

## What do patients and parents say about parents' ability to choose early genital surgery for girls with CAH?

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**Introduction:** To determine opinions of CAH females, and parents, about parent's ability to choose early genital surgery for CAH girls.

**Methods:** Online survey of CAH females (46XX, 16+ years old) and independently recruited parents (2019-2020) diagnosed in first year of life in the United States. A multidisciplinary medical team, CAH women, and parents drafted the survey. Fisher's exact test was used.

**Results:** Of 57 CAH females (median age: 39 years), 93.0% underwent genital surgery (median 1-2 years old). Most females (79.0%) believed legislation prohibiting surgery in childhood would cause harm. Most (64.9%) believed a ban "would have been harmful to me" (24.6% not harmful, 10.5% neutral). Most females (70.2%) believed a ban undermined parental rights to make medical decisions in their child's best interest. While 75.4% did not believe a ban was in the best interest of CAH females, 14.0% did (10.5% neutral).

For 132 parents of CAH females (parent/child median ages: 40/11), 78.8% of children underwent surgery (median <1 year old). Most parents (93.6%) believed legislation prohibiting surgery in childhood would cause harm. Most (77.3%) believed a ban "would have harmed my daughter" (12.1% no harm, 5.3% neutral, 5.3% no answer). Parents were more likely than CAH females to oppose a ban ( $p \leq 0.02$ ). Most parents (90.9%) believed a ban undermined parental rights. While 93.9% did not believe a ban was in the best interest of CAH females, 3.8% did (2.3% neutral).

**Conclusions:** Majority of CAH females and parents support parents' ability to decide about potential genital surgery, opposing moratoria on surgery in childhood.

**Word count:** 2498

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## Introduction

In the past five years, genital surgery has been scrutinized in children born with atypical genitalia due to disorders of sex development (DSD), sometimes referred to as intersex. Malta and Portugal have banned non-emergent genital surgery in these children.<sup>1, 2</sup> Both the Parliamentary Assembly of the Council of Europe<sup>3</sup> and the United Nations Human Rights Office advised governments to “prohibit medically unnecessary surgery and procedures on the sex characteristics of intersex children.”<sup>4</sup> Several state legislatures in the United States have proposed legislation banning genital surgery in intersex children.<sup>5-7</sup>

Congenital adrenal hyperplasia (CAH) is the most common cause of atypical genitalia in newborns, accounting for ~50% of all cases<sup>8, 9</sup> and ~90% of cases with a 46XX karyotype.<sup>8</sup> Thus, laws restricting surgery in childhood would predominantly impact females (46XX) with CAH. Surprisingly, neither the CAH community, nor any group born with atypical genitalia, has been surveyed regarding potential bans. In other words, laws have been proposed, and even enacted, which disproportionately affect a specific patient population, without having garnered their input.

Studies have found that females with CAH and parents of females with CAH consider themselves distinct from the DSD or intersex designation,<sup>10, 11</sup> wishing to be excluded from any legislation regulating childhood surgery.<sup>12</sup> We hypothesized that the majority of CAH patients and parents would be against prohibiting genital surgery in childhood. We aimed to determine opinions of females with CAH, and parents of CAH females, about parent's ability to choose early genital surgery for girls with CAH.

## Methods

The Life with CAH Study Group methodology has been described.<sup>12</sup> We conducted an anonymous, Qualtrics-based online survey of CAH females (46XX,  $\geq 16$  years old) and parents of CAH females living in the United States (2019-2020). To capture attitudes of individuals most affected by a potential surgical ban, we studied CAH females, and parents of females, diagnosed in the first year of life.

The study aim was previously identified by focus groups of CAH females, and parents of CAH females, as an important research topic deserving further investigation (unpublished). Study questions were generated, reduced and reformatted for content validity by a multidisciplinary international team of endocrinologists, psychologists, ethicist, sexual/reproductive health researcher, urologists and nurses involved in CAH care (see **Acknowledgements**). Questions were pretested with CAH women (n=3) and parents (n=3) to ensure they captured what was intended. Feedback provided by participants at the end of the study supported questionnaire face validity (data not shown).

### *Questionnaire content*

Survey respondents answered four study questions following the stem: “Some advocacy and lobby groups seek laws to ban all genital surgery in children born with atypical genitalia (including CAH) unless they are medically necessary. If such laws were passed and included children with CAH, it is likely that very few, if any, surgeries would be performed in childhood, even if parents believed it would be in the child’s best interest. Instead, these groups propose waiting until children reach an age at which they themselves can provide informed consent for any genital surgery. Please choose the answer that best describes you. You may choose to skip any question that makes you feel uncomfortable by clicking on prefer not to answer.”

Question 1: “What effect would a law prohibiting surgery in childhood have on girls and women with CAH?” (3-point Likert scale: cause harm, neutral, prevent harm)

Question 2: “Do you agree with the statement: If a law prohibiting surgery in childhood was in place when I (or my daughter) was a child, it would have been harmful to me (or my daughter)?” (3-point Likert scale: disagree, neutral, agree)

Question 3: “Do you agree that a law prohibiting surgery in childhood respects rights of parents of girls with CAH to make medical decisions they believe are in the best interest of their child?” (3-point Likert scale: yes it respects parents’ rights, neutral, no it undermines parents’ rights)

Question 4: “Do you agree that a law prohibiting surgery in childhood has the best interest of girls and women with CAH in mind?” (3-point Likert scale: disagree, neutral, agree)

#### *Questionnaire administration and security*

The questionnaire was serially distributed via three multidisciplinary CAH clinics in the Midwest, East, and West coasts of the United States, and to members of the CARES Foundation to open the study to individuals affiliated and not affiliated with an advocacy group. As previously described,<sup>12</sup> multiple design, administration, and analysis safeguards were used to protect the integrity of the study.<sup>13</sup> We provided no incentive payments, embedded testing questions to detect automatic data entry, and used a long questionnaire to discourage poorly motivated fraudsters (response rate: 34.9%, completion rate: 79.3%, median completion time: 24min.). Each group was recruited separately, 1-2 months apart, via unique time-sensitive password-protected links. Secure follow-up invitations were sent one month later. No serial repeating IP addresses or answers were observed.

#### *Statistics*

National reference values were obtained from 2018 United States Census.<sup>14</sup> Since some data suggest responses may differ based on support group affiliation, we compared these groups.<sup>15</sup> Non-parametric statistics were used: Fisher’s exact and Wilcoxon rank sum tests ( $p=0.05$ , Stata, StataCorp, College Station, TX, USA).

## **Results**

### *Females with CAH*

Fifty-seven females with CAH were enrolled (median age: 39 years, IQR 27-48) (**Table 1**).

They lived in 24 states, with 35.1% residing in California, Texas, Florida, New York, and Pennsylvania, mirroring national census data ( $p=0.79$ ). Among females 25 years old or older, 75.5% had a post-secondary degree (above national values,  $p<0.001$ ). Median annual household income was similar to the national average (\$60,000-79,999,  $p=0.60$ ). Females with CAH were recruited through the CARES Foundation (84.3%) and clinics (15.7%).

All females had classical CAH and 93.0% underwent genital surgery. Median age at surgery was 1-2 years old. Surgery involved both vagina and clitoris (66.7%), vagina only (21.1%), or clitoris only (1.5%).

Most females with CAH (79.0%) believed legislation prohibiting surgery in childhood would cause harm (12.3% prevent harm, 8.8% neutral) (**Figure 1**). Most (64.9%) believed a ban “would have been harmful to me” had it been in place when they were children (24.6% not harmful, 10.5% neutral).

Most females (70.2%) believed a ban undermined parental rights to make medical decisions in their child’s best interest (17.5% respected parental rights, 8.8% neutral, 3.5% no answer) (**Figure 2**). While 75.4% did not believe a ban was in the best interest of CAH females, 14.0% did (10.5% neutral). There were no statistically significant differences between responses from females recruited via CARES Foundation vs. clinics for any study questions ( $p=0.66, 0.99, 0.71, 0.99$ , respectively).

#### *Parents of females with CAH*

Parents provided 132 responses (81.8% mothers) at a median parental age of 40 years (IQR 35-48) (**Table 1**). They lived in 37 states, with 43.9% residing in the five most populous states, consistent with national census data ( $p=0.13$ ). Among parents at least 25 years old, 81.7% had a post-secondary degree (above national values,  $p<0.001$ ). Median annual household income exceeded the national average (over \$100,000,  $p<0.001$ ). Parents were recruited through the CARES Foundation (77.3%) and clinics (22.7%).

Median age of daughters with CAH was 11 years old (IQR 7-18.5). All the daughters had classical CAH and 78.8% underwent genital surgery. Median age at surgery was <1 year old. Surgery involved both vagina and clitoris (59.4%), vagina only (18.2%) or clitoris only (0.8%).

Most parents (93.9%) believed legislation prohibiting surgery in childhood would cause harm (3.1% prevent harm, 2.3% neutral, 0.8% no answer) (**Figure 1**). Most (77.3%) believed a ban “would have harmed my daughter” had it been in place when she was younger (12.1% no harm, 5.3% neutral, 5.3% no answer). Parents were more likely than CAH females to oppose a ban ( $p\leq 0.02$ ).

Most parents (90.9%) believed a ban undermined parental rights (6.8% respected parental rights, 1.5% neutral, 0.8% no answer) (**Figure 2**). While 93.9% did not believe a ban was in the best interest of CAH females, 3.8% did (2.3% neutral). Parents were more likely to report that a ban would undermine parental rights and not be in the best interest of females with CAH ( $p\leq 0.002$ ). There were no statistically significant differences between responses from parents recruited via CARES Foundation vs. clinics for any study questions ( $p=0.11, 0.15, 0.23, 0.30$ , respectively).

## Discussion

We report the first assessment of patient- and parent-communicated opinions about potential legislative bans on genital surgery in childhood affecting females with CAH. First, the majority of women with CAH (8 out of 10) and parents of CAH females (9 out of 10) believed such a ban would cause harm. Moreover, they believed that, was a ban in effect, it would have personally harmed them or their daughter. Second, our findings highlight how individuals with a personal experience of atypical genitalia balance parental and child rights in this setting. Over 70% of women with CAH, and over 90% of parents, felt a ban undermined parental rights to make healthcare decisions in the best interest of their daughter. Furthermore, they believe a ban was not in the best interest of females with CAH. Finally, similar to previously reported findings,<sup>12</sup> most females with CAH and parents share similar opinions, but further study of reasons behind small differences between female and parental responses is warranted.

We believe this study contributes to the growing body of scientific literature drawing on the actual experience of those born with, and living with, atypical genitalia, rather than perspectives of those who do not. This distinction is critical as opinions differ between these two groups. At least one study found that women with CAH were far more likely to support the option of childhood genital surgery, and parents' ability to make the decision, compared to the general public.<sup>16</sup> Significantly, females with CAH do not identify with the intersex designation for clinical care<sup>10</sup>,<sup>11</sup> or legal identification<sup>12</sup> and prefer a separate designation from the larger heterogeneous DSD group. In this study, we noted an overwhelming opposition to a legislative ban on genital surgery in childhood among CAH females and parents of females with CAH, the people predominantly affected by such laws. This supports multiple previous studies reporting that, whether they underwent genital surgery in childhood or later, women with CAH<sup>16-19</sup> and parents<sup>16, 19-21</sup> strongly support genital surgery be permitted in childhood. This growing body of consistent evidence lends strong support to excluding females with CAH from policies and legislation pertaining to the intersex or DSD community, especially restrictions on genital surgery in childhood.

We have used the language of "parents' rights" in the study. We acknowledge that more technically appropriate terms when referring to surrogate decision making by parents on behalf of their child would be parental authority, or parental responsibility, when honoring individuals' rights to bodily integrity, privacy, and protection.<sup>22</sup> We used the language of "rights" because it is the language primarily used to discuss the controversy of managing children with conditions classified under DSD.<sup>23</sup> In addition, it has a more readily understood meaning, particularly when utilizing frameworks that respect parents' right to share in any decision making process affecting their child. Furthermore, children carry the right to have a competent person make decisions for them, a responsibility that defaults to a parent, barring extenuating circumstances. We aimed to use language commonly understood by women with CAH, their families, medical experts, human rights activists, and politicians. Moreover, "rights" language may add to the misunderstanding that decisions surrounding genital surgery in this population are either right or wrong. Adopting terms like authority and responsibility highlights the complexities and nuances these very challenging decisions involve.

Our findings are a sobering reminder that policies meant to help resolve or prevent injustices must involve the affected group(s), lest they have serious negative unintended consequences. Unfortunately, proposed and enacted legislative prohibitions on genital surgery in childhood have taken insufficient account of beliefs of the people they will directly impact. Additionally, such legislation was drafted in the absence of conclusive supportive scientific evidence. Multiple observational studies of long-term results of childhood genital surgery in CAH exist.<sup>24</sup> While some individuals and groups imply benefits of avoiding genital surgery in childhood, no data exists describing long-term implications of proposals forgoing early surgery.

While most CAH females, and parents of CAH females, opposed a ban on genital surgery in childhood, it does not translate that they were “pro-surgery.” Rather, participants endorsed maintaining options and choices for treatment without government intrusion. Parents deciding to proceed with surgery make a choice. Parents deciding to raise their daughter without early surgery make a choice as well. Since not all study participants underwent surgery, we included women with CAH who lived with the consequences of either choice their parents made before they could participate in the decision. The views expressed by study participants echo the 2018 American Medical Association report supporting the right of parents to make treatment decisions they believe are best for their children, while emphasizing shared decision-making as doctors help parents make empowered choices carefully personalized for their child.<sup>25</sup>

Any discussion of mandating treatment decisions warrants a brief discussion of this topic from the broader perspective of health.<sup>26</sup> A proposal to restrict non-emergent surgical interventions among children born with atypical genitalia opposes the World Health Organization definition of health as more than merely the absence of disease.<sup>27</sup> Instead, health is much broader, encompassing physical, mental and social dimensions. This is especially true in children, who thrive under optimal physical, mental and social conditions. Parents are the most important adults in a child’s life and their relationship forms the basis of their child’s future. It only follows that the optimal management of females with CAH be patient- and family-centered in a multidisciplinary setting.

Collecting anonymous self-reported data has led to several limitations. This limited our ability to classify CAH subtypes and to link potentially related female-parent responses. While noting a response rate comparable to similar studies, anonymous data made a sampling bias analysis impossible, since responder to non-responder characteristics could not be compared.

It would be ideal to compare opinions of females with similar degrees of baseline virilization who did and did not undergo early genital surgery. Unfortunately, objective information on baseline virilization was unavailable. Females born with more virilized genitalia are more likely to undergo early surgery and may hold different views about the potential ban. Therefore, comparing opinions of participants who did and did not undergo genital surgery in childhood without this data risked a biased comparison.

Our findings may not apply to other cultures or countries. We believe our results may be generalizable to the United States female CAH population. This is despite most study participants being recruited through an advocacy group, particularly since participants provided similar re-

sponses whether they were recruited through hospital-based clinics or not. Multiple characteristics of study participants, especially women with CAH, reflected the general population. In addition, participants were recruited from different social clusters, especially clinics, informing heterogeneous experiences of genital surgery and opinions about a ban on genital surgery in childhood.

## Conclusions

While responses were not unanimous, the majority of females with CAH and parents of females with CAH overwhelmingly support parents' ability to choose timing of potential genital surgery, opposing a moratorium on genital surgery in childhood.

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

**Keywords:** adrenal hyperplasia, congenital; differences of sex development; patient reported outcome measures

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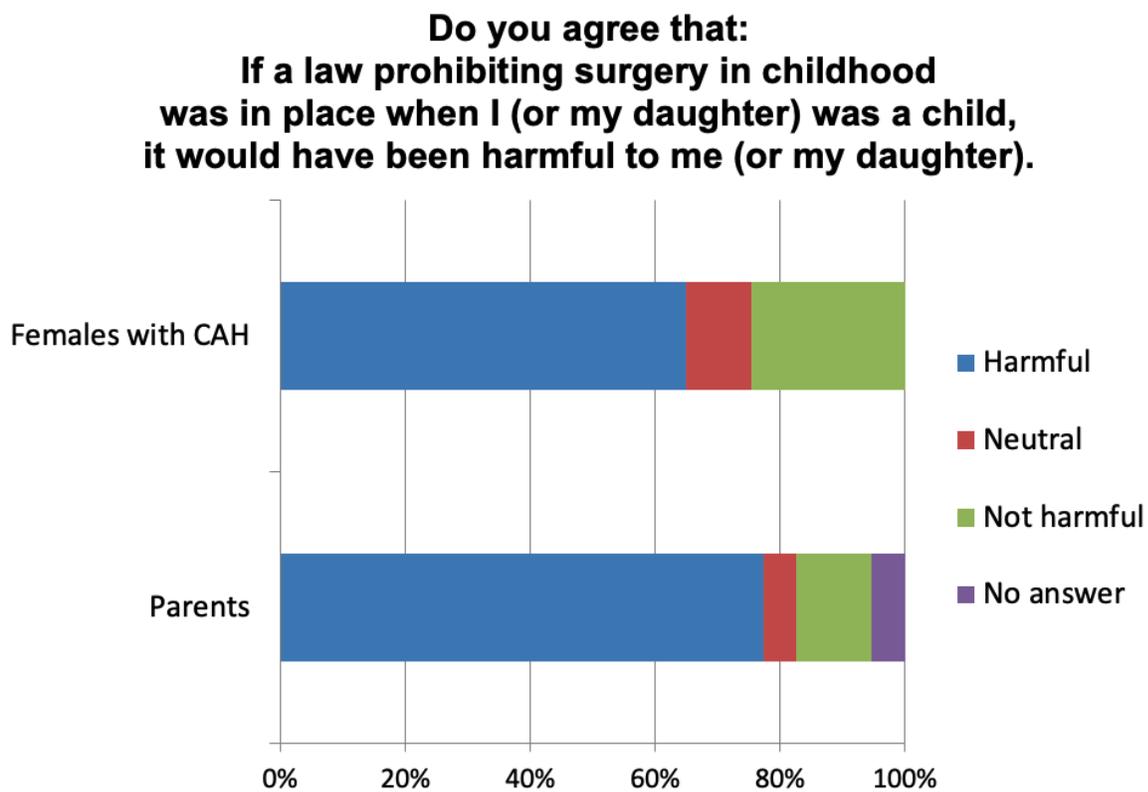
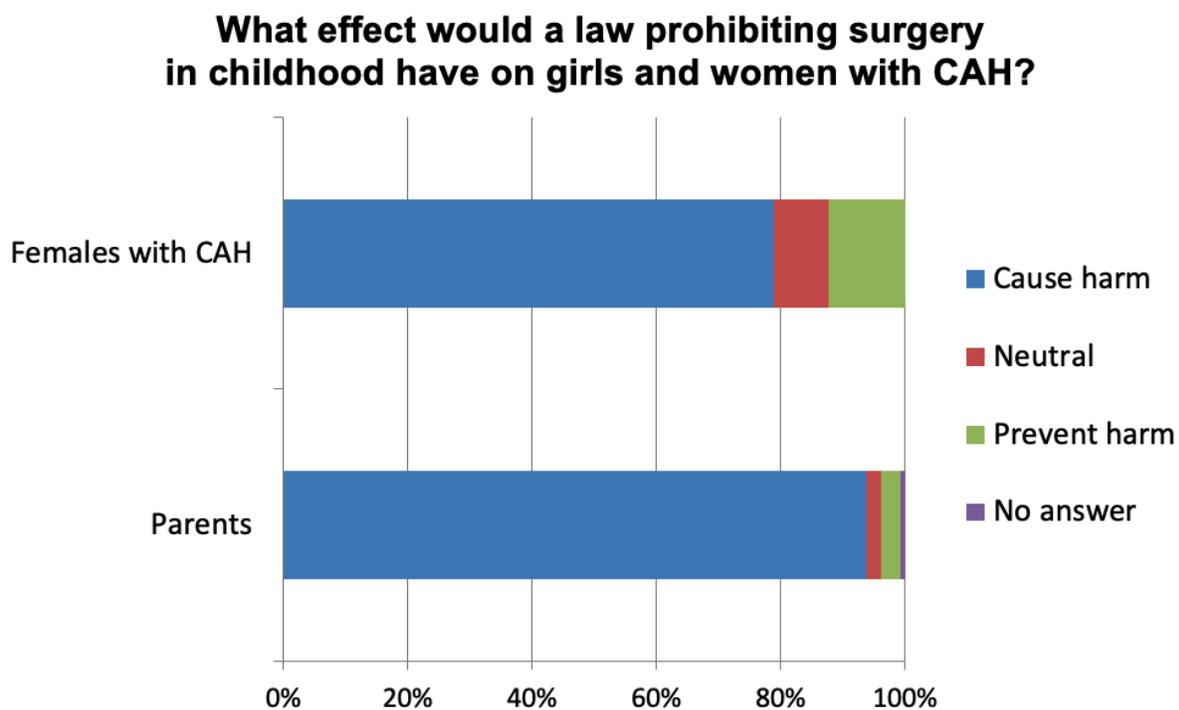
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**Table 1.** Participant characteristics.

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%)	
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%)	

**Figure 1.** Potential effects of a legal prohibition on genital surgery in childhood for girls with CAH.



**Figure 2.** Potential implications on parental rights and child best interest of a legal prohibition on genital surgery in childhood for girls with CAH.

