

Waiting for the next Shoe to Drop: The Experience of Parents of Children with Fanconi Anemia

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Received: 10 March 2011 / Accepted: 11 July 2011
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Abstract Fanconi Anemia (FA) is a rare genetic disease that generally affects children and results in bone marrow failure requiring blood or marrow transplantation for survival. A unique feature of the condition is the long, often many years, waiting period between genetic diagnosis and treatment. This qualitative study looked at the lived experience of parents confronting their child's diagnosis of FA. We aimed to describe factors which parents found helpful or detrimental during the waiting time period and to recommend strategies to support families who will have these experiences in the future. Categories that emerged were: parents' emotional responses, thoughts about FA (which occurred daily for most parents), sources of stress, mechanisms of coping, family dynamics and responses that were supportive and non-supportive. We found that most parents experience stress, uncertainty, and active surveillance throughout the course of the illness. Healthcare professionals, and especially physicians, were agents of both the most and least supportive experiences of parents. Parents

described family centered team care as helpful throughout the illness and health professional education as a priority need.

Keywords Fanconi anemia · Parents · Psychosocial · Waiting · Coping · Stressors · Uncertainty · Bone marrow transplantation

Introduction

Fanconi anemia (FA) is a rare genetic disease first described by Guido Fanconi, a Swiss pediatrician, in 1927 (Fanconi 1927). The condition affects approximately 1 in 350,000 people worldwide (Schroeder et al. 1976). Individuals with FA may have an assortment of congenital anomalies including short stature, absent thumbs, kidney and heart defects (Kutler et al. 2003). Almost inevitably a person with FA will have bone marrow failure where the bone marrow stops producing one or more blood cells such as red and white blood cells or platelets. Bone marrow failure routinely occurs in the first decade of life affecting 90% of individuals by the age of 40 (Alter et al. 2003; Kutler et al. 2003). When bone marrow failure occurs cord blood or marrow transplantation (BMT) is usually necessary. BMT uses chemotherapy and/or radiation to destroy a patient's bone marrow cells. The patient's immune system is severely suppressed to allow for blood-forming stem cells to be intravenously administered replacing the missing or defective marrow, blood, and immune cells. Due to the possible complications and risks associated with BMT, the procedure is typically delayed as long as possible. Individuals with FA may wait years to decades from the time they are diagnosed until they require BMT. During this time period, individuals are monitored by complete blood counts every 3–4 months, annual bone

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marrow biopsies, and personalized screening for affected organ systems. After BMT, medical surveillance is mandatory for on-going medical concerns especially focused on screening for squamous cell carcinomas of the head and neck, gastrointestinal or gynecological system as early as the teenage years (Alter et al. 2003). Thus, even after successful BMT there may be indefinite uncertainty associated with a potential cancer diagnosis.

Waiting is a common phenomenon experienced by individuals with genetic and chronic diseases such as FA. In general the act of waiting is associated with frustration, suffering, discomfort, anxiety and restlessness (Naef and Bournes 2009). For parents of children undergoing donor transplantation for kidney, heart, and liver transplantation, the waiting period is associated with great psychological distress (Naef and Bournes 2009; Simons et al. 2007). Almost 80% of parents of children waiting for a cardiac transplant and 50% of parents of children waiting for a liver transplant report moderate distress (Suddaby et al. 1997; Tarbell and Kosmach 1998). Clinically significant levels of stress were reported in about 20% of parents with children undergoing a variety of organ transplantations (Rodrigue et al. 1997; Simons et al. 2007). A recent review by Packman et al. summarizes the body of knowledge on BMT related psychological effects (Packman et al. 2010). BMT is traditionally used for life-threatening diseases in need of immediate treatment with a relatively short waiting period of months. In this acute time-period, one study found 50% of mothers experienced anxiety and 8% were clinically depressed (Barrera et al. 2000). The level of depression is much lower than that detected in a study done by Nelson et al. which showed 66% of mothers experienced depression pre-BMT (Nelson et al. 1997). Futterman et al. conducted a study of BMT patients and spouses to evaluate mechanisms of psychological distress and noted that patients and spouses experienced greatest negative and psychological stress during the period prior to admission for BMT (Futterman et al. 1996). The body of literature on psychosocial issues prior to BMT suggests a high level of stress in this acute time-period, but little is known about the long term effects of stress, anxiety and depression associated with illness uncertainty and anticipation of BMT over long periods of time.

Given the likely challenges of raising a child with FA and the contexts in which they occur, we looked at the phenomenon of waiting through a biopsychosocial lens. This approach recognizes the reciprocal influences and complex system dynamics of the family, health care system, and the environment in which health care is provided (McDaniel et al. 1992). Key differences related to the individual, the family, the illness and the environment may play vital roles in understanding stress and coping associated with waiting in childhood genetic and

chronic diseases (Stewart and Mishel 2000). Availability of genetic testing prior to bone marrow failure has extended the concept of illness to include a pre-symptomatic crisis phase. Challenges that need to be met in this phase include creating meaning about genetic information that preserves a family's sense of mastery and competency and to develop flexibility to face an uncertain future (Rolland 1999).

Family dynamics play an important role in dealing with illness-related stress. Gender-related differences in response to stresses of illness may complicate parental communication (McDaniel and Cambell 1999). Women tend to discuss and express their feelings more than men and may grieve for a longer time. Men tend to be less likely to express emotions and to return more quickly to work or other activities (Weil 2008). One study interviewed siblings and found that a qualitative research design provided rich descriptions and novel insights into the "hardships" of living within an FA family (Hutson and Alter 2007). Furthermore, results from a survey of FA families completed by the Fanconi Anemia Research Fund described waiting as an extremely arduous and taxing experience (FA scientific symposium, 2008). Although each of the studies suggested major psychosocial risks to patients and families during the waiting period, none of them examined the issues encountered during the extended nature of waiting in the context of having a child with FA nor recommended strategies for addressing them.

We designed a qualitative descriptive study to address this knowledge gap. This approach is phenomenological in that it addresses the meaning of living with FA between diagnosis and treatment for bone marrow failure (i.e. steroid treatment and/or BMT) and to identify strategies to address any challenges. We hypothesized that this may be the most stressful period over the course of the family experience with FA. The first aim was to describe the lived experience by interviewing parents of children with FA who are currently, or who have, experienced waiting. The second aim was to identify factors which parents found helpful or detrimental during the specified time-period. Lastly we sought to provide strategies for health care professionals to aid in supporting parents and to offer advice to other parents with a child who has FA.

Methods

Study Design

A series of six demographic and fourteen open-ended questions was created for a semi-structured interview guide. Main areas of investigation included family life before and after learning about FA, stressful times, means of coping,

resources used during the waiting period, and suggestions for parents and providers. The study used qualitative approaches to understand family systems phenomena linking qualitative research to family systems theory (Rosenblatt and Fischer 1993). Qualitative methods allow inductive processes that look for patterns of congruence or incongruence. A major advantage to using such a method is that researchers make sense of the data without imposing pre-existing constraints (Patton 1990).

Participant Recruitment and Data Collection

The study was approved by University of Minnesota Institutional Review Board (IRB). Following IRB approval, parents of children with FA visiting the University of Minnesota for a return clinic visit in the next 2 months were sent a letter of invitation and consent form describing the study. No financial or other incentives were offered for participation. New referrals and patients less than 6 months post-BMT were excluded from the study. The average number of return patients fitting these criteria was estimated to be about three per month. Following the original 2 month recruitment period, a second batch of study invitations was sent out approximately 6 months after the first. The possible participants were asked to return the signed consent form or contact the co-investigator to schedule an interview during their regularly scheduled time at the clinic. Participants who had not responded to the initial mailing were called 1 week prior to their visit to determine interest in study participation. Parents who agreed to be in the study were offered the option of participating in an in-person interview during their clinic visit at the University of Minnesota Medical Center Fanconi Anemia Comprehensive Care Program or to schedule a phone interview. All interviews were recorded and transcribed.

Data Analysis

An inductive content analysis of parental responses to the interview questions was conducted. The first step in analysis was to select passages, either sentences or paragraphs, that described family experiences related to waiting and assign them a code to describe the content. Initially, each investigator coded the first two interviews independently and then met to agree on the codes and categories they derived. The goal was to be comprehensive in describing all data related to family experiences during the waiting time. The two investigators then met to review the remaining interviews, to assign codes, and to agree on language to describe major categories. One investigator then entered the content and codes for all of the interviews into the qualitative data-analysis software Atlas/ti Version 6.2. Finally, both investigators met again to review all codes

and assignments and to create a chart that included all categories and codes that represented sub-categories. Codes were added, deleted, and revised in an iterative process of moving between codes and the data they reflected. Finally, a complete list of categories was created and codes enumerated to reflect all data most relevant to parents' experience with FA. Quotes were not attributed in order to maintain privacy for families with this rare syndrome.

Results

Participants

Parents of nine patients were invited to participate in the study. At least one parent from a total of seven out of a possible nine patients' (78%) agreed to participate in the study. Four interviews were conducted in person and three were conducted by phone. Interviews lasted approximately 1–1.5 h. Of the two individuals not interviewed, one parent noted that she did not feel as though she would have much to contribute since the time from diagnosis to transplant was short. The other family did not return the consent forms and could not be contacted prior to their appointment. After reviewing the consent form and granting permission to record the conversation, a total of two couples interviewed together, four mothers and one father participated individually in semi-structured interviews (Table 1). After interview five, no new categories arose from data analysis. Therefore, we did not pursue additional interviews after the initial seven were conducted.

The participants had varying age at diagnosis, time between diagnosis to BMT (waiting period), and time post-transplant (Table 1). Children with FA had been diagnosed from age 6 months to 11 years. Five of the patients had had a BMT and were between 6 months and 2 years post-BMT. Time from diagnosis to BMT ranged from 1 month to 4 years. The remaining two patients had not yet needed treatment for bone marrow failure and were 3.5 and 4 years

Table 1 Characteristics of study participants

	Parent #1	Parent #2	Age at diagnosis (years)	Time waiting (years)	Time post-transplantation (years)
Child #1	Mother	Father	0.5	4	0.5
Child #2	Mother		9	1	2
Child #3	Mother		8	2	1
Child #4		Father	11	4	N/A
Child #5	Mother		0.5	3.5	N/A
Child #6	Mother		3	1	2
Child #7	Mother	Father	10	9	2

past diagnosis at the time of the interview. The majority of respondents were Caucasian and married. For the purposes of privacy, the ages, marriage status, and ethnic backgrounds of parents were not included.

Parents' Experiences from Diagnosis to Bone Marrow Transplantation and Beyond

Seven major categories were identified and a list of parents' suggestions for other parents and medical professionals was created. The categories that arose from the descriptive interviews included emotional responses to the diagnosis, thoughts about FA, sources of stress, mechanisms of coping, family dynamics, support systems, and unsupportive responses.

Emotional Responses: Acute and Chronic

The initial diagnosis of FA was described as a time of acute intense distress.

Parents stated,

"After we found out about FA I had sleepless nights; It just engulfs the whole person. You can't do your job, can't concentrate so everything suffers."

"Yeah, you know the first 6 months was a shock. Every day I would say to my husband, why can't we just go to transplant now, why can't we just get it over with now. I hate waiting, I really do"

"The first 6 months, I was literally to the point where I wasn't doing my hair, I wasn't putting on make-up. I was coming home taking my uniform off and putting on sweatpants and I would hold him and I wouldn't put him down."

Some families experience blame and anxiety surrounding the diagnosis. For example,

"I had a really tough time dealing with the fact that I had a child that was different and had a child with a life-threatening disease. I blamed it on myself of course and I blamed it on my husband so I had a really tough time at first so at that point I was being forced to go seek medical attention for mental health. I didn't take it seriously. I didn't think anyone could help me. I had to accept it and I had to do that on my own, no one else could help me do that. My husband couldn't even help me do that."

Overtime a transition into the realization of the chronicity of the disease takes place. One parent described the experience,

"What was very interesting is when you first get diagnosed, it's more of an acute grief, and then you come more to terms with it but there's still a chronicity to it, and anxiety, definitely anxiety."

The grief associated with the disease persists as described by one mother.

"There's definitely a grieving process that goes on... I think that even continues post-transplant, because you realize things are never truly normal in the sense of what we call 'normal,'... I very much cried, probably about every day."

Although the initial aim was to understand the time between diagnoses to treatment for bone marrow failure, the parents reinforced the notion that the grief and thinking about FA continues indefinitely.

Thoughts about FA

All parents stated that they thought about FA everyday and the concerns regarding the health of their children continued with as much, if not more intensity, after BMT. The sentiment is summarized in the following quotes,

"You're always kind of waiting for the other shoe to drop, because you know that it is coming because that is pretty much how the disease works. You know what the future holds but you just don't know when that is going to happen."

"It's like a ticking time bomb. You think about it every day. You know it's going to happen; you just don't know when. But we don't obsess about it. If you get a bad doctor's report 1 month, you're going to obsess about it for a couple days."

"Life has never been normal since then. Every day there's the burden in your heart."

What was somewhat unexpected was the continued magnitude of worrying about FA post-BMT. Although we did not specifically ask about this time period in our interview, many parents commented on their unrelenting worries. For example, two parents describe their feelings post-BMT.

"I'm a nervous wreck, to be honest. I'm waiting on something. She's done so well, it's kind of ironic to say that I'm just waiting on the ball to drop"

"But of course he's not cured. The next step is cancer on the horizon, so you've got to keep your eye out for that constantly."

Stressors

Stressors during the waiting period were predominantly medically related. Other stressors included financial matters and telling others about the disease. Examples of medical

stressors included searching for a donor, working with uninformed doctors, undergoing treatments, and the process of preimplantation genetic diagnosis.

Searching for a donor:

“Last year we were in a crisis mode because we were looking to find a donor really quickly, and we did not know how the idea was going to work. It was really tough last year to maintain a normal life at home because there were times when my wife and I were away to the marrow drives.” (Searching for a donor)

Working with uninformed doctors:

“And then the obstacle of doctors, us having to tell them what to do, at home not here.”

Undergoing treatments:

“The worst day for us I think, was knowing when he was getting radiated... It’s almost like you’re killing your child is the way I felt about it, even though it’s invisible and he’s laying there.”

Preimplantation genetic diagnosis:

“That year-and-a-half was probably the hardest because I was on bed rest a lot of the times. We would travel to Chicago to try to get it (PGD) done one time. I was gone for 11 days from my babies; that was awful.”

Financial matters made spending time with sick children difficult. One parent stated:

“We were pretty much broke, so that was a big stress on me before we came here, especially when I found out we only had \$250 K for transplant and it cost \$300 K-some. We went and got a loan from the bank. They gave us that. We had enough equity in the house and this, that and the other to be able to cover everything. I should probably be playing with him all day instead of working till after he goes to bed at night, and that was a big deal. That was my hardest thing.”

Talking about a sick child to others also brought about stress. Another parent described his experience:

“A lot of times when there is a serious health issue, you don’t want other people to know about it in the community. You kind of try to hide it because you don’t want people to be asking you about this all the time. But then how are you going to be able to get help?”

Coping

Families interviewed articulated many ways in which they cope with being a parent of a child with FA (Table 2). The

most frequently stated ways to cope were positive thinking, delaying worry, followed by informing children with FA about their medical needs. Positive thinking included quotes focusing on being optimistic, cherishing each day, and finding good in the situation. One parent describes her experience as follows,

“We’ve got a lot of really negative things and I just don’t want us to get stuck there because we have a lot of things to be hopeful for too. It took me a long time to allow myself to feel that way. I don’t know, I guess I just wanted to be realistic about our situation. So it took me a long time to figure out that balance between being realistic but also being hopeful and optimistic.”

Similar to positive thinking, many parents stated that they were not going to worry about things until a situation arose that necessitated concern. For example,

“Well, I think prior to the start of bone marrow failure and very obviously needing something, I was happy being ignorant”

A notable number of parents identified educating their children about their medical condition as a stress reliever. One mother said,

“We let him know everything that was going on. We didn’t hide one thing from him. He knows when he’s getting a shot; he knows when he’s getting an IV. A lot of kids, we noticed over at Ronald McDonald House, their parents never told them, they threw holy fits and they were very uncontrollable. It was awful.”

Another mother remarked,

“(Child with FA) came in to me and said, ‘now, this thing I have, it will kill me, right?’ Yes, it will. He had heard enough. I said, ‘you’ve been there. You’ve heard it. You know.’ I think probably one of the most freeing moments.”

Other coping strategies were informing the general public and siblings, normalizing the condition so that the kid was ‘not just a sick kid.’ Some parents identified information seeking as a way to reduce stress while others found avoiding information to be a better way to cope. Lastly some families used fundraising, religion, and medications to promote coping. Two participants mentioned that they did not currently use counseling but had considered counseling, if necessary in the future, for either themselves or their entire family.

Family Dynamics

The family responses to FA focused on differences in parental coping methods, interactions of parents with

Table 2 Coping subcategories and representative quotes

Subcategory	Representative Quote(s)
Positive Thinking	<p>“It just brings to the forefront that there’s no guarantee in life, so you learn to cherish every day.”</p> <p>“Of course it’s grave when you first find out about it. It’s this horrible, grave description of what could possibly come and what you may have to deal with in the future. But then as you learn more about it, and you study more about it, you get confident in the terminology and the language of it, it changed. I’m a very, very positive person. I rarely see the negative in things.”</p> <p>“Obviously, you don’t wish anybody to have a condition like FA. That’s something we’ve been dealt and we’ve just been trying to take positives out of that.”</p>
Delaying Worry	<p>“We’re not going to worry until there’s something to worry about.”</p> <p>“Just taking one thing at a time which makes it manageable.”</p>
Normalization	<p>“We’re always trying to make sure he is having the best day that he could have which is not something that you might necessarily think about if you didn’t have a child with a chronic illness. We have to work really hard to make sure he’s not just a sick kid, so that he is not defined by FA.”</p> <p>“You struggle as you go through post-transplant is when you try and regain that sense of normalcy... You start getting to that juncture... where you’re very, very protective of your child and taking care of them, and then you find yourself, you get to a stage where you’re comfortable maybe to try to start pushing them away again, and that did not exist previously... You have the chance to grow up and the chance to live your own life.”</p>
Informing Others	
General	<p>“We’ve always been really honest about our friends and family, we’ve always felt like that was best route for us so that if everyone understood it, he wouldn’t have to be a just the kid with FA. If his friends parents know about it and his friends know about it then it will be (son’s name) who also has FA.”</p> <p>““I love talking about it. I love telling people about my son about his disease, because his disease is so rare. I love it. I get on a high horse.”</p>
Children with FA	<p>“We’re always honest with him that he’s going to have to do things that his brother doesn’t have to do and that his friends won’t have to do. We’re just trying to be honest with him at a 5 year old level. What his expectations should be and what his parameters are and why. Just little things like why he has to always wear a hat everyday if he is in the sun. To big things like, he may not be able to play football.”</p> <p>“He’s seen something on the Discovery Channel about sperm banks. I say, ‘sperm banks? Yeah, where men deposit their sperm.’ He goes, ‘that’s just weird!’ So we’re talking about stuff. Yeah, you deal with things like that.”</p> <p>“We let him know everything that was going on. We didn’t hide one thing from him. He knows when he’s getting a shot; He knows when he’s getting an IV. A lot of kids, we noticed over at Ronald McDonald House, their parents never told them, they threw holy fits and they were very uncontrollable. It was awful.”</p>
Siblings	<p>“One of the things, too, we try to do is we always involve her. If we had a doctor’s appointment she would go. We wanted to make her aware of what was going on. If she was there I felt like she could sort of process it in her own little bitty mind as to what exactly is going on. I think it helped.”</p>
Fundraising	<p>“We started getting fundraisers and that has been really powerful tools for us. I think as a parent you feel really helpless because you are kind of at the mercy of the disease and the treatment and there is really not much that you can do about it...As a parent, you are suppose to protect your child and keep them well and keep them healthy. You’re in a situation where you are kind of powerless to do those things so one thing that has made us feel powerful and like we are actually doing something is starting to do fund raisers and raise money. I think that has been really helpful for us emotionally.”</p>
Avoiding some information	<p>“I did it once and it [look at websites] was a mistake and it will not happen again because everyone’s journey is different and I think that’s the reason why I haven’t reached out to other families that have children with FA. I’m not ready to learn about someone else’s journey just yet. I want to just focus on what we have going on with (child with FA). I don’t feel like I have the emotional ‘where-with-all’ to deal with all the other things that might be happening to other people.”</p>
Information seeking	<p>“I read whatever I could, the FARF website, and I read a lot of the Caring Bridge logs and I looked for the ones that are most current, and to me, that was very helpful, not so much for the horror stories, because some people go way off on it. I think you find one that matches your temperament as a family, and that’s what I did. I found ones that were fairly current and that I could tell matched our temperament on things.”</p>
Medication	<p>“I’ll be honest. I had to go on medication to help me with all this, too. That was something I didn’t want to do but I had to do. There wasn’t any other choice.”</p>
Religion	<p>“We’ve always viewed children as gifts of god. In a way, we talked about it even before we had our children. I don’t know why I was so philosophical about it, that if you’re going to have children we will always consider them as gifts of god.”</p>

children, identifying children with FA as extraordinary, sibling animosity and isolation (Table 3). Four of the seven interviews identified parental differences in dealing with having a child with FA. One couple explained, Father:

“From the outset I said I’m a realist. I want to expect the worst and then I’ll be excited when we get good news. I don’t want to be hoping and then be devastated. That’s the way I live my whole life. She’s completely opposite.”

Mother:

“I’m like, everything is great. [Laughs].”

Another wife stated,

“My husband I took very different approaches. I tried to read and get as much information as I possibly could. He preferred the approach of ignorance, relying totally on the doctors here.”

Parents noted that their interactions with their children with FA were different or altered as a result of the diagnosis. One mother noted,

“I do everything in my power to have my daughter have the best life but I think I do more for my son

[who has FA], which is terrible but I think that’s how it is.”

They also talked extensively about the remarkable-ness of their child with FA. They stated the dramatic impact their child had made on their community or their own perception of life. One parent described this phenomenon,

“He’s restored and renewed a lot of faith for a lot of people. That’s the best thing he could have ever done for so many of those folks. It’s been pretty incredible.”

Another parent described their son’s impact on their life,

“Yes, my son is amazing. He has taught me, he has taught me everything. He’s changed my views on so many different things in life.”

We found, similar to Hutson and Alter (2007), that siblings felt both animosity towards their brother or sister with FA and in some cases felt isolated from the family. One mother summarizes her experience with sibling animosity and how other people treat her child with FA and child without FA different,

“I think the worst thing of the whole thing is really the way my daughter acts sometimes, as far as she has

Table 3 Family dynamics subcategories and representative quotes

Subcategory	Representative Quote(s)
Couple	“I think we have definitely handled things differently throughout the process somehow it has worked for us. We have divided and conquered the whole thing, not conquered but kind of battled it and we never really talked about it”
Parent-Children	“ We’ve had to work really hard to make sure that he’s healthy and happy. You know we probably think of things a little bit different than, than other parents will. We’re always trying to make sure he is having the best day that he could have which is not something that you might necessarily think about if you didn’t have a child with a chronic illness.” “Am I going to be taking something away from him because so much more time and energy is focused on (son with FA) and what emotionally, psychologically, he will have to work through as he gets older and has a better understanding of what exactly the disease is. I’ll have to be cognizant of how he is feeling.”
Special children	“He’s 3.5 years old and all I have to do is look at him and I feel so much better about life. Because he is so full of life. He goes through his bone marrow biopsies, he goes through getting poked in his veins every month, getting his physical and occupational therapy, every doctor under the sun and he will sit there and say thank you. He’s amazing, absolutely amazing. “
Sibling animosity	“He’ll (sibling) get so mad and say, ‘by the time I was your age, I had to try trumpet, I had to try soccer, I had to try... and I hated them all.’ He (child with FA) doesn’t do any of them now, so I’m going, how much of it’s just having a second child and going, phttt! And how much of it’s...? I don’t know.” “[Sibling] did a report on FA last year in 8th grade because they were studying genetics...This summer I come out on the porch and they’re talking about things. [Sibling]’s saying, ‘[child with FA], your wiener’s fried. That’s a good thing. He [Child with FA] started to get so mad, and I know he’s not so mad about the whole infertility thing as much as he’s mad about his brother just talking that way to him. It’s like, God, Jiminy Cricket!”
Sibling isolation	“My sons, I know their big thing is they feel left out and like they’re not getting enough attention, but then they feel guilty because to ask for it...”

animosity towards him. To be honest, I don't think it's the way we treat them because we do treat them equally. I think it's because of everybody else because everybody..."

Another parent talked about the isolation that can result in families,

"People will come up and speak to him [affected child] and won't even notice her. It's gotten better now, but the last couple of years that's the way it's been, and you can tell in her attitude; she's very shy. We try and force her out of her shell. Maybe that's not the right thing to do, but we don't want her to be a recluse."

Support Systems

Parents used many different means to gain support during the waiting period (Table 4). By far the largest source of support was doctors familiar with FA. In every interview, physicians were seen as one of the largest means of support. In most cases, the doctor they were referring to was their primary BMT doctor and in some cases also their local hematologist. One parent explains her first experience meeting with a FA BMT doctor:

"[BMT doctor] made us comfortable in the first five minutes. He said *"Let's educate you about FA"*, then talked about the success rates and finally said that [son's name] needs a transplant now. The way he talks about it is more like an educator. We felt better after that half hour than in all the time since diagnosis."

The doctors were also described as working collaboratively with the family. One parent described her appreciation of being kept in the loop during her child's care:

"Well, just the fact that a lot of the doctors have been really nice, especially up here. I might be an over-reactive mom, but they'll listen to me to let me get that off my chest so that I feel better, and then make me feel like I'm part of the loop and not isolated from everything. That really helped."

Religion was a strong area of support for about half of the participants. One father states,

"We have not felt the need to get in touch with counseling, just because we're blessed with a strong faith. Huge, especially my wife. She attends church every morning, and during the summer she goes almost every evening. But almost every day she is there early in the morning."

Other sources of support were spouses, blogs-websites, Camp Sunshine (an annual meeting for FA families in

Maine), Fanconi Anemia Research Fund (FARF), community, employers, friends, and other FA families.

Unsupportive

Uneducated or non-empathetic doctors were identified as the largest source of unsupportive experiences (Table 4). Several families described having to repeatedly explain the disease to local doctors. In addition doctors neglected to really understand the disease or their child's specific needs. One parent noted,

"I did have an issue with one pediatrician where she asked me if [child's name] sucked his thumb and that bothered me. I understand that, that is probably a typical question that she asks everybody, but as a pediatrician you're suppose to know that he doesn't have thumbs."

Another complaint of parents was not being in let 'into the loop.' For instance,

"I like our doctors [local hematologists] but they tend to keep us out. They don't want to tell me what her counts are or what other... I'm used to hearing and knowing what's going on so I wouldn't be shocked if somebody tells me something. I would rather know than be slammed like I did the first time [with the diagnosis]. That was very traumatic."

Interestingly, some unsupportive responses during the waiting period were also sources of support to other individuals. Camp Sunshine and the internet resources were not a positive experience for all parents. Attending camp was described as "too much overload for me" and "in your face." The timing of camp attendance may play a role as one parent explained that their child was newly diagnosed. Overall parents reported many more supportive than non-supportive responses.

Recommendations

Although our original question and aim was to identify suggestions for healthcare professionals, parents spoke extensively about their suggestions for other parents and less so about their recommendations for professionals (Table 5). Many of the suggestions focused on education from FA specialists and from FA families. Other recommendation focused on the psychological consequences of waiting with FA. Parents suggested limiting worry, counseling, attending Camp Sunshine, informing your child, and finding a blog that matches your personality. Recommendations for health care professionals were focused primarily on physicians requesting better education on the disease, better education of families, and increased empathy in regards to giving bad news.

Table 4 Supportive and unsupportive systems subcategories and representative quotes

Subcategory	Supportive Representative Quote(s)	Unsupportive Representative Quote(s)
Doctors	<p>“We just feel really lucky that the hematologist here had seen patients with FA before, so he was able to help guide us through everything prior to getting to Minnesota and a relief to know that we have that waiting when we got back.”</p> <p>“We met with [BMT doctor]. That had to be my best doctor’s appointment because [BMT doctor] and his staff knew that there was everything that there was to know about this disease that I had been researching for 2.5 years.”</p>	<p>“For awhile I had to go in and explain to doctors what FA is. I had an issue with that at first, because I’m like you’re a doctor, you know that we’re coming, you should have researched the disease and known. Yeah, I had a problem with that.”</p> <p>“(Son with FA) was a picky eater and he bruised easily. The pediatrician said “Boys will be boys.””</p>
Blog – Website	<p>“We did it (website) for our friends and family to keep updated on what was going on with us without having to make daily phone calls, that just wasn’t really a feasible option. I was really reluctant to do it because I’m kind of a private person and that was putting all my emotions out there for everyone to read but I actually found it to be really cathartic. We got so much support for that website it was unbelievable...”</p>	<p>“There’s an on-line group through yahoo and I get the e-mails but I do not participate in them because it’s all kind of, how can I explain it? It’s like a click almost... When I have a question that I just cannot find the answer to, my last resort is that I’ll go to the group. I never ever get the response that the people get that are normally on it.”</p> <p>“The internet- The success rates with unrelated donors (which were old) freaked us out the most.”</p>
Camp Sunshine	<p>“Camp Sunshine was phenomenal. A lot of FA kids don’t have the thumb, so they’ve come up with this thing, high four instead of high five. Seeing it and it’s everyday life. You’re not this one in a million person any more. You’re a community and a family and that’s why we always write Family with a big capital FA. We write FAmily on that.”</p> <p>“She’s always been so tiny, she actually felt alone with the way she looked; and then finding out about this, she felt even more alone because it was such a rare disease that nobody else had it. Going up there [camp] and seeing the kids helped her to understand that she’s not alone with it. It really did. She ran up to [spouse] and I and said, ‘There’s other kids like me!’”</p>	<p>“When we first found out, we went to that [Camp]. They stayed through the whole camp out playing, but I couldn’t go through all the doctors. I went to probably the first two days and then I couldn’t handle it. It was too much overload for me, so I had to go.”</p> <p>“I didn’t enjoy the FA group as far as the parents went. I didn’t enjoy the camp up in Maine. I DID NOT AT ALL enjoy that. It maybe was just me. It was kind of ‘in your face.’”</p>
Religion	<p>“Of course religion’s been a big thing. I don’t think that we would have made it through as well as we did if we didn’t have as many prayers as we did.”</p> <p>“Our church has been extremely supportive throughout the whole thing... He’s restored and renewed a lot of faith for a lot of people.”</p>	
Spouses	<p>“If I had a concern, he had that concern right along with me, and vice-versa with him. That’s been the biggest help”</p> <p>“I guess my husband (is source of support). I really think you’ve got to be able to talk, and even though we deal with things differently, you’ve got to feel that love. You’ve got to feel that security.”</p>	
FARF	<p>“The FARF contacted me and sent me information, which was fantastic.”</p>	
Community	<p>“They (community) had put together a little fundraiser for us when we first came up here, and they did a whole big program and spoiled (child with FA) rotten.”</p>	
Other medical professional	<p>“She [Genetic counselor] did a good job, a really good job, and she did it in a way that we could understand it and in a way that we could remember it so that we could explain it to other people. She was great. She also did an amazing thing and job reaching out to us on a personal level too.”</p>	
Employers	<p>“My employer was very, very supportive and then post-FMLA, very supportive if I needed to work from home. They gave me a laptop and connected.”</p>	
Friends	<p>“Our friends formed a support group. They are the ones that help us do the fund raisers... We’ve got really good friends”</p>	
FA families	<p>“Fortunately enough, that very night [mother of a child with FA], called us. That put me at ease so much more because I just felt like I had an outlet, a little gab book. I could pick at her brain and she could hold our hand and follow us along.”</p>	

Discussion

After diagnosis the lives of FA families are never the same. A similar sentiment has been seen in childhood cancer survivors where children and their parents describe the experience as “never over with...always a waiting game” (Woodgate 2006). Koocher and O’Malley (1981) described this phenomenon as *Damocles syndrome*, a state of persistent uncertainty and dread of disease recurrence (Koocher and O’Malley 1981). Parents in this study described similar feelings as “waiting for the next shoe to drop” and “a ticking time bomb.” Fanconi anemia is at the forefront of their thoughts and radiates throughout their family. Similar categories have been noted in children with other chronic illnesses and described as ‘illness at the foreground’ and ‘a transformative experience’ (Knafl and Zoeller 2000). We hypothesized that the ‘waiting’ period between diagnosis and transplant would lead to maximum stress. In reality we found that waiting related stressors were continuous and for some parents heightened after BMT. The experience of living with chronic illness has been noted as involving constant but subconscious thinking about uncertainty (Cohen 1993, 1995). Thinking about FA is however a constant, unrelenting, arduous, conscious experience which impacts every aspect of life.

Uncertainty and Medical Stressors

Families engendered and verbalized the stress triggers and stress responses described in years of childhood illness and waiting. Parents’ main stressors identified in this study were related to their child’s illness and their interactions with the healthcare system that persisted overtime. Cohen (1995) described seven triggers which heighten the level of uncertainty. Parents of children with FA repeatedly exemplified each of the potential triggers of uncertainty (M. H. Cohen 1995). First, parents experience routine medical appointments (e.g. complete blood counts or bone marrow biopsies). Second, parents monitor their child’s well-being and notice minor changes to their child’s symptoms (e.g. bruises, bleeding, or fatigue) that may or may not be related to FA. Third, some medical words or phrases caused increased stress especially when the words were not explained. Fourth, changes to the therapeutic regimen such as increased frequency of doctors’ visits and blood draws created increased stress. Transplant delays and quick moves to transplant were particularly stressful. Fifth, announcements of a person with FA’s death on-line or at a fund-raiser were immensely impactful to parents. Sixth, changes in the child’s developmental stages also increased uncertainty, especially the transition from adolescence when children must start to take personal responsibility of their

health (e.g. use of sun screen, smoking and alcohol) and are at increased risk for cancer associated with these environmental factors. Lastly, in the absence of distraction, nighttime has been described as a trigger of uncertainty. One mother described her inability to sleep at night due to thinking about FA. Parents routinely experienced triggers of uncertainty which constantly reminded them of their child’s diagnosis. Professionals can assume that stress is an ongoing feature of life-long medical care for parents of children with FA.

Effective Coping with Disease Uncertainty

The large number and types of coping and supportive systems identified in this study implied that parents were positively coping and seeking support. Positive thinking was the most common form of coping in our population and has also been noted as the main source of coping in patients awaiting heart transplant (Cupples et al. 1998; Porter et al. 1994). An optimistic interpretation of the disease is also noted in children with another chronic genetic disease, cystic fibrosis (CF) (McCubbin 1984; Venters 1981). Delaying worry was described as an effective process of coping and adapting by our population and has also been used by CF families (Bywater 1981; Kellerman et al. 1980). The act of delaying worry may allow time for parents to mourn the loss of their ‘healthy’ child and focus on coping with the new diagnosis and immediate medical needs (Holaday 1984). Cohen described the act of ‘managed’ information as a means to reduce uncertainty and minimize incapacitation related to overloading on information (M. H. Cohen 1993). Interestingly, parents stated substantial stress relief after informing children about the implications of their disease and about specific medical procedures required to treat their disease. Some of the topics discussed by parents and children may be very complicated to conceive (e.g. infertility and mortality) but discussing these issues ultimately led to parental liberation as well as understanding and trust between parents and children. Open communication has been shown to increase psychological adjustment following BMT (Ho et al. 2002). Ho et al. (2002) showed that pre-transplant family relationships and coping resources associated moderately with psychological distress during the immediate period following BMT. Our findings would suggest that informing children regarding FA may be a very effective mechanism of coping throughout life. Helping children adapt to the medical stressors associated with FA may be beneficial especially surrounding a major medical treatment such as BMT.

Substantial Support

Parents’ main sources of support during the time from diagnosis to treatment included physicians, spouses, com-

Table 5 Recommendations from parents of children with FA to other parents and health care professionals

Suggestion to:	Type of Suggestion	Respective Quotation(s)
Other parents	Get education	<p>“I came here (Minnesota) a couple years ago to attend a parent conference, which was very informative”.</p> <p>“(It was helpful) the very first time we came here, they had us meet with a social worker. At that time we did not know when she would need a bone marrow transplant, so she went over some of the situations that we might come across when we come down here for the transplant, like living arrangements and things like that. She gave us some books on the marrow donor program to read.”</p>
	FA Specialty Center	<p>“I loved it, [BMT doctor] knew everything, obviously he’s like the best doctor in the country. He knew everything, [Genetic counselor] knew everything, his nurses knew everything. Everybody had an answer and nobody tried to beat around giving us an answer. I asked a question and the answer came right out.”</p> <p>“The initial visit is very important, and they’re doing it the right way over here (Minnesota FA Program). The very first time we came, I mentioned that we met with the social worker, but we also met with (genetic counselor) and we met with (transplant coordinator). I met the whole team. That was very useful.”</p>
	Talking with experts	<p>“It makes it a whole lot easier when you can actually talk to someone that knows what’s going on. When we go home we could talk to doctors, but they don’t have a clue.”</p> <p>“Go get the real facts from the real people instead of just trying to get something [from internet] that wasn’t as truthful as what you need to know.”</p>
	Talk with other families	<p>“Being open to people; to help; to knowledge from [other’s] experiences. Reach out.”</p> <p>“Yeah, and other parents. They’re the resources that you need.”</p>
	Limit worry	<p>“Just enjoying your children would be our biggest thing, enjoying who they are as people and not labeling them something different than just this amazing little person.”</p> <p>“Hang on, it’s a bumpy ride. It is, there will be ups and downs. Like I said, I had to figure it out on my own. I couldn’t have anyone else tell me, you’ll be ok, you’ll be ok. I had to come to grips with it on my own. Take a deep breath, sit back and it will be ok, because that little perfect child that looks up at you, needs you more than anything.”</p>
	Attend Camp Sunshine	<p>“That is the one thing (Camp Sunshine) if we could tell any family, that’s what we’d tell them.”</p>
	Counseling	<p>“You’re dealing with so much emotions that you just don’t know where to deal with them all. I didn’t. I would sit in the room crying just over everything because I just didn’t know how to release everything, so I think that (family counseling) would help somebody quite a bit..”</p>
	Child involvement in medical care	<p>“We started working with (child and family life) here and teaching (child with FA) how to swallow pills because I knew that (child) hated the liquid medicines, and that was huge, just to make it so he could swallow pills. That’s probably the biggest solid piece of advice.”</p>
	Blogs	<p>“I looked for the ones (blogs) that are most current, and to me, that was very helpful, not so much for the horror stories, because some people go way off on it. I think you find one that matches your temperament as a family, and that’s what I did. Some of them get very deeply religious, that’s for them and that’s fine. I’m not like that, so I found some that very much matched my temperament, and for me it was helpful because it helped me to grasp the rhythm of the process. I’d get the flow and the rhythm.”</p>
	Doctors	Better education
Giving bad news		<p>“It’s not blaming anybody, but the news should have not been conveyed over the telephone.”</p> <p>“Make them understand, first of all, that it’s going to be a while before you accept that this has happened, so talk to them about the process of grief that people go through, the different stages of that. Everybody is going to take the news differently.”</p>
Educating parents		<p>The one thing I was going to add that would have been helpful to me, to maybe have a little bit more explanation... I can’t think of the clinical term so I’ll just say the different... like what’s a normal hemoglobin? What’s your child’s hemoglobin? The platelets...”</p>

munities (websites, physical, and religious), family and friends. A substantial portion of the interviews focused on the multiple means that families gained support. Doctors appear to play a key role in supporting the medical care and well-being of parents during this time. Health care professionals such as genetic counselors and social workers also provided support. For some, the parents' employer was a significant source of support. It is worthy to note that six out of the seven parents spoke in depth about the extraordinary nature of their child as an exceptional source of support. The children were described as having distinctive personalities and abilities that greatly influenced their parents' lives as well as their community. Many of the support mechanisms were quite variable and even contradictory. Religion and the religious community was a central support system for some families but other parents noted that they had no identifiable faith and used other means for strength. The FA community (FAF, Camp Sunshine) were recognized as being a supportive group for some but also a source of stress to others.

Recommendations

Physician knowledge and communication played the largest role in creating emotional challenges but also in supporting parents of children with FA. Doctors with expertise in FA provided information and medical care that calmed parents, whereas doctors with little understanding of the disease or empathy for the families led to increased concern. In general parents of sick children have a need for information and partnering with physicians (Fisher 2001). Several parents described their preferred method of health care delivery as a family-centered approach. Parents wanted to be involved in decision making and the particulars of their child's medical treatment. A literature review of the needs of parents with chronically sick children found that the need for partnerships is critical (Fisher 2001; Fisher 2001). Family-centered health care provides a way to gain control through collaborating on decision-making with health care professionals in the care of their child (Jerrett and Costello 1996). It has also been noted that when partnerships do not exist, or are perceived to be broken, that parents can become vigilant and assertive (Diehl et al. 1991; Scharer and Dixon 1989). Several parents discussed their positive experiences with family-centered care. One mother even acknowledged that after not receiving the same standards of care at other facilities she did become very upset but always felt better after insisting and finally being included in her child's care. All of the recommendations to health care professionals in our study focused on parents' needs for better education from physicians and communication between physicians and parents reinforcing the need for a family-centered approach to caring for children with FA.

Suggestions to other parents with children who have FA focused on learning about the disease from other parents, experts, and getting a general understanding of disease and healthcare resources. Parents recognized their own need for obtaining accurate information about FA and thought that other families would benefit from more fully understanding the condition from a variety of viewpoints. Gaining information is a necessity that must be met for parents of chronically sick children but how and when this requirement is met appears to be an important, fragile balancing act (Fisher 2001). Other specific suggestions were to start a website, limit worry, and consider counseling.

Genetic counselors can be a bridge to foster communication of difficult technological information and the psychosocial implications of the knowledge to mediate some of the medically-related stressors and assist with appropriate resources. As described by one parent, "She [Genetic counselor] did a good job, a really good job, and she did it in a way that we could understand it and in a way that we could remember it so that we could explain it to other people. She was great. She also did an amazing thing and job reaching out to us on a personal level too." The balance between information being beneficial and harmful may be especially important for FA families. Understanding this equilibrium of need for information and delaying/limiting worry may foster better patient relationships and greater satisfaction with the health care system. Genetic counselors may provide anticipatory education about the difficult psychosocial and physiological issues that will arise throughout the lifespan. Parents of children with FA spoke about their experiences with depression and anxiety as well as their current need or future desire to have counseling. Connecting families with these resources may be another important role of genetic counselors working with FA families.

Limitations

The population of parents in the study may not be generalizable to all parents of children with FA. Recruitment of participants occurred from a FA specialty center. Volunteer participants may have added resources and/or increased coping levels which enabled them to locate the specialized center and therefore be a part of this study. The participants were randomly selected based on month of appointments and included parents who were past the initial period of waiting. Therefore the extraction of information on the waiting period was retrospective for some parents. Many of these parents reflected on the waiting period but also commented on their current

emotional state. Focusing primarily on families who are still waiting for treatment may have resulted in a narrower but truer to life experience. The study criteria excluded newly diagnosed, participants undergoing BMT, and adults with FA. The study does not reflect the experiences of those individuals. The resulting participants were predominantly married Caucasians. Thus the findings may not be generalizable to single parents and individuals from other ethnicities. Our sample, although small, did represent a national cross-section of mothers and fathers from throughout the United States with a range of experiences, length of waiting, and years since BMT. Lastly, the timing of interviews in respect to length of time pre- or post- BMT varied. The specific timing of the interviews may influence the emotional responses to the survey and the results obtained.

Conclusions

Despite their heightened stresses, parents of children with FA can describe, reflect, and recommend strategies for dealing with waiting and treating a chronic genetic disease. Parents supplied rich images of their experiences stressing the lifelong, continuous, conscious thinking about FA where they are always waiting for the next shoe to drop. They articulated many medical stressors, positive coping methods, and a myriad of support systems. We found substantial variability and in some cases contradictory mechanisms of support. Specifically, there appears to be a fine balance between the need for information and delaying/limiting worry. Major parental recommendations focused on educating and partnering with the healthcare system to care for children with FA. Future studies may examine the implementation of family-centered strategies and focus on the experience of waiting in children and adults with FA.

Acknowledgements The authors want to sincerely thank the parents for their participation in the study and the insights they shared. We also want to thank our departments for supporting these research efforts.

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