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Parental decisional regret and views about optimal timing of female genital restoration surgery in congenital adrenal hyperplasia

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Summary

Purpose
The role of female genital restoration surgery (FGRS) in girls with congenital adrenal hyperplasia (CAH) is controversial, with no long-term parent-reported outcomes available. Decisional regret (DR) affects most parents after their children's treatment of pediatric conditions, including hypospadias. We aimed to assess parental DR after FGRS in infancy or toddlerhood and explore optimal timing for surgery.

Materials and methods
One-hundred and six parents of females with CAH undergoing FGRS before 3 years old and followed at our institution (1999–2017) were invited to enroll online. Higher Decision Regret Scale (DRS) scores indicated greater DR (range 0–100). Participants also reported preferred FGRS timing relative to their surgery (earlier, same, later/delayed). Non-parametric statistical tests were used.

Results
Thirty-nine parents (median 4.4 years after FGRS) participated (36.8% response rate). Median age at FGRS was 9 months. Median DRS score was 0 (mean: 5.0). Overall, 20.5% of parents reported some regret (all mild-moderate) (Figure). Fewer parents reported DR after FGRS compared with published DR after hypospadias repair (50–92%, $p \leq 0.001$) or adenotonsillectomy (41–45%, $p \leq 0.03$). No parent preferred delayed FGRS. Seven parents (18.1%) preferred earlier surgery, especially when performed after birthday (80.0% vs. 8.8%, $p = 0.004$).

Discussion
We present the first report of validated long-term parent-reported outcomes after FGRS in infant and toddler girls with CAH. One limitation is that this is largely a single surgeon series. Reasons for the observed low levels of DR are likely multifactorial. Far from a definitive study, we aimed to provide parents willing to share about their experience an opportunity to do so. For that reason, selection bias may exist in our study. While parents with higher DR were potentially less likely to participate because of mistrust of the medical establishment, those with a negative experience may in fact be more likely to voice their opinions. A low participation rate was likely a result of the sensitive nature of FGRS, a desire for privacy, and inability to locate parents. A larger study will be required to assess how DR is affected by sexual function, genital appearance and complications, and DR among women with CAH.

Conclusions
Parents of females with CAH report low levels of DR after FGRS in infancy and toddlerhood. This appears to be lower than after other genital and non-genital pediatric procedures. When present, parental DR is usually mild. No parents preferred delayed surgery, even among those with DR. Some preferred earlier surgery.

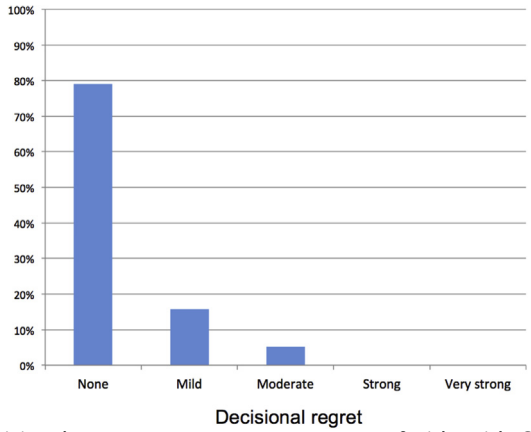

Figure Histogram of decisional regret scores among parents of girls with CAH after female genital restoration surgery.

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Introduction

Congenital adrenal hyperplasia (CAH) is the most common cause of ambiguous genitalia in newborns [1–3]. Historically, female genital restoration surgery (FGRS) was performed in infancy to prevent urinary pooling and to match external genitalia to chromosomal sex. Psychological benefits to the child, family, and caregivers [3–6], often cited as a reason for early surgery, remain to be proven. Recently, some advocacy groups, ethicists, and physicians have challenged early surgery, some calling for moratoria on genital surgery in all disorders of sexual development, until children are old enough to decide for themselves [7]. Adding to this controversy are concerns that clinical outcomes may differ for surgeries performed in infancy versus after puberty [8,9]. Reliable data to support either approach are limited, as the literature consists primarily of surgeon-reported outcomes of historical procedures [10,11].

Long-term patient- or parent-reported outcomes (PROs) are lacking to support any position regarding the role of FGRS in CAH [10]. One of the goals of any CAH management strategy is to minimize patient and parental decisional regret (DR). DR can follow any treatment decision, including a decision to forgo treatment. DR has been reported in parents of children undergoing various treatments, and in fact, DR affects 50–92% of parents after their sons' hypospadias surgery [12,13].

We aimed to assess DR of parents of females with CAH after FGRS performed in infancy and toddlerhood and to explore their opinion of optimal timing for surgery. We hypothesized that prevalence of DR after FGRS is similar to other procedures performed in childhood and few prefer delayed surgery.

Methods

We performed an IRB-approved cross-sectional online study of parents of girls with CAH followed at our institution after FGRS by age 3, mostly performed by a single surgeon (1999–2017). A minimum of 3 months since FGRS was required for inclusion. Of 118 potential parents, 106 with contact information were eligible. Adult women with CAH were also invited to participate. This report focuses on the results form parents, due to a very low response rate from women (women's responses are summarized at the end of the Results section).

Eligible participants were mailed generic letters inviting them to participate without disclosing the diagnosis or treatment history. Interested participants were emailed an individualized link to the online survey, with a reminder emailed a week later. Study data were managed using REDCap, a secure web-based platform [14].

Decisional regret

DR was assessed using the validated Decision Regret Scale (DRS) [15,16]. DRS consists of five items scored on a 5-point Likert scale (strongly disagree, disagree, neither disagree nor agree, agree, strongly agree). Scores range from 0 to 100, with higher scores indicating greater DR. Scores were classified as no DR (0), mild DR (1–25), moderate (26–50), as previously described [17], further dividing higher DR into strong (51–75) and very strong (76–100).

Sensitivity analysis

DRS is a sensitive instrument, classifying the slightest answer indicating potential regret as regretful, with no established clinically meaningful cutoff (0 = no DR vs. 1–100 = some DR). To adjust for this low threshold, sensitivity analyses were performed. First, we used cutoffs which may be more clinically meaningful: >10 [18], >25 [17,19], and >30 [20]. Second, we used a non-neutral regret definition: DR present in those who either disagreed/strongly disagreed with any of items 1, 3, or 5, or agreed/strongly agreed with items 2 or 4 [18].

Risk factors of parental decisional regret (exploratory)

Several potential predictors of higher DR were selected *a priori*: age at FGRS (<1 year old vs. 1–2 vs. >2), preoperative Prader scale (3 vs. 4–5), being a mother (vs. other), undergoing another genital surgery, and time since FGRS (<5 years, 5–10, >10 years).

Parental decisional regret after other procedures

A systematic search was completed in April 2017 to identify relevant articles in Medline (from 1950), PubMed (from 1946), Embase (from 1949), and GoogleScholar (from 1990). Combinations of the terms: decisional regret, parent, child were used. Included publications needed to use the DRS. To determine the average percentage of parents reporting DR for all reported conditions, a weighed mean of DR was calculated. For each study, the percentage of parents reporting DR was multiplied by the number of participants. The sum of these values was divided by the total number of participants.

Preferred timing of surgery

Participants were asked about their preferred timing of surgery relative to when FGRS was actually performed ("Looking back, when would you have done the original surgery for your child?"). Answer options included: earlier in life, same time, later in life.

Risk factors of earlier or later surgery (exploratory)

Similar to the analysis of DR, we screened the following predictors: age at FGRS, preoperative Prader scale, being a mother, additional surgery, time since FGRS and DR.

Power calculation

Although a power calculation was not carried out at study inception, we performed it at study completion. To detect a 50% difference in DR reported by parents of girls with CAH

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