

Genital sensation after feminizing genitoplasty for congenital adrenal hyperplasia: a pilot study

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OBJECTIVE

To assess sensation in the clitoris and vagina in women with congenital adrenal hyperplasia (CAH) who have previously had genital surgery, and to evaluate sexual function in this group as the latter, and particularly the experience of orgasm, appear to be closely related to sensitivity.

PATIENTS AND METHODS

Six women were recruited from a multidisciplinary clinic specialising in intersex conditions, and representing an initial cohort from a larger ongoing study. The patients were asked to complete a postal questionnaire with a specialized sexual function assessment. Thermal, vibratory and light-touch sensory thresholds were assessed

in the clitoris and vagina using a genito-sensory analyser and Von Frey filaments.

RESULTS

All six women had highly abnormal results for sensation in the clitoris. Only three of them had an introitus capable of admitting the vaginal probe, and the vaginal sensory data of all three were within the validated ranges. A self-administered sexual function assessment was completed by the five women who were sexually active. The scores indicated sexual difficulties, particularly in the areas of infrequency of intercourse and anorgasmia.

CONCLUSIONS

The sensory data for all six women were outside the normal range for the clitoris. The

results for the upper vagina, which had not had surgery, were within normal ranges. These findings suggest that genital surgery may disrupt sensory input. Sexual function also appears to be impaired and this may relate to the compromised sensitivity and restricted introitus. The possibility that women with CAH have deficient clitoral sensation ab initio cannot be excluded. These striking findings must be evaluated further in the light of the controversy about the issue of genital surgery in children with CAH.

KEYWORDS

sexual function, female genitalia, congenital adrenal hyperplasia, clitoris, sensation

INTRODUCTION

Surgical correction of the enlarged clitoris is currently common practice in the management of conditions such as congenital adrenal hyperplasia (CAH), and other intersex conditions. Techniques originally consisted of complete removal of the clitoris, but have developed to allow preservation of the glans and neurovascular bundle in the hope that this will preserve sensation to this sensitive area.

It is now accepted that the clitoris is important in the female sexual response and in achieving orgasm. It is thought that pleasurable sensations and orgasm are mediated largely through sensory input. However, there are few data on the long-term effects of surgery to this sensitive organ. As part of a larger ongoing study investigating genital sensory function after feminizing genital surgery, we assessed the sensation in the clitoris in patients with CAH and compared it with published normal values

from women without CAH, or any history of genital surgery. The results are from the first six patients in the pilot study, and are presented because the results were markedly abnormal.

PATIENTS AND METHODS

Patients were identified using the database for a tertiary referral clinic for intersex conditions, based in a teaching hospital. All patients who were aged ≥ 16 years with a diagnosis of 21-hydroxylase deficient CAH were eligible for recruitment. Patients were excluded if they had additional medical conditions which might alter sexual function, e.g. diabetes or multiple sclerosis. Recruitment was via a postal invitation with a study information sheet. Respondents were then sent a questionnaire to complete before attending the genital assessment clinic, which incorporated the Golombok Rust Inventory of Sexual Satisfaction (GRISS) sexual functioning questionnaire [1]. The

GRISS is a multidimensional instrument which has been validated in the UK. It provides an overall score of sexual function, and seven further subscale scores reflecting sexual function in the following areas: frequency, communication, satisfaction, avoidance, sensuality, vaginal penetration difficulties, and orgasm.

The clinical examination consisted of cosmetic and anatomical evaluation, using a scoring sheet, and neurological assessment using the genito-sensory analyser (GSA, Medoc Ltd., Israel) and Von Frey filaments (Semmes Weinstein Von Frey anaesthesiometer). The GSA has been designed and tested on normal women, and those with female sexual dysfunction. Normal values were derived from a group of 89 healthy volunteers and 61 women (none intersex) with sexual dysfunction. Of these 150 women, 62 were re-tested to confirm the reproducibility for both normal and abnormal responses [2]. Normal values were obtained for warmth, cold and vibration, and varied with age; these

TABLE 1 Sensation first felt, with normal values in brackets, and the GRISS scores

Variable	Patient						Mean (SD)
	1	2	3	4	5	6	
Mean (upper limit of normal)							
Clitoris:							
warmth °C	44 (38.4)	42.8 (38.5)	38.3 (38.4)*	47.7 (38.5)	49.6 (38.4)	46.1 (38.3)	44.8 (4.0)
cold °C	28.9 (33.3)	24.7 (33.3)	30.7 (33.3)	30.2 (33.3)	20.1 (33.3)	25.5 (33.3)	26.7 (4.1)
vibration amplitude, µm	3.13 (1.6)	8.09 (1.8)	1.95 (1.6)	1.26 (1.8)*	5.5 (1.6)	3.3 (1.5)	3.9 (2.5)
Vaginal vibration amplitude, µm	–	–	5.82 (6.0)*	–	4.6 (6.0)*	3.5 (5.0)*	4.64 (1.16)
Von Frey, g	0.4	0.6	0.04	0.6	1.0	0.16	0.5 (0.35)
Surgery	CR	CR	CR	CR	CR+	CR+	–
GRISS scores†							
Overall sexual function	4	2	6	6	9		
Infrequency of intercourse	6	8	9	5	8		
Communication problems	3	5	9	8	5		
Dissatisfaction	3	6	4	7	2		
Avoidance	5	1	7	8	9		
Sensuality	3	1	6	6	8		
Vaginal penetration difficulties	2	1	5	6	8		
Anorgasmia	7	3	6	6	8		

* value within the normal range; †the original operation was clitoral reduction (CR), but on examination no glans or clitoral tissue could be identified, the appearance being consistent with that of total clitorrectomy. ‡A score of ≥ 5 in any category indicates a problem.

represent the control group in the present study. The GSA was fully calibrated and maintained by the parent company since arrival in our unit.

The GSA machine consists of two separate probes, one for temperature measurements and one for vibratory testing, with a feedback patient-response switch. Each probe can be used externally on the clitoris and inside the vagina. The temperature probe has a thermal button on the end for applying to the clitoris and a flat element on the cylinder for the vaginal contact. The vibration probe vibrates throughout its length, with an end button for clitoral application, or is placed in the vagina to assess vaginal sensation.

The method of limits was applied, whereby the stimulus is changed in linear increments until the first sensation is felt. At this point the subject presses a button to register sensation and stop the test. The tests are repeated six times each for warmth, cold and vibration, and the mean of the six readings calculated. Sensation to light touch was evaluated using Von Frey filaments; these deliver a consistent reproducible force to the area and are available in a series of grades. Increasing strengths of filament were applied until first sensation was felt. Ethical approval

was granted by the joint university and hospital ethics committee.

RESULTS

Of 38 women contacted, 20 agreed to participate; the present results are from the first six tested. The individual results are shown in Table 1, with the normal values for comparison. The six women were aged 17–31 years; all had 21-hydroxylase CAH, five of whom were salt-losing and one (no. 6) who was not. All patients had undergone clitoral reduction, in five of them one clitoroplasty and in one (no. 3) two procedures. Two patients had no identifiable glans or clitoral tissue, with an appearance consistent with total clitorrectomy, although the original operation was noted as clitoral reduction (nos 5 and 6). Where no clitoral tissue was visible the position of the clitoris could still be identified from the palpable scar tissue, and the probe was placed against this.

Five patients had abnormal results for sensation of warmth to the clitoris, and all had abnormal results for sensation of cold; five of the six had abnormal results for vibration to the clitoris. None of the patients had a sufficient introital opening to

accommodate the thermal probe vaginally, so vaginal temperature could not be assessed. The thermal probe is 2.8 cm in diameter and 13.5 cm long, of which two-thirds is designed to be entered into the vagina. The vibration probe is smaller, at 10.3 cm long and 2.4 cm diameter, and this could be introduced into the vagina in three of the six women. All results for vaginal vibration were within normal limits.

The GRISS was completed by the five women who were sexually active and results are also shown in Table 1. A score of ≥ 5 in any of the subsets indicates a problem with that area. All five women had problems with infrequency of intercourse. Four of them had problems with achieving orgasm and tended to avoid intercourse. Three had vaginal penetration difficulties and issues with sensuality, although only two of the five expressed dissatisfaction with their sexual relationships.

DISCUSSION

The surgical management of infants born with ambiguous genitalia is controversial; the idea that a gender should be assigned at birth and the genitalia constructed to conform with it is being questioned. There are doubts about

several aspects of this policy, but not least is the possibility that surgery in infancy may irrevocably damage sexual function.

The anatomy of the clitoris is not well understood, and studies have shown that most anatomy texts are inaccurate in the size and precise location of this organ [3]. Anatomical studies show dense innervation to almost all areas of the normal clitoris. Even with careful surgical techniques, cut nerves were detected in corporeal specimens obtained from women with CAH [4]. The densest innervation in the glans clitoris is on the dorsal (top) aspect, with nerves fanning out extensively around the dorsal and lateral aspects of the clitoral body. Stimulating the dorsal aspect of the clitoris has been suggested to be most important for achieving orgasm in normal women [5]. A different approach to surgery was suggested based on these anatomical studies, to preserve as many nerves as possible, with a view to maximising sensation.

Innervation to the clitoris is supplied by the pelvic, pudendal and hypogastric nerves, and is connected to L5–S1, T12–L4, and S2–S4 [2,6]. Light pressure and vibration are conducted by large myelinated fibres to the dorsal columns of the spinal cord. Temperature and pain are carried by small unmyelinated fibres to the spinothalamic tracts. Therefore, assessing temperature, vibration and light touch, the sensation of the clitoris may be objectively evaluated.

The present study is the first to use objective sensory testing studies on patients with a history of genital surgery. Previous studies assessed pudendal evoked responses before and after surgery. Gearhart *et al.* [7] evaluated six patients before and after surgery, and showed that evoked responses were not significantly altered. They concluded that nerve conduction and sensation were preserved. However, we dispute the use of evoked responses as appropriate methods to assess sensation. Pudendal evoked responses assess the large myelinated fibres only, whereas sensation is also carried in unmyelinated C and small myelinated A δ fibres. Chase [8] argues that subjects may have normal evoked responses, yet have little or no sensation, or capacity for orgasm.

The clitoris is the most densely innervated part of the human body; in the present study temperatures of up to 50 °C were applied to

this sensitive area. It is likely that most people would be unable to tolerate such temperatures on other areas of body, e.g. the palm of the hand, but the patients only responded when the probe reached this temperature, showing their degree of sensory impairment. This study suggests that sensation to the clitoral area is impaired after surgery. It is possible that the innervation of the clitoris in CAH is abnormal from birth, although this seems a less likely explanation for our findings.

It is often argued that the results of genital surgery carried out 15 or 20 years ago should be interpreted cautiously. The recent consensus statement on the management of 21-hydroxylase deficiency states there is reason for optimism that future outcomes will be better with current surgical techniques [9]. Although five of the six patients had initial surgery 15 years earlier, one (no. 6) had initial surgery only a year before the study and yet showed markedly abnormal results in temperature and vibration sensation in the clitoris. We are unaware of any data which show that the outcome is improved with modern techniques.

Two of the present patients had minimal clitoral tissue, with an appearance consistent with total clitorectomy rather than clitoral reduction, although that was the stated operation. We feel it is appropriate and valid to include these women in our study. First, many women will have had a clitorectomy, either intentionally, in keeping with previous practice, or unintentionally, through surgical trauma. In our clinical practice we note that even patients documented as having a clitoral reduction may have a very small, scarred or absent clitoris in adult life, presumably through vascular damage or necrosis. Not to include these patients is inappropriate and introduces bias. Second, although clitorectomy is no longer thought to be used in the UK or USA, it is still practised elsewhere, and the present study highlights the long-term problems with this approach [10].

An additional finding was that none of the patients could insert the temperature probe into the vagina. Vaginoplasties can result in stenosis and further operations are routinely required to permit subsequent intercourse [11]. This has led to suggestions that vaginoplasties should be undertaken during adolescence, to reduce the number of operations required by patients [12]. Three of

the six patients were able to insert the smaller vibration probe, which assesses vibration to the upper vagina. All three results were within normal limits, and none of them had had surgery to the upper vagina.

At present there are no data for values of Von Frey filament sensation to the clitoris and vagina in normal controls, so it is difficult to interpret the results for the present patients. However, assessing light touch is an important part of a neurological examination, to evaluate nerves carried in the dorsal columns, and normal values need to be developed to allow a thorough assessment.

Sexual function is an important area which has been neglected in the past. Women with CAH have been reported to have a higher incidence of sexual dysfunction than the normal population [13]. They tend to reach sexual milestones later than matched peers, and have more issues with body image [14]. Penetration difficulties are common, which may be related to a high incidence of vaginal stenosis [15]. In addition, dyspareunia is common and may be a result of clitoral tissue being trapped during arousal. Normal female sexual function is multifactorial and it is not known to what extent hormonal changes may affect sexual function. Women with CAH often have fluctuating testosterone levels in the normal to high range. Current research does not suggest that high androgen levels contribute significantly to female sexual dysfunction, although the role of androgens in female sexual function is still being evaluated [16]. Data on sexual function should form part of the long-term follow-up of patients who have had genital surgery. The present data shows overall sexual difficulties, especially for infrequency of intercourse and compromised capacity for orgasm. Even so, interestingly only two patients reported dissatisfaction with the perceived quality of their sexual relationship. If sex were difficult and pleasure compromised, then infrequency of intercourse would be unlikely to be an common complaint; rather it may be welcomed. In addition, this evidence suggests that sexual function cannot be assessed purely on reported satisfaction, elicited during a routine clinic consultation.

In women with a vagina that is totally inadequate for intercourse the goals of reconstruction may be limited; perhaps the construction of a neovagina that allowed penetrative intercourse might be better than

no intercourse. However, different criteria apply to the clitoris. If the clitoris is functionally normal at birth but simply enlarged, it is questionable whether surgery that irretrievably impairs function is acceptable. We recognize that raising a female infant with a prominent clitoris may be difficult, but on reaching adulthood the woman may resent the destruction of a major part of her sexual potential. The present results should, at the very least, draw attention to the poor functional results of present protocols.

The current results are preliminary and from an ongoing research project. Future results will be fully reported, including further evaluation of sexual function, and patient attitudes to genital surgery.

In conclusion, we evaluated sensation in the clitoris, the results of which were profoundly abnormal in women with CAH who have had clitoral surgery. However, vaginal sensation appeared to be within normal limits, suggesting that the abnormal clitoral findings may be a result of surgery in this delicate area, rather than an effect of CAH. There is currently no justification for the optimism that modern surgical techniques are better for preserving clitoral sensation than previous operations. Further results from the current study will help to evaluate these striking findings; meanwhile, parents and patients need to be advised that sensation to the clitoris may be compromised after corrective surgery, and that this may impair sexual function.

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Abbreviations: CAH, congenital adrenal hyperplasia; GRISS, Golombok Rust Inventory of Sexual Satisfaction; GSA, genito-sensory analyser.