Girls born with genital anomalies: Are they receiving optimal care for their sexual and reproductive life?

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Background:
Genital anomalies at birth include anomalies that affect the genitalia, the urinary tract and the lower bowel. At birth, the priorities for these children is correction of their urinary and bowel problems as these may have immediate and potentially life-threatening consequences.

In girls, genital anomalies often include absence of, or failed fusion of the reproductive tract with potential obstruction of one or both sides. These reproductive tract anomalies are not always recognised or corrected during childhood when the other problems are being addressed. Failure to identify these problems may result in acute presentation with menstrual outflow obstruction or puberty, resulting in presentation with pain and requiring urgent surgery. Other issues relating to the consequences of having altered genitalia which need to be dealt with include those relating to the potential use of tampons, to sexual activity, as well as issues around body image. Failure to address these issues may have negative consequences on the adolescent developing positive sexual relationships. Discussion regarding childbearing also needs to occur with the young woman.

At present, limited treatment pathways exist for young women with genital anomalies at the Royal Children's Hospital (RCH) in Melbourne, Australia or other paediatric centres internationally.

Aims
This audit aimed to review the experiences of girls born with some specific genital anomalies (congenital adrenal hyperplasia (CAH), cloacal anomalies, bladder exstrophy and ano-rectal malformations (ARM)) seen at RCH. The rate of timely identification of reproductive tract anomalies, referral for appropriate advice and support regarding menstruation (in view of potential difficulties with the vaginal opening), and the opportunity for discussion regarding sexual activity were assessed.

Results
- 241 medical records were reviewed and 145 were included
- Ano-rectal malformations (imperforate anus) n = 49
- Cloaca n = 11
- Bladder exstrophy n = 21
- Congenital adrenal hyperplasia (CAH)

Obstructive reproductive tract anomalies were identified in 35% of girls with cloacal anomalies (50% with acute presentations) and in 75% of girls with bladder exstrophy (2/3 of these presenting acutely).

Referrals for timing of sexual health counselling (girls age 10+) are shown in Figure 1 below.

Sexual health topics that were noted to have been discussed with the adolescent girls are shown in Figure 2 below.

Conclusion
- There needs to be an increased awareness of potential reproductive tract anomalies to avoid the acute presentation post menarche requiring emergency surgery in girls with cloacal anomalies and cloacal exstrophy.
- Many girls born with genital anomalies were not referred for sexual health counseling or were referred after puberty.
- Many girls did not have a discussion on all sexual health topics noted in their records.
- The potential to intervene in a preventative and proactive way exists for this group of girls to decrease future sexual health problems.

References: