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Practice changes in childhood surgery for ambiguous genitalia?

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Abstract *Objective:* In 2001, this team published an observational study of the clinical outcomes of a cohort of adolescent girls born with ambiguous genitalia. The poor outcomes observed represented a major scientific challenge to the standard practice of childhood feminising genital surgery. That publication was one of several contributing to a call for change in surgical practice, which culminated in the publication of the Chicago Consensus Document in 2006. The aim of this current study was to repeat the same evaluation of clinical outcomes on a recent cohort of adolescent girls and compare the two cohorts to identify differences in adolescent outcomes which may indicate a change in paediatric surgical practice.
Methods: This was an observational study of a current cohort of adolescent girls treated in childhood for ambiguous genitalia and referred to a specialist adolescent disorders of sex development (DSD) service for assessment. Data were collected on surgical history, genital examination findings and treatment recommendations for 30 consecutive adolescents over a 5-year period. Findings were compared with those of a similar cohort of adolescent girls published over a decade previously.
Results: Clitoral surgery remained common (93% vs 100%, current cohort vs historical cohort). However, concomitant vaginoplasty was performed less frequently (80 vs 100% current vs historical). Vaginoplasty revision surgery was also less commonly required (65 vs 81%), although 24% of the recent cohort still required major revision surgery prior to intercourse. There was some improvement to the cosmetic outcomes as deemed by the surgical team using the same criteria as the previous report.
Conclusions: This study provides some slight evidence of recent practice change. There was a small reduction in the number of vaginoplasties performed in childhood and an improvement in vaginoplasty outcomes and cosmesis. However, there was no identifiable change in management of clitoromegaly and the numbers of clitoral reduction operations remained high. This is surprising given the clear evidence of a detrimental impact of surgery on clitoral sensation and sexual function.

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Introduction

Ambiguous genitalia are anatomical signs associated with certain forms of 'disorders of sex development' (DSD) — a controversial term encompassing atypical development of chromosomal, gonadal or anatomical sex [1]. It has been estimated that about 1 in 4500 live born infants have external genitals that are sufficiently ambiguous to render gender assignment not obvious [2]. Elective feminising genitoplasty for female assigned infants and children continues to be the most debated aspect of clinical management [3].

Reports of feminising surgery can be identified from the 1880s [4]. Clitoral surgery has evolved from clitoridectomy to clitoral recession and clitoral reduction. The highly popular 'one stage genitoplasty' means creating a vaginal opening at the same time as clitoral surgery [5]. The type of vaginoplasty carried out depends on anatomical factors and the underlying medical condition. Operations thus range from a simple introitoplasty to major surgery, such as intestinal vaginoplasties. Paediatric urologists in favour of early childhood genital surgery have tended to report short-term outcomes, at least relative to the intended lifelong impact. Pre-pubertal girls do not menstruate, are not sexually active and are generally not preoccupied about their genital appearance, so that meaningful outcome measures are not available in childhood. Outcomes that are pleasing to paediatricians are not easily extrapolated to adult successes.

In 2001, an observational study of the surgical outcomes of a cohort of adolescent girls born with ambiguous genitalia was published by the current clinical team [6]. In that study, childhood feminising genital surgery was found to be universal and multiple episodes were common. Furthermore, the majority of the postpubertal girls required further reconstructive surgery to permit menstrual flow and facilitate sexual intercourse. The report became widely cited in the medical and user literature. It resonated with many adult women who had been recipients of childhood surgery and whose dissatisfaction had hitherto been considered exceptional and therefore dismissed [7]. Since then the topic has received further scientific research and ethical analysis [6,8,9].

By 2005, sufficient doubt had been cast so that the first and only international consensus statement published in 2006 recommended a far more cautious approach to childhood feminising surgery [1]. A larger clitoris is medically benign, a vaginal opening has no purpose for a prepubertal girl, and the evidence of the presumed long-term benefits for these interventions is missing. The consensus view was that delaying these interventions until gender orientation and sexual preferences are clearer and the individual can participate in the consenting process appeared to be a reasonable course of action. The caution advocated by the consensus statement has continued to be challenged by some paediatric surgeons who remain optimistic that refinement of surgical techniques will overcome the problems of vaginal stenosis and clitoral numbness [10,11].

The current study was designed to perform the same clinical evaluation on a new cohort of adolescents with ambiguous genitalia caused by DSD who had been treated a decade later. Their surgical trajectories were compared with those of the previously published cohort. The aim was to identify any differences in adolescent clinical outcomes which may reflect a change in paediatric surgical practice.

Methods

The study took place at a multidisciplinary service for adolescents and adults with DSD diagnoses. The service does not manage paediatric patients with DSD but is the largest referral centre in the UK for adolescent and adult DSD. The study was an observational study of the surgical history, current anatomical findings and recommendations for further management of all adolescent patients referred to the service between 2007 and 2011. The protocol was approved as an audit by the hospital research ethics committee.

Examination under anaesthetic (EUA) is an aspect of routine care for adolescents with a history of genital ambiguity at birth whether or not childhood surgery has been performed. Anatomical and appearance findings for females are documented in the standard format detailed below and any treatment options are discussed by the team at the time of the EUA.

The anatomical assessment focuses on vaginal and clitoral dimensions. Vaginal length is measured in centimetres from the posterior fourchette to the posterior fornix or vaginal vault if no cervix is present and capacity and suppleness are recorded. Capacity is measured by insertion of vaginal dilators of graduating size. Suppleness is the ease with which the vagina stretches and to what extent this is affected by scar tissue. The clitoris is measured using a sterile ruler from the base to the tip of the glans clitoris on the dorsal surface and deemed normal if less than 3.5 cm [12]. It is deemed absent when not visible and no corporal tissue is palpable.

Cosmetic criteria include the appearance of the glans and clitoral corpora, the presence of a clitoral hood, the appearance of labia minora and majora (rugosity, scarring, pigmentation), vaginal introital position and appearance along with overall genital proportions. The overall cosmesis is graded by the surgical team as: Good (external genitalia more or less typical for women; unusual features not identified), Satisfactory (up to two mildly unusual features identified) or Poor (highly atypical, three or more unusual features identified). Examples of unusual features would include an absent clitoral hood or absent or markedly asymmetrical labia minora.

Treatment recommendations are made according to established local practice [6]. Dilation is the primary intervention to increase vaginal volume in the absence of scarring and capacity for at least partial insertion of the smallest Femmax® Dilator. Minor surgery is recommended to those with mild stenosis or the presence of scar tissue at the introitus (Fenton's introitoplasty). Major surgery may be indicated in the presence of severe vaginal stenosis and includes flap, pull through and intestinal vaginoplasty techniques depending on genital findings.

Clitoral surgery is offered to girls and women whose clitoris is enlarged and in the presence of distressing symptoms. Psychological assessment and interventions are available to patients who opt in.