tan et al., 2018), and stress may interfere with treatment adherence by affecting how the family monitors the adolescent's symptoms and makes treatment decisions (Continisio et al., 2020; Wood et al., 2015).

There are currently few studies analysing CF and PCD together and among caregivers and patients. Therefore, this study aims to analyse the emotional impact of CF or PCD on adolescents and their caregivers through predictors such as parental stress, disease threat and quality of life. The hypotheses are H1. adolescents and caregivers will show elevated depression and anxiety rates; H2. there will be no differences based on diagnosis; H3. the adolescent emotional adjustment will be positively associated with quality of life and negatively associated with disease threat.; H4. caregiver emotional adjustment will be positively associated with stress; H5. A positive association is expected between caregiver and adolescent emotional adjustment.

Methods

Participants

A total of 39 caregiver-patient dyads participated. In the case of adolescents with CF and PCD, their ages ranged from 9 to 18 years ($M=12.23$; $SD=3.16$), with 48.7% being male and 51.3% female. For caregivers, the main caregiver was a relative of the patient diagnosed with CF or PCD, the family member who was mainly responsible for the care of the adolescent both at home and regarding the disease. The most frequent relationship in the study was that of the paediatric patient's mother, with 88.60% ($n=31$). The mean age of the main caregivers was 43.18 years ($SD=5.18$).

Measures

Emotional adjustment: the Hospital Anxiety and Depression Scale (HADS) was used to assess emotional adjustment, which was measured through anxious-depressive symptomatology in the case of caregivers (Zigmond & Snaith, 1983), and the version for chronically ill adolescents was used in the case of adolescents (Author 1 et al., 2019). Regarding the internal consistency of the scale, a validation study in the Spanish adult population found that the values for the anxiety scale ranged between .68 and .93 and for the depression scale between .67 and .90 (Ibáñez & Caro, 1992). For the adolescent population, in the corresponding study (Author 1 et al., 2019), the values for the anxiety scale were .77, and for the depression scale .61.

Quality of life (QoL): The Chronic Respiratory Questionnaire (CRQ), in its self-administered version translated into Spanish, was used to assess health-related quality of life in adolescents (Vigil et al., 2011). The present study used a new version adapted and validated for patients with paediatric chronic diseases (9-18 years) (Author 1 et al., 2019), showing adequate reliability indices of .85 for the total scale.

Threat of disease: the Brief Illness Perception Questionnaire (B-IPQ) was used (Broadbent et al., 2006). It measures the patient's cognitive and emotional representations of their illness. A new version was adapted and validated for adolescents (9-16 years) (Author 1, Lacomba-Trejo., 2020). The results of the analyses performed showed good internal consistency (Broadbent et al., 2006; Author 1 Lacomba-Trejo, et al., 2020).

Perceived stress: the Pediatric Inventory for Parents (PIP) was used for caregivers (Casaña-Granell et al., 2018; Streisand et al., 2001). It assesses the stress of parents taking care of a chronically ill child. Optimal reliability coefficients were obtained, being 0.78 for the total frequency scale and 0.81 for the total effort scale (Streisand et al., 2001).

An ad hoc questionnaire was used to review the patient's medical records in consultation with a pulmonologist to record data related to medical issues and their interpretation adequately. The variables recorded were:

- Characteristics of the disease itself: for CF, the presence of pancreatic insufficiency, the level of pulmonary insufficiency, the presence of pseudomonas infection and the type of pseudomonas infection (chronic or not) were recorded. For both CF and PCD,

spirometry values and the presence of bronchiectasis were recorded.

- Medical treatment: the type of treatment taken was recorded, differentiating between aerosol, nebulised and pill therapy.
- Other: time since diagnosis (months) and number of hospital admissions.

Design

The study consisted of a one-pass, cross-sectional design. The procedure involved identifying all patients with CF or PCD who attended paediatric pulmonology consultations at the hospital. For paediatric patients, the inclusion criteria were to be aged between 9 and 18 years and diagnosed with one of the above-mentioned rare diseases. Participants who met these diagnostic criteria and agreed to participate in the study by signing the informed consent form and their caregivers were included. For exclusion criteria, patients were excluded if, despite meeting the criteria, they had a previously diagnosed physical or psychological pathology, or cognitive difficulties. This study was reviewed and approved by our institution's ethics committee.

Data Analysis

First, descriptive analyses of the variables assessed were carried out. Subsequently, mean comparisons were performed according to diagnosis. Thirdly, the relationships between the variables under study were examined within the dyad, and, finally, the qualitative comparative analysis (QCA) was performed.

QCA allows for quantitative analysis of a small number of cases, using Boolean algebra as a formal tool to identify which of a series of factors (causal conditions) are associated with the presence of a given outcome (outcome condition). It proposes pathways (combining a given interaction between the study variables) to optimise the prediction of the independent variable under analysis (Ragin, 2008). The IBM SPSS Statistics 26 software package and fsQCA 3.0 software package were used.

Results

Descriptive data of the study cohort

69.20% (n=27) of the adolescents were diagnosed with CF, compared to 30.80% (n=12) for PCD. Among patients with CF, 77.80% (n=21) had pancreatic insufficiency and 42.80% (n=11) had mild pulmonary insufficiency, 53.80% (n=14) had moderate pulmonary insufficiency and 3.80% (n=1) had no pulmonary insufficiency. For CF, 76.80% (n=24) suffered pseudomonas infection at the time of the assessment, and 60% (n=20) had chronic pseudomonas infection. Altogether 54.1% (n=21) of the patients had bronchiectasis. Regarding the type of treatment, 61.50% (n=24) were taking aerosols, 66.70% (n=26) were on nebulised therapy, and 74.40% (n=29) took medications in pill doses such as antibiotics, creon and others.

Regarding the time of diagnosis, these adolescents were diagnosed with a mucociliary bacillary disease for a mean of 99.55 months (around 8 years) (SD=51.73). Finally, for the number of hospital admissions, the mean number of admissions was 2.88 (SD=2.99), with a minimum of 0 and a maximum of 15.

The mean spirometry values among patients were: 99.43 for FVC (minimum 65.30 and maximum 133.10), 95.36 for FEV₁ (minimum 60.70 and maximum 133.10) and 83.62 for the ratio between FVC and FEV₁ (minimum 57.62 and maximum 101.80).

Descriptive analyses of the psychological variables of the dyad

Adolescents

Quality of life

Overall, adolescents reported high QoL, with a minimum score of 1 and a maximum score of 7, a mean score of 5.94 for total QoL (SD=0.77). For the remaining dimensions, the subscale with the lowest score for the remaining dimensions was emotional functioning (M=5.62; SD=0.97). The dimension with the highest score was perceived dyspnea (M= 6.62; SD=0.85).

Anxious-depressive symptomatology

The mean scores obtained for anxious symptomatology were 4.15 (SD=3.66; Range 0-18; 1.46 for depressive symptomatology (SD=2.39; Range 0-15) and 5.61 for general emotional distress (SD=5.48; Range 0-33). When scores were recorded under the cut-off values to obtain the categories, it was found that 79.50% (n=31) presented an absence of anxious symptoms, 94.90% (n=37) absence of depressive symptoms and 89.70% (n=35) an absence of emotional discomfort. For the presence of anxious symptomatology, 12.80% (n=5) fitted into this category. Finally, regarding the percentage of cases presenting a psychological disorder that required psychological care, 7.70% (n=3) for anxiety, 5.10% (n=2) for depression and 10.30% (n=4) for general distress.

Perceived threat of disease

Disease threat levels were 22.46 (SD=11.16; Range 0-50). This suggests an underestimation of the threat level. When asked about the causes of the disease, the responses were: 56.40% (n=22) indicated that they were unaware of the cause of their disease; 28.20% (n=11) indicated genetic issues; 7.8% (n=3) indicated that they were born this way and the remaining 7.8% (n=3) reported respiratory problems related to mucus accumulation or respiratory malfunction.

Caregivers

Anxious-depressive symptomatology

The mean scores obtained for anxious symptomatology were 8.06 (SD=34.05; Range 0-21; 4.53 for depressive symptomatology (SD=3.42; Range 0-21) and 12.59 for general emotional distress (SD=7.07; Range 0-42). When the scores were recorded according to the cut-off values to determine the categories, 50% (n=17) showed an absence of anxious symptoms, 82.40% (n=28) an absence of depressive symptoms and 88.20% (n=30) an absence of emotional distress. For the presence of anxious symptomatology, 35.30% (n=12) fitted into this category; 8.80% (n=3) for depressive symptomatology. Finally, for the percentage of cases presenting a psychological disorder requiring psychological care, 14.70% for anxiety (n=5); 8.80% (n=3) for depression and 11.80% for general distress (n=4).

Stress

Total stress scores on the frequency dimension (referring to the number of occasions they encounter such situations) were 3.33 (SD = 0.69; Range 1-5): indicating moderate stress scores. On the effort or discomfort dimension (how much discomfort or interference it

causes to the caregiver), the score was: 3.97 ($SD=0.95$; Range 1-5), slightly higher for this dimension. The rest of the sub-dimension scores are shown in Figure 1.

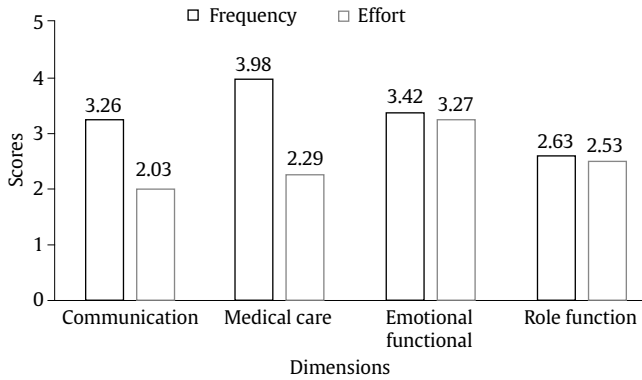


Figure 1. Mean stress scores on the PIP sub-dimensions
 Note. The minimum score is 1 and the maximum score is 5, with a higher number being a higher stress score.

Mean comparisons according to diagnosis

Statistically significant differences were only found between PCD and CF in the anxious symptomatology of adolescents. Adolescents with PCD were found to have higher mean scores than adolescents with CF on anxious symptomatology ($t=-2.18$, $p=0.047$, $d=0.99$ $M_{CF}=3.15$, $SD_{CF}=2.43$; $M_{PCD}=6.42$, $SD_{PCD}=4.93$).

To investigate differences in anxious-depressive symptomatology in adolescents and their primary caregivers, a chi-square test was conducted across categories according to diagnosis, as shown in Table 1. Differences were found in both anxious and depressive symptomatology, once again showing that a greater number of cases of adolescents with PCD fitted into categories related to

greater severity of this symptomatology compared to their peers diagnosed with CF.

Mean comparison according to emotional adjustment of the dyad

First, mean differences were analysed for adolescent variables according to the categories of caregiver anxiety and caregiver depression. Regarding the threat of disease, it was found that adolescents whose caregivers belonged to the psychological anxiety disorder category had a greater perceived threat of disease.

The same procedure indicated in table 2 was performed for depressive symptomatology. The results showed that when caregivers fitted in the higher severity categories, adolescents reported higher scores on anxious symptomatology and a lower QoL in general. Thus, the results indicate that the caregiver's emotional state may influence the adolescent's disease burden.

Next, mean differences in caregiver variables were analysed according to the adolescent's emotional state. Again, when adolescents presented higher levels of anxious symptomatology, their caregivers had higher mean scores on anxiety and depression. Therefore, preliminary evidence suggests a certain degree of emotional contagion between adolescents and caregivers.

Comparison of means based on disease characteristics

Means were compared based on the presence of pseudomonas and no differences were found in any of the variables. In the case of adolescents with CF, we compared means for pancreatic insufficiency and pulmonary insufficiency and found no differences either. The only difference found was for the presence of bronchiectasis. In the case of the presence of bronchiectasis, statistically significant differences were found in total QoL ($t= -1.99$, $p=.034$), with lower QoL scores in patients with bronchiectasis. In the same line, higher levels of anxiety were found in adolescents with bronchiectasis ($t= 2.36$, $p=.011$).

Table 1. Chi-square tests as a function of diagnosis for anxious-depressive symptomatology in the dyad.

		Emotional adjustment in dyad					
		Anxiety in adolescent					
		Absence of symptomatology % (n)	Presence of symptomatology % (n)	Possible psychological diagnosis % (n)	χ^2	P	
Diagnosis	Cystic fibrosis	88.90 (24)	11 (3)	-	7.93	.02	
	Primary ciliary dyskinesia	58.30 (7)	16.70 (2)	25 (3)			
			Depression in adolescent				
			Absence of symptomatology % (n)	Presence of symptomatology % (n)	Possible psychological diagnosis % (n)		
	Cystic fibrosis	100 (27)	-	-	4.74	.03	
	Primary ciliary dyskinesia	83.30 (10)	-	2 (16.70)			
			Anxiety in caregiver				
			Absence of symptomatology % (n)	Presence of symptomatology % (n)	Possible psychological diagnosis % (n)		
	Cystic fibrosis	47.80 (11)	39.10 (9)	13(3)	0.50	.78	
	Primary ciliary dyskinesia	54.50 (6)	27.30 (3)	18.20 (2)			
			Depression in caregiver				
			Absence of symptomatology % (n)	Presence of symptomatology % (n)	Possible psychological diagnosis % (n)		
Cystic fibrosis	87 (20)	8.70 (2)	4.30 (1)	1.80	.41		
Primary ciliary dyskinesia	72.70 (8)	9.10 (1)	18.20 (2)				

Table 2. One-factor ANOVA to compare dyad means as a function of the categories of emotional adjustment of both dyads.

		Emotional adjustment in dyad			F	η²	
		Absence of symptomatology M (SD)	Presence of symptomatology M (SD)	Possible psychological diagnosis M (SD)			
Variables of adolescent	Anxiety in caregivers						
	Threat of disease		3.29(3.38)	3.42 (2.19)	6.60 (6.07)	4.03*	0.21
		Depression in caregivers					
	Anxiety		2.82 (2.16)	7(6.08)	10 (5.29)	10.46***	0.40
		Fatigue	5.96 (0.75)	5 (1.64)	4.08 (1.18)	7.47**	0.33
		Emotional functioning	5.88 (0.73)	5.17 (1.45)	4.28 (1.51)	5.17*	0.25
Quality of life		6.11 (0.66)	5.74 (1.12)	4.85 (1.25)	2.68*	0.22	
Variables of caregiver	Anxiety in adolescent						
	Anxiety		7.41 (3.25)	7.50 (3.54)	14.67 (6.66)	5.62**	0.27
		Depression	3.86 (2.94)	4.50 (0.71)	14.67 (6.66)	8.66***	0.36

*p ≤ .05; **p ≤ .01; ***p ≤ .001.

Correlations

Variable correlations indicated that the adolescent’s age was positively associated with the perceived threat of the disease. Correlations were made with variables associated with the disease (time since diagnosis, hospital admissions, daily dose, aerosol, nebulized and tablet doses, and spirometry values), only the significant ones are shown in Table 3. The number of hospital admissions was positively associated with patient and caregiver anxious symptomatology and negatively associated with the adolescent’s QoL. In addition, caregivers reported higher stress scores. Disease threat was positively associated with aerosol doses and negatively associated with spirometry values. Quality of life was positively associated with spirometry values.

In terms of emotional symptomatology, adolescent anxiety was positively associated with caregiver anxiety and depression and negatively associated with adolescent QoL. Adolescent

depression was positively associated with caregiver depression. Finally, the caregiver’s emotional state was positively associated with perceived stress. The outcomes for these correlations can be found in Table 3.

Explanatory models (QCA)

First, the main descriptors and calibration values of the study variables are presented (Table 4).

Analysis of necessity

From the results obtained, the analysis of necessity indicated that there was no necessary condition for anxious-depressive symptomatology in the dyad since the consistency values were below .90 (Claude & Christopher, 2014; Ragin, 2008)

Table 3. Correlations between dyad psychological variables, dyad age, and medical indicators.

	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15
Age adolescent	1														
Age caregiver	.44**	1													
Hospital admissions	.12	-.13	1												
Daily dose	-.22	-.11	.10	1											
Anxiety adolescent	.16	.03	.40*	-.31	1										
Depression adolescent	-.05	-.04	.24	-.09	.63***	1									
Threat of disease	.33*	-.29	.26	.02	.22	.04	1								
Quality of life	-.20	-.02	-.59***	-.14	-.53***	-.28	-.39*	1							
Anxiety caregiver	.28	-.31	.42*	-.14	.45**	.19	.50**	-.35*	1						
Depression caregiver	.17	-.21	.29	-.23	.62***	.41*	.30	-.41*	.79***	1					
Stress frequency	.01	-.28	.36*	.12	-.09	-.06	.38*	-.04	.37**	.14	1				
Effort stress	.15	.02	.34	-.11	.19	-.01	.12	-.35	.55**	.53**	.36*	1			
Aerosol Doses	-.17	-.18	.04	.30	.10	.09	.32*	-.18	-.05	-.08	.34	-.03	1		
FVC/FEV1	-.53***	.03	-.32	-.30	-.14	-.19	-.36*	.42*	-.22	-.01	-.26	.04	-.30	1	
Diagnosis time	-.38*	.27	.09	.43*	-.25	-.08	.18	-.09	.21	.11	-.02	.26	-.15	.05	1

*p ≤ .05; **p ≤ .01; ***p ≤ .001

Table 4.
Main descriptions and calibration values

	Anxiety patient	Depression patient	Threat of disease	Quality of life	Anxiety caregiver	Depression caregiver	Stress frequency	Effort stress
<i>M</i>	46.11	4.74	11701.77	1332.35	787.29	115.44	7828311.13	1433289.94
<i>SD</i>	115.01	11.58	18930.72	599.52	1866.31	224.08	24782586.53	5761282.06
Min.	1	1	1	130.63	8	1	432	8
Max.	576	72	78408	2343.83	9216	972	156250000	36000000
<i>Calibration values</i>								
P10	2	1	12	542.67	16	2	4860	384
P50	6	2	2268	1298.60	144	24	2160000	23040
P90	72	12	49280	2115.17	1944	288	115200000	1433289.94

Note. *M*: mean; *SD*: standard deviation; min: minimum; max: maximum; P10=:10th percentile; =P50=50th percentile; P90=:90th percentile.

Analysis of sufficiency

As for the analysis of sufficiency, the combination of conditions that led to emotional adjustment was calculated (Tables 5 and 6). Based on the premise that in the fsQCA, a model is informative when consistency is around or above .74 (Eng & Woodside, 2012), all the proposed models were consistent as their values were all higher.

Emotional adjustment in adolescents

The main combination for predicting high levels of anxious symptomatology in adolescents, explaining 47% of cases (Consistency=.89; Coverage=.47), was the interaction between high caregiver emotional distress, high effort stress scores and low levels of quality of life. In the second and third pathways, as shown in table 4, a diagnosis of CF was found to be a variable that also explained high levels in combination with other variables. The main predictor for low levels of anxious symptomatology was the combination of a CF diagnosis, low caregiver emotional distress scores and low levels of effort stress, explaining 35% (Consistency=.76; Coverage=.35).

The main combination for predicting high levels of depressive symptomatology in the adolescent, explaining 51% of cases (Consistency=.84; Coverage=.51), is the same pathway found for high levels of anxiety. Additionally, the main predictor for low depressive symptomatology was the combination of a CF diagnosis, low scores on caregiver emotional distress and low

levels of effort stress, explaining 64% of cases (Consistency=.83; Coverage=.64).

Emotional adjustment in caregivers

The main combination for predicting high levels of caregiver anxious symptomatology, explaining 45% of cases (Consistency=.86; Coverage=.45), was the interaction between a diagnosis of CF in their children, high effort stress scores and low levels of quality of life. The main pathway for predicting low levels of anxious symptomatology was the combination of a CF diagnosis and lower stress frequency levels, explaining 54% (Consistency=.83; Coverage=.54).

The main combination for predicting high levels of depressive symptomatology in caregivers, explaining 41% of cases (Consistency=.88; Coverage=.41), was the interaction between their children being diagnosed with CF, high stress frequency scores and high levels of threat of disease. Additionally, the main pathway predicting low levels of depressive symptomatology was the combination of low scores on adolescent emotional distress, low levels of effort stress, high quality of life and low disease threat explaining 43% of cases (Consistency=.98; Coverage=.43).

Discussion

The present study aimed to examine exploratory predictors of emotional adjustment in adolescents with rare respiratory diseases such as CF and PCD and their caregivers.

Table 5.
Sufficiency analysis for emotional adjustment in adolescents

Frequency cutt-off 1	High levels of anxiety in adolescents			Low levels of anxiety in adolescents			High levels of depression in adolescents			Low levels of depression in adolescents		
	Frequency cut-off .76			Frequency cut-off .83			Frequency cut-off .79			Frequency cut-off .89		
	1	2	3	1	2	3	1	2	3	1	2	3
Diagnosis		o	o	o	o	o		o	•		o	o
Emotional discomfort caregiver	•		•	o			•		•	o	o	
Stress frequency		•	•		o			•			o	o
Stress effort	•		•	o			•	•		o		
Quality of life	o	o				•	o	•	o			•
Threat of disease			o	o	•			o			o	o
Raw coverage	.47	.35	.24	.46	.45	.28	.51	.30	.23	.64	.39	.33
Unique coverage	.05	.09	.05	.08	.05	.04	.05	.12	.06	.14	.01	.01
Consistency	.89	.76	.81	.79	.86	.94	.84	.87	.81	.83	.96	.91
Total coverage			.67			.82			.77			.89
Total consistency			.79			.79			.74			.81

Note. •= presence or high levels; o= absence or low levels. All paths are consistent because the consistency is above .74. Expected vector for high levels of anxiety and depression in the adolescent 1.1.1.1.0.1

Expected vector for low levels of anxiety and depression in the adolescent 0.0.0.0.1.0.

Table 6.

Sufficiency analysis for emotional adjustment in caregivers

Frequency cut-off 1	High levels of anxiety in caregivers Frequency cut-off .90			Low levels of anxiety in caregivers Frequency cut-off .90			High levels of depression in caregivers Frequency cut-off .84			Low levels of depression in caregivers Frequency cut-off .87		
	1	2	3	1	2	3	1	2	3	1	2	3
Diagnosis	○		○	○			○		○			○
Emotional adjustment adolescent		●			○			●		○	○	
Stress frequency		●		○	○	○	●	●	●		○	○
Stress effort	●	●	●		○	○		●	●	○	○	
Quality of life	○	○	○		●	●		○	○	●	●	
Threat of disease						○	●			○		●
Raw coverage	.45	.39	.38	.54	.39	.38	.41	.39	.32	.43	.40	.23
Unique coverage	.07	.08	.07	.15	.01	.02	.14	.07	.03	.02	.01	.03
Consistency	.86	.98	.87	.83	.92	.90	.85	.99	.92	.98	.95	.85
Total coverage			.75			.84			.79			.69
Total consistency			.88			.85			.88			.89

Note. ● = presence or high levels; ○ = absence or low levels. All paths are consistent because the consistency is above .74. Expected vector for high levels of anxiety and depression in the adolescent (Fiss, 2011) 1.1.1.1.0.1

Expected vector for low levels of anxiety and depression in the adolescent 0.0.0.0.1.0

Regarding the hypothesis on emotional symptomatology in caregivers and adolescents with these diseases, studies indicate high anxiety-depressive symptomatology rates in adolescents (Catastini et al., 2016; Georgiopoulos, 2020; Guta et al., 2021) EMBASE, MEDLINE, Cochrane, Scopus, Web of Science, CINAHL, and manually on Google Scholar. This meta-analysis follows the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA and caregivers (Graziano et al., 2020)). The results indicate a greater presence of anxious-depressive symptoms in the caregiver than in the adolescent. In both adolescents and caregivers, scores for anxious symptomatology were higher than those for depressive symptomatology. In the case of anxiety, the percentages ranged between 11-25% for adolescents and between 14-40% for caregivers. Therefore, this would be in line with previous research, and H1 would be accepted. However, the percentages for depression are slightly lower.

As for the second hypothesis, no differences in emotional adjustment were expected, although no previous studies have compared both diseases. However, previous research indicated that emotional impairment was associated with chronic respiratory diseases (Carotenuto et al., 2013; Catastini et al., 2016; Graziano et al., 2020). The results found indicate that there were no differences in anxious-depressive symptomatology based on diagnosis for caregivers. However, higher levels of anxiety and depression were observed in adolescents suffering from PCD. A possible explanation for this is that, while the type of treatment and routines are very similar to those for CF patients, the diagnosis of PCD is often longer and may influence the feeling of lack of control or uncertainty about the course of the disease. Therefore, the hypothesis would be partially accepted.

In H3, adolescents' emotional adjustment was expected to correlate negatively with quality of life and positively with the threat of disease. Correlation results showed that disease threat was not negatively associated with anxious-depressive symptomatology. The QCA models' findings do not show this variable's impact on the adolescent's emotional adjustment. Therefore, despite being a relevant variable, further studies should be conducted to determine its role in coping with the illness.

Quality of life was negatively associated with anxious symptomatology but not with depression. However, in QCA models,

it was observed that one of the predictor variables of high and low levels of anxious-depressive symptomatology was the adolescent's quality of life. This would be in line with findings from other studies (Graziano et al., 2020; Tickner et al., 2022). Quality of life was negatively associated with anxious symptomatology but not with depression.

However, in QCA models, it was observed that one of the predictor variables of high and low levels of anxious-depressive symptomatology was the adolescent's quality of life. This would be in line with findings from other studies (Graziano et al., 2020; Tickner et al., 2022). Therefore, H3 could also be partially accepted.

The results found in both the correlations and the QCA models are in line with the initial hypothesis. Positive associations were found between stress and anxious-depressive symptomatology. On the other hand, when the caregiver experienced stress-related discomfort or tension, it was significantly associated with poorer emotional adjustment. Stress frequency was only associated with anxiety but not depression. Again, in the QCA models, it is confirmed that caregiver stress is positively associated with emotional adjustment, but caregiver effort-related stress may be a more relevant variable. Therefore, H4 would be accepted, and the proposed approach would be in line with previous observations (Casaña-Granell et al., 2021; Pinquart, 2018; Tan et al., 2018), which found that caregivers with these characteristics often report moderate stress levels and may also negatively influence their children's adjustment to the disease.

Finally, regarding H5, there was an influence between the emotional state of the adolescent and that of the caregiver. Differences were observed in the adolescent's perceived threat of the disease based on parental anxiety. Adolescents may perceive their illness as more threatening if their parents show higher anxiety levels. On the other hand, mean differences were observed for parental depression, finding lower scores in quality of life and higher anxiety scores among adolescents within those groups of caregivers who could have a psychological disorder. Moreover, correlations found a relationship between adolescent and caregiver variables, with caregiver anxiety being positively associated with adolescent anxiety and perceived disease threat and negatively associated with adolescent quality of life. Similarly, for depressive symptomatology, caregiver depression was positively associated

with adolescent emotional symptomatology. These results are in line with previous studies (Continisio et al., 2020; Wood et al., 2015; Yuksekgonul et al., 2020) we aimed to determine the psychosocial status of primary care givers of children with cystic fibrosis (CF), which indicate that the caregiver's emotional state may influence how the adolescent copes with the disease and its progression.

In the QCA models, the adolescent's anxious-depressive symptomatology was associated with the caregiver's stress levels and mainly with the caregiver's stress levels in combination with other variables. For caregivers, their emotional adjustment was influenced by their own variables but also by their child's quality of life, as well as by their perceived threat and, to a lesser extent, by their emotional adjustment. Therefore, given the above, the hypothesis could be accepted.

This study is one of the first attempts to perform an exploratory comparative study of both rare respiratory diseases in the adolescent population, particularly in Europe. Few similar studies have been conducted before, and even fewer in Spain. Despite its contributions, the study is not without its limitations. The first limitation is that the information was gathered in a specific area of Spain, so it would be necessary to increase the sample to generalise the results. Furthermore, although the study is exploratory, it has a cross-sectional design, so it would be interesting to assess whether there are changes over time and the evolution of the disease through longitudinal studies and strive to increase the sample size to enhance the generalizability of the results. It would also be interesting to include other disease-related variables such as duration of hospital admissions. All these aspects will be considered in future research.

In conclusion, the results found are relevant because they provide a better understanding of the psychosocial aspects associated with diseases such as CF and PCD. They also allow observing the relationship between caregivers and adolescents. These studies help health professionals explore other aspects of the disease in greater depth and could improve the decision-making process between the patient and the professional.

Relevance for clinical practice

The study provides clinically relevant results by exploring the psychosocial aspects associated with diseases such as cystic fibrosis (CF) and primary ciliary dyskinesia (PCD). These findings offer a better understanding of the relationship between caregivers and affected adolescents, enabling healthcare professionals to address the emotional and social needs of patients more comprehensively. The research also emphasizes the importance of exploring additional aspects of the disease and suggests improvements in the shared decision-making process between patients and healthcare professionals. In summary, the findings have practical applications, including the enhancement of medical care, the design of psychosocial interventions, the training of healthcare professionals, and the facilitation of patient-centered decision-making, thereby contributing to a more holistic and personalized approach to healthcare.

Data Statement

The data that support the findings of this study are available from the corresponding author, [Author 3], upon reasonable request.

Conflict of interest

The authors have no conflict of interest.

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