



# How the Child's Gender Matters for Families Having a Child With Congenital Adrenal Hyperplasia

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## Abstract

Children with congenital adrenal hyperplasia (CAH) are exposed to high levels of testosterone in utero often resulting in nontypical genitalia at birth for girls. The purpose of this analysis, which draws on data from a larger study, was to examine, based on the gender of the child, the family experiences of having a child with CAH. Sixteen parents were interviewed, and comparisons were made across all categories coded in the parents' interviews to examine similarities and differences in the experiences of families based on the child's gender. Families having a daughter with CAH experienced additional challenges when compared with families having a son. These include fear of stigmatization, challenging surgical decisions, and concerns regarding disclosure of the condition. Acknowledging this and creating support programs tailored for girls that address surgical procedures and complications resulting from being born with elevated testosterone are critical to promoting a healthy family and child response.

## Keywords

nontypical genitalia, congenital adrenal hyperplasia, family management

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Congenital adrenal hyperplasia (CAH) is an inherited, endocrine disorder that occurs in approximately 1 in 15,000 live births and affects males and females equally (Speiser et al., 2010). Both boys and girls born with CAH are exposed to high concentrations of testosterone in utero; while boys typically do not show any outward physical signs of this exposure, girls often have nontypical genitalia to varying degrees, which typically leads to a diagnosis shortly after birth (Witchel & Azziz, 2011). The degree of virilization is graded according to the Prader score (with a 0 appearing as a typical female and a 5 appearing as a typical male). Gender assignment in these affected females can pose both a medical and family emergency (Merke & Bornstein, 2005; Speiser et al., 2010). A surgical procedure, often known as reconstructive feminizing genitoplasty, is recommended by some providers at an early age and can involve multiple surgeries into adolescence (Karkazis, 2008; Witchel & Azziz, 2011). Although girls with CAH typically identify as female (Berenbaum & Bailey, 2003; Dessens, Slijper, & Drop, 2005; Meyer-Bahlburg, 2011), they can show increased preference for male toys and activities (Nordenstrom, Servin, Bohlin, Larsson, & Wedell, 2002; Pasterski et al., 2005; Wong, Pasterski, Hindmarsh, Geffner, & Hines, 2013), increased aggression (Mathews, Fane, Conway, Brook, & Hines, 2009; Pasterski et al., 2007), increased male-typical sexual orientation (Nordenstrom et al., 2010), and differences when compared with non-CAH girls in spatial abilities (Berenbaum, Bryk, & Beltz, 2012; Hines et al., 2003). Having a daughter born with nontypical genitalia presents unique challenges to families including uncertainty about the sex of the child at birth and complicated decisions regarding the possibility of genital surgery. This analysis contributes to a better understanding of a situation that both families and health care professionals find extremely challenging to manage. Despite the large body of research addressing family response to childhood chronic conditions, we have limited understanding of the challenges these families face and the ways in which providers contribute to or impede family management and decision making.

All children with CAH are at significant risk for adrenal crisis, defined as an abrupt, life-threatening state with symptoms including hypotension, pallor, fatigue, headache, tachycardia, and vomiting, due to steroid deficiency (Speiser et al., 2010). Parents must administer steroids, typically oral hydrocortisone, up to 3 times daily to replace deficient levels of cortisol, and "stress dose," meaning giving additional oral steroid doses or an intramuscular injection of hydrocortisone, during times of acute physical illnesses due to common viral and bacterial causes as well as during surgeries or traumas. Despite the complex nature of CAH management, both during times of maintenance and emergency, as well as the especially challenging situations and decisions that girls and their families experience, prior studies have focused on the

medical management of CAH and the gender preferences of girls with CAH. Little is known about how families incorporate CAH management into their daily life and the distinct management challenges of having a daughter with CAH. With improved understanding of family life in the context of CAH, health care providers will be better equipped to provide appropriate, family-focused care and guidance to these families.

This secondary analysis comes from data collected during the qualitative phase of a cross-sectional mixed-methods (Phase 1, quantitative; Phase 2, qualitative) study conducted by the first author and guided by the Family Management Style Framework (FMSF). The FMSF provides a structure for understanding family responses to a child's chronic condition (Knafl, Deatrick, & Havill, 2012). The framework is comprised of three components: *definition of the situation* (the ways in which the child and child's condition is viewed by the parent(s), management mind-set, and parental mutuality), *management behaviors* (parenting goals, strategies, and behaviors linked to caring for a chronically ill child), and *perceived consequences* (actual or expected family, child, and illness outcomes that shape management behaviors; Knafl et al., 2012; Knafl et al., 2013). The purpose of the larger study was to examine parental management of adrenal crisis in children with CAH; however, during the analysis of data during Phase 2 of the study, it became evident that certain aspects of the family experience of having a child with CAH were profoundly different based on the gender of the child. The purpose of this analysis, which draws on data from the larger, mixed-methods study, was to examine, based on the gender of the child, the varying family experiences of having a child with CAH.

## Method

### *Participants*

To be included in the larger study, parents needed to be over the age of 18, English speaking, and have a child between the ages of birth and 18 years diagnosed with classic, salt-wasting CAH and free from any other complex health conditions. Furthermore, parents needed to have access to a telephone, computer, and email account. After receiving the university's Institutional Review Board (IRB) approval, parents were recruited through the CARES Foundation (CARES), a nonprofit organization based in New Jersey, which provides support to families in which there is a member with CAH (CARES Foundation, 2014). Recruitment for the study consisted of an "Invitation to Participate" letter that CARES emailed to its members who had previously expressed an interest in participating in research. In addition, CARES

provided a brief description of this research study within the research section of their website. Parents who contacted the first author were emailed the measures and consented to participate online.

Sixteen parents, seven mother/father dyads and two single mothers, consented to participate in the qualitative portion of the parent study. Participants contacted the primary author via email and specified, when completing the online measures, that they were willing to be interviewed in addition to completing the online measures. The purposive intent of the Phase 2 interview sample in the primary study was to achieve maximum variation in terms of demographics (e.g., parents of boys and girls, varying child age, etc.) and parents' experiences managing adrenal crisis events. Scores on the structured measures in Phase 1 were also examined so that parents reporting both high and low impact of the condition on their family as well as differing levels of management ability were included in the second phase of the study. The final sample for the qualitative portion included parents from five families having a girl with CAH and four having a boy. Child age ranged from 2 to 15 years and geographical location, family income, marital status of the parents, and scores on the quantitative measures were varied in keeping with the intent of the purposive sample. The interviews were conducted over telephone. Telephone interviews can be used as an effective tool in research, and their credibility in qualitative research has been supported (Novick, 2008; Saura & Balsas, 2014). Some advantages to telephone interviews include decreased cost, increased access to multiple geographical areas, decreased interviewer bias, and a faster pace of interviewing (Novick, 2008). However, there are certain challenges associated with telephone interviews such as maintaining participant involvement, clear communication, and lack of visual cues (Musselwhite, Cuff, McGregor, & King, 2007); yet, when comparing in-person and telephone interviews, there is considerable evidence that telephone interviews generate data of comparable quality and depth to in-person interviews. (Novick, 2008).

### *Analysis of Data*

A professional transcriptionist transcribed the audio recordings verbatim, and the transcriptions, which the first author checked for accuracy against the recording, were reviewed multiple times to get a broad understanding of the content. MAXQDA (VERBI Software, 2015), a qualitative software program, was used to aid with the coding and retrieval of coded data. Conventional content analysis was used to identify descriptive and/or conceptual codes to guide the analysis of data from Phase 2 of the parent study (Hsieh & Shannon, 2005; Vaismoradi, Turunen, & Bondas, 2013). Coded data were then reviewed

across individual respondents as well as dyads to identify themes characterizing the families' response to the condition and case summaries were developed. Case summaries are a useful strategy for grounding the analysis of individual codes in the context of the respondent's overall experience (Ayres, Kavanaugh, & Knafl, 2003; Sandelowski, 2011).

When performing the secondary analysis, the first author reviewed the transcripts and identified themes that were present in the interviews with parents of the girls that were not present in the interviews with parents of the boys. Distinct, gender-based differences emerged from the 16 interviews involving (a) how the condition was defined initially and over time and (b) the perceived consequences of having a child with CAH. There were two interview questions that elicited the richest data in the context of gender differences. The first was "Tell me how you felt soon after your child was diagnosed with CAH and has that changed in the years since diagnosis?" This question prompted parents to discuss the time of diagnosis, which was a particularly confusing time for parents of girls. The second question was "How do you see CAH impacting your child and family in the future if at all?" This question further showed clear and important distinctions in the family experiences of parents of boys and girls with CAH.

## **Findings**

Of the 16 parents (nine families), five had a girl with CAH ranging in age from 2 to 14, and all five girls were born with nontypical genitalia. These parents, four mother/father dyads and one single mother, detailed experiences much different than parents in the four families having a boy between the ages of 3 and 15. These differences included parents' descriptions of the time of diagnosis, their accounts of challenging surgical, treatment-related decision making, and perceptions of stigma surrounding CAH.

### *Timing of Diagnosis*

For boys, the CAH diagnosis was made within 2 weeks of birth, after the child had been discharged from the hospital setting, via newborn screening. None of the parents of the boys stated they were dissatisfied with the manner in which health care providers informed them of the diagnosis: "I guess the very first thing was the newborn screening. I remember vividly the day. I remember exactly what I was wearing. What I was doing. It shattered our whole world basically."

Unlike parents of boys, parents of girls experienced an extended hospital stay after birth because of the child's nontypical genitalia and learned

of the condition during the hospitalization. Providers suspected a diagnosis of CAH or some other condition from the beginning, and these parents stayed in the hospital with their daughters until a definitive diagnosis was made. For parents of girls, the news was somewhat of a relief, as parents were searching for reasons as to why their daughter had nontypical genitalia and had concerns over other possible explanations. The news of CAH provided an answer after several hours or days of confusion, which was welcomed: “So when she was first diagnosed I think that I was . . . I was relieved mostly. The testing confirmed that the condition was something that the doctors understood and that it was something that could be treated.”

Four families of girls expressed frustration with the way in which the health care providers in the delivery room (obstetricians, nurses, pediatricians, nurse midwives) as well as pediatric endocrinologists responded to and explained their daughters’ nontypical genitalia.

They [health care providers] seemed to be much more preoccupied about her genitalia in a way that my partner and I absolutely were not . . . So it was really intimidating. And you know I just had a baby. I was feeling awful and it was, you know, a pediatric gynecologist, a pediatric social worker, pediatric psychologist . . .

Parents in only one family were satisfied with the way providers shared the news of their daughter’s diagnosis. The father remembers this positive encounter:

He could tell by doing some sonograms what the internal physiology was because the outside physiology could have gone either way . . . He said it could be—it was ambiguous enough that he could not make a call from that. So they set up, even on a Sunday morning, they set up the sonogram for us. It was probably like around 9:30, 10 o’clock in the morning, with a big smile on his face and said you have a beautiful, healthy daughter . . .

All five of the parents were told that their child’s gender was uncertain immediately after birth, but the terminology varied. For some parents, the terminology used by providers to describe their daughter in the delivery room was painful and traumatizing.

My husband is in the room, and our nurse takes a peek and says it’s a boy! Then the midwife turns her over, looks at me and says “I’m not so sure.” The NICU doctor and my midwife turned to me and said “we need to talk about the white elephant—that this child might be a ‘hermaphrodite.’” Oh my God! It was

awful, the shock. All I could think was “hermaphrodite.” I’m lying in the hospital thinking can I raise this baby? It was so traumatic.

A father added,

One of the things one of the nurses said when my daughter was first born . . . We thought maybe she was a boy and then like one of the nurses was like, “oh maybe this is a transvestite?” And you hear that and you’re like, really?

Following the diagnosis and within the first month, parents of boys did not report any concerns regarding telling family and friends of their child’s birth and condition. Most described extended family as an initial source of support providing child care for the baby and/or siblings as well as accompanying the parents to doctor appointments. However, for families having a girl with CAH, the experience of who to tell after their daughter’s birth was much more complicated. Parents explained that deciding whether or not to tell friends was difficult, as they were concerned that as their daughter grew, she might feel uncomfortable with others knowing that she was born with nontypical genitalia. One father stated that he didn’t tell certain family members about his daughter’s nontypical genitalia initially because he felt like they would never change her diaper so they didn’t need to know. A mother described not wanting to announce the birth of her daughter on social media because she was unsure at that time if her child was a boy or a girl. Keeping the birth so quiet for the first week, especially when family and friends knew she had gone to the hospital to have the baby, was very stressful and awkward for her and her husband and created an environment of secrecy regarding an occasion that she thought would be joyous and open: “But talking about the ambiguous genitalia with our friends and family . . . pretty much we decided in the beginning we would not tell everybody because what if she didn’t want anyone to know?”

For parents of girls, the way the condition was initially described to them by health care providers was, for the most part, traumatizing and began to shape the way they viewed CAH. A prolonged hospital stay combined with confusion over the condition and stigmatizing language differentiated the time surrounding the diagnosis of CAH for parents of girls versus boys.

### *Surgical Treatment Decisions*

Of the five families having a daughter with nontypical genitalia, parents in four families decided in favor of surgical intervention within the first year. The goals of feminizing genitoplasty are to facilitate genital appearance

compatible with gender, relieve urinary obstructions that result in episodes of incontinence and infections, and to preserve adult sexual and reproductive function (Speiser et al., 2010). All parents described this as a very difficult decision, although none of them currently expressed regret. One mother and father of a teenage daughter who has had two surgeries to date and continues to require daily vaginal dilating interventions at home stated,

We talked about it . . . we went over it a million times in our head and we said you know she may hate us for it later, but we feel this is the best decision we can make for our child.

Another mother of a preteen daughter with CAH explained that she and her husband voiced their concerns regarding trying to preserve later sexual function to their daughter's pediatric urologist.

We, you know, had read about the doctors that focused more on the aesthetic of it and then these . . . children grow up and don't have the nerve endings there because they were cut to make it look perfect and they have no sexual satisfaction. So we expressed to him several times, and got our point across, that we wanted her to . . . have sexual satisfaction.

Finally, another mother of a teenage daughter with CAH shared that although she was currently satisfied with the results of the surgery from a cosmetic standpoint, her daughter had undergone three surgeries and will need at least one more. These surgeries were extremely stressful for this mother and contributed to her decision not to have any more children.

Oh yeah it's really been a struggle for me. I'm not the same person, but it was worth it to me. It was absolutely worth it. I was a wreck. She had to be put to sleep a total of three times for that. Each time I thought I was going to lose my mind with worry so I couldn't do that again.

One family of a 3-year-old girl made the decision not to have feminizing genitoplasty. They felt strongly that there were too many potential problems later in life related to genital surgery and believed that waiting until their daughter was a young adult and could make an informed decision herself was a better approach. They were disappointed in the way in which their daughter's initial pediatric urologist explained the surgical option to them.

He said "well you know this is just the time to do it." And I said wouldn't you have to have like revisions later? And he said "yeah but, you know, it's better now."



He ended up like kind of divorcing us from his practice because we kept saying no to surgery, and he said “well fine then we’ll see you in five years.”

These parents went on to explain that, for their daughter, there was no medical reason to have the surgery. Therefore, it was frustrating and upsetting to them that they felt the medical team did not honor or respect their opinion to forego the procedure, at least until their daughter was older.

Unless there is a medical reason, I would never touch her genitalia because she might need it . . . I don’t know. I think it’s a challenge because you kind of could delay some of these things until even like five or six or when your kid can articulate what they’re feeling, you know?

### **Stigma**

Parents of both boys and girls with CAH described experiencing some degree of stress and stigmatization involving their child’s condition. Wearing a medic alert tag identifying their child as “different,” taking medication during school hours, and having to explain a rare condition that is complex to manage to others in their child’s social circle were mentioned by all parents. However, the stigma for families having a daughter with nontypical genitalia was pervasive and intense and contributed to how these families viewed CAH in general.

We decided in the beginning we would not tell everybody because what if she didn’t want anyone to know? But then we thought that kind creates an atmosphere that there’s something wrong if we keep it a secret . . .

We don’t want to create a freak show environment . . . where people are talking about it all the time because people don’t really talk about their own genitalia all the time. So we kind of want to normalize it, and it’s really hard to know how to do that. . . . There’s nobody to ask.

When parents of both boys and girls were asked what they felt the most important topic to discuss at a support group meeting for families having a child newly diagnosed with CAH, parents of the boys overwhelmingly focused on preparation for times of illness and adrenal crisis. Parents of girls mentioned this as well, but focused heavily on trying to normalize, gender identity, and managing stigma. The life-threatening aspect of CAH appears to be something that parents of girls are clearly aware of and concerned about, but view as something that is clear-cut and manageable. Parents of boys did not view potential adrenal crisis events in this way. Parents of girls described

potential adrenal crisis events as somewhat in the background of daily life. For these parents, issues related to nontypical genitalia were the predominant concern.

How to deal with the ambiguous genitalia is something that's very much ongoing and not as cut and dry as the adrenal condition as it stands because we're sort of constantly running this risk of providing private information about our child. You know we can't untell people. On the other hand, we're also [cognizant] of the fact that there's nothing to be ashamed about this condition and we have to treat it like we would treat anything else. There's nothing to be ashamed of.

In addition, several parents of girls described stress dosing or giving the injection during times of illness as daunting, but easier to manage and explain to others when compared with nontypical genitalia. For stress dosing, there are clear treatment guidelines such as tripling the oral dose if their daughter has a high fever but there were few, if any, guidelines for parenting a child with nontypical genitalia. However, parents of boys all remarked on how the fear of adrenal crisis has affected family life and shaped their view of the condition.

Finally, whether or not society would accept and understand their daughter's past and/or present nontypical genitalia was unknown to parents and described as a major worry. This concern was evident from birth for parents of girls and is reflected in their hesitancy to announce the birth, share information about the diagnosis, and in deliberate attempts by parents to protect their child from possible embarrassment or ridicule.

## Discussion

Rare childhood conditions, such as CAH, are often misunderstood, which creates isolation and stress for those affected as well as their families (Dellve, Samuelsson, Tallborn, Fasth, & Hallberg, 2006; Zurynski, Frith, Leonard, & Elliott, 2008). Family-related challenges, such as access to care, stress during times of acute illness, and school and work disruptions contribute to parental feelings of loneliness and exclusion, which was evident in the larger, parent study (Zurynski et al., 2008). However, families having a daughter with CAH experience significant, additional challenges associated with the condition when compared with families having a son with CAH. These include the fear of stigmatization, challenging surgical decisions, and concerns regarding disclosure to others of their daughter's condition.

The initial time period following the diagnosis of CAH, whether that is in the delivery room or by a newborn screening days later, produced varied parental responses based on the gender of the child. How the child's

condition is initially portrayed to the family by health care providers is critical in shaping the lens through which the family views the condition. For parents of girls with CAH, the view of the condition begins to emerge shortly after birth and is shaped in part by health care providers' responses to the associated nontypical genitalia as well as what terminology is used to communicate the diagnosis to parents. The term "intersex" has been used in the past to describe children with congenital conditions, such as CAH, that result in nontypical sex development; however, in the mid-2000s, it was determined that this term had negative connotations and was not preferred by parents. "Intersex" was then replaced by "disorders of sex development" (DSD; Davies, Malone, & Fairhurst, 2012). Despite this new term, a more recent study showed the terminology most supported by the CAH community was "non-typical genitalia" (Lin-Su, Lekarev, Poppas & Vogiatzi, 2015) over "ambiguous genitalia" or "disorder of sex development"; hence, "non-typical genitalia" is the appropriate terminology for health care providers to use today.

Parents of daughters with nontypical genitalia struggle with sharing information about their child's condition with others, not only at birth, but throughout their child's life (Crissman et al., 2011; Sanders, Carter, & Goodacre, 2012). For parents having a child with nontypical genitalia, the decision of whether or not to disclose information and to whom is based on the likelihood of stigmatization; who they felt had a "right" to the information; a personal ease with the terminology associated with the affected anatomy; and parents' perceptions of their ability to accurately describe the condition (Crissman et al., 2011). If parents decide to share information with others, they often choose to limit the amount and details due to the fear of stigmatization; deciding how much information to share is an added stress for parents.

In the primary study, parents of both genders described difficulties in how much information to provide to individuals in their child's social life such as family and friends. They explained not wanting to "scare" others about the possibility of adrenal crisis events when the likelihood of occurrence is relatively small. For example, determining if injection training is needed for adults watching their child for an occasional, brief period of time was problematic as was apprising their child's adolescent friends of condition management. However, fear that not informing others might result in a life-threatening situation if their child became ill or sustained a traumatic injury was also distressing. Despite these concerns, parents of boys did not describe concerns about stigma surrounding disclosure decisions, unlike parents of girls. As in previous studies, the parents in this current study guarded the knowledge that their daughter was born with nontypical genitalia very closely in an effort to protect their child's relationships now and in the future

and shield them from potential embarrassment and stigmatization (Sanders et al., 2012). However, in doing so, parents worried that keeping this part of their child's life a secret was, in and of itself, stigmatizing.

For families of girls with CAH considering feminizing genitoplasty, perceptions of stigma were described as a major factor in parental surgical decision making. If the degree of virilization is less, meaning minimal clitoromegaly with the junction between the vagina and urethra near the perineum, surgery might not be necessary from a medical perspective. However, some parents may still opt for surgical intervention from a more cosmetic perspective (Speiser et al., 2010). Parents are typically told that if they opt for surgery, it is best to do so between 2 months and 1 year of age due to the assumed child psychological benefits associated with having normal genitalia, less likelihood of associated stigma related to nontypical genitalia, and the inability of the child to remember the surgery as a traumatic childhood event (Gollu et al., 2007). In addition, early surgery is thought to reduce parental anxiety associated with having a child with nontypical genitalia (Crawford, Warne, Grover, Southwell, & Hutson, 2009). Few studies have included adult women with CAH regarding their satisfaction with having surgery as an infant; however, those who have participated in retrospective studies stated that they preferred early surgery to late surgery. It is possible that this is related to effective coping over time. (Binet, Lardy, Geslin, Froncois-Fiquet & Poli-Merol, 2016; Fagerholm et al., 2011). Furthermore, parents of girls who received early genital surgery have reported that their daughters have satisfactory quality of life and experience a female gender identity (Cassia Amaral et al., 2015; Crawford et al., 2009). It is important to note that for some girls born with CAH, particularly those with a higher Prader score, some type of genital surgery is necessary to prevent genitourinary tract infections and to allow for menstruation as they mature.

Any type of surgery that reduces clitoral size is a delicate one, as without preservation of the neurovascular bundle, the glans, and the preputial skin associated with the glans, sensitivity in the genital area may be comprised, which has the potential to permanently affect sexual satisfaction in adulthood (Gollu et al., 2007). In addition, some of these girls will need additional, corrective surgery as they age into adulthood to successfully wear tampons and have sexual intercourse. Having repeated clitoral surgery has the potential to cause increased damage to sexual functioning and some providers recommend waiting to make the decision to have surgery until the child is old enough to participate in the decision-making process (Creighton, Minto, & Steele, 2001; Gollu et al., 2007). Other studies of adult women who have had cosmetic surgery for genital ambiguity as children found that women report feeling dissatisfied with having had the surgery, not necessarily related to

ultimate cosmetic appearance, but rather sexual satisfaction (Creighton et al., 2001; Gollu et al., 2007; Rangescroft, 2003). Parents of daughters with nontypical genitalia report a struggle between wanting to protect their child and do “what is right,” but realizing that their decision for their child may be in conflict with what their child as an adolescent or adult may have chosen for themselves (Sanders et al., 2012). It is evident in this study and others that the decision regarding whether or not their daughters should have corrective, genital surgery is a very difficult one for parents, filled with concerns over stigmatization, future sexual satisfaction, and gender identity (Crissman et al., 2011; Lundberg, Lindström, Roen, & Hegarty, 2016; Sanders et al., 2012). Health care providers must be aware of this significant family struggle from social, ethical, and practical perspectives and provide not just information, but also compassionate and supportive care to families having a daughter with CAH. In addition, this care should not just be focused on the time surrounding the surgery and the eventual surgical decision. Thoughtful care should continue throughout the child’s life, as early surgical intervention may alleviate some concerns surrounding genital appearance initially, but over time, parents continue to worry about their child’s future sex development and function as well as whether additional surgeries will be needed.

## **Conclusion**

Although there were some limitations to this study such the small sample size and lack of children’s perspectives, it is evident that family management is different, and more complex, for families of girls when compared with boys with CAH. Acknowledging the significant different experiences for families having a boy versus families having a girl with CAH and creating support systems specifically for girls born with CAH that address both the surgical procedures often associated with nontypical genitalia as well as possible long-term complications resulting from being born with elevated testosterone is critical to promoting a healthy family response and child adaptation to the disorder.

Health care providers need to evaluate not only the physical aspects of health for girls with CAH such as growth and endocrine lab values, but also the psychosocial effects related to being born with nontypical genitalia. Nurses are uniquely positioned to assist these families, as they have a commitment to support family and societal health and interact with diverse families in many health care settings (International Family Nursing Association, 2015). In addition, the terminology related to nontypical genitalia that is used by health care providers is critical in shaping not only how parents define their child’s CAH, but also how they perceive the consequences of the

condition for their daughter. When health care providers approach the topic of feminizing genitoplasty, it must be done in a way that allows the decision to rest in the hands of the parents, not the provider. Health care providers have the ability to positively influence caregiver management of their child's condition by motivating and empowering parents, and when providers fail to meet parental expectations, parental distress is possible (McKechnie, Pridham, & Tluczek, 2016). Providers must be careful to ensure that the narrative that is given is without judgment, filled with support regardless of the decision, and allows for adequate time for informed parental decision making. Whether or not parents chose surgery for their daughter with CAH, providers should realize that the decision-making process is inherently stressful and likely is concurrent with learning about the management of a condition that is lifelong and life threatening.

Conducting further research into the possible relationship between parents of girls deciding to keep the condition confidential in an effort to protect the child and family's privacy and the risk of people in the child's social environment not knowing of the diagnosis, thus not responding appropriately during times of acute illness and/or adrenal crisis, is warranted. In addition, recruitment through pediatric endocrinology centers that specialize in treating children with CAH would contribute to a larger, more diverse sample. Finally, longitudinal research exploring the satisfaction of females who had reconstructive surgery as they age is critical, whether it was performed in infancy or into adolescence, so that parents can make surgical decisions based on a more complete understanding of the medical, psychological, and social implications of the surgery.

In summation, future studies with an emphasis on family experiences and management, including differences between families having a boy versus a girl, would enhance the current state of the science and provide a much-needed window into needed prospective interventions aimed at improving the lives of all families and children with CAH.

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