

# “I know I have it, but I don’t have it like I used to”: A qualitative exploration of the experiences of adolescents with cystic fibrosis recently commenced on elexacaftor/tezacaftor/ivacaftor (Kaftrio)

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## Introduction

Psychological concomitants of cystic fibrosis (CF) include psychosocial adjustment difficulties; depression and anxiety (parents & children); negative impact of hospitalisation, treatment burden on quality of life and identity confusion related to improvements in health secondary to new treatments (Havermans & Duff, 2020).

Kaftrio has the potential to transform CF from a life-limiting illness to a chronic condition (Coverstone & Ferkol, 2021).

There is now a query about how children and adolescents’ are adjusting to the prospect of a better quality of life and improved prognosis.

## Aim

- To explore the expectations and experiences of adolescents on the RECOVER trial to provide important insights and enhance the quantitative findings from the main RECOVER study, exploring the ‘why’ and the ‘how’ of the impact of this new treatment.

## Methods

17 Participants Aged 12-17 years were recruited following 1 year of Kaftrio treatment. Exclusion criteria included those whose participation in the current study may cause adverse effects to their wellbeing.

Semi-structured interviews were developed asking; general experience of having CF; what their experience has been since starting Kaftrio (including how it has affected their wellbeing and treatment adherence) whether it has changed how they see their future. The data were transcribed and analysed using the principles and six phases of reflexive thematic analysis (Braun & Clarke, 2021)

Participants completed The Patient Health Questionnaire (PHQ-9; Johnson et al., 2002) and the Generalised Anxiety Disorder (GAD-7; Mossman et al., 2017) as well as three unstandardized scales, with scores from 0 to 10 assessing the perceived importance of carrying out the Kaftrio treatment and all usual treatments, as well as their confidence in managing their current treatment regime.

## Interview Themes and Sample Quotes

### ‘Kaftrio changed things for me’

“Then I just got better and better, no hospital. And also I remember I went for an Xray a while ago and your man couldn’t believe I have CF ‘cause my lungs were completely clear....so there’s no kind of mucus or anything and he was so shocked he actually googled “CF lung” to show me a picture of what average CF lungs look like. In my one they were completely clear after Kaftrio, so that was just brilliant.” [P10]

“It’s the small things I notice. When I cycle home from school you have to cycle up this hill...last summer...I would have to walk up the rest of the way, but now I can do it no problem.” [P5]

“I’d be disappointed [if Kaftrio didn’t work] but I wouldn’t be overly disappointed ‘cause I set myself up to expect that.” [P3]

### ‘How that’s changed how I view myself and my CF’

“But when I was first starting it I was kind of scared because I wasn’t scared of what it would do but I was scared CF is all I’ve ever known. it’s a massive part of my personality and as people grow up they figure out at a young age not who they are, but like what their life is like and I did that with CF and I feel like I’m gonna have to have that realisation later like I am gonna have to re-realise who I am. [P16]

“In a way I don’t know what it’s like to have it anymore, but I know I have it, but I don’t have it like I used to...” [P3]

“I don’t want to see myself as any different...I have it fairly well boxed off in my head that you know taking Kaftrio...it’s in a neat corner and I have to go into that corner in the morning and in the evening. And then I come out and just, live my life.” [P2]

“Another thing I realised with Creon ...whenever I do [miss] it, it doesn’t affect me as much as it would before, like before I’d be in a lot of pain...the only thing about the Creon NOT hurting as much when I don’t take it, it kind of makes me feel at the start it was like I was kinda not being as strict with myself and it was no, you still need to take them. but like before I was almost getting scared into taking them.” [P16]

### ‘Reframing how I manage my CF’

“It’s amazing I kinda feel invincible and then sometimes I may slack on doing this treatment or something and my mam would remind me, “I know you feel amazing but you still have to keep doing everything you’re doing” [P9]

“I still kind of resist the sinus rinse...I don’t do the physio anymore...any day I do any kind of training I won’t have to do it [the physio], but I do training every day, so it’s kind of ...I just don’t do it anymore.” [P10]

## Results

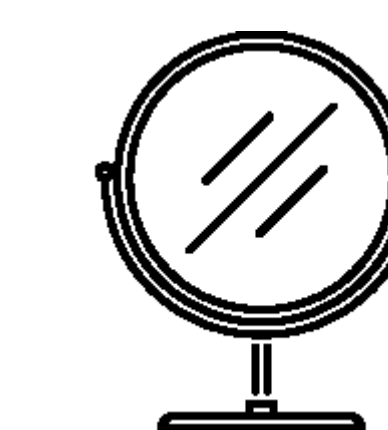
Variable	N	%	M	SD	Min	Max
Gender	Male	9				
	Female	8				
Age			13.8	1.01	13	17
PHQ-A Total			4.4	3.25	0	10
Normative population (Comparison)			2.3	3.3		
GAD-7 Total			3.53	3.07	0	11
Normative population (Comparison)			2.76	3.49		
Importance of Kaftrio Regime (0=not important, 10=extremely important)			9.33	1.39	5	10
Importance of all other treatments (0= not important, 10=extremely important)			8.73	2.28	3	10
Confidence in Managing current routine (0=not confident, 10=extremely confident)			8.47	1.3	5	10
BMI pre-Kaftrio			19.63	2.68	15.58	24.19
BMI Kaftrio 12 months			21.12	3.13	15.66	26.37
FEV1% predicted pre-Kaftrio			87.99	11.5	56.8	104.72
FEV1% predicted Kaftrio 12 months			94.29	21.93	23.37	120.82

Table 1; Participant Demographics and Clinical Data

## Discussion



‘Kaftrio Changed Things for Me’ captures the physical and symptomatic improvements Reflecting on perceived changes to their own and others’ expectations for Kaftrio; some were underwhelmed by the actual results while others had started with reserved expectations and were surprised by the improvement.



‘How That’s Changed How I View Myself and my CF’ reflects shifting views of identification with CF and how they cope, in light of a changing prognosis due to the effectiveness of Kaftrio. Participants coped using compartmentalisation and downward social comparison, conceptualised CF as something they do (i.e. treatments) rather than something they have. If they didn’t do the treatment, would their CF be gone? Participants reported a changing relationship to their illness, while acknowledging it part of their identity.



‘Reframing how I manage my CF’ reflects a shift in attitudes and behaviours to CF following health improvements related to Kaftrio. Participants decrease routine treatments, particularly physiotherapy and nebulisers, They also described more autonomy and feeling their family was less worried about them.

## Conclusion

Adolescents who have commenced on Kaftrio report improved quality of life and a reduction of the impact of CF. As they feel better they report reducing or altering treatments which negatively impacts on treatment adherence. Clinical implications include support for treatment adherence and psychosocial interventions to support identity change and lifespan development.

# Adherence Summary at 1 year for People aged >12

## 1 MPR



Baseline MPR of hypertonic saline, azithromycin, salbutamol and dornase alpha was low to moderate. Further reduction at 12-months [hypertonic saline ( $p=0.0002$ ) and dornase alpha ( $p=0.0015$ )].

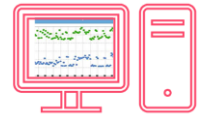
Modulator adherence (dual) was good at baseline with further improvement at 12-months (ETI) ( $p=0.0009$ ).

## 2 TAQ



No. of good adherers reduced from baseline to 12 months across airway clearance ( $p<0.001$ ), dornase alpha, hypertonic saline

## 3 MEMS<sup>®</sup>



ETI adherence: **82.7%**  
IVA adherence: **83.1%**  
Avg. adherence: **82.9%**

Self report considerably overestimated actual adherence