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Thriving alongside CF: Developing a Grounded Theory of Empowerment in Children and Young People with Cystic Fibrosis During Key Life Transitions

Short title: A Grounded Theory of Empowerment in CF

Naomi H. Fairweather, DClinPsychol [1,2]

Fergal W. Jones, PhD, PsychD [1,2 *]

Sally A. Harris, PhD, ClinPsyD [3]

Martha Deiros Collado, ClinPsyD [4]

Amy Shayle, ClinPsyD [4]

1. *Salomons Institute for Applied Psychology, Canterbury Christ Church University, 1 Meadow Road, Tunbridge Wells, Kent, UK*
2. *Sussex Partnership NHS Foundation Trust, Sussex, UK*
3. *Royal Alexandra Children's Hospital, Brighton, UK*
4. *Addenbrooke's Hospital, Cambridge, UK*

*Corresponding author:

fergal.jones@canterbury.ac.uk

ORCID: 0000-0001-9459-6631

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Abstract

Background

Although the importance of patient empowerment is increasingly recognised, little is known about empowerment in children and young people (CYP) with long-term conditions. Empowerment may be particularly important in CYP with cystic fibrosis (CF) due to high treatment burden and limited opportunities for peer support.

Methods

A Grounded Theory method was employed to develop a preliminary theory of empowerment in CYP with CF. Seven CYP with CF, five parents and four professionals were interviewed.

Results and conclusions

The emerging model suggests that 'thriving alongside CF' may be supported by interactions between 'having a team' and 'taking charge and having a voice', leading to 'being able to just be a child', that 'concealing self' may get in the way of 'thriving alongside CF' and that these processes occur within wider medical and developmental contexts. Study limitations, clinical and research implications are discussed.

Key words: Cystic Fibrosis, Empowerment, Adolescents, Children

Introduction

Cystic fibrosis (CF) is a multisystemic condition primarily affecting the lungs and digestive tract (Cystic Fibrosis Trust, 2020a). Approximately one in every 2,500 babies in the UK is born with CF (Cystic Fibrosis Trust, 2020a). Fifty years ago it was unlikely for a child with CF to live to their 10th birthday (Coulthard, 2018). With universal screening, earlier diagnosis, new understandings and treatments, life expectancy has increased dramatically, with over half of people with CF living beyond 47 years (Cystic Fibrosis Trust, 2020a).

CF research intensified following discovery of the CF transmembrane conductance regulator (CFTR) gene in 1989 (De Boeck, 2020; Kerem et al., 1990). There are over 2000 mutations of CFTR. For patients with common mutations, precision medications known as CFTR modulators have recently been developed to tackle the underlying causes of CF (De Boeck, 2020). In the UK, Orkambi and Symkevi were made available through the NHS at the end of 2019 (Cystic Fibrosis Trust, 2020b). Kaftrio received its European licence in 2020 (Cystic Fibrosis Trust, 2020b). For those with rarer CFTR mutations, treatments are still in development (Cystic Fibrosis Trust, 2020b; De Boeck, 2020).

Increased life expectancy opens up both possibilities and problems for children and young people (CYP) with CF. Empowering CYP with CF is important due to the significant treatment burden, rapid changes in life expectancy and barriers to face-to-face peer support associated with cross-infection risks (Cystic Fibrosis Trust, 2020a).

Patient empowerment

Most models of patient empowerment, draw on community psychology principles (Bravo et al., 2015; Rappaport, 1984; Zimmerman, 1995), much of the existing literature

focuses on adults (Zimmerman, 1995). Empowerment is thought to encompass self-efficacy, knowledge and skills, perceived control, a sense of meaning, and feeling respected have been identified as indicators of empowerment in adults with long-term conditions (Bravo et al., 2015). Empowered adult patients have been found to participate in shared decision-making, self-manage, and utilise support groups (Bravo et al., 2015) and there is some evidence of better health outcomes (Yeh, Wu, & Tung, 2018).

There is growing interest in adolescent patient empowerment. The Gothenburg Young Persons Empowerment Scale (GYPES) has recently been developed for CYP with long-term conditions (Acuña Mora et al., 2018). GYPES is made up of five domains: knowledge and understanding, personal control, identity, decision-making and enabling others. However, the GYPES is based on adult patient empowerment models (Acuña Mora et al., 2018) and it is unclear how relevant the scale is to CYP with CF. Moreover, patient empowerment in CYP is complex due to developmental changes, and the impact of systemic influences including parents and peers (Kirk et al., 2013).

Developmental perspectives

Little is known about the influences on and impact of empowerment in CYP with CF during adolescence, a period associated with life transitions including school changes, increased independence, developing relationships, identity formation and future planning (Erikson, 1959). Prior to availability of precision medicines, for adolescents with CF this period of rapid developmental change often coincided with worsening CF symptoms, e.g. cough and fatigue, increasingly frequent pulmonary exacerbations and associated increases in treatment burden (Hegarty, MacDonald, Watter, & Wilson, 2009). At a time when CYP would otherwise be increasing independence and spending time with friends, for those unable to benefit from new precision medications, deteriorating health could lead to isolation, more

dependence on family, needing to stay home or be hospitalised (Cystic Fibrosis Trust, 2020b; De Boeck, 2020; Rowbotham, Palser, Smith, & Smyth, 2019).

Peer support features in many existing models of patient empowerment (Bravo et al., 2015; Jørgensen et al., 2018; Small, Bower, Chew-Graham, Whalley, & Protheroe, 2013). Historically, CYP with CF benefited from peer support through residential social events. This reduced isolation, protected against stigmatisation, and increased quality of life through sharing of adaptive coping strategies. In the mid-1990s, the dangers of cross-infection were recognised, resulting in strict infection control policies, including guidance that patients should never meet in person (Rowbotham et al., 2019). Whilst CYP with CF can seek peer support online (Kirk & Milnes, 2016), and there is some evidence that online support can serve as a tool of empowerment (Gavrila, Garrity, Hirschfeld, Edwards, & Lee, 2019; Kirk & Milnes, 2016), studies of other patient groups suggest that high quality face-to-face interactions are more likely to promote psychological wellbeing (Subrahmanyam, Frison, & Michikyan, 2020).

Therefore, research and theory regarding empowerment cannot be assumed to generalise from adults or children with other long-term conditions to CYP with CF. To the authors' knowledge, there are no existing studies of empowerment in CYP with CF. Hence, this study aimed to develop a grounded theory of perceived facilitators of, barriers to, and consequences of empowerment in CYP with CF during key life transitions.

Methodology and Methods

Design

A Critical Realist Grounded Theory Methodology (Anonymised for peer review, 2020; Birks & Mills, 2015; Corbin & Strauss, 2015; Oliver, 2012) was selected as little is known about empowerment in CYP with CF, the phenomena of interest involves a process, and developing new theory with explanatory power was a desired outcome. For further details, please see the online supplementary material.

Recruitment and Participants

Participants were continuously recruited from two NHS paediatric CF services in England until no new concepts arose during analysis of successive interviews. Data collection and analysis was conducted concurrently, later participants were recruited according to the likelihood that they would be able to answer questions raised by earlier interviews and fill gaps in the emerging theory (theoretical sampling). Twenty-two participants were approached, 16 of whom participated (5 CYP and 2 parents declined). Key adolescent life transitions represented included: starting secondary school, looking ahead to college, increased time with peers, taking on more responsibility for managing CF, considering career options, and thinking about moving on to adult CF services. Demographic and health information are shown in Tables 1-3.

Interviews

Semi-structured interviews were conducted by the first author in clinic or via telephone. Topics included: times when CYP feel most confident, accessing information, handling change, managing CF treatment, hopes for the future and messages for people

wanting to help CYP feel empowered. Interviews ranged between 17-56 minutes and were audio-transcribed.

Patient and public involvement and Ethical considerations

Feedback from the Cystic Fibrosis Trust and a hospital-based youth advisory panel was incorporated into the project. Approval was granted by an NHS research ethics committee. Children under 16 whose parent/guardian had given consent were asked for their assent. Over 16s consented for themselves. CF specific infection control policies were complied with.

Data analysis

Data were analysed using grounded theory as described by Corbin and Strauss (Corbin & Strauss, 2015), supported by NVivo 12 software. This method utilises three stages of coding: open, axial and selective. This process is outlined in the supplementary material along with a detailed coding table. Data were initially analysed by the first author and the analysis was reviewed and developed in consultation with the other authors. Quality of analysis was supported through use of triangulation, research diaries, bracketing interviews, active listening, adhering closely to the data and making constant comparisons within and between transcripts.

Results

Due to space constraint, some subcategories will be described more fully than others. The focus will be on categories addressing the research questions most directly, aspects not explained in previous literature, and those that are most relevant for clinical practice, future research and policy.

Overview of the model

The model presented in Figure 1 illustrates the phenomenon **'thriving alongside CF'**. Categories and subcategories are highlighted in bold. The quotations included exemplify the interpretation of the derived categories.

The core category within the data was **'thriving alongside CF'**. According to the grounded theory five higher level categories influenced **'thriving alongside CF': having a team, taking charge and having a voice, being able to just be a child, concealing self** and **the wider context.** Two higher level categories supported **'thriving alongside CF'**: **having a team**, and **taking charge and having a voice**. These categories interlinked with each other and enable **being able to just be a child. Concealing self** appeared to obstruct **'thriving alongside CF'** because having to conceal oneself limits **having a team** and **being able to just be a child**. **'Thriving alongside CF'** was influenced by **wider contextual factors**, occurring in the context of constant developmental changes, changes in the condition and treatments at the individual level, and constant developments in biomedical understandings and CF treatment.

Core category: Thriving alongside CF

The core category linking all the data together **'thriving alongside CF'** refers to CYP being able to live without their diagnosis defining them or preventing them from having influence over their life.

> *'they're happy… they live their life, and CF is a part of it, but CF isn't their whole life'* (Professional)

Professionals noted that there is a group of CYP with CF who aren't **'thriving alongside CF'**. It is unclear, what proportion (if any) of these CYP would be found in the

42% who declined to be interviewed. These voices remain unreported and are referred to as ***'undocumented voices'***.

Having a team

All participants referred to the importance of CYP with CF having a support network. CYP, parents and professionals all used 'team language' for example *'we're very much a team'* giving rise to the in vivo code 'having a team'.

Team membership

Family, friends, medical teams, and school were key members of the team for many CYP. For some, a connection with God and a faith community were important, for others, sports clubs or pets. Whilst team size and membership varied, all participants made reference to the importance of at least one significant confiding relationship.

> *'what we've found helps most successfully to manage that transition is ensuring that they have somebody to talk to about it'* (Professional)

Trusting relationships

All participants referred to the importance of trusting relationships. CYP and their families valued enduring friendships and consistency of medical team members.

> *'it's a good feeling when there's someone there who's been with you since you were born'* (YP 17, M)

Having a team of trusted people facilitated open and honest conversations. Whilst having friends in general was beneficial having **'at least one good friend'** seemed particularly important and links to the subcategory of **'explaining to others'.**

> *'Get a really close friend… one person you can really trust and tell them'* (YP 15, F)

CYP varied in how open they were about CF. Some chose to tell one or two carefully selected friends or teachers on a strictly need-to-know basis.

> *'…we've got some children who… have… a couple of best friends who know… they don't want it blazoned across… for all in sundry to hear.'* (Professional)

Others spoke proudly of the confidence they had gained through standing up in front of large groups and raising awareness of CF, for example in school assemblies.

> *'Once it was off my chest I felt so much better… they know what I'm going through and why I'm missing school'* (YP, 13 F)

Having **'at least one good friend'** protects against **'concealing self'.**

> *'we definitely see a difference in the children who… haven't told their friends at school and have to take their Creon in secret and they find that it's… always stressful'* (Professional)

Sharing the burden of CF

CF was described by CYP and parents as too much for any one person to manage.

> *'… it's not just down to us parents, it's not just down to [child], it's a team effort, work with the CF team as well, you've got their support, their backing, their help….'* (Parent)

The CYP's team has several roles which may be fulfilled in different ways by different members at different times. Several participants described how having a team helped share the burden of CF through practical and emotional support.

'If I'm down… the dumps, my family and my friends always help me out. And my little pet lizard always keeps me calm' (YP, 12, M)

Several CYP explained that although they could complete all aspects of their CF regime, doing all the CF-related tasks completely independently was not their priority. Sharing the burden of CF with help from family, friends and the medical team to complete tasks provided CYP with the time and energy to do other things that were important to them.

'I can do my IVs myself, but I choose not to… My mum will do them if I'm at home. But if I'm in hospital, the nurses will.' (YP, 18, F)

For some CYP the CF community formed an important part of their team

'It's nice to know that people have the same thing and can relate to… what we're going through' (YP, 14 M)

Others expressed a yearning for connection with other CYP with CF.

'I know absolutely no-one whatsoever… I would love to know someone my age and talk to them… – not necessarily about CF – but talk to someone daily, how you would talk to anyone else that's in the same position as you. Because you can have really supportive people around you, really amazing people, but they're not in the same position as you' (YP, 14 F)

Providing a sense of acceptance/'normalising'

One of the team's roles was providing a sense of acceptance. Many CYP spoke of the importance of feeling 'normal'. Feeling accepted at home, with friends and at school was experienced as 'normalising' by many CYP.

'I'd class my friends as normal, when I'm out with them not worrying about my health… I'm more classed as normal, and that's quite nice.' (YP 18, F)

Making room for mistakes and learning from experience

Another theme that emerged, primarily from interviews with parents and professionals, was the team's role in allowing mistakes and letting CYP learn from experience. This was the case both in relation to lessons CYP need to learn for themselves in managing CF, as well as generic learning experiences which would have an added layer of complication for those with CF.

'I've certainly had a couple of chats about alcohol with our older teenagers… They're teenagers, they're going to experiment…. I try really hard to show that I'm not judging them and I'm not going to tell them off.' (Professional)

Professionals reflected on how taking on the role of coach facilitated CYP taking charge.

'*team members to act like coaches, and to allow the children… to feel like they can ask really simple questions.... be honest about their compliance with treatments… be honest about when they're really struggling.'* (Professional)

Many adult participants described how allowing CYP to learn from experience both required and built trusting relationships.

'Whatever's going on, we can support you and if you want to make a change or get back on track… we're here to support you' (Professional)

Several parents and the three oldest CYP with CF reflected on the role their families and medical team had in enabling learning from experience.

> *'he knows if he doesn't take his medication he will get poorly… cos he's tried it once before, he's hidden his tablets when he was younger'* (Parent)

> '*stop badgering...Remind me a few times, but it's on me if I don't do it'* (YP, 17 M)

Taking charge and having a voice

'Having a team' supports **'taking charge & having a voice.'** All participants referred to taking charge and developing a voice, both directly in relation to the medical context and more broadly. This interacted with developmental changes and evolved over time. Taking charge and 'having a voice' appeared to both require and develop information and understanding.

> *'[I said] "I don't know how to explain it to my friends, can you help me?" Then [doctors] helped'* (YP, 13, F)

Professionals spoke of a wide range in levels of understanding about CF amongst CYP.

> '*there's a huge range of … level of detail they know about their condition'* (Professional)

Professionals and parents noted the role of parents in passing on information about CF to CYP.

> *'Some of our parents are very open with their children about the condition and complications... Others are very secretive… they don't want to worry their child …'* (Professional)

All professionals described strategies developed to encourage CYP to take ownership of their understanding of CF/CF treatments – for example developing tailored information sheets and encouraging CYP to ask questions. Professionals in particular reflected on how the informational needs of CYP change over time. Some CYP made references to the added value of getting information from others with lived experience as opposed to solely from professionals and information sheets.

> *'someone who has the experience… instead of… people who have either known people with CF or have just found things out from reading.'* (CYP 14, M)

Approximately half of the CYP interviewed were connected to other CYP with CF through internet support groups which acted as a source of information and social interaction. For those who did not have contact with others with CF, this was experienced as a loss with CYP feeling they were missing out on an important source of information and connection.

> *'I'm not allowed near any other person with it… it's a bit concerning, and upsetting… I can't talk to people who understand what I'm going through'* (CYP 12, M)

A range of views were expressed by parents and professionals about information from the internet, including from support groups. Some had their own positive experiences of connecting with others in similar circumstances; others feared support groups could hold frightening or inaccurate information, or encourage non-compliance.

'she's quite well informed but I don't think she knows everything, all the different paths that the future could lead to … she's still very young, in terms of having to sort out if some people are rebelling against regimes… I'm just wondering if that's unhelpful as well as helpful.' (Parent)

Having information and understanding seemed to enable taking charge and having a voice which in turn seemed to enable active participation in decision-making.

'I think the first step is understanding what it is… and from there, you can make choices' (CYP 15, F)

Many participants stressed the importance of having information so that CYP could make decisions based on individual preferences and circumstances. This included both treatment-related decisions such as medication regime timings and broader life decisions such as career choices.

'Taking control of CF and being able to have a voice in the management and direction…being able to manage it alongside all the rest of their life' (Professional)

Taking charge and 'having a voice' appeared to enable 'finding ways round & through'/practical problem solving. All the CYP and parents gave examples of finding ways of doing things that are important to them despite the significant challenges that CF and treatment regimens can present. For many, having a routine facilitated this. Others described emotional coping strategies and creative solutions to practical problems.

'We've done things like administering IV antibiotics in the car, before she went to a party' (Parent)

Taking charge and 'having a voice' enabled young people to explain to others X,Y,Z and this helped counteract other's assumptions.

'I basically said, "If I cough, it's not contagious." And as the weeks went on, they found out a little bit more' (YP 18, F)

Taking charge and 'having a voice' also enabled 'being able to just be a child'.

Being able to just be a child/getting on with life

'Being able to just be a child' was connected to 'thriving alongside CF'. This was something that many of the CYP and their families strived for.

'our attitude is, that if you've done all your treatments then we can do anything else… forget about it, and crack on with normal stuff' (Parent)

This required 'acceptance of difference'.

'If I never had CF, I would never be me' (YP, 13 F)

Acceptance of difference appeared to enable 'achieving in relation to own goals' and 'having adventures and fun' both of which were described as examples of 'being able to just be a child'. For some CYP, 'achieving in relation to own goals' meant spending time with friends, others accomplished this through sport or academic achievements, or through raising awareness about CF.

'to be able to do all the things they want to do in life… relationship building or …a particular career pathway' (Professional)

Concealing self

'Other people's assumptions' and/or 'difficulties fitting it all in' can put CYP in the position of having to make difficult choices and may lead to having to keep secrets or concealing aspects of themselves. 'Other people's assumptions' described by participants

included misunderstandings about what CF is (e.g. contagious), low expectations (e.g. in relation to sport), and judgements (e.g. teachers and peers making comments about CYP with CF eating 'unhealthy' foods, when in fact, a high fat / high calorie diet is medically necessary).

All participants referred to the 'difficulties fitting it all in', and many CYP described arriving home exhausted and making choices between completing homework or their full CF regimen.

'it is hard to fit homework in as well as medicines' (YP 13, F)

Some referred to difficulties having honest conversations with the medical team about struggling to fit everything in. This seemed to relate to fear of disappointing professionals whom they valued as part of their team.

'She hates so many of her treatments… I think she finds it hard to talk honestly about that without offending them' (Parent)

All professionals and some parents reflected on CYP 'having to keep secrets.' This was described as stressful for the CYP and appeared to get in the way of 'thriving alongside CF'

'*they feel embarrassed about some of the things they have to do, and so they don't tell people… just increases the stress'* (Professional)

'Concealing self' appeared to get in the way of developing 'trusting relationships' and seemed to limit 'having a team' as CYP may be unable to test whether they will be accepted.

'*she felt quite embarrassed about that* [side effect of medication] *and so we were thinking about ways you could open up and own that… without feeling mortified.'* (Parent)

The accounts of professionals and some parents suggested that 'having to keep secrets' may limit 'active participation in decision-making'

> *'She will just nod and say yes and be polite. I don't know how much she really feels able to really tell them* [CF team] *truthfully'* (Parent)

Whilst some CYP reflected on times they felt the need to conceal aspects of themselves and keep secrets, they spoke of this as something they had done in the past. A sense of the CYP currently feeling they have to conceal aspects of themselves (as opposed to those making an active choice to keep some information private) came through the professionals and parents of CYP who opted not to participate. This gave rise to the category 'undocumented voices.'

> *'It's a bit of a shame… it is the young people that do quite like to talk that got involved* [in the research] *… it would be nice to hear* [those that declined] *and for them to say, "Actually, I'm just completely pissed off with having CF and I don't want to talk about it"'* (Professional)

Wider context

Thriving alongside CF was described by participants as occurring in the wider context of all CYP with CF being unique individuals, with unique constellations of experiences, hopes, fears, challenges, preferences and goals.

> *'It's one of those things that people have their own… definition of…what is empowering to one person won't be to the other'* (YP 18, F)

There also seemed to be interactions with developmental stages and changes. CYP growing up with CF are constantly changing in terms of levels of cognitive, emotional and physical

maturity. All participants spoke of the additional struggles faced by CYP with CF in navigating the typical challenges of adolescence. Professionals in particular, highlighted the impact of uncertainty and the decline in health that older adolescents may face just when peers without CF will be in their physical prime.

> *'Puberty, changing peer groups, family dynamics, all those [things that]… children without CF, have to deal with…changes in health that they may or may not suffer with'* (Professional)

In addition to the constant changes in the condition and treatments at the individual level, 'thriving alongside CF' also occurs in the context of constant developments in biomedical understandings and treatments for CF. Although this appeared important in maintaining a sense of optimism for the future for some participants, many CYP spoke of acceptance and living more in the moment. Some spoke of difficulties in keeping up with medical developments

> *'It's a bit confusing… you're used to the old stuff, then they give you new stuff, I kind of get a bit worried that it's gonna be hurting or… it's not gonna work because it's brand new, and they've not tested it properly yet'* (YP, M, 12)

Discussion

This study developed a grounded theory of empowerment in CYP with CF with a core category of **'thriving alongside CF'**. The model is consistent with descriptions of empowerment in the community psychology literature (Rappaport, 1987; Zimmerman, 1995). Both Rappaport's (1984) empowerment theory and the 'thriving alongside CF' model

describe a process by which people gain mastery over their lives. Rappaport asked the question 'under what conditions do we find people reporting a sense of control over their lives?' The current model provides some answers to this question in relation to CYP with CF. Similarly to Rappaport, the current model suggested that there are multiple possible solutions to challenges, that different solutions suit different individuals and that professional input may open up more possibilities.

The model is also consistent with Zimmerman's (1995) conceptualisation of psychological empowerment, in that 'thriving alongside CF' was found to take different forms in different CYP and different contexts (for example at home, at school and in clinic), and fluctuates over time.

There is overlap between the current model and models of patient empowerment based on research with adult patients (e.g. Bravo et al., 2015; Small et al., 2013) and reflected in the newly developed GYPES measure (Acuña Mora et al., 2018). This is particularly the case in relation to having information and decision-making. There are also important differences. Some of these relate to developmental stages – for example feeling or being perceived to be 'different' and confronting other people's assumptions are likely to be more of a challenge for CYP than for adults, and children are more likely to be dependent on adults for reliable information (Ernst, Johnson, & Stark, 2011). Other differences were specific to CF. For example the impact of infection control guidelines and the limited opportunities for CYP to connect with others with CF (Vines, Fisher, Conniff, & Young, 2018) meant the role of peer support was more complex than is often reflected in models of empowerment based on other patient groups.

The model is consistent with previous research involving CYP with CF. For example, Barker, Driscoll, Modi, Light, and Quittner (2012) described the importance of family and

friends in successfully managing CF during adolescence. Parents in the Sawicki, Heller, Demars, and Robinson (2015) study made reference to allowing adolescents to make and learn from mistakes in promoting adherence. Adolescents and their parents in Dashiff, Suzuki-Crumly, Kracke, Britton, and Moreland (2013) acknowledged the importance of understanding medical aspects of CF when making decisions, and highlighted a need to make information more accessible to children and families. Children participating in D'Auria, Christian, and Richardson (1997) spoke of the process of deciding if and who to share CF information with. However, in contrast to the current study, participants in previous studies (e.g. D'Auria et al. 1997) generally agreed that it was better to keep CF a secret. One explanation for this difference is that participants in the current study were older, indeed some reflected on being afraid to tell peers when they were younger.

There are numerous references in the literature to limited time and the burden of CF and CF treatment within the context of short lives (e.g. Barker et al., 2012). Whilst this clearly featured in the current study, there was a sense of more optimism and expectation of surviving and thriving into adulthood. In contrast to previous studies focusing on adherence in CF (e.g. Sawicki et al., 2015) the current study suggested CYP taking full responsibility for CF care may be neither achievable nor desirable. Participants in the current study consistently indicated that CYP feel more empowered when the full responsibility of CF care is shared with their team rather than dealt with alone. Increasing longevity associated with recent medical advances is affording many more CYP with CF opportunities not previously considered possible. All participants made references to hopes for the future, including hopes for careers, getting married and having families – all of which would have been less likely until relatively recently and therefore perhaps not accounted for in previous literature.

Given the prominence of patient peer support in previous models of patient empowerment, it is particularly important to highlight the impact of infection control guidelines limiting physical contact between patients with CF. These guidelines are still relatively recent and whilst other research on the impact of isolation is beginning to emerge (Vines et al., 2018) the current study adds an additional perspective. In addition, to the authors' knowledge, the impacts of developmental stages and changes, the progressive nature of CF and the particularly rapid nature of current advances in CF care have not been accounted for in previous models of patient empowerment. Increasing longevity associated with recent medical advances is affording many more CYP living with CF new opportunities in life. All participants made references to hopes for the futures of the CYP with CF. This included hopes for careers, getting married and having families of their own – all of which would have been less likely until relatively recently and are therefore perhaps not accounted for in much of the previous literature. As new treatments such as modulator therapies become increasingly available and the longer-term impact of these is known, it will be important to revisit the influence on empowerment in CYP with CF.

Limitations

Although each category identified in the data was deemed to reach theoretical sufficiency, the theoretical model's validity could be enhanced with a larger sample and respondent validation (Mays & Pope, 2000). Furthermore, while participants were recruited from two NHS sites and attempts were made to recruit via CF charities and social media, there remained a lack of diversity. All participants were white British, to an extent this is consistent with the genetic predisposition of CF (Williams & Barker, 2010). Due to complexities of measuring socio-economic status in CYP, this was not recorded, though socio-economic status is likely to impact CYP's experiences of empowerment.

The nature of the research is likely to have meant that CYP who are not currently 'thriving alongside CF' would have been less likely to have time, energy, confidence or inclination to participate. This group was alluded to by several participating professionals and captured in the model under the category of 'undocumented voices'. This is an important group to consider in future research.

Implications

Research to establish the acceptability, reliability and validity of the GYPES (Acuña Mora et al., 2018) for CYP with CF is warranted. CYP who are not currently 'thriving alongside CF', whose voices are rarely heard in clinic and whom professionals struggle to support provide a clear focus for future qualitative research. Creativity will be key in considering how best to access this population, for example use of peer researchers as in (Gathercole, 2019) or anonymous questionnaires may mitigate power differentials between researcher and participants.

Given the importance ascribed by participants to 'having a team' it may be valuable for medical professionals to routinely ask CYP who is in their support network and how CYP would like them to be involved. This could include questions about whether CYP have access to support from other CYP with CF. Access to peer support could be facilitated through development of internet-based group interventions.

The current study adds weight to recommendations made by Sawicki et al. (2015) for example developing communication tools for use by CYP and professionals that incorporate a mutual understanding of competing priorities and time pressures, and developing problem-solving skills. This could involve parents and professionals (including teachers), allowing choices, noting initiative and encouraging opinions and problem-solving. Clinicians may find it helpful to continuously review CYP's understanding of CF and to develop a stock of

accessible resources. Service evaluations could help understand the extent to which CYP feel they have opportunity for ‘taking charge’/ ‘having a voice’.

Data availability statement

We do not have participants' consent or ethical approval to share the raw data.

Key messages

- Empowerment related experiences in children and young people with cystic fibrosis differ from those of adults with various other long-term conditions.
- Empowerment appears to be facilitated by a support network, access to information and involvement in decision making.
- Having to conceal aspects of self appears to be a barrier to empowerment.
- Empowered patients appear more able to ‘thrive alongside CF’
- Further research is needed to explore perspectives of children and young people with CF whose voices are seldom heard and who may not be ‘thriving alongside CF’.

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Table 1

Young person demographic and health information

Age	Range: 12-18 Mean: 14.7
Gender	Male: 3 Female: 4
Ethnicity	White British: 7
Age at CF diagnosis	Newborn: 4 ≤ 2 years: 2 ? ≤ 5 years: 2
FEV-1[a]	Range: 79 – 122% Mean: 100.5%
CF related comorbidities & procedures	CF related diabetes; intestinal obstruction; gastrostomy; ACE procedure; ileostomy; lobectomy; microcytic anaemia
Number of episodes of IVs in previous year	Range: 0 – 4 Mean: 1.5
Number medications	Range: 5 – 15 Mean: 11

[a] FEV_1 (forced expiratory volume in 1 second) is a measure of CF disease severity. An FEV_1 of 100% means that lung function is as would be expected for a person of the same age and height who doesn't have CF. $FEV_1 \geq 85\%$ is considered normal; 70-84% indicates mild lung disease, 41-69% indicates moderate lung disease; ≤ 40% suggests severe lung disease (24).

Table 2

Parent/carer demographic information

Relationship to child with CF	Father: 1 Mother: 4
Gender	Male: 1 Female: 4
Ethnicity	White British: 5
Age of children	Range: 11-18 Mean: 13.8

Table 3

Professional demographic information

Profession	Dietician Physiotherapist Specialist nurse Pharmacist
Years of experience	1-5 years: 1 6-10 years: 1 11+ years: 2

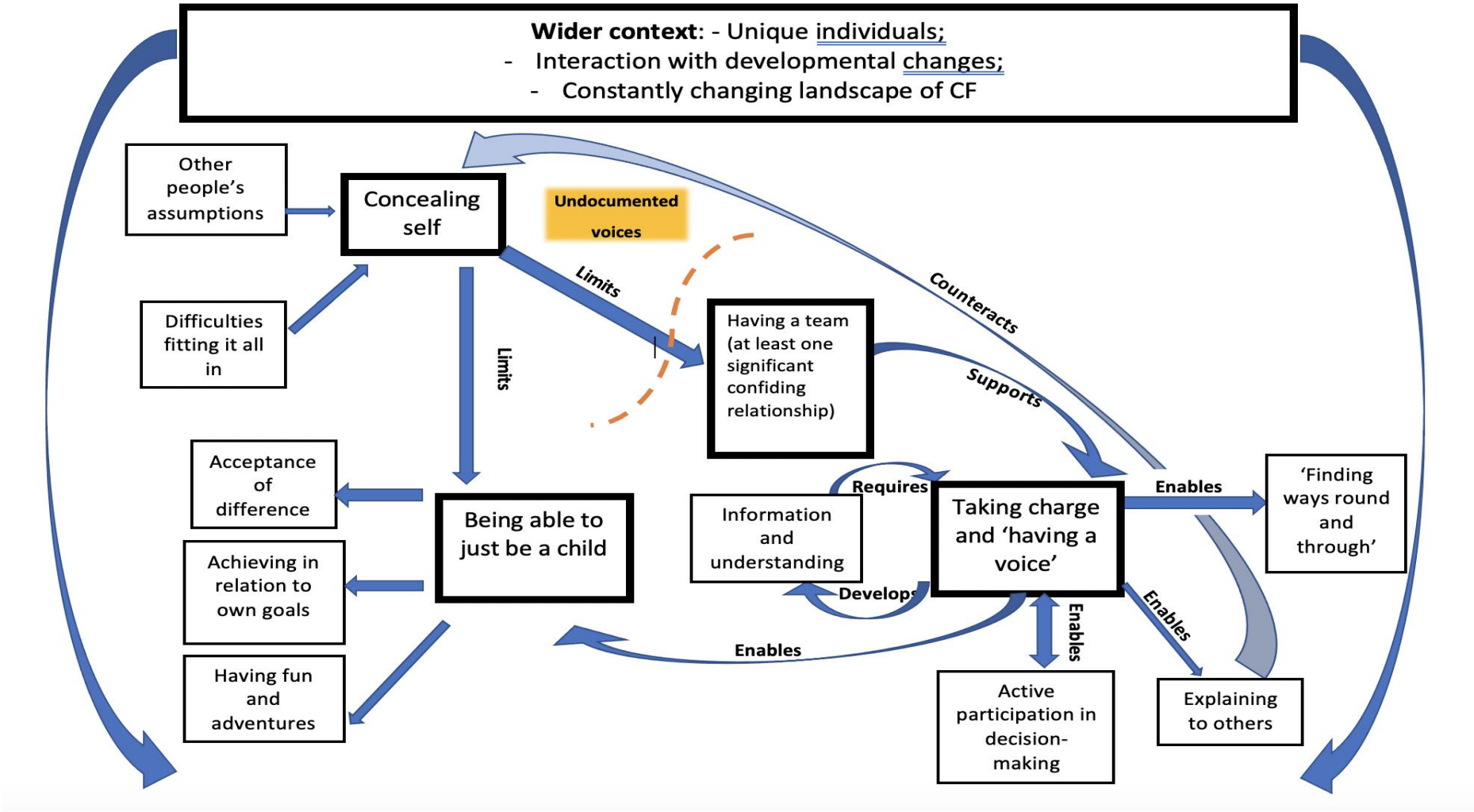

Figure 1 A diagrammatic representation of the grounded theory of thriving alongside CF.

Online supplementary material

Description of the analytic process	
Initial 'open' coding	Audio files were checked against verbatim transcripts and coded line-by-line enabling the researcher to become thoroughly immersed in the data. This initial coding was used to break the data down and allow segments to be compared with other segments, between and across transcripts (Corbin & Strauss, 2015). During open coding the researcher stuck closely to the data and was particularly careful to reflect on their own assumptions when making early analytical decisions (Corbin & Strauss, 2015).
Concurrent data collection and analysis	Data was analysed after each interview. In each case the researcher asked 'what is the data saying and where are the gaps?' (Corbin & Strauss, 2015). On the basis of this, interview questions were refined and 'theoretical sampling' was used to identify which other potential participants to interview.
Theoretical sampling	Additional participants were recruited according to likelihood that they would be able to answer some of the questions raised by other interviews (Birks & Mills, 2015).
Intermediate 'axial' coding	Axial coding was used to reassemble the data that was fractured during open coding at a more conceptually abstract level (Corbin & Strauss, 2015). First categories were developed by linking subcategories and considering their properties and dimensions. Categories were then linked together (Birks & Mills, 2015). The main question here was what are the relationships between categories (who, what, when, and how) (Corbin & Strauss, 2015).
Selective coding and theoretical integration	Central phenomena identified and systematically related to other categories (Corbin & Strauss, 2015). Storyline technique used as a mechanism for theoretical integration (Corbin & Strauss, 2015).
Theoretical sensitivity	Idea that all researchers have their own way of seeing the world, influenced by their own experiences and things they've read.

	As data analysis progresses, they will become more sensitive to emerging concepts that resonate with their own previous experience
Memo writing and diagrams	Diagrams and memos were used to note emerging ideas and track theory development, acting as a bridge between data collection and theoretical analysis. Memos are used throughout the process to record initial reflections, and as analysis continued links between increasingly abstract concepts (Birks & Mills, 2015).
Constant comparison	Throughout the process data was constantly compared within and between all three levels of coding and across memos.

For references, please see the main paper.

As hundreds of open codes were produced, the following table contains examples of from each category rather than an exhaustive list.

Coding table

Selective code	**Axial codes**	**Open codes**
Having a team	Team membership	• Ref to having a team • YP ref to role of family • Parent ref to role of family • Prof ref to role of family • Ref to at least one good friend • YP ref to role of friends • Parent ref to role of friends • Prof ref to role of friends • Ref to role of medical team • Ref to role of school • Ref to CF community
	Provides a sense of acceptance/ 'normalising'	• Parent ref to belonging • Prof ref to belonging • YP ref to being accepted • YP ref to fitting in
	Share burden of CF	• Ref to sharing responsibility • Ref to practical support • Ref to emotional support • Ref to multiple supportive roles

	Makes room for mistakes/ learning from experience	• Prof describing role as coaching • Parent talking about YP learning from experience • Prof reflecting on role of mistakes
	Trusting relationships	• YP reflecting on trusting relationships • Parent ref to trusting relationships • Prof ref to trusting relationships
Taking charge and 'having a voice'	Information and understanding	• YP ref to interacting with information about CF • Parent ref to information and understanding • Prof ref to information and understanding • Ref to technology
	Active participation in decision-making	• YP reflecting on making decisions • Parent reflecting on making decisions • YP reflecting on having a voice • Parent highlighting importance of YP having a voice • Parent ref to YP confidence • Prof reflecting on making decisions
	Explaining to others	• YP reflecting on explaining to others • YP reflecting on sharing personal information • YP ref to raising awareness • Parent reflecting on YP explaining to others • Prof ref to YP explaining to others
	'Finding ways round and through'/ practical problem solving	• YP reflecting on asking for help • YP describing coping strategies • YP describing ways around limitations • Parent describing YP's coping strategies • Parent ref to YP facing challenges • YP making the most of things • YP ref to routine • YP talking about importance of hope • Ref to preparing for future

Being able to just be a child	Acceptance of difference	• Ref to dealing with difference • YP expressing acceptance • YP reflecting on developing confidence • YP sharing stories of survival • YP showing wisdom • YP talking about who they are
	Achieving in relation to own goals	• YP describing achieving • YP showing determination • YP sharing hopes for the future • YP ref to non-CF challenges • Parent hopes for their child's future • Prof ref to YP goals • Prof ref to YP being determined • Parent ref to YP helping others • Prof hopes for the future of YP they support • Ref to priorities
	Having adventures and fun	• YP ref to enjoyment • YP describing having adventures • Prof ref to YP having adventures • Parent ref to child's life being more than CF
Concealing self	Other people's assumptions	• Reflecting on other people's assumptions • Parent ref to responses of other's • Ref to lack of flexibility
	Time/difficulties fitting it all in	• YP ref to time • YP describing the challenge of CF • Prof ref to logistical challenges
	Having to keep secrets/concealing self	• Struggling with being different • Having to keep secrets
	Undocumented voices	• Undocumented voices
Wider context	Unique individuals	• YP explaining it's all about the individual • Ref to individual differences

	Interaction with developmental stuff: changing emotional, physical, cognitive (maturation)	• Ref change over time – responsibility • Ref to change – getting older • Prof reflection on interaction with developmental norms
	Constantly changing landscape of CF	• YP reflecting on change over time in own CF • Prof reflecting on change over time in disease • Ref to changing landscape of CF