

Contemporary Surgical Approach in CAH 46XX – Results from the I-DSD/I-CAH Registries

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Introduction

Genitoplasty in girls with congenital adrenal hyperplasia (CAH) remains controversial and, globally, little consensus exists on the indication as well as the timing of these procedures. Currently, it is unclear whether this increased controversy has led to any notable shifts in practice.

Objective

To evaluate recent international trends in surgical practice in girls with CAH.

Patients and Methods

The I-DSD and the I-CAH registries are international registries that use a common platform approved by the National Research Ethics Service of the United Kingdom for storage of pseudo-anonymised data. All cases that had been classified in the I-CAH/I-DSD registry as 46XX CAH and who were born prior to 2017 were identified (555 participants from 41 centres, born 1953-2016). Centres were approached to obtain information on factors that influenced the option of performing surgery, timing and type of surgery.

Results

Of the 330 participants, 208 (63%) presented within the first month of life. 326 (99%) cases were assigned female. Genital surgery had been performed in 251 (76%). The proportion of girls that had undergone surgery showed geographical variation with 64%, 89% and 97% of cases residing in Europe, South America and Asia, respectively (figure 1). Median age at first surgery was 1 year (0, 15), with clitoral surgery and vaginal surgery at 1 year (0, 15) and 1 year (0, 38), respectively. In a logistic regression model, the incidence of surgery showed a trend to reduced number of surgeries over time (figure 2). However, in those cases who did have surgery, there was an increased trend over time of undergoing surgery before 24 months of life (figure 3). Limitations: Data were extracted from a huge database and there may be reporting and/or selection bias.

Conclusion

Surgery in DSD remains a controversial topic. Despite the widespread controversy in the medical community, media and general population, there is a significant lack of reliable and prospective data. This large international study provides a clear snapshot of current practice and reveals geographical and temporal differences. Whilst there is a trend towards avoiding surgery in Europe, there also seems to be a significant trend towards aiming for surgery within the first two years of life.

Surgery age by continent and time

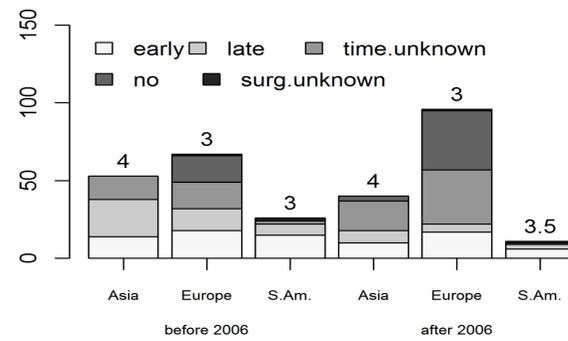


Figure 1 Surgery before and after the consensus statement 2006 in different regions and median prader score

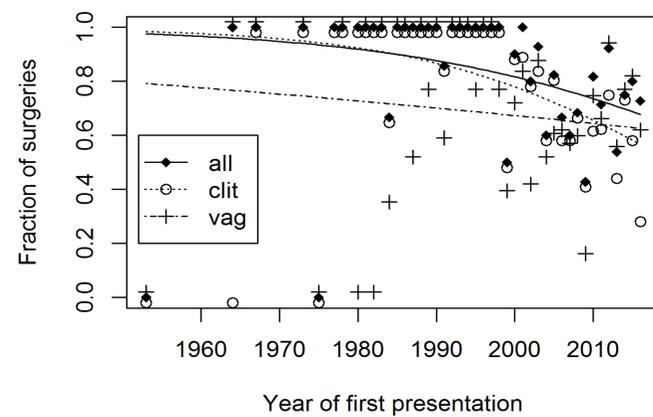


Figure 2 The percentage (0-1) surgeries and type of surgery over the years (all=surgery; clit=clitoral surgery; vag=vaginoplasty)

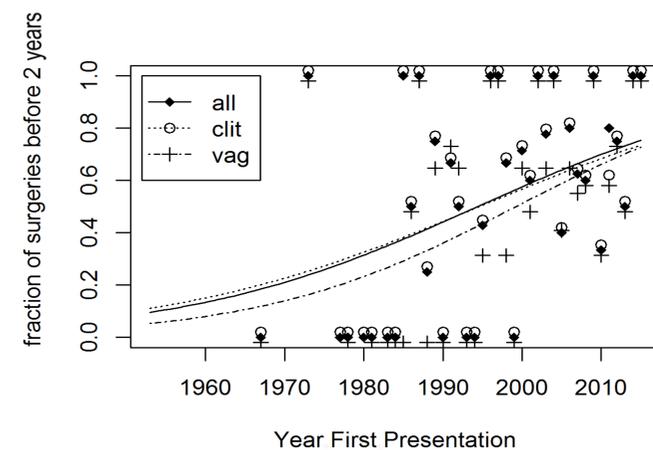


Figure 3 Percentage (0-1) of surgery before 2 years of age over time (all=surgery; clit=clitoral surgery; vag=vaginoplasty)

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