

Objective cosmetic and anatomical outcomes at adolescence of feminising surgery for ambiguous genitalia done in childhood

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There are few, if any data on the long-term outcome of feminising genital surgery for children with ambiguous genitalia. We present a retrospective study of cosmetic and anatomical outcomes in 44 adolescent patients who had ambiguous genitalia in childhood and underwent feminising genital surgery. Cosmetic result was judged as poor in 18 (41%) of these patients. 43 (98%) of 44 needed further treatment to the genitalia for cosmesis, tampon use, or intercourse. 23 (89%) of 26 of genitoplasties planned as one-stage procedures required further major surgery. This information must be available to parents and clinicians planning such surgery. Cosmetic genital surgery in infancy needs to be reassessed in the light of these results.

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Genital appearance has been cited as a fundamental factor in childhood gender and psychosexual development,¹ and has led to the current management of infant genital cosmetic surgery for genital ambiguity. For people brought up as girls, surgery aims to promote female gender development and avoid psychological distress, whilst long-term aims are to allow normal-looking and functioning adult female genitalia. However, there is increasing evidence that long-term there is patient dissatisfaction with surgery outcomes² and debate over timing of vaginoplasty is on-going.³ Irreversible infant cosmetic genital surgery will have life-long impact on social, psychological, and sexual function. Central to this debate is the question of whether feminising surgery in childhood is

successful cosmetically and functionally. We report our retrospective study of childhood feminising genital surgery.

The notes of all paediatric and adolescent patients with a history of ambiguous genitalia and feminising childhood surgery were reviewed at the Department of Gynaecology, Elizabeth Garrett Anderson Hospital, London, UK. All had undergone planned detailed genital examination before menstruation and sexual activity. All UK hospital notes were retrieved and analysed for previous surgical details including type, timing, and number of genital operations (table 1). Patients were allocated one of three diagnostic groups: A, B, or C. The genital assessment included: cosmetic appearance, anatomical assessment, and treatment recommendations. No standard methodology exists for genital cosmetic assessment. We assigned one of three categories for cosmesis: good (genitalia appear normal; no abnormal features), satisfactory (up to two minor abnormalities, unlikely to be judged abnormal by a non-medically trained person), or poor (genitalia appear abnormal; three or more abnormal features). Our cosmetic assessment criteria included: genital proportions and symmetry, pubic hair distribution, clitoral hood size and shape, glans clitoris shape and prominence, clitoral body size and prominence, labial positions and proportions, vaginal introital position and appearance, and genital skin quality (scarring, pigmentation, and rugosity). Anatomical dimensions were assessed against normal values for clitoral and vaginal size.⁴ Treatment recommendations were made according to our treatment protocol. Dilatation is the first choice for vaginal enlargement whereas surgery is indicated for those with fibrotic scar tissue not amenable to dilatation, or an absent vaginal introitus. Minor surgery included labial refashioning and introital enlargement (episiotomy or Fentons procedures). Major surgery included all other operations. We divided overall outcome into two groups: acceptable outcome (no intervention required, vaginal dilators, minor surgery, surgery for clitoral regrowth, or major surgery electively deferred until

Surgery type	Diagnostic group		
	A: Congenital adrenal hyperplasia, 21-hydroxylase deficiency (n=21)	B: Miscellaneous conditions causing ambiguous genitalia (n=12)*	C: Abnormalities of cloacal and/or urogenital development (n=11)†
Clitoral surgery n=38 (86%)‡			
Age (years) at clitoral surgery (median; mean range)	0·8 (1·7 [0·1–10·4])	1·2 (2·3 [0·1–8·1])	0·4 (2·3 [0·1–7·2])
Number of patients			
Total	21	12	5
Clitoral reduction	18	11	2
Clitoral recession or plication	1	1	0
Total clitoral excision	2	0	0
Other clitoral procedures	0	0	3
Labial surgery n=16 (36%)			
Number of patients			
Total	9	7	0
Preputial skin constructed labia minora	9	4	0
Labia majora revised	0	3	0
Vaginal surgery n=39 (89%)§			
Age (years) at vaginal surgery (median; mean range)	0·8 (1·7 [0·1–10·4])	1·6 (3·1 [0·1–10])	1 (3·9 [0–16])
Number of subjects			
Total	20	8	11
Sagittal cutback to open urogenital sinus	5	0	0
Perineal fortunoff flap	12	5	1
High take off urethra-vagina with vaginal pull through	3	2	2
Other vaginoplasty**	0	1	8

*Diagnoses: 17 ketosteroid-3-reductase deficiency (n=1), XXY and ambiguous genitalia (n=1), true hermaphrodite (n=2), XY female of unknown aetiology (n=4), virilisation because of maternal drugs (n=1), mixed gonadal dysgenesis (n=3). †Diagnoses: urogenital sinus anomaly (n=2), exstrophy variant (XX; n=1), female epispadias-classical (n=1), cloacal exstrophy (XY) female (n=1), cloacal anomaly (XX; n=4), cloacal anomaly (XY) female (n=1), bladder exstrophy (XY; n=1). ‡26% had repeated clitoral surgery. §We do not have information on how many patients were asked to perform vaginal dilation post-operatively. 31% underwent vaginal revision surgery at least once. **Types: intestinal transposition (n=3), McIndoe (n=2), remodelling the introitus (n=2), complex vaginoplasty incorporating cloacal and urethral elements (n=2).

Table 1: Details of diagnostic group and previous genital surgery

Outcome	Result
Anatomical assessment	
Clitoris	
Absent	3 (7%)
Small	3 (7%)
Normal	26 (59%)
Large	3 (7%)
Excessive	9 (20%)
Vaginal introitus	
Absent	4 (9%)
Small	32 (73%)*
Normal	8 (18%)
Vaginal length	
Absent	3 (7%)
Short	9 (20%) [†]
Normal	32 (73%)
Labia [‡]	
Normal	27 (61%)
Poor/scrotal	13 (30%)
Partial fusion	5 (11%)
Total fusion	1 (2%)
Overall cosmetic result	
Good	8 (18%)
Satisfactory	18 (41%)
Poor	18 (41%)
Further treatment recommendations[‡]	
None	1 (2%)
Vaginal surgery	
Major	30 (68%)
Minor	3 (7%)
Clitoral surgery	11 (25%)
Labial surgery	5 (11%)
Vaginal dilation	10 (23%)

*20 of 32 patients were <1 cm diameter. [†]Three 0.5–3 cm and six 3.5–6 cm long. [‡]More than one category may be applicable for each patient.

Table 2: Results of cosmetic and anatomical genital examination, and recommendations for further treatment

after puberty); and poor outcome (further major surgery recommended).

Over 4.5 years, 44 patients fitting the study criteria were referred for further investigation. 42 patients underwent examination under anaesthesia and two requested outpatient examination. 37 patients were given routine referrals for adolescent genital assessment, and seven were referred for clinical reasons (two for clitoromegaly, two for abdominal pain, one for primary amenorrhoea, one for menorrhagia, and one for vaginal discharge). Mean age at examination was 15.1 (range 7.5–19.4) years. Most first surgical procedures were done during 1979–89 (range 1979–95). Mean follow-up time was 13.2 (4.3–19.3) years. Median age of first surgery was 0.8 (2 weeks–10.4 years) years. In two (5%) of 44 patients, major vaginal surgery was deferred until after puberty. In 42 (95%) of 44 patients, childhood genital surgery was intended to be definitive with seven (17%) of the 42 having additional surgery planned later in childhood but for most, 35 (83%) of the 42, the first procedure was intended to be single and definitive (26 of 35 were planned one-stage clitorovaginoplasties).

In 26 (59%) of the 44 patients the cosmetic appearance was good or satisfactory (table 2). Overall 15 (34%) had an acceptable outcome and 29 (66%) had a poor outcome. Analysing outcome by diagnostic group, 24% of Group A, 33% of Group B, and 55% of group C had acceptable outcomes. Of those undergoing single-stage feminising genitoplasty, nine (35%) of 26 had already had undergone at least one further major genital surgical procedure before referral to our department. Of the remaining 17 (65%) of 26 patients, none would be able to use tampons without further treatment. Of these, three (18%) of 17 required dilators or minor surgery and 14 (82%) of 17 needed major surgery. Ten (26%) of 38 had undergone two or more clitoral procedures with one having undergone four clitoral reductions.

The outcomes of childhood genital surgery are substantially poorer than reported previously⁵ with nearly all children requiring further treatment. All surgery was done at specialist units and should give the best results available. This study is retrospective, but because of the numbers of patients involved, we believe it is representative. Nevertheless, despite planned routine referrals for all relevant adolescents, these patients may be those with the poorest outcomes. Additionally these children had surgery between 1979 and 1995. There have been changes in surgical techniques, equipment, sutures, and antibiotics since that time. More up-to-date procedures may have better outcomes, although there are few data to support this.

This study prompts a re-evaluation of cosmetic genital surgery in children. Most vaginal surgery can be deferred until after adolescence unless haematocolpos is a risk. Repeated clitoral surgery may be more damaging to sexual function than a single procedure. Clitoral regrowth occurred in 39% of patients. Children with mild clitoromegaly should have surgery deferred until they are old enough to be involved in the decision. Surgery should not damage genital sensitivity and sexual expression should be pleasurable with the ability for orgasm undiminished; there are currently no objective data for these outcomes. It is important that clinicians and parents understand that genital ambiguity cannot be corrected in infancy by a single procedure. For most individuals further treatment will be necessary in adolescence and the long-term impact of such treatment on adult sexual function is still unknown.

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Cold feet and prolonged sleep-onset latency in vasospastic syndrome

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People with vasospastic syndrome have cold hands and feet and abnormal vasoconstriction after local cold exposure. Normally there is a circadian rhythm of distal vasodilation, with onset in the early evening, which directly influences ability to fall asleep. We gave a sleep questionnaire to 32 patients with primary vasospastic syndrome and 31 healthy controls. People with vasospasticity had significantly prolonged sleep-onset latency both at onset of night-time sleep and after nocturnal disturbance. This prolonged latency could be associated with impaired capacity for distal vasodilation.

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An association between thermoregulatory processes and the initiation of sleep has long been recognised. The degree of