Gender Assignment for Newborns With 46XY Cloacal Exstrophy: A 6-Year Followup Survey of Pediatric Urologists

David A. Diamond,* Jeffrey P. Burns, Lin Huang, Ilina Rosoklija and Alan B. Retik

From the Department of Urology, Children's Hospital, Boston, Massachusetts

Study received Children's Hospital institutional review board approval.

* Correspondence: Department of Urology, Children's Hospital, 300 Longwood Ave., Boston, Massachusetts 02115.

Purpose: Gender assignment for newborns with ambiguous genitalia remains a challenge. An initial survey of colleagues on this subject was performed in 2004. Our objective was to understand the basis for the attitudes and practices of pediatric urologists in regard to gender assignment for 46XY cloacal exstrophy in a 6-year followup survey.

Materials and Methods: A survey on a case of 46XY cloacal exstrophy was completed by 191 of the 263 fellows (73%) in the Urology Section, American Academy of Pediatrics. Questions referred to gender assignment, surgery timing, clinical outcomes and respondent demographics.

Results: Of the fellows 79% favored male gender assignment. The most important factor in male assignment remained androgen brain imprinting (97%) while in female assignment it was surgical success in creating functional genitalia (96%). Respondent characteristics associated with assigning female gender were longer practice duration (greater than 15 years) ($p < 0.03$), having trained in programs where female gender was always or usually assigned ($p < 0.02$) and not being a fellowship program director (0 of 27 respondents, $p < 0.03$). There was an evolution among respondents from female gender assignment earlier in the career to male assignment currently ($p < 0.0001$).

Conclusions: Most pediatric urologists favor male gender assignment for 46XY cloacal exstrophy, which is a significant increase in 6 years. This change represents an evolution from female to male gender assignment and virtual unanimity among fellowship directors to gender assign male. Longer practicing clinicians perceived better outcomes for female gender assignment. If this reflects true clinical outcomes, the trend toward the eventual disappearance of female gender assignment for 46XY cloacal exstrophy is concerning.

Key Words: abnormalities, cloaca, genitalia, male, female

GENDER assignment for newborns with ambiguous genitalia remains 1 of the most controversial issues in pediatrics. In 2006 we published the findings of our survey of attitudes and practices of fellows of the Section on Urology, American Academy of Pediatrics.[1] For masculinized 46XX CAH there was 99.5% support for female gender assignment. On the other hand, for 46XY cloacal exstrophy there was considerable disagreement with 70% of respondents favoring male gender assignment (fig. 1). These choices were strongly influenced by demographic variables since clinicians with greater than 15 years of experience and those teaching higher levels of trainees (residents/fellows) were 4 times as likely to assign female gender. The reason for these differences was unclear but included the possibilities that longer

0022-5347/11/1864-1642/0
THE JOURNAL OF UROLOGY®

Vol. 186, 1642-1648, October 2011
Printed in U.S.A.
DOI:10.1016/j.juro.2011.03.101

Figure 1. Photograph of patient with 46XY cloacal exstrophy with rudimentary phallic structures presented in previous and current surveys.

practicing surgeons had seen greater success with female or less success with male gender assignment, or they trained in a prior era during which sex assignment was largely driven by anatomical limitations and phallic size. The distinction seemed important since to our knowledge psychosexual outcome data are lacking and the correct approach remains indeterminate.

The purpose of this study was to better understand factors responsible for differences in the attitudes and practices of gender assignment for 46XY cloacal exstrophy, and determine whether any significant changes had occurred in the 6 years since the original survey.

MATERIALS AND METHODS

The questionnaire was developed with attention to clarity and a range of response options in a manner similar to the original survey instrument. It was pilot tested for content validity and clarity with internal pediatric urology faculty but not formally validated.

The questionnaire consisted of 2 sections with a total of 15 questions. Section 1 included questions on a representative case of 46XY cloacal exstrophy with rudimentary phallic structures, identical to that presented in the original survey. Questions related to gender assignment, supporting reasons and the proposed timing of surgery, and asked respondents to describe their training in gender assignment for 46XY cloacal exstrophy and personal clinical experience. They were asked to rate their perception of overall success with female and male gender assignment, and describe the evolution of their practice. The final section gathered data on respondent characteristics, such as age, geographic location, years in clinical practice, presence of an experienced partner, affiliation with training programs and whether the respondent was a pediatric urology fellowship director. After data analysis a single followup survey question was sent to 33 respondents who had noted that female gender assignment was more successful than male assignment but who would still currently assign male gender.

Study Population

The sample was determined using the current roster of the Section on Urology, American Academy of Pediatrics. A self-administered anonymous questionnaire was sent electronically with a cover letter using SurveyMonkey® in June and July 2009. For nonrespondents to the electronic survey this was followed by a mailed packet that included a cover letter from the principal investigator and a stamped, self-addressed return envelope. The questionnaire did not elicit any personal information linking responses to specific individuals. Nonrespondents were tracked by code numbers. All responses submitted by November 2009 were analyzed. The single followup question was sent electronically in August 2010, followed by a mail packet to nonrespondents. The protocol was approved by the Children's Hospital institutional review board.

Of the 263 fellows of the Section on Urology who were sent questionnaires 191 (73%) completed the survey. Not all respondents answered each question. A total of 27 individuals identified themselves as program directors or co-directors, which included each pediatric urology fellowship program in North America. All of them completed the survey. For the followup survey the response rate was 90% (30 of 33 fellows).

Data Analysis

The frequency and percent of answers to each question were calculated and compared to those of the previous survey by the chi-square test. We used a 1-sided p value since we set the null hypothesis of the new survey as an equal or lesser rate of preferring male gender assignment. Factors affecting the physician choice of male over female were analyzed by the chi-square or Fisher exact test. The Cochran-Armitage Trend test was used to analyze the association between 2 categorical variables, of which 1 had ordinal categories. Friedman's chi-square test was used to determine whether certain groups of physicians rated the overall success differently in patients assigned a female or male gender. Two-sided p values were used with $p < 0.05$ considered statistically significant. All analyses were done with SAS® 9.2.

The questionnaire was designed to include a wide range of variables about respondents, such as age, practice environment and clinical experience, for which we performed statistical tests. Most variables showed no association with the outcome, gender assignment. Furthermore, stratification analysis and logistic regression were also done to measure the effects of potential confounding variables.

RESULTS

46XY Cloacal Exstrophy

Of the respondents 148 (79%) agreed or strongly agreed that male assignment was most appropriate, which was a significant increase from the 70% in the previous survey ($p = 0.041$, fig. 1). The factor most highly rated as important or very important was the