

# Adult cystic fibrosis patients' experiences of primary care consultations: a qualitative study

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## ABSTRACT

### Background

'Expert patient' programmes have been introduced in the UK as a new approach to chronic disease management for the 21st century. The average survival age of those with cystic fibrosis (CF) has steadily increased such that the majority of those with the condition now live into adulthood. Currently, specialist CF centres deliver the core of medical care, with primary care providing access to prescribed medicines, referral to other services, and care of non-CF needs, however, it is necessary to provide a more comprehensive service for adult CF patients, involving both specialist centres and primary care. To date, little is known about these expert patients' experiences of primary care.

### Aim

To investigate how young adults with CF perceive and experience primary healthcare services.

### Design of study

Qualitative study.

### Setting

One specialist CF centre in southeast England.

### Method

Interview study of 31 patients with CF, aged 18 years or over.

### Results

Adults with CF consult in primary care on two distinct levels: as lay and expert patients. When consulting as experts, patients tend to operate as consumers of health care and perceive a satisfactory doctor-patient relationship to be influenced by three factors: GPs' understanding of how people live with CF, GPs' ability to prescribe certain specialist medications, and sensitive management of the cost of health care for adults with CF. A doctor-patient relationship based on trust and understanding is seen as desirable, but requires that these factors are addressed both by the GP and the patient.

### Conclusion

Expert patient policy has focused on the role of patients with common chronic conditions in secondary and tertiary care, with little consideration of how adults with rare chronic illness and their GPs manage health problems that can be addressed in primary care. Enabling easy access to holistic care, as well as establishing successful trusting relationships with people with long-term rare conditions, is a necessary foundation for expert patients to take an active role in their care.

### Keywords

cystic fibrosis; doctor-patient relations; expert patient; primary health care; qualitative research.

## INTRODUCTION

The increasing prevalence of chronic illness has led to a heightened emphasis on the key role of primary care and also on the development of a range of self-care initiatives for those with long-term conditions. In the UK, the government's expert patient policy<sup>1</sup> was introduced to encourage and enable patients with chronic diseases to take an active role in their own care within the remit of chronic disease management. This shift towards greater patient involvement in healthcare decisions has provoked concerns, particularly surrounding the potentially detrimental effect on the doctor-patient relationship.<sup>2</sup> Indeed, it has been suggested that the provision of 'expert' patients, trained to help others with similar illnesses, might even serve to reduce access to a health professional.<sup>3</sup>

Although the government's expert patient policy is still in the formative stages of development, for many years, patients with relatively rare long-term diseases, who receive the majority of their treatment and care from a secondary or tertiary service, have taken on the expert role when consulting in primary care. Knowledge of these patients' experiences of primary care, therefore, can help inform a more successful 'expert patient' approach to health care. In this article, we examine the experiences of adults with cystic fibrosis (CF) when consulting in primary care, and the impact that their expertise in their illness has on the doctor-patient relationship.

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Although CF is the UK's most common autosomal recessive genetic disease, its estimated prevalence is currently only 7750.<sup>4</sup> However, the average survival age has been steadily rising over the past three decades such that patients are now predicted to live, on average, for 40 years;<sup>5</sup> it is estimated that over 20 000 people will be living with CF and treated by a secondary or tertiary service in the UK when survival into middle adulthood becomes the norm.<sup>6</sup> As more patients survive to adulthood and experience age-related conditions associated with CF, particularly CF-related diabetes,<sup>7</sup> they are likely to rely more and more on primary care. Their increasing age also means that they will not only take over responsibility for their treatment and care from their parents but, having been encouraged throughout their lives to be active participants in their own care, they are also likely to consult within primary care as patients with specialised expertise.

To date, clinical guidelines for CF treatment recommend that patients be seen at specialist centres<sup>8</sup> as these have been shown to result in a better clinical outcome for patients.<sup>9</sup> Much of the routine treatment, however, including intravenous antibiotics, takes place in the home with family members assisting the patient.<sup>10</sup> With the increasing longevity of patients with CF, it is now important to integrate primary and secondary care services to ensure seamless care delivery.

Although the role of the primary care team in antenatal genetic CF screening, for example, has been well documented,<sup>11</sup> GPs have expressed concerns about the provision of care for patients with CF and, in particular, the current arrangements for their prescribing of specialist medicines of which they may have little clinical experience.<sup>12</sup> So far, the part that GPs play in the continuing care of adult CF patients has received little attention. Specifically, adult patients' views on the relationship that they have with their GP and their experiences of being what could be considered an 'expert patient' within primary care, have not previously been reported.

This paper reports on adult patients' experiences of primary care. The findings reported here are part of a wider study that used mixed methods to explore the health and quality of life of adults with cystic fibrosis,<sup>13</sup> and the perceived risks and benefits of other current and future potential treatments and care,<sup>14</sup> including relationships with healthcare professionals providing these in primary and secondary care.

## METHOD

The sample was drawn from patients living in southeast England and attending a specialist CF centre. A letter inviting participation was sent to 198 patients aged 18 years or over, with reminder letters

## How this fits in

In the UK, the government's expert patient policy has been introduced to enable patients with chronic disease to take an active role in their management. This study offers evidence that those with rare long-term conditions act on two distinct levels in primary care: either as lay or expert patients. When consulting as experts, patients act as consumers of health care and perceive a satisfactory doctor-patient relationship in primary care to be based on the GP's understanding of how patients live with a long-term condition, ability to prescribe specialist medicines, and sensitive management of healthcare costs.

sent to non-responders after 4 weeks. There was an overall acceptance rate of 26% ( $n = 51$ ). Thirty-one patients (14 male, 17 female) completed in-depth interviews, with the remainder completing questionnaires, subsequently withdrawing consent, or becoming too ill to participate.

The interviewer was a medical sociologist and, although attached to the specialist centre for research purposes, was not involved directly in any patient's care. Interviews were conducted in patients' own homes using an interview guide drawn from CF and sociological literature as a prompt for discussion. A summary of the topic guide is shown in Box 1. Interviews, lasted between 1 and 1.5 hours, on average.

During their interviews participants were specifically asked to describe their experiences and relationship with their current and any previous GPs. Where interviewees had not already raised the issue earlier in the interview without prompting from the interviewer, participants were asked a general question such as 'What is your GP like?', with follow-on opening questions about the participant's perception of their GP's knowledge and their use of primary care, phrased for example as, 'Do you feel your GP has a good knowledge of CF?', and, 'Do you go to your GP often?' Probing questions were asked to explore responders' initial answers. All interviews were audiotape recorded and transcribed verbatim. Confidentiality was assured between the patient and their healthcare providers.

### Box 1. Summary of topic guide used in interviews.

- ▶ Perceived quality of life (quantitative and qualitative account)
- ▶ Family support (including living arrangements and lay carers)
- ▶ Medical history (including diagnosis of CF and CF-related disease, symptoms, and treatments)
- ▶ Contact with healthcare services (including specialist care, local hospital care, and primary care)
- ▶ Education and employment history (including disclosure of disease)
- ▶ Sources of information obtained (including CF Trust, NHS, others with CF)

### Analysis

Data analysis has been described in depth elsewhere.<sup>13</sup> Following transcription, interview material was imported into ATLAS-ti, a software programme for the analysis of qualitative data. Codes were assigned to a word, phrase, paragraph, or sentence to assist with the detection of categories, themes, and patterns.<sup>15</sup> Data relating to each of the codes were then retrieved and analysed for content, focusing particularly on links and variances within the dataset. The coding frame was developed and the subsequent analysis of the coded data was discussed by the authors who agreed the analytic framework and assessed the explanatory value of the categories against the retrieved data. The majority of conceptual categories had started to be developed during the course of the interviews in the style of theoretical conceptualisation, whereby patterns of action and interaction between and among various types of social units could be worked out.<sup>16</sup>

### RESULTS

Of the 31 patients who were interviewed, the median age of the 14 men was 33 years (interquartile range [IQR] = 26–37 years) and the median age of the 17 women was 30 years (IQR = 22.5–33.5 years). Participants represented a range of personal experiences of living with CF; some perceived themselves to be in relatively good health while others were aware that their health had severely deteriorated over the past few years. All patients attended an annual review at the specialist centre; outpatient's appointments ranged from 6-weekly to 3-monthly. The range of health states experienced by patients is shown in Table 1.

When asked at interview to describe their experiences with their current and any previous GPs, participants suggested they had two distinct roles:

- as 'lay' patients, consulting for non CF-related problems in which they had little experience or expertise; and
- as 'expert' patients, seeking practical help for CF-related problems.

When operating as a 'lay' patient, participants perceived the GP role as consultative, where they sought advice and guidance from the GP, acknowledging their role as the medical 'expert', yet simultaneously playing down their GP's role in the care for their condition:

*'He's a nice doctor, but I don't really deal with him on the CF side. I deal with him more on the, "oh, I've got a broken ankle", or something like that.'* (F7, 18 years)

*'I only go to my GP for other things apart from CF. [Later] He's okay for a sprained ankle or something in your eye or something like that.'* (M4, 40 years)

*'Mainly I deal with my specialist when it comes to CF, but anything else, like I've got the flu or a cold or something, I go and see the GP and he's fine.'* (F14, 18 years)

As the interviews continued, however, it was evident that despite their initial claims, many of these patients did operate as an expert patient in CF-related consultations with their GP, if only to gain access to treatment and care. This suggested that a claim of non-contact for CF-related problems may have been made initially to emphasise their own expertise in CF. The GP, however, did not always appear to recognise that the patient was operating as a lay patient, seeking help for problems unrelated to their CF. This resulted in practical treatments being offered for what patients believed the GP saw as CF-related problems:

*'I remember for the appendix [appendicitis] she gave me cramp-reducing tablets or something, and said that I was to eat a lot, and that actually nearly forced it to burst. I think she just put it all down to CF.'* (F4, 21 years)

When consulting as an expert patient (for CF-related problems), participants appeared to perceive the GP role as being limited to providing practical help. Primary care was valued as a useful means of getting prompt access to repeat prescriptions and minor procedures, such as venepuncture and re-siting of in-dwelling cannulae for intravenous antibiotics. All participants had had some contact with their general practice in the month preceding the

**Table 1. Health status of patients at time of interviews.**

Health status	Male (n = 14)	Female (n = 17)	All
Pancreatic insufficiency (taking enzymes)	12	14	26
History of intravenous antibiotics	12	16	28
CF-related diabetes	6	5	11
CF-related arthritis	1	-	1
Post lung/heart-lung transplant	2	2	4
Post living related donor lung transplant	-	1	1
Post liver transplant	-	1	1
Awaiting transplantation (on transplant list)	1	1	2
Infected with <i>Burkholderia cepacia</i>	-	1	1
With children	1	5	6
Undergoing/failed in vitro fertilisation	2	2	4

interview, illustrating that they were regular users of the service. Many participants, however, tended to describe GPs in terms of the functional role that they played. As can be seen from the examples below, the doctor–patient relationship in this context appeared to be characterised by a consumer approach to health care, with patients operating as the experts in their condition and viewing the GP as little more than a gatekeeper for treatments:

*'Oh, brilliant, he's great, I mean I go up and tell him what I want, and he gives it to me.'* (M1, 26 years)

*'The GP for me is there to dispense the drugs and sick-notes.'* (F1, 33 years)

*'I just write him a list, tell him what I want and he writes it for me.'* (F2, 30 years)

*'I just phone up and order loads of drugs from him. I'm always phoning up. I hardly ever see him.'* (M2, 21 years)

*'GPs are only good for repeat prescriptions really [laughs]. Not much use beyond that.'* (F8, 23 years)

One identified area of practical assistance from the GP was in performing venepuncture and re-inserting in-dwelling cannulae so that patients would not have to attend their local hospital. However, participants did not consider whether GPs should take on roles that may be outside their remit or whether they have the time to perform such tasks. Of key importance was the convenience of being able to receive this prompt treatment from the GP:

*'The first time I ever had them [intravenous antibiotics] the line went down about 5 days in, and the GP just put another venflon in for me.'* (F3, 27 years)

*'He takes my blood so I don't have to go up to the local hospital ... [he] will put venflons in and things like that.'* (F4, 26 years)

Indeed, seeing themselves as expert judges of their health care, some participants felt able to determine whether the GP was competent to perform the requested task:

*'My local GP has had to do so many [venflons] now for me that he's actually getting quite good at it.'* (F5, 38 years)

Although GPs were clearly seen by participants to play an important role in access to treatment, the

dynamics of the doctor–patient relationship were influenced by participants' use of the primary care services for meeting purely functional needs. Although primary care aspires to provide a personalised continuity of care that takes into account the patient's physical, psychological, and social needs, its ability to do this appears to be limited when patients not only consult as experts, but also as consumers focused solely on the practical aspects of care. In choosing to be recipients of a 'functional' rather than holistic primary care service, patients effectively prevented their GPs from developing a more general and fuller understanding of their needs. In considering this problem, patients highlighted three key factors that they perceived to influence the doctor–patient relationship, all attributed to attitudes of GPs, rather than those of adults with CF. These factors are discussed below.

#### ***The perception that GPs needed to understand how people lived with CF***

Many of the participants felt that the GP lacked understanding about their experiences of living with CF and the different ways in which it affected their lives. Acknowledging that people with CF needed regular medical services performed locally, rather than at the specialist centre, that they also suffer from 'normal' health problems in which they had little or no expertise, and that, through fertility treatment, many were now able to become parents, was seen as being particularly important when patients consulted the GP for health concerns:

*'When I had to have a line out ... he said no ... It was at Christmas time and I wasn't well and I had the hump and I thought ... that means I've got to go all the way into [specialist centre], which is a real trek for me, if you're not feeling 100% anyway. Didn't give me any reasons. I wonder if he understood fully what I'd asked him. I don't think he did.'* (F8, 30 years)

*'He said, "Oh no, you mustn't have a baby" he said, "You don't want to waste 31 years of your life, you've got one child already".'* (F9, 31 years)

Patients with CF spoke quite clearly about the importance that they attached to having a good relationship with the GP. Some viewed their GP as becoming more knowledgeable about the challenges of living with CF and perceived the GP's efforts to increase their knowledge about CF as enhancing the doctor–patient relationship. Encompassing this was not only knowledge of the disease process itself, but also understanding that adults with CF are people trying to lead a full, active life:

*'It's incredibly important to have a good relationship with the doctor, where you can actually sit and you think that they're listening to you.'* (F8, 30 years)

*'It's not just the knowledge of cystic fibrosis, the disease itself, but it's a knowledge of the people ... he knows that I've lived with my disease for 26 years.'* (M1, 26 years)

*'Whenever I see her [GP] now she has a very intelligent discussion with me concerning my health, she asks me [about] my lung functions and to her credit I think she's investigated it at the moment with me and my family.'* (M6, 26 years)

*'[My GP is] Brilliant. He's really good. I would say he's, as far as listening and trying to help you, 100%.'* (M9, 35 years)

Although all participants stated that specific detailed information about CF would be sought from the specialist centre or the CF Trust, in operating as an expert patient within primary care, they also valued the GP's input into their holistic care.

#### **Willingness to prescribe certain doses and types of specialist medications**

Although participants acknowledged that the GP did not see many patients with CF and was, therefore, unlikely to have comprehensive knowledge about the condition. They generally felt that the GP was not adequately informed about CF and, as such, they would be unable to fully meet their primary care needs. Patients perceived this as affecting the type and amount of drugs that the GP prescribed, especially the antibiotics needed to treat chest infections. This often appeared to create dissension between the patient and the GP, with patients reporting that they had to resort to asking doctors at the specialist hospital to act as an advocate for them by writing directly to the GP to request that they prescribe certain drugs:

*'He obviously understands nothing about CF at all 'cause he just wouldn't give me any more antibiotics and the only way I get antibiotics is if the [specialist centre] phone him up and say, "she must go on these antibiotics".'* (F9, 31 years)

Participants did not consider that their GP's hesitance to prescribe some medications, for example Pulmozyme® (Genentech), may be due to clinical inexperience. Instead, some patients interpreted unwillingness on the part of the GP to prescribe

treatment as reflecting their disinterest in providing care and, at times, a useful way of referring them back to the specialist hospital:

*'I went [to the GP] this morning 'cause I keep getting these temperatures and I've had a sore throat and my voice went 2 days ago, my chest has been hurting. And he just sat there and he said, "Well, I don't know". And I thought "great, what am I supposed to do? I've got a temperature of 102 and what am I supposed to do?" And he just said to me, "Carry on taking the Disprin® and if it gets any worse, come back". So it's useless.'* (F5, 25 years)

*'When I do see my own doctor he always says, "Have you phoned [the specialist centre]?" Or, "Are you going to go up there?" So he just gets rid of me, really.'* (F5, 25 years)

*'My other GP [partner] sort of says to me, "You're not very well, are you? You'd better ring [the specialist centre] and book yourself in".'* (F8, 23 years)

For some patients, however, previous difficulties relating to prescribing appeared to be largely resolved. This arose either through the GP's contact with the specialist centre or by the patient becoming more involved in the prescribing decisions when the GP acknowledged that, through their experience of managing their condition, the patients had become experts in living with the disease. In turn, this seemed to result in an improved doctor-patient relationship:

*'I've got a brilliant GP and I don't even have to see him, I just ring him up and say, "Look, I don't feel well" and he said, "Well, what would you like to go on?" And I go, "Well, last time I was on erythromycin so this time I think we'll try flucloxacillin" and that's where it works.'* (F8, 23 years)

*'He says, "What do you want?" And I tell him what I want and he just does it. 'Cause you go in there and he says, "Well, you obviously know more about this than I do so if you let me know".'* (M10, 36 years)

*'He's not frightened to say that he doesn't know, he's not an expert on it, you know, and he's happy to leave us to do what we think we need to do.'* (M10, 36 years)

Over time, as patients gained confidence in taking on the expert patient role, they became included in

the hospital/GP partnership, often facilitating communication between the specialist centre and the GP:

*'[The specialist centre] really do guide me. I tend to guide my GP, like [the specialist centre] will say to me, "Oh, you're sensitive to this [antibiotic]". I had three courses of ciprofloxacin and I kept saying [to the GP] "Well, I'm sensitive to that" or, "That's working". So my GP said, "Oh, alright then".'* (F6, 31 years)

### **Sensitivity towards the cost of health care**

Due to the type, quantity, and frequency of prescribed medications, the cost of treatment for CF tends to be fairly high, particularly for recently licensed drugs such as Pulmozyme® (Genentech). Responders were very aware of this and at interview often referred to themselves as 'expensive' patients. They remained concerned that their GPs might be denying them medication because of its cost. The emphasis on cost appeared to stem, in part, from numerous previous encounters with the GP in which the cost of care was frequently raised:

*'I used to get Post-It® notes on the ordinary prescriptions, "Do you realise the cost of Colomycin® [Pharmax], do you realise the cost of Pulmicort® [Astra Zeneca]?" ... He had me in the office and said, "Do you realise you're our dearest patient? You cost this practice £20 000 a year, how long do you expect to live?".'* (M4, 40 years)

*'There was a time, I think it was about a year ago, there was a bit of confusion over the Pulmozyme prescriptions, and he wrote to me and basically said, you know, the [specialist centre] have prescribed this, so they've got to give it to you, we're not going to be able to afford to do it.'* (M7, 33 years)

A heightened awareness of the financial constraints surrounding health care was a particular feature of early consultations with the GP and appeared to be of more concern to older responders (in their 30s and 40s) and those in worse health, with a perception that care might be compromised as they lived past their predicted survival age. The concerns around difficulties obtaining routine treatments also extended to situations where patients hoped for referral to other services:

*'It was when we were applying for fertility [treatment]. He said, "Well, you can't really expect that because the NHS has been very good to you, and spent a lot of money on you, so don't expect that".'* (M8, 32 years)

*'Obviously I'm a very expensive patient, yeah, that's come up, because my parents deal with it, they would know more than I do. I think it's been brought up in the past.'* (M3, 37 years)

This knowledge that patients with CF were 'expensive' often prevented participants from changing their GP if their current one was not perceived to be providing an acceptable level of care. Patients, therefore, appeared to generally change their GP only when moving area:

*'If I tried now [to change GP] it might be a bit of a job because of the drugs being expensive and things.'* (F2, 30 years)

**Researcher:** *'Have you not tried to change your GP?'*

**M8:** *'I don't think anybody else would have me'* (32 years)

For all but one of the responders, however, many of the financial difficulties appeared to have been resolved over time as GPs applied for special allowances and responders felt that the relationship between themselves and their GPs had improved:

*But then that [confusion over prescribing Pulmozyme] was cleared up and he phoned me up and said, basically, "Anything you want you can have".'* (M7, 33 years)

*'There's not a problem with me, see, he turned round and said, "It's not a problem with you, we can apply for having expensive ...", I don't know what it is, but expensive patients, he said, "Yeah, we can apply for that, no problem".'* (M1, 26 years)

*'They're very good about paying for everything.'* (F12, 35 years)

Although the introduction of additional resources for patients who are chronically ill appears to have contributed to an improved doctor-patient relationship, the data suggest that a previously negative experience surrounding financial concerns over care may sometimes leave a lasting effect on the relationship that patients have with their GP. Practices, such as prescribing cheaper drugs, were understood to be part of primary care's usual conduct, but seen within the context of being an 'expensive' patient:

*'I was on Zantac® [GlaxoWellcome] and he changed me over to Axid® [Lilly] 'cause it's*

*cheaper. Not that it worries me, 'cause if it does the same thing, it's the same thing. But there is the phrase that, "Oh yes, it's cheaper so I'll put you on the other one". I suppose it's not penny-pinching, that's like normal balancing the books, isn't it? But coming from him with a past history of CF.'* (M8, 32 years)

Although participants appeared to understand the need for good financial management in health care, due to past experiences, those with a rare, life-limiting disease are likely to be more sensitive to discussions about cost.

## DISCUSSION

### Summary of main findings

Although properties of good adult CF specialist care have been noted previously,<sup>9</sup> very little is known about experiences of primary care for adults with CF. This study illustrates that patients with CF operate within two distinct roles when consulting in primary care:

- as lay patients, and
- as expert patients.

Both of these roles have characteristics that are potentially problematic for the doctor–patient relationship.

Within the lay patient role, where doctors are consulted about problems unrelated to CF, patients have no expert knowledge and want to be guided by the medical experts. If the doctor is used to the patient being an 'expert' — as has been the case during consultation for CF-related problems — this switch in roles may go unrecognised by GPs. This can result in delays in the diagnosis of symptoms as they may have been incorrectly attributed to CF.

Within the expert patient role, where participants consulted for CF-related problems, patients saw primary care as simply providing access to treatments — GPs were afforded little more than a functional role. As experts in living and dealing with CF, participants tended to take a consumerist approach to primary care, and from this perspective reported experiencing a doctor–patient relationship shaped by the GP's ability to understand how people lived with CF, their willingness to prescribe specialist medicines, and their focus on the cost of health care.

Although many participants reported a sense that GPs did not initially fully understand how CF affected their lives, to some extent, this appeared to be changing, with some patients stating that the GP had developed greater knowledge about CF over time and was more willing to prescribe specialist medicines. Moreover, previous negative experiences of a heightened awareness of the financial

constraints surrounding CF care had largely been resolved by the introduction of additional resources, such as special allowances, for patients with long-standing illnesses.

The findings of this study suggest that although patients with rare long-term conditions may be encouraged to take on the expert patient role, this may promote a consumer approach to health care and, in particular, a perception of primary care as little more than a gatekeeper of services. Currently, the juxtaposition between the secondary care provision of specialist services and the provision of personalised continuity of care within primary care, provides a potential source of conflict within the doctor–patient relationship.

### Strengths and limitations of the study

The responders in this study were not chosen to be representative of patients with CF, but rather so that they might provide information on the diversity of experiences involved in being treated for CF. Nevertheless, our sample was older than the average patient with CF, and this is likely to have resulted in wider ranges of experiences, particularly in relation to the organisational structure of primary care, which has undergone many changes over the past two decades.

Just over a quarter of patients invited to participate in the study agreed to participate, resulting in a self-selected group who may be particularly disaffected by the services they receive from primary care. It is possible, therefore that our findings represent more extreme views. It is also important to note that these views are solely the perspective of individual patients, as GPs were not interviewed and consultations were not recorded.

### Comparison with existing literature

The government's expert patient policy aims to encourage and enable patients with chronic diseases to take an active role in their own care within the remit of chronic disease management.<sup>1</sup> However, GPs have reported dissatisfaction with current arrangements for prescribing specialist medicines and with being the means of obtaining a prescription;<sup>12</sup> certainly in the present study many patients reported using their GP in this manner. We suggest that patients clearly have a duty to maintain regular contact with their GP and not rely solely on obtaining repeat prescriptions if primary care services for adults with rare long-term conditions are to be improved. Nevertheless, our data suggest that in shared decision-making, the balance of inputs differs within each patient according to the problem being discussed, that is whether they are consulting as an expert on their long-term condition, or as a lay

patient on a problem that they perceive to be unrelated to their underlying condition.

Communication issues raised by these participants tended to reflect the wide-ranging concerns of patients with other chronic health conditions.<sup>17</sup> However, adults with long-term rare conditions may be particularly sensitive to communication difficulties as they have a high level of contact with health professionals in secondary and/or tertiary care.

Clinical unfamiliarity has been noted as being a factor in not prescribing medication;<sup>12</sup> however participants in this study often appeared more likely to view GPs' hesitance as being due to the high financial costs of CF care. GPs are unlikely to see more than one or two patients with a rare condition such as CF during their careers and so unfamiliarity with certain aspects of their care is to be expected. Like others with long-term rare diseases, the number of adults with CF is increasing and will require more primary care services as the amount of patients, CF-related conditions, and survival age increase. Specialist services are already reporting being stretched<sup>18</sup> and more patients are receiving traditionally hospital-based care in the community.<sup>10</sup>

### Implications for clinical practice and future research

This study suggests that financial and prescribing issues had, to some extent, been resolved over time, particularly with the introduction of financial allowances for chronic illness care and a recent policy focus on the management of patients with long-term conditions. The findings, however, raise some important questions about the impact of advances in medical treatments, such as in vitro fertilisation, for people with long-term conditions and suggests that health professionals need to reframe how they make decisions surrounding the appropriate care for these patients.

Expert patient policy has focused on the role of patients with common chronic conditions in secondary and tertiary care, with little consideration of the dynamics of how adults with rare chronic illness and their GPs manage health problems that are addressed in primary care. Enabling easy access to holistic care and treatment, as well as establishing successful trusting relationships with people with long-term rare conditions, is a vital part of primary care and a necessary foundation for the expert patient to take an active role in his or her care.

The perceived financial burden that adults with CF feel they represent may also be pertinent for other young people with traditional rare diseases of childhood who are experiencing increased longevity and who, as adults, bear the responsibility for their health and health care. Future research must focus

more closely on how these 'expert patients' and their GPs experience the interface between primary and secondary care, how they can best be supported in managing the conditions, and how young people's care can be improved most effectively.

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### Ethics committee

Royal Brompton Hospital/National Heart & Lung Institute Research Ethics Committee

### Competing interests

The authors have stated that there are none

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