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What about my daughter's future? Parental concerns when considering female genital restoration surgery in girls with congenital adrenal hyperplasia

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Abbreviations
CAH, congenital adrenal hyperplasia; FGRS, female genital restoration surgery; DSD, disorders of sex development

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Summary

Purpose
The parental decision-making process regarding female genital restoration surgery (FGRS) for girls with congenital adrenal hyperplasia (CAH) is controversial and poorly understood. The aim of the study aim was to evaluate parental concerns related to their child's future and parental plans about disclosure prior to FGRS.

Materials and methods
The authors performed an online survey of consecutive parents presenting at a tertiary referral center for consultation regarding FGRS for their daughter with CAH before 3 years of age (2016–2018). Twenty issues initially identified by three families and six clinicians were rated on a 6-point Likert scale of importance ('not at all' to 'extremely').

Results
Sixteen consecutive families participated (Prader 3/4/5: 43.8%/43.8%/12.5%). Fourteen girls (87.5%) subsequently underwent FGRS at a median age of 8 months. Most issues (19/20, 95.0%) were ranked 'quite a bit' to 'extremely' important (Table). Top issues were not surgical: Normal physical/mental development, adrenal crisis and side-effects of medications. Surgery-related and self-image concerns followed in importance. Least prioritized issues were multiple genital exams ('quite a bit' important) and the child not being involved in the decision to proceed with FGRS ('somewhat' important). On average, no issues were considered 'not at all' or 'a little' important.

Disclosure of FGRS to their daughter was the 15th prioritized issues. Almost all families (93.8%, 1 unsure) planned to disclose the surgery to their daughter, although many were unsure when and how to do it (33.3% and 37.5%, respectively).

Comment
Initial efforts to understand the complex process of parental decision-making regarding FGRS in the context of CAH, a complex, multifactorial disease, are presented. Parents of infant girls with CAH simultaneously weigh multiple life-threatening concerns with a decision about FGRS. While issues of genital ambiguity and surgery are important, they are not overriding concerns for parents of girls with CAH. Parents report significant uncertainty about appropriate timing and approach to disclosing FGRS to their daughters. Unfortunately, best practice guidelines for this process are lacking. The findings are not based on actual history of disclosure but on parents' anticipated behavior. Further data are need from parents, children, and women with CAH about successful disclosure. Being a single-center series, these data may not correspond to the wider CAH community.

Conclusions
Parental decision-making regarding FGRS is multifactorial. Even when considering FGRS, parents' largest concerns remain focused on the life-threatening and developmental effects of CAH and side-effects of its medical treatment. The disclosure process deserves further attention.

Table Parent-reported importance of issues prior to female genital restoration surgery (no issue had a mean important score below 'somewhat' important, $n = 16$).

Rank	Category	Issue	Importance score	Importance category
1	Developmental	Normal physical and mental development	92.5	Extremely (90.0–100.0)
2	Medical	My child having an adrenal crisis	88.8	Very much (70.0–89.9)
3	Medical	Side-effects from medications	88.8	
4	Surgery/genital	My child's future ability to have children	85.0	
5	Surgery/genital	My child's future ability for sexual intercourse	80.0	
6	Developmental	My child having problems with her self-image	80.0	
7	Surgery/genital	My child having a complication after surgery	78.8	
8	Developmental	Not letting CAH define her life	77.5	
9	Surgery/genital	General appearance of my child's genitalia	76.7	
10	Surgery/genital	My child needing possible future surgery	76.7	
11	Surgery/genital	Decreased sensation of my child's clitoris	72.5	
12	Developmental	My child's future gender identity	67.5	Quite a bit (50.0–69.9)
13	Medical	Privacy about my child's medical history	65.0	
14	Surgery/genital	Size of my child's clitoris	65.0	
15	Surgery/genital	Disclosing the surgery to my child	63.8	
16	Family stress	Stress on our family	61.3	
17	Developmental	How other people will view my child	58.8	
18	Developmental	My child finding love and acceptance in the future	57.5	
19	Surgery/genital	My child having multiple genital exams by doctors in the future	50.0	
20	Surgery/genital	My child not having had a voice in choosing surgery	28.8	Somewhat (30.0–49.9)
n/a	n/a	n/a	n/a	A little (10.0–29.9)
n/a	n/a	n/a	n/a	Not at all (0.0–9.9)

CAH, congenital adrenal hyperplasia.

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Introduction

Congenital adrenal hyperplasia (CAH) is the most common cause of ambiguous genitalia in newborns [1—3]. The parental decision-making process regarding female genital restoration surgery (FGRS) for girls with CAH is controversial and poorly understood. The timing of FGRS is controversial due to ethical concerns about patient and parental rights [4], and the fact that very limited reliable data exist to support either an early or delayed surgical approach [5,6].

Voices of parents of girls with CAH are largely absent from the discussion of the most appropriate approach to FGRS for females with CAH. This is despite the fact that it is the parents who first face this challenging decision, which will significantly impact their daughter's future, regardless of what choice they make. In addition, regardless of whether they opt for FGRS in infancy or allow their daughter to decide for herself later in life, it is also the parents who will bear the burden of explaining their decision to their daughter when she is older. Their views should, therefore, be taken into account in this debate.

The aim of this study was to evaluate parental concerns related to their daughter's future and parental plans about disclosure prior to FGRS. The authors hypothesized that parents' primary concerns would be focused on genital ambiguity and FGRS and that most parents would be uncertain about when and how to approach the disclosure process with their daughter in the future.

Methods

An Internal Review Board (IRB)-approved cross-sectional questionnaire was administered to consecutive parents scheduling a consultation at a tertiary referral center regarding FGRS for their daughter with CAH before 3 years of age (2016—2018). After providing informed consent, eligible participants were emailed an individualized link to the online survey, with a reminder emailed a week later. Each family provided a single set of answers. Study data were managed using REDCap, a secure web-based platform [7].

Parental concerns prior to FGRS

The questionnaire listed 20 issues identified by three families, one nurse, and five pediatric urologists with experience in the care of patients with CAH during a series of semistructured interviews carried out in person and over the phone with one of the authors (K.M.S.). Interviews continued until the saturation point was reached, and no new issues were identified (mean time per interview: 20 min). Issues were rephrased in an iterative fashion during the interviews to ensure comprehension and clarity. Ten of the issues were related to FGRS and genitalia, while others were related to development (6 issues), medical care (3), and family stress (1).

Participants were asked to rate each issue in response to the question: 'Looking at your child's future as she grows up, how important are these concerns to you?' Responses were rated on a 6-point Likert scale of importance ('not at all' to 'extremely'). For analysis, answers were converted to numerical values ('not at all' = 0, 'a little' = 20, 'somewhat' = 40, 'quite a bit' = 60, 'very much' = 80, and 'extremely' = 100). Mean scores for each issue were calculated and used to rank their relative importance.

Anticipated parental disclosure of FGRS

Based on semistructured interviews regarding parents' plans regarding telling their daughter about her CAH surgery, the anticipated timing and approach to and impact of disclosure were assessed. First, parents were asked about the age at which they planned to disclose FGRS to their daughter, starting with the first mention and then when they would provide all details (<9 years old, 9—11, 12—14, 15—17, or 18 and older).

Parents were then asked how they planned to frame the disclosure to their daughter. Yes/no options included 'I believed it was best not to wait,' 'It was a problem that needed to be fixed,' 'not sure,' and 'other,' which was a free-text option.

Finally, parents reported the level of their concerns about possible consequences of disclosure to their daughter. These included fears that telling a child about her CAH surgery will (1) confuse her, (2) change how she feels about herself (sexual identity, body integrity, self-worth, etc.), (3) about her parents, and (4) about her doctors. Level of concern was noted on a 5-point Likert scale was used ('not at all' to 'very much').

Results

Population characteristics

Parents of 16 consecutively assessed girls with salt-wasting 21-hydroxylase deficiency participated, completing the questionnaire at a median of 3 days prior to FGRS (interquartile range 2—7). Overall, 81.3% of children were white, 12.5% Asian, and 6.3% African American, while 75.0% had private and 25.0% had public insurance. One family refused to participate. Half the children were Prader 3 (43.8%), followed by 4 (43.8%) and 5 (12.5%). Fourteen (87.5%) girls underwent FGRS at a median age of 8 months (range 6 months—3 years), an 11-month's parents have not yet decided about FGRS, and a 19-month-old did not undergo it due to other, unrelated medical problems. Among those who underwent surgery, 12 underwent vaginoplasty (posterior skin flap: 7, posterior sinus flap: 4 [8], pull-through: 1) and all a partial urogenital sinus mobilization, clitoroplasty and labioplasty.

Parental concerns prior to FGRS

Most issues (19/20, 95.0%) were ranked 'quite a bit' to 'extremely' important (Table). Top issues were not surgical: Normal physical/mental development, having an adrenal crisis, and side-effects of medications. Surgery-related concerns and family stress followed in importance. Most prioritized surgery-related issues included (in order) fertility, ability to have future intercourse, complications, genital appearance, requiring future surgery, and

Table 1 Age at which parents anticipate to disclose genital surgery to a daughter with CAH ($n = 15$, one family did not answer this question).

Anticipated age (years)	First tell child about genital surgery	Disclose all the details about genital surgery
Younger than 9	1	0
9–11	7	3
12–14	2	5
15–17	0	1
18 or older	1	1
Not sure	4	5

CAH, congenital adrenal hyperplasia.

decreased clitoral sensation. Developmental concerns about the child's self-image and not letting CAH define her life and future gender identity were similarly highly ranked. Only then did concerns regarding privacy, clitoral size, disclosure of genital surgery, and family stress follow. The two least prioritized issues were multiple genital exams ('quite a bit important') and the child not being involved in the decision to proceed with FGRS (only issue to be 'somewhat important').

Anticipated parental disclosure of FGRS

Disclosure of FGRS to their daughter was the 15th prioritized issue. Almost all parents (93.8% [15/16]) planned to disclose the surgery to their daughter. One family (6.2%) was unsure whether to disclose, and no family (0.0%) planned withholding disclosure altogether. Many families were unsure when and how to disclose FGRS (33.3% and 37.5%, respectively). Among those who suggested potential ages, the most common age to start the conversation about FGRS was 9–11 years old, with full disclosure of all details at 12–14 years old, although suggested ages varied (Table 1).

Regarding how best to frame reasons for choosing surgery during disclosure to their daughter, most parents (62.5%) thought the idea that 'I believed it was best not to wait' would help in the disclosure process. Fewer (25.0%) thought the idea that 'It was a problem that needed to be fixed' would be helpful. A third (37.5%) thought neither idea would be helpful.

Concerns regarding four potential consequences of disclosing FGRS were assessed. A third of parents (37.5%) had completely no concerns that disclosure would confuse their daughter, change her self-perception, or relationship with her parents (Table 2). Most parents (62.5%) were not concerned it would affect how she felt about physicians. A third of parents (31.3%) were 'quite a bit' to 'very much' concerned about how the disclosure would change their daughter's self-perception, and 18.8% reported similar levels of concern regarding confusing her and her perception of her parents. No parents (0.0%) reported similar levels of concern regarding her perception of physicians.

The one family which was unsure about disclosure was 'very much' concerned that it would confuse her and change how she feels about herself and 'somewhat' concerned about a disclosure changing how she feels about her parents and doctors.

Discussion

Parents of infants and toddlers with CAH considering FGRS have multiple significant concerns. Contrary to the study's hypothesis, the most significant issues prior to surgery focused on the life-threatening and developmental effects of CAH as well as side-effects of its medical treatment, rather than surgery. As suspected, much uncertainty exists among parents about disclosing FGRS to their daughters. A third of parents were unsure about the timing, and half were unsure about the best approach. This is consistent with a previous report that 34% of parents of children, mostly boys, after urological surgery would find disclosure guidance helpful [9]. Similarly, most parents of children conceived using assisted reproductive technologies are also unclear about the timing and approach of disclosure [10]. Unfortunately, best practice guidelines for the disclosure process are lacking. Aspiring quality, patient- and family-centered care of children with CAH, the authors' hope this data will help in future work to develop aids for parents in the decision-making process regarding FGRS and subsequent disclosure of the decision to their daughters.

Women with CAH and families of children with CAH identify as a separate entity from disorders of sex development (DSD) [11,12]. As the findings underline, issues of genital ambiguity in CAH exist in a complex context of

Table 2 Parental concerns about disclosure of genital surgery to a daughter with CAH ($n = 16$).

Parental concerns about disclosure	Not at all	A little	Somewhat	Quite a bit	Very much
Telling my child about CAH surgery will confuse her.	6 (37.5%)	4 (25.0%)	3 (18.8%)	2 (12.5%)	1 (6.3%)
Telling my child about CAH surgery will change how she feels about herself (sexual identity, body integrity, self-worth, etc.).	6 (37.5%)	4 (25.0%)	1 (6.3%)	2 (12.5%)	3 (18.8%)
Telling my child about CAH surgery will change how my child will feel about her parents.	6 (37.5%)	3 (18.8%)	4 (25.0%)	1 (6.3%)	2 (12.5%)
Telling my child about CAH surgery will change how they will feel about doctors.	10 (62.5%)	4 (25.0%)	2 (12.5%)	0 (0.0%)	0 (0.0%)

CAH, congenital adrenal hyperplasia.

medical and developmental concerns directly linked to the underlying metabolic abnormality. Even immediately before undergoing FGRS, when one would expect surgery-related concerns be at an all-time high, parents were most concerned about developmental and medical issues for their daughter. Parents of girls with CAH must simultaneously juggle concerns about a life-threatening possibility of an adrenal crisis and unknown effects of long-term medical management with a decision about FGRS. It appears that while issues of genital ambiguity and surgery are very important, they are not the overriding concerns for parents of girls with CAH.

It is now widely recognized that full disclosure of the decision to undergo or forgo FGRS in childhood is paramount [13,14]. Early disclosure encourages honesty and transparency, normalizes the child's experience, enhances good psychosocial adjustment, and creates space for open dialog, allowing a young woman to become a more active participant in her care and empowering her to seek attention for possible late complications of surgery [9,15,16].

Rather than considering disclosure of a sensitive topic as a single event at a particular age, it is better thought of as an age-appropriate and developmentally appropriate process [14,15,17,18]. On average, parents of girls with CAH anticipated initiating and completing full disclosure between 9 and 14 years of age. This concurs with the suggested guidelines of the Sexuality Information and Education Council of the United States modified for children with DSD, which suggest for disclosure process to start after 8 years of age and continue through pre-adolescence/adolescence [18]. This age appears to be somewhat older than that the mean age of 9 years reported by parents of children undergoing urological surgery [9]. Importantly, most children in that study were boys, and few, if any, had genital ambiguity.

In this study, the majority of parents of girls with CAH reported at least some concern about the effect disclosure may have on her self-perception and their relationship with her. This is in contrast to parents of mostly boys undergoing urological surgery, where only 14% were worried about the disclosure process [9]. Not surprisingly, this suggests that parental concerns about disclosure of FGRS may be quite distinct from other urological procedures.

Almost all parents in this study planned on telling their daughter about the surgery, with only one family being unsure (7%). This appears to be lower than the 20% of 20 parents of boys after hypospadias surgery who did not plan a disclosure [9]. This difference was not statistically significant given the small size of both study populations (Fisher's exact test: $P = 0.37$). At the same time, as the authors of the study suggest, parents may be more likely to disclose more significant genital surgery to children, as only parents after distal shaft hypospadias, and not more proximal repairs, planned on no disclosure.

The concerns assessed in this study were identified by parents and urologists and therefore reflect wording they proposed. Therefore, some overlap exists between individual concerns. Further work will be required to more precisely and exhaustively define them.

Since this study is a small single-center series using a non-validated questionnaire, a larger study is required to determine if findings of this study are generalizable to the wider CAH community. Children presenting for consultation to the tertiary referral center are likely more virilized, and their parents are more interested in FGRS. Since almost all parents opted for FGRS, it is unclear if their responses would correspond to those of parents who forgo FGRS in infancy or toddlerhood. Although a small series of parents of girls with CAH are presented, almost all consecutive families participated, minimizing potential selection bias.

As data were collected close to surgery, it is plausible that counseling received from the multidisciplinary team at the authors' institution may have influenced parental opinions. The fact that the majority of families arrive for their consultation with a prior broad, in-depth understanding of the issues pertaining to CAH, including FGRS, would suggest otherwise. In addition, the author's team spends a considerable amount of time with the family discussing the controversies of decision-making regarding FGRS. Although the authors do not suspect this to be occurring, if postcounseling bias overemphasizing parental concerns about FGRS was indeed present, actual parental concerns in this area may be lower.

Importantly, this work represents initial efforts to understand the complex process of disclosure of a sensitive issue like FGRS in the context of a complex, multifactorial disease such as CAH. It is recognized that the findings are not based on actual history of disclosure but are rather parent's anticipated behavior. Further data are needed from parents who had completed disclosure to their daughters, its timing, and rationale. In order to develop best practice guidelines in this area, successful and less than successful disclosure approaches need to be delineated based on perspectives from parents, children, and, most importantly, women with CAH.

Conclusions

Parental decision-making regarding FGRS is multifactorial. Even when considering FGRS, parents' largest concerns remain focused on the life-threatening and developmental effects of CAH and side-effects of its medical treatment. The disclosure process deserves further attention.

Author statements

Ethical approval

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None declared.

Internal review board approval

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