Information needs of parents of infants diagnosed with cystic fibrosis: Results of a pilot study

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Abstract
This study investigated the information needs, priorities and information-seeking behaviours of parents of infants recently diagnosed with cystic fibrosis (CF) following newborn screening, by piloting the 'Care of Cystic Fibrosis Families Survey'. The questionnaires were posted to eligible parents (n = 66) attending CF clinics in hospitals in two Australian states; reply-paid envelopes were provided for return of the questionnaires. Twenty-six were returned (response rate 39.4%). The most common questions to which parents required answers during their initial education period related to what CF is, how it is treated and how to care for their child. Parents preferred face-to-face consultations to deliver information, and yet all reported using the Internet to search for more information at some point during the education period. Many parents provided negative feedback about being given their child’s CF diagnosis via telephone. The timing, content and method of information delivery can all affect the initial education experience. We can deliver education to better suit the information needs and priorities for education of parents of infants recently diagnosed with CF. The Care of Cystic Fibrosis Families Survey was successfully piloted and recommendations for amendments have been made for use in a larger study across Australia.

Keywords
Communication, cystic fibrosis, education of parents, information needs, newborn screening

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Introduction

Cystic fibrosis (CF), the most common genetic condition in Caucasians, is generally identified through routine newborn screening followed by other confirmatory diagnostic tests. Parents may find a diagnosis of CF in their infant to be unexpected and confronting, and have little knowledge of the ramifications of such a diagnosis on their families. In Australia, this information is imparted to them when they come under the care of specialist staff. Cystic fibrosis outpatient centres are established in the major children’s hospitals; it is here that parents of infants newly diagnosed with CF will receive initial education from the interdisciplinary team about the disease and its management. Many parents from regional, rural or remote locations will need to attend a CF centre located in the capital cities to receive this education, although ongoing clinical management of their child may be undertaken on an outreach basis.

There is limited research about the information needs and priorities of parents of infants newly diagnosed with CF following newborn screening, although some previous studies suggested that such needs were not being met (Hummelinck and Pollock, 2006; Jessup et al., 2016; Sawyer and Glazner, 2004). Some studies have identified that parents experienced information overload, distress, shock, grief or disbelief during the initial diagnosis and education period (Hummelinck and Pollock, 2006; Jessup et al., 2016; Tluczek et al., 2006). Any education program needs to consider the content, length of education, timing of education following diagnosis, as well as delivery modes. Through interviews of parents of children with CF, Jessup et al. (2016) concluded that parents preferred a short time frame between the diagnosis of their infant and initial education.

Effective communication is essential when disclosing newborn screening results and will promote understanding and retention of information, lower parental stress and increase participation and shared understanding (Duff and Brownlee, 2008; Hummelinck and Pollock, 2006; Salm et al., 2012; Sweet and Mannix, 2012; Tluczek and De Luca, 2013; Tluczek et al., 2011). Health professionals have suggested strategies for how communication could be improved during the initial disclosure of screening results (Finan et al., 2015). Their recommendations included training on how best to break bad news, listing the most common questions from parents at such a time with sample responses and developing visual aids and written information to reinforce the information provided verbally (Finan et al., 2015).

Aims

This study aimed to investigate the information needs, priorities and information-seeking behaviours of parents of infants newly diagnosed with CF following newborn screening, by piloting the ‘Care of Cystic Fibrosis Families Survey’.

Method

Design

A cross-sectional postal survey design was used, collecting self-reported data from relevant participants who were asked to recall their experience of education provided at an earlier time (retrospective). Both quantitative and qualitative data were collected and analysed.
Participants
Participants were parents (or guardians) attending the tertiary specialist CF centres in two Australian states, called centre A and centre B in this article. There were three convenience samples: parents of children aged one to five years, attending centre A ($n = 16$); parents of children aged from birth to one year ($n = 20$), attending centre B; and parents of children aged one to five years ($n = 30$), attending centre B. Parents were excluded if their infants did not yet have a definitive diagnosis of CF, or if clinical staff deemed them to be at risk of psychological deterioration should they participate.

Data collection tool
The Care of Cystic Fibrosis Families Survey, informed by a previous qualitative study (Jessup et al., 2016), was developed specifically for this study. Development took place over one year at the main CF centre in Western Australia. Jessup et al.’s findings were combined with the relatively sparse existing literature about the psychosocial care of children and families with CF, and with the experiences of the multidisciplinary team members, to guide the design of the initial version of the survey. Various iterations of the questionnaire followed, based on responses from parents on a CF advisory panel that had been set up to oversee a series of studies, from health professionals working with families affected by CF and from members of the general public. Given that CF parents are bombarded with requests to be part of research studies, it was deemed appropriate to do the initial development away from them. Once responses from the various steps of the development were completed, statistical advice was sought to ensure the phrasing of questions would generate data that could be analysed effectively.

This pilot study was the first application of the survey in a ‘real’ sample. There were two slightly different versions of the survey used in this pilot, with the questionnaire distributed to parents with children aged one to five years attending centre B having some additional questions and minor wording amendments, based on initial sampling at centre A. Thus, there were unequal sample sizes for those additional questions. The survey consisted of five sections:

- **Section A** – questions about the parent and the family
- **Section B** – questions about the initial CF education period following their baby’s diagnosis, when parents met the health professionals in the CF team and first learned about their child’s CF care
- **Section C** – questions about how the initial CF education was organized and how information was given to the parents
- **Section D** – questions about ways that parents searched for information about CF
- **Section E** – asked for any other comments the parents may have had.

To date, there has been no psychometric testing of the tool; rather, we tested content and face validity, internal consistency of the questions within their sections and clarity of each item as perceived by the respondents.

Ethical considerations
The study was approved by the research ethics committees of the relevant institutions (approval numbers 1870EP and HREC-14-QRCH-327). The questionnaires were completed anonymously;
the researchers had no access to the names of the parents who had been invited to participate, nor to the names of the parents who returned their questionnaires. All study documentation was stored securely, with access to data limited to the researchers, as per the conditions of the ethics approvals.

**Study process**

The survey packs were assembled by the researchers and consisted of a participant information sheet, the questionnaire and an addressed postage-paid envelope for returning the completed questionnaire directly to the researchers. Staff from the CF centres were asked to identify appropriate potential participants, and then distribute the survey packs to those parents.

**Data analysis**

Responses were analysed using descriptive statistics. Responses to the open-ended questions were used to provide depth to the quantitative data. Responses were reviewed within each question and across questions, with words/phrases that denoted similar concepts grouped together and represented in reporting by the word/phrase used most often and/or that best captured the collated meaning. Since this was also a pilot study of the survey, responses to the questions were reviewed for internal consistency, and areas where the survey could be improved for future use were identified.

**Results**

**Respondents**

Twenty-six questionnaires were returned, an overall response rate of 39.4%. Response rates varied between the three groups: 10/16 (63%) parents of children aged one to five years attending centre A responded; 2/20 (10%) parents of children aged from birth to one year attending centre B responded; 14/30 (47%) parents of children aged one to five years attending centre B responded. The mean age of respondents was 31 years; all but one respondent was female; all but one spoke only English; and 17 (65%) respondents had completed additional post-secondary school education.

**Components of the initial education period**

The length of initial CF education varied – from less than three days (n = 18, 69.2%), between three days and five days (n = 5, 19.2%), to more than five days (n = 3, 11.5%). During that time of initial education, most parents met with the specialist CF doctor, the CF nurse specialist and a specialist physiotherapist. Most (n = 19, 73%) attended the initial education session with their partner, with only one attending without a support person. This person commented that being there alone was very difficult. Respondents were asked to rate how important the various education topics (refer to Table 1) were at that initial session. The topics that were most frequently rated ‘essential to know’ were treatment of CF, such as antibiotics, hospital visits and medications (n = 25, 96.2%); caring for their child, such as daily routines at home, and mealtimes (n = 25, 96.2%); and information about the disease pathophysiology and how it will affect the child (n = 24, 92.3%).
The topics included in the initial education sessions that were less frequently rated ‘essential to know’ were genetics of CF, including how it is passed on and having more children ($n = 10, 38.5\%$); social aspects of CF, such as social benefits and financial implications ($n = 11, 42.3\%$); emotional aspects of having a child with CF, such as how to deal with feelings and get support ($n = 15, 57.7\%$); the impact CF may have on their home life, family and quality of life ($n = 16, 61.6\%$); and research and new advances in CF ($n = 16, 61.6\%$). The genetics of CF, impact of CF on the family and quality of life, and emotional aspects of having a child with CF were each rated ‘not that important’ by one respondent. Social aspects of CF were rated ‘not that important’ to know by three respondents.

**Organization of the initial education sessions**

The majority of the respondents ($n = 19, 73.1\%$) indicated that they had received the right amount of information during the initial education session, with a minority indicating that they had not received enough information about some very specific aspects of CF. For example, one parent ($3.8\%$) indicated they had insufficient information to be able to explain their child’s diagnosis of CF to friends or family and three parents ($11.5\%$) indicated they did not know about where to access further information about CF. Only one person said they were given too much information. There was considerable variation in preferences as to how the respondents would have preferred the initial education period to be organized – from all at once over a few days or a week ($n = 11, 42.3\%$), at intervals over the first month after diagnosis ($n = 6, 23.1\%$) or at intervals over the first 6–12 months ($n = 4, 15.4\%$). Other options were suggested by the remaining five respondents.

Sixteen parents ($61.5\%$) said they would have liked the opportunity to meet with other parents of children with CF during this initial education period, although seven parents ($26.9\%$) did not want such an opportunity. One of those parents qualified their response by stating that they would have liked an offer to meet with other parents at a later time. One parent suggested an online forum to connect with other Australian parents and families.

A free-text question within section C of the questionnaire (organization and delivery of education) asked about which information sources from the CF team were most valued. Table 2 summarizes these responses. The written responses gave some explanations as to why individual consultations were considered most valuable. For example, respondents explained that during face-to-face consultations they could have their questions answered immediately, and that the practical demonstrations offered during such consultations helped to clarify any misunderstandings, and...
they appreciated meeting with health professionals from the various disciplines and with different experiences.

**Initial information about child’s CF diagnosis**

An additional question included in the amended questionnaire asked about how parents were informed about their child’s CF positive screening diagnosis. The majority ($n = 11/14, 78.6\%$) were told via telephone, and the remaining ($n = 3/14, 21.4\%$) were told face-to-face. This was considered unsatisfactory because there was a lack of sensitivity as to where the parent might be when they were having that initial crucial phone conversation, who might be with them at that time or the inappropriateness of making that phone call late on a Friday afternoon when the parent obviously had no opportunity to receive relevant credible information from the health service. Also, some of these phone calls were delivered by people with poor English skills, or who had no specific knowledge about the implications of a CF diagnosis.

**Searching for information about CF**

All respondents searched the Internet for more information about CF, with respective state CF associations being the second most frequent source of additional information. The topics about which parents most often reported seeking more information were life expectancy, treatment (including lung transplants), what CF is, quality of life and the daily routine of caring for a child with CF. The parents reported that searching for such information assisted them greatly to fill in gaps in their knowledge and prepared them to ask more questions of the specialist CF team. However, their searching was less useful in assisting them to prepare for learning about CF and working with the CF team, or to be more involved in decisions about their child’s care. Parents were also asked a free-text question about how searching for information about CF on the Internet made them feel. Comments were more frequently associated with negative emotions, such as fearful, horrible, upset and overwhelmed ($n = 27$) compared to positive emotions such as better informed and empowered ($n = 16$).

**The questionnaire**

The majority of the questions asked within the Care of Cystic Fibrosis Families Survey elicited consistent responses. Recommendations were made to modify the wording of seven questions to increase clarity, add additional questions to expand upon the meaning of two questions in the survey,
one about the participant’s prior involvement with CF, which may have affected the responses, and the other asked about satisfaction with the education provided by the CF team. Both were broken into two questions. One question was recommended to be reworded to address apparent misunderstandings.

**Discussion**

The majority of the parents who returned their surveys indicated that they considered factual information about CF itself to be more important than some of the psychosocial topics usually included at the time of initial education. They indicated that they needed answers about what CF is, how it is treated and how they could best care for their child. At that time, information about the genetics of CF was not considered ‘essential to know’ by the majority of respondents, consistent with findings of Sawyer and Glazner (2004). It may also reflect other literature that parents need practical information and the skills to care for their child day to day at this initial time, rather than dwelling on the negative aspects of CF (Jessup et al., 2016). Perhaps those topics that respondents thought less essential to be imparted during the initial education session could be left to a later stage during the child’s ongoing care. If that was the case, parents could be provided with written information to be taken away and perused at their leisure in their own homes, and told that the topics would definitely be addressed at a future visit.

Consistent with other studies (e.g. Collins et al., 2013; Duff and Brownlee, 2008; Hayeems et al., 2008; Jessup et al., 2013; Salm et al., 2012), respondents to this survey clearly preferred face-to-face communication, especially about the newborn screening results, with a health professional with expertise about CF. Although Collins et al. (2013) identified that the positive aspect of disclosure of results by telephone was the provision of immediate answers, face-to-face disclosure of information was more helpful, because it was reassuring, comfortable and personable. They suggested the need to prioritize the information to be provided at the time of disclosing the CF diagnosis. From the responses of parents in this current study, it was clear that the delivery of information about their child’s diagnosis via telephone was not appreciated. It is recommended that actions be taken by CF centres to consider the most appropriate ways to deliver such potentially distressing news. Use of technologies such as Face time and Skype may partially address some of the concerns raised regarding telephone notification, while also catering for service access challenges experienced by rural and remote families.

It was also clear from the responses to many of the questions that it is extremely difficult to arrange education in a standardized way. For example, the considerable variation in preferences about when the initial education should be delivered highlights that there needs to be ways to ascertain the essentials, and then to deliver additional ‘important to know’ information at intervals to meet the needs of the individuals. These preferences may well be influenced by the distance from the CF centre. For example, parents living in rural areas may prefer to go to the capital city for a week to acquire information at a sufficient level for them to feel confident to care for their child at home. However, it would also be likely that it might be more difficult for a partner to attend for all of that time, for example, if they lived on a farm or had a larger family that needed care. Parent-driven educational programs, in which the educational information is delivered in formats preferred by the parents, may be effective and enhance parents’ confidence and skills (McDonald et al., 2013). Such tailored programs might also help to address comments from some parents in this study that more detailed information was not provided when it was asked for, and who felt that the information was not tailored to their level of intellect.
The common use of the Internet for additional information about CF by the parents in this study is similar to findings by other researchers (e.g. Dillard et al., 2010; Duff and Brownlee, 2008; Jessup et al., 2016). Given that parents sought additional information via the Internet, healthcare professionals need to give careful consideration as to how parents can be guided to credible information about CF (Jordan and Chambers, 2016; Pimentel et al., 2013). Written information could include a list of suggested credible websites with embedded hyperlinks that can be accessed with one click, for documents being emailed to families and/or posted on the healthcare organization’s website or social media platform(s). Jordan and Chambers (2016) have suggested ways to make healthcare information more accessible to non-professionals who do not have access to academic journals. These include existing social media platforms rather than specific websites, as well as more traditional sources such as leaflets and videos. The provision of information via different media acknowledges that there is no single best way to deliver education. Parents wrote that they would welcome the opportunity to be introduced to other parents of children with CF. If it was not possible for them to meet in person, then the use of online blogs and social media groups may provide another source of support. The hosting and moderating of social media groups by healthcare organizations could serve to provide a source of credible information, while also monitoring and correcting any misinformation posted by families.

Strengths and limitations
This pilot study tested the use and appropriateness of a questionnaire specifically designed to use with parents of infants newly diagnosed with CF, through newborn screening. To the best of our knowledge, this is the first questionnaire to do so. Once the adaptations found in this pilot are made, it will undergo further testing with a small sample (to ensure the adaptations work) and it will be rolled out across Australia.

This pilot will be relevant to countries where testing for CF is part of routine newborn screening. Other countries could possibly use it to survey parents whose child is diagnosed through different methods, but some of the questions may have to be adapted to reflect that different diagnostic process.

The anonymous nature of the questionnaire, and its return directly to the research team rather than the clinical team, were both strengths of the study’s design. It was considered essential to clearly differentiate between the research team and the clinical team, so as not to disrupt the ongoing relationship between the parents and the staff who would help them over a prolonged period to care for their child. The piloting of the Care of Cystic Fibrosis Families Survey in two Australian states has resulted in identification of ways that it can be improved for use in a broader survey.

The relative rareness of CF will always restrict the potential sample size. Efforts were made to encourage responses, and the inclusion of the three groups of parents was one strategy to increase the overall sample size. The small convenience sample also restricted the types of analyses that could be undertaken. Given the specific demographics of this respondent group (e.g. predominantly post-secondary educated women), widespread generalization would not be appropriate.

A key limitation was the possibility of recall bias, particularly for those parents who responded to the questionnaires some months or years after disclosure of the initial newborn screening result. However, this was a consequence of expanding the age ranges of the children in order to increase the number of questionnaires distributed. Since the parents of infants with CF have ongoing involvement with the healthcare system, this additional time frame may have elicited some
considered responses about the type and mode of delivery of information they would have valued when their baby was first diagnosed.

It was too early to undertake psychometric testing. This pilot afforded the team the opportunity to test content and face validity, internal consistency of the questions by sections, and item clarity. The study will be rolled out in a larger, national sample and appropriate psychometric testing will be done then.

**Recommendations for future research**

A national study with a longer time frame for distribution of surveys would increase the sample size. Reminders to prompt parents to complete the questionnaire, along with a small incentive may enhance the response rate. It would be appropriate to use inferential statistics for subgroup analyses if there was a larger number of completed questionnaires. The survey was successful in eliciting the intended input from the small target population of parents of children with CF, and will provide an effective tool for further national and international research.

**Implications for clinical practice**

From the responses to this survey, it would seem reasonable to recommend that the initial diagnosis of CF be given face-to-face by a healthcare professional knowledgeable about CF, rather than parents being delivered this ‘bad’ news via telephone by a health professional without such specialist knowledge. If a face-to-face appointment is not possible due to access issues such as distance or cost, then using videoconferencing could add visual cues that would provide better emotional and educational support during this crucial conversation. Since parents accessed the Internet for information about CF, it is recommended that health professionals provide details of relevant and reputable websites to parents and families of children with CF at the earliest opportunity.

**Conclusion**

The Care of Cystic Fibrosis Families Survey was successfully piloted with 26 parents of children diagnosed with CF. It elicited information about the information needs and priorities of those parents when their infants were initially diagnosed. The most important questions parents required answers to during their initial education period were in relation to learning about the disease, treatment and how to care for their child. Parents highly valued face-to-face consultations with their health professionals. This study contributes to the goal of developing adequate, appropriate and timely information packages tailored to the needs of individuals at a particular time. It is recognized that parents’ information preferences and priorities will change as their child grows.

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Authors’ Contributions
DJE carried out all aspects of the study, under the supervision of KW, WS, LS and TD. All authors contributed equally to manuscript preparation.

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