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Majority of females with a life-long experience of CAH and parents do not consider females with CAH to be intersex

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Abbreviations
CAH, congenital adrenal hyperplasia; IQR, interquartile range; DSD, differences of sex development

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Summary

Introduction
To assess opinions of females with CAH, and parents of females with CAH, about designating this population "intersex," particularly in legislation about genital surgery during childhood.

Methods
We conducted a mixed-methods (quantitative and qualitative) anonymous cross-sectional online survey of females with CAH (46XX, 16+years old) and independently recruited parents of girls with CAH (2019–2020) diagnosed in first year of life from the United States. A multidisciplinary CAH team drafted the survey in collaboration with women with CAH and parents. Fisher's exact test was used to compare female and parent responses. A qualitative thematic approach was used to analyze open-ended answers for emergent categories of reasons why CAH females should or should not be considered as intersex.

Results
Of 57 females with CAH participating (median age: 39 years, 75.5% of ≥25year olds had post-secondary degree), all had classical CAH and 93.0% underwent genital surgery at median 1–2 years old. While 89.5% did not endorse the intersex designation for CAH, the remaining 5.3% did (5.3% provided no answer, Summary Figure). Most CAH females (63.2%) believed CAH females should be considered separately in "any laws banning or allowing surgery of children's genitals" (19.3% disagreed, 17.5% neutral, 0.0% no answer). Most common themes identified by females with CAH not endorsing an intersex designation were: normal female internal organs, sex chromosomes, personal identity, genital appearance, issues with language, hormones, and those endorsing it: genital appearance, community/group experiences, topic complexity.

Overall, 132 parents of females with CAH participated (parent/child median ages: 40/11 years, 81.7% of ≥25year olds had post-secondary degree). All children had classical CAH and 78.8% underwent surgery at median <1 year old. While 95.5% of parents did not endorse the intersex designation for CAH, 2.3% did (2.3% no answer), similar to females ($p = 0.29$). Most parents (81.1%) believed CAH females should be considered separately in legislation (9.1% disagreed, 6.1% neutral, 3.8% no answer), a slightly higher percentage than females ($p = 0.01$).

Discussion
Echoing previously published disagreement with clinically designating CAH females as intersex, majority of CAH females and parents oppose a legal intersex designation. Differing opinions among females and parents strengthen concern about a one-size-fits-all approach to legislation about childhood genital surgery. Differences in opinions between female and parent responses, while statistically significant, were relatively small.

Conclusion
Majority of females with CAH and parents believe CAH should be excluded from the intersex designation, and should be considered separately in legislation pertaining to childhood genital surgery.

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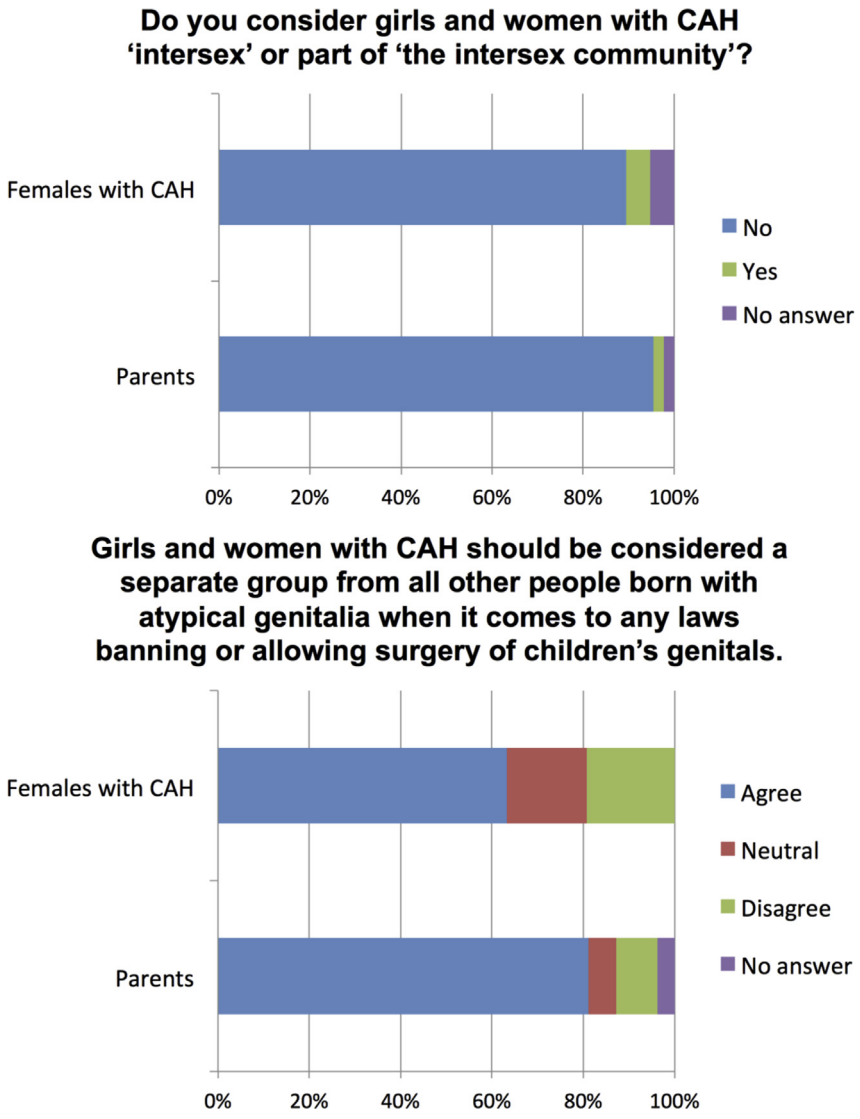

Summary Figure Opinions about females with CAH being legally designated as intersex.

Introduction

Congenital adrenal hyperplasia (CAH) is an endocrine disorder, which, when present in females (46XX), is the most common cause of atypical genitalia in newborns [1]. Several publications report that females with CAH consider themselves distinct from the heterogeneous group classified under the umbrella term Differences of Sex Development (DSD) [2,3]. Ignoring this physiological and self-declared distinction, several legislatures recently proposed sweeping changes to the clinical care of children born with DSD, typically referring to this group as intersex, a term no longer in clinical use [4—6]. Importantly, no studies previously investigated the attitudes of CAH females, or any other group born with atypical genitalia, toward being legally designated as intersex, subsequently dictating medical and surgical care.

CAH is a heterogeneous condition. Females with classic CAH, rather than non-classic CAH, tend to be diagnosed as newborns, are far more likely to have atypical genitalia and a more severe condition, likely leading to different life experience of CAH [1]. Studies pertaining to atypical genitalia should exclude people without CAH, CAH males (46XY) and CAH females diagnosed later in life, as they typically present without atypical genitalia and would be unaffected by these laws.

We aimed to assess opinions of females with CAH, and parents of females with CAH, regarding being designated as intersex, particularly with respect to laws about childhood genital surgery. We also sought to explore reasons for these opinions using qualitative methods. Based on studies demonstrating their preference for diagnosis-specific clinical care, we hypothesized that most participants would oppose CAH females being designated intersex, particularly with respect to legislation regarding their care.

Methods

We conducted an IRB-approved, anonymous online survey of CAH females (46XX) $\geq$16 years old and parents of CAH

girls living in the United States (2019—2020). To capture attitudes of those most affected by potential legal designations, we restricted analysis to females diagnosed in the first year of life, those most likely to be born with virilized genitalia.

Study group focused on patient- and parent-centered outcomes

The Life with Congenital Adrenal Hyperplasia Study Group formed in 2019 to explore patient-/parent-centered CAH research topics. People directly affected by CAH, rather than clinicians or researchers, deemed these topics, including the current study, as both important and insufficiently researched (research questions were prioritized by people with CAH, not clinicians caring for them). Topics were identified during a series of focus groups of 60 participants living with CAH (children, adults, parents) held in Indianapolis, Indiana over a 2-day period in 2016 (unpublished). Sessions were facilitated and data subsequently analyzed by the Patient Engagement Core at Indiana University, which specializes in applying user-centered design to health services research and creating patient-centered study interventions [7,8].

Questionnaire development

The questionnaire addressed multiple research questions, to be reported separately. It was drafted by a multidisciplinary international medical team (endocrinologists, psychologist, ethicist, sexual/reproductive health researcher, urologists and nurses involved in CAH care, see **Acknowledgements**). Question phrasing and comprehension were finalized by iterative feedback from women with CAH (n = 3) and parents (n = 3).

Questionnaire content

The questionnaire was similar for CAH females and parents, with one additional question (#2) for females. Questions detailed demographics and CAH care, allowing for a CAH classification. We classified as classical CAH those females: diagnosed in the first 5 years of life by atypical genitalia or adrenal crisis, and those diagnosed at a later time (or for a different reason) and taking fludrocortisone or salt. Discrepancies were resolved by consensus among endocrinologists.

Questions were introduced by the stem: "When it comes to making laws, some people believe that girls and women with CAH are part of a larger group of all people born with atypical genitalia. They often refer to this larger group as 'intersex' or 'the intersex community.' They believe that the same laws, such as bans on surgery, should apply to everyone who they label as 'intersex,' including girls and women with CAH."

Question 1: "Do you consider girls and women with CAH a part of a group of people often referred to as 'intersex' or 'the intersex community'?" (answers: yes, no).

Question 2: "Why do you feel that females with CAH are/are not intersex?" (free text).

Question 3: "Girls and women with CAH should be considered a separate group from all other people born with atypical genitalia when it comes to any laws banning or allowing surgery of children's genitals" (3-point Likert scale: disagree, neutral, agree).

Questionnaire administration and security

The questionnaire was serially distributed to four groups living with CAH: three multidisciplinary CAH clinics in the United States and CARES Foundation members. CARES Foundation is the largest patient and family advocacy group for people affected by CAH in the world. Recruitment through clinics in the Midwest, East and West coasts of the United States opened the study to individuals potentially not affiliated with this advocacy group. Recruitment was not offered to non-CAH specific support groups to limit participation to only those with CAH and prevent inclusion of those without CAH.

We took precautions to safeguard the integrity of the questionnaire during questionnaire design, administration and analysis [9]. Study data were managed using Qualtrics, an online platform for building and managing online surveys and databases. To minimize false entries, we offered no incentive payments, embedded testing questions to detect automatic data entry and the questionnaire was lengthy, discouraging poorly motivated fraudsters (34.9% response rate, median completion: 24min., 79.3% completion rate). Recruitment of each group occurred separately, 1—2 months apart. The questionnaire was accessible using a password-protected link, which was live only for a week. A follow-up invitation was emailed a month later with another individualized, password-protected link, which was live for one week. During analysis, serial repeating answers and IP addresses were screened to detect data dumping (none observed).

Qualitative analysis of statements by females with CAH

We used a thematic approach to analyze the content of open-ended answers to examine emergent categories of why females with CAH do or do not believe that females with CAH are intersex [10]. All open-ended answers were downloaded onto a spreadsheet and carefully examined. Responses with multiple sentences or statements were separated for unique analysis. A total of 92 statements were contained in 64 sentences. Each statement was categorized into nine categories by one researcher (DJH). These were discussed by two researchers (DJH, KMS) with knowledge and expertise of the clinical and social experiences associated with CAH. Seventeen statements (18.5%) were reclassified based on this discussion. After finalizing this process, the categorical distribution of all statements was examined.

Statistics

National reference values were obtained from 2018 United States Census [11]. Non-parametric statistics were used:

Fisher's exact test for categorical and Wilcoxon rank sum test for continuous data. A critical $p = 0.05$ was used (software: Stata, StataCorp, College Station, TX, USA).

Results

Females with CAH

Median age of 57 females with CAH participating was 39 years (IQR 27–48). Overall, 87.7% were white, slightly higher than 76% nationally ($p = 0.04$) (Table 1). They lived in 24 states. Specifically, 35.1% lived in the 5 most populous states (California, Texas, Florida, New York, Pennsylvania), which represent 37% of the country's population. This was consistent with national census data ($p = 0.79$). Among those at least 25 years old, 75.5% had post-secondary degree (higher than national values, $p < 0.001$). Median annual household income was $60,000–79,999, similar to the national average ($p = 0.60$). Whether participants lived with parents, spouse/partner or alone was not assessed. One in six participating women was recruited through a clinic, remaining 84.3% were recruited through the CARES Foundation.

All females had classical CAH. Overall, 93.0% underwent genital surgery at median 1–2 years old. Most commonly, surgery involved both the vagina and clitoris (66.7%). Another 21.1% had only vaginal and 1.5% had only clitoral surgery.

While 89.5% of females with CAH did not endorse the intersex designation for CAH, 5.3% did (5.3% provided no answer, Summary Figure). Most CAH females (63.2%) believed CAH females should be considered separately in "any laws banning or allowing surgery of children's genitals" (19.3% disagreed, 17.5% neutral, 0.0% no answer). There were no statistically significant differences between responses from females recruited via CARES Foundation vs. clinics ($p \geq 0.66$).

More written feedback was provided against the intersex designation (81 statements, 88.0% of all statements) rather than supporting it (11, 12.0%) (Table 2). Most evidence against an intersex designation focused on comparisons to existing knowledge: normal female internal organs (22.2%, example quote: "Internally, they have all the female parts such as ovaries, uterus, vagina"), genetics/sex chromosomes (17.3%, "They are 46XX") and personal identity (13.6%, "We are born women and we are women"). Other themes included genital appearance (12.3%, "Females with CAH have a large range of genitalia"), issues with language (11.1%, "I don't think intersex is a useful or compassionate term because it [labels her] as other") and hormones (9.9%, "[Females with CAH are] all female except for their hormones being unbalanced in the early weeks [of pregnancy due to] … malfunction of the adrenal gland"). Remaining themes included medical need/surgery (4.9%), comparison to boys with CAH (2.5%), community/group experiences (2.5%), politics (2.8%) and topic complexity (1.2%).

Evidence in favor of females with CAH being intersex focused on genital appearance (45.5%, "Their genitalia fall somewhere between a textbook man and a textbook woman"), community/group experiences (18.2%, "[If they] choose the term intersex then they are part of that community") and topic complexity (18.2%, "I think this depends strongly on the situation and is a more nuanced question than a simple yes or no answer") (Table 2). Remaining themes included medical need/surgery and issues with language (9.1% each).

Parents of females with CAH

A total of 132 parental responses were analyzed (81.8% mothers, 12.9% fathers 4.6% both, 0.8% female guardian, Table 1). Median parental age was 40 years old (IQR 35–48). About 83.3% were white, similar to national values ($p = 0.06$). Parents lived in 37 states. Specifically, 43.9% lived in the five most populous states, which was consistent with national census data ($p = 0.13$). Among parents 25 years old or older, 81.7% post-secondary degree (higher than national values, $p < 0.001$). Median annual household income was over $100,000, higher than national average ($p < 0.001$). Parents and females with CAH were comparable in terms of age, race, ethnicity, language spoken at home, state of residence and education level ($p = 0.07$). One in four participating parents were recruited through a clinic, while 77.3% were recruited through the CARES Foundation.

As parents answered the survey, their daughters' median age was 11 years (IQR 7–18.5). The daughters all had classical CAH and 78.8% underwent genital surgery at median age <1 year old. Most common surgery involved both the vagina and clitoris (59.4%). Another 18.2% had only vaginal and 0.8% had only clitoral surgery.

While 95.5% of parents did not endorse the intersex designation for CAH, 2.3% did (2.3% no answer), similar to females with CAH ($p = 0.29$) (Summary Figure). Most parents (81.1%) believed CAH females should be considered separately in legislation (9.1% disagreed, 6.1% neutral, 3.8% no answer), a slightly higher percentage than females ($p = 0.01$). Slightly more parents recruited via CARES Foundation vs. clinics disagreed with an intersex designation (98.0% vs. 86.7%, $p = 0.01$), but similar percentages supported a separate legal designation (84.3% vs. 70.0%, $p = 0.10$).

Discussion

We report findings of quantitative and qualitative analysis of responses directly from individuals with a lived experience of CAH, who represent the largest group born with atypical genitalia. First, the majority of females with CAH (9 out of 10), and the majority of parents of females with CAH (19 out of 20), oppose being included under an intersex designation. This was in line with results by Binet et al.: 86% of 21 CAH females born with atypical genitalia did not consider themselves intersex [12]. The majority also do not endorse being considered as intersex in legislation regarding genital surgery in childhood. Second, differences of opinion exist both among females and among parents. This lack of a unanimous view by groups who would be most affected by potential legislation strongly supports opposing any blanket legal measures regarding childhood genital surgery. Third, while agreement exists between the majority of females with CAH and majority of parents of

Table 1 Participant characteristics.

Variable	Females with CAH (n = 57)	Parents of females with CAH (n = 132)	p-value
Age (median, IQR, years)	39 (27–48)	40 (35–48)	0.17
Type of parent			
Mother	n/a	108 (81.8%)	n/a
Father		17 (12.9%)	
Both mother and father		6 (4.6%)	
Female guardian		1 (0.8%)	
Age of child with CAH (median, IQR, years)	n/a	11 (7–18.5)	n/a
Race of person with CAH			
White	50 (87.7%)	110 (83.3%)	0.76
Black or African American	0 (0.0%)	2 (1.5%)	
Asian	2 (3.5%)	5 (3.8%)	
Native Hawaiian or Pacific Islander	0 (0.0%)	1 (0.8%)	
More than one race	2 (3.5%)	9 (6.8%)	
Other	1 (1.8%)	4 (3.0%)	
Prefer not to answer	2 (3.5%)	1 (0.8%)	
Hispanic/Latino	4 (7.0%)	11 (8.3%)	0.99
Primary language at home			
English	56 (98.3%)	131 (99.2%)	0.51
Spanish	1 (1.8%)	0 (0.0%)	
Other	0 (0.0%)	1 (0.8%)	
State of residence			
California	6 (10.5%)	14 (10.6%)	0.25
Texas	4 (7.0%)	12 (9.1%)	
Florida	5 (8.8%)	6 (4.6%)	
Pennsylvania	4 (7.0%)	13 (9.9%)	
New York	1 (1.8%)	13 (9.9%)	
Alabama, Arizona, Arkansas, Colorado, Connecticut, District of Columbia, Georgia, Idaho, Illinois, Indiana, Iowa, Kentucky, Louisiana, Maryland, Massachusetts, Michigan, Minnesota, Mississippi, Missouri, Nebraska, New Jersey, North Carolina, Ohio, Oklahoma, Oregon, South Carolina, Tennessee, Utah, Virginia, Washington, West Virginia, Wisconsin	37 (64.9%)	74 (56.1%)	
Annual household income			
<20,000	2 (3.5%)	2 (1.5%)	<0.001
20,000–39,999	18 (31.6%)	6 (4.5%)	
40,000–59,999	5 (8.8%)	10 (7.6%)	
60,000–79,000	6 (10.5%)	9 (6.8%)	
80,000–99,999	9 (15.8%)	16 (12.1%)	
100,000 or more	11 (19.3%)	76 (57.6%)	
Not sure or prefer not to answer	6 (10.5%)	13 (9.8%)	
Educational level among those 25 years old or older			
Less than high school	0 (0.0%)	0 (0.0%)	0.09
High school or equivalent	4 (4.1%)	9 (6.9%)	
Some college	10 (20.4%)	15 (11.5%)	
Associate degree	8 (16.3%)	9 (6.9%)	
Bachelor degree	18 (36.7%)	50 (38.2%)	
Graduate degree	11 (22.5%)	48 (36.6%)	
Currently studying	8 (14.0%)	7 (5.3%)	0.07
Classical CAH	57 (100.0%)	132 (100.0%)	0.99
History of genital surgery			
Yes	53 (93.0%)	104 (78.8%)	
Vaginal and clitoral surgery	38 (66.7%)	79 (59.4%)	

Table 1 (*continued*)

Variable	Females with CAH (n = 57)	Parents of females with CAH (n = 132)	p-value
Vaginal only	12 (21.1%)	24 (18.2%)	
Clitoral only	3 (1.4%)	1 (0.8%)	
No	3 (5.3%)	23 (17.4%)	
Not sure or prefer not to answer	1 (1.8%)	5 (3.8%)	
Age at surgery			
<1 year old	20 (37.8%)	74 (71.2%)	
1–2 years old	13 (24.5%)	17 (16.3%)	
2–3 years old	6 (11.3%)	4 (3.8%)	
3–4 years old	3 (5.7%)	1 (1.0%)	
4–6 years old	0 (0.0%)	3 (2.9%)	
6–9 years old	0 (0.0%)	1 (1.0%)	
9–17 years old	5 (9.4%)	1 (1.0%)	
18 years old or older	1 (1.9%)	1 (1.0%)	
Not sure	5 (9.4%)	2 (2.0%)	

females with CAH, small differences in opinion exist. Future research is needed to verify and explore these.

We were surprised that while the vast majority of both females with CAH and parents did not identify with the intersex designation, a slightly smaller majority preferred being considered a legally separate group. The reason for this is unclear, but may be related to question wording. The question about a legal designation did not specifically use the term intersex; some participants may not have connected their degree of support for an intersex designation to the question about a legal designation, or vice versa. Additionally, many people affected with CAH have a desire not to be treated as a group separate from the general population. Some participants may have simply expressed not wanting to be considered a separate group in general, rather than a separate group from the intersex community.

Our results provide the first insights into why females with CAH may support or oppose an intersex designation. Most of the 90% of females with CAH who opposed this designation did so due to normal female internal anatomy, normal female sex chromosomes, personal experience of female identity and/or CAH being solely due to a hormonal imbalance. On the other hand, most of the 5% of females with CAH who favored an intersex designation cited atypical genital appearance and group/community experiences as evidence. It appears that individuals emphasize different aspects of life with CAH, underlining that no single approach applies to everyone. These findings also raise questions of how clinical, personal and family factors influence the self-perception of a young woman living with CAH.

In medicine, people presenting similarly, whether with loss of consciousness, chest pain or fever, are managed based on the underlying cause, not their presentation. It may be detrimental to people's health if an endocrine disease like CAH would be managed like other diagnoses under the DSD umbrella simply because females present with atypical genitalia. Given its unique pathophysiology [1] and psychosocial impact [3,13], it would be prudent to consider CAH separate from DSD in order to provide appropriately individualized multidisciplinary care.

While the 2006 Chicago Consensus Statement provided an umbrella term DSD [14], several papers published since then underline a lack of consensus in its use. Our results corroborate previous studies demonstrating that affected individuals, and their families, prefer diagnosis-specific designations. Lin-Su et al. reported that 84% of females with CAH, and parents, did not identify with an undifferentiated DSD designation [2]. Our group previously reported that parents of girls with CAH preferred a diagnosis-specific designation for CAH [3]. In addition, Johnson et al. found that 81% of individuals with DSD diagnoses other than CAH, and their parents, also prefer diagnosis-specific nomenclature for self-identification [15].

It is important to note that 63% of females with CAH and 81% of parents in our study did not endorse a legal intersex designation for CAH. It is possible, although unproven, that the legal designation may be more acceptable to individuals with atypical genitalia not due to CAH. This may be indicated by the finding of Johnson et al., that the majority of people with DSD diagnosis other than CAH endorse the intersex designation for clinical care or research studies [15]. Future work is needed to determine if groups other than CAH classified as intersex, or DSD, such as complete androgen insensitivity, reject or support a legal intersex designation.

There has been concern that legislative opinions come from physicians rather than those living with the condition. Ours is the first study directly reporting opinions of people living with CAH regarding being designated as intersex in legislation. We strongly believe their experience and opinions should inform any legislation that may affect them. Moreover, their voices should be considered to a higher degree than those of people born without atypical genitalia.

Despite being one of the larger studies of the CAH population, the actual number of participants is relatively small. This is partly because we restricted analysis based on

Table 2 Theme categories of explanations provided by 57 females with CAH about why they feel that females with CAH are or are not intersex (n = 92 statements).

Theme category	Overall (n, %)	Females with CAH are not intersex (81 statements, 88.0% of total)	Example quote	Females with CAH are intersex (11 statements, 12.0% of total)	Example quote
		n (%)		n (%)	
Normal female internal organs	18 (19.6%)	18 (22.2%)	We were born with ovaries and uterus, so that makes us female.	0 (0.0%)	—
Genetics/sex chromosomes	14 (15.2%)	14 (17.3%)	Because they were born with the female sex chromosomes, making them female.	0 (0.0%)	—
Personal identity	11 (12.0%)	11 (13.6%)	We are born women and we are women.	0 (0.0%)	—
Genital appearance	15 (16.3%)	10 (12.3%)	Females with CAH have a large range of genitalia.	5 (45.5%)	Their genitalia fall somewhere between a textbook man and a textbook woman.
Issues with language	10 (10.9%)	9 (11.1%)	I don't think intersex is a useful or compassionate term because it [labels her] as "other," neither male nor female.	1 (9.1%)	If you are altering genitals to fit a "norm" then medicine should consider XX CAH as intersex.
Hormones	8 (8.7%)	8 (9.9%)	[Differences are] simply due to having no cortisol and no access to replacement steroids yet.	0 (0.0%)	—
Medical need/ surgery	5 (5.4%)	4 (4.9%)	Because they can grow up to be perfectly normal if surgery is performed.	1 (9.1%)	Surgery would include surgery to open my daughter's pre-pubescent vagina which she has done at 10.
Comparison to boys with CAH	2 (2.2%)	2 (2.5%)	Are males born with CAH considered [intersex]? No — so how/ why is the exact same medical condition considered to be [different] for females?	0 (0.0%)	—
Community/group experiences	4 (4.3%)	2 (2.5%)	[Intersex] lumps a groups of women with different extremes into one drastic group.	2 (18.2%)	[If they] choose the term intersex then they are part of that community.
Politics	2 (2.2%)	2 (2.5%)	This nonsense needs to stop, and until it does, it will give our politicians free range to make laws as they please	0 (0.0%)	—
Topic complexity	3 (3.3%)	1 (1.2%)	Based on what I know, someone described as "intersex" means that they are any number of variations in the sex/gender characteristics.	2 (18.2%)	I think this depends strongly on the situation and is a more nuanced question than a simple yes or no answer.

gender, age of diagnosis and geography. However, being born with atypical genitalia is a *rare entity* and populations affected tend to be small. Nonetheless, CAH is the *most* common cause of this entity. We hope our collaborative approach can serve as an example of tackling a controversial and politically charged research topic in a vulnerable population.

We believe our results may be generalizable to the female CAH population. Despite most study participants being recruited through an advocacy group, it does not imply direct influence. A significant number of participants were independently recruited through clinics. Moreover, responses from CAH females were similar, regardless of how they were recruited. Presuming the CAH population has similar characteristics to the general population, our study sample certainty overrepresented white and higher educated participants. At the same time, participants, especially the women with CAH, were of diverse ages, socioeconomic groups, receiving medical care at multiple centers with a nationwide geographical distribution, reflecting the general population. Participants were recruited from different social clusters: hospital-based clinics and a patient advocacy group. We hope this multipronged recruitment strategy gave an opportunity to participate to some of "the silent majority" which typically does not participate in studies [16].

Several study limitations were related to the anonymous self-reported data collected. Without access to laboratory or genetic testing, classification of CAH was limited. We were unable to link participant responses to objective clinical measures, particularly the degree of genital virilization at birth. Subsequently, we did not compare responses between females who did and did not undergo genital surgery. Such a comparison would be significantly confounded by virilization at birth. This is because females with more virilized genitalia were more likely to undergo surgery and may hold different opinions about being designated as intersex. Since all responses were entered independently, we could not verify whether any parents and female participants were related.

Conclusion

The majority of CAH females and parents believe CAH should be excluded from the intersex designation. Additionally, CAH should be considered separately in legislation pertaining to genital surgery in childhood.

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Internal review board approval

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Conflicts of interest

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References

[1] Schaeffer TL, Tryggestad JB, Mallappa A, Hanna AE, Krishnan S, Chernausek SD, et al. An evidence-based model of multidisciplinary care for patients and families affected by classical congenital adrenal hyperplasia due to 21-hydroxylase deficiency. Int J Pediatr Endocrinol 2010;2010:692439.

[2] Lin-Su K, Lekarev O, Poppas DP, Vogiatzi MG. Congenital adrenal hyperplasia patient perception of 'disorders of sex development' nomenclature. Int J Pediatr Endocrinol 2015; 2015:9.

[3] Szymanski KM, Salama AK, Whittam B, Frady H, Cain MP, Rink RC. Beyond changing diapers: stress and decision-making among parents of girls with congenital adrenal hyperplasia seeking consultation about feminizing genital restoration surgery. J Pediatr Urol 2019;15:653—8.

[4] SB 201 California: medical procedures: treatment or intervention: sex characteristics of a minor. 2019-2020. ed2020.

[5] SB 408 Nevada: an act relating to health care; establishing conditions for the performance on a child of any surgical procedure to assign anatomical sex; providing for disciplinary action against certain providers of health care for any violation of those conditions; and providing other matters properly relating thereto. 2017. ed2017.

[6] Bill 388 Connecticut: an act concerning a person's intersex status or characteristics. 2019.

[7] Sanematsu H, Hong Y, Cole L, Zollinger T. Listening through seeing: using design methods to learn about the health perceptions of Garden on the Go® customers. Indianapolis, Indiana: IUPUI Research Day; 2014. 2014.

[8] Sanematsu H, Wiehe S. Learning to look: design in health services research. Touchpoint; 2014. July:TP06-2P82.

[9] Teitcher JE, Bockting WO, Bauermeister JA, Hoefer CJ, Miner MH, Klitzman RL. Detecting, preventing, and responding to "fraudsters" in internet research: ethics and tradeoffs. J Law Med Ethics 2015;43:116—33.

[10] Terry G, Hayfield N, Clarke V, Braun V. Thematic analysis. The SAGE Handbook of Qualitative Research in Psychology: SAGE Publications Ltd.; 2017. p. 17—37.

[11] UnitedStatesCensusBureau. United States census bureau. 2020. https://www.census.gov/en.html.

[12] Binet A, Lardy H, Geslin D, Francois-Fiquet C, Poli-Merol ML. Should we question early feminizing genitoplasty for patients with congenital adrenal hyperplasia and XX karyotype? J Pediatr Surg 2016;51:465—8.

[13] Szymanski KM, Whittam B, Kaefer M, Frady H, Cain MP, Rink RC. What about my daughter's future? Parental concerns when considering female genital restoration surgery in girls with congenital adrenal hyperplasia. J Pediatr Urol 2018; 14(5):417.e1—5.

[14] Lee PA, Houk CP, Ahmed SF, Hughes IA. International consensus conference on intersex organized by the Lawson Wilkins pediatric endocrine S, the European society for paediatric E. Consensus statement on management of intersex disorders. International consensus conference on intersex. Pediatrics 2006;118:e488—500.

[15] Johnson EK, Rosoklija I, Finlayson C, Chen D, Yerkes EB, Madonna MB, et al. Attitudes towards "disorders of sex development" nomenclature among affected individuals. J Pediatr Urol 2017;13:608 e1—e8.

[16] Szymanski KM, Cain MP, Rink RC. A proposal for a New approach to differences of sexual development research. J Urol 2020;203:42—3.