Received: 10 April 2018 | Revised: 23 May 2018 | Accepted: 10 June 2018

DOI: 10.1002/pbc.27350

RESEARCH ARTICLE

WILEY Pediatric Blood & Cancer | SIOP INTERNATIONAL SOCIETY OF PAEDIATRIC ONCOLOGY | aspho The American Society of Pediatric Hematology/Oncology

Parent–child communication surrounding genetic testing for Li–Fraumeni syndrome: Living under the cloud of cancer

Jessica M. Valdez[1,2] | Breya Walker[3] | Susan Ogg[3] | Jami Gattuso[3] | Melissa A. Alderfer[4] | Kristin Zelley[5] | Carol A. Ford[6] | Justin N. Baker[7] | Belinda N. Mandrell[3] | Kim E. Nichols[1,2]

[1]Division of Cancer Predisposition, St. Jude Children's Research Hospital, Memphis, Tennessee

[2]Department of Oncology, St. Jude Children's Research Hospital, Memphis, Tennessee

[3]Division of Nursing Research, St. Jude Children's Research Hospital, Memphis, Tennessee

[4]Center for Healthcare Delivery Science, Nemours Children's Health System, Wilmington, Delaware

[5]Division of Oncology, The Children's Hospital of Philadelphia, Philadelphia, Philadelphia

[6]Division of Adolescent Medicine, The Children's Hospital of Philadelphia, Philadelphia, Philadelphia

[7]Division of Quality of Life and Palliative Care, St. Jude Children's Research Hospital, Memphis, Tennessee

Correspondence
Kim E. Nichols, Division of Cancer Predisposition, St. Jude Children's Research Hospital, Chili's Care Center I3311, MS1170, 262 Danny Thomas Place, Memphis, TN 38105.
Email: Kim.Nichols@stjude.org

Abstract

Background: Advances in the application of genetic technologies reveal a growing number of heritable disorders associated with an increased risk to develop cancer during childhood. As genetic testing is increasingly employed in the clinical setting, it is essential to understand whether parents communicate with their children about test results and to elucidate the factors that influence the content and outcomes of these conversations.

Methods: Semistructured interviews were conducted with 14 parents whose children tested positive for Li–Fraumeni syndrome (LFS). Semantic content analysis was performed on transcribed interviews, focusing on questions related to parent–child conversations about the genetic testing process and disclosure of positive test results.

Results: All parents emphasized the importance of involving children in conversations about LFS. The majority (93%) identified as being part of "cancer families" in which prior experiences with cancer created opportunities for communication. While all had spoken with their children about cancer, only seven (50%) specifically disclosed to their children that they had tested positive for LFS. The most common reason cited for nondisclosure at the time of this study was the young age of the children.

Conclusion: Parents of children with LFS desire open conversations about genetic testing and cancer risk. These conversations are challenging yet essential to enable child understanding of genetic risk status and enhance compliance with health-promoting and cancer surveillance measures. Development of age-appropriate educational materials and novel clinical models to facilitate parent–child conversations about genetic test results and risk status for cancer are needed.

KEYWORDS

cancer predisposition, communication, genetic testing, Li–Fraumeni syndrome, pediatric oncology

1 | INTRODUCTION

"Knowing this 'cloud of Li–Fraumeni' is over us has made things different… We don't put things off… We don't take each other for granted, and we tell each other that we love each other every single day. It made us live as a family unit a bit better."—Parent of child who tested positive for Li–Fraumeni syndrome.

The last three decades have witnessed the discovery of a growing number of heritable disorders associated with an increased risk of developing cancer during childhood.[1] Often, the optimal management of affected children relies upon clinical genetic testing followed by targeted surveillance for the early detection and treatment of tumors. However, for many conditions, uncertainty remains regarding if, when, and what type of cancer will occur as the result of the heritable predisposition. This uncertainty often leaves parents with the difficult decision of whether to have their children tested, and when to share genetic risk information with them.[2]

This uncertainty is well exemplified for families with Li–Fraumeni syndrome (LFS), a cancer-predisposing condition caused by germline

Abbreviations: CCM, constant comparative method; CHOP, Children's Hospital of Philadelphia; FAP, familial adenomatous polyposis; LFS, Li–Fraumeni syndrome

pathogenic variants in the *TP53* tumor suppressor gene. Over the course of their lives, most individuals with LFS will develop cancer, with 20% presenting with a malignancy before age 20.[3–5] Individuals with LFS are prone to develop a variety of cancers, including soft tissue and bone sarcomas, early-onset breast cancers, adrenocortical carcinomas, acute leukemias and brain tumors, and many develop multiple primary tumors.[4] Additionally, LFS follows an autosomal dominant inheritance pattern making understanding of carrier status an important consideration for family planning and additional familial testing.

Following the identification of pathogenic variants in the *TP53* gene as the cause of LFS, there was much debate about the clinical utility of genetic testing for this condition.[6,7] Many believed that there was little to be done to mitigate the negative outcomes from cancer, and thus, there was concern that testing would only increase stress and anxiety, straining family relations and potentially causing stigmatization of the individual who tested positive. However, subsequent studies have shown that the negative impacts of LFS testing (if any) are generally transient,[8,9] and new data reveal that adherence to specific tumor screening protocols can improve outcomes for those living with LFS.[10,11] Thus, attitudes have changed, and testing has now become commonplace, even for children.

Through a prior research study on parental decision-making around *TP53* testing for children, we observed high rates of parental acceptance of testing.[12] In a follow-up evaluation of adolescent understanding and attitudes surrounding such testing, we found that most adolescents and young adults believe that *TP53* testing should be offered to children as a way to learn about risk status, allow for disease prevention strategies, and reduce uncertainty and anxiety.[9] Together, these prior studies demonstrated that many parents prefer to have their children tested, and that most children support their parent's decision to be tested and learn about the test results. However, it remained unclear whether, when, and how parents speak with their children about the genetic testing process and the final LFS test results. By providing accurate information, parents can enhance child understanding and minimize misconceptions about cancer risk. They can also provide support during adolescence, a time of rapid and often tumultuous emotional and psychological development. Finally, parents can encourage simple but positive lifestyle changes aimed at lifelong preventative healthcare behaviors (such as use of sunscreen and avoidance of tobacco products and alcohol). To gain further insights into these poorly understood domains, we interviewed parents about their discussions with children regarding LFS genetic testing and cancer risk. We also evaluated the factors that influenced whether or not (and if so, when) parents decided to speak with their children and examined the barriers and facilitators to these conversations.

2 | MATERIALS AND METHODS

This study was approved by the institutional review board at The Children's Hospital of Philadelphia (CHOP) where data were collected. The analysis was deemed nonhuman research by the St. Jude Children's Research Hospital (SJCRH) institutional review board. This is a multiinstitutional study that recruited participants from CHOP (Philadelphia, PA), Sick Kids (Toronto, ON), Dana-Farber Cancer Institute (Boston, MA), Huntsman Cancer Institute (Salt Lake City, UT), National Cancer Institute (Bethesda, MD), Texas Children's Hospital (Houston, TX), and Stanford University (Stanford, CA).

Detailed methods regarding patient recruitment and interviews have been previously published.[12] Briefly, English-speaking families for which at least one child less than 22 years of age had been offered *TP53* genetic testing were recruited via letter.[12] This cut off was selected in order to include ages at which most centers accept patients into their pediatric oncology clinics and where parental involvement in health care is most likely. Families interested in participating were contacted by the study team via telephone or e-mail. Interviews were conducted individually for each parent, recorded, and transcribed.[12] Of 46 parent interviews from 39 families, 14 involved parents from 10 families for whom a child tested positive for LFS. We examined the transcripts of these 14 interviews with an emphasis on questions related to parent–child communication.

2.1 | Data analysis

Parental responses pertaining to interview questions gauged toward aspects of communication were extracted from the semistructured interview transcript. These pertinent sections of transcripts were agreed upon by three independent study team members with extensive experience in qualitative research methodology. Each response underwent inductive semantic content analysis[13–15] using MAXQDA software version 12.2.3 (MAXQDA, Berlin, Germany). The same three study team members utilized elements of grounded theory and the constant comparative method (CCM) for semantic content analysis. Insuch a manner, codes were established based on common themes that appeared throughout parental responses. Consensus for code assignment to parental responses was met based on CCM, as described above. Thus, 100% agreement among the three study team members was met for all codes assigned to parental responses during content analysis. Interrater reliability was maintained above 90% for all questions through consensus.

Code frequencies and demographic characteristics were summarized using descriptive statistics (mean, percentage). The frequency of each code was tallied. To compensate for multiple occurrences of a code in a single interview, the percentage of parents for whom the code appeared at least once was also tallied. For each question, multiple codes coincided or overlapped in meaning and were consolidated to arrive at final themes.[13]

3 | RESULTS

3.1 | Demographics

Analysis was completed on interviews with 14 parents (10 mothers and four fathers) from 10 families who had at least one child who tested positive for LFS (Figure 1). The 10 families included 20 children who were tested, of whom 17 were positive for a pathogenic variant in *TP53*. Three of the 20 children (15%) underwent diagnostic testing (i.e., these children had a cancer consistent with the LFS spectrum of

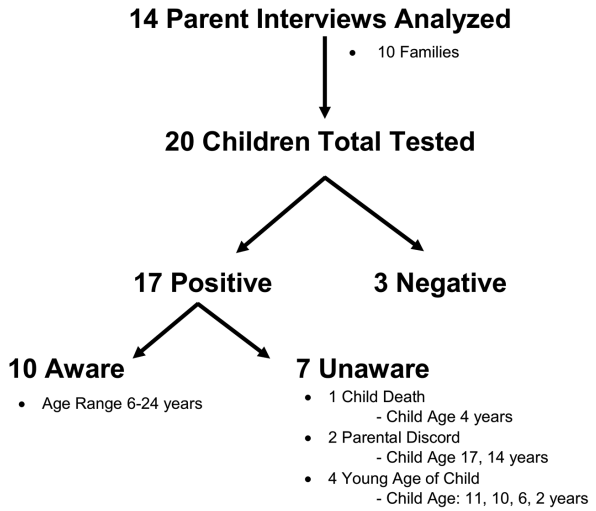

FIGURE 1 Schema depicting the parents interviewed and children tested for Li–Fraumeni syndrome

tumors), and 17 children (85%) underwent predictive testing (i.e., these children did not have cancer but were siblings or offspring of those known to have LFS; Table 1). Tested children ranged in age from newborn to 20 years at the time of testing (median age, 8 years). At the time of the parent interview, 10 of the 17 children (59%) who tested positive were aware of their LFS diagnosis (i.e., their parents had one or more conversations with them about the positive *TP53* result). Seven children (41%) remained unaware of their diagnosis (i.e., the parents had not disclosed their child's positive *TP53* result). Seven of the 14 parents (50%) interviewed were positive for LFS and six (43%; all known to have LFS) had a prior history of cancer.

3.2 | Content analysis

Analysis of code frequency endorsed by individual parents across Questions 1 and 2 revealed that the codes were evenly distributed among all parents interviewed and multiple parents endorsed each code. Tables 2 and 3 include a comprehensive summary of the themes that emerged. Selected themes, highlighting key quotations pertaining to each question, are shown below.

Question 1. Have you talked with your child(ren) about the results of genetic tests or genetic risks of cancer in your family?

Prompts: When and how do the conversations occur? What information do you include? How do the conversations go?

All 14 parent interviews were analyzed for responses to Question 1. All parents spoke in at least general terms about cancer with their child(ren). This communication was independent of the disclosure of the specific *TP53* test results, where only 10 of 17 children were made aware of this information (Figure 1). A total of 268 segments of text were coded and organized into 10 themes that are shown in Table 2.

3.3 | Selected themes from question 1 (N = 14)

All 14 parents endorsed the theme of *child involvement*: the importance of having children included in conversations about the LFS diagnosis, genetic testing, and management through surveillance. All parents had already had or planned to have these discussions with their children:

> *"We recognized that we needed to lay a solid foundation of open communication. The kids need to know that they can come to us with questions and concerns … building this foundation of trust and communication was important."*

While all parents endorsed the importance of *child involvement*, 10 of 14 parents (71%) indicated that discussions need to take place in *a developmentally appropriate manner.* Nine (64%) portrayed the conversations as *ongoing* and evolving as their children get older. Parents described discussing small portions of the subject matter as issues arose in their families or if the child asked. None of the parents endorsed sitting down and covering all information during one conversation.

TABLE 1 Demographic characteristics of parents (N = 14) and children (N = 20)

	No.	%	Range	Median
Characteristics of Adults (N = 14)				
Age			36–56	45
Sex of parent				
Female	10	71		
Male	4	29		
Race/ethnicity				
White	14	100		
Other	0	0		
Both parents of child interviewed	8	57		
Individual parent of child interviewed	6	43		
Parent positive for Li–Fraumeni syndrome	7	50		
Parent history of cancer	6	43		
Education				
Some college	2	14		
2-Year college	1	7		
Bachelor's degree	5	36		
Professional/graduate degree	6	43		
Characteristics of children (N = 20)				
Child age at time of testing			0–20	8
Child age at time of interview			2–24	12.1
Sex of child				
Female	16	80		
Male	4	20		
Child positive for Li–Fraumeni syndrome	17	85		
Child history of cancer	3	15		
Diagnostic testing	3	15		
Predictive testing	17	85		

TABLE 2 Themes generated from responses to question 1: "Have you talked with your child(ren) about the results of genetic tests or genetic risks of cancer in your family?"

Theme	Definition	Example quote	No. of parents using code N = 14 (%)	No. of text segments coded N = 268
Child involvement	Parent endorses open, honest, communication. Parents felt that they could make the best decision for the child but wanted to include the child as much as possible.	*We told them all about the testing. We actually let them help make the decision but encouraged them that it was the right thing to do.*	14 (100)	55
Cancer family	Parent describes family as "cancer family" living under the cloud of Li–Fraumeni. Communication occurs due to multiple diagnoses of cancer in immediate and distant family members.	*When my boys were about those preteen years; …they were asking a lot of questions. And so that's when we told them…we are a "cancer family"—it runs in the family.*	13 (93)	50
Being proactive	Parent feels it is important to try to prevent illness as much and as early as possible. Parent encourages child to communicate problems and make healthy decisions.	*We've taken the power in our own hands. We're going all the time to get you tested, to make sure as soon as the day comes as soon as it—we'll know and we'll do what we need to do.*	12 (86)	29
Emotional distress	Parent/child expresses negative emotions (depression/sadness/guilt) with communication/results.	*…the guilt, knowing that they got this from me and that this burden that they have to bear came from me.*	10 (71)	28
Developmentally appropriate	Parent presents information to child as is based on typically developing ages and stages.	*We've tried to be pretty open with the kids, though age appropriate, we don't want to scare them or tell them information that they really—we didn't think that they could handle.*	10 (71)	27
Parental responsibility	Parent expresses accountability for making tough decisions for child and/or providing information parent thinks child needs. This is a parental burden.	*It's like any other adult conversation you have to have with your kids, you know, I mean, they're all tough but you know, it's part of the job of being a parent.*	9 (64)	23
Ongoing communication	Parent describes communication as a continuous process and will communicate with child as situation requires.	*We've kinda made it an ongoing process so it wouldn't be a, you know, a shock at the end when they do get the information. So, we kinda tried to take a graduated approach where, you know, they get a little bit at a time.*	9 (64)	17
Knowledge is power	Parent/child feels having this information provides a better understanding of what they are dealing with may give them peace of mind and not knowing may be worse.	*They both understand really well what it is and why it is and how it's passed down through generations and what the ramifications are.*	8 (57)	18
Conversations went well	Parent felt the discussion/communication were successful or was not as difficult as they expected to be.	*We've approached it to their level; they seem to go pretty well. They would usually listen pretty intently and then every once in a while, they'd have a few questions and we also found out some of their anxieties through the process.*	8 (57)	12
Avoid communication	Parent/child coped by ignoring or just not thinking about it.	*My husband on the other hand wants to pretend it's not there. He's like, yeah, yeah, it's over with, yeah, well let's not talk about it.*	6 (43)	9

> *"(The conversations are) more age specific… we tried to take a graduated approach where, you know, they get a little bit at a time. I mean there was no way to deny that they knew that their family members were getting ill, so we had to deal with it that way."*

> *"I was going through my cancer treatment so I was bald and sick, they knew mom was going through cancer treatments, they knew that their aunt had just finished it, and my six-year-old asked me if I had caught cancer from his aunt, and if he was going to catch cancer from me… That's when we told them… yes, you know, we are a cancer family."*

The theme of being a *cancer family* was raised by 13 of 14 parents (93%) interviewed. Parents identified that having a family or personal experience with cancer created opportunities for conversations with their children about cancer risk or the diagnosis of LFS:

When prompted, eight of 14 parents (57%) stated that the conversations with their child *went well*, indicating they were able to effectively convey the intended information. They described the conversations as "comfortable," "straightforward," and "initially hard but

TABLE 3 Themes generated from responses to question 2: "If you have not disclosed the results of the genetic testing with your child what is preventing you from doing so?"

Theme	Definition	Example quote	No. of parents using code N = 7 (%)	No. of text segments coded N = 63
Disclosure	Parents involve child in general medical discussions and plan to include the child in future conversations regarding diagnosis and care.	*Right now, we left it open if he has any questions to ask, go right ahead. We also promote when he goes to Doctor, if at any point he has any questions to ask them.*	7 (100)	18
Age appropriate	Parent plans to communicate with child in the future based on the developmental age as to avoid increased burden at what they consider young age.	*Definitely when they get to be teenage, which is around the corner with my son… preparing them for their adult lives, when this is something they're gonna have to take care of.*	7 (100)	17
Future discussion	Parent feels child will understand more in the future and that will be the time to discuss details.	*Once they get to be close to adult, I think we're have to definitely discuss that they were tested, and they actually do have this mutation*	7 (100)	13
Parental distress	Parent expresses negative emotions (frustration/worry/dread) with situation.	*It's tough for the parents too and to try and negotiate and make sure you're on the same page with things… that's been a challenge for my husband and I.*	4 (57)	11
Avoid communication	Parent does not engage in communication about diagnosis or medical condition with the child due to personal preference or discomfort.	*Right now, I can say – she'll be like Dad, why do I go get my belly rubbed? Oh, because the doctor has to check you out. Oh my God, do you wanna go to Toys-R-Us? She's like okay…because I do not want to get into it with a six-year old.*	4 (57)	4

becoming easier over time." Six parents (43%) reported that the conversations were "hard" and described instances when they *avoided* them altogether. They cited reasons such as not being a family that is very open, fear of how the discussion might proceed, and one family stated, "because they (the parents) just didn't want to." Ten of 14 parents (71%) described having *emotional distress* or feeling overwhelmed when asked about having these conversations. Parents described waiting for the child to initiate the conversation or waiting for new treatments or recommendations regarding LFS as a reason to start the conversation. Parents expressed a lack of guidance or support about how best to initiate these conversations. Interestingly, of the nine interviewees who have disclosed results to at least one of their children, only three reported having a genetic counselor or a physician involved in the disclosure conversation.

Question 2. If you have not disclosed the results of the genetic testing with your child what is preventing you from doing so?

Prompts: Do you plan to disclose the results? When and how do you plan to have the conversation? Is there anything getting in the way of having that conversation?

The analysis of Question 2 was limited to seven parents with at least one child who was unaware of their positive *TP53* test result. Two of the interviewees, a married couple, had two children with LFS; however, only one of their children was aware of the diagnosis. The parents explained that the other child had not been told due to young age (the child was 6 years old at the time of the interview). These parents were included in the analysis of interview Question 2 because they had at least one child to whom they chose not to disclose the results. An additional parent whose child died prior to the interview was included in the analysis of Question 2 because the parent had not disclosed the test results to the child prior to the child's death and had not yet disclosed the results to the child's sibling (sibling was not yet tested at the time of the interview).

3.4 | Selected themes from question 2 (N = 7)

A total of 63 text segments were coded and organized into five separate themes (Table 3). The most common reason for nondisclosure was the child's age at the time of the interview (Figure 1). All seven parents who had not yet disclosed results reported that they would do so when their child was older and it was more *age appropriate*:

> *"The littler one, she's really not ready to discuss it. She's just too little, certain ages are just too young to really understand. But across the board, they understand that we have cancer in our family and it's something we have to be careful of."*

While these seven parents had different expectations of the *appropriate time* to have conversations, all endorsed the importance of *disclosure* and including their children in discussions related to health, medical care, and family illness. Furthermore, all seven are planning on having *future discussions* with their children regarding the LFS diagnosis, test result, and cancer risk. When asked about the timing of *future discussions*, parents did not describe an identifiable age but alluded to having discussions when the children started asking questions or seeking information.

> *"I don't know that I can say that there is an exact age. Every kid's different, the maturity level is different, so I think you got to weigh that. When they're ready to discuss it, or as a parent you're feeling there's a need to discuss it, then it should be addressed."*

Two of seven parents cited concern for social functioning and worried that disclosure of test results would negatively influence their child's adolescence and transition to college. Four parents (57%) endorsed *parental distress* and *avoidance of communication* when asked about the disclosure of test results. These parents reported not wanting to take away their children's "childhood" due to worry about having LFS and they wanted their children to be aware that the LFS diagnosis was not a "death sentence." They described delaying disclosure in order to keep things "normal" for as long as possible.

4 | DISCUSSION

In this study, we explored parent–child communication surrounding genetic testing for LFS. All 14 parents interviewed expressed a desire for open conversations with their children about LFS testing and associated cancer risks. Based on their own personal or family experiences with cancer, most parents described feeling better equipped to have these discussions. Furthermore, most parents wanted to have conversations with their children that were better than the ones that they had experienced themselves when they were young. Despite these intentions, only half of the parents had spoken directly with their children about the LFS test results, citing factors such as young child age, concern for social adjustment, and parental distress as reasons for nondisclosure.

5 | BENEFITS OF COMMUNICATION

Current research supports disclosure of genetic test results to family members.[16–18] The importance of passing this information to children so they can be empowered with their own health care knowledge has been supported by several studies examining adolescent involvement in cancer genetic testing.[9,19,20] Communication about genetic risk to pediatric patients and their families has been shown to have benefits that include improved medical management and health outcomes, decreased uncertainty, and increased ability to obtain healthcare and complete family planning.[21,22] Additionally, childhood is a vital time in which to discuss any possible interventions that may help protect against and prevent disease.[21] The importance of healthy behaviors should be communicated early and reinforced during vital periods such as adolescence and transition to adulthood. Currently, no guidelines or recommendations exist as to how or when parents should speak with their child(ren) about cancer genetic testing and heritable cancer risk. It is unclear at what age a child is best able to comprehend the complicated nature of having a predisposition to cancer; however, this likely represents a continuum that varies from child to child depending upon their intellectual and emotional maturity and the support systems available to them.

6 | PROCESSES OF COMMUNICATION

Conversations about genetic risk between family members are often unplanned, ongoing, and prompted by the child or a particular event related to the diagnosis in question.[23] In prior studies, children reported wanting to receive small amounts of information (with less formal conversations) during routine activities.[23] Our families echoed this sentiment and commented that discussions often occurred when driving to appointments for surveillance, working on a biology project for school, or when a family member was ill. Delaying conversations and worrying about the "best time" to have them can be a source of distress for parents.[23] Knowing that they do not need to have a long, sit-down and often uncomfortable talk may take the pressure off of parents. This may allow parents to have conversations that occur naturally, empowering them to look for opportunities to bring up the topic and foster an environment of open, ongoing communication.

Parents are the primary gatekeepers of medical knowledge and communication with their children.[24] This is particularly challenging when children are perceived to be too young to understand the implications of testing or participate in decisions about testing.[6,22,25–28] In this study, parents expressed a desire to hold conversations when they felt their children were "ready." Deciding upon the appropriate developmental stage at which to disclose vital health information is often left to parents and wrought with uncertainty.[24] When children receive information in a gradual fashion over time, information is often less shocking and easier to handle.[23] In the current study, parents also preferred ongoing conversations that were often prompted by the children. This approach allowed the child to drive the level of detail provided.

7 | CONCERNS

Parents often describe conversations about genetic risk with their children as difficult and emotionally painful.[23] In this study, the majority of parents noted that holding these discussions with their children was emotionally distressing with six of 14 (43%) indicating that they actively avoided conversations about LFS or cancer, often citing fear of not knowing what to say. Many parents expressed concern that the conversations would cause distress and worry in their children. However, systematic reviews of psychosocial distress regarding genetic testing and discussion with children for other inherited disorders such as Huntington's disease and familial adenomatous polyposis (FAP) indicate that children do not suffer clinical harms in terms of well-being, self-perception, and sociobehavioral functioning.[21,29,30] Additionally, studies have shown that children desire to be involved in discussions surrounding their medical care.[9,23,31] It is interesting to note that the parents among our cohort who felt the conversations "went well" described these conversations as an opportunity to address concerns directly while continuing to encourage future discussions.

8 | LIMITATIONS

The current study was limited by a small sample size and thus it was not possible to determine whether parent decisions to disclose test results were influenced based on a parent's diagnosis with cancer or LFS. The cohort was homogenous with no racial/ethnic variability. Therefore, cultural factors that could influence parent–child communications may

have been missed. The study was limited by the length of time from testing to interview period, which for some families was many years[12] and may have led to recall bias. Finally, this study was limited to a single cancer predisposition syndrome, namely, LFS. Despite this fact, it is reasonable to extrapolate these results to other genetic disorders and cancer-predisposing syndromes based on published literature on communications around Huntington's disease,[32] FAP,[20] and melanoma.[21,23]

9 | CONCLUSION

Here, we show that parents of children who tested positive for LFS express a strong desire to discuss the test results and their implications with their children. Nevertheless, these discussions can be challenging and a source of distress, particularly when parents have differing views regarding what information to communicate to each of their and when to have these conversations. It is important that medical professionals remain aware of these challenges and provide individualized support and guidance to patients and parents. Providers can gently remind parents that these discussions need not occur all at once. Rather, they can be carried out in an ongoing fashion, relaying smaller units of information. Parents should remain attentive to their children's comments and questions, which can serve as triggers for these conversations.

It is important that discussions occur during adolescence, when emerging adults struggle to establish independence and a personal identity, and are more likely to participate in risky behaviors.[33] To enable these discussions, we suggest a simple clinical model in which genetic counselors or providers familiar with cancer predisposition syndromes such as LFS re-engage with patients and their families prior to the transition to the adult medical setting. During this meeting, young adults can be reminded about their cancer risks and the approaches available to screen for or even to prevent any cancers from occurring. They can be educated about recurrence risks and discuss family planning options. They can receive guidance about how to navigate the medical system, including finding adult care providers and obtaining insurance. Finally, they can be encouraged to advocate for their own healthcare as they transition to independence.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

ACKNOWLEDGMENTS

This work was supported by funding from the Center for Childhood Cancer Research (Children's Hospital of Philadelphia), Center for the Integration of Genomic Healthcare Technologies (University of Pennsylvania Perelman School of Medicine), and the American Lebanese Syrian Associated Charities (SJCRH). We thank Emily Quinn, Lynn Harrison, and Kayla Hamilton for their critical review and helpful comments regarding this manuscript. Finally, we are grateful to our colleagues at the collaborating sites and the families who participated in this study. Without their support, this study would not have been possible.

ORCID

Jessica M. Valdez http://orcid.org/0000-0001-5480-3827

REFERENCES

1. Zhang J, Walsh MF, Wu G, et al. Germline mutations in predisposition genes in pediatric cancer. *N Engl J Med*. 2015;373:2336–2346.
2. Clarke A. What is at stake in the predictive genetic testing of children. *Fam Cancer*. 2010;9:19–22.
3. Strahm B, Malkin D. Hereditary cancer predisposition in children: genetic basis and clinical implications. *Int J Cancer*. 2006;119:2001–2006.
4. Valdez JM, Nichols KE, Kesserwan C. Li-Fraumeni syndrome: a paradigm for the understanding of hereditary cancer predisposition. *Br J Haematol*. 2017;176:539–552.
5. Bougeard G, Renaux-Petel M, Flaman JM, et al. Revisiting Li-Fraumeni syndrome from TP53 mutation carriers. *J Clin Oncol*. 2015;33:2345–2352.
6. Evans DG, Lunt P, Clancy T, Eeles R. Childhood predictive genetic testing for Li-Fraumeni syndrome. *Fam Cancer*. 2010;9:65–69.
7. Eng C, Hampel H, de la Chapelle A. Genetic testing for cancer predisposition. *Annu Rev Med*. 2001;52:371–400.
8. Lammens CR, Aaronson NK, Wagner A, et al. Genetic testing in Li-Fraumeni syndrome: uptake and psychosocial consequences. *J Clin Oncol*. 2010;28:3008–3014.
9. Alderfer MA, Lindell RB, Viadro CI, et al. Should genetic testing be offered for children? The perspectives of adolescents and emerging adults in families with Li-Fraumeni syndrome. *J Genet Counsel*. 2017;26:1106–1115.
10. Villani A, Shore A, Wasserman JD, et al. Biochemical and imaging surveillance in germline TP53 mutation carriers with Li-Fraumeni syndrome: 11 year follow-up of a prospective observational study. *Lancet Oncol*. 2016;17:1295–1305.
11. Villani A, Tabori U, Schiffman J, et al. Biochemical and imaging surveillance in germline TP53 mutation carriers with Li-Fraumeni syndrome: a prospective observational study. *Lancet Oncol*. 2011;12:559–567.
12. Alderfer MA, Zelley K, Lindell RB, et al. Parent decision-making around the genetic testing of children for germline TP53 mutations. *Cancer*. 2015;121:286–293.
13. Krippendorff K. Content analysis: An introduction to its methodology. Thousand Oaks, CA: Sage; 2012.
14. Krippendorff K. Content analysis: An introduction to its methodology. Thousand Oaks, CA: Sage; 2004.
15. Boeije H. A purposeful approach to the constant comparative method in the analysis of qualitative interviews. *Qual Quant*. 2002;36:391–409.
16. Forrest LE, Delatycki MB, Skene L, Aitken M. Communicating genetic information in families—a review of guidelines and position papers. *Eur J Hum Genet*. 2007;15:612–618.
17. Graves KD, Sinicrope PS, Esplen MJ, et al. Communication of genetic test results to family and health care providers following disclosure of research results. *Genet Med*. 2014;16:294–301.
18. Breitkopf CR, Petersen GM, Wolf SM, et al. Preferences regarding return of genomic results to relatives of research participants, including after participant death: empirical results from a cancer biobank. *J Law Med Ethics*. 2015;43:464–475.
19. Harel A, Abuelo D, Kazura A. Adolescents and genetic testing: what do they think about it. *J Adolesc Health*. 2003;33:489–494.

20. Peterson SK, Watts BG, Koehly LM, et al. How families communicate about HNPCC genetic testing: findings from a qualitative study. *Am J Med Genet C*. 2003;119c:78–86.
21. Wu YP, Mays D, Kohlmann W, Tercyak KP. Pediatric predispositional genetic risk communication: potential utility for prevention and control of melanoma risk as an exemplar. *J Genet Counsel*. 2017;26:887–893.
22. Ross LF, Saal HM, David KL, Anderson RR. Technical report: ethical and policy issues in genetic testing and screening of children. *Genet Med*. 2013;15:234–245.
23. Metcalfe A, Plumridge G, Coad J, Shanks A, Gill P. Parents' and children's communication about genetic risk: a qualitative study, learning from families' experiences. *Eur J Hum Genet*. 2011;19:640–646.
24. Rafferty KA, Hutton K, Heller S. I will communicate with you, but let me be in control": understanding how parents manage private information about their chronically ill children. *Health Commun*. 2017:1–10. http://doi.org/10.1080/10410236.2017.1384432
25. Clayton EW, McCullough LB, Biesecker LG, Joffe S, Ross LF, Wolf SM. Addressing the ethical challenges in genetic testing and sequencing of children. *Am J Bioethics*. 2014;14:3–9.
26. Johnson LM, Hamilton KV, Valdez JM, Knapp E, Baker JN, Nichols KE. Ethical considerations surrounding germline next-generation sequencing of children with cancer. *Expert Rev Mol Diagn*. 2017;17:523–534.
27. Botkin JR, Belmont JW, Berg JS, et al. Points to consider: ethical, legal, and psychosocial implications of genetic testing in children and adolescents. *Am J Hum Genet*. 2015;97:6–21.
28. Hens K, Nys H, Cassiman J-J, Dierickx K. The return of individual research findings in paediatric genetic research. *J Med Ethics*. 2011;37:179–183.
29. Wade CH, Wilfond BS, McBride CM. Effects of genetic risk information on children's psychosocial wellbeing: a systematic review of the literature. *Genet Med*. 2010;12:317–326.
30. Wakefield CE, Hanlon LV, Tucker KM, et al. The psychological impact of genetic information on children: a systematic review. *Genet Med*. 2016;18:755–762.
31. Metcalfe A, Coad J, Plumridge GM, Gill P, Farndon P. Family communication between children and their parents about inherited genetic conditions: a meta-synthesis of the research. *Eur J Hum Genet*. 2008;16:1193–1200.
32. Hartelius L, Jonsson M, Rickeberg A, Laakso K. Communication and Huntington's disease: qualitative interviews and focus groups with persons with Huntington's disease, family members, and carers. *Int J Lang Commun Disord*. 2010;45:381–393.
33. Fanos JH. Developmental tasks of childhood and adolescence: implications for genetic testing. *Am J Med Genet*. 1997;71:22–28.

How to cite this article: Valdez JM, Walker B, Ogg O, et al. Parent–child communication surrounding genetic testing for Li–Fraumeni syndrome: Living under the cloud of cancer. *Pediatr Blood Cancer*. 2018;e27350. https://doi.org/10.1002/pbc.27350