

 **Family Management Style and
Diagnosis Disclosure in Klinefelter
Syndrome: Preliminary Findings** 

Looking to the Future
For X and Y Chromosome Variations
July 2013 Conference
Denver, Colorado

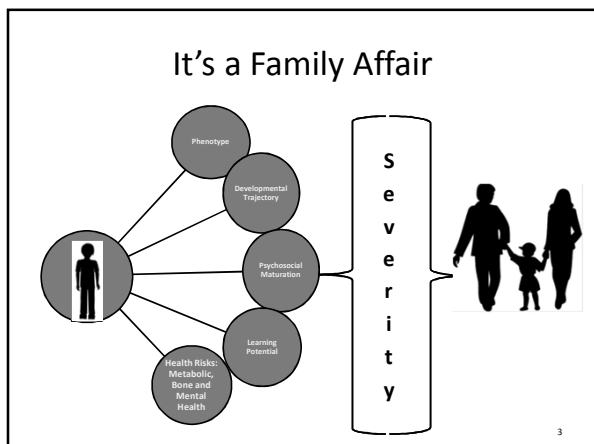
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1

Learning Objectives

- Explore how KS affects the child and the family system
- Describe the vulnerabilities and needs of parents of boys with KS and how they manage the health of their sons
- Discuss how to initiate conversation with health care providers concerning how we can optimize clinical support for KS families.

2



Families of Children with Chronic Conditions

- Parental attitude shapes the way families manage the health of the affected child. (Gallo et al. 2005)
- How family life is affected by childhood chronic conditions:
 - Systems
 - Developmental Life course
 - Symbolic Interaction
 - Stress and Coping
 - Resilience
 - Family Management Style (Knafli, VanRiper & Knafli, 2012)

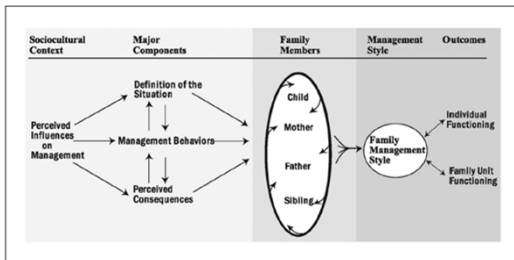
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Family Management

- Families have different responses to chronic illness based upon real and perceived capabilities. (Summers et al. 2005)
- Management styles may vary from
 - Accommodating
 - Enduring
 - Struggling
 - Floundering (Knafli et al. 1996)

5

Theoretical Framework: Family Management Style



Knafli, Deatrick & Havill, 2012

6

Previous Study

"An Exploratory Study of Physical Phenotype, Biomarkers and Psychosocial Health in Boys with Klinefelter Syndrome"

Close et al. (2011)

- Recruitment Challenges:
 - Some parents had not disclosed the diagnosis
 - Difficult to reach target population with traditional recruitment means
- Collection of demographic information and medical history of boys
 - Parents wanted to talk about their stories
 - Expressed frustration about lack of information
 - Parents complained that they did not know how to care for their sons

7

Current Study:

Family Management and Diagnosis Disclosure in Klinefelter Syndrome

- Challenges from the previous study led to new research question:
 - "What is it like to have a son with KS?"***
 - How do parents manage information?
 - How do parents care for their sons?
 - How do parents decide to disclose the diagnosis their sons and others?

8

Purpose: Aim 1

- To explore the needs of families who have sons with KS
- How they feel about raising a son with KS
- How they manage the health of their son
- How they make the decision to disclose the diagnosis to their sons

9

Purpose: Aim 2

- To describe family management style, family quality of life and perceived stress in parents
- To examine the relationships among parental disclosure of diagnosis and these factors

10

Methods

- Design:
Concurrent Triangulated Mixed Method
- Qualitative interviews
- Quantitative Surveys
- Sample: 40 families of boys with KS using purposive sampling for maximum variation

11

Data Collection

Qualitative Interviews

- Interviews conducted using the semi-structured interview guide
- 45 minute interviews conducted via SKYPE or telephone
- All interviews were recorded and transcribed.
- Analysis by Interpretive Description Method (Thorne, 2008) using Atlas.ti software

Quantitative

- Survey's were self-administered via Qualtrics® Survey software.
- Participants were invited by email link.
- Responses were recorded and downloaded to an Excel database.

12

Quantitative Measures: 3 Self-Report Surveys

- **Family Quality of Life Survey** (Hoffman et al. 2006)
 - 25 Item Survey
 - Measures family quality of life.
 - 5 subscales: family interaction, parenting, emotional well-being, physical/material well-being and disability-related support.
- **Family Management Measure** (Knaff et al 2011)
 - 45 Item Survey
 - Measures how families manage caring for a child with a chronic condition and how they incorporate condition management in everyday lives.
 - Subscales: Child's Daily life, Condition Management Ability, Management Effort, Family Life Difficulty, View of Impact & Parent Mutuality
- **Stress** (Cohen et al., 1983)
 - 10 Item survey
 - The degree to which situations in one's life is appraised as stressful

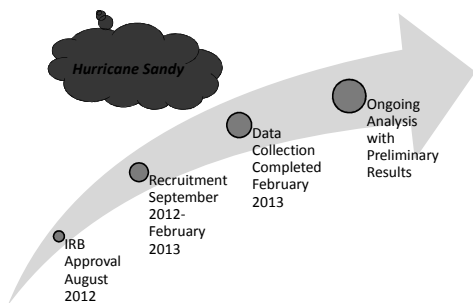
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Qualitative Approach Semi-Structured Interview

- What is it like to have a son with KS?
- How do you find information about KS?
- How do you manage your son's health care?
- How did you disclose the diagnosis to your son?
- What do you worry about?
- What do families of sons with KS need?

14

Timeline to date



15

Results: Characteristics of the Sample

N=40

| Age | Mean ± SD | Range |
|------------------|------------------|----------------|
| Parent | 48.6 years ± 7.6 | 32 – 62 years |
| Son | 13.2 years ± 6.9 | 0.3 – 25 years |
| Gender | Frequency | Percent |
| Female | 33 | 82.5% |
| Male | 7 | 17.5% |
| Marital Status | | |
| Married | 35 | 87.5% |
| Divorced | 5 | 12.5% |
| Race | | |
| Caucasian | 35 | 87.5% |
| African American | 1 | 2.5% |
| Hispanic | 2 | 5% |
| Other | 2 | 5% |

16

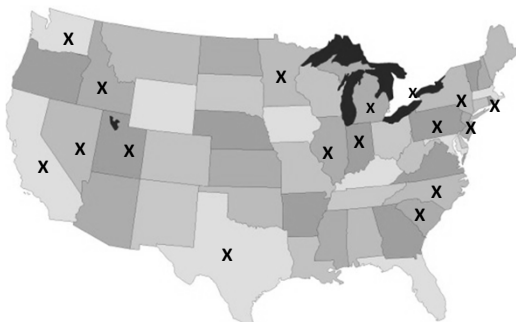
Results: Characteristics of the Sample

N = 40

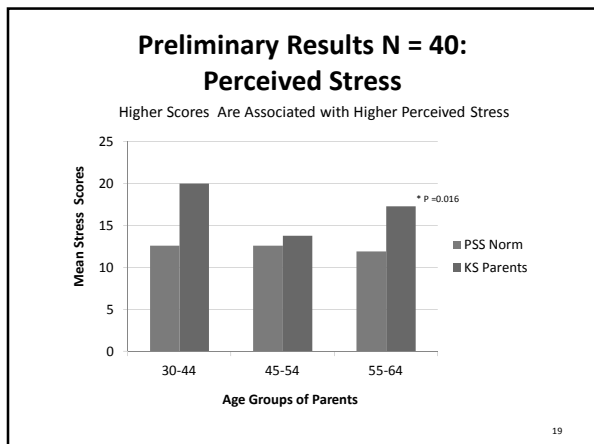
| Parent Education | Frequency | Percent |
|--------------------------------|-----------|---------|
| HS Diploma or less | 7 | 17.5% |
| College Degree | 23 | 57.5% |
| Graduate Degree | 10 | 25% |
| Son's Karyotype | | |
| 47, XXY | 37 | 92.5% |
| 47, XXY/46XY | 3 | 7.5% |
| Diagnosis of KS | | |
| Pre-natal | 21 | 52.5% |
| Post-natal | 19 | 47.5% |
| Disclosure of Diagnosis to Son | | |
| Yes | 25 | 62.5% |
| No | 15 | 37.5% |

17

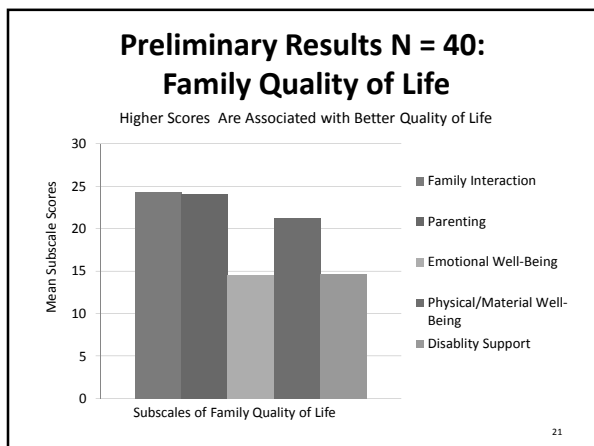
Geographic Distribution of Recruitment N = 40

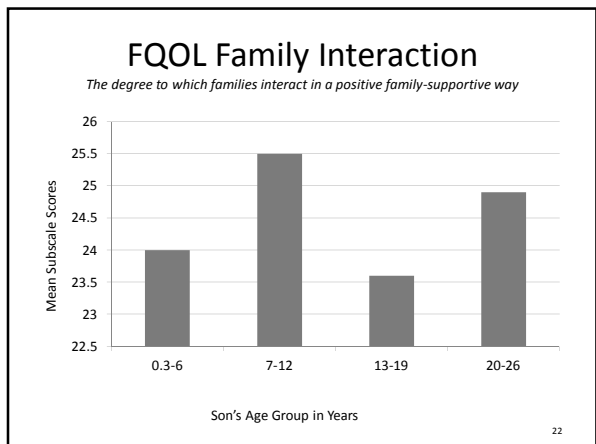


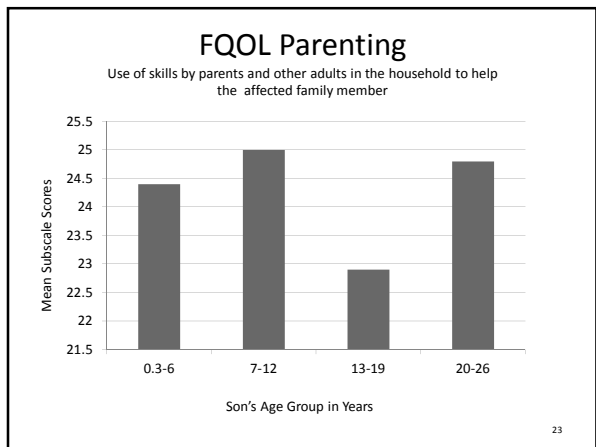
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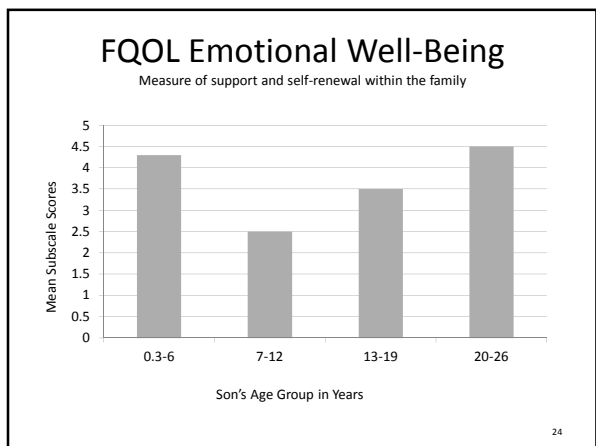


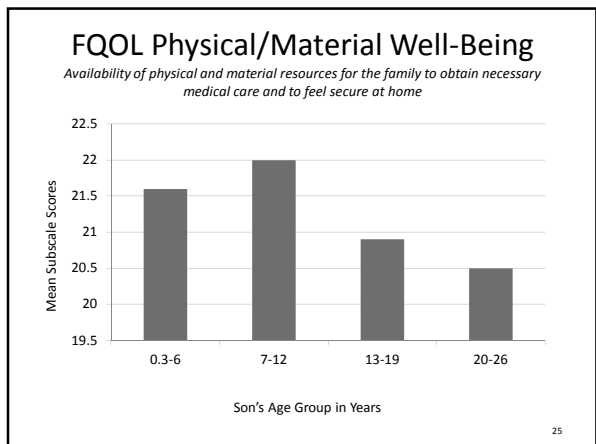


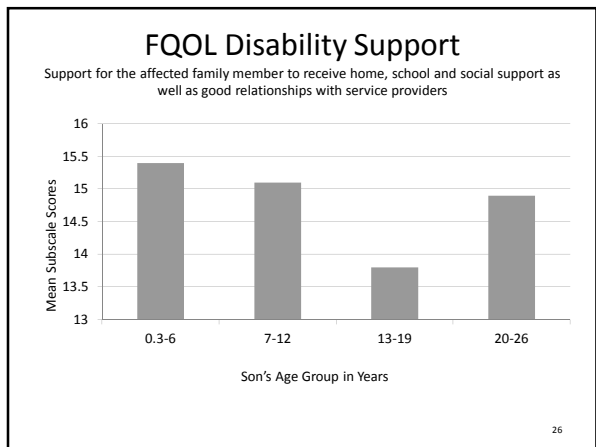


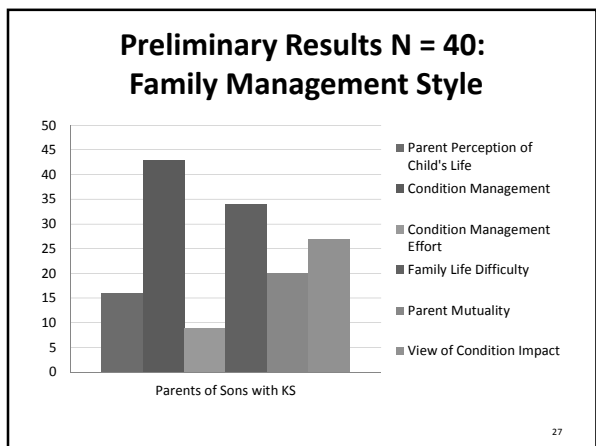


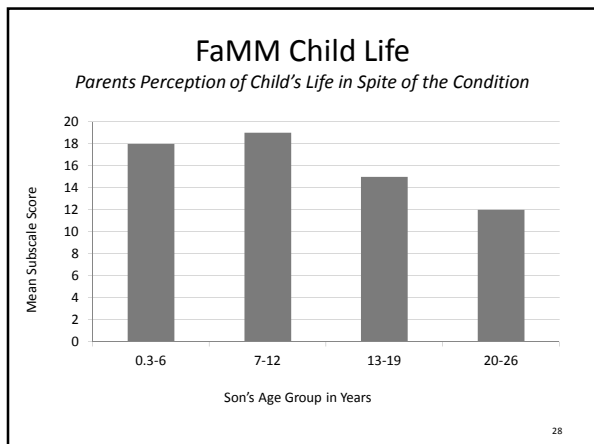


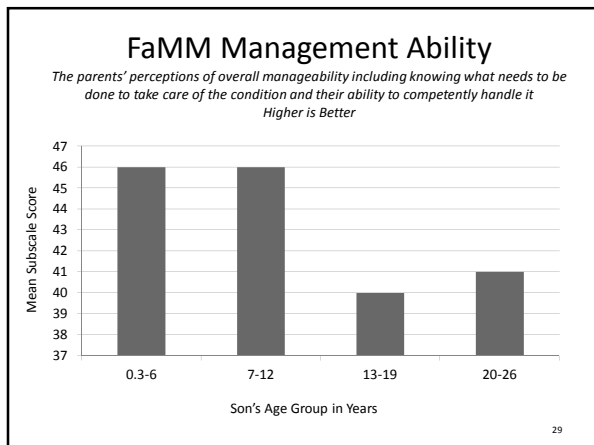


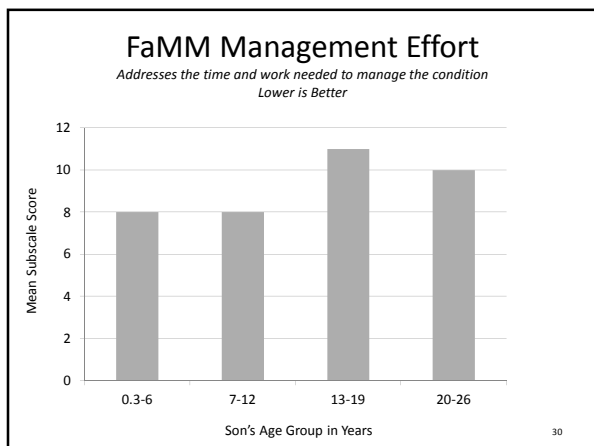


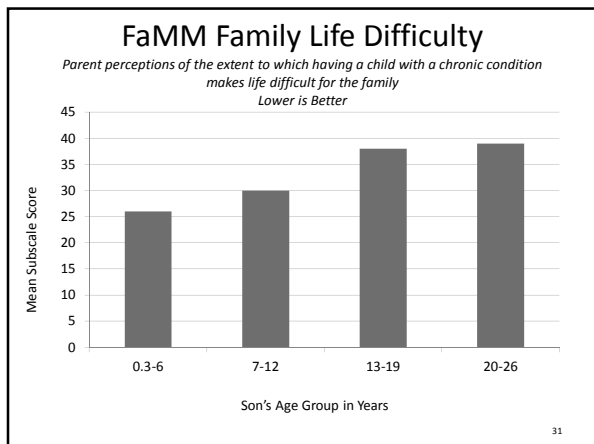


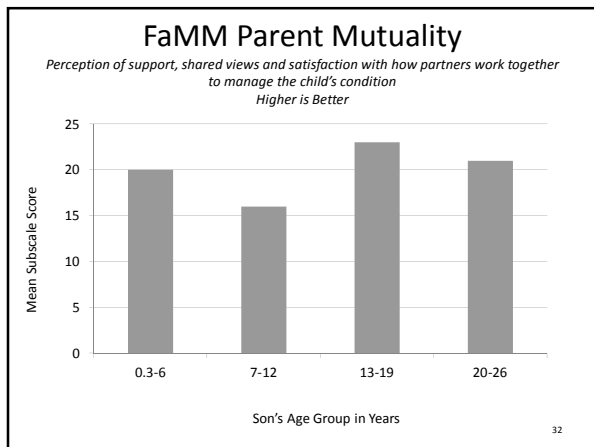


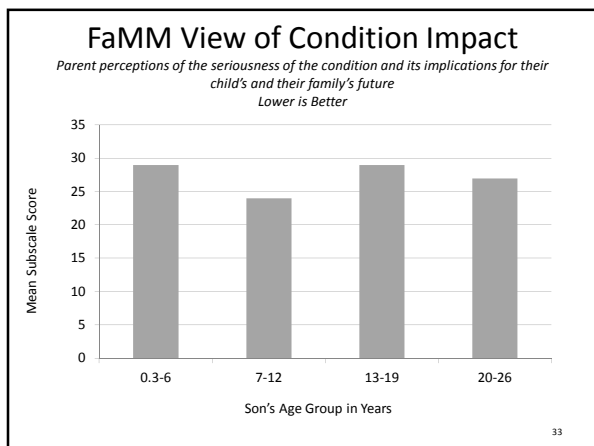












Preliminary Results N = 40 Qualitative Themes

- Double Edge Sword
- In the Dark
- Internalized Worry
- Diagnosis Disclosure Dilemma
- Need for a Roadmap



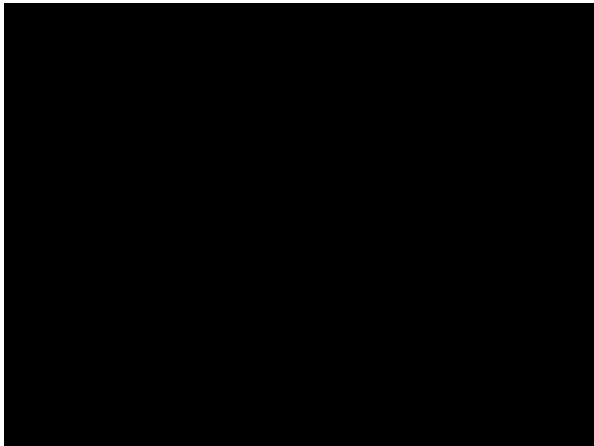
34

What Is It Like to Have a Son with KS?

"It's a Double-Edge Sword" Mother of a 16 year old



35



What Parents Worry About

Life Course



Misunderstanding

Labeling



Education

Stigma


Independence

Social Relationships, including marriage

Infertility

37

What Parents Think About Disclosing the Diagnosis of KS to Their Sons



"And so we were **battling over this**, but each time we talked about telling him, you know, you **don't want to tell him in middle school because the kids are all mean and nasty**, you know? Then you're in high school and they're dealing with all these other issues, so **you don't want to disrupt that**."

But what ended up **finally pushing us over the edge** to tell him was an article that I read about them **finding sperm in teenage boys**, and that you could extract it and bank it for the future in the event that you could get sterile as an adult, and that the earlier you did that, the less likely the extraction procedure would be invasive. And so **we finally decided to tell him** so that we could take him and have him bank his sperm in case that he wanted to have children later. **And that's why we told him.**"

Father of a 16 year old son

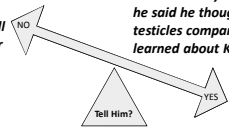
38

Tipping Point: Diagnosis Disclosure

"We debated numerous times as to whether to tell him when he was little, or tell him when he was 12, or when he was 15, or maybe not tell him at all and let him discover it when he was married and was going to be sterile."

Father of a 16 year old

NO



YES

"He was studying about Biology and he said he thought he had small testicles compared to other kids. He learned about KS in Biology Class"

Mother of a 14 year old

"He'd actually asked me. He was like, 'Mom, why do I have to keep going to all these doctor's appointments?' That's kind of how it started"

Mother of a 16 year old

"So we told him in bits and pieces, I would say, as much that seemed like enough"

Mother of a 15 year old

39

Observing the **HOW** to Disclose the KS Diagnosis

"I said to him, son, you know how you've always wondered why things don't come easy to you when you're learning and you say you can't remember things as easily as other kids?"

Well, the good news is that you are not crazy and it's real and it's called Klinefelter Syndrome....and now we're going to put our game faces on and we're going to figure it out with you."

Mom of a 14 year old

40

What Parents and Families Need: A Roadmap



"When do we start Testosterone?"
Mother of a 6 year old

"What part of this is KS and what part is my son?"
Mother of an 8 year old

"How can we give our son his best shot at life?" Father of a 3 year old

"How do we talk to him about fertility?" Mother of a 12 year old

"Who is supposed to be in charge of my son's care?"

41

Preliminary Impressions of This Study

Explore how KS affects the child and the family system

- Parents are **distressed** and **frustrated** about how to help their sons **throughout the life span**
- **Confusion** leads to **uncertainty** and **unpredictability** for how to help their sons.
- The **seriousness** and **chronic** nature of KS symptoms contributes to the families' inability to define their **new "normal"**.
- Parents feel lost (**in the dark**) about the ability to manage their son's health.

42

Preliminary Impressions

Describe the vulnerabilities and needs of parents of boys with KS and how they manage the health of their sons

- Stress is higher: parents of younger & older children
- Family quality of life is decreased for emotional well-being and disability support: scores are strong for family interaction and parenting
- Family Management Measure shows interesting patterns and we are in the process of linking those scales with the qualitative findings.

43

Potential Implications for Practice

Initiate discussion with health care providers, concerning how we can optimize clinical support for KS families

- **Awareness and training** of HCPs for higher index of suspicion.
- **Need for more research** on decision support for how to treat KS.
- **Need to develop interventions** to help with individual & family management of KS through the life course
- **Parents need help and support for how to disclose** all aspects of KS to their sons and to others

44

Significance

- Since one in 450-500 male births results in having a son with KS, there is a need to understand how parents care for the physical and psychosocial needs of their sons.
- This study adds to the understanding:
 - How families manage KS
 - How families choose to disclose the KS diagnosis
- Specific problem identification leads to the development of an intervention to help families in the future

45

Limitations

- Results are preliminary, so conclusions are not final
- Potential for biased sampling
- Self-report questionnaires
- Full qualitative rigor has not been assessed yet
 - Dependability, Credibility, Trustworthiness, Triangulation

46

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Much Appreciation
To
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47

Questions Are Welcome



"All I can say is that we need to find ways to help more families because KS occurs in one in 500 boys. That's a lot of kids."

Mother of a 16 year old son

48
