



Common Stressors for Families Having a Child with Congenital Adrenal Hyperplasia

Louise Fleming, PhD, RN

The University of North Carolina at Chapel Hill
School of Nursing



UNC
SCHOOL OF NURSING

My Family



Family Challenges Associated with CAH

---What We Already Know.....

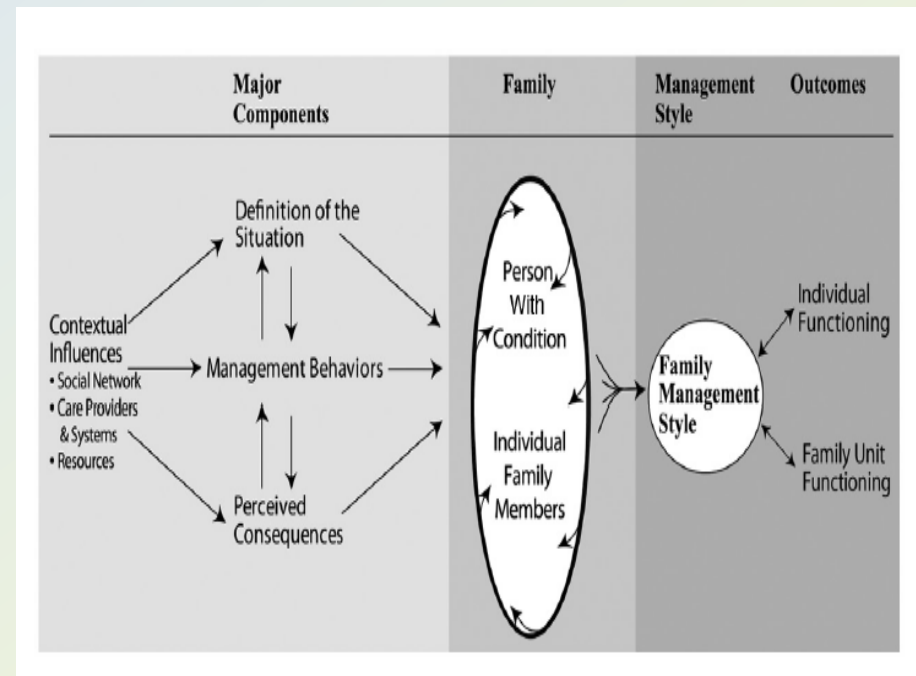
- Stress and uncertainty related to having to perform emergency measures is present for these families, as it is with other conditions such as type 1 diabetes and life threatening food allergies
- Females born with CAH experience ambiguous genitalia
 - Additional stressor
 - Stigma
- Instruction from healthcare professionals to parents on how to respond to crisis events lacks uniformity
- Family isolation occurs in children with chronic illness

Aims of a Recent 2016 Study

- **AIM 1.** Describe circumstances surrounding adrenal crises in children with CAH
- **AIM 2.** Explore parents' perceptions of the consequences, for themselves and their family, of their child's CAH
- **AIM 3.** Examine a possible relationship between parents' management ability and the impact CAH has on the family

Theoretical Framework-Family Management Style Framework (FMSF)

- The FMSF was developed to accommodate multiple patterns of family response to a chronic illness.
- Families dealing with a chronically ill child attempt to normalize based on the:
 - Family's definition of the situation
 - The way they manage often complex health needs
 - The way they perceive future consequences related to their child's condition



Participants

- Recruited through the CARES Foundation
- Inclusion:
 - Parent over the age of 18
 - English speaking
 - Have a child between 0-18 years of age with classic CAH
 - Access to telephone, computer, email account
- Invitation to Participate letter was emailed to all CARES members that had expressed an interest in participating in research when they joined
- 77 parents took the survey and 16 were interviewed

Parent Characteristics-Phase 1

Child Characteristics-Phase 1

Parenting Role	<u>n (%)</u>	Racial Identification	<u>n (%)</u>
Mother	60 (78)	American Indian/Alaskan Native	1 (1.3)
Father	15 (19)	Asian	2 (2.7)
Grandmother	2 (3)	Black	1 (1.3)
		Hawaiian/Pacific Islander	0
Racial Identification	<u>n (%)</u>		
American Indian/Alaskan Native	1 (1.3)	White	71 (94.7)
Asian	2 (2.7)		
Black	1 (1.3)	Ethnicity	<u>n (%)</u>
Hawaiian/Pacific Islander	0	Hispanic	6 (7.9)
White	71 (94.7)	Non-Hispanic	1 (1.3)
Ethnicity	<u>n (%)</u>		
Hispanic	4 (5.3)	Wears Medical Alert Tag	<u>n (%)</u>
Non-Hispanic	72 (94.7)	Yes	51 (66)
		No	26(34)
Yearly Family Income	<u>n (%)</u>	Gender of Child	<u>n (%)</u>
<\$50,000	10 (13.7)	Male	33 (43)
\$50,000-80,000	12 (16.4)	Female	44 (57)
\$80,000-100,000	15 (20.6)		
\$100,000-150,000	14 (19.2)	Age of Child	
>\$150,000	22 (30.1)	Mean (SD)	8.4 (5.3)
		Range	1 month-18 years
Age of Mother			
Mean (SD)	41.98 (7.4)		
Range	26-54		
Age of Father			
Mean (SD)	40.71 (4.9)		
Range	33-47		
Age of Grandmother			
Mean (SD)	59.5 (21.9)		
Range	44-75		

Family Stressors For Families Having a Child with CAH-- What we found out...

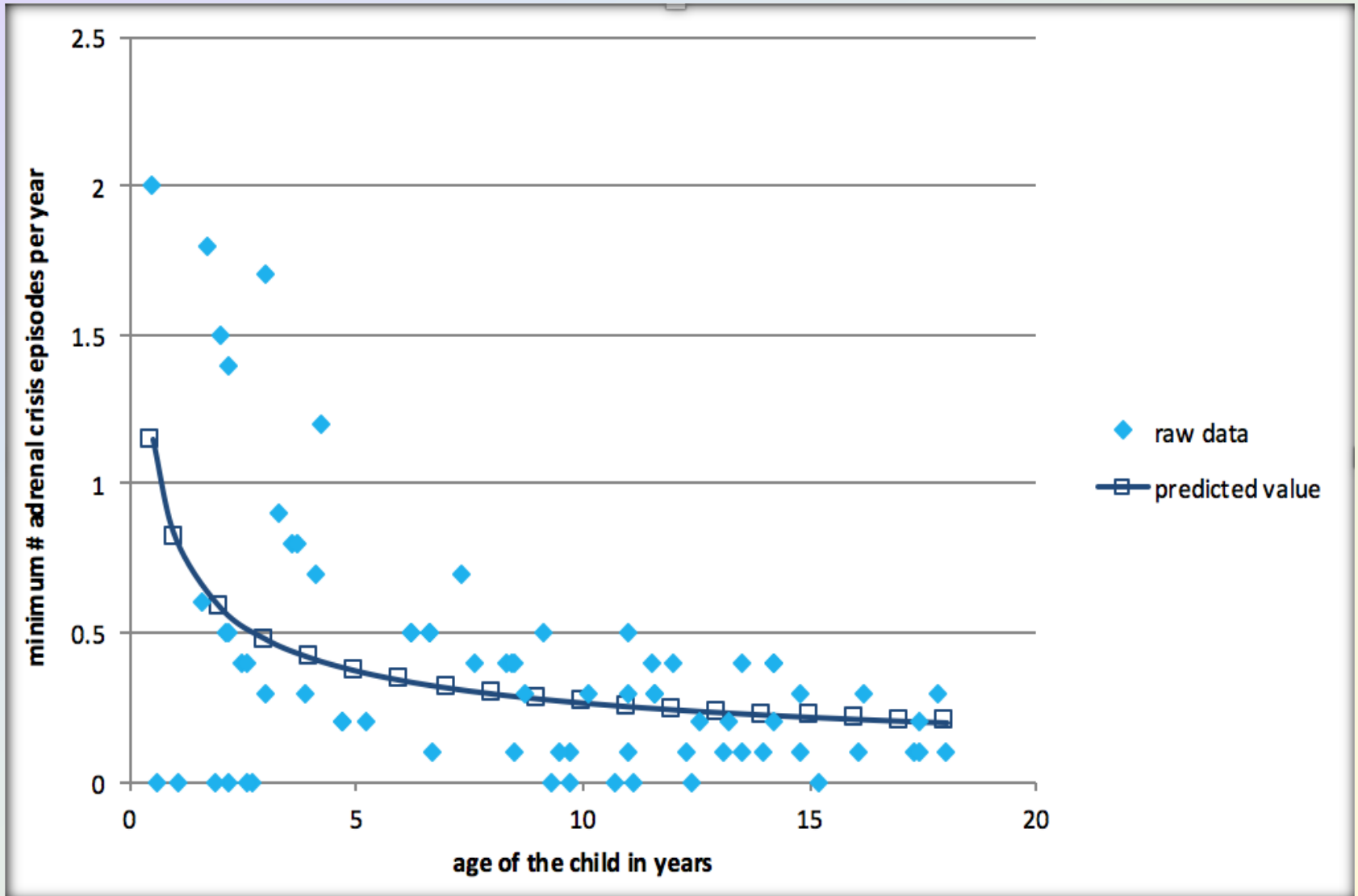
- Adrenal crisis events and the potential for such events
- Interchanges with healthcare providers and school personnel
- Social network
- Access to resources
- Stigma



Adrenal Crisis Events

- Adrenal crisis and the fear of adrenal crisis are paramount in the lives of families having a child with CAH
- Parents reported children having a minimum number of 1 adrenal crisis episode every 1.3 years in the first five years of life; after the age of five, parents reported a minimum average of 1 adrenal crisis episode every 3.7 years
- As children get older, parents feel more able to manage their child's CAH and feel like CAH has less of an impact on the family

Phase 1-Minimum Number of Adrenal Crisis Episodes and Age of the Child



Interchanges with Healthcare Providers

- Some providers do not offer emotional support and did not refer parents to family support groups---local nor national
- Most parents felt their current providers educated them on the emergency injection of hydrocortisone, but were only shown how once. However, parents voiced concern that one-time instruction was insufficient in terms of feeling confident in their ability to effectively manage an adrenal crisis.

“As far as [providers] being there emotionally, I don’t know if his doctors ever really played that role. I think - I just don’t see them – or I can’t cite an example where there’s the pat on the back or the hug. Nothing like that.”

“They [initial pediatric endocrinologist office] did not demonstrate anything. They actually could not even give us a correct dosage of how much to put in the syringe. They gave us a piece of paper that had instructions on it and sent us on our way.”



Interchanges with School Personnel

- Parents described educating their child's school nurse or daycare provider on CAH and reviewing injection instructions with school nurses on a yearly basis, but described the process as lengthy
- Parents reported favorable experiences with school personnel regarding nurse preparation and teacher understanding of the condition

"But each time when she has change schools, you know moved up from you know kindergarten to first grade to elementary, we have to start all over with the nurse and how is that nurse going to respond? But, it's usually gone well."

Social Network

- **Babysitters**

- How much to tell
- Train them on injection?
- Prefer to use family members

- **Extended family**

- Some described them as helpful
- Others said they “didn’t get it” and were afraid to help watch the child

- **Extracurricular Activities/Sports**

- Do they tell the coach?
- Afraid of the intensity of the sport and child’s health but desire for child to participate like “other kids”
- Does a parent need to be at every practice and/or game?

“I have taught the babysitter how to give the injection. And I write it down on a piece of paper ... like really simple steps if they would have to. But I would always emphasize to just call 911.”

“This is my son, so if you’re going to be around my son, you’re going to know what to do. You’re going to know what to look for. You’re going to understand. Now what might happen to him if I leave the room, if I run to the store, if whatever. If you’re with my child you’re going to know what he has and how to do the basic treatment of it.”

Access to Resources

- Many left initial provider because provider didn't seem to have the correct CAH knowledge to properly treat
- Some parents drive 3-8 hours to see provider

"Yeah now we've been stable now for I think two years with the one we got but we still have to drive three and a half hours away [to see new pediatric endocrinologist]."

"Because what I was finding was that it could be fatal if not treated correctly. And the endocrinologist we had at the time was not somebody we liked at all and did not explain this to us."



Stigma



- Both boys and girls experienced stigma related to having a chronic condition
- Differences in the view of the condition and what is the biggest challenge associated with the condition
- For families having a girl, the stigma was pervasive and profound

How Parents Define CAH

- Overall, children are doing well in administering daily medication, child's current height and weight, and child's performances in school.
- All felt that their child was thriving, happy, and well-adjusted



Perceived Consequences

- Unpredictable, acute illnesses and the possibility of adrenal crisis often disrupts routine
- Parents are hopeful for their child's future, but expressed concerns during puberty/adolescence
- Worry as the child transitions into a young adult
- Minimal concern for effect on siblings except during crisis events
- Future family planning challenges

“Yeah and I think about that when she goes to college, you know? Is she going to be equipped? Are the medical centers going to be equipped if she gets hurt or if she drinks too much and she's vomiting the next day? You know I think about those things.”

“My expectation is she's going to have a normal life. And she's got a pretty good head on her shoulders.”



Strategies that Parents Described as Helpful-Adrenal Crisis Events

- Child wearing a medic-alert bracelet
- Having the hydrocortisone injection kit with their child at all times
- Frequent review with their providers on how to give the injection as well as researching injection technique on their own/through CARES
- Having a solid understanding of when and how to stress dose in general

Strategies that Parents Described as Helpful-- Interchanges with healthcare providers and school personnel

- Meeting with all teachers and principal at the beginning of each year and doing a “training” session—CARES video (now on a flash drive as well as website)—what to look for
- Meeting and/or emailing with teachers throughout the school year
- 504 plan-accommodations including absences, not restricting fluids or restroom (esp. during standardized tests), not restricting sodium
- Having open communication between their child’s pediatric endocrinologist and general pediatrician
- Asking pediatric endocrinologists and/or staff to review injection training at well child checks
- Keep a “notebook” that shows labs, growth, times when child was stress dosed-some keep on a flash drive

Strategies that Parents Described as Helpful-- Social Network

- Having a scripted email to send to parents who will be watching their child –birthday parties, sleepovers, etc.
- Some parents had coaches that were willing to learn how to give the injection, but most did not go that route-they attended all practices and games
- Most tried to get family to babysit, especially with younger children
- If they did use a teenage/college age babysitter, they either:
 - Trained them how to give injection
 - Didn't travel too far

Strategies that Parents Described as Helpful-- Social Network--Continued

- Parents prepared their child to answer questions from other children regarding their medical bracelet or why they took medicine. Be simple and direct.
 - Instructed child to explain that they are “healthy” but need to take medicine every day to stay “healthy.” Also if they were to get sick, and adult would need to know quickly
- A fine line in letting others know that their child has CAH but not scaring those in their social network—emphasis on how the vast majority of the time, their child with CAH was just like any child.

Strategies that Parents Described as Helpful— Access to Resources

- Seek second opinions-sometimes out of state
- Look for research studies (CARES website is a good source)
- CARES designated Comprehensive Care Centers
- Join support groups
- Ask provider how many children with CAH he/she has treated in their career
- Have a clear path on which provider to take child to when acutely ill-- Ped Endo or Pediatrician?

Strategies that Parents Described as Helpful– Stigma

- Disclosing only on a “need to know” basis to others at times that the family is comfortable with
- Joining support groups
- Explaining CAH in terms of factual, medical, pertinent information
- Describing to their child that their condition is very manageable and that the expectation is that their future is filled with awesome possibilities
- Understanding the frustrations that children with chronic conditions sometimes experience, especially related to being “different” (ie. Wearing a medical tag, frequent doctor appointments, taking daily medications, a cautious approach when acutely ill)

Important Conclusions & Future Directions

- Parents adapt to the management challenges of having a child with CAH, but the threat of adrenal crisis is constant
- Lack of standard, repeated education on adrenal crisis management remains a problem
- Parents have an increased sense of management ability and children have less adrenal crisis episodes after their child turns 5 years old
- Contextual influences related to the child's CAH are a significant challenge to families
- Stigma is present in families that have boys and girls with CAH, but it is more profound in families having a girl

....A special thanks to



Questions???

