Published 2007 Wiley-Liss, Inc. American Journal of Medical Genetics Part A 143A:2089–2097 (2007)
†This article is a US Government work and, as such, is in the public domain in the United States of America.

Parenting Children With Proteus Syndrome: Experiences With, and Adaptation to, Courtesy Stigma

Joyce Turner,[1]* Barbara Biesecker,[2,3] Jennifer Leib,[4,6] Leslie Biesecker,[1] and Kathryn F. Peters[3,5]

[1]National Human Genome Research Institute, Genetic Disease Research Branch, Bethesda, Maryland
[2]National Human Genome Research Institute, Social Behavioral Branch, Bethesda, Maryland
[3]National Human Genome Research Institute, Medical Genetics Branch, Bethesda, Maryland
[4]National Institute for Neurological Disorders and Stroke, Neurogenetics Branch, Bethesda, Maryland
[5]The Pennsylvania State University, Center for Developmental and Health Genetics, University Park, Pennsylvania
[6]Current affiliation is Affymetrix Incorporated

Received 29 September 2006; Accepted 22 May 2007

Courtesy stigma refers to the stigmatization an unaffected person experiences due to his or her relationship with a person who bears a stigma. Parents of children with genetic conditions are particularly vulnerable to courtesy stigma, but little research has been done to explore this phenomenon. The purpose of this study was to investigate the courtesy stigma experiences of parents of children with Proteus syndrome (PS) and related overgrowth conditions. Thematic analysis of transcripts from 31 parents identified three distinct themes: stigma experiences, social-emotional reactions to stigmatizing encounters, and coping responses. Four types of stigmatizing experiences were identified: intrusive inquires, staring and pointing, devaluing remarks, and social withdrawal. Additionally, we uncovered eight strategies parents used to cope with courtesy stigma: attributing cause, assigning meaning to social exchanges, concealing, withdrawing socially, taking the offensive, employing indifference, instructing and learning from family, and educating others. Parents' choices of strategy type were found to be context dependent and evolved over time. This is the first study to document the adaptive evolution of coping strategies to offset courtesy stigma by parents of children with genetic conditions. These results provide groundwork for genetic counseling interventions aimed at addressing issues of courtesy stigma and further investigation of the phenomenon itself. Published 2007 Wiley-Liss, Inc.†

Key words: courtesy stigma; stigma; coping; Proteus syndrome; parents; adaptation; psychosocial; genetic counseling

How to cite this article: Turner J, Biesecker B, Leib J, Biesecker L, Peters KF. 2007. Parenting children with Proteus syndrome: Experiences with and adaptation to courtesy stigma. Am J Med Genet Part A 143A:2089–2097.

INTRODUCTION

Stigmatization is a complex, social phenomenon, and since the 1960s a number of models have been developed to describe the stigmatization process and related outcomes. Common to these models are two beliefs. For stigmatization to occur an attribute must (1) be perceived to negatively stray (deviate) from socially construed norms and expectations and (2) evoke undesirable (negative) responses by the perceiver [Goffman, 1963; Jones et al., 1984; Susman, 1994; Dovidio et al., 2000; Link and Phelan, 2001]. It is important to note that perceptions and responses vary, as they are dependent upon individual/group views (e.g., culture), needs, values, and objectives, as well as the time and place in which they occur [Goffman, 1963; Jones et al., 1984; Groce and Zola, 1993; Susman, 1994; Dovidio et al., 2000; Link and Phelan, 2001]. Additionally, perceptions shape individuals' affective (emotional), cognitive, and behavioral responses to stigmatization. This holds true for those who stigmatize and for those who are the targets of stigmatization [Dovidio et al., 2000].

Courtesy stigma is the stigmatization an unaffected person faces due to his/her association with a person who bears a stigma [Goffman, 1963]. Dealing with courtesy stigma can be difficult. One can disengage

Grant sponsor: Intramural Research Program of the National Human Genome Research Institute, National Institutes of Health.

*Correspondence to: Joyce Turner, MS, NIH/NHGRI, 10 Center Dr., Bldg 10/3C710, Bethesda, MD 20892-1253. E-mail: jturner1@mail.nih.gov

DOI 10.1002/ajmg.a.31904

WILEY InterScience® DISCOVER SOMETHING GREAT

socially from the stigmatized person. However, for parents of a child with a stigmatizing condition, dissociation is not practical, socially permissible, or desired. Consequently, parents of children with stigmatizing genetic conditions must develop strategies to cope with courtesy stigma.

Few studies have been done to understand this phenomenon and to provide coping strategies for families, particularly among parents of children with visible differences. Chapple et al. [1995] and Kessler et al. [1984] found that parents may harbor feelings of guilt and shame secondary to their social experiences with having a child with a genetic condition. Mothers of children with mental retardation have reported weakened friendships, severed relations, and condescending remarks from friends and neighbors [Birenbaum, 1970]. The social awareness of an affected child's speech, aberrant behavior, and behavioral management difficulties have been associated with parental stress [Baxter, 1989]. This resulted from others displaying discomfort, drawing other's attention, and staring. Mothers of children with chronic disabilities have described being repeatedly asked intrusive questions and blamed for their child's disabilities [Green, 2003]. The mothers perceived their affected children to be devalued and discriminated against, which contributed to their emotional distress. The mothers also described tactics they used to minimize stigmatizing encounters. These included educating others, saying something positive about their child to make others feel comfortable around their child, and using humor to counter offensive questions.

In 1994, a natural history study and molecular investigation into the cause of Proteus syndrome (PS) began at the National Human Genome Research Institute (NHGRI). PS is a variable, progressively disfiguring condition. It is characterized by asymmetric and disproportionate bony and soft tissue overgrowth, skin lesions, and vascular malformations [Cohen and Hayden, 1979; Wiedemann et al., 1983; Turner et al., 2004]. The condition is thought to be genetic but not inherited, and the cause is unknown [Happle, 1987; Barker et al., 2001; Biesecker et al., 2001; Cohen et al., 2002, 2003]. The diagnosis of PS is based on clinical presentation, and the physical features of the condition overlap with other overgrowth conditions [Biesecker et al., 1998; Turner et al., 2004]. The management of PS is complex and surgical intervention outcomes can be cosmetically unsatisfactory [Biesecker, 2005].

After enrolling the first few families into our study, it became clear that concerns related to social adjustment and stigmatization were particularly prominent. We noted that although parenting a child with PS shares many of the psychosocial challenges of parenting children with other congenital syndromes, the severity of PS and its progressive nature caused significant psychosocial burden and stigmatization for some parents. Thus, the goal of the current study was to investigate how courtesy stigma affects parents of a child with a visibly apparent, progressive, genetic disorder such as PS. More specifically, we conducted a qualitative study in which we sought to understand ways that parents encounter and respond to courtesy stigma. The results of this study suggest that there are added psychological and social dimensions faced by parents of children with visible differences. The results provide groundwork required to develop counseling interventions for those affected by courtesy stigma.

MATERIALS AND METHODS

Study Population and Recruitment

This study was conducted at the National Institutes of Health (NIH) and was approved by an Institutional Review Board. Parents were eligible to participate if their child was believed to have PS. The study population included 31 parents from 20 families: 29 parents of children who were evaluated at the Warren Grant Magnuson Clinical Center at the NIH, and two parents of a deceased, affected child. Four parents of three children were told prior to their interview that their child was affected with another condition. The affected children of all of the other parents carried the diagnosis of PS at the time of the interview.

Data Collection

K.F. Peters conducted 25 in-person and six telephone, semi-structured interviews designed to elicit parents' experiences with stigmatization. Previously reported dimensions of stigma [Jones et al., 1984] served as the theoretical basis for the interview guide. Each interview lasted about 60 min. In families in which both parents participated, interviews were done separately but in immediate succession to avoid cross talk. Parents were also asked to rate the severity of their child's condition on a scale of 0 (unaffected) to 10 (severely affected).

Data Analysis

Interviews were audiotaped and transcribed. A codebook was collectively drafted by four investigators (K.F. Peters, J.T. Turner, J.R. Leib, and B.B. Biesecker). Coding of six transcripts was done in duplicate by three investigators to check for coding inconsistencies and to validate the coding schemata. The inter-coder reliability was 84%. Coding discrepancies were rectified and adjusted accordingly. Data were entered into QSR NUD*IST Rev 4.0 (Qualitative Solutions and Research Pty Ltd., Sage Publications, Inc., Thousand Oaks, CA) a qualitative analysis software package. Because of the limited number of participants, all data were

analyzed collectively for prevalent themes with no within or between couple, gender, or overgrowth condition type comparisons.

RESULTS

Demographics

All 31 parents invited to participate in the interview study elected to participate. Table IA reports the demographic characteristics of the parent-participants. Their average age was 36.9 years (Median = 37 years). Table IB describes characteristics of the parents' affected children, including the parents' perceptions of the severity of their child's condition.

TABLE IA. Characteristics of Parents N = 31

Participants	n (%[a])
Females	19 (61)
Age range (years)	22–55
<32	6 (19)
32–35	7 (23)
36–40	7 (23)
41–55	11(35)
Ethnicity	
Caucasian	29 (93)
Other	2 (7)
Education	
Graduate degree	2 (7)
College diploma	14 (45)
Some college/ Technical school	9 (29)
≤High school diploma	6 (19)
Employment	
Full time	18 (58)
Part time	5 (16)
Not employed	7 (23)
Retired	1 (3)
Marital status	
Married	27 (87)
Divorced	2 (7)
Single	1 (3)
Widow	1 (3)

n, Number of parents.
[a]Rounded to the nearest percent.

Stigma Experiences

Thirty (97%) parents reported at least one personally stigmatizing experience related to their affected child. Analysis revealed four types of stigmatizing experiences with varying frequencies (Table II). The following is a description of each:

Intrusive inquiries. Parents readily reported that their child's appearance prompted inquiries when out in public: "Every time we go anywhere... [people ask] what's wrong with your boy...You're stopped and talk to total strangers about...your kid...And they just keep bringing it up over and over...again (F3[1])."

Staring and pointing. Parents described staring and pointing as common reactions by strangers to their child because of his or her appearance. One parent estimated that she observed others staring 75–80% of the time she is out in public. Another parent concurred that staring occurs on a daily basis. Although staring was usually aimed at their child, parents stated that staring affected them personally.

Devaluing remarks. Parents reported that their child was often the object of others' insensitive remarks. They also said that they were not immune to this. Rather, 15 parents (48%) provided 16 examples of devaluing remarks that were directed at them. Collectively, these remarks fit into four categories. The first category includes remarks centered on the parents' decisions regarding their child's medical care:

> One sister felt very strongly we should have [my son's foot] corrected, and we were leaning against having any kind of [cosmetic] surgery, and all the doctors we were talking to were...saying don't put him through the surgery...she very strongly felt that we should have corrected his foot...She felt like we were being...negligent...to not make his foot look really nice.... (M4)

The second category suggested that parents should feel shameful about their child's appearance. This includes comments made to parents encouraging them to remove or conceal their child's affected body part(s) from public view: "I had nurses coming in bringing me awful pictures of...deformed children and telling me that I could home school him... (M6)."

The third category of devaluing comments includes those that suggested that the parents

TABLE IB. Characteristics of Children N = 20

Children	n (%[a])
Females	13 (65)
Age range (years)	10 months to 26 years
<1	1 (5)
1–6	9 (45)
7–11	9 (45)
>11	1 (5)
Ethnicity	
Caucasian	18 (90)
Other	2 (10)
Age at diagnosis (years.)	
≤1	8 (40)
>1≤5	9 (45)
>5≤10	2 (10)
>10	1 (5)
Severity of appearance as reported by parents**	
Average	4.5
Mode and median	4.0
Number of children with siblings	17 (85)

n, Number of parents.
[a]Rounded to the nearest percent.
**Based on a severity scale from zero unaffected to 10 (severely affected).

[1]*F/*M—father or mother of an X-year-old child.

TABLE II. Four Types of Stigma Experiences

Type	n (%[a])
Intrusive inquiries	25 (81)
Staring and pointing	21 (68)
Devaluing remarks	21 (68)
Social withdrawal	6 (19)

n, Number of parents
[a]Rounded to the nearest percent.

were responsible for having an affected child. For example, parents reported being told that their child's condition was their fault, "God's punishment (F5)," or as a result of something that the parent should have known to avoid, such as "too much anesthetic gas. . . (M10)."

The fourth category of comments suggested that the parents were special people. For example, "[My husband's] brother said, 'I don't know how you guys do it. I could never do it. I'd rather be dead (M6)." Although parents recognized that the intention of such a comment was to praise the parent, it often made the parent feel singled out.

Social withdrawal of family and friends. Some parents said that they no longer had the same relationships with family and friends. The sign of the demise of their relationships was the distancing from or failure to include the family in social gatherings:

> I don't see my parents very often. They hardly ever come see the kids. . .A great example, . . .[they] completely forgot their birthdays. Granted they got invitations, but they forgot the date. . .I only talk to them. . .once a month on the phone. And they live [near us]. (M3)

Social-Emotional Responses to Stigmatizing Encounters

Overall, seven (23%) parents described experiencing persistent worry that their child would be treated differently by others.

Responses to intrusive inquires. Most parents (n = 23, 74%) stated that they answered the questions that were posed to them in public about their child. However, 15 parents (48%) indicated that at times they felt less willing to discuss their child's condition than at other times. This ambivalence was reported to stem largely from the parents' own difficulty in concisely and clearly explaining their child's condition:

> When you try to explain what it is nobody has ever heard of it. The only thing they know of is the Elephant Man. . .Then you say he has the same thing the Elephant Man had. . .I just don't know really how to explain what he has. (M1.5*)

Additionally, parents questioned the motives for queries, and were more open to those deemed genuine:

> You can tell when people are. . .genuinely asking and when they're just asking out of courtesy. When. . .you. . .get the feeling that they were truly interested, then it was okay. . .If somebody says, "Oh, just tell me about that" and. . .they would look at their watch. . .you just don't want to do it. (F5)

Some parents said that their mood and their child's current health status influenced their willingness to answer questions. Parents reported that over time they grew tired of responding to queries. As a result, they created abbreviated responses: "We. . .get tired of being asked [about it]. 'What is that. . .[Is] that a burn?' It's. . .more convenient not to have to stop and answer everybody's questions. . .We. . .programmed ourselves to say, 'No. It's. . .a birthmark (M7).'"

Parents rarely became overly upset or angered by queries themselves. Rather, they reported that they became incensed by the insensitive or judgmental manner in which the queries were phrased. Parents reported that others' questioning negatively affected them more readily when they had just begun the process of adjusting to their child's diagnosis. Four parents reported their perspective on answering queries changed over time: "I did not like when people would come up and say, 'What is wrong?'. . .It doesn't bother me now, but I did not like it before. . .I was trying to deal with it myself, and it made it harder (M7)."

A third of the parents (29%) reported that they preferred people asking questions to other devaluing behaviors: "I don't like it when people stare. I would [rather] they. . .ask me a question. It is usually that kids come up and ask the questions. . .[not] adults. . .(M2)."

Responses to staring. Eight parents (26%) stated that when others stared at their child, it affected them (the parents) negatively. Parents reported that they became upset, angered and even outraged by others' stares: "I think. . .everyone looks. It's one thing to look. Don't point and gawk. . .If somebody points and gawks. . .that's going to be it. . .[I will] punch them right in the face. . .It's just respect. Just respect *me and my family* (F1)."

Among those angered by stares, two described how their emotional responses changed over time: "You get lots of funny looks. . .Initially. . .it hurt, but now it doesn't. It doesn't faze me (M 3)." Lastly, four parents (13%) said that staring did not bother them.

Responses to devaluing remarks. Almost half the parents (52%) reported becoming angered when they or their child were the objects of devaluing comments.

American Journal of Medical Genetics Part A: DOI 10.1002/ajmg.a

> When somebody would say, "...This is God's punishment"...just one stupid statement like that to think that God would do this to my son as a punishment...if that's the case, then there is no God because we believe God is a just God. And to think that he would use this as a punishment was just, oh, boy.... (F5)

Four parents stated that they adapted to others' insensitive comments, "If someone makes a rude comment...some people are very insensitive...But I learned how to overlook people...It's not really something that you learn. It's just something that you do (M7)."

Responses to social withdrawal. Parents described disappointment and anger in response to the social withdrawal of family and friends: "'Oh, we had a party in October; forgot to invite you.' Yeah, right. Does it look like 'stupid' on the front of my head...? 'You [did] it on purpose because she's not perfect (M6).'" None of the parents reported that their response to this type of social stigmatization had changed over time.

Coping Responses to Stigmatizing Encounters

Parents reported eight distinct coping strategies in response to and/or in anticipation of stigmatizing encounters. The frequencies of these experiences are listed in Table III. These strategies were primarily cognitive responses (attributing cause, assigning meaning, employing indifference), or behavioral responses (concealing, withdrawing, taking the offensive, instructing family, educating others). The following summarizes each:

Attributing cause. Parents stated that they mulled over possible causes of their child's condition and primarily blamed themselves for their child's differences. Nine parents described how beliefs about the origin of their child's condition had changed over time: "I was...concerned that...I had caused [it]...But it is getting to the point where I [feel] like I wasn't negligent...I don't feel that guilt over it anymore (M10.5)."

TABLE III. Eight Types of Coping Responses

Strategy	n (%[a,b])
Attributing cause	
Blame self	17 (57)
Genetics	11 (36)
Do not know	10 (32)
Environment	6 (19)
Chance	4 (13)
Inheritance	1 (3)
God	1 (3)
Stress	1 (3)
Assigning meaning to social exchanges	15 (48)
Employing indifference	9 (29)
Concealing	8 (26)
Withdrawing socially	7 (23)
Taking the offensive	10 (33)
Instructing and learning from family	10 (32)
Educating others	11 (36)

n, Number of parents.
[a]Percentages do not add up to 100%, as parents may endorse more than one strategy.
[b]Rounded to the nearest percent.

Assigning meaning to social exchanges. Parents reported assigning malevolent meaning to others' looks, questions, comments, and perceived thoughts. In reviewing these examples collectively, several themes arose. The first theme includes those parents who reported feeling that their child's differences lead others to question their ability to properly care for their child: "When he was in casts...people [asked] me, 'How did he break his legs?'...implying I was the bad mother, that I might have dropped him...It was more or less...'What did you do to him... (M1.5).'"

The second theme that arose includes those parents who expressed feeling insecure since having a child with PS. Yet this insecurity was not static:

> I was born with normal features...and everybody I knew was...at 26 years old all of a sudden people [were] looking at me with different eyes than they used to...It was hard, but...now...I look at them, and I don't even pay attention.... (M6)

The third theme parents expressed was that they perceived that others pitied them: "There is a pity [factor]...people spend time with you and your son and enjoy it but then go back and think, 'Oh, God, thank God it is them and not me...(M3).'"

The fourth theme includes parents who reported that they perceived that others blamed them for the cause of the child's condition: "You talk to people...[and] they say, '...Was there something you did...?' They were trying to help you figure it out. And you wondered...if they were...accusing you or if they were...trying to help (M5)."

Employing indifference. Some parents reported coping with courtesy stigma by adopting an indifferent attitude. These parents stated that they "don't care," "overlook," or "do not concern themselves with how others think about them." One parent clarified that this attitude was adopted over time:

> I think strangers [looked at me differently because of my son's appearance]...And that's something I have [had] to work on...[I used to worry] about what people thought of me... I learned that what people think of me is not as [important] as it used to be.... (M5)

Concealing. Parents described hiding affected parts of their child to prevent having aberrant interactions with others: "I was embarrassed because

my child wasn't....normal...[I] put big socks on her...[carried] her with her foot behind me...put my hand over her foot...(M7)." Other parents elected to cover their child's affected areas inconspicuously. For example, they had their child wear a t-shirt when swimming. Three of these parents described how their hiding behaviors changed with time:

> When she was [first] born I hid her...I was embarrassed because my child wasn't...normal...I don't know what hit me, but it was like, "What are you doing?"...I had a talk with myself...it was like if you are going to teach this child to be strong, you have got to be strong, and so I just started doing it, and it was hard. (M7)

Seventeen parents (55%) reported that they did not try to conceal their child's differences. Eight of these felt that concealment was not appropriate and four did not try because the overgrowth was too extensive.

Withdrawing socially. Although 42% of the parents reported that their child's condition does not limit them socially, some offered that they had withdrawn socially to avoid negative reactions and intrusive questions. Two parents shared that their views regarding social withdrawal changed over time:

> I wouldn't want to deal with the stares...and people just bugging me I wouldn't want to take him to the store...it is just hard.... You get all tense inside...I am pretty much over that...I find that I don't...think about what people are thinking anymore. (M6)

Taking the offensive. Parents described becoming angry and taking the offensive in response to stigmatizing encounters. This included staring back, responding with sarcasm, reprimanding and intentionally revealing their child's disfigurement to shock the offender. Parents shared that they did not initially, routinely, or indefinitely use these tactics. As one mother said, "I was at the [playground]...and this little girl was staring at him...I told her, 'This is what happens when you lie to your parents'...but that was when I was younger...(M6)." Other parents tried to calmly respond to devaluing encounters with an explanation: "Sometimes, I'd be very calm and explain to them that God would not do this. He's not punishing [one] human being to punish another...maybe not too politely (F5)."

Instructing and learning from family. A number of parents reported that they talked with their children about stigma related issues, yet they also learned from and adopted their child's coping tactics. Six parents (19%) noted that they talked openly with their children about looking and being different. Although, eight parents (26%) said that they felt the need to teach their children how to respond appropriately to stigmatizing encounters, three parents (10%) also learned from their children how to better cope with courtesy stigma: "I don't...think about what people are thinking anymore...I started doing that by watching [my son]...he didn't care what people thought...I just started watching [him] and I...started doing what he did (M6)."

Educating others. For some parents, responding to inquiries was disruptive, but for others, these conversations served as a chance for parents to talk about their child and to educate others. This helped reduce stigmatization and prevent malicious assumptions:

> [One person asked], "What's happened to his hands, has he broken his fingers...?" "No, he has PS." "Oh, what's that?" I was sitting there talking to her about it...It was good... to explain what was wrong...Some people might think he's been beaten because of his hands...I like to...explain what's wrong...rather than them...look and guess. (F3)

Parents provided 15 examples in which they took the initiative to educate others about their child's condition. These parents explained that openly discussing their child's condition enabled them to prepare or instruct others about their child's needs. Moreover, it warded off stigmatization and promoted inclusion of themselves and their affected child, "After I explain he has this condition, I can...see a difference...Most people, like at church, they forget...that he has anything wrong with him. That's kind of nice. We need that (M1.5)."

DISCUSSION

Investigation into stigmatization of persons with genetic conditions is an area of growing research interest [Lapham et al., 1996; Rothenberg and Terry, 2002; Shinaman et al., 2003; Apse et al., 2004; Peters et al., 2005; Sankar et al., 2006]. Although affected persons are the primary victims of social stigmatization, their relatives are at risk for experiencing courtesy stigma. The goal of this study was to explore how courtesy stigma affects parents of children with PS. The results demonstrate that parents of children with PS are subject to courtesy stigma in a variety of ways and they employ a number of strategies to cope with stigmatization. Importantly, the results show an evolution in the strategies that parents adopt and their emotional responses to such encounters. Finally, these results show that both mothers and fathers can positively adapt to their roles as parents of children with PS.

Courtesy Stigma Experienced

In this study, all but one parent reported having had experienced a devaluing social encounter secondary to having a child with PS. Thematic analysis identified three distinct themes: stigma experiences, social-emotional reactions to stigmatizing encounters, and coping responses. More specifically, parents encountered four types of stigma experiences: intrusive inquiries, staring and pointing, devaluing remarks, and the social withdrawal of family and friends. Although parents reported that they were most commonly confronted with intrusive inquiries, parents also reported that this type of stigma experience was the least emotionally burdensome. Parents reported growing tired of discussing their child's condition or at times were reluctant to talk about it. However, parents also reported that they actually appreciated genuine queries from others. One explanation for this finding may be that while parents interpret others' questions as an attempt to satisfy the questioner's curiosity, in other cases parents view inquiries as an expression of concern and sympathy. Parents may feel empowered and supported when others take a genuine interest in their child. Moreover, parents may feel less pressured and more in control when given an authentic opportunity to openly discuss their child's condition. The act of telling one's story and providing an explanation to others can serve to relieve stress and aid in the parental coping process [Baxter, 1991; Blum, 1991; Peters et al., 2005]. Thus, being asked and responding to sincere inquiries may assist parents in their own emotional adjustment.

On the other hand, the other three types of stigma experiences (staring and pointing, devaluing remarks, and withdrawal of family and friends) were reported to be hurtful for parents. We hypothesize that in contrast to intrusive inquiries, parents have limited liberty to deflect or diffuse the devaluing experiences of being stared at, the subject of discrediting remarks, and/or intentionally excluded from social gatherings. A sense of being in control is central to the process of adaptation and adjustment to social and health threats, particularly for persons adjusting to genetic conditions [Berkenstadt et al., 1999]. Because these three types of stigmatizing experiences do not inherently include opportunities for parents to respond to, and regain control of, their situation, parents may view these types of encounters as the most compromising. These encounters may be viewed as stressful and threatening to parents, as parents may not initially have a repertoire of coping strategies immediately at hand to mitigate the discomfort and frustration of stigmatizing encounters [Miller and Major, 2000]. Further research is needed to better understand the social outcomes of the differing types of stigmatizing encounters and the emotional implications each have for parents of children with genetic conditions. Research into effective strategies to address these psychosocial issues is also necessary in order to provide optimal care for families.

Taking an Active Role in Coping and Adaptation

The results of early studies on stigma coping posited that people who are targets of stigma are passive and unable to control their experience [for a review, see Siegel et al., 1998] However, the results from the present study are consistent with the results of more recent studies of stigma experiences [Siegel et al., 1998; Swim et al., 1998; Dovidio et al., 2000], which report that stigmatized individuals can be active participants in the social exchange, rather than victims. In this active role, stigmatized individuals may cope with discreditors or potential discreditors by choosing where, when, with whom and how they socialize. This finding is demonstrated in the present study by the fact that parents reported using eight distinct coping strategies to minimize stigma and delineated variables that influence their strategy choice. These include the status of their relationship with the discreditor, context and frequency of the situation, and the parent's own personality or personal code of ethics.

The coping strategies that stigmatized individuals enlist have been described as ranging from reactive to proactive [Siegel et al., 1998]. The data from the present study provide further evidence for this. That is, *reactive strategies* include those that aim to avoid or lessen the effects of stigmatization without challenging the norms or values that support such acts. Examples of such strategies in our study include withdrawing socially, concealing, attributing cause, and assigning meaning to social exchanges. *Proactive strategies* on the other extreme attempt to reject and confront the social norms and values, which are the foundation for stigmatization, by taking action to curb or prevent such occurrences. Proactive strategies seek greater empowerment for the stigmatized individual. Examples of such strategies in the current study include instructing and learning from family members and educating others. *Intermediate strategies* include tactics with both proactive and reactive characteristics. These types of strategies reject the source of stigma but do not necessarily provide discreditors with something constructive. In this study, employing indifference and taking the offensive are examples of intermediate strategies. This classification scheme provides a framework for clinicians to think about how their clients are coping and adapting to social stigma and provides health professionals with insight into other avenues they can explore with their clients.

There is an abundance of data describing the ways in which stigmatized individuals cope with stigma

[Link et al., 1989, 2002, 2004; Siegel et al., 1998; Swim et al., 1998; Miller and Major, 2000; Puhl and Brownell, 2003]. Yet there is a paucity of data describing how individuals' perceptions of and emotional reactions to stigmatizing encounters, and their strategies used to cope with stigma, *change* over time. Clearly, coping with stigma is not a static process. Our results provide evidence that coping with courtesy stigma is an evolving process. For example, parents described how their emotional reactions to stigmatizing encounters had changed from feeling upset and angry to being comfortable with and not bothered by others' reactions. Parents' responses to stigmatizing encounters had evolved from blaming oneself and being shameful to having a more open and self-confident attitude. Further, parental responses evolved from assigning meaning, concealing, and withdrawing socially (reactive strategies) to employing indifference and taking the offensive (intermediate strategies). Overall, parents viewed these changes as tangible signs that they were positively adapting to the social implications of their child's condition.

Recommendations for Clinical Care

As the psychosocial ramifications of genetic conditions may be as debilitating as the medical condition itself [Boutte, 1987; Gollust et al., 2003; Peters et al., 2005], it is important for genetics professionals to address issues of courtesy stigma with parents of children with PS. Given the implications that stigma experiences can have for parents and the extent to which the significance of such experiences can change over time, clinicians should routinely assess how parents are responding to the social aspects of their child's condition. Parents who indicate that they are having significant difficulties with stigmatization may benefit from targeted psychological support and counseling. Additionally, clinicians should routinely refer parents of children with PS for peer support (http://www.proteus-syndrome.org). Involvement provides parents with an opportunity to identify implicitly and explicitly with a group who has the same concerns, which can aid in the coping process [Miller and Major, 2000]. Clinicians should not be disheartened by the fact that some parents may choose not to follow-up on referrals to genetic support groups. Seeking a community of others with the same stigmatizing condition can be hindered by parents' desire to conceal their child's condition [Siegel et al., 1998]. Thus, referrals to support groups should be offered repeatedly over time.

Future Studies

Future studies of courtesy stigma among parents of children with a range of genetic conditions are warranted to confirm and expand the list of ways that parents experience and cope with courtesy stigma. Investigation into situational, developmental, and parent-specific variables is needed to increase the understanding of the evolution of parents' courtesy stigma coping process and ways that clinicians and support groups may enhance this transformation. Because parents reported learning how to cope with courtesy stigma from their children, the extent to which siblings and other family members encounter and cope with courtesy stigma is also warranted.

Study Limitations

Participants include those who came to enroll in a natural history study at the NIH, and none declined participation. Results may reflect parents who were farther along in the adaptation process. Since this study was conducted, we have learned that PS is more progressive compared to other overgrowth conditions and is associated with premature death [Slavotinek et al., 2000; Turner et al., 2004]. Because of this, the emphasis of one's stigma perceptions and encounters may have shifted or expanded. Future work needs to assess how this affects parents' social interactions. Participants were predominantly Caucasian, college-educated, working individuals and thus, limits the findings to a sociodemographic group. Lastly, like most retrospective studies, recall bias limits interpretation of our results.

ACKNOWLEDGMENTS

This research was supported by the Intramural Research Program of the National Human Genome Research Institute, National Institutes of Health. We would like to thank Yelena Kemel, MS and Lisa Pike-Buchanan, MS for their assistance with the study design and an initial analysis of the data. We would also like to thank Aideen McInerney-Leo, MS and an anonymous reviewer for their constructive and challenging feedback on this manuscript. Finally, we are most grateful to the Proteus Syndrome Foundation (http://www.proteus-syndrome.org) and the parents of children with Proteus syndrome who enrolled in this study, without whose support and participation this study would not be possible.

REFERENCES

Apse KA, Biesecker BB, Giardiello FM, Fuller BP, Bernhardt BA. 2004. Perceptions of genetic discrimination among at-risk relatives of colorectal cancer patients. Genet Med 6:510–516.

Barker K, Martinez A, Wang R, Bevan S, Murday V, Shipley J, Houlston R, Harper J. 2001. PTEN mutations are uncommon in Proteus syndrome. J Med Genet 38:480–481.

Baxter C. 1989. Investigating stigma as stress in social interactions of parents. J Ment Defic Res 33:455–466.

Baxter C. 1991. Investigating stigma within the stress process. Int J Rehabil Res 14:244–245.

Berkenstadt M, Shiloh S, Barkai G, Katznelson M, Goldman B. 1999. Perceived personal control (PPC): A new concept in measuring outcome of genetic counseling. Am J Med Genet 82:53–59.

Biesecker LG. 2005. Proteus syndrome. In: Cassidy SB, Allanson JE, editors. Management of Genetic Syndromes. 2nd edition. Hoboken, NJ: Wiley-Liss. p 449–456.

Biesecker LG, Peters KF, Darling TN, Choyke P, Hill S, Schimke N, Cunningham M, Meltzer P, Cohen MM Jr. 1998. Clinical differentiation between Proteus syndrome and hemihyperplasia: Description of a distinct form of hemihyperplasia. Am J Med Genet 79:311–318.

Biesecker LG, Rosenberg MJ, Vacha S, Turner JT, Cohen MM. 2001. PTEN mutations and Proteus syndrome. Lancet 358: 2079–2080.

Birenbaum A. 1970. On managing a courtesy stigma. J Health Soc Behav 11:196–206.

Blum N. 1991. The management of stigma by Alzheimer family caregivers. J Contemp Ethnog 20:263–284.

Boutte M. 1987. 'The stumbling disease': A case study of stigma among Azorean-Portuguese. Soc Sci Med 24:209–217.

Chapple A, Carl M, Campion P. 1995. Parental guilt: The part played by the clinical geneticist. J Genet Counsel 4:179–191.

Cohen MM, Hayden PW. 1979. A newly recognized hamartomatous syndrome. Birth Defects Orig Artic Ser 15:291–296.

Cohen MM, Neri G, Weksberg R. 2002. Proteus syndrome. In: Cohen MM, Neri G, Weksberg R, editors. Overgrowth Syndromes. New York: Oxford University Press. p 75–110.

Cohen MM, Turner JT, Biesecker LG. 2003. Proteus syndrome: Misdiagnosis with PTEN mutations. Am J Med Genet Part A 122A:323–324.

Dovidio JF, Major B, Crocker J. 2000. Stigma: Introduction and overview. In: Heatherton TF, Kleck RE, Hebl MR, Hull JG, editors. The Social Psychology of Stigma. New York: Guilford Press. p 1–28.

Goffman E. 1963. Stigma: Notes on the Management of Spoiled Identity. Englewood Cliffs: Prentice-Hall. 147 p.

Gollust SE, Thompson RE, Gooding HC, Biesecker BB. 2003. Living with achondroplasia in an average-sized world: An assessment of quality of life. Am J Med Genet Part A 120A:447–458.

Green SE. 2003. "What do you mean 'what's wrong with her?'": Stigma and the lives of families of children with disabilities. Soc Sci Med 57:1361–1374.

Groce NE, Zola IK. 1993. Multiculturalism, chronic illness. And disability. Pediatrics 91:1048–1055.

Happle R. 1987. Lethal genes surviving by mosaicism: A possible explanation for sporadic birth defects involving the skin. J Am Acad Derm. 16:899–906.

Jones EE, Farina A, Hastorf A, Markus H, Miller D, Scott R. 1984. Social Stigma: The Psychology of Marked Relationships. New York: W.H. Freeman. 347 p.

Kessler S, Kessler H, Ward P. 1984. Psychological aspects of genetic counseling. III. Management of guilt and shame. Am J Med Genet 17:673–697.

Lapham E, Kozma C, Weiss J. 1996. Genetic discrimination: Perspectives of consumers. Science 274:621–624.

Link BG, Phelan JC. 2001. Conceptualizing stigma. Annu Rev Sociol 27:363–385.

Link BG, Struening EL, Dohrenwend BP, Cullen FT, Shrout PE. 1989. A modified labeling theory approach to mental disorders" an empirical assessment. Am Sociol Rev 54:400–423.

Link BG, Stuening EL, Neese-Todd S, Asmussen S, Phelan JC. 2002. On describing and seeking to change the experience of stigma. Psych Rehab Skills 6:201–231.

Link BG, Yang L, Phelan JC, Collins PY. 2004. Measuring mental illness stigma. Schizophr Bull 30:511–541.

Miller CT, Major B. 2000. Coping with stigma and prejudice. In: Heatherton TF, editor. The Social Psychology of Stigma. New York: Guilford Press. 450 p.

Peters K, Apse K, Blackford A, McHugh B, Michalic D, Biesecker B. 2005. Living with Marfan syndrome: Coping with stigma. Clin Genet 68:6–14.

Puhl R, Brownell KD. 2003. Ways of coping with obesity stigma: Review and conceptual analysis. Eat Behav 4:53–78.

Rothenberg K, Terry S. 2002. Before it is too late—Addressing fear genetic discrimination. Science 297:196–197.

Sankar P, Cho MK, Wolpe PR, Schairer C. 2006. What is in a cause? Exploring the relationship between genetic cause and felt stigma. Genet Med 8:33–42.

Shinaman A, Bain L, Shoulson I. 2003. Preempting genetic discrimination and assaults on privacy: Report of a symposium. Am J Med Genet Part A 120A:589–593.

Siegel K, Lune H, Meyer I. 1998. Stigma management among gay/bisexual men with HIV/AIDS. Qual Soc 21:3–24.

Slavotinek AM, Vacha SJ, Peters KF, Biesecker LG. 2000. Sudden death caused by pulmonary thromboembolism in Proteus syndrome. Clin Genet 58:386–389.

Susman J. 1994. Disability, stigma and deviance. Soc Sci Med 38: 15–22.

Swim JK, Cohen LL, Hyers LL. 1998. Experiencing everyday prejudice and discrimination. In: Swim JK, Stangor C, editors. Prejudice: The Target's Perspective. San Diego, CA: Academic Press. p 37–59.

Turner JT, Cohen MM, Biesecker LG. 2004. Reassessment of the Proteus syndrome literature: Application of diagnostic criteria to published cases. Am J Med Genet Part A 130A:111–122.

Wiedemann HR, Burgio GR, Aldenhoff P, Kunze J, Kaufmann HJ, Schirg E. 1983. The Proteus syndrome. Partial gigantism of the hands and/or feet, nevi, hemihypertrophy, subcutaneous tumors, macrocephaly or other skull anomalies and possible accelerated growth and visceral affections. Eur J Pediatr 140: 5–12.